

GUIDELINES FOR THE
CARE OF PEOPLE WITH
SPINA BIFIDA



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Guidelines for the Care of People with Spina Bifida

An Initiative of the Spina Bifida Association

Preface to the Fourth Edition

Spina Bifida is the most commonly-occurring complex congenital birth defect associated with long-term survival. With this understanding, along with the knowledge of the multiple medical and psychosocial issues that people with Spina Bifida face, *the Guidelines for Spina Bifida Health Care Services Throughout the Lifespan* were first published by the Spina Bifida Association of America (now known as the Spina Bifida Association, SBA) in 1990 and revised in 1995. Both editions were the culmination of several years of work by the SBA's Professional Advisory Council (PAC), as well as numerous consultants under the editorial leadership of Karen Rauen, RN, MSN. These guidelines were based on limited contemporary knowledge and expert opinion.

Research on outcomes in Spina Bifida has been sparse. In that light, a symposium entitled "Evidence-Based Practice in Spina Bifida: Developing a Research Agenda" was convened May 9-10, 2003 to identify the current evidence related to Spina Bifida, identify research gaps and priorities, and to foster new directions and funding for research. Sponsors included the Centers for Disease Control and Prevention, Agency for Healthcare Research and Quality, the National Institutes of Health (Office of Rare Diseases), and the U. S. Department of Education. Additional supporting agencies included the National Institute of Child Health and Human Development, the Interagency Committee on Disability Research, and the Spina Bifida Association. (A summary manuscript, edited by Gregory Liptak, MD, MPH, is available from the Spina Bifida Association, 1600 Wilson Blvd, Suite 800, Arlington, VA 22209.)

This meeting highlighted that much of the research in Spina Bifida was based on case series; very few randomized control trials or representative cohort studies had been performed on any topic on people with Spina Bifida. Research related to adults with Spina Bifida was nearly non-existent. The primary goal of the evidence-based review was achieved: directions for research were clarified. The third edition of the *Guidelines for Spina Bifida Health Care Services Throughout the Lifespan*, edited by Mark Merkens, MD, was published in 2006 by SBA. Guidance included in the third edition was based on reviews generated from the Evidence-Based conference as well as expert consensus.

Guidelines for the Care of People with Spina Bifida are the fourth edition of the Guidelines, and the result of three years of planning, literature review and content development by nearly 100 volunteers. The new Guidelines were needed to ensure that all people living with Spina Bifida receive the best and most up-to-date care possible, and because previous versions did not have robust coverage of the care needs of adults. Additionally, the fourth version features a new title that reflects greater respect and understanding for the people who are impacted by living with Spina Bifida. In other words, the fourth edition Guidelines were developed to treat and care for *the people* who live with Spina Bifida, not just *the conditions* associated with this birth defect. Finally, this fourth edition features a number of new topics, including Transition and Quality of Life, important to the health and well-being for all people living with Spina Bifida.

Despite the efforts resulting from the "Evidence-Based Practice in Spina Bifida: Developing a Research Agenda" conference in 2003, the extensive literature review done for the fourth

edition of the Guidelines continues to identify that research in Spina Bifida remains limited. Where evidence exists, it is included. For other recommendations the collective judgement of expert working groups determined the appropriateness of assessments and interventions to be considered. The workgroups used the consensus-building methodologies of Single Text Procedure and Nominal Group Techniques. These recognized guidelines development methodologies allow the inclusion of expert opinion for aspects of care for which medical evidence does not exist or is not robust¹⁻⁵

Moving forward, these recommendations will be updated as new data become available. As such, these should be considered as guidelines and options, not standards of care. Currently available reported research findings are not sufficiently strong and robust to set standards of care. Guidelines are not meant to be legal requirements but rather provide the practitioner with recommended directions for assessments and interventions for their patients with Spina Bifida based on the current best available research findings and expert consensus. It is hoped that these Guidelines will not only guide health care providers but also patients and families, so that people with Spina Bifida can have the best and most scientifically-based care and treatments throughout their ever-longer and higher-quality lives.

Since the publication of the third edition of the Guidelines, there have been advances in health care service delivery concepts related to improving the care of children with a medical complexity, including Spina Bifida. These concepts will be important in ensuring the full implementation of the fourth edition of the *Guidelines for the Care of People with Spina Bifida*. The first is that care coordination is an essential component of health care delivery.⁶ At the core, patient- and family-centered care within a medical home is a foundational component; outcomes are optimized when there is cross-sector collaboration among the multiple medical systems and providers, community services, and support agencies with whom families and people with Spina Bifida interact. While effective care coordination typically requires dedicated paid personnel, care coordination activities are not the sole responsibility of a single individual or provider.⁷ Rather, all people who interact with patients and families have a role to play in care coordination. The second concept, in the context of patient- and family-centered care, is that for people with Spina Bifida, care provision may be provided via a medical neighborhood⁸ with team-based care.⁹ Within this framework is co-management with defined roles, data sharing, and collaborative care protocols among primary care, community-based services, and subspecialty care. Full implementation of these Guidelines to optimize outcomes for people with Spina Bifida cannot rest with the Spina Bifida clinic alone. Indeed, guidance provided on many topics should be implemented through primary care providers and efforts of community services. While the Spina Bifida clinic may direct the overall health care planning in many cases, optimal care is best achieved as a partnership between families and people with Spina Bifida, primary and subspecialty care providers, health systems, and community services.

These Guidelines were developed to serve people with Spina Bifida and those who care for them. It is essential to remember that several factors influence how an individual or family member uses the education and written information they are provided. This is imperative, particularly when reaching across potential obstacles such as cultural and/or language differences. It is known that the dynamics that modify the incidence of Spina Bifida are multifactorial, such as the well-documented higher incidence of Spina Bifida among people of Hispanic origin. Thus, it is increasingly critical for health care and community service providers to consider how a family's language, level of acculturation, and cultural constructs of care (e.g. concept of self-management and independence from others) directly influence their understanding and reception of the health care message along with their willingness to change behavior.¹⁰ Moreover, since over 20% of the US population older than five years of age speaks

a language other than English at home, when possible, all families with limited English proficiency ought to be supported with additional health care navigation services, along with oral and written information provided in their preferred language.¹¹

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Care Coordination

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Introduction

“Care coordination is the deliberate organization of patient care activities between two or more participants (including the patient) involved in a patient’s care to facilitate the appropriate delivery of health care services. Organizing care involves the marshalling of personnel and other resources needed to carry out all required patient care activities, and is often managed by the exchange of information among participants responsible for different aspects of care.” — International Journal of Care Coordination¹

Care coordination (also described as case management services) in the case of people with Spina Bifida and their families, is a process that links them to services and resources in a coordinated effort to maximize their potential by providing optimal health care. However, care coordination for people with Spina Bifida and their families can be complicated due to the medical complexities of the condition and the need for multidisciplinary care, as well as economic and sociocultural barriers to coordination of care. Care coordination is often a shared responsibility by the multidisciplinary Spina Bifida team.² For this reason, the Spina Bifida Care Coordinator has the primary responsibility for overseeing the overall treatment plan for the individual with Spina Bifida.³ Care coordination includes communication with the primary care provider in a patient’s medical home.^{2,4-5}

Care coordination is an essential part of the multidisciplinary Spina Bifida care team and vital to improving the health care and wellness outcomes for people living with Spina Bifida. It is recommended, if possible that Spina Bifida care programs dedicate the necessary financial resources and fund sufficient full-time equivalent staff so that optimal care coordination can be provided by designated, trained, and paid health care professionals.

There are very few database studies that demonstrate the benefits of Spina Bifida care coordination programs resulting in improved health outcomes, decreased morbidity and mortality, higher quality of life, improved success and independence in adulthood and decreased cost of care for people with Spina Bifida. More research needs to be completed to compile scientific evidence of the effectiveness of care coordination programs to develop a best-practices model of care coordination for the person with Spina Bifida.

A pediatric medical home is a family-centered partnership within a community-based system that provides uninterrupted care with appropriate payment to support and sustain optimal health outcomes.⁶ In their important role of providing a medical home for people with Spina Bifida, primary care providers also have a vital role in the process of care coordination, in concert with the family, and the Spina Bifida team.^{2,4}

Over the past 50 years, advances in medicine have resulted in increased survival of children with Spina Bifida.⁷ Many of these people, now adults, require long-term coordinated services from a variety of health care professionals and organizations. Great variability exists among programs with services for people with Spina Bifida and their families. During the past 10 to 20

years, people with Spina Bifida and their families have had greater access to care coordination, in part due to systems of care consisting of a variety of organizations and agencies that include independent health care professionals and third-party payers, often with different missions. However, despite increased access in some areas, not all individuals receive appropriate care coordination services, especially as they transition from pediatric to adult care.

Generally, the goals of care coordination are the following:

- gain access to and integrate services and resources,
- link service systems with the family,
- avoid duplication and unnecessary cost, and
- advocate for improved individual outcomes.

Outcomes

Primary

1. Maximize the overall health and functioning of individuals living with Spina Bifida throughout the lifespan by improved access to team-based, patient- and family-centered coordinated care for medical, social, educational, equipment needs, and other developmentally relevant related services.

Secondary

1. Promote comprehensive, coordinated and uninterrupted access to medical, subspecialty, and allied health professional services throughout the lifespan with appropriate communication between the person with Spina Bifida and members of their care team.⁸
2. Promote routine screenings and testing congruent with Spina Bifida guidelines for specific secondary conditions.

Tertiary

1. Maintain up-to-date coordinated care for individuals living with Spina Bifida to minimize medical complication rates, help control cost of care, and minimize emergency room use and unanticipated hospitalization, morbidity, and mortality.⁹

Pregnancy to 0-11 months

Clinical Questions

1. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as people with Spina Bifida age?
2. How do Spina Bifida Care Coordinators collaborate with team members, allied health services, and community partners to optimize opportunities and overall quality of life?
3. What is the best way to communicate effectively between the multiple Spina Bifida care team members and the family so as to best serve the child's needs, prevent complications, and improve the overall experience of care?
4. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers include insufficient training, logistical difficulties, and unavailability of personnel and community resources.
5. What aspects of a care coordination program do families with a pregnancy or children with Spina Bifida find most helpful and improve their perception of the care they receive?

Guidelines

1. After the Spina Bifida diagnosis has been made, it is recommended that the Spina Bifida Care Coordinator should be readily available to the family to provide support

and education throughout the pregnancy. These consults may take place as part of a maternal fetal health visit in a high-risk pregnancy center. The goals of the consults may include to:

- assist the family with coping with the new diagnosis,
- provide overall education on what the family can expect ages 0-11 months and while in their stay in the neonatal intensive care unit (NICU) stay, and
- provide general information on the signs and symptoms related to Spina Bifida.

The Spina Bifida Care Coordinator may assist in synchronizing prenatal visits for other subspecialties that could include neurosurgery, urology, and orthopedics. The primary role of the Spina Bifida Care Coordinator during this stressful time for families is to convey the message that the family is not alone because a well-prepared team will be on hand to provide them with the support they need to help care for their child.¹⁰

2. It is recommended that the Spina Bifida Care Coordinator should use the time during pregnancy or 0-11 months to introduce the family to the Spina Bifida clinic and multidisciplinary team (when one is available) and begin the process of arranging post-discharge follow-up. Through counseling and encouragement, the Spina Bifida Care Coordinator:
 - assists the family to accept the diagnosis, and
 - contacts the medical home of the family and infant with Spina Bifida and identifies the specific lead professional or nurse case manager who will serve as the point of contact for the family to provide education, resources, and support.¹⁰⁻¹¹
3. It is recommended that the Spina Bifida Care Coordinator provide families with a broad and appropriate early education across the spectrum of symptoms and conditions related to Spina Bifida. This may include educating the family on early urologic work-up and management and possibly teaching them about clean intermittent catheterization (CIC). Other topics may include education on latex allergy and precautions, education regarding early orthopedic interventions, and education to help families recognize potential neurosurgical complications. (clinical consensus) (Latex and Latex Allergy in Spina Bifida Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)
4. It is recommended that the Spina Bifida Care Coordinator work closely with the NICU staff to ensure that parents have the necessary skills and education for discharge and a smooth transition to home care. (clinical consensus)
5. It is recommended that the Spina Bifida Care Coordinator should communicate and collaborate between the family and the multidisciplinary and sub-specialty Spina Bifida team members to arrange and execute the child's follow-up appointments, monitoring, and care plan.^{2,12}
6. When applicable, it is recommended that the Spina Bifida Care Coordinator should update the child's primary care provider and/or medical home on the current care goals and recommendations of the Spina Bifida multidisciplinary care team. Use two-way communications to identify and address medical concerns and obtain updated records from the medical home, such as immunizations, growth charts, developmental screenings, and other materials.^{4,11}
7. When appropriate, it is recommended that the Spina Bifida Care Coordinator should refer families to early intervention services. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

8. It is recommended that the Spina Bifida Care Coordinator make referrals to local Spina Bifida Association Chapters and parent support groups, as available. (clinical consensus)
9. It is recommended that the Spina Bifida Care Coordinator should monitor the parent's and caregiver's compliance with appointments, and problem-solve with them if non-compliance is noted. (clinical consensus)
10. When appropriate, it is recommended that the Spina Bifida Care Coordinator should assess family dynamics in how they are coping with the diagnosis, evaluate psychosocial stressors for the family, and assist them with referrals to mental health and social services professionals. (clinical consensus) (Mental Health Guidelines)

1-2 years 111 months

Clinical Questions

1. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as people with Spina Bifida age?
2. How do Spina Bifida Care Coordinators collaborate with team members, allied health services, and community partners to optimize the opportunities and overall quality of life of the child?
3. What is the best way to communicate effectively between the multiple Spina Bifida care team members, the child, and the family so as to best serve the child's needs, prevent complications, and improve the overall experience of care?
4. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers could include insufficient training, logistical difficulties, and unavailability of personnel and community resources.
5. What aspects of a care coordination program do families and their children find most helpful and improve their perception of the care they receive?

Guidelines

1. It is recommended that the Spina Bifida Care Coordinator work with the family and the multidisciplinary Spina Bifida care team to ensure that the child with Spina Bifida is up-to-date on all sub-specialty care visits, imaging, monitoring, and equipment needs where appropriate. This may include assistance with insurance authorization or referrals.¹²
2. It is recommended that the Spina Bifida Care Coordinator provide education across the spectrum of symptoms and conditions related to Spina Bifida to empower families and children to manage their own care and recognize complications and emergencies. The Spina Bifida Care Coordinator should also identify gaps in the family knowledge base. (clinical consensus) (Family Functioning Guidelines, Self-Management and Independence Guidelines)
3. It is recommended that the Spina Bifida Care Coordinator monitor and document family enrollment in and progress with therapies and treatments and encourage continued participation in early intervention services.¹³
4. It is recommended that the Spina Bifida Care Coordinator collaborate with team members to identify gaps or barriers to achieving the goals of the person's care plan and assist with additional referrals as appropriate.¹²
5. When applicable, it is recommended that the Spina Bifida Care Coordinator update the primary care provider and/or medical home on the current care goals and recommendations of the Spina Bifida multidisciplinary care team. Use two-way communications to identify and address medical concerns and obtain updated

- records from the medical home, such as immunizations, growth charts, developmental screenings, and other materials.^{4,11}
6. It is recommended that the Spina Bifida Care Coordinator begin emphasizing the child's path towards independence with the family. Encouraging activities such as learning to help put on shoes and braces will promote greater independence and autonomy and promote further discussions of independence as the child ages. (clinical consensus) (Self-Management and Independence Guidelines)
 7. It is recommended that the Spina Bifida Care Coordinator assess family dynamics in how they are coping with the diagnosis, evaluate psychosocial stressors for the family, and assist them with referrals to mental health and social services professionals when appropriate. (clinical consensus) (Mental Health Guidelines)

3-5 years 11 months

Clinical Questions

1. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as people with Spina Bifida age?
2. What evidence exists to show the success of the care coordination program in improving the overall health of children with Spina Bifida?
3. What literature is available to support optimal teaching and education of children and their caregivers throughout the lifespan to maximize early independence?
4. What is the best way to effectively communicate between multiple Spina Bifida care team members, people with Spina Bifida and their families to best serve their needs, prevent complications, and improve their overall experience of care?
5. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers could include insufficient training, logistical difficulties, and unavailability of personnel and community resources.
6. What aspects of a care coordination program do families and their children find most helpful and improve their perception of the care they receive?

Guidelines

1. It is recommended that the Spina Bifida Care Coordinator provide developmentally-appropriate care education across the spectrum of symptoms and conditions related to Spina Bifida to empower families and children to manage their own care and be able to recognize when complications and emergencies arise. The coordinator should also identify and/or improve gaps in the family knowledge base specifically related to the preschool period (mobility progress, skin inspection, bowel and bladder care, speech/cognitive development, and more). (clinical consensus) (Bowel Function and Care Guidelines, Mental Health Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Skin (Integument) Guidelines, Urology Guidelines)
2. It is recommended that the Spina Bifida Care Coordinator prepare the family for early independence, entering preschool, and planning special education and health-related services in the school. (clinical consensus)
3. It is recommended that the Spina Bifida Care Coordinator coordinate with the family and multidisciplinary Spina Bifida care team to ensure that the child with Spina Bifida is up-to-date on all sub-specialty care visits including, imaging, monitoring, and equipment needs, where appropriate, including assistance with insurance authorization when needed.^{2,12}
4. It is recommended that the Spina Bifida Care Coordinator assess family dynamics in coping with the diagnosis and evaluate psychosocial stressors for the family. The

- Spina Bifida Care Coordinator should also assist with referrals to mental health and social services when appropriate. (clinical consensus) (Mental Health Guidelines)
8. When applicable, it is recommended that the Spina Bifida Care Coordinator update the primary care provider on the current care goals and recommendations of the Spina Bifida multidisciplinary care team. Use two-way communications to identify and address medical concerns and obtain updated records from the person's primary care provider such as immunizations, growth charts, developmental screenings, and other materials.^{4,11}
 5. It is recommended that the Spina Bifida Care Coordinator serve the family as the lead contact person and information-provider for the multidisciplinary medical services for the child with Spina Bifida. The Spina Bifida Care Coordinator should monitor the family's needs and prescriptions for durable medical equipment, supplies, and medications, as needed.¹¹

6-12 years 11 months

Clinical Questions

1. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as people with Spina Bifida age?
2. What evidence exists to show the success of care coordination programs in improving the overall health of children with Spina Bifida?
3. How do Spina Bifida Care Coordinators collaborate with team members, allied health services, and community partners to optimize the opportunities and overall quality of life of people with Spina Bifida?
4. What is the best way to effectively communicate between multiple team members, people with Spina Bifida and their families to best serve their needs, prevent complications, and improve the overall experience of care?
5. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers could include insufficient training, logistical difficulties, and unavailability of personnel and community resources.
6. What aspects of a care coordination program do people with Spina Bifida find most helpful and improve their perception of the care they receive?

Guidelines

1. It is recommended that the Spina Bifida Care Coordinator provide developmentally-appropriate care education across spectrum of symptoms and conditions related to Spina Bifida to better empower children and their families to manage their own care and be able to recognize complications and emergencies. Identify and/or improve gaps in the family knowledge base specifically related to the school age period (mobility progress, skin inspection, bowel and bladder care, academic/cognitive development, school and social functioning, and more). (clinical consensus) (Bowel Function and Care Guidelines, Mental Health Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Skin (Integument) Guidelines, Urology Guidelines)
2. It is recommended that the Spina Bifida Care Coordinator monitor primary school functioning and update school education and health plans. Encourage participation in age-appropriate activities outside of school with peers, with and without Spina Bifida. Encourage participation in activities such as camps or special family weekends that provide safe places to develop peer relationships with children who may have similar medical challenges. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

3. It is recommended that the Spina Bifida Care Coordinator coordinate with the family and multidisciplinary Spina Bifida care team to ensure that the child is up-to-date on all sub-specialty care visits, imaging, monitoring, and equipment needs, where appropriate. This may include assistance with insurance authorization when needed.^{2,12}
4. It is recommended that the Spina Bifida Care Coordinator assess family dynamics in coping with the diagnosis and evaluate psychosocial stressors for the family. Assess for depression and anxiety and assist with referrals to mental health and social services when appropriate. (clinical consensus) (Mental Health Guidelines)
5. It is recommended that the Spina Bifida Care Coordinator should work with the child, his or her family and Spina Bifida team members and therapists to start progress on self-management goals and education. Monitor family progress in self-management at regular intervals and clinic visits. Engage the school nurse to help facilitate self-management and independence.¹⁴ Teach self-advocacy and encourage the child to participate as much as possible in his or her own self-management.¹⁵ (Self-Management and Independence Guidelines)
6. When applicable, it is recommended that the Spina Bifida Care Coordinator update the primary care provider on the current care goals and recommendations of the Spina Bifida multidisciplinary care team. The Spina Bifida Care Coordinator should use two-way communications to identify and address medical concerns and obtain updated records from the primary care provider and/or medical home such as immunizations, growth charts, developmental screenings, and other materials.^{4,11}
7. It is recommended that the Spina Bifida Care Coordinator serve the family as the lead contact person and information-provider for the multidisciplinary medical services for the child with Spina Bifida and monitor family needs and prescriptions for durable medical equipment, supplies, and medications, as needed.^{11,16}

13-17 years 11 months

Clinical Questions

1. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as people with Spina Bifida age?
2. What evidence exists to show the success of care coordination programs in improving the overall health of people with Spina Bifida?
3. How do Spina Bifida Care Coordinators collaborate with team members, allied health services, and community partners to optimize the opportunities and overall quality of life of people with Spina Bifida?
4. What is the best way to effectively communicate between multiple team members, people with Spina Bifida and their families to best serve their needs, prevent complications, and improve their overall experience of care?
5. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers could include insufficient training, logistical difficulties, and unavailability of personnel and community resources.
6. What aspects of a care coordination program do people find most helpful and improve their perception of the care they receive?
7. What is the Spina Bifida Care Coordinator's role in 1) educating and bringing adult providers into the care team to ensure seamless transition of care and in 2) developing transition goals and processes for people as they age out of the pediatric system to ensure continuity of care?

Guidelines

1. It is recommended that the Spina Bifida Care Coordinator provide developmentally-appropriate care education across the spectrum of symptoms and conditions related to Spina Bifida to better empower children and families to manage their own care and recognize complications and emergencies. Identify and or improve gaps in the family knowledge base specifically related to the teenage age period (mobility progress, skin inspection, bowel and bladder care, sexuality, academic/cognitive development, social functioning at school and with peers, high risk behaviors, and more). (clinical consensus) (Bowel Function and Care Guidelines, Mental Health Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Skin (Integument) Guidelines, Urology Guidelines)
2. It is recommended that the Spina Bifida Care Coordinator monitor secondary school functioning and update the school education and health plan so that it includes preparation for college or other higher education opportunities. Encourage participation in age-appropriate activities with peers outside of school. Where appropriate, provide information for driver education and training programs for the teenager with Spina Bifida. (clinical consensus)
3. It is recommended that the Spina Bifida Care Coordinator communicate with the family and multidisciplinary Spina Bifida care team to ensure the individual with Spina Bifida is up-to-date on all sub-specialty care visits, imaging, monitoring, and equipment needs where appropriate. This may include assistance with insurance authorization when needed.^{2,12}
4. It is recommended that the Spina Bifida Care Coordinator update the primary care provider on the current care goals and recommendations of the Spina Bifida multidisciplinary care team. The coordinator should use two-way communications to identify and address medical concerns and obtain updated records from the primary care provider such as immunizations, growth charts, developmental screenings, and other materials.^{4,11}
5. It is recommended that the Spina Bifida Care Coordinator work with the teenager and his/her families, Spina Bifida team members, and therapists to continue progress on self-management goals and education. Monitor family progress at regular intervals in self-management and clinic visits with the goal of achieving as much independence as is realistically possible. Engage the school nurse to help facilitate self-management and independence.¹⁴ Teach self-advocacy and encourage the teenager to participate as much as possible in his or her own self-management. When appropriate, discuss what limitations to independence the teenager may have due to deficits in memory, cognition, and executive functioning and provide the parents with additional resources and support services, as needed.¹⁷⁻¹⁹ (Self-Management and Independence Guidelines)
6. It is recommended that the Spina Bifida Care Coordinator begin preparing the teenager for transition to adult health care, including familiarizing them and their family with the Transition Guidelines and Self-Management and Independence Guidelines. Encourage the family to develop and assemble their own health care folder and records for use during travel, appointments in hospitals that are away from their home area, and other occasions when they will be away from the medical home. Encourage the person to make his or her own medical appointments once she or he is capable of doing so, and to start leading the conversation with specialists and other providers during clinic visits. Assist the family by making them aware that transition to adult life involves many aspects beyond health care, including educational planning or job training, making arrangements to live independently, and financial planning.^{15,20-21} (Self-Management and Independence Guidelines, Transition Guidelines)

7. It is recommended that the Spina Bifida Care Coordinator assess family dynamics in coping with the diagnosis and evaluate their psychosocial stressors. Collaborate with primary care provider to review age-appropriate screenings and assist with referrals to mental health and social services when appropriate. (clinical consensus) (Mental Health Guidelines)
8. It is recommended that the Spina Bifida Care Coordinator serve the family as the lead contact person and information provider for the multidisciplinary medical services for the person with Spina Bifida and monitor family needs and prescriptions for durable medical equipment, supplies, and medications, as needed. (clinical consensus)
9. It is recommended that the Spina Bifida Care Coordinator conduct an inventory of the person's ability to provide self-management, complete activities of daily living, and manage mobility equipment and transportation needs. For a person with a significant intellectual disability who may not be able to live independently, assist the family with the conservatorship process prior to age 18, and with maintaining Supplemental Security Insurance (SSI) and other types of insurance coverage. (clinical consensus) (Self-Management and Independence Guidelines)

18+ years

Clinical Questions

1. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as people with Spina Bifida age?
2. What evidence exists to show the success of care coordination program in improving the overall health of people with Spina Bifida?
3. How do Spina Bifida Care Coordinators collaborate with team members, allied health services, and community partners to optimize the opportunities and overall quality of life of people with Spina Bifida?
4. What is the best way to effectively communicate between multiple team members, people with Spina Bifida and their families to best serve their needs, prevent complications, and improve their overall experience of care?
5. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers could include insufficient training, logistical difficulties and unavailability of personnel and community resources.
6. What aspects of a care coordination program do people find most helpful and improve their perception of the care they receive?
7. What is the Spina Bifida Care Coordinator's role in 1) educating and bringing adult providers into the care team to ensure seamless transition of care and in 2) developing transition goals and processes for people as they age out of the pediatric system to ensure continuity of care?

Guidelines

1. It is recommended that the Spina Bifida Care Coordinator be knowledgeable about the resources for adults with Spina Bifida in their geographic area and coordinate the successful transition from pediatric to adult providers for adults with Spina Bifida. Where appropriate, educate adults with Spina Bifida about the key differences between adult and pediatric providers, including the possibility that there may not be an adult multidisciplinary Spina Bifida team nearby. Inform adults with Spina Bifida on the importance of having a primary care provider.¹⁵ (Transition Guidelines)
2. It is recommended that the Spina Bifida Care Coordinator conduct an inventory of the adult's ability to provide self-management, complete activities of daily living, and

- manage mobility equipment and transportation needs. For those who have a significant intellectual disability and may be unable to live independently, assist the family with the conservatorship process and with maintaining Supplemental Security Insurance (SSI) and other types of insurance coverage. (clinical consensus) (Self-Management and Independence Guidelines)
3. It is recommended that the Spina Bifida Care Coordinator assist with referrals to vocational and employment training opportunities, as needed. (clinical consensus)
 4. It is recommended that the Spina Bifida Care Coordinator assist with referrals to driver education training opportunities, as needed and/or referral for appropriate pre-driver training evaluations including vision, ability to use lower extremities vs. hand controls and other driving requirements. (clinical consensus)
 5. It is recommended that the Spina Bifida Care Coordinator coordinate with Spina Bifida care providers to determine if the person is up-to-date on all sub-specialty care visits, imaging and monitoring and equipment needs, where appropriate. This may include assistance with insurance authorization.^{2,12,17} (Mobility Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, and Urology Guidelines)
 6. It is recommended that the Spina Bifida Care Coordinator assess individual dynamics in coping with living with Spina Bifida and evaluate psychosocial stressors for the individual. Collaborate with primary care provider to review age-appropriate screenings and assist with referrals to mental health and social services, when appropriate. (clinical consensus) (Mental Health Guidelines)
 7. It is recommended that the Spina Bifida Care Coordinator should serve as the lead contact person and information provider for the Spina Bifida clinic and monitor individual needs and prescriptions for durable medical equipment, supplies, and medications as needed. Special considerations may be needed to apply care coordination principles to assist adults who see multiple providers independently.¹⁵
 8. It is recommended that the Spina Bifida Care Coordinator assess and monitor for clinical deterioration, loss of mobility, chronic pain, obesity, and use two-way communication between the Spina Bifida Care Coordinator and the primary care provider and/or medical home to assess and address concerns and assist with medical referrals, as appropriate.^{17,20} (Health Promotion and Preventive Medicine Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Nutrition, Metabolic Syndrome, and Obesity Guidelines, Orthopedics Guidelines, Urology Guidelines)

Research Gaps

1. What database studies demonstrate the benefits of Spina Bifida care coordination programs, specifically regarding improved health outcomes, decreased morbidity and mortality, higher quality of life, improved success and independence in adulthood and decreased cost of care?
2. What research exists regarding the effectiveness of care coordination programs to develop a best-practices model of care coordination for the person with Spina Bifida?
3. How do the roles and responsibilities of the Spina Bifida Care Coordinator evolve over time as the person with Spina Bifida ages?
4. What are the common barriers to creating an effective patient-centered care coordination program within the multidisciplinary Spina Bifida clinic? Examples of barriers could include insufficient training, logistical difficulties, and unavailability of personnel and community resources.
5. What aspects of a care coordination program do families and individuals with Spina Bifida find most helpful and improve their perception of the care they receive?

6. What evidence exists to show the success of the care coordination program in improving the overall health of people with Spina Bifida?
7. What literature is available to support optimal teaching and education of children and their caregivers throughout the lifespan to maximize early independence?
8. What is the Spina Bifida Care Coordinator's role in 1) educating and bringing adult providers into the care team to ensure seamless transition of care and in 2) developing transition goals and processes for people as they age out of the pediatric system to ensure continuity of care?

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Health Promotion and Preventive Health Care Services

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Introduction

Individuals with Spina Bifida have unique and specific medical and social needs when compared to the general population, especially over a lifetime. They often have issues with pain, including the shoulders¹ and back, as well as neuropathic pain.² Neurogenic bowel and neurogenic bladder are common medical problems for people with Spina Bifida. Without routine monitoring, neurogenic bladders can result in kidney failure and require dialysis.³ Persons with Spina Bifida are also at increased risk for hypertension.^{4,5} Depending on the subtype of Spina Bifida there is also the risk of hydrocephalus, either with or without shunting, and this requires lifelong management.^{6,7} Self-management of personal medical care by adults with Spina Bifida may be difficult because of the frequent presence of cognitive impairment and the number and complexity of possible medical issues to be managed.⁸

Because of challenges with self-management and the need for environmental and attitudinal accessibility, individuals with Spina Bifida may find it difficult to access both routine and preventive health care services fully. Health professionals must be willing and able to take the time needed to appropriately communicate with and manage the sometimes complex health care needs of people with Spina Bifida.

Persons with Spina Bifida need the same routine screening procedures that any other person their age would need.⁹⁻¹⁰ However, many medical offices do not follow universal design principles. For example, examination rooms may be far from the reception area and/or otherwise inaccessible; examination tables may be too high or not be adjustable to allow for a transfer for a physical examination; scales may not be wheelchair accessible or safe for people with limited balance.

Persons with Spina Bifida also have difficulty with medical professionals' lack of knowledge about or recognition of common medical conditions related to Spina Bifida. Medical professionals who do not routinely care for people with Spina Bifida are less likely to recognize these unique situations and address them appropriately. Routine histories may not always include questions about possible changes in mobility or other bodily functions.

The goal of improving access for people with Spina Bifida is to promote better use of routine and preventive health. Health promotion supports improved long-term health and well-being, improved quality of life, and decreased utilization of emergency services and hospitalizations.^{7,11}

Outcomes

Primary

1. Maximize physical and mental health for individuals with Spina Bifida within the context of the underlying condition.
2. Identify risks and presence of common or known secondary conditions early.

Secondary

1. Limit preventable emergency department visits and hospitalizations for individuals with Spina Bifida.
2. Monitor trends of identified and newly-emerging secondary conditions.

Tertiary

1. Provide patient-centered general health monitoring based on the United States Preventive Services Task Force (USPSTF) recommendations and Spina Bifida-specific biomedical information.

0-11 months

Clinical Questions

1. Do children with Spina Bifida receive preventive health care recommended by the USPSTF?¹

Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations and vision and hearing screens).⁹⁻¹⁰
2. Provide age-typical health promotion counseling (e.g. counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition).⁹⁻¹⁰ Counseling should be individualized to accommodate for Spina Bifida comorbidities such as having a shunt, mobility impairments, orthopedic deformities, developmental delays, and bowel and bladder management.^{10,12} (Bowel Function and Care Guidelines, Mobility Guidelines, , Neuropsychology Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)
3. Counsel families about possible future medical and social needs related to living with Spina Bifida. Needs might include latex allergies,^{5,13} chronic urinary issues,^{3,13} problems with shunts, and achieving an inclusive environment.^{2,13} (Latex Allergy Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Urology Guidelines)
4. Monitor the child for neglect and/or abuse.⁹⁻¹⁰ (Family Functioning Guidelines)

1-2 years 11 months

Clinical Questions

1. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?¹

Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations, vision and hearing screens).⁹⁻¹⁰
2. Provide age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition).⁹⁻¹⁰ Counseling should be individualized to accommodate for Spina Bifida comorbidities such as having a shunt, bowel and bladder management, mobility impairments, orthopedic deformities and developmental delays.^{10,12} (Bowel Function and Care Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)
3. Counsel families about possible future medical and social needs related to living with Spina Bifida. Needs might include latex allergies,¹² chronic urinary issues,^{3,12} problems with shunts, achieving an inclusive environment, overweight/obesity risk,⁹ importance of physical and recreational activity,¹ managing unexpected changes in function, keeping regular medical appointments, and pain.^{2,13} (Bowel Function and Care Guidelines, Latex Guidelines, Nutrition, Metabolic Syndrome, and Obesity Guidelines, Physical Activity Guidelines, Urology Guidelines)
4. Monitor the child for neglect and/or abuse.⁹⁻¹⁰ (Family Functioning Guidelines)

3-5 years 11 months

Clinical Questions

1. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?¹

Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations and vision and hearing screens).⁹⁻¹⁰
2. Promote age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and skill development).⁹⁻¹⁰
Counseling should be individualized to accommodate for Spina Bifida comorbidities such as having a shunt, mobility impairments, orthopedic deformities, developmental delays, and bowel and bladder management.⁹⁻¹⁰ (Bowel Function and Care Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)
3. Counsel families about possible future medical and social needs related to living with Spina Bifida. Needs might include latex allergies⁵, chronic urinary issues,^{5,13} problems with shunts, achieving an inclusive environment, overweight/obesity risk,⁹ importance of physical and recreational activity,⁹ managing unexpected changes in function, keeping regular medical appointments, and pain.^{2,13} (Bowel Function and Care Guidelines, Latex Guidelines, Nutrition, Metabolic Syndrome, and Obesity Guidelines, Physical Activity Guidelines, Urology Guidelines)
4. Monitor the child for neglect and abuse.⁹⁻¹⁰ (Family Functioning Guidelines)

6-12 years 11 months

Clinical Questions

1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities?
2. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?⁹
3. When does pain become a common secondary condition? What are the characteristics that increase risk for pain complaints?
4. What are the characteristics that increase the risk for hypertension?

Guidelines

1. Monitor that the child is making routine well-child visits to their primary care provider to receive age-appropriate health promotion and preventive services, including age-appropriate screenings for: (clinical consensus)
 - Hypertension.^{4,10,14} Since there is no agreement on blood pressure targets for patients with Spina Bifida, it is recommended that baseline blood pressure be monitored to know what is considered hypertensive for a particular child. (clinical consensus)
 - Iron deficiency.⁹⁻¹⁰
 - Lipid disorders.⁹⁻¹⁰
 - Overweight/obesity, including the role in hypertension.⁹ (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
 - Abuse, neglect, and/or violence.¹⁰ (Family Functioning Guidelines)
 - Social isolation, anxiety, and depression.⁹⁻¹⁰ (Mental Health Guidelines)
 - Motor vehicle and wheelchair safety.^{4,9-10} (Mobility Guidelines)
2. Provide counseling about tobacco and illicit drug use and refer the family to an

- appropriate treatment program if needed.⁹⁻¹⁰
3. Provide guidance on skin cancer prevention.⁹⁻¹⁰
 4. Provide information about adaptive physical and recreational activities keeping in mind the particular child's degree of mobility.⁷ (Physical Activity Guidelines)
 5. Provide information about accessible physical activity and recreational opportunities in the community.⁹ (Physical Activity Guidelines)
 6. Monitor for pain and changes in pain using an appropriate pain scale for the child's level of cognition and communication, as pain may not be clearly recognized due to the unique neurologic status of children with Spina Bifida. Be aware that depending on their cognitive status, the child may not be able to give specific answers to questions such as the severity, frequency and duration of the pain. Proceed with appropriate evaluation and treatment.^{3,9,16}
 7. Monitor for comorbid conditions that are specific to children with Spina Bifida, both during visits specifically-intended to monitor Spina Bifida conditions as well during well-child visits. (Bowel Function and Care Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Skin Care Guidelines, Urology Guidelines)
 - Shunt concerns.⁵ Ask about any neurologic changes.
 - Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep.⁹ (Sleep Related Breathing Disorders Guidelines)
 - Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. (clinical consensus)
 - Constipation, urinary tract infections (UTIs), renal function, and problems with bowel and bladder regimens. Provide prescriptions for routine bowel medications, treatment of recurring UTIs, monitor for adherence to bowel and bladder management program and changes in bowel and bladder function.³ (Bowel Function and Care Guidelines, Urology Guidelines)
 - Skin breakdown and pressure injury.⁷ Urge the family and child (if appropriate) to perform daily skin checks. Recommend that the child's skin is properly moisturized, and that appropriate weight shifting is taking place based on the child's neurologic level. (Mobility Guidelines, Skin Care Guidelines)
 - Adaptive equipment needs, including for orthoses, crutches, walkers, and wheelchairs.^{3,6-7,9} Make referrals to necessary subspecialists. (clinical consensus) (Mobility Guidelines)
 - Osteoporosis. Encourage weight-bearing activities for at least one hour per day to promote bone health as well as for its social benefits. (clinical consensus) (Mobility Guidelines, Orthopedics Guidelines, Physical Activity Guidelines)
 8. Promote care coordination between Spina Bifida-specific subspecialists and primary care providers.⁷ (Care Coordination Guidelines]
 9. Educate families on early signs of chronic conditions related to Spina Bifida.^{5,7}

13-17 years 11 months

Clinical Questions

1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities?
2. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?⁹

3. What are the parameters to begin screening for metabolic syndrome?
4. What are parameters and modifications are needed (if any) to begin education related to sexually transmitted infections, partner violence, and human papillomavirus (HPV) immunization?
5. What characteristics may put a child in this age range at risk for low self-rated health and health-related quality of life (HRQOL)? Do increasing acute medical conditions affect this?⁵ Are there interventions or supports that may mitigate this?
6. When does sleep apnea become notable and what are the risks?⁷
7. Do interventions make a difference in weight control, participation in physical or recreational activities, and pain control?
8. Can emergency department or hospital admissions be avoided?^{3,9,6}
9. Do hypertension interventions make a difference?

Guidelines

1. Monitor that the child is making routine well-child visits to his or her primary care provider to receive age-appropriate health promotion and preventive services, including age-appropriate screenings for:
 - Hypertension.^{4,9-10} Since there is no agreement on blood pressure targets for patients with Spina Bifida, it is recommended that baseline blood pressure is monitored to know what is considered hypertensive for the particular child. (clinical consensus)
 - Iron deficiency.⁹⁻¹⁰
 - Lipid disorders.⁹⁻¹⁰
 - Overweight/obesity, including the role in hypertension.⁹⁻¹⁰ (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
 - Abuse, neglect, and/or violence.¹⁰ (Family Functioning Guidelines)
 - Social Isolation, Anxiety, Depression.⁹⁻¹⁰ (Mental Health Guidelines)
 - Motor vehicle and wheelchair safety.^{4,9-10} (Mobility Guidelines)
 - Contraceptive use, pregnancy, and sexually transmitted diseases. (as age appropriate) ¹ (Sexual Health and Education Guidelines, Women's Health Guidelines, Men's Healthcare Guidelines)
2. Provide counseling about tobacco and illicit drug use and refer the family to an appropriate treatment program if needed.⁹⁻¹⁰
3. Provide guidance on skin cancer prevention.⁹⁻¹⁰
4. Provide information about adaptive physical and recreational activities keeping in mind the particular child's degree of mobility.⁷ (Physical Activity Guidelines)
5. Provide information about accessible physical activity and recreational opportunities in the community.⁹ (Physical Activity Guidelines)
6. Monitor for pain and changes in pain using an appropriate pain scale for the child's level of cognition and communication, as pain may not be clearly recognized due to the unique neurologic status of children with Spina Bifida. Be aware that depending on their cognitive status, the child may not be able to give specific answers to questions such as the severity, frequency and duration of the pain. Proceed with appropriate evaluation and treatment.^{2-3,6}
7. Monitor for comorbid conditions that are specific to children with Spina Bifida, both during visits that are specifically intended to monitor Spina Bifida conditions as well during well-child visits. (Neurosurgery Guidelines, Orthopedics Guidelines, Bowel Function and Care Guidelines, Urology Guidelines, Mobility Guidelines, Skin Care Guidelines)
 - Shunt concerns.⁴ Ask about any neurologic changes.
 - Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep.⁹

- Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. (clinical consensus)
 - Constipation, urinary tract infections (UTIs), renal function, and problems with bowel and bladder regimens. Provide prescriptions for routine bowel medications, treatment of recurring UTIs, monitor for adherence to bowel and bladder management program and changes in bowel/bladder function.³ (Bowel Function and Care Guidelines, Urology Guidelines)
 - Skin breakdown and pressure injury.⁷ Urge the family and child (if appropriate) to perform daily skin checks. Recommend that the child's skin is properly moisturized, and that appropriate weight-shifting is taking place based on the child's neurologic level. (Mobility Guidelines, Skin Care Guidelines)
 - Adaptive equipment needs, including for orthoses, crutches, walkers, and wheelchairs.^{3,6-7,9} Make referrals to necessary subspecialists. (clinical consensus) (Mobility Guidelines)
 - Osteoporosis. Encourage weight-bearing activities for at least one hour per day to promote bone health as well as for its social benefits. (clinical consensus) (Mobility Guidelines, Orthopedics Guidelines, Physical Activity Guidelines)
8. Promote care coordination between Spina Bifida-specific subspecialists and primary care providers.⁷ (Care Coordination Guidelines)
 9. Educate families on early signs of chronic conditions related to Spina Bifida.^{5,7}

18+ years

Clinical Questions

1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities?
2. Do adults with Spina Bifida receive preventive health care recommended by the USPSTF?¹
3. What characteristics may put the adult with Spina Bifida at risk of low self-rated health and health-related quality of life? Do increasing acute medical conditions affect this?⁵ Are there interventions or supports that may mitigate this?
4. Can future health issues and health care utilization be predicted? What is the prevalence and risks for common and Spina Bifida-related conditions? What is the expected cost of care?
5. Can the number of emergency department or hospital admissions be mitigated?^{3,9,6}

Guidelines

1. Monitor that the adult is receiving typical and age-related health promotion and preventive services, including screening or counseling about:
 - Hypertension.^{4,9-10} Since there is no agreement on blood pressure targets for patients with Spina Bifida, it is recommended that baseline blood pressure be monitored to know what is considered hypertensive for the particular child.
 - Lipid disorders.⁹ Treat as needed.
 - Overweight/obesity. Counsel as to healthy diet and exercise habits.¹ (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
 - Cancer, including skin cancer.⁹ Promote age-appropriate screening.
 - Diabetes and metabolic syndrome.⁹ Screening and treatment as needed.
 - Fall prevention.⁹
 - Adaptive physical activity.⁷ (Physical Activity Guidelines)
 - Depression and anxiety.⁹ (Mental Health Guidelines)
 - Smoking and illicit drug use.¹
 - Hearing and vision.⁹

2. Monitor for comorbid conditions that are specific to adults with Spina Bifida, both during visits that are specifically intended to monitor Spina Bifida conditions as well as routine visits to their PCP. (Bowel Function and Care Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Skin Care Guidelines, Urology Guidelines) Check for:
 - Shunt concerns.⁴ Monitor for neurologic changes.
 - Sleep apnea.⁹ May need a referral for pulmonary evaluation and sleep study.
 - Skeletal and limb deformity.⁷ Check for new problems with positioning or brace use and new pain.
 - Osteoporosis. Counsel about the need for weight-bearing activities.¹
 - Pain.^{2,15} Use age- and cognition-appropriate pain scale to assess.
 - Constipation, urinary tract infections (UTIs), renal function, and problems with bowel and bladder regimens.^{3,7} Provide prescriptions for routine bowel medications, treatment of recurring UTIs, monitor for adherence to bowel and bladder management program and for changes in bowel/bladder function³ (Bowel Function and Care Guidelines, Urology Guidelines)
 - Skin breakdown and pressure injury.⁹ Encourage adults to conduct frequent skin checks and to shift their weight at least every two hours.
 - Lymphedema.¹⁶
 - Adaptive equipment needs such as for orthoses, crutches, walkers, and wheelchairs.^{2-3,7,9,16-19} Make referrals to necessary subspecialists.
3. Promote care coordination between Spina Bifida-specific subspecialists and primary care providers.⁷ (Care Coordination Guidelines)
4. Educate adults about early signs of chronic conditions related to Spina Bifida.^{5,7}
5. Counsel about and monitor for sexually transmitted infections, use of different types of contraceptives, and violence among intimate partners. (clinical consensus) (Men's Health Guidelines, Sexual Health and Education Guidelines, Women's Health Guidelines)
6. Provide counseling about family planning and possible fertility and genetic counseling to individuals interested in pregnancy. Recommend counseling about prenatal vitamins and folic acid. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
7. Promote self-management for health and health care services. Assess the adult's ability to perform routine care needs such as bowel, bladder, and skin-check regimens, their ability to detect changes in their health status, and their awareness of their need for provider services to maximize their independence.^{6-7,9} (Self-Management and Independence Guidelines)

Research Gaps

1. Can future health issues and health care utilization be predicted? What are the common health conditions that are preventable or easily amenable to interventions? What is prevalence and risks for common and Spina Bifida-related conditions? What are the common causes for preventable or unanticipated mortality? What is the expected cost of care?
2. What are the common physical and mental health conditions associated with emergency department visits and hospitalizations? What are key anticipatory guidance or management strategies to prevent the need for higher levels of care? Can emergency department or hospital admissions be avoided, and how? Does a medical home help to prevent admissions for all age groups?¹⁸

3. What are the characteristics that may put an individual with Spina Bifida at risk of low self-rated health and HRQOL? Are there preventive care interventions or supports that may mitigate this?
4. What adaptations to general care practice and the USPSTF recommendations do individuals with Spina Bifida need, taking into account patient-centered perspectives and biomedical information?
5. What long-term care planning is needed to support the best health for adults with Spina Bifida?

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Prenatal Counseling

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Introduction

The prenatal diagnosis of Spina Bifida or a neural tube defect (NTD) is a pivotal moment for families. NTDs can be open or closed, and the type of finding determines important counselling considerations, particularly as closed NTDs often do not need surgical intervention in the neonatal period. The manner in which information about NTDs is conveyed is critical to the family's future and provides the basis for important family decisions (e.g. possible fetal surgery versus postnatal closure for open NTDs, adoption and termination).¹ Families may experience intense emotions such as uncertainty, confusion, grief, anxiety and anger. At the same time, families need to absorb a great deal of information about their options and to understand the risks and benefits of those options. Differing cultures have disparate perceptions around disability, and this may impact families' expectations about treatment options and/or their understanding of the condition. It is essential that parents and caregivers are told about the clinical course and the anticipated strengths and challenges associated with Spina Bifida. This information should be individualized and provided in a neutral and collaborative manner that meets the needs, values, and beliefs of each family.²⁻³ Families should be offered the opportunity to meet with key members of the Spina Bifida team.^{1,4-5} Neurosurgeons experienced with and dedicated to caring for patients with NTDs are uniquely qualified to discuss both short term and realistic long-term expectations and challenges facing a child born with Spina Bifida.

Outcomes

Primary

1. Convey information about medical care and lifelong impact of Spina Bifida in a value-neutral, collaborative manner while seeking from families an understanding of their needs, values and beliefs.

Secondary

1. Provide education regarding all treatment options available to women carrying fetuses affected by Spina Bifida, including fetal surgery, postnatal closure, adoption and termination.

Tertiary

1. Provide families with information about providers and resources that will assist in the caring for their child with Spina Bifida.

Clinical Questions

1. What are the essential components of prenatal consultation for any child affected by a potentially disabling condition?
2. What are the essential treatment options that are to be discussed as part of the prenatal consultation for a family with a fetus affected by a NTD?
3. Are there critical language elements to be considered?
4. Are there critical and specific medical providers that should participate in the prenatal consultation for a family affected by a fetus with a NTD?

Guidelines

1. Convey information about the medical care and lifelong functional impact of Spina Bifida in a value-neutral, collaborative manner while seeking from families an understanding of their needs, values, and beliefs.
 - Ideally, consultations with the parents will take place shortly after identification of the NTD is made at the 18th week and before the 24th week of gestation.⁵⁻⁶
 - Efficient consultation is essential and should happen quickly, soon after the identification of the NTD, to allow parents the broadest array of options and to provide enough time to consider the option for fetal surgery. (clinical consensus)
 - Base consultation with families on a collaborative, shared decision-making model that includes the medical team and parents. (clinical consensus)
 - Avoid using words that assign value or bias, such as “risk,” “bad news,” and “bad outcomes.” Use words that impart the importance of the decision, such as “important news,” “significant outcome,” and “potential challenges.” This allows parents to assign their own values to the news provided.³⁻⁴
 - Offer a review of prenatal testing and results to clarify any misunderstandings or confusion that may exist.¹ Typically, the diagnosis is made by a high-resolution ultrasound examination that is performed during the second trimester at a maternal–fetal medicine unit. An ultrasound can define the location and size of the lesion, whether it is open or closed (in most instances), and secondary findings such as hydrocephalus.⁷ Given the increased risk of other abnormalities, fetal echocardiography should be considered. Genetic evaluation by amniocentesis for chromosomal microarray should be recommended because the identification of a genetic abnormality in a fetus with an NTD has important implications for counseling regarding prognosis, pregnancy management, and determining whether the patient is a candidate for in-utero NTD repair.⁸⁻⁹ Measurement of amniotic fluid acetylcholinesterase helps to differentiate between open and closed NTDs and is a component of many preoperative evaluations for fetal repair. Fetal MRI also may be considered for assessment of unclear findings on ultrasonography.¹⁰
 - Expect to provide critical information about the likelihood of survival and the spectrum of outcomes (i.e. neurosurgical, cognitive, developmental, urologic, orthopedic, dermatologic) for children with NTDs.^{5,11}
 - Discuss disability. Provide information on outcomes with a lifespan approach.³
 - Review general principles associated with lesion levels, as well as the difficulty with providing specific predictions based on lesion level.¹²
 - Review treatment options for conditions associated with NTDs with an emphasis on functional outcomes.¹¹
2. Review evidence-based treatment options with the family, including fetal surgery.
 - Treatment options should include prenatal closure for open NTDs offered at treatment centers with expertise in the surgical and obstetrical management of NTDs.^{4-6,13} It is recommended that fetal surgery and the details of the surgical and obstetrical impacts should be reviewed by surgeons/obstetricians with experience managing high-risk pregnancies and/or providing care to infants with NTDs.^{4-6,11,13}
 - In addition to the option for prenatal closure, also present the option for term delivery and postnatal closure for open NTDs. Explain to parents and caregivers that caesarian delivery at 37 weeks and closure within 24 hours of delivery is generally recommended when the decision is made for postnatal closure. Ensure that the parents are aware of what to expect at birth and after the surgery.¹²

- Although closed NTDs usually do not require surgical intervention in the newborn period, it is recommended that they have the same monitoring and investigations in the newborn period as open NTDs. (clinical consensus)
 - Present adoption as an option for parents who are not open to termination but are not able to raise a child with a disability. (clinical consensus)
 - Review termination of the pregnancy as another option for the family.⁴
3. Offer families the opportunity to meet with key members of the Spina Bifida care team:
- Specialists in fetal medicine and/or obstetrical medicine are familiar with managing pregnancies complicated by a prenatal diagnosis of NTD. These providers are first to share the results of the testing. The prenatal diagnosis of Spina Bifida should be made in a value-neutral manner.^{4,13}
 - Neurosurgeons provide information about management approaches such as fetal surgery and postnatal closure.⁴⁻⁵ Neurosurgeons experienced with and dedicated to caring for patients with neural tube defects (NTDs) are uniquely qualified to discuss both short term and realistic long-term expectations and challenges facing a child born with Spina Bifida.
 - Experts in clinical genetics can clarify test results, discuss the genetics of NTDs, provide information about folic acid, and discuss recurrence risk and potential impact on future pregnancies.^{1,3}
 - Developmental pediatricians, advanced practice nurses, and physiatrists focus on childhood disabilities and how to optimize function. These specialists can provide essential insight into potential medical needs and functional goals across the lifespan. They provide parents with evidence-based, up-to-date information. They can also assist with resource identification, access to care, and utilization, including caregiver support and mental health resources.^{1,14} (Family Functioning Guidelines, Mental Health Guidelines)
 - Social workers provide critical emotional support and screening for parental mental health and are recommended to be included in all consultations. They provide families with links to important financial resources and sources of emotional support, including caregiver support and mental health resources.^{5,13} They can also provide information to families about local, national, and international sources.¹ (Family Functioning Guidelines, Mental Health Guidelines)
 - Urologists and orthopedists can provide more detailed discussions on interventions available for optimizing functional outcomes.⁵ (Orthopedic Guidelines, Urology Guidelines)
 - Neonatologists can provide information and resources in advance about the child's immediate care needs such as breastfeeding, skin-to-skin care, and tours of the neonatal intensive care unit (NICU).^{5,13}
4. Offer information about what to expect at birth.
- Review that the child may need to be admitted to a special care or intensive care nursery (all with open NTDs, some closed NTDs may be cared for collaboratively with the nursery and community teams) and that psychosocial support is available to them in that setting.⁴ (Family Functioning Guidelines, Mental Health Guidelines)
 - Help families to anticipate that specialists in Spina Bifida will need to be present at the delivery to examine the child, that there exists the possibility that intravenous fluids/antibiotics will be necessary, and of the possibility that the child will need additional support.⁴

- If fetal surgery is not an option, review the timing of delivery by 37 weeks¹² and post-natal management. The first step should be the closure of the spinal defect within 24 hours¹² followed by attention to hydrocephalus as indicated.¹⁵
 - Emphasize typical aspects of newborn care, including breastfeeding and skin-to-skin care.¹⁶⁻¹⁷
 - Counsel families who choose to continue the pregnancy that there are many normal aspects of pregnancy, caregiving for the newborn and parenting across the lifespan. (clinical consensus)
5. Discuss folic acid and recurrence risk.
- Reassure the parents that while NTDs are not completely understood, this birth defect was not something that was caused by their actions.¹
 - Reassure the parents that while folic acid can help diminish the chance of a pregnancy being affected by NTD, it is not entirely preventative.¹⁸⁻¹⁹
 - Counsel women about their recurrence risk and review the 1991 U.S. Public Health Service guideline for daily consumption of 4 milligrams (4000 micrograms) of folic acid beginning at least one month (but preferably 3 months) before they start trying to get pregnant and continuing through the first three months of pregnancy. (Women's Health Guidelines)

Research Gaps

There exist limited studies describing parental experiences and the variables that influence decision-making. While the short-term outcome data for prenatal surgery is promising, long-term outcome data are not yet available. In addition, there is limited data on trauma informed care and this approach to prenatal counselling. There is also limited data on intervention strategies to facilitate coping by parents and families as they navigate the health care system.

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Transition

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Introduction

The primary goal of transition from pediatric to adult health care is to maximize lifelong functioning through the provision of quality, developmentally-appropriate health care that continues uninterrupted as the individual moves from adolescence to adulthood.¹ Coordination between the individual, family, health care providers, school, and allied health services throughout the transition process enables young adults with Spina Bifida to optimize their ability to eventually assume adult roles and activities.

Health care transition is a process that includes:

1. Preparation for engaging in adult health care and adult responsibilities through planning and coaching during pediatric years, school, and home settings.
2. Handoff of care responsibilities from pediatric to adult providers and from parents to young adults as they are developmentally able.
3. Transfer of care to adult providers and health care settings between the ages of 18-21.²

Adolescents and young adults with Spina Bifida have increased hospitalizations for chronic condition exacerbations, such as urinary tract infections and skin breakdown, and more difficulty accessing health care services than their age-matched peers.³ Furthermore, they are less likely to achieve emerging adult milestones such as leaving home, attending college, finding employment, developing romantic relationships, and having multiple friendships. Executive function, socioeconomic status, intrinsic motivation, and parental fostering of independence are significant predictors of successful transition to adulthood.⁴ Patient-centered, comprehensive transition care is needed to address the chronic health condition, funding, care coordination, self-management, and social challenges that adolescents and young adults with Spina Bifida face when transferring to adult care and life. (Neuropsychology Guidelines, Self-Management and Independence Guidelines)

The following key elements have been identified as essential for transition programs for adolescents with chronic conditions:

1. Designate a transition program to support care coordination and transition to adult-oriented care;
2. Ensure flexibility regarding transfer timing based on the individual's cognitive development, physical abilities, social and financial situation, and health status;
3. Begin transition planning at ages,¹²⁻¹⁴ including creating a medical summary, identifying insurance coverage plans, and designating care providers throughout the transition process;
4. Support self-management development for health care navigation and chronic condition management (Neuropsychology Guidelines, Self-Management and Independence Guidelines);
5. Consider the adolescent's views and preferences regarding transition plans;
6. Designate time alone with the adolescent for at least part of their visit, if developmentally appropriate;

7. Identify adult providers to assume care prior to the transition; and
8. Provide chronic condition management and age-appropriate preventive care throughout transition.^{1,5-6} (Health Promotion Guidelines)

While transition focuses on adolescent age groups, the trajectory to maximize adult function and independence is fostered throughout the lifespan by setting expectations for adult independence and making the transition to adult care. While survival to adulthood for individuals with Spina Bifida now exceeds 85%, the degree of adult independence in the population varies.⁷ In general, individuals with higher lesions (i.e., above L2) and hydrocephalus are more dependent on others for bowel and bladder management, mobility, self-care, transfers, and activities of daily living.⁸⁻⁹ Individualized goals and interventions should be emphasized for adolescents with Spina Bifida because they tend to experience a two- to five-year delay in developing autonomy skills compared with their typically-developing peers.¹⁰ By age 30, approximately one-third of individuals with Spina Bifida are independent, one-third need supervision and occasional help, and one-third routinely need assistance for daily care needs.¹¹ Thus, lifelong assessment and interventions to maximize chronic condition management, family function, socialization, cognitive function and school performance, mental health, and self-management/self-care are foundational to participation, function, and quality of life in adulthood.^{9,12-15} (Self-Management and Independence Guidelines).

Transition resources and assessment tools can be found at www.gottransition.org.

Outcomes

Primary

1. Maximize health and participation in emerging adult milestones throughout the transition process for individuals with Spina Bifida.

Secondary

1. Provide patient-centered, comprehensive transition care that includes transition planning and care coordination beginning by age;¹⁴ self-management coaching; decision-making support; education and employment resources; and independent-living support.

Tertiary

1. Promote access to uninterrupted, developmentally-appropriate Spina Bifida condition management and preventative care throughout transition, specifically ages.¹⁴⁻²¹

0-11 months

Clinical Questions

1. How can a child's probable trajectory regarding future adult function and independence be identified?
2. What are barriers and facilitators to participating in emerging adult milestones for children with Spina Bifida?
3. What are the essential transition planning elements to promote uninterrupted access to care once children with Spina Bifida need adult care?

Guidelines

1. Provide families with a realistic, long-term orientation that includes a probable trajectory for adult function and expectations for optimal independence according to the child's abilities.^{7-11,16}
2. Provide information for families regarding long-term financial, insurance, and supportive living planning based on the child's probable trajectory into adult function.¹⁷

3. Set expectations as to where individuals with Spina Bifida can access comprehensive care throughout the lifespan, including transition care.¹⁸

1-2 years 11 months

Clinical Questions

1. How can a child's probable trajectory regarding future adult function and independence be identified?
2. What are barriers and facilitators to participating in emerging adult milestones for children with Spina Bifida?
3. What are the essential transition planning elements to promote uninterrupted access to care once children with Spina Bifida need adult care?

Guidelines

1. Provide updates for families regarding a probable trajectory for adult function and expectations for optimal independence according to the child's abilities and chronic condition status.^{7-11,16}
2. Provide updates for families on information regarding long-term financial, insurance, and supportive living planning based on the child's probable trajectory into adult function.¹⁷
3. Review expectations as to where individuals with Spina Bifida can access comprehensive care throughout the lifespan, including transition care.¹⁸

3-5 years 11 months

Clinical Questions

1. How can a child's probable trajectory regarding future adult function and independence be identified?
2. What are barriers and facilitators to participating in emerging adult milestones for children with Spina Bifida?
3. What are the essential transition planning elements to promote uninterrupted access to care once children with Spina Bifida need adult care?

Guidelines

1. Provide updates for families regarding a probable trajectory for adult function and expectations for optimal independence according to the child's abilities and chronic condition status.^{7-11,16}
2. Provide updates for families on information regarding long-term financial, insurance, and supportive living planning based on the child's probable trajectory into adult function.¹⁷
3. Review expectations as to where individuals with Spina Bifida can access comprehensive care throughout the lifespan including chronic condition management, preventative care, and transition care.¹⁸

6-12 years 11 months

Clinical Questions

1. How can a child's probable trajectory regarding future adult function and independence be identified?
2. What are barriers and facilitators to participating in emerging adult milestones for children with Spina Bifida?
3. What are the essential transition planning elements to promote uninterrupted access to care once children with Spina Bifida need adult care?

Guidelines

1. Provide updates for families regarding a probable trajectory for adult function and expectations for optimal independence according to the individual's abilities and chronic condition status.^{7-11,16}
2. Consider unique neuropsychological issues in transition and self-management. (Neuropsychology Guidelines, Self-Management and Independence Guidelines)
3. Provide updates for families on information regarding long-term financial, insurance, and supportive living planning based on the child's probable trajectory into adult function.¹⁷
4. Review expectations where individuals with Spina Bifida can access comprehensive care throughout the lifespan including chronic condition management, preventative care, and transition care.¹⁸
5. Review the clinic's transition policy with patients and families at age 12.¹⁹

13-17 years 11 months

Clinical Questions

1. How can a child's probable trajectory regarding future independence be identified?
2. What are barriers and facilitators to participating in emerging adult milestones for children with Spina Bifida?
3. What are child-centered perceptions of a successful transition experience?
4. What are the systems level barriers to successful transition and strategies that have effectively mitigated them?
5. What are the key transition readiness parameters for patients with Spina Bifida that can be measured over time?
6. What are the preventative and chronic condition management considerations in the transition age group, ages 14-21?
7. What are the essential transition planning elements to promote uninterrupted access to care once children with Spina Bifida need adult care?

Guidelines

1. Provide updates for children and families regarding a probable trajectory for adult function and expectations for optimal independence according to the individual's abilities and chronic condition status.^{7-11,16}
2. Consider neurocognitive assessment to identify cognitive, adaptive, or learning support needs if knowledge or skill gaps are identified. (Neuropsychology Guidelines, Self-Management and Independence Guidelines)
3. Discuss transition planning with children and families including:^{5,17,20-27}
 - a. Expectations of when the transfer to adult care will occur based on the individual's health condition, insurance/funding, cognitive development, and personal/family needs.
 - b. Expectations of who will provide care throughout the transition process.
 - c. Counselling regarding long-term financial, insurance, and supportive living (housing and transportation) plans, based on the individual's current needs and probable trajectory of adult function.
 - d. Information regarding the child's education and employment needs, such as vocational rehabilitation services, school transition planning as part of the Individualized Educational Plan [INSERT LINK], and adaptive vocational needs.
 - e. Preparation for decision-making supports and modalities that maximize the individual's ability to participate in decisions for themselves once they are age 18, such as a medical power of attorney, supportive decision-making, or

- guardianship. Referral for neurocognitive testing and to medical legal partnerships may be needed.
- f. Information regarding the Social Security Administration's Disability Determination Services before age 18, as applicable.
 - g. Creation of a medical summary including past medical and surgical history, current care plans, medications, allergies, vaccines, and current providers.
 - h. Self-management support. Consider using transition and self-management assessment tools to direct goals and interventions.¹⁹ (Self-Management and Independence Guidelines)
4. Ensure that the patient's views and preferences are included in transition planning.⁶
 5. Designate time alone with the child for at least part of their visit, if developmentally appropriate.⁶
 6. Consider having a designated transition clinic or care coordinator to support transition planning and coordination.⁵
 7. Ensure patient-centered and developmentally appropriate preventive and chronic condition management services are provided throughout transition. (Health Promotion Guidelines) Evaluate management plans and assess for necessary adaptive equipment and supplies to maximize independent function.^{18,22-23}

18+ years

Clinical Questions

1. What are barriers and facilitators to participating in emerging adult milestones for individuals with Spina Bifida?
2. What are the preventative and chronic condition management considerations in the transition age group?
3. What are the essential transition planning elements to promote uninterrupted access to care once children with Spina Bifida need adult care?
4. What are the best strategies to find and engage adult providers?

Guidelines

1. Provide updates for adults and families regarding a probable trajectory for adult function and expectation for optimal independence according to the individual's abilities and chronic condition status.^{7-11,16}
2. Consider unique neuropsychological issues in transition and self-management. (Neuropsychology Guidelines, Self-Management and Independence Guidelines)
3. Continue to assist with transition coordination as applicable including:^{5,13,20-25,28}
 - a. Assistance in identifying adult providers who accept the patient's insurance and can assume his/her care.
 - b. Counselling regarding long-term financial, insurance, and supportive living plans (housing, transportation, etc.) based on the individual's current needs and probable trajectory of adult function.
 - c. Information for education/employment transition support as applicable to the individual's need such as vocational rehabilitation services, school transition planning, as part of the Individualized Educational Plan [INSERT LINK], and adaptive vocational needs.
 - d. Decision-making supports and modalities that maximize the individual's ability to participate in decisions for themselves, such as a medical power of attorney, supportive decision-making, or guardianship. Referral to medical legal partnerships may be needed.
 - e. Adult disability determination information, if applicable.

- f. Creation of a medical summary including past medical and surgical history, current care plans, medications, allergies, vaccines, and current providers.
 - g. Self-management support. (Self-Management and Independence Guidelines) Consider using transition and self-management assessment tools to direct goals and interventions.¹⁹
4. Ensure that patient-centered and developmentally appropriate preventive and chronic condition management services are provided throughout transition. Evaluate management plans and assess for necessary adaptive equipment and supplies to maximize independent function.^{18,22-23}

Research Gaps

1. There is a need for studies that explore comprehensive care and best practices for children with Spina Bifida ages 13+ that address demographic and functional variables that influence transition, preventive care and condition management, strategies to improve access to quality health care throughout transition, and evaluation of effective self-management intervention programs.²⁸⁻³¹
2. Educational programs are needed to improve health care professionals' awareness and knowledge of the medical and social issues related to the transition from child to adult life and health care for people living with Spina Bifida.
3. Studies are needed to identify the risks and facilitators of secondary education and workforce and social participation for adults 18+ with Spina Bifida, in order to inform transition counseling and intervention.^{29,31}
4. Studies are needed to examine internet and technology applications for education and transition should be further explored.³²
5. Studies are needed to determine what barriers/facilitators adult health care providers experience in caring for adults with Spina Bifida, and how they can best support health care services for adults with Spina Bifida.
6. Studies are needed to determine the best assessments to measure transition readiness for adolescents with Spina Bifida.
7. There is a need to identify models of care for adults living with Spina Bifida that consider the specific needs of the individual, such as proximity to appropriate specialty and primary care services, transportation accessibility, and personal preference about the service delivery desired.

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Family Functioning

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Introduction

The impact on the family of having a child with Spina Bifida varies considerably.¹⁻² Overall, there are relatively low rates of family-level dysfunction (10-15%) and high levels of family resilience. Families of children with Spina Bifida show few differences in marital function compared to families of typically-developing children.^{4,13} The quality of the marital relationship prior to the birth of a child with Spina Bifida is an important predictor of parental adjustment. Findings suggest both positive and negative effects of having a sibling with Spina Bifida.¹⁴ Positive family attitudes toward Spina Bifida, overall family satisfaction, and the degree of sibling conflicts are important predictors of sibling adjustment.¹⁵⁻¹⁶ Family cohesion appears to be somewhat lower in families with children who have more significant cognitive impairment.³

Parental stress in families who are raising children with Spina Bifida is higher than in the general population, particularly among mothers, single parents, older parents, and/or economically disadvantaged and culturally-diverse parents. This is an example of cumulative risk, which is an important construct relevant to both family functioning and Spina Bifida care.³⁻⁴ The complexity of the child's condition and parental personality traits (e.g., extraversion, agreeableness, emotional stability) have also been found to be significant predictors of parental stress.⁹

It is common for parents of children with Spina Bifida to feel less satisfied and competent as parents, to have a lower quality of life, to have smaller social networks, to be less optimistic about the future, and to feel more isolated.⁵⁻⁶ Spina Bifida has a significant impact on parental adjustment and outcomes such as sleep duration, especially for mothers.⁷⁻⁸ Parents may also experience Post Traumatic Stress Disorder (PTSD) and depression.^{5,7,19} The alterations in parent functioning that occur as a result of having a child with Spina Bifida impact the psychosocial adjustment of children.¹⁰

Parents of youth with Spina Bifida exhibit higher levels of parental intrusiveness (i.e., overprotectiveness) and these behaviors are often linked with less functional child outcomes (e.g., lower levels of independent decision-making). However, these effects are often modified by the child's cognitive level (e.g., children with lower IQs have parents who are more controlling).¹¹ Parents of children with Spina Bifida tend to be less responsive to pubertal development than is the case in families of typically-developing children.¹²

Adolescence is a challenging time as parents and teens negotiate the gradual transfer of medical management from parent to child.⁴ Family conflict is associated with diminished medical adherence in this age group.¹⁷ Few family intervention studies specific to Spina Bifida have been conducted to better understand this important topic.¹⁸ (Self-Management and Independence Guidelines, Transition Guidelines)

Outcomes

Primary

1. Maximize family resilience and adaptation to multiple Spina Bifida-related and normative stressors as appropriate for developmental level.

Secondary

1. Maximize parental adaptation, expectations, and responsiveness to the changing developmental level of the child by identifying and reinforcing effective parenting techniques.
2. Maximize independence of the child within the family context, given developmental level and condition-related constraints.

Tertiary

1. Minimize parental and marital stress and maladaptation when raising a child with a serious chronic health condition.
2. Maximize family engagement in social activities, including parental self-care activities.
3. Maximize parental knowledge of Spina Bifida and advocacy.

0-11 months

Clinical Questions

1. What is the impact of having a child with Spina Bifida on family functioning (including parental adjustment, marital functioning, effect on the siblings and extended family, and familial participation in social activities) and how does this impact change as children move through various stages of lifespan development?
2. What resilience and vulnerability factors are predictive of familial adaptation at each level of child development?
3. What parenting behaviors facilitate adaptive child outcomes (including independence-related outcomes such as self-management and the transfer of health care responsibilities from parent to child) and how do these adaptive parenting behaviors vary developmentally?
4. What interventions or approaches can promote family functioning?

Guidelines

1. Refer families who have received a prenatal diagnosis of Spina Bifida for prenatal counseling and consultation with members of a Spina Bifida multidisciplinary clinical team. Assess family dynamics and adjustment in response to diagnosis.¹⁹ (Prenatal Counseling Guidelines)
2. Assess for postpartum depression. Provide information about Spina Bifida, parenting, treatments, support groups, and the Spina Bifida Association. (clinical consensus)
3. Coordinate services during the transition from the hospital stay to subsequent clinic follow-up, stressing the need for ongoing multi-specialty care.¹⁹
4. Teach necessary home care procedures such as post- surgical care, skin care, and clean intermittent catheterization, as needed.¹⁹
5. Assess family dynamics and adjustment (e.g., post-traumatic stress disorder in parents) during infancy.^{5,7,19}
6. Refer the parents or caregivers to infant intervention and appropriate state programs (e.g., Supplemental Security Income) and financial resources as needed. Provide financial counseling if necessary. (clinical consensus)
7. Provide support and ongoing counseling as needed to parents, the child, and siblings.^{16,19}

8. Provide anticipatory guidance for parents regarding strengths and possible cognitive and behavioral challenges in children with Spina Bifida and their siblings.^{16,19}
9. Teach parents to advocate for themselves and their child when working with medical, educational, and agency staff. (clinical consensus)
10. Be aware that although interventions should target all families, some families are particularly at-risk for adjustment and adherence difficulties (e.g., those from lower socioeconomic backgrounds and single-parent families).¹⁸
11. Assess the family's ability to carry out medical regimens, and identify possible barriers to adherence, such as need for caregiver support and parental beliefs regarding alternative therapies.¹⁷

1-2 years 11 months

Clinical Questions

1. What is the impact of having a child with Spina Bifida on family functioning (including parental adjustment, marital functioning, effect on the siblings and extended family, and familial participation in social activities) and how does this impact change as children move through various stages of lifespan development?
2. What resilience and vulnerability factors are predictive of familial adaptation at each level of child development?
3. What parenting behaviors facilitate adaptive child outcomes (including independence-related outcomes such as self-management and the transfer of health care responsibilities from parent to child) and how do these adaptive parenting behaviors vary developmentally?
4. What interventions or approaches can promote family functioning?

Guidelines

1. Provide support and ongoing counseling as needed to parents, the child, and siblings.^{16,19} (Mental Health Guidelines)
2. Be aware that although interventions should target all families, some families are particularly at-risk for adjustment and adherence difficulties (e.g., those from lower socioeconomic backgrounds and single-parent families).¹⁸
3. Promote effective parenting techniques or provide referral for such services.¹¹
4. Provide anticipatory guidance for parents regarding possible behavioral challenges and autonomy needs in children with Spina Bifida and their siblings.^{16,19} (Mental Health Guidelines, Neuropsychology Guidelines)
5. Assess family's need for additional counseling, financial resources, or other support services. (clinical consensus)
6. Inform families of advocacy resources and encourage them to contact the appropriate governmental and non-governmental authorities to obtain additional information, referrals, and support. (clinical consensus)
7. Encourage the parents or other primary caregivers to teach other family members or close friends how to provide for the child's specialized care needs and how to access other needed services. Alternatively, families can arrange for child care by trained professionals. (clinical consensus)
8. Educate parents about the importance of engaging in personal activities that promote parental well-being.⁵
9. Refer the parents to early intervention services, if these are not already in place. (clinical consensus)
10. Assess the family's ability to carry out medical regimens, and identify possible barriers to adherence, such as need for caregiver support and parental beliefs regarding alternative therapies.¹⁷

3-5 years 11 months

Clinical Questions

1. What is the impact of having a child with Spina Bifida on family functioning (including parental adjustment, marital functioning, effect on the siblings and extended family, and familial participation in social activities) and how does this impact change as children move through various stages of lifespan development?
2. What resilience and vulnerability factors are predictive of familial adaptation at each level of child development?
3. What parenting behaviors facilitate adaptive child outcomes (including independence-related outcomes such as self-management and the transfer of health care responsibilities from parent to child) and how do these adaptive parenting behaviors vary developmentally?
4. What interventions or approaches can promote family functioning?

Guidelines

1. Provide support and ongoing counseling as needed to parents, the child, and siblings.^{16,19}
2. Provide anticipatory guidance for parents regarding possible behavioral challenges and autonomy needs in children with Spina Bifida and their siblings.^{16,19} (Mental Health Guidelines, Neuropsychology Guidelines)
3. Teach parents to advocate for themselves and their child when working with medical, educational, and agency staff. (clinical consensus)
4. Be aware that although interventions should target all families, some families are particularly at-risk for adjustment and adherence difficulties (e.g., those from lower socioeconomic backgrounds and single-parent families).¹⁸
5. Re-assess parenting skills such as discipline, behavior management, and sibling relationships.¹¹
6. In the context of family functioning, address self-care abilities and refer to therapies (OT, PT). (clinical consensus)
7. Discuss issues that affect children with Spina Bifida when they transition to school. (clinical consensus)
8. Advise parents of their child's' right to free and appropriate education in the least restrictive environment through the public schools (i.e., explain services available under the Individuals with Disabilities Education Act). (clinical consensus)
9. Assess the family context for helping the child to develop self-management skills and to carry out medical regimens and identify possible barriers to adherence.¹⁷ (Self-Management and Independence Guidelines, Transition Guidelines)

6-12 years 11 months

Clinical Questions

1. What is the impact of having a child with Spina Bifida on family functioning (including parental adjustment, marital functioning, effect on the siblings and extended family, and familial participation in social activities) and how does this impact change as children move through various stages of lifespan development?
2. What resilience and vulnerability factors are predictive of familial adaptation at each level of child development?
3. What parenting behaviors facilitate adaptive child outcomes (including independence-related outcomes such as self-management and the transfer of health care responsibilities from parent to child) and how do these adaptive parenting behaviors vary developmentally?

4. What interventions or approaches can promote family functioning?

Guidelines

1. Provide support and ongoing counseling for parents, the child, and siblings, as needed.^{16,19}
2. Provide anticipatory guidance for parents regarding possible behavioral challenges and autonomy needs in children with Spina Bifida and their siblings.^{16,19} (Mental Health Guidelines, Neuropsychology Guidelines)
3. Be aware that although interventions should target all families, some families are particularly at-risk for adjustment and adherence difficulties (e.g., those from lower socioeconomic backgrounds and single-parent families).¹⁸
4. Assess family dynamics and relationships with school staff. (clinical consensus)
5. Have detailed discussions about appropriate interventions to address academic and social difficulties. Provide parents with current and accurate information about various school settings. For each type of setting, identify potential gaps and determine the impact that such a setting has on family members and the family system. (clinical consensus)
6. Encourage advocacy activities and resources and motivate parents to advocate for themselves and their children with medical, educational, and agency staff. (clinical consensus)
7. Advise parents of their child's right to free and appropriate education in the least restrictive environment through the public schools (i.e., explain services available under the Individuals with Disabilities Education Act and Section 504 of Vocational Rehabilitation Act of 1973). (clinical consensus)
8. Serve as a resource to school systems regarding health issues, individualized educational planning [INSERT LINK TO IEP/504], and socialization. (clinical consensus)
9. Reinforce appropriate family leisure activities. (clinical consensus)
10. Reinforce effective parental discipline, behavioral management, and expectations.¹¹
11. Encourage the family to facilitate medical self-management in their children with Spina Bifida, as developmentally appropriate.^{17,20-21} (Self-Management and Independence Guidelines)
12. Facilitate parents' understanding of the importance of fostering their child's independence and participating in chores and other activities of daily living.¹⁹ (Self-Management and Independence Guidelines)
13. Encourage social activities such as sleepovers, camp overnights, dating, and social and recreational activities outside the home. Encourage development and maintenance of friendships.²²
14. Emphasize positive attitudes, assertiveness, and self-empowerment of family members. (clinical consensus)
15. Encourage the family to develop strategies that gradually empower their children toward independence such as decision-making and problem-solving.^{11,20-21} (Self-Management and Independence Guidelines)
16. Assess the family context for helping the child to develop self-management skills and to carry out medical regimens and identify possible barriers to adherence.¹⁷ (Self-Management and Independence Guidelines, Transition Guidelines)

13-17 years 11 months

Clinical Questions

1. What is the impact of having a child with Spina Bifida on family functioning (including parental adjustment, marital functioning, effect on the siblings and extended family,

- and familial participation in social activities) and how does this impact change as children move through various stages of lifespan development?
2. What resilience and vulnerability factors are predictive of familial adaptation at each level of child development?
 3. What parenting behaviors facilitate adaptive child outcomes (including independence-related outcomes such as self-management and the transfer of health care responsibilities from parent to child) and how do these adaptive parenting behaviors vary developmentally?
 4. What interventions or approaches can promote family functioning?

Guidelines

1. Provide support and ongoing counseling for parents, child, and siblings, as needed.^{16,19}
2. Be aware that although interventions should target all families, some families are particularly at-risk for adjustment and adherence difficulties (e.g., those from lower socioeconomic backgrounds and single-parent families).¹⁸
3. Assess parent-child communication and their relationship. Aid parents to encourage the development of autonomy in their child with Spina Bifida.¹¹ (Self-Management and Independence Guidelines)
4. Encourage the family to begin planning for their child's transition to adult health care. (Transition Guidelines)
5. Begin discussions of other important developmental milestones, including educational and vocational achievement, living independently, and community participation. (Transition Guidelines)
6. Give advice to the child and family about the right to free and appropriate education in the least restrictive environment through the public schools (i.e., explain services available under the Individuals with Disabilities Education Act and Section 504 of Vocational Rehabilitation Act of 1973). (clinical consensus)
7. Assist with normative sexual education, as well as specific issues relevant to the teen's condition. Work with the teen to navigate sexual expression in a safe and mature fashion. (Sexual Health and Education Guidelines)
8. Continue to encourage the family to facilitate medical self-management in their child with Spina Bifida.²⁰⁻²¹ (Self-Management and Independence Guidelines)
9. Assess the family context for helping the child to develop self-management skills and to carry out medical regimens and identify possible barriers to adherence.¹⁷ (Self-Management and Independence Guidelines, Transition Guidelines)

18+ years

Clinical Questions

1. What is the impact of having a young adult with Spina Bifida on family functioning (including parental adjustment, marital functioning, effect on the siblings and extended family, and familial participation in social activities) and how does this impact change as children move through various stages of lifespan development?
2. What resilience and vulnerability factors are predictive of familial adaptation at each level of child development?
3. What parenting behaviors facilitate adaptive child and adult outcomes (including independence-related outcomes such as self-management and the transfer of health care responsibilities from parent to child) and how do these adaptive parenting behaviors vary developmentally?
4. What interventions or approaches can promote family functioning?

Guidelines

1. Provide support and ongoing counseling for parents, young adults, and siblings, as well as older adults with Spina Bifida, as needed.^{16,19}
2. Work with families to support the development of maximal vocational and social independence.¹⁹
3. Continue to work with the family to support medical self-management in their young adult.²⁰⁻²¹
4. Continue working with the family to ensure a successful transition to adult health care. (Transition Guidelines)
5. Work with the young and older adults to navigate sexual expression in a safe and mature fashion. (Sexual Health and Education Guidelines)
6. Assess the family context for helping the young adult to develop self-management skills and to carry out medical regimens and identify possible barriers to adherence.¹⁷ (Self-Management and Independence Guidelines, Transition Guidelines)

Research Gaps

1. What interventions are available to maximize familial resilience and adaptation at each level of the child's development?
2. What interventions are available to facilitate adaptive parenting behavior?
3. What interventions are available to enhance familial, marital, and parental adjustment outcomes?
4. What interventions are available to support families as they transfer medical management from parent to child, and the transition from pediatric to adult health care?
5. How does the characteristic cognitive profile of children and young adults with Spina Bifida complicate the unfolding of self-management within the family context?

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Mental Health

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Introduction

Multiple studies have shown that children with Spina Bifida have lower Health Related Quality of Life (HRQOL) than both typically developing individuals without Spina Bifida and children with other chronic health conditions.¹⁻⁵ Variables such as resilience (e.g., attitude towards Spina Bifida, hope and future expectations, coping skills) have been strongly related to higher HRQOL and quality of life (QOL). In contrast, depression, a lack of optimism and reduced executive functioning are related to lower QOL/HRQOL.⁵ The interplay between the neuropsychological patterns of development in children, family functioning and quality of life is the context within which the mental health of children with Spina Bifida is best understood.

Children with Spina Bifida tend to score below average on measures of neuropsychological functioning that involve the construction or integration of information.⁶⁻⁷ The ability to shift attention appropriately (sometimes referred to as executive functioning) is important to social development. Impairments in this area are associated with subsequent internalizing of symptoms (i.e., depressive and anxiety symptoms).⁸

Children with Spina Bifida also tend to have social difficulties, including social immaturity and passivity, fewer friends, and fewer social contacts outside of school. They also have fewer romantic relationships during adolescence.⁹⁻¹¹ These social difficulties appear to continue into adulthood.^{9,12} Youth with Spina Bifida may also exhibit lower levels of sexual maturation, knowledge, and experience.¹³⁻¹⁵

Children with Spina Bifida are more dependent on their parents for guidance, show less intrinsic motivation at school and exhibit less behavioral autonomy at home.^{9,12,17} Levels of decision-making autonomy lag behind typically developing peers by about two years.¹⁷ Pain and depressive symptoms interfere with social involvement.¹⁸

Children with Spina Bifida exhibit lower levels of participation in physical activities and activities of daily living.¹⁹⁻²⁰ Higher levels of physical activities are related to adaptive outcomes (i.e., participation and HRQOL). Some evidence exists that weight management interventions that include physical activities are effective in this population.²¹

The transition from pediatric to adult Spina Bifida health care poses significant challenges.²³ For instance, the reported quality of health tends to decline from adolescence to young adulthood, presumably due to difficulties in navigating the transition to health care for adults with Spina Bifida.^{21,23}

Regarding psychosocial adjustment during emerging adulthood, young adults with Spina Bifida, like their younger counterparts, are at-risk for depressive symptoms and anxiety²³⁻²⁴, but they are less likely to engage in at-risk behaviors than their typically developing peers (e.g., using alcohol and having multiple sexual partners).²⁵ With respect to relationship quality, 43 to 77% live with their parents²⁶⁻²⁷ but over half (52-68%) have had a romantic relationship, although this latter rate is lower than in typically developing young adults.²⁷ The lowest level of life satisfaction is in the areas of romantic relationships, employment, and financial

independence.²⁸⁻³¹

Regarding educational and vocational outcomes, emerging adults with Spina Bifida are less likely to go to college than typically developing youth.^{22,24,25,27,31} With respect to employment, recent studies report rates of full- or part-time employment ranging from 36-48%^{1,31}, which are significantly lower than those found in typically developing youth and in those with other chronic conditions. With respect to community participation and social integration, participation in leisure and recreational activities tends to be low, with over 50% participating in no activities.²⁸ Studies that examine the employment and community participation of middle-aged adults with Spina Bifida document a decline in workforce participation over time, particularly among those with high levels of motor impairment and lower educational levels.³²⁻³³ Bowel and bladder incontinence is also associated with unemployment and social isolation among adults with Spina Bifida.³⁴⁻³⁵

Access to mental health services is a critical issue throughout the lifespan for children with Spina Bifida and their parents and other family members. Such services could begin just after birth for parents as they adjust to having a child with Spina Bifida. During the school years counseling for learning and emotional issues can be accessed via the child's IEP or 504 Plan. Camp programs can also provide emotional support and a context where children and youth can learn independence and self-management skills. Individual psychotherapy by skilled pediatric psychologists and social workers may be needed during adolescence and adulthood for emotional, educational, and vocational issues related to the transition to adulthood. Regional Independent Living Centers can offer peer counseling and referral to mental health services for adults with Spina Bifida.

Outcomes

Primary

1. Achieve optimal mental health throughout the lifespan as evidenced by adaptive psychological, social, and participation outcomes.

Secondary

1. Maximize adaptation across all factors that are predictive of mental health outcomes (including neuropsychological, family, peer, academic, biological, and condition-related predictors). Access services and supports across appropriate domains to optimize mental health throughout the lifespan.

Tertiary

1. Maximize self-management, independence, quality of life, and transition-to-adulthood outcomes by addressing mental health challenges.

0-11 months

Clinical Questions

1. What parenting interventions can promote mental health for parents and children?

Guidelines

1. Assess family functioning, stressors and supports. Identify strengths and build on resources and supports that encourage resilience.^{10,24}
2. Provide parents with detailed information about Spina Bifida.¹ (Prenatal Counseling Guidelines)
3. Connect families with contact information of local Spina Bifida Association (SBA) Chapters, community resources, and the SBA's National Resource Center. (clinical consensus) (<http://spinabifidaassociation.org/chapters/>)

- (<http://spinabifidaassociation.org/national-resource-directory/>)
4. Address developmental strengths and concerns through information and support. (clinical consensus)
 5. Refer to early intervention services [INSERT LINK TO EARLY INTERVENTION] and the American Academy of Pediatrics. (clinical consensus) (INSERT LINK TO <https://www.aap.org/en-us/Pages/Default.aspx>)
 6. Assess quality of parent-child attachment.¹⁰
 7. Promote effective parenting skills in stimulation, caregiving, and enjoyment of the child to optimize typical child development.^{1,10} (Neuropsychology Guidelines)
 8. Screen for post-partum depression and post-traumatic stress disorder. (Prenatal Counseling Guidelines)

1-2 years 11 months

Clinical Questions

1. What is the psychosocial impact of having Spina Bifida on mental health and adaptation across the lifespan?
2. Which domains of mental health are most adversely affected in individuals with Spina Bifida and in what areas of mental health are individuals with Spina Bifida most resilient?
3. What are some common maladaptive behaviors that can negatively impact persons with Spina Bifida across the lifespan?
4. What resources or practices are most effective at mitigating mental health issues in this population?

Guidelines

1. Address developmental concerns and optimize typical child development by building on resilience, resources, and supports.¹ (Self-Management and Independence Guidelines)
2. Encourage families to offer developmentally-appropriate choices in daily life activities, including such things as picking up toys, cleaning up, and doing imitative housework.¹⁰
3. Encourage developmentally-appropriate play and social opportunities. (clinical consensus) (Family Functioning Guidelines, Physical Activity Guidelines)
4. Assess parenting skills and provide education on parenting strategies and behavior management techniques as needed.¹⁰
5. Provide additional age-appropriate information about Spina Bifida as the child grows.¹
6. Continue participation in early intervention services, as appropriate. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)
7. Consider referrals for parent-to-parent support opportunities. (clinical consensus)
8. Encourage families to participate in SBA and SBA Chapter-related activities and events (e.g., Spina Bifida Education Days, Walk-N-Roll for Spina Bifida, and other activities organized by local Chapters). (clinical consensus) (<http://spinabifidaassociation.org/chapters/>)

3-5 years 11 months

Clinical Questions

1. What is the psychosocial impact of having Spina Bifida on mental health and adaptation across the lifespan?
2. Which domains of mental health are most adversely affected in individuals with

- Spina Bifida and in what areas of mental health are individuals with Spina Bifida most resilient?
3. What are some common maladaptive behaviors that can negatively impact persons with Spina Bifida across the lifespan?
 4. What resources or practices are most effective at mitigating mental health issues in this population?

Guidelines

1. Discuss with parents the importance of their child's socialization and developing friendships with their peers and taking advantage of opportunities for activities. (36, 37) (Family Functioning Guidelines, Neuropsychology Guidelines)
2. Encourage participation in preschool educational programs. (clinical consensus)
3. Discuss the importance of making and keeping schedules/routines, doing chores, modeling behaviors, and making age-appropriate choices and decisions. (clinical consensus)
4. Assess social and psychological development and identify resources that build on strengths and encourage resilience.^{9,36}
5. Refer for social skills training as indicated.³⁶
6. Include optimization of mental health when developing an Individualized Education Program (IEP, IFSP or 504 Plan). (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)
7. Provide additional age-appropriate information about Spina Bifida as the child grows.¹
8. Refer parents to the local school district to begin the process of requesting special education or classroom support (IEP, IFSP, or 504 Plan) needed to optimize their child's participation in school. (clinical consensus)) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

6-12 years 1 month

Clinical Questions

1. What is the psychosocial impact of having Spina Bifida on mental health and adaptation across the lifespan?
2. Which domains of mental health are most adversely affected in individuals with Spina Bifida and in what areas of mental health are individuals with Spina Bifida most resilient?
3. What are some common maladaptive behaviors that can negatively impact persons with Spina Bifida across the lifespan?
4. What resources or practices are most effective at mitigating mental health issues in this population?

Guidelines

1. Encourage participation in community activities for recreation.^{19-20,28,36,37,38} (Physical Activity Guidelines)
2. Promote the development of friendships by helping families to identify social opportunities (e.g., participation in camps, adaptive sports programs/events, Walk-N-Roll for Spina Bifida, Boy and Girl Scouts, church youth groups, YMCA activities, and SBA and SBA Chapter social events).³⁶
3. Assess the child for depression, anxiety, bullying (including cyber bullying), and social participation. Similarly, identify the child's strengths and build on resources that encourage resilience. Initiate individual and family interventions when appropriate.^{9,23}

4. Encourage activities and hobbies that improve face-to-face social contact. (clinical consensus)⁴⁰
5. Promote transfer of age-appropriate medical responsibility from parent to child in those who have the requisite abilities and cognitive capacity.¹⁷ (Family Functioning Guidelines)
6. Discuss the importance of increasing household responsibilities that are appropriately modified to account for mobility and cognitive limitations. (clinical consensus)
7. Refer children with emotional and/or behavioral difficulties for psychological support and counseling. Identify community resources for social and psychological development (e.g., camps, recreation centers and more).^{9,12}
8. Assess the family's relationship with their child's school and encourage parents to be advocates for their children in the school setting. (clinical consensus) (Family Functioning Guidelines)
9. Promote the child's independence and choice in social activities. Promote self-care so that the child is able to be independent in social settings.³⁶ (Self-Management and Independence Guidelines)
10. Promote appropriate after-school sports and club activities.³⁸
11. Provide additional age-appropriate information/knowledge about Spina Bifida as the child grows. Begin to include child in clinical decision-making.³⁹ (Neuropsychology Guidelines)
9. Promote and encourage participation in community and SBA and SBA Chapter-related activities. (clinical consensus) (<http://spinabifidaassociation.org/chapters/>) (<http://spinabifidaassociation.org/national-resource-directory/>)

13-17 years 11 months

Clinical Questions

1. What is the psychosocial impact of having Spina Bifida on mental health and adaptation across the lifespan?
2. Which domains of mental health are most adversely affected in individuals with Spina Bifida and in what areas of mental health are individuals with Spina Bifida most resilient?
3. What are some common maladaptive behaviors that can negatively impact persons with Spina Bifida across the lifespan?
4. What resources or practices are most effective at mitigating mental health issues in this population?

Guidelines

1. Assess peer relationships and friendships.³⁶⁻³⁷
2. Assess for at-risk behaviors (alcohol, drug, or tobacco use and unsafe or unprotected sex), and identify areas of strength and build on resources that encourage resilience.²⁸
3. Screen for depression or anxiety and initiate individual and family interventions when appropriate.^{9,23}
4. Provide counseling and/or behavioral support as needed. (clinical consensus)
5. Promote transfer of medical responsibility from parent to child in those who have the requisite abilities and cognitive capacity.¹⁷ (Self-Management and Independence Guidelines)
6. Refer for social skills training as needed.³⁶
7. Encourage activities and hobbies that improve face-to-face social contact.⁴⁰ (clinical consensus)

8. Provide counseling regarding sexuality, sexual functioning, fertility, and contraception. Focus on sexual safety issues.¹³⁻¹⁵ (Sexual Health and Education Guidelines)
9. Discuss the safe use of and choices around drugs and alcohol and conduct risk assessment in this domain.²⁵
10. Discuss the importance of initiating and organizing opportunities for social activities.³⁶
11. Discuss the relationship between independence and interdependence and mental health. (clinical consensus) (Self-Management and Independence Guidelines)
12. Facilitate the child's involvement with a peer role model, such as a teen with Spina Bifida who is of a similar age). (clinical consensus)
13. Provide or refer to opportunities for formal or informal mentoring. (clinical consensus)
14. Encourage the teen to participate in the school's IEP transition team meeting. (clinical consensus)
15. Develop a plan for the teen's transition to independent living, post-secondary education, vocational training, and career interests. (clinical consensus) (Transition Guidelines)
16. Develop a plan for transition from pediatric to adult health care. (clinical consensus) (Transition Guidelines)

18 + years

Clinical Questions

1. What is the psychosocial impact of having Spina Bifida on mental health and adaptation across the lifespan?
2. Which domains of mental health are most adversely affected in individuals with Spina Bifida and in what areas of mental health are individuals with Spina Bifida most resilient?
3. What are some common maladaptive behaviors that can negatively impact persons with Spina Bifida across the lifespan?
4. What resources or practices are most effective at mitigating mental health issues in this population?

Guidelines

1. Screen for depression or anxiety and initiate interventions when appropriate.^{6-7,11}
2. Continue the transfer of medical responsibilities in young adults with Spina Bifida who have the requisite abilities and cognitive capacity.¹⁶
3. Encourage activities and hobbies that improve face-to-face social contact. (clinical consensus)
4. Encourage ongoing efforts to promote friendship and social intimacy.³⁷
5. Encourage and promote vocational or occupational goals and pursuits. (clinical consensus) (Transition Guidelines)
6. Maintain efforts for good general health promotion and exercise, as well as specialized Spina Bifida care. Optimize health to reduce the risk of obesity and maximize social opportunities and mental health.^{39,41} (Physical Activity Guidelines)
7. Recommend SBA resources (<http://spinabifidaassociation.org/learn-about-sb/adults/>). (clinical consensus)
8. Continue to refine the plan to ease transition from pediatric to adult health care. (Transition Guidelines)

Research Gaps

1. What services and supports can be utilized to mitigate barriers to optimal mental health throughout the lifespan?
2. What are the links between mental health and the following outcomes: self-management, independence, continence, quality of life, and the transition from pediatric to adult health care?
3. What interventions are available to enhance mental health across the lifespan in individuals with Spina Bifida?
4. What methods have been implemented by providers who care for children and adults with Spina Bifida and have an identified mental health diagnosis to guide their transition to adult health care?
5. What resilience factors mediate mental health outcomes in children and adults with Spina Bifida?

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Quality of Life

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Introduction

Quality of Life is defined as “an individual’s perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations and concerns.”¹⁻² Health Related Quality of Life (HRQOL) is considered a sub-domain of Quality of Life (QOL) and measures a subjective perception of the impact of a health condition and/or its treatment on the individual.³⁻⁶ HRQOL is most often multidimensional encompassing physical, emotional, social, and cognitive/occupational status. The focus of this guideline is to mitigate the factors that negatively impact QOL/HRQOL and enhancing the factors positively related to QOL/HRQOL.

The measurement of QOL and HRQOL in Spina Bifida is early in its development. The quality of most studies are limited by sample size, diversity and response rate of participants, measures that may not capture all domains of HRQOL (e.g., impact of scoliosis and scoliosis repair on HRQOL) and measures the lack sensitivity to capture changes in QOL or HRQOL or reflect cultural differences.^{3,7} Thus, the evidence that follows is preliminary and may be incomplete, but summarizes the current state of the literature at the time that these guidelines were created.

QOL/HRQOL should be measured by condition and age-related instruments, both the parent and child/adolescent perception should be measured and the child perception valued.^{3,7} Parent report is often but not always lower than child/adolescent report.⁸⁻⁹ Children as young as eight can report on their QOL/HRQOL.¹⁰ Use of HRQOL measures has been found useful in other chronic health conditions.^{7,11} New age- and Spina Bifida-specific HRQOL instruments have been recently created (QUALAS-C, QUALAS-T, QUALAS-A) (Appendix 1) but not been used extensively.⁸⁻⁹ If time is limited, the adolescent self-report should be used over parent report.

When deciding on an instrument to use to measure QOL/HRQOL, it should be understood that some QOL measures and most HRQOL measures equate the ability to function to QOL/HRQOL such that any individual with a disability will have, by nature of the questionnaire, lower HRQOL than peers without disabilities (Appendix 1). This conceptual equation devalues the lives of people with disabilities by automatically declaring that a person with a disability cannot have as good a quality of life as someone without disabilities. Measures that capture the individual’s perception of how their condition (i.e., Spina Bifida) impacts their life are preferred.¹² This focus on function is evident in the literature where QOL/HRQOL assessments of children with Spina Bifida are consistently lower in the physical domain as function (i.e. walking upstairs, running a distance) not perception, are measured.^{4,7}

Tools such as World Health Organization Quality of Life (WHOQOL) Brief uses items addressing perceived energy to do physical activities important to the individual and thus avoid this problem.¹³ Similarly, new Spina Bifida and age specific measures address perception (e.g. bother, worry), not function.⁸⁻⁹ Findings regarding the impact of Spina Bifida on other domains of QOL/HRQOL for children, adolescents and adults (social, emotional, cognitive/school/work) are inconsistent, although one review of qualitative studies indicated more issues in the psychosocial domain of QOL than physical domain.^{3,6-7} Evidence regarding most Spina Bifida factors (e.g. level of lesion, severity of Spina Bifida, ambulation) have generally had no or small

associations with youth report QOL/HRQOL and only a modest relationship to parent report of generic QOL.^{3,7,14-16} Pain has consistently been related in all ages, by both parent and self-report and across varied instruments.¹⁸

Other factors related to QOL/HRQOL found in recent literature include:

- Urinary tract infections and pressure injuries in children.
- Pressure injuries and latex allergy in adults.
- Level of lesion and hydrocephalus.
- Although Spina Bifida variables have been inconsistently related to QOL/HRQOL in children, some evidence indicates that level of lesion, full time wheelchair use, and hydrocephalus was associated with reduced HRQOL in adults.¹⁹⁻²⁴

Evidence consistently supports that bowel incontinence is associated with lower HRQOL and satisfaction with a bowel program is associated with higher HRQOL.^{15,24-25} Data on the relationship of bladder incontinence to QOL in children is inconsistent, but studies of adolescents and adults report that support for urinary continence contributes to overall HRQOL.²⁶⁻²⁹

Using a new instrument (QUALAS-A) that specifically measures the impact of continence on adult HRQOL,⁸ any bowel continence and the amount, but not frequency of urinary incontinence, were related to the “Bladder and Bowel HRQOL subscale” but not to the “Health/Relationship or Esteem/Sexuality HRQOL subscales.”³⁰ There is little literature on sexuality and QOL and using generic measures there was no relationship.³¹ In studies to date, scoliosis status³²⁻³³ has not been related to HRQOL. Only one study found obesity related to HRQOL in Spina Bifida.³⁴⁻³⁵ In contrast, obesity was related to HRQOL in typically developing children and those with other chronic health conditions.^{23,34-36}

Variables such as resilience (e.g., attitude towards Spina Bifida, hope and future expectations, coping skills) have been strongly related to higher HRQOL and QOL.^{15-16,37} In contrast, depression, a lack of optimism and reduced executive functioning were related to lower QOL/HRQOL.¹³ Similarly, family variables such as higher family satisfaction and family resources have been related to higher QOL for adolescents and those over 18 years of age.^{13,15-16} In order to foster QOL/HRQOL clinicians should develop strategies to optimize psychosocial wellbeing, bowel and bladder continence, and minimize the impact of pain, if present.

QOL or HRQOL should not be measured in isolation. There may be components of HRQOL that are not measured by current instruments. If clinicians are going to address QOL they also need to address the factors important to the individual with Spina Bifida and their family. An emerging concept, Family QOL (FQOL) may have usefulness in the care of individuals and families with Spina Bifida.³⁸⁻³⁹ FQOL has been measured with domain-specific instruments⁴⁰ and a generic FQOL tool (Appendix 1). There is not enough experience with the concept or the tools to include FQOL in the guidelines but future investigation is warranted.

Outcomes

Primary

1. Improve QOL across the lifespan in individuals with Spina Bifida.

Secondary

1. The information provided in this guideline gives the health care providers a better understanding of QOL and HRQOL measurement, potential issues related to available tools or tool development, and other factors related to QOL or HRQOL.
2. Increase QOL assessments in clinical practice.

Tertiary

1. Clinicians of every specialty integrate assessment of QOL and intervention to address QOL into clinical practice.

0-11 months

Clinical Questions

1. What factors are related to QOL?

Guidelines

1. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children. (clinical consensus) (Family Functioning Guidelines)
2. Address constipation because long-term constipation impedes the development of an effective bowel program. (clinical consensus) (Bowel Function and Care Guidelines)

1-2 years 11 months

Clinical Questions

1. What factors are related to QOL?

Guidelines

1. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children. (clinical consensus) (Family Functioning Guidelines)
2. Address constipation because long-term constipation impedes the development of an effective bowel program. (clinical consensus) (Bowel Function and Care Guidelines)

3-5 years 11 months

Clinical Questions

1. What factors are related to QOL?
2. What measures of QOL/HRQOL are the most efficient and useful?

Guidelines

1. Assist families in their efforts to facilitate the development of protective psychosocial behaviors (e.g. showing affection, bouncing back when things don't go the child's way, showing interest in learning new things). Encourage independence, praise for accomplishment, and provide opportunities for fun. (clinical consensus) (Family Functioning Guidelines, Mental Health Guidelines)
2. Address assessment of executive function. (clinical consensus) (Neuropsychology Guidelines)
3. Target strategies to optimize the child's bowel program because bowel incontinence is consistently related to HRQOL. (clinical consensus) (Bowel Function and Care Guidelines)

6-12 years 11 months

Clinical Questions

1. What factors are related to QOL?
2. What might QOL assessment and improvement activities look like in clinical practice?
3. What measures of QOL and HRQOL are the most efficient and useful?

Guidelines

Psychosocial well-being

1. Assist families in their efforts to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships.^{3,7,15-16,23} (Mental Health Guidelines)
2. Consider strategies to optimize peer relationships.⁴⁵ (Mental Health Guidelines)
3. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children.^{13,15-16} (Family Functioning Guidelines)
4. Refer to community resources that enhance protective factors, such as sports, camps, scouts, and other community programs. (Self-Management and Independence Guidelines)
5. Address assessment of executive function.⁴¹ (Neuropsychology Guidelines)

Continence

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL.^{24-25,28} (Bowel Function and Care Guidelines)
2. Assess both volume and frequency of urinary incontinence, as volume may be more distressing than frequency.³⁰ (Urology Guidelines)

Pain

1. Evaluate presence and characteristics of any pain experienced.^{7,13,42-43}
2. Develop strategies to address pain and its impact on school, work, recreation, and social activities. (clinical consensus)

Measurement of QOL

1. Use a systematic approach to evaluating QOL/HRQOL.^{4-5,7,44}
2. Consider using both self and parent-report instruments.^{3,7}
3. If feasible, use Spina Bifida and age-specific HRQOLs instruments that measure perception ("concerned about," "worried about," "avoid") and avoids the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) when assessing children with Spina Bifida. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful.^{3-4,7,9-10,12} (Appendix 1)
4. Consider using a single-item QOL question^{7,15-16} such as "How would you rate your quality of life?" on a scale of 0-100 with 0=poor and 100=excellent? (Appendix 1) Individual and family factors associated with HRQOL in adolescents and young adults with Spina Bifida should be explored with follow up assessment if needed.

13-17 years 11 months

Clinical Questions

1. What factors are related to QOL?
2. What might QOL assessment and improvement activities look like in clinical practice?
3. What measures of QOL and HRQOL are the most efficient and useful?

Guidelines

Psychosocial well-being

1. Assist families in their efforts to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships.^{3,7,15-16,23} (Mental Health Guidelines, especially the section on peer relationships)
2. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children.^{13,15-16} (Family Functioning Guidelines)
3. Consider strategies to optimize peer relationships.⁴⁵ (Mental Health Guidelines)
4. Consider each individual's unique priorities important in QOL. (clinical consensus)
5. Refer to community resources such as sports, camps, scouts, and other community programs that enhance protective factors. (clinical consensus) (Self-Management and Independence Guidelines)
6. Address strategies to compensate for executive functioning challenges.⁴¹ (Neuropsychology Guidelines)

Continence/mobility

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL.^{24-25,28}
2. Investigate the child's satisfaction with her or his bowel program. Address concerns that will help to optimize program.¹⁵ (Bowel Function and Care Guidelines)
3. Assess both volume and frequency of urinary incontinence as volume may be more distressing than frequency.³⁰ (Urology Guidelines).
4. Consider functional mobility options that optimize societal participation. (clinical consensus) (Mobility Guidelines)

Pain

1. Evaluate presence and characteristics of any pain experienced.^{7,13,42-43}
2. Develop strategies to address pain and its impact on school, work, recreation, and social activities. (clinical consensus)

Measurement

1. Use a systematic approach to evaluating QOL/HRQOL.^{3-5,7,44}
2. Consider using both self and parent-report instruments.^{3,7}
3. Use the new Spina Bifida HRQOL instrument that measures perception ("concerned about," "worried about") and avoids the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) when assessing children with Spina Bifida. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful.^{4,7,12} (Appendix 1)
4. Use an age- and condition-specific instrument to assess QOL/HRQOL.^{3,7-8,10} (Appendix 1)
5. Evaluate both the child's self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report.^{3,7,10}
6. Consider using a single-item QOL question(s) with follow up assessment if needed.^{7,15,16} (Appendix 1) For example:
 - "How would you rate your quality of life?"
 - "What makes up QOL for you?"
 - "What do you think would make your QOL better?"

18+ years

Clinical Questions

1. What factors are related to QOL?
2. What might QOL assessment and improvement activities look like in clinical practice?
3. What measures of QOL and HRQOL are the most efficient and useful?

Guidelines

Psychosocial well-being

1. Identify strategies or resources to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships.^{3,7,15-16,23} (clinical consensus) (Mental Health Guidelines, especially the section on peer relationships)
2. Explore satisfaction with relationships and their sexuality. (clinical consensus) (Sexual Health and Education Guidelines)
3. Consider strategies to optimize peer relationships. (clinical consensus) (Mental Health Guidelines)
4. Consider the importance of each individual's QOL unique priorities. (clinical consensus)
5. Refer to community resources such as sports, camps, community advocacy groups, universities with strong programs to support students with disabilities, and other community programs that enhance protective factors. (clinical consensus) (Self-Management and Independence Guidelines)
6. Address strategies to compensate for executive functioning challenges.⁴¹ (clinical consensus) (Neuropsychology Guidelines)
7. Consider strategies to enhance self-management behaviors.⁴⁷ (Self-Management and Independence Guidelines)

Continenence/mobility

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL in adults, especially in social domains.^{24-25,28}
2. Investigate the adult's satisfaction with her/his bowel program.¹⁵ Address concerns to optimize program.
3. Assess both volume and frequency of urinary incontinence in adults, as volume may be more distressing than frequency.³⁰
4. Consider functional mobility options that optimize societal participation.²⁰ (Mobility Guidelines)

Pain

1. Evaluate the presence and characteristics of any pain experienced.^{7,13,42-43}
2. Develop strategies to address pain and its impact on school, work, recreation, and social activities. (clinical consensus)

Measurement

1. Use a systematic approach to evaluating QOL/HRQOL.^{4-5,7,44}
2. Consider using both self and parent-report instruments.^{3,7}
3. Use an age-and condition-specific instrument to assess HRQOL. Instruments that measures perception ("concerned about," "worried about," "avoid") and avoid the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) are preferred. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful.^{4,7,12} (Appendix 1). Instruments like the WHOQOL-BREF (Appendix 1)^{1,2,19} avoid this issue using questions such as "Do you

- have enough energy for everyday activities?” or “To what extent do you feel that physical pain prevents you from doing what you need to do?” Spina Bifida-and-adult-specific measures also assess perception and avoid this issue.⁸
4. Evaluate both the adult’s self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report of QOL/HRQOL.^{3,7,10}
 5. Consider using a single-item QOL^{7,15-16} question(s) with follow up assessment if needed. (Appendix 1). For example:
 - “How would you rate your quality of life?”
 - “What makes up QOL for you?”
 - “What do you think would make your QOL better?”

Research Gaps

1. Need continued refinement of HRQOL and QOL measurement including the relationship of individual and parent proxy reports.
2. Continued research is needed to identify the factors related to QOL/HRQOL and how change in these factors across time impacts QOL/HRQOL. Especially needed is to extend the exploration of current factors to include whether finances, ethnic identity, religion and spirituality or aging with play a role in QOL/HRQOL.
3. Research is needed to determine if measuring QOL/HRQOL in clinical practice actually leads to activities that improve QOL/HQOL.
4. Research is needed to identify QOL/HRQOL during transition to adulthood and adult health care.⁴⁸
5. Implementation research is needed to evaluate if emerging evidence on QOL/HRQOL is integrated into practice. If the emerging evidence is not being integrated into practice, there is a need to identify and address the barriers to implementing the findings.
6. Need further research on the emerging concept of QOL in families and its association with child outcomes.

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Appendix 1: Summary and Assessment of QOL/HRQOL/FQOL Instruments

Summary and Assessment of QOL Instruments used in children, adolescents, and adults with Chronic Health Conditions (CHC) and their potential use in the population with Spina Bifida. The instrument uses criteria developed by Waters et al.⁴ and has been expanded to include additional instruments.

QOL/HRQOL Assessment Criteria Coding Table

1.	HF/QOL:	Original purpose of instrument	Health/functioning=1; midrange=2; QOL=3
2.	Fam:	Origin of items	Low involvement of family=1 midrange=2; High involvement of family=3
3.	Focus:	Actual focus of the instrument	Functioning=1; midrange=2; well-being=3
4.	Opp:	Opportunity to self-report	No opportunity to self-report=1 midrange=2; self-report version available=3
5.	Self-est:	Potential threat to self-esteem	Negative wording =1; midrange=2; positive wording=3
6.	# Items:	Length	Large number of items=1; midrange=2; small number of items=3
7.	Reliability & Validity	Psychometric Properties	Poor or not demonstrated=1; midrange=2; excellent and demonstrated adequately=3

Summary and Assessment of QOL/HRQOL/FQOL Instruments

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/Q OL	Fam	Focus	Opp	Self-est	# Items	R & V	
Generic instruments										
PedsQL™ (Varni) (child or parent SR) ⁴⁹ 20 items	Versions (age): -Child (5-12) -Adolescent (13-18) -Young adult (18 + years) Parent and child report.	Physical, Emotional, Social, Cognitive (school /work).	1	2	1	3	1	3	3	Do not use physical scale. Emotional, social and cognitive scales may be useful especially if comparing to typically developing youth. However, heavy focus on functioning. Strong psychometrics across many CHC and typically-developing peers

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
CHQ Child (N=187) or parent (N=50) SR ⁵⁰	Parent and child version developed by experts using literature and other instruments.	Behavior, bodily pain, general health, mental health, parent impact, emotional, physical functioning, parent impact time, emotional/behavioral role, physical and self-esteem. Physical and psycho-social summary scores.	1	1	2	1	2	3	2	Long; may be useful if specific subscales are of interest. No data on sensitivity to change. Multiple items with floor and ceiling effects. Not used extensively in Spina Bifida. Cerebral palsy comparison indicated it was outperformed by other measures. ⁴
KID SCREEN ⁵¹	Ages 8-18. 27, 10 and 57-item versions available.	5 domains: -Physical well-being - Psychological well-being -Support -Peers -Financial resources	3	3	3	3	3	3	3	Only child generic instrument rated as 3 in all categories by Waters et al., 2009 ⁴ No known use to date in US in children with Spina Bifida. ⁷ Focus groups, cognitive interviews and pilot testing 52 and 27-item versions. Validated in 12 European countries using over 22,000 children. Supported with internal and test-retest reliability.
WHO QOL-BRIEF The WHO QOL Group, 1998 ¹	26 items in four domains. Shorter version of the 100-item, 1997 original instrument.	4 domains: -Physical health -Psycho-social health -Social Environment 2 single items: -overall perception of QOL	3	3	2	3	3	2	3	Preferred generic scale for adults with SB. Physical scale: while assessing the impact of physical status on QOL does so with items that do not automatically disadvantage individuals with a specific mobility-related impairment. Reference period: Last 2 weeks. Positively-worded and flexible for all conditions. (e.g. "Do you have

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
		-overall perception of health								enough energy for everyday life"?) Available at: http://www.who.int/mental_health/media/en/76.pdf
Spina Bifida-Specific Instruments										
HRQOL-SB Parent and teen report ⁵²	2 versions: -child (parent report, 44 items) -adolescent (adolescent report, 47 items)		3	3	2	3	3	2	1	Positively-phrased items; many items with ceiling effect; strong internal reliability. No factor structure, test-retest reliability, or sensitivity to change analyses. May be more appropriate for general assessment of younger child by parent. Only total score supported; no domain assessment possible. Use with caution. May be useful to assess HRQOL if previously used in a longitudinal study.
HOQ ⁵³	For children with hydrocephalus. Measures functional status. No factor analysis.	Originated from focus groups	1	2	1	1	3	2	1	Not a priority instrument for use in Spina Bifida. Limited psychometric properties.
Quality of Life Assessments (QUALAS) with child, teen, and adult versions SR QUALAS Child (QUALAS-C) ⁹	A family of three instruments created to evaluate living with Spina Bifida: child, teen, adult. Child (ages 8-12), 10 items.	Reference: last 4 weeks. Responses are "never" to "always." Five options plus alternative. Two scales: -Esteem/Independence -Bladder and Bowel	3	3	3	3	2	3	2	Recommended for use with individuals who have SB. All three age versions: Based on qualitative and cognitive interviews. Strong input from families/those with Spina Bifida. Assessed using appropriately large enough samples to assess construct validity. Good factor structure, internal and test-retest reliability.

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
QUALAS-Teen (QUALAS-T) ⁵⁴	Teen version (ages 13-17) 10 items	Two scales: -Family/Independence -Bladder and Bowel	3	3	3	3	2	3	2	<p>Typical question: "Did it annoy you if you could not do what other teenagers could do?" Besides "never" to "always" there is an alternative answer, "I could do what other teens do."</p> <p>Some negative wording ("upset," "embarrassed," "bother you") but generated from qualitative interviews and affirmed by cognitive interviews.</p> <p>No physical scale in child, teen or adult (could be an asset or liability).</p> <p>Child: Useful to assess how self-esteem and bowel and bladder status is perceived.</p> <p>Teen: Useful to assess the two domains. May not be a useful measure of overall HRQOL.</p>
QUALAS-Adult (QUALAS-A) ⁸	Adult version (ages 18 and above) 15 items - all in health and relationships. Positively-worded.	3 scales -Health/Relationships -Esteem / Sexuality -Bladder and Bowel	3	3	3	3	2	3	2	<p>Useful measure of domains assessed.</p> <p>May not be a useful measure of overall HRQOL.</p> <p>Important inclusion on items on sexuality (only instrument that does). Sexuality items might be also appropriate for older teens.</p> <p>Internal consistency and test-retest reliability were high for all domains (Cronbach's alpha ≥ 0.70, ICC ≥ 0.77). Correlations between QUALAS-A and WHOQOL-BREF were low except for high correlations with Health and Relationships domain ($0.63 \leq r \leq 0.71$), which supports the ability of the QUALAS-A.</p> <p>Bowel and Bladder scale same for teens and adults so can use same scale for those 13 or older</p>

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
Spina Bifida PQ QOL questionnaire for children with Spina Bifida ⁵⁵	<p>Dutch scale developed by using existing items from other instruments (PedsQL and Fecal Incontinence QOL survey n=10) and qualitative interviews.</p> <p>Yielded additional 25 items for total of 35 items for children 6-18 years mental age.</p> <p>Questions address last three months, 11 minutes to complete.</p>	<p>-Physical, social, and emotional function -School -Home -Hospital</p> <p>Child and parent versions available with picture book for children.</p> <p>Includes questions on pain and energy (e.g. "Have you been too tired to do your regular activity?")</p>	3	3	2	3	2	2	1	<p>Only 62 patients used for initial assessment.</p> <p>Internal reliability good for most scales and ICC for stability.</p> <p>No factor analysis to confirm domains.</p> <p>Authors identify three items that "stand out" as negatively impacting QOL: -Feeling angry in the emotional domain (unclear whether this is related to SB) -the use of colon enemas in physical domain -missing activities as a result of doctors' visit/surgeries etc.</p> <p>Available in English but no data on English samples. Needs more psychometric evaluation before broad use.</p>
Development of a tool to describe overall health, social independence and activity limitation in AYA with disability ⁵⁶	<p>QOL tool for adolescents with a disability.</p> <p>Used with 174 adolescents with Spina Bifida, (38%); Muscular Dystrophy, and Fragile X syndrome to develop tool.</p>	<p>-Emotional health -Physical health - Independence -Activity limitation - Community participation</p>	3	1	3	3	1-2	1	1	<p>Use with caution.</p> <p>Should avoid physical scale that addresses specific tasks (vigorous activities, running, heavy lifting).</p> <p>Community participation scale may be useful.</p> <p>Activities scale would be more useful if stated in a positive manner (what the individual can do rather than focus on limitations).</p> <p>Instrument developed from other instruments. Preliminary psychometrics. No involvement of individuals or family members.</p> <p>Many items in article appendix are useful and worth reviewing for those addressing transition.</p>
Single-item QOL ^{1, 7, 15, 16}	<p>A part of many instruments. Overall how</p>	<p>Allows individual to determine</p>	3	1	3	3	3	3	2	<p>Good for an overall perception; the person</p>

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
	would you rate your QOL?	domains important to them and prioritize domains based on personal perception.								<p>can give priority to their domains of importance.</p> <p>Does not help the health care provider identify what determines QOL for the individual.</p> <p>Would need a follow-up question to identify domains important to the individual.</p> <p>Some evidence of validity (related to variables as expected).</p> <p>Factor analysis and internal reliability – not applicable.</p>
Fecal Incontinence (FIC) QOL survey not included as it addresses only one aspect of HRQOL. Refer to Sawin & Bellin, 2010 ⁷ for additional information.										
Family QOL (for full discussion of FQOL scales see HU et al, 2011)⁴⁴										
PedsQL™ Family Impact Model (parents SR) ⁴⁹	<p>Impact of pediatric CHC on parent's functioning.</p> <p>Family functioning subscale.</p> <p>One factor of general negative impact of pediatric CHC on social and familial systems.</p>	<p>Problems with physical, emotional, social, and cognitive functioning; communication; worry.</p> <p>Problems with family activities and relationships.</p>	1	1	1	3	2	2	2	<p>Some have used for FQOL. Authors indicate that it can be used for QOL assessment.</p> <p>Assesses impact on the family but may not be assessment of QOL.</p> <p>Heavy emphasis on function.</p>
FQOL generic tool ³⁹	<p>Created for use with family with AYA with Spina Bifida.</p> <p>Parent and teen self-report</p> <p>3 items: -How would you rate your QOL? -How would you rate your child/parent QOL? -How would you rate FQOL"? Rated from 0-100; summed</p>	<p>Items allow responder to include domains important to them and to rank domains according to their own priorities.</p> <p>Rated from 0-100; summed and added.</p>	3	2	3	3	3	3	1	<p>The instrument has been evaluated in a sample of AYA with Spina Bifida (n=120), a comparison sample (n=98) and parents of the AYA sample (n=instrument was found to have strong preliminary psychometrics including support for a single factor and high internal reliability.</p>

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
	and mean used.									
Beach Family QOL Scale (FQOL) <small>38, 40</small>	Measures several aspects of perceived satisfaction. 5 domains: -Family Interaction -Parenting -Emotional well-being -Physical / material well-being -Disability-related support	25-item questionnaire; 5-point Likert-type response pattern. "Very dissatisfied" to "Very satisfied." Available from Beach: https://www.midss.org/sites/default/files/fqol_survey.pdf	1	2	2	3	3	2	2	Widely used in the field of intellectual disabilities and families with children who have special needs. Developed at the Beach Center. Heavy emphasis on function. Detailed and long; may limit use in clinical practice.

AYA = Adolescent/Young Adults; CHQ= Child Health Questionnaire; FQOL=Family Quality of Life. KIDSCREEN= (10, 27, 52 version); HoQ=Hydrocephalus Quality of Life; PedsQL = Pediatric Quality of Life—Varni family of tools; SR = Self-Report; QUALAS= Quality of life Assessments in SB for Child, Teen, Adult.
For comprehensive assessment of generic QOL/HRQOL measures used in Spina Bifida see Bakaniene, et al., 2016; Sawin & Bellin, 2010, and Waters et al., 2009.^{3,4,7}

Self-Management and Independence

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Introduction

Self-management and independence are closely related constructs and are presented together. Self-management for youth and emerging adults with Spina Bifida is an active daily and flexible process in which youth and their parents share responsibility and decision-making for managing their condition, health, and well-being through a wide range of knowledge, attitudes, activities, and skills. The goal of this increasing responsibility is to develop the self-management behaviors needed to achieve independence and transition to adulthood and independent living.¹⁻² Self-management for all is the interaction of health behaviors and related processes that patients and families engage in to care for a chronic condition.³

Child autonomy provides a critical foundation for developing self-management and independence. For all children, autonomy begins early and is fostered by opportunities to make choices and to develop a sense of mastery. Most children with Spina Bifida achieve basic self-management and independence behaviors, (e.g., dressing appropriately, planning activities with peers, or cooking pre-planned meals) yet often lag 2-5 years behind their typically-developing peers in these behaviors.⁴ This gap may be due to the child's difficulties performing common everyday motor and processing activities in efficient and independent ways.⁵ Adaptation of performance and initiation of new steps may be especially challenging.⁵

Social skills in children are also important building blocks for independence. Many children with Spina Bifida need assistance with building adaptive social behaviors in peer interactions, specifically basic social skills such as reading social cues, clarity of thought and collaboration.⁶ Monitoring self-management learning is needed for all with cognitive functioning challenges, especially those with executive functioning, inattention and working memory issues.⁶⁻¹⁰ Educational programs in the home, school and broader community that offer opportunities to practice new behaviors are critical.

Youth do not enter adolescence with comprehensive knowledge of self-management (i.e. watching for signs of skin breakdown, bowel problems, shunt failure, and urinary tract infections), yet most develop this knowledge before age eighteen.^{1,4,11} Advanced self-management behaviors achieved by peers but not by individuals with Spina Bifida by age 18 are broad in scope and include doing their own laundry, cooking independently, managing their bank account without assistance, managing their allowance, and making their own appointments.⁴ It is not clear if these delays in skill development are developmentally appropriate for youths with Spina Bifida or due to a lack of expectations and support in the home, school, health system, or broader community. However, healthy family functioning was consistently related to better self-management outcomes across all developmental stages.^{9,11} Since there is evidence that responsibility in the home (e.g., chores and general decision-making) promotes self-management skill-building, individuals and families should be encouraged to expand their range of everyday living skills and responsibilities.^{4-5,11-14} Potential self-management skill-building challenges identified from longitudinal research include older school-age children with Spina Bifida perceiving themselves as being more independent

relative to parent assessment.¹⁵ Health care providers expect school-age and older children to perform self-management behaviors related to bladder programs, bowel programs, skin checks, and prevention of other secondary conditions.¹⁶ Yet, older children have reported that self-catheterization and bowel programs were a challenge, which often needed parent involvement¹. In addition, both the family and child have had difficulty carrying out diet recommendations, bowel programs, and skin care.¹⁷ Thus, tailored interventions are needed to support growth in these areas.¹⁷

Children with Spina Bifida transitioning to adulthood are generally poorly prepared to self-manage their condition or live independently and enter young adulthood with preventable secondary conditions.¹⁸ Unhealthy behaviors continue into adulthood.¹⁹ Skin breakdown, along with fewer self-management behaviors, predicted hospitalization in this population.^{13,20} However, there is also evidence that improved self-management in young adults impacts health outcomes.

Adults were often without access to a usual source of health care or had gone without care due to barriers.²¹ Most adults over 18 years of age have not achieved optimal independence milestones in education, employment, and independent living.²²⁻²³ However, they were reported to have higher independence than those with other severe conditions.²⁴

Self-management interventions for youth with Spina Bifida and other Chronic Health Conditions (CHC) generally show at least one significant improvement, although a short workshop-based intervention for older children with Spina Bifida yielded no significant differences in groups.²⁵⁻²⁶ Family-oriented self-management interventions may be most effective in younger adolescents.²⁷ Camp-based psychosocial interventions promoting skills-development in goal-setting and problem-solving have shown promise in developmentally-diverse samples of children and adults with Spina Bifida.²⁸⁻²⁹ Other interventions using a problem-solving model combining education and home/community practice similarly improved child self-management skills and decreased parent burden. In addition, there is evidence that improved self-management in adults impacts health outcomes.³⁰ Rehabilitation interventions in young and middle-aged adults have improved all aspects of self-management and independence, with moderate- to large-effect sizes including self-efficacy, management of bowel and bladder incontinence, cognitive function, and psychosocial symptoms.^{13,31} Although tested mostly in adults, technology-based interventions hold promise for expanding self-management behaviors in youth as well.^{13-14,32-36}

Clinicians should consider using one of the valid and reliable generic or Spina Bifida-specific measures of self-management and independence.^{8,37-39} (Appendix A). Clinical assessment of the level of self-management and independence in those with Spina Bifida should specifically distinguish between the skills and behaviors the individual knows how to do and the behaviors they actually execute independently.⁴⁰ The evidence supports the need to have a structured, planned, and incremental approach to building self-management and independence skills beginning in early childhood, conveying expectations for developmentally-appropriate household responsibilities and increasingly involving the child in their care. Plans that accommodate cognitive learning styles or executive functioning status and purposefully, incrementally increase skills with multiple opportunities to practice new behaviors are central to achieve successful self-management and independence.

Outcomes

Primary

1. Perform effective self-management behaviors at the highest level of their abilities.

2. Achieve optimal independent living and employment, as well as maximal participation in society.
3. Young children develop autonomy, responsibility, and other foundational skills for self-management and independent living.

Secondary

1. Interventions that address the foundational skills necessary for complex self-management and independence behaviors are introduced throughout the lifespan, as appropriate.
2. Target foundational skills should include executive functioning skills, self-efficacy, self-regulation, and engaging in social activities.
3. Self-management and independence goals are evaluated yearly with the family, child, adolescent, and adult.

Tertiary

1. Adults with Spina Bifida over 18 who have a guardian responsible for their health care should perform self-management behaviors in the areas of medication management, prevention of complications, implementation of bladder and bowel programs, skin surveillance, and be able to communicate their findings to their guardian and/or health care providers at their highest level of ability.
2. Adults with Spina Bifida over 18 who do not need a guardian are fully responsible to self-manage their condition and independence (e.g., making appointments, ordering medications, arranging for transportation, conducting basic living skills like cooking and doing the laundry, managing money, managing insurance, and communicating with their health care provider).
3. Individuals with Spina Bifida interact effectively with family, health care providers, and others in the external environment in an independent manner.

0-11 months

Clinical Questions

1. What approaches optimize individual and family self-management and eventual independence?

Guidelines

1. Provide instruction and support to families regarding knowledge and skills needed to manage their child's Spina Bifida and related issues. (clinical consensus)
2. Provide orientation to families that include the expectation for eventual self-management and independence according to the individual's age and the status of their Spina Bifida. (Prenatal Counseling Guidelines)
3. Encourage families to expect participation in activities of daily life including tasks such as picking up toys, cleaning up, and imitative housework. (clinical consensus)
4. Evaluate and support family function. (Family Functioning Guidelines)
5. Identify and make referrals to early intervention programs. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

1-2 years 11 months

Clinical Questions

1. What are the approaches that optimize individual and family self-management and eventual independence?

Guidelines

1. Provide instruction and support to families regarding knowledge and skills needed to

- manage their child's Spina Bifida and related issues. (clinical consensus)
- 2. Provide anticipatory guidance regarding developmental needs of children (such as exploration of environment, routines, and age-appropriate choices). (clinical consensus)
- 3. Teach families to offer daily age-appropriate choices such as choosing between two articles of clothes, two cereals for breakfast, and two books to read. (clinical consensus)
- 4. Encourage families to expect participation in daily life activities, including tasks such as picking up toys, cleaning up, and imitating housework. (clinical consensus)
- 5. Identify and make referrals to early education programs. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

3-5 years 11 months

Clinical Questions

- 1. What approaches optimize independence and individual and family self-management in children with Spina Bifida?

Guidelines

- 1. Provide instruction and support to families regarding knowledge, skills, and behaviors needed to manage their child's Spina Bifida and related issues. (clinical consensus)
- 2. Discuss the need to expand the range of daily life activities and chores, as well as strategies to accommodate the child's learning style and/or mobility.^{41,42}
- 3. Provide anticipatory guidance that autonomy skills are maximized when positive behaviors are reinforced and clear and consistent consequences for inappropriate behavior are used. (clinical consensus) (Mental Health Guidelines, Neuropsychology Guidelines)
- 4. Refer to community resources such as early education programs that promote autonomy, self-efficacy, and other foundational independence skills. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

6-12 years 11 months

Clinical Questions

- 1. What skills, abilities, and self-management behaviors should be targeted during age 6-12 years?
- 2. What are the most effective approaches to teach these skills and behaviors to children with Spina Bifida and their families?
- 3. Does specific skill training improve self-management behaviors (e.g., taking medication) and other independence behaviors?
- 4. What are optimal age expectations for specific self-management skills and behaviors (e.g. ability to self-catheterize; conduct skin checks; ability to describe their medication, its uses and side effects, and to take it on schedule; and describe their condition to a new professional) in children with Spina Bifida?
- 5. What instruments are available to measure self-management skills, abilities, and behaviors in children?

Guidelines

- 1. Provide instruction and support to children and families regarding the knowledge and skills needed to manage Spina Bifida and related independence issues. Teach the child basic self-management skills, including skills to prevent secondary conditions

- (clean intermittent catheterization, skin care, equipment care, bowel and bladder care, wheel chair maintenance and propulsion) based on individual abilities. Focus on self-efficacy.⁴³ Children with Spina Bifida may develop foundational skills and self-management behaviors at a slightly later age (2-5 year delay) and may need more deliberate practice. However, most self-management behaviors are achievable by adults with Spina Bifida.^{4-5,12,17,43,45-46} (Neuropsychology Guidelines)
2. Assist families in learning how to incrementally involve the child in organizing school work and self-management activities and how to begin to transition from parents doing to child doing with parental oversight to eventually child doing without parent oversight.^{5,7,12,44}
 3. Discuss the need to expand the range of daily life activities and chores as well as strategies to accommodate the child's learning style and/or mobility.^{4,7-8,11-12,14}
 4. Serve as a resource to school systems regarding transportation, learning skills, health issues, and development of self-management skills. (clinical consensus)
 5. Emphasize positive attitudes, self-esteem, assertiveness, self-efficacy and self-empowerment. (clinical consensus)
 6. Assess peer relationships and encourage peer social involvement.^{1,6} (Mental Health Guidelines, Neuropsychology Guidelines)
 7. Assess for potential patient, family, or environmental barriers to developing autonomy and independence, including family stress and conflict, and address in action plan.^{15,47} (Family Functioning Guidelines)
 8. Assess bladder and bowel management programs for eventual independent self-management (Bowel Function and Care Guidelines, Urology Guidelines).
 9. Consider using an age-and condition-appropriate assessment instrument (Appendix A) especially if the child has executive-functioning impairments.^{4,7-9,41-42}
 10. Discuss with parents the need to help their child develop basic money management skills.⁴ If the child has an Individualized Educational Plan (IEP), encourage parents and the school to include money management skills in the child's IEP. (clinical consensus)
 11. Encourage families to facilitate their child's language performance by creating intellectually- and culturally-enhancing activities in the child's typical environment.⁴⁸
 12. Set beginning expectations for independent living. (clinical consensus)
 13. Encourage use of technology to enhance self-management. (clinical consensus)

13-17 years 11 months

Clinical Questions

1. What approaches optimize individual and family self-management?
2. What skills, abilities and self-management behaviors should be targeted at age 13-17 years?
3. What are the most effective approaches to teaching these skills and behaviors to children age 13-17 years with Spina Bifida and their families?
4. Does specific skill training improve self-management behaviors (e.g., taking medication) and other independence behaviors?
5. What are optimal age expectations for specific self-management skills and behaviors (e.g. ability to self-catheterize; conduct skin checks; ability to describe their medication, its uses and side effects, take it on schedule, and describe their condition to a new professional) in children with Spina Bifida?
6. What instruments are available to measure self-management skills, abilities, and behaviors?

Guidelines

1. Evaluate self-management in appropriate areas (e.g. managing medications, prevention of complications, skin care, equipment care, bowel and bladder care, and making health care appointments). Assess self-efficacy⁴³ for these activities, considering that the child's ability to assume responsibility for health care encounters and other self-management of Spina Bifida is sequential. Full responsibility for self-management is critical for successful transition.^{4-5,12-13,17,45-46}
2. Assist families in knowing how to incrementally involve the child in organizing self-management activities and how to transition from parents doing to child doing with parental oversight to eventually child doing without parent oversight.^{5,7,12,44}
3. Initiate a discussion and develop action plans to address deficits in self-management and independence skills, abilities, and behaviors, as needed.
 - Use a valid and reliable instrument to assess self-management skills, abilities, and performance of self-management and independence behaviors.^{8,41,49-50}
 - Encourage increasing responsibility for behaviors such as management of medication, bowel and bladder programs, and skin-monitoring.^{15,17} (Bowel Function and Care Guidelines, Integument (Skin) Guidelines, Urology Guidelines)
 - Support development of skills necessary for self-management (e.g., decision-making, goal-setting, self-regulation, and communication).^{3,11}
 - Evaluate and monitor cognitive functions as they underpin decision-making, goal-setting, self-regulation, self-management, socialization, and transition issues.⁷⁻¹⁰ (Neuropsychology Guidelines)
 - Assess the child's ability to use transportation. Encourage their enrollment in driver's education (adaptive, if needed) if the teen possesses the necessary cognitive and motor abilities. If driving is not realistic, teach (or encourage the family to teach) them how to use public transportation, van services for individuals with disabilities, or other transportation options.⁵¹ (clinical consensus)
 - Expand self-management interventions to encompass everyday living activities such as laundry, meal preparation, money management, managing finances, and making health care appointments.^{4,11,22,39,52}
 - Encourage the family to expand the range of responsibilities for daily life activities, chores, and jobs.¹¹
 - Evaluate the potential to eventually live independently (for those later in this age range) and connect them with housing resources (e.g. Centers for Independent Living). (clinical consensus)
4. Encourage participation in IEP/504 planning that addresses self-management and transition skills. For those with an IEP, transition planning must be initiated by age 14. (Transition Guidelines)
5. Support family functioning strengths related to self-management (navigating family stress, conflict, satisfaction, and family resources).^{47,49} (Family Functioning Guidelines)
6. Involve the local Department of Vocational Rehabilitation and include vocational counseling in transition team planning. (clinical consensus)
7. When it is developmentally appropriate, include time alone with the child to discuss self-management and independence topics as part of the visit. (clinical consensus)
8. Discuss sexuality, contraception (including latex allergy precautions), marriage, childbearing issues, genetic counseling, and folic acid supplementation. (Latex and Latex Allergy in Spina Bifida, Men's Health Guidelines, Sexual Health and Education Guidelines, Women's Health Guidelines)
9. Assess individual and system barriers to self-management and transition from pediatric to adult health care (e.g., responsibility for health management, advocacy,

- assertiveness, and insufficient adult services).^{21,52}
10. Encourage the use of technology to enhance self-management.^{13,32-33,36,53}
 11. Share expectations and resources for future independent living, transition to college or employment.^{18,44}
 12. Provide consultation to adult providers with limited skill in providing care to those with congenital conditions such as Spina Bifida.⁵² (Transition Guidelines)

18+ years

Clinical Questions

1. What approaches optimize individual and family self-management?
2. What self-management skills, abilities, and behaviors lead to self-management and independent living in adults?
3. Does specific self-management skill training improve independence with self-management behaviors (e.g., taking medication and monitoring skin status)?
4. Is performing more self-management behaviors independently related to improved or positive health or functional outcomes (depression, quality of life, secondary conditions such as urinary tract infections, and pain)?
5. What health care and community supports optimize self-management, independence, and health outcomes?
6. Does increased independence with self-management increase community participation?
7. How can comprehensive preparation for self-management and independence be integrated into primary or specialty health care settings?
8. What instruments measure the individual's performance of self-management and independence behaviors in adulthood?

Guidelines

1. Evaluate full responsibility for implementing condition-specific self-management behaviors in appropriate areas, as needed (e.g. managing medications, preventing complications, monitoring skin care, maintaining equipment, bowel and bladder care, and ability to make health care appointments).^{4-5,12-13,17,41-42,45-46}
2. Reinforce the need for daily skin assessment, given the high incidence of skin breakdown on lower extremities (e.g. due to poor fitting leg braces) and risk for wound-related hospitalization.^{13,17,20} (Integument (Skin) Guidelines)
3. Evaluate if the adult has expanded self-management to encompass everyday living activities such as laundry, meal preparation, managing finances, making health care appointment and ordering supplies.
4. Initiate a discussion and develop an action plans to address deficits in self-management skills, abilities and behaviors, as needed. (clinical consensus)
 - Use a valid and reliable instrument to assess self-management skills, abilities and performance of self-management or independence behaviors over time in adults.^{8,41,49-50}
 - Support development of knowledge and skills necessary for self-management (e.g., self-efficacy, decision-making, goal setting, self-regulation, and communication).^{11,43}
 - Evaluate and monitor cognitive functions, as they underpin decision-making and self-management.^{1,18,45} (Neuropsychology Guidelines)
 - Assess the adult's ability to use transportation; encourage enrollment in driver's education (adaptive, if needed) if the adult possesses the necessary cognitive and motor abilities and has not done so already. If driving is not realistic, teach (or encourage the family to teach) the adult how to use transportation (e.g.,

- public transportation, van services for individuals with disabilities, or other transportation options). (clinical consensus)
- Evaluate the young adult's ability to live independently and connect with him or her with housing resources, such as Centers for Independent Living. (clinical consensus)
5. Encourage the use of technology in developing basic self-management skills. For instance, using email or a personal online health record, or patient portal to contact the clinic coordinator and physician with questions. Offer alternatives if this form of access is not available or appropriate.
 6. Encourage the use of technology programs to enhance self-management outcomes (e.g. using mobile health (mHealth) or telehealth tools to monitor skin breakdown or report response to medication for UTI).^{13,32-33,36,53}
 7. Expand the discussion of sexuality, contraception (including latex allergy precautions), marriage, childbearing issues, genetic counseling, and folic acid supplementation. (Sexual Health and Education Guidelines)
 8. Expand the discussion on child rearing and parenting issues and resources as appropriate. (clinical consensus)
 9. Discuss strategies for safe infant handling (e.g., holding an infant if you use a wheelchair or accessing a crib or car seat) with parents or expectant parents with mobility limitations. (clinical consensus)
 10. Encourage involvement in empowerment activities and organizations (e.g., sports, mentoring, camps, and local, national and international Spina Bifida, and other disability organizations).²⁸
 11. Support family functioning strengths related to self-management including family satisfaction and family resources.^{47,49} (Family Functioning Guidelines)
 12. Assess individual and system barriers to self-management (e.g., difficulties with self-advocacy, assertiveness, insufficient adult services).^{14-15,29,49}
 13. Refer to vocational rehabilitation, independent living centers, or other community agencies as appropriate. (clinical consensus)
 14. Provide information about accessible housing, financing, and appropriate outside agencies. (clinical consensus)
 15. Encourage planning and use of support services (e.g., in a college setting, services for students with disabilities) for self-management and independence in new environments. (Transition Guidelines)
 16. Encourage the use of wellness programs.³²
 17. Evaluate and support patients as their parents and caregivers age and assist individuals with Spina Bifida plan for changes in self-management and independence when their parents and caregivers will not be available. (clinical consensus)

Research Gaps

1. What are the foundational skills and abilities in young children that facilitate the development of self-management and independence behaviors in later childhood and adulthood?
2. What interventions optimize the development of these foundational skills and abilities early in childhood?
3. What are the barriers and facilitators for people with Spina Bifida to developing autonomy, self-management, and independent living skills?
4. What interventions are effective in closing the gap between self-management behaviors in individuals with Spina Bifida and their typically-developing peers? What

- interventions need to be targeted to patients and their parents to facilitate parental roles in self-management transitioning to coach and consultant as the patient nears adulthood?
5. Do interventions to enhance self-management and independence need to be delivered outside of clinical care?
 6. What structure(s) of clinical services are optimal for coordinated, comprehensive transition to adult care?
 7. Do routine clinical assessments of self-management behaviors, along with the development of action plans, in cooperation with the adolescent and their family yield improved outcomes?
 8. What successful strategies can health care workers use to facilitate behaviors that encourage independence in children and adults with Spina Bifida and their parents?
 9. What supports within the family and health care system lead to positive health and independence outcomes for children and adults with Spina Bifida?
 10. How do the needs related to self-management and independence of young, middle-aged, and older adults change as they age with Spina Bifida?

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Appendix A. Self-Management Instruments

Name of instrument <i>Citation</i>	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: Reliability and validity	Recommended use
Spina Bifida-Specific Instrument Developed with Samples of Youth with Spina Bifida			
<p>KKIS-SB Kennedy Krieger Independence Scales-Spina Bifida (KKIS-SB)</p> <p><i>The Kennedy Krieger Independence Scales-Spina Bifida Version: A Measure of Executive Components of Self-Management (KKIS-SB).</i>⁸</p>	<ul style="list-style-type: none"> • Twenty-two items. • Caregiver-reported measure of self-care skills for individuals ages 10 and above. • Based on assumption that self-care skills require adequate executive functioning and that other scales do not assess the executive burden of these tasks. • Four response pattern options: <ul style="list-style-type: none"> ○ More than 90% of the time ○ 10-90% of the time ○ Less than 10% of the time ○ Not necessary or no opportunity • Two subscales: Initiation of Routines And Prospective Memory. • Initiation of Routines subscale items: <ul style="list-style-type: none"> ○ Keep room clean ○ Finish chores ○ Catheterize on time ○ Get out of bed on time ○ Hygiene on time ○ Take medication on time • Prospective Memory subscale items: <ul style="list-style-type: none"> ○ Arrive at appointments on time ○ Arrange transportation ○ Look for skin breakdown ○ Start bowel program ○ Perform pressure relief ○ Write scheduled appointments 	<p>Evidence reported⁸</p> <ul style="list-style-type: none"> • Psychometric analysis with a sample of 122 parents of individuals with Spina Bifida ages 10-29. <p>Reliability</p> <ul style="list-style-type: none"> • Internal reliability $\alpha = 0.89$ • Test-retest not reported <p>Validity</p> <ul style="list-style-type: none"> • Exploratory factor analysis, reliability and construct validity using BRIEF (Behavior Report Inventory of Executive Function) were conducted. • Factor analysis supported two subscales. • Correlations between KISS-SB initiation of routines subscale and BRIEF summary scales ($r = -.031 - 0.56$) as well as five of the 8 BRIEF subscales (inhibit, shift, working memory and monitor) ($r = -.29$ to $-.62$) support validity of the KKIS-SB. • Age-related changes and correlation with the Adaptive Behavior Assessment System scales also support validity of both KKIS-SB subscales 	<ul style="list-style-type: none"> • Parent scales are recommended. • Child report version exists but psychometric data have not been published. • Contact developers for more information on child-report KKIS-SB. • KKIS-SB provides a highly-specialized assessment of self-management abilities based on a known area of challenge in individuals with Spina Bifida (executive functioning).
<p>AMIS II (Interview)</p> <p><i>The Development of the Adolescent/Young Adult Self-Management and Independence Scale-AMIS II: Psychometric Data.</i>³⁹</p>	<ul style="list-style-type: none"> • AMIS II is a 17-item structured interview instrument that measures self-management behaviors in individuals ages 12 to adult. • Parallel versions are available: <ul style="list-style-type: none"> ○ parent ○ adolescent/young adult/adult. • These generic instruments have 3 questions (complication prevention, medication, and knowledge) that can be tailored to a specific condition. • The individual is rated on how much of the behavior they actually perform and thus can be used as an outcomes measure. • Response pattern: 7 options from 0%-100%. • Two subscales: Condition Self-Management and Independent Living Self-Management. • Condition Self-Management subscale: <ul style="list-style-type: none"> ○ condition knowledge ○ medication management ○ complication prevention 	<p>Evidence reported³⁹</p> <p>Initial psychometric analysis with a sample of 201 adolescents/young adults ages 12-25 with Spina Bifida and 129 of their parents.</p> <p>Reliability</p> <ul style="list-style-type: none"> • Internal reliability $\alpha = .72-.89$. • Test-retest intraclass correlation (ICC)=0.82 supporting test-retest <p>Validity</p> <ul style="list-style-type: none"> • Factor analyses supported the two-factor AMIS II • Validity supported by age-related changes and by moderate correlations with other related variables (parent-reported chores, responsibility and functional status and adolescent/young adult report of decision-making and functional status. $r = 0.30-0.61$) • Validity also supported by use in published studies of transition-aged 	<ul style="list-style-type: none"> • Parent and adolescent/young adult/adult versions are recommended. • Scoring manual available from authors. • Self-report version now available but no psychometric evidence to date • Additionally, self-report instruments have been developed and are available for field testing. <ul style="list-style-type: none"> ○ AMIS II-SR-SB: 35-item Spina Bifida-specific instrument. ○ AMIS II-SR-G: 28-item generic instrument.

Name of instrument <i>Citation</i>	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: Reliability and validity	Recommended use
	<ul style="list-style-type: none"> ○ advocacy ○ family involvement ● Independent Living Self-Management subscale: <ul style="list-style-type: none"> ○ Making health care appointments ○ Ordering supplies ○ Household skills ○ Community living skills ○ Managing finances ○ Managing insurance 	<p>young adults. Increase in self-management over one year was related to decrease in depressive symptoms.</p>	
<p>Medical Self-Management and Transition Readiness</p> <p><i>Measurement of medical self-management and transition readiness among Canadian adolescents with special health care needs.</i>⁵⁵</p>	<ul style="list-style-type: none"> ● Twenty-one-item measure of self-management and transition readiness for individuals ages 11-18 and their parents aimed at assessing awareness of their health care condition and ability to make decisions relative to health care. ● Response pattern: 1-5 Likert-type “strongly agree” to “strongly disagree” ● Item stems are primarily “know, can, understand.” For example: <ul style="list-style-type: none"> ○ “I know what medical insurance I have.” ○ “I can get myself to medical appointments.” ○ “I have discussed sexuality-related topics with my medical professionals.” ● Others are behaviors. For example: <ul style="list-style-type: none"> ○ “I take part.” ○ “I keep track.” ○ “I have discussed.” 	<p>Evidence reported:⁵⁵</p> <ul style="list-style-type: none"> ● Psychometric study with a sample of 49 patients and their parents from a neurology clinic in Canada (only 1 person with SB) <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability strong (0.89 to 0.93 adolescent/parent). ● No stability assessment. <p>Validity</p> <ul style="list-style-type: none"> ● Means, Standard Deviations (SD) and correlations to demographic and Scales of Independent Behavior (SIB). ● No factor analysis. ● Moderate correlations between adolescent/parent (r=0.56). ● Concurrent validity: <ul style="list-style-type: none"> ● Weak relationship to age in the parent report only. ● No relationship to parent education or transition program. ● Moderate relationship to a measure of skills needed to function at home, school, or work by adolescent and parent report. ● Major finding: medical self-management closely related to independence in other domains. 	<ul style="list-style-type: none"> ● Use with caution as no psychometrics established. ● Not as well developed as KKIS-SB or AMIS II.
<p>Spina Bifida Self-Management Profile (SBSMP)</p> <p>Sharing of Spina Bifida Responsibilities Scale (SOSBMR)</p> <p>Spina Bifida Independence Scales (SBIS)</p> <p><i>Condition self-</i></p>	<ul style="list-style-type: none"> ● A series of the instruments adapted from diabetes measures. Each measure collected from mother, father, and child. ● SBSMP: 14-item structured interview of adherence to treatments (diet, catheterization, bowel program, skin checks and exercise subscales). Indicates that the task is being completed but not by whom it was done. Items scored as either adherent or non-adherent (1,0). ● Sharing of Spina Bifida Responsibilities Scale (SOSBMR): 34 items showing who is primarily responsible for each task. Scoring (1=parent, 2= equal, or child=3). 	<p>Evidence reported:</p> <ul style="list-style-type: none"> ● These measures reported in a study of 140 children with SB and their families. <p>Reliability</p> <ul style="list-style-type: none"> ● No internal reliability computed for SBSMP due to large number of “not applicable items.” Internal reliability for SOSBMR reported as Cronbach’s alpha>.60) for the 9 subscales. ● No-test-retest reliability reported for any of the scales. <p>Validity</p>	<ul style="list-style-type: none"> ● May have promise for future use, especially to compare to diabetes. ● Several scales are needed to measure these concepts and may have item burden in clinical practice.

Name of instrument <i>Citation</i>	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: Reliability and validity	Recommended use
<i>management in pediatric spina bifida: A longitudinal investigation of medical adherence, responsibility-sharing, and independence skills.</i> ¹⁷	<ul style="list-style-type: none"> Spina Bifida Independence Scales (SBIS). Parent evaluation using 50 items of child's knowledge and ability to do skills to manage Spina Bifida care (yes, no, not sure, or NA) but does not measure if the child does them on a consistent basis. 	<ul style="list-style-type: none"> Validity was generally supported by several relationships in the expected direction. Increased age was related to increased ability and responsibility. Increased ability was related to increased responsibility. However, the relationship between adherence and age was more variable. Although these scales have been used extensively in studies of children with diabetes, no formal assessment of validity in populations of children with Spina Bifida were reported. 	
Generic Self-Management/Transition Readiness Instruments. No reported use in Spina Bifida			
TRAQ (Transition Readiness Assessment Questionnaire) <i>Measures the transition-readiness of youth with special health care needs.</i> ⁵⁰	<ul style="list-style-type: none"> Twenty-item scale created to reflect Stages of Change Theory (pre-contemplation, contemplation, preparation, action, maintenance) in individuals ages 12 years and older. Response Pattern: <ul style="list-style-type: none"> 1= I do not know how to do this 2 = I do not know how, but I want to learn 3 = I am learning how to do this 4 = I have started doing this 5 = I always do this when I need to Five subscales: <ul style="list-style-type: none"> Managing medications Appointment-keeping Tracking health issues Talking to providers Managing daily activities 	Evidence reported ⁵⁰ <ul style="list-style-type: none"> Psychometric studies in three stages using 269, 178 and 526 participants respectively ranging in age from 12 to 26 years. Reliability <ul style="list-style-type: none"> Internal reliability strong=0.97 for total scale; 0.77-0.90 for subscales. No test-retest data reported. Validity <ul style="list-style-type: none"> Content validity: ethnographic interviews with adolescents/ family members to assess relevance, wording/ literacy level, intelligibility. Exploratory and Confirmatory Factor Analyses (RMSEA =0.23; GFI 0.92) support 5 subscales. All 5 subscales increase with age (p < 0.005). Gender differences found (females > males). 	<ul style="list-style-type: none"> Widely used and translated into multiple other languages. Excellent instrument for beginning discussions and facilitating movement to self-management. Does not measure increments of responsibility for implementing the behavior.
UNC TR(x)ANSITION scale. <i>A clinical tool to measure health care transition components from pediatric to adult care.</i> ⁵⁷	<ul style="list-style-type: none"> Thirty-three-item scale with 10 domains that uses a semi-structured interview format to measure issues in transition for those 12-20 years of age. Does not rely solely on patient report. Verified with information from the medical record. Ten domains: <ul style="list-style-type: none"> Type of illness Rx-Medications Adherence Nutrition Self-management Informed reproduction Trade/school Insurance Ongoing support 	Evidence reported: ⁵⁷ <ul style="list-style-type: none"> Pilot tested with a sample of 185 children/adults ages 12-20 with different chronic illnesses. Reliability <ul style="list-style-type: none"> Internal reliability supported by Item-total correlation scores (0.34 - 0.74). Inter-rater reliability was strong (kappa 0.71). No test-retest data reported. Validity <ul style="list-style-type: none"> Content and construct validity were satisfactory. Factor analysis not available Overall score was sensitive to 	<ul style="list-style-type: none"> Promising generic tool. Each program should review items and determine if interview version is compatible with their clinic to determine use.

Name of instrument <i>Citation</i>	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: Reliability and validity	Recommended use
	<ul style="list-style-type: none"> ○ New health providers 	advancing age (about a one-point increase in total score for each year of age).	
PEDI-CAT <i>Pediatric Evaluation of Disability Inventory (PEDI) Computer Aided Test (CAT).</i> ^{41, 42}	<ul style="list-style-type: none"> ● PEDI-CAT for children and adults ages 0-21. Instrument uses Item-Response Theory to measure basic activities of daily living, mobility, social/cognitive and a new scale – responsibility by youth based on parent report. ● Responsibility scale has 51 item-bank. ● Five, 10 or 15 items are based on previous answers. ● Extension of the previous paper measure; addresses functional outcomes and adds responsibility ● Response pattern for Responsibility domain: <ul style="list-style-type: none"> ○ Adult/caregiver has full responsibility; the child does not take any responsibility. ○ Adult/caregiver has most responsibility and child takes a little responsibility. ○ Adult/caregiver and child share responsibility about equally. ○ Child has most responsibility with a little direction, supervision or guidance from an adult or caregiver. ○ Child takes full responsibility without any direction, supervision or guidance from an adult or caregiver. ○ I don't know. 	Evidence reported: ⁴¹ <ul style="list-style-type: none"> ● Psychometric assessment included a sample of parents of children and adolescents with disabilities (n=2205) as well as typically developing children (n=703). ● Only responsibility (self-management) scale data reported here. Reliability <ul style="list-style-type: none"> ● PEDI-CAT is more reliable and valid than the legacy (paper) PEDI measures. Validity <ul style="list-style-type: none"> ● Confirmatory factor analysis confirmed unidimensionality of the responsibility scale (CFI<.0.99, RMSEA=0.057). ● 5,10, or 15-item scales highly correlated with total item bank (r=>0.99). ● Paper version addressing functional outcomes has been used for children with Spina Bifida.⁵⁶ ● Additional validity testing needed on responsiveness and feasibility of use by parents with limited English. 	<ul style="list-style-type: none"> ● Excellent test of independence. One of few to span the 0-21 age group. ● Useful for measuring many daily and social foundational skills as well as the incremental performance of independence of behaviors. ● Recommended if clinic/program/organization has purchased technology and if technology available on routine basis. ● Theoretically could be used with young adults without intellectual disabilities as the reporter. ● Use of Item-Response Theory means a small number of items can tap a domain, e.g. responsibility. ● Limitation: Only validated with a parent reporter at the time this guideline was written.
STARx Self-Management and Transition to Adulthood with Rx=Treatment <u><i>Self-management and transition readiness assessment: development, reliability, and factor structure of the STARx questionnaire.</i></u> ⁵⁸	<ul style="list-style-type: none"> ● Eighteen-item self-report survey for adolescents/ young adult (AYA) and parent report of three areas of transition readiness, disease knowledge, communication with medical provider, and self-management. ● Both paper and web-based administration versions available. ● Response pattern: 1-5 with “never” to “always” for behaviors; “nothing” to “a lot” for knowledge and “very hard” to “very easy” for self-management ● The instrument was developed in three phases including interviews of 29 AYA with a variety of chronic health conditions studies with strong input from AYA. <ul style="list-style-type: none"> ○ Subscales: 	Evidence Reported: ^{58, 59} <ul style="list-style-type: none"> ● Initial psychometric assessment using sample of 194 AYA for reliability and factor structure after extensive item generation and pilot studies. Samples from 8 sites were for concurrent (n=267) and predictive validity (n=847). Reliability <ul style="list-style-type: none"> ● Internal reliability of total scale was strong (Cronbach alpha =0.80). Subscale reliabilities were moderate (α=0.44 to 0.77 with half below 0.70), ● Stability (n=26) was supported by ANOVCA analysis finding of no significant difference in two administrations of STARx. 	<ul style="list-style-type: none"> ● Strong support for a brief measure of overall transition readiness. ● Recommended for self-report of AYA perceptions of knowledge, communication and select self-management behaviors in the last three months. ● Heavy emphasis on medication management.

Name of instrument <i>Citation</i>	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: Reliability and validity	Recommended use
	<ul style="list-style-type: none"> ○ Medical Management ○ Provider Communication ○ Disease Knowledge ○ Adult Health Responsibilities ○ Resource Utilization 	<p>Validity</p> <ul style="list-style-type: none"> ● Exploratory analysis yielded 6 factors: <ul style="list-style-type: none"> ○ 4 factors had 3 items. ○ 1 factor had 4 items (medication). ○ 1 factor had 2 items (resources). ○ Factor loadings were .31 to .88. ● Concurrent validity supported by strong relationships to other transition measures (e.g. TRAQ $r=0.78$). Relationship of subscales of STARx to medication use, number of hospitalizations and length of hospitalizations support STARx predictive validity. 	

Resources:

The SBA “Beyond Crayons” resources are useful in developing the knowledge, self-efficacy, attitudes and skills necessary for self-management. They can be found here: <http://spinabifidaassociation.org/beyond-crayons>

Neuropsychology

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Introduction

Neuropsychological studies show a pattern of strengths and weaknesses involving motor, cognitive, academic, and adaptive functions in individuals with Spina Bifida.¹⁻³ This pattern is most commonly seen among individuals with Spina Bifida who are born with an open spinal lesion (myelomeningocele) and usually have a Chiari II malformation and other congenital brain malformations involving the cerebellum, midbrain, and corpus callosum.⁴ Most of the existing literature is focused on patients who had hydrocephalus treated with surgical implantation of a shunt; however, the literature is just emerging on younger populations treated for hydrocephalus with different modalities, many of whom have similar cognitive profiles. It is important to identify this subgroup of patients with myelomeningocele, which makes up 90% of the population with Spina Bifida because individuals born with other types of Spina Bifida do not have these changes in neuroanatomical development and often have more typical cognitive development.⁵ The Spina Bifida Myelomeningocele (SBM) neurocognitive pattern involves strengths in learning skills and performing tasks that rely on associative, rule-based processing (e.g., math fact retrieval and vocabulary), and weaknesses when learning and performance involves the construction or integration of information (e.g. math problem-solving, reading comprehension). Many of these strengths and weaknesses are discernable across the lifespan.⁶⁻⁷

Motor Function: Children with SBM have difficulty with controlled motor performance tasks that require adaptive matching of a motor response to changing visual information, which involves the cerebellum,² and is associated with the Chiari II malformation. However, they can learn motor skills through repetition and correction of errors, which involves the relatively preserved basal ganglia,⁴ even though this type of learning may require more repetition and feedback than for typically-developing children.⁸⁻¹⁰

Perception: The ventral, object-based system involves detection of visual features and perception of categories (e.g., faces) and is critical for word reading. The dorsal, action-based system is responsible for the construction of visual representations and the linking of these representations to movement. This system relies on the posterior parietal region, which is disrupted by hydrocephalus. Children with SBM can identify faces and read words, but have difficulty with visual spatial reasoning and visually-guided goal-directed action.¹¹

Language: Strengths are noted in vocabulary and grammar. However, children with SBM experience challenges at the level of oral discourse, comprehension, and the use of language in context (pragmatics).¹² Individuals with SBM have difficulty in matching language output to a changing social language context.¹³⁻¹⁴ This has been linked to anatomic changes in the corpus callosum.¹⁵

Reading: Word recognition is often well developed¹⁶⁻¹⁷ reflecting compensation in middle temporal lobes.¹⁸ Difficulties in reading comprehension parallel those in oral language.

Mathematics: Children with SBM can learn math facts. However, complex procedures that require multiple steps and algorithms are an area of challenge. They often experience difficulties

with estimating quantities and have impaired problem-solving skills.^{17 19-20} Problems with math, a long-term predictor of adult independence, are common in adults and children with SBM.^{2,21}

Attention: Many children with SBM meet criteria for Attention Deficit Disorder, Predominantly Inattentive Type (ADD).²²⁻²³ However, in contrast to children with ADD related to frontal lobe dysfunction, the attention profile of children with SBM is characterized by under-arousal and excessive persistence in controlling attentional focus. These difficulties in alerting and orienting to external stimuli are related to disruptions in midbrain and posterior cortex and are discernable from infancy.²⁴⁻²⁶ Attention deficits involving posterior brain pathways may be a better way of understanding the self-regulation and organizational problems of individuals with SBM than is “executive dysfunction,” such as with traumatic brain injury and other injuries affecting frontal lobe function. Indeed, the frontal lobes are relatively spared in SBM. With sufficient repetition and error correction, people with SBM who have attention deficits can regulate their attention on specific tasks and learn content with persistence and over time.

Variability in the Typical Neuropsychological Profile: Understanding the variability in neuro-anatomic deficits, ethnicity, and the environment (socio-economic status and education) is the key to understanding individual (rather than group) differences in outcomes. Neurological status, including more severe hydrocephalus, repeated shunt malfunctions, and ethnicity predict poorer outcomes.⁵ Individuals with higher lesion levels have more severe neuro-anatomic brain malformations and higher rates of intellectual disability. Spinal defects at T12 and above are more frequent among individuals of Hispanic/Latino ethnicity. These populations also often have lower socio-economic status, diminished access to care and adverse outcomes attributable to social determinants of health.⁵

Psycho-educational testing provided by school districts identifies some, but not all, aspects of the SBM neurocognitive profile. A full neuropsychological assessment is recommended, when available, to document and monitor cognitive functioning in individuals with SBM.

Outcomes

Primary

1. Optimal development of language, academic, and other learning skills.
2. Optimal performance in school, university, and vocational settings.

Secondary

1. Maximize independence according to individual capabilities.
2. Maximize participation in society.

Tertiary

1. Acquisition of a job.
2. Utilization of learning skills is apparent in a variety of contexts.

0-11 months

Clinical Questions

1. What early interventions in infancy are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?
2. How are new treatments such as prenatal repair in the Management of Myelomeningocele Study (MOMS) and the Endoscopic Third Ventriculostomy/Choroid Plexus Cauterization (ETV/CPC) affecting the health and development of infants?
3. How can teams use early Magnetic Resonance Imaging (MRI) findings (e.g.,

malformations, dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?

Guidelines

1. Provide parents with formal teaching and intervention around the development of effective parenting practices for fostering developmentally appropriate and responsive parent-child interactions. Teach parents more interactive parenting strategies, as research has shown that doing so in infancy results in significantly stronger cognitive and social language outcomes (at age 3) and better social problem-solving skills (at age 7).²⁷ (clinical consensus)
2. Closely monitor infants who have undergone prenatal treatment, given the paucity of literature on their long-term outcomes.²⁸
3. Use infant development scales that assess cognition, language, motor, and social development for all infants with SBM, including those who have not been surgically treated for hydrocephalus. Adaptive behavior assessments that are interview-based are easy to complete and sensitive to growth trajectories in development.⁷

1-2 years 11 months

Clinical Questions

1. What early interventions are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?
2. How is the health and development of children changing with prenatal surgery (MOMS trials)?
3. How is the health and development of children changing with the use of new surgical procedures such as ETV/CPC rather than shunting?
4. How does monitoring for hydrocephalus and delayed shunting alter development?

Guidelines

1. Monitor and evaluate onset and progression of physical, cognitive, communicative, and social development; refer all children in this age group with SBM to an early intervention program [INSERT EARLY INTERVENTION LINK]. If children are discharged or are receiving private services, any changes in development warrant a re-referral to a formal program for early intervention/birth-three years. (clinical consensus)
2. Implement formal early intervention supports for language (delayed in onset, articulation difficulties, or unusual in pattern of development such as excessive imitation, difficulties in language comprehension), as well as physical and occupational therapy for independent mobility, strengthening, and functional activities that are essential for most children with SBM, along with parental involvement. (clinical consensus)
3. Teach and encourage parents to engage in effective interactions that facilitate the child's movement and exploration, language and communication, and play. Children of parents with higher expectations who facilitate attention, require movement, and support language development have better outcomes later in development.²⁷ (Family Functioning Guidelines)
4. Encourage the use of equipment that facilitates object exploration and manipulation because it can be essential to providing access to their environment. This may include seating to support the trunk with a large enough tray to catch objects that are dropped and parent assistance with maintaining attention to objects that are able to be manipulated and explored by the child. These supports can often be obtained through early intervention programs/birth to three [INSERT EARLY INTERVENTION LINK], occupational/physical therapy services, or from a physiatrist. (clinical

- consensus) (Mobility Guidelines)
5. Provide encouragement to participate in group learning experiences for children, especially when families are unable to find available day care that attends to necessary medical needs. These group learning experiences can be provided through either community groups or early intervention [INSERT EARLY INTERVENTION LINK]. (clinical consensus)
 6. Monitor developmental progress based on thorough assessments beyond determination of milestones, which are weak indicators of developmental difficulties. Shifts in the rate of skill development and skill regressions can reflect changes in medical status that warrant urgent follow up. (clinical consensus)
 7. Conduct periodic assessments with age-appropriate measures of early language skills because these can help identify more subtle difficulties of development. Monitor coordinated upper limb movement and attention multiple times per year in children with severe Chiari malformation, tectal beaking, and callosal hypogenesis.²

3-5 years 11 months

Clinical Questions

1. How does the relation between the nervous system and mental functions among children with SBM affect their learning in reading, mathematics, writing, social science, and science? How does it affect them at different developmental stages?
2. What do teachers, psychologists, and other professionals need to know about the development of individuals with SBM?

Guidelines

1. It is essential to carefully monitor the development of attention and self-regulation skills; these begin to emerge as a separate domain and directly affect the subsequent development of cognitive, academic, and social skills. Expectations for independent problem solving, responsibility, and social interactions are critical for school performance and psychosocial adjustment. Preschoolers with SBM show early manifestations of attention, pragmatic language, and math difficulties that subsequently emerge as major factors in academic and social adjustment.²⁹ Patients with identified concerns, even if mild, require timely referrals to the local special education preschool program and/or outpatient providers (e.g., psychologist, developmental pediatrician). (clinical consensus)
2. Monitor language comprehension problems because interventions may facilitate the development of vocabulary and conversational speech that are essential for reading comprehension later in school.³
3. Carefully observe children with more severe hydrocephalus, hypogenesis and/or severe hypoplasia of the corpus callosum and history of central nervous system infection because they are at greater risk for difficulties involving construction of meaning from language.^{13,30} These skills need to be carefully tracked by preschool education teams or through formal assessments with neuropsychologists, developmental specialists, or speech and language pathologists. (clinical consensus)
4. As part of the child's medical team, advocate for children to have access to high quality public education with related services that support the development of attention, self-regulation, social interaction skills, and independence. If parents choose private school or decide to home school, then formal assessments and recommendations for support services and supplemental resources should be provided in those settings as well. All children, regardless of placement, can and should be evaluated for eligibility for special educational services when learning problems are present. (clinical consensus)

5. Although it should not matter as to which of the 13 categories of special education a child is identified, “other health impaired” (or neurological disorder classifications in some states) helps schools understand that potential learning difficulties are related to the underlying neurological disorder. Help all individuals who interact with the child understand that SBM is not simply an orthopedic condition or “physical disability.” Brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as language, reading comprehension, and mathematics.^{16,20-21}
6. Monitor development with assessments of early math and literacy skills to help establish more subtle difficulties with development³⁰ and the need for more tailored educational supports.
7. When available, consider full neuropsychological evaluations that include the assessment of early literacy and numeracy skills. Neuropsychological assessments provide a more comprehensive understanding of strength and weakness, as well as significant discrepancies that may not be captured by psycho-educational testing that is provided by school districts. (clinical consensus)

6-12 years 11 months

Clinical Questions

1. How does the relation between the nervous system and mental functions among children with SBM affect their learning in reading, mathematics, writing, social science, and science? How does it affect them at different developmental stages?
2. What interventions support their cognitive development and academic achievement?
3. What do teachers, psychologists, and other professionals need to know about the development of individuals with SBM?

Guidelines

1. Orient health care professionals that an individual with SBM does not simply have an orthopedic impairment. Explain to them that brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as self-management skills. Learning is facilitated when it is based on rules that can be verbally mediated and rehearsed, much like a recipe. This is especially important for bladder and bowel interventions for which the child’s participation at an early age facilitates independence and social adjustment³¹ and adherence to dietary regimens. Abstract concepts and global guidelines about self-care are ineffective for skill acquisition. It is essential to create routines, so that practice and repetition of self-management tasks can become rote activities. Coach clinical teams to carefully formulate clinical instructions to be verbally mediated and to emphasize rule-based learning with repetition and rehearsal. (clinical consensus) (Health Promotion and Preventive Services Guidelines, Nutrition and Obesity Guidelines)
2. Orient educators and school-based professionals that an individual with SBM does not simply have an orthopedic impairment and that brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as language, reading comprehension, and math problem-solving. Psycho-educational assessments can track global intellectual and academic progression, but rarely assess the development of essential skills in attention, executive functioning, coordinated upper limb, and memory domains, as well as adaptive skill acquisition. Children with SBM benefit from a full neuropsychological assessment, when available. (clinical consensus)

3. Monitor school age children carefully for the onset of academic, attention, and behavioral difficulties. These problems tend to be identified later in school, partly because of the early development of word recognition, rote numerical skills, and vocabulary skills (usually in children who are not from socially- and economically-disadvantaged settings) that mask the presence of difficulties with math and reading comprehension. (clinical consensus)
4. Carefully monitor children for the onset of attention problems, as they are often interpreted as motivational or behavioral issues and are often manifested as lack of focus, slow cognitive tempo, failure to initiate, and infrequently with hyperactivity or impulsivity.²³ Attention problems are correlated with the Chiari malformation, tectal beaking, and hypogenesis of the corpus callosum.^{24-26,30,32-33}
5. Follow American Academy of Pediatrics guidelines when evaluating for ADHD.³⁴ One-third of individuals meet criteria for ADHD, predominantly Inattentive Type on parent rating scales.⁵ Interventions for attention problems that involve medications may be tried, but clinical experience suggests that lower doses are effective and that many children with SBM do not respond robustly to stimulants³⁵, most likely because the underlying attention problem emerges from posterior components of the attention network and not from the frontal-striatal networks (as in developmental ADHD). (clinical consensus)
6. Monitor children for the development of language and reading problems. The severity of hydrocephalus and corpus callosum malformations affects the child's ability to integrate information and to construct meaning from language.^{15,30} Over 25% of children with SBM have significant language and reading comprehension problems, which tend to be present both for listening and reading comprehension.⁵ Because of these common academic difficulties in children with SBM, formal assessment should include text-level reading comprehension and not just word reading accuracy and fluency.¹⁶
7. Monitor children for the development of math problems. Over 50% of children with SBM develop math difficulties.⁵ Assessment of mathematics should include assessment of complex calculation skills and, in the later grade school years, math word problems.^{19,20}
8. Implement interventions like those used with children with learning disabilities when a child has a problem with reading or math, as these are often effective. For example, although problems with word reading and phonological awareness are rare in children with SBM, treatment programs like those used with children with dyslexia have been shown to be effective. Another example is the successful use of math problem-solving interventions designed for children with math disabilities. Take advantage of children's strengths in rule-based learning by providing explicit, well-structured instruction.³⁶
9. Use assistive devices as early as possible when developing writing programs. Keyboarding is a viable alternative to handwriting, although some practice with paper and pencil skills is useful through most of elementary school. Keyboarding must be taught and rehearsed if it is to be useful. Accommodations for writing difficulties are critical components of the educational plan. (clinical consensus)
10. When available, consider full neuropsychological evaluations that include the assessment of early literacy and numeracy skills. Neuropsychological assessments provide a more comprehensive understanding of strength and weakness, as well as significant discrepancies that may not be captured by psycho-educational testing that is provided by school districts. (clinical consensus)

13-17 years 11 months

Clinical Questions

1. How does the relationship between the nervous system and mental functions among individuals with SBM affect their learning in reading, mathematics, writing, social science, and science? How does it affect them at different developmental stages?
2. What interventions and programs provide smooth transitions to post-secondary education and/or career and vocational training?
3. What do teachers, psychologists, and other professionals need to know about the development of people with SBM?
4. How do treatment teams help prepare all of their patients for the transition to adulthood, and to take on their own medical care? What indicators are helpful to a team in identifying individuals who may require ongoing support in order to have adequate management of their medical conditions?

Guidelines

1. Promote interventions that address integration and assimilation of information with a specific focus on reading comprehension and mathematics problem-solving, as well as other areas of applied mathematics and functional numeracy.^{11,19} Intervention programs should be maintained because the absence of intervention is associated with plateaus in skill development in most populations with disabilities.³⁶
2. Encourage participation in school-related and extracurricular activities and create vocational plans and transitional services with a focus on functional independence. (clinical consensus)
3. For students receiving special education services, the Individualized Education Plan [INSERT EARLY INTERVENTION LINK] (IEP) is required to include a formal transition plan to address vocational, occupational, and life skill domains by 14 to 16 years of age. Coach parents to ask about educational transition plans and to request evaluations to bolster the plans. Early transition plans are essential to develop the capacity to assume the roles and responsibilities of the post high school environment and achieve optimal independence. They are also needed to ensure that appropriate referrals are made to adult agencies, that there is suitable life and vocational skill training, and that there are discussions about plans after high school. Educate families on the need for a transition plan and check to ensure a comprehensive plan is created. If needed, refer to state-based educational advocacy programs (e.g., the ARC) that can provide support and education. (Transition Guidelines)
4. Because social skills of individuals with SBM are strongly related to neuropsychological variables, namely attention and executive function,³⁸ consider using psycho-educational and/or neuropsychological assessments to inform psychosocial interventions and mental health supports.³⁹ (Mental Health Guidelines)
5. Be aware that in addition to the cognitive and learning problems associated with the underlying neurological disorder, persons with SBM may experience reduced quantity and quality of social interactions. Encourage structured opportunities for social interaction through school, church, and afterschool opportunities. (clinical consensus) Conduct yearly screening and timely referrals for appropriate diagnosis and treatment of anxiety and/or depression with psychotherapy and/or medication treatment as needed.³⁹ (Mental Health Guidelines, Quality of Life Guidelines)
6. Identify cognitive strengths and weaknesses for those who are assuming responsibility for their own medical care. This may require formal assessment, particularly if children are unable to assume responsibility for their own medical decision making and will require guardianship. Efforts to assess and build

communication skills, increase knowledge about their medical condition and history, and develop medical triaging skills needs to begin as early as possible because it may take children in this age group over several years to learn the skills necessary to understand and take responsibility for their own medical care. Address bladder and bowel incontinence, as both can be major issues affecting social adjustment.³⁴ (clinical consensus) (Bowel Function and Care Guidelines, Transition Guidelines, Urology Guidelines)

7. Advise children and/or their parents/guardians to obtain copies of psycho-educational and/or neuropsychological assessments. Explain that documentation of intellectual disability and/or learning disability prior to age 18 is needed to qualify for services in adulthood. A diagnosis of intellectual disability requires thorough assessment of adaptive skills. This is an important point because school programs and special education service evaluations may not always include formal assessment of adaptive skills.⁴⁰

18+ years

Clinical Questions

1. How do treatment teams help prepare all of their patients for the transition to adulthood, and the assumption of their own medical care?
2. What indicators are helpful to a team in identifying those who may require ongoing support for adequate management of their medical conditions?

Guidelines

1. Many patients with intellectual disabilities or significant learning challenges will remain eligible for services through their local school districts until 21 or 22 years of age. When young adults are eligible, these services provide access to both vocational and life skills training that are essential to support the development of stronger functional independence skills. (clinical consensus)
2. Encourage that vocational services addressing job skills, additional education, and related activities be provided to appropriate individuals in a timely manner. Referrals to state-based agencies are commonly included in transition programs, and found in special education documentation/IEPs. (clinical consensus)
3. For students who received special education (IEP) or 504 Plan accommodations in high school, ongoing supports under the Americans with Disabilities Act (ADA)/Section 504 of the Rehabilitation Act are necessary. For those attending college, refer them to their college's office of disability services for ongoing educational supports. Many students will also require an updated neuropsychological assessment to support eligibility. For those in workplace environments, refer to the state-based rehabilitation/vocational commission for additional support. (clinical consensus)
4. In preparation for the transition to adult care models, where often times less coordination of medical care is provided, medical team members must take an active teaching and training role to build the necessary skills to support transition. Teach the person with Spina Bifida the skills necessary to effectively communicate with staff, recognizing that they may prefer a different method than their parents (e.g., phone calls vs. internet portal). Test patients on important aspects of their medical conditions, regimens, and allergies. Rehearse triaging medical symptomology, with clear guidelines on when to seek medical care, to mastery (e.g., not when they first get it right, but when they always get it right). (clinical consensus) (Self-Management and Independence Guidelines, Transition Guidelines)
5. Continuously monitor cognitive skills, especially math, memory, and attention, to

- ensure the maintenance of learning skills essential for work and independence.^{21,24} Changes in these areas may be a sign of unidentified shunt failure or shunt dependency, or other significant medical problem requiring intervention. (clinical consensus). Full neuropsychological assessment is recommended for adults with SBM who experience cognitive decline and suspected shunt failure. (clinical consensus)
6. Monitor for mental health concerns and potential cognitive decline with aging. (Mental Health Guidelines)

Research Gaps

1. How can teams use early Magnetic Resonance Imaging (MRI) findings (e.g., malformations, dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?
2. What early interventions in infancy are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?
3. What is the long-term effect of sequential monitoring of hydrocephalus on development? Is it better to shunt early and control hydrocephalus or to monitor ventricular expansion over time? What are the best indicators of the need for shunt diversion?
4. How well do interventions used across the lifespan involving cognition, learning, and social skills work with persons living with Spina Bifida?
5. How are attention problems best treated from pharmacological and non-pharmacological perspectives?
6. How are new treatments such as prenatal repair in the Management of Myelomeningocele Study (MOMS) and the Endoscopic Third Ventriculostomy/Choroid Plexus Cauterization (ETV/CPC) affecting the health and development of infants?

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Neurosurgery

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Introduction

Myelomeningocele (MMC) is the most common and most serious congenital anomaly of the human nervous system that is compatible with long term survival.¹⁻⁵ It arises from an error in neural development early in embryonic life and results in a variety of structural abnormalities and associated functional neurologic deficits. In open MMC the caudal spinal cord is open and exposed and distal neurologic function is lost. As such, neurologic issues last the lifespan of the individual and are central to virtually all clinical problems.²⁻⁴ Other variant forms of dysraphism are less severe and result in skin-covered anomalies that are collectively referred as occult spinal dysraphism.⁵ Spina Bifida properly refers to the full spectrum of dysraphic conditions but by convention has evolved to refer primarily to open MMC.

The publication of the Management of Myelomeningocele study (MOMs) trial galvanized the clinical landscape of neurosurgical care in MMC and cast prenatal neurosurgical issues to the forefront.⁶ This prospective, randomized, multi-center trial demonstrated improved outcomes in multiple neurological domains associated with prenatal closure including:⁶

- 1) A pronounced reduction (82% compared to 40%) in the need for ventricular shunts.
- 2) A reduction in both radiographic and symptomatic Chiari II malformations (C2M).
- 3) Improved lower extremity motor function scores that exceeded those predicted from the anatomical lesion level (on average by a single level).
- 4) A significant improvement in the composite score of neurodevelopmental outcomes.⁶
This was a secondary outcome measure and was a composite score for which the primary scores did not show significant improvement.

These improvements in fetal/infantile outcomes were offset by higher maternal morbidity, a higher incidence of premature delivery and increased risk for invasive care and obstetrical complications in subsequent pregnancies.⁶⁻¹⁰ Subsequent research by the MOMs centers has centered on refinement of surgical technique and protocols to reduce and minimize these complications.¹¹⁻¹⁵ These efforts have been fruitful and recent outcome studies suggest reductions in prematurity and maternal morbidity.¹³⁻¹⁵ There has been an associated increase in the number of centers offering Intra-Uterine Myelomeningocele Closure (IUMC). However, there remain issues and challenges that:

- mandate that these results are interpreted with caution, and
- limit the widespread availability and utility of IUMC techniques.

These issues and challenges include but are not limited to the following:

- The procedure is costly and as such is of limited contribution in environments of resource constraint (where the incidence of dysraphism is highest). Despite recent expansion in centers performing pre-natal closure, there is still limited availability of centers and access remains limited and potentially subject to disparities.
- Longitudinal outcome studies are not yet available to assess whether the favorable results are durable, lasting, and not offset by evolving new problems related to IUMC. Best available, current studies on the original MOMS cohort suggest that improvements in hydrocephalus, Chiari II malformation/brainstem dysfunction, motor function and learning are persistent.^{7,11,15} The incidence of tethered cord in infants

- who undergo IUMC appears higher than those closed by conventional techniques.¹⁸ Neurologic loss from tethered cord has some potential to reduce and offset gains seen in lower extremity motor function and bladder control observed in the original MOMS cohort. IUMC did not result in a decrease in need for clean intermittent catheterization in the most recent follow up from the MOMs cohort.¹⁸⁻²⁰
- The original maternal cohort was homogeneous and dissimilar to many of the demographics of mothers that typify mothers and families with a pregnancy with Spina Bifida.²²
 - Prematurity has been reduced but not eliminated.^{14-15,17}
 - Maternal factors remain significant. Uterine closure remains a difficult challenge and infers some risk to subsequent pregnancies and insures that delivery by cesarean section will be required for this and all subsequent pregnancies.^{16-17,21}
 - As experience with and the number of centers offering IUMC has increased there has been a simultaneous evolution of techniques such that several surgical approaches (i.e., open vs. endoscopic repair, and dural or skin patching techniques) now exist.¹⁷ It is unclear which techniques will result in the best long-term outcomes with lowest complications, morbidity and mortality. As center number increases each center is likely to see fewer cases and thereby reduce sample size associated with a given technique which may challenge studies that assessing outcome and guide technical evolution of IUMC.

Beyond pre-natal closure decisions, neurosurgical prenatal counselling of parents with a fetus with Spina Bifida is important for all families. Neurosurgeons experienced with and dedicated to caring for patients with neural tube defects (NTDs) are uniquely qualified to discuss with families the realistic long-term expectations and challenges facing a child born with open Spina Bifida (Prenatal Counselling Guidelines). Route of delivery remains a controversial issue in open MMC but significant evidence for one route of delivery over another, such as cesarean versus vaginal delivery, remains lacking.²³⁻²⁴

Neurosurgical care for most infants who are born with MMC begins with closure of the spinal defect and subsequent evaluation for the need to treat associated hydrocephalus.²⁵⁻³⁰ Ventricular shunts remain the cornerstone of treatment for hydrocephalus in Spina Bifida but there are active controversies and research surrounding:

- the thresholds for initiating treatment,
- the evolving role of endoscopic third ventriculostomy with choroid plexus coagulation (ETV/CPC).^{3-4,33-35}

Traditionally about 80% of patients with open MMC require treatment of hydrocephalus with a shunt, but the frequently problematic and troublesome natural history of shunts has fostered several experienced centers to challenge conventional thresholds for treatment.³⁻⁴ By tolerating larger ventricles and performing more local wound care several experienced centers have reduced shunt rates to 55-65%.⁵ Long-term follow up studies of the neuro-cognitive impact of these changes are unknown but appear limited in short term evaluation. Most importantly, these patients are spared the morbidity of repeated shunt operations and infections.

Endoscopic third ventriculostomy with choroid plexus coagulation (ETV-CPC) is a recently developed, promising alternative to shunts for treating hydrocephalus. Warf and colleagues refined traditional techniques of endoscopic third ventriculostomy (ETV) by adding choroid plexus coagulation (CPC) and reported initial high efficacy in a cohort of East African children with hydrocephalus from a variety of etiologies.³³ Both in the original cohort and subsequent

work by Warf's team in the United States, cohorts of children with hydrocephalus from Spina Bifida did the best of all etiologies with success rates of 70-75%.³³⁻³⁴ This led to enthusiasm and rapid expansion of the number of centers performing and offering ETV-CPC. A grading scale for success has been developed and is widely utilized to predict success of ETV-CPC.³⁵ Extensive research is underway to assess ETV-CPC but other centers appear to be struggling to attain the high rates of effectiveness observed and reported by Warf and colleagues.³³⁻³⁵

C2M remains an important issue for children with open MMC.³⁶⁻⁴⁰ By definition, every child with open MMC has a C2M, which properly refers to the entire abnormality of the brainstem and posterior fossa which is characterized by anatomic distortion with elongation and caudal displacement of the medulla and cerebellar vermis into the cervical spinal canal. This distortion imparts or is associated with brainstem dysfunction that can range widely in its clinical severity. Controversy regarding surgical management prevails but there has been a decline in the frequency with which Chiari II malformation surgical decompression (C2MD) of the posterior fossa for the C2M is performed. This decline has been in part due to:

- growing awareness of the inconsistent impact of posterior fossa decompression upon symptomatic C2M,
- the frequency with which a symptomatic C2M is precipitated by hydrocephalus or shunt failure,³⁶ and
- the recognition that some children have underlying irreversible brainstem pathology.³⁷⁻⁴⁰

Tethered Spinal Cord (TSC) is another important neurosurgical issue in Spina Bifida. Ongoing research efforts have focused on understanding the optimal thresholds and triggers for intervention, and improving technical aspects of untethering procedures to reduce re-tethering. This problem will require particular attention as children undergoing IUMC mature due to the potential for increased risk of TSC from IUMC.⁴¹⁻⁴⁴

There is increasing interest in transitional and adult care for patients with Spina Bifida.⁴⁵⁻⁴⁶ With increased survival, there are more adults than children alive with Spina Bifida, and there is a growing need for ongoing research to define optimum protocols and paradigms to maintain quality care.⁴⁵⁻⁴⁸ Early results suggest that there is a wide spectrum of quality of life for adults with Spina Bifida and that issues such as bowel management and the pursuit of personal, volunteer or job activities outside the home are associated with higher quality of life.⁴⁵⁻⁴⁹ More centers in North America are developing transition protocols and programs but much work in this domain remains.

Outcomes

Primary

1. Protect neurologic function and neurocognitive development by optimizing cerebrospinal fluid (CSF) dynamics throughout the lifespan, and by using the following parameters to balance the risks of ongoing hydrocephalus against the risks
2. of treatment:
 - presence or absence of neurological symptoms or signs (including those referable to CM2 such as stridor and poor secretion management) or tethering as manifestations of hydrocephalus and/or shunt malfunction;
 - ventricular size/morphology (particularly changes in ventricular size on serial imaging studies), yet retain the crucial awareness that important and threatening

- clinical changes can occur from shunt malfunction in the absence of demonstrable changes in ventricular size;
- head size for age as compared with normal head growth curves, and status of fontanelle(s) when applicable.
3. Perform or order adjunctive tests as necessary including ventricular imaging studies (MRI or CT), shunt taps, shunt X-rays, shunt settings (for programmable shunts), radionuclide studies, manual muscle testing, swallowing evaluations, direct laryngoscopy, sleep studies and neuropsychological testing.
 4. Preserve and sustain spinal cord function using the following interventions:
 - perform regular and ongoing assessments of spinal cord function,
 - refer to and collaborate with urology colleagues for urodynamic studies to support assessment for possible TSC,
 - recognize and diagnose tethered cord syndrome (clinically with consideration for supporting evidence from urodynamic function studies) and perform surgical tethered cord release to preserve spinal cord function and minimize recurrent spinal cord tethering,
 - optimize surveillance and treatment for symptomatic syringomyelia
 - maintain stability of brain stem and lower cranial nerve function,
 - recognize the importance of hydrocephalus and shunt failure in promoting symptomatic CM2, and
 - optimize hydrocephalus before considering Chiari decompression operations (C2MD).
 5. Improve overall mortality and morbidity of open Spina Bifida by increasing attentiveness of patient/family/medical providers to the broad clinical spectrum of neurologic decline.
 5. Educate the medical community regarding the full spectrum of signs and symptoms of ventricular shunt failure.

Secondary

1. Determine short- and long-term efficacy of intra-uterine closure to prevent recognized morbidities and mortality.
2. Define and disseminate the following quality metrics among established IUMC programs:
 - fetal morbidity metrics,
 - maternal metrics, and
 - neurological outcome metrics.
3. Minimize occurrence of shunt obstruction and infection by taking steps to:
 - reduce overall dependence upon ventricular shunts to manage hydrocephalus,
 - define and refine optimal thresholds for initial treatment of hydrocephalus, and
 - refine and optimize candidacy criteria for ETV/CPC.
4. Identify optimal strategies to prevent, diagnose, and treat symptomatic tethered cord.
5. Determine the optimum timing, frequency, and role of adjunctive studies both for surveillance and in evaluating neurologic deterioration. Maximize and protect neurologic outcome while minimizing expense and risk of diagnostic studies.
6. Establish a lifetime care model program that allows for successful transition to independent health decision-making in adulthood.

0-11 months

Clinical Questions

1. How can IUMC strategies evolve to minimize maternal risks and reduce premature delivery? What is the role for IUMC of MMC and what are its short- and long-term benefits and risks?
2. In what economic situations is IUMC a cost-effective strategy?
3. Does surgical pia-to-pia re-approximation of the neural placode (surgical “neurulation”) reduce the risk for Tethered Cord Syndrome (TCS)?
4. Does concomitant or staged closure and shunt placement reduce complications and cost?
5. What are appropriate criteria for shunt placement in infancy?
6. Are there surgical techniques that optimize shunt performance?
7. Are there optimal metrics to evaluate brain stem function?
8. What are the optimal metrics to assure optimized CSF dynamics (head growth, frequency of follow-up imaging studies and adjunctive testing)?
9. What is the appropriate role for ETV/CPC in infants with MMC?
10. What is the role for operative decompression of the posterior fossa (C2MD) for symptomatic C2M in the neonatal period?
11. What is the appropriate role, timing, and frequency of ventricular imaging in the assessment of the child from 0-11 months with open Spina Bifida?

Guidelines

Patient/Family

1. Consult with a multi-disciplinary team prior to birth to establish joint delivery plan and a plan of care. (clinical consensus)
2. Learn about regional centers that could provide evaluations for the suitability of IUMC upon prenatal diagnosis of NTD if desired. (clinical consensus)
3. Support and encourage periconceptional dietary consumption of folate to minimize the incidence of folate-related Spina Bifida.³⁹ (Women’s Health Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Meet with the parents of patients with fetal Spina Bifida soon after the diagnosis to discuss the impact of the Spina Bifida on the child and family. Review options with regard to continuation versus termination of pregnancy and IUMC and provide information on newborn care management. Provide prognosis for neurologic capabilities and limitations and explain the need for long-term multidisciplinary care. (clinical consensus) (Prenatal Counselling Guidelines)
2. Recognize indications for IUMC when infants are prenatally diagnosed with MMC, discuss this with families and refer them to regional centers that provide IUMC. (clinical consensus)
3. Define and disseminate quality outcomes for IUMC. (clinical consensus)
4. Encourage IUMC centers to seek, use, and continue to refine best available techniques to minimize premature delivery and other risks of IUMC.
5. Deliver babies with MMC who are being carried to term via cesarean or vaginal delivery. Babies undergoing IUMC are uniformly delivered via cesarean delivery. Despite the lack of consistent evidence of superiority there appears a clinical preference toward cesarean delivery.^{33,38}
6. Coordinate care with local and regional medical centers to optimize delivery, immediate care, transfer to centers with subspecialty availability and optimize early care for infant and mother. (clinical consensus)
7. Protect newborn MMC patient placode with clean, moist dressings.¹³⁻¹⁵
8. Close new MMC within 48 hours of birth in viable newborns.^{16,18}
9. Surgically re-approximate the pial edges of the neural placode (“surgical neurulation”) and close the wound in sequential layers.¹³⁻¹⁵

10. Manage CSF dynamics and acute hydrocephalus. Consider the following signs and symptoms as criteria for shunt placement or ETV/CPC:
 - increasing intracranial pressure (accelerating head growth, bulging fontanelle(s),
 - splitting sutures,
 - increasing irritability,
 - declining oral intake and/or vomiting,
 - extraocular palsies or sun setting eyes,
 - alteration in mental status,
 - brainstem signs (stridor, opisthotonus, silent cry, poor control of oral secretions, hypopnea/apnea), and
 - CSF leak from the back wound.^{2,12-13}
11. Consider C2MD for neonates in setting of brainstem crisis and only after operatively confirming the presence of functioning shunt or other adequate CSF diversion technique.^{23,25-27}
12. Encourage and help families to develop a relationship with a multidisciplinary Spina Bifida clinic.¹⁵
13. Follow infants younger than 12 months in clinic, at three to four month intervals. (clinical consensus)

1-2 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brain stem function, such as swallow and sleep studies?
3. What are the optimal metrics to assure optimized CSF dynamics (head growth, frequency of follow-up imaging studies and adjunctive testing)?
4. How does ventricular size and morphology correlate with neurocognitive outcomes?
5. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes?
6. What is the optimal frequency of clinic visits and neuroimaging during ages 1-2 years 11 months?

Guidelines

Patient/Family

1. Learn about and observe the child for clinical signs of brainstem dysfunction (stridor/silent cry/ failure to control secretions), shunt failure, and TSC. (clinical consensus)
2. Foster and develop working relationship with the team of Spina Bifida providers. (clinical consensus)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow children of 1-2 years 11 months at 6-month intervals for routine care in the Spina Bifida clinic and remain available in event of clinical change. (clinical consensus)
2. Teach families the signs of acute shunt failure (headache, vomiting, and lethargy/sleepiness) and chronic shunt failure (accelerated head growth, loss of developmental milestones or neurological deterioration). Follow the child clinically to observe for these signs. (clinical consensus)
3. Teach families the signs of brain stem failure that might occur in this age range (poor control of oral secretions, swallowing dysfunction, stridor, and impaired language acquisition). Follow the child clinically to observe for these signs. (clinical consensus)

4. Teach families the signs of TSC (back pain, declining lower extremity sensorimotor function). Follow the child clinically to observe for these signs.³⁰⁻³¹
5. Use adjunctive studies judiciously (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) to augment clinical decision-making according to clinical experience and judgment.³⁶ (clinical consensus)

3-5 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brain stem function, such as swallow and sleep studies?
3. How does ventricular size and morphology correlate with neurocognitive outcomes?
4. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes?
5. What is the optimal frequency of clinic follow-up and neuroimaging?
6. What are the optimal metrics to assure optimized CSF dynamics (head growth trajectory no longer contributory)?
7. What are the clinical presentations, surgical indications, and optimal surgical management for syringomyelia?
 - Holocord syrinx
 - Cervical syrinx
 - Thoracolumbar syrinx

Guidelines

Patient/Family

1. Teach the family to learn about and observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)
2. Foster and develop working relationship with the team of Spina Bifida providers.^{1,15} (clinical consensus)

Providers/neurosurgeons/Spina Bifida clinic

1. Follow children aged 3-5 years 11 months at intervals of 6-12 months in the Spina Bifida clinic. (clinical consensus)
2. Teach families about and review the signs of acute shunt failure (headache, vomiting, and lethargy/sleepiness), and chronic shunt failure (low grade recurring headache and neck pain, loss of developmental milestones). Follow the child clinically to observe for these signs. (clinical consensus)
3. Teach families the signs of brain stem dysfunction that might occur in this age range (poor control of oral secretions, swallowing dysfunction, stridor, and impaired language acquisition). Follow the child clinically observing for these signs. (clinical consensus)
4. Teach families the signs of TSC (back pain, declining lower extremity sensorimotor function) and urologic dysfunction. Follow the child clinically to observe for these signs.²⁹⁻³³
5. Teach families of signs of syringomyelia (back pain, sensory changes in hands). Follow the child clinically to observe for these signs. (clinical consensus)
6. Use adjunctive studies judiciously (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well child, according to experience, preference and best clinical judgment, to augment clinical decision-making. (clinical consensus)³⁶

6-12 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brain stem function, such as swallow and sleep studies?
3. How does ventricular size and morphology correlate with neurocognitive outcomes?
4. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes?
5. What is the optimal frequency of clinic visits and neuroimaging during ages 6-12 years 11 months?
6. What are the optimal metrics to assure optimized CSF dynamics (head growth trajectory no longer contributory)?
7. What are the clinical presentations, surgical indications, and optimal surgical management for syringomyelia?
 - Holocord syrinx
 - Cervical syrinx
 - Thoracolumbar syrinx
8. Does a more aggressive approach to diagnosis and surgical intervention reduce morbidity from symptomatic TSC?
9. What is the best algorithm for assessing bladder function and interpreting changes in response to somatic growth and/or tethering?

Guidelines

Patient/Family

1. Continue to encourage the family to observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)
2. Foster and develop working relationship with the team of Spina Bifida providers.^{1,15} (clinical consensus)
3. Motivate the family to establish working relationships with their child's educational system including teachers and other educational professionals. (clinical consensus)
4. Urge the family to collaborate with the clinic coordinator and/or social worker to optimize resources in the setting of potential neurocognitive dysfunction, and to identify and relay neurocognitive changes to the medical team. (clinical consensus) (Neuropsychiatry Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow children ages 6-12 years 11 months at 12-month intervals in the Spina Bifida clinic. (clinical consensus)
2. Review the signs of acute shunt failure (headache, neck pain, vomiting, and lethargy/sleepiness), and chronic shunt failure (recurring low grade headache and neck pain; loss of developmental milestones; cognitive, behavioral, or neurological decline; and orthopedic or urological regression) with the family. Follow the child clinically to observe for these signs.^{2,4,13}
3. Teach or review with the family and urge them to observe for the signs of TSC (back pain, declining lower extremity sensorimotor function, bladder or bowel control decline and progressive orthopedic deformities and/or scoliosis). Follow the child clinically to observe for these signs.²⁸⁻³³
4. Teach or review with the family and urge them to observe for signs of syringomyelia (neck or back pain and sensorimotor changes in arms and hands). Follow clinically to observe for these signs. (clinical consensus)
5. Review the signs of brain stem dysfunction that might occur in this age range (poor control of secretions, swallowing dysfunction, stridor, and declining language)

- function) with the family. Follow clinically to observe for these signs. (clinical consensus)
6. To augment clinical decision-making, use adjunctive studies during routine visits with the well child (for example, imaging such as MRI/CT and urodynamic and sleep and swallow studies), doing so judiciously and according to experience, preference, and best clinical judgment.³⁶ (clinical consensus)

13-17 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brain stem function, such as swallow and sleep studies?
3. How does ventricular size and morphology correlate with neurocognitive outcomes?
4. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes?
5. What is the optimal frequency of clinic visits and neuroimaging during ages 13-17 years 11 months?
6. What are the optimal metrics to assure optimized CSF dynamics (head growth trajectory no longer contributory)?
7. What are the clinical presentations, surgical indications, and optimal surgical management for syringomyelia?
 - Holocord syrinx
 - Cervical syrinx
 - Thoracolumbar syrinx
8. Does a more aggressive approach to diagnosis and surgical intervention reduce morbidity from symptomatic TSC?
9. What is the best algorithm for assessing bladder function and interpreting changes in response to somatic growth and/or tethering?
10. What is the cause of the observed temporal increase in shunt failure rates in children aged 13-17 years 11 months?
11. What are the neurosurgical barriers to beginning the transition process? What are the optimal strategies to assure successful transition to adult care?

Guidelines

Patient/Family

1. Observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC, and/or syringomyelia. (clinical consensus)
2. Continue to foster a working relationship with the team of Spina Bifida providers. (clinical consensus)
3. Neurosurgery should assist child and family in learning the concept of transition to adult care and in identifying an adult neurosurgery provider. (clinical consensus) (Transition Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow children ages 13-17 years 11 months at 12-month intervals in a Spina Bifida clinic. (clinical consensus)
2. Begin to address transition to adult neurosurgical provider early in teen years to promote self-knowledge and functional independence and encourage teen self-monitoring.^{32,37} (See Transition and Self-Management and Independence Guidelines)
3. Review and observe for signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), and chronic shunt failure (recurring low grade headache and neck pain, behavioral and/or cognitive changes, neurological decline, urological

- changes, and increasing orthopedic deformities and/or progressive scoliosis). Follow the child clinically to observe for these signs.^{2,4,13}
4. Review with the family and child the signs of brain stem dysfunction that might occur in this age range (poor control of secretions, swallowing dysfunction, stridor, and declining language function). Follow the child clinically to observe for these signs. (clinical consensus)
 5. Teach or review with the family and child and urge them to observe for signs of TSC (back pain, declining sensorimotor function, urological changes, and progressive orthopedic deformities and/or scoliosis). Follow the child clinically to observe for these signs.²⁸⁻³³
 6. Teach or review with the family and child and urge them to observe for signs of syringomyelia (back pain and sensorimotor changes in arms and hands). Follow the child clinically to observing for these signs. (clinical consensus)
 7. Use adjunctive studies judiciously (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well child, according to experience, preference and best clinical judgment, to augment clinical decision-making.³⁶ (clinical consensus)

18+ years

Clinical Questions

1. Does the incidence of symptomatic shunt failure change or decline in adulthood? Does a lower risk for shunt malfunction impact algorithms for monitoring shunt function?
2. What variables are associated with the highest quality of life for adults living with Spina Bifida?
3. What are the clinical presentations and optimal management of TCS in adulthood? How do these differ from TCS during childhood?
4. What is the evidence that multidisciplinary care in adulthood improves overall outcomes? Do all adults with Spina Bifida need to be followed in a multidisciplinary clinic? What is the most judicious use of neurosurgical resources in this population?

Guidelines

Patient/Family

1. Observe the adult for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)
2. Continue fostering a working relationship with the team of Spina Bifida providers. (clinical consensus)
3. Adult and family should be encouraged to review information about transitioning to adult care, including:^{34,37} (Self-Management and Independence Guidelines, Transition Guidelines,)
 - Knowledge and autonomy for personal health decisions.
 - Awareness of own body symptoms/signs.
 - Knowledge about predictors of good quality of life in adulthood.

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow adults of 18+ years at 12-month intervals in an adult Spina Bifida clinic setting. (clinical consensus)
2. Neurosurgery should assist the patient and family in identifying an adult neurosurgery provider and facilitate and support completion of transitional care. (clinical consensus) (Transition Guidelines)
3. Review with the adult and family the signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), and chronic shunt failure (recurring low grade

- headache/neck pain and changes in behavioral or cognitive function). Follow clinically to observe for these signs. (clinical consensus)
4. Review with the adult and family the signs of brain stem dysfunction in adults (poor control of secretions, swallowing dysfunction, stridor, and declining language function). Follow the adult clinically to observe for these signs. (clinical consensus)
 5. Teach or review with the adult and family and urge them to observe for signs of TSC (back pain, declining sensorimotor function, and urologic dysfunction). Follow the adult clinically to observe for these signs.²⁸⁻³³
 6. Teach or review with the adult and family and urge them to observe for signs of syringomyelia (back pain and sensorimotor changes in arms and hands). Follow the adult clinically to observe for these signs. (clinical consensus)
 7. Use adjunctive studies judiciously to augment clinical decision-making (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well adult, according to experience, preference, and best clinical judgment.³⁶ (clinical consensus)
 8. Encourage pediatric neurosurgeons to be available for education and teaching opportunities from the adult Spina Bifida team in order to learn how to provide care to adults with Spina Bifida.

Research Gaps

1. Will the long-term results and continued evolution of surgical technique in IUMC support broadening the use of this treatment? How will the results differ when IUMC is performed by a larger number of institutions and providers? How will quality be monitored, and with what indicators? How will quality metrics be disseminated to providers and families?
2. What clinical and/or radiological parameters should be used in deciding the need to treat hydrocephalus? What is the relationship between ventricular size and volume and long-term neurocognitive outcomes? Can morbidity and mortality be reduced - and quality of life improved - by reducing the use of ventricular shunts to manage hydrocephalus, without compromising long-term neurocognitive development?
3. What is the appropriate role for ETV-CPC?
4. What are the most meaningful and cost-effective studies to surveil and evaluate neurological decline, and how should these be used throughout the lifespan to optimize neurologic function?
5. How frequently does shunt malfunction occur without a demonstrable change in neuroimaging, and how does this population differ from those having ventricular enlargement?
6. Does shunt revision for radiographic change alone improve outcomes and prevent morbidity or mortality from emergent shunt failure later, or does the increased morbidity of such a strategy outweigh the benefits?
7. What is the optimum strategy to untether the spinal cord to protect and support spinal cord function throughout the lifespan?
8. What is the role of posterior fossa decompressive surgery for symptomatic C2M in infancy, childhood, or adulthood?

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Mobility

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Introduction

Mobility is defined as the ability for a person to move within his or her environment and community. Mobility can be achieved by a variety of methods. Some individuals are able to ambulate with minimal impact from their Spina Bifida while others may require orthoses or assistive devices. In children and adults with Spina Bifida, the neurological level and motor impairment are the main factors that define an individual's potential for mobility.^{1,2}

It is well known that mobility enhances development and participation in the family and community across the age spectrum. In young children mobility positively impacts cognitive, physical, and social skills. Mobility for all ages provides a way to be involved in physical activity, exercise and promote a healthy lifestyle. (Physical Activity Guidelines) Mobility may change with age especially as physical and social demands increase.

The benefits of mobility include: (clinical consensus) (Physical Activity Guidelines)

- contracture management
- exercise: cardiovascular-respiratory effects
- strength effects and endurance
- community engagement/household mobility
- bone density
- bowel and bladder evacuation
- facilitates ability to perform self-care activities
- pressure reduction/redistribution

Neurologic Level of Lesion and Anticipated Mobility

Thoracic/upper lumbar level (L1):

- Walking at this level is not common, and mobility is typically at a wheelchair level. There is no quadriceps function.^{3,4}
- Household/therapeutic ambulation requires the use of a hip-knee-ankle-foot-orthosis (HKFO) or reciprocating gait orthosis (RGO). Standing may be achieved using passive or dynamic standers.

Mid to high lumbar level (L2-L3):

- Mobility is predominantly using a wheelchair for community distances although those with an L3 level (quadriceps function) have more ability to use a mixed pattern of assisted ambulation and a wheelchair.^{3,4}
- Hip flexion is present.
- No gluteus medius/maximus function is present.
- Ambulation requires ankle-foot orthoses (AFO) or knee-foot orthoses (KAFO) and usually crutches or a walker.

Lower lumbar (L4-L5):

- Fair to good ambulation potential for both household and community.^{3,4}

- Use of AFOs is common, and an individual may use forearm crutches to improve gait pattern.
- May use a wheelchair for long distances or sports participation.
- Foot dorsiflexion is present.

Sacral level:

- Ability to ambulate but may need orthoses.^{3,4}
- May involve nerve injury of the cauda equina, so there may be sparing of some distal motor and sensory function (e.g., foot plantarflexion may be present).

The following guidelines were developed to provide a framework for care providers when discussing mobility with families. There is limited evidence, and many of the comments are based on clinical expertise.

Outcomes

Primary

1. Develop expectations for mobility based on age and neurologic level.
2. Understand and utilize appropriate mobility devices and therapy interventions to optimize mobility across the age spectrum.

Secondary

1. Reduce the threats and effects of pain, aging, neurologic deterioration, and obesity on mobility.
2. Reduce risk of pressure injuries. (Integument (Skin) Guidelines)
3. Maximize safe functional mobility and acquisition of developmental milestones for social and environmental exploration.
4. Maximize safe and functional mobility for Activities of Daily Living (ADL), as well as, social, recreational, and functional pre-vocational/vocational goals.

Tertiary

1. Understand how primary and secondary outcomes affect quality of life.

0-11 months

Clinical Questions

1. What are expected developmental milestones based on the early neurological exam related to motor skills?
2. If early mobility is delayed, do mobility devices improve developmental outcomes such as cognitive performance, social skills, and visual attention? Types of early mobility devices would include caster carts, pediatric cars, and age-appropriate manual wheelchairs.
3. Do such mobility devices help with contracture prevention?

Guidelines

1. Assess neurologic and motor level using standardized assessment tools so there is a baseline to monitor for neurologic changes. (clinical consensus)
2. Assess multi-domain developmental milestone progress using standardized tools. (clinical consensus)
3. Refer to early intervention programs and implement physical and occupational therapy programs to optimize skill attainment in fine motor and gross motor domains. (clinical consensus)
4. Maximize motor development using good body alignment with an emphasis on trunk control as a first key goal.^{5,6}

5. Use the “Back to Sleep, Prone to Play” model that emphasizes postural control acquisition as the foundation of movement. Focus on antigravity muscle activity that engages the trunk extensors before the trunk flexors. Lack of prone positioning is linked to developmental delays in typical infants and therefore has an impact on children with disabilities.⁷
6. Provide a family-centered approach and, in conjunction with the family, develop strategies to incorporate mobility within the home environment and daily routine. (clinical consensus)
7. Use casting, splinting, and orthoses to support and maintain alignment and movement. Monitor skin according to recommended guidelines. (clinical consensus) (Integument (Skin) Guidelines)
8. Collaborate with orthopedic specialists to monitor for age specific musculoskeletal problems. (Orthopedic Guidelines)
9. Encourage weight-bearing activities every day to promote bone health. (clinical consensus)

1-5 years 11 months

Clinical Questions

1. Does being overweight or obese impede the development of mobility?
2. Does a positioning/stretching program prevent contractures and how long does it need to be implemented?
3. What is the usual trajectory of gait development by neurologic level, including specific gait parameters such as cadence and efficiency?
4. What is the role of treadmill training on gait development and fitness?
5. What are the long-term consequences of walking with or without orthoses/crutches on the joints in the lower extremities and the spine?

Guidelines

1. Assess neurologic level and strength changes using standardized assessment tools at each clinic visit. Monitor for changes in gait, sensation, bowel and bladder function, and musculoskeletal changes.⁹ (clinical consensus)
2. If the child is not pulling to stand, consider using a standing frame or mobility device to get him or her upright and weight bearing. (clinical consensus)
3. Emphasize mobility options for all children including ambulation and wheelchairs. (10) Make sure parents are aware that all children who have the potential to walk may have some delay in achieving this milestone.⁵
4. Use appropriate bracing to assist weak muscles and protect the lower limbs from torque and shear forces.¹¹
5. Ensure proper wheelchair fit, posture, and technique in children who use wheelchairs, in order to reduce energy expenditure and promote long-term function. (clinical consensus)
6. Have an understanding of the coverage for durable medical equipment (DME) and how this relates to current and future DME needs. (clinical consensus)
7. Encourage weight-bearing activities every day to promote bone health. (clinical consensus)
8. Collaborate with orthopedic specialists to monitor for age specific musculoskeletal problems. (Orthopedic Guidelines)

6-12 years 11 months

Clinical Questions

1. What is the usual trajectory of mobility based on the neurologic level?

2. What are the factors that influence the transition from ambulation to wheelchair mobility for different neurologic levels?
3. What are typical gait parameters and patterns for different neurologic levels?
4. What is the role of gait analysis to monitor gait and make recommendations to optimize function?
5. Is there a benefit of early use of forearm crutches or KAFOs to protect the knee joint?
6. What is the impact of scoliosis on gait, transfers and wheeled mobility? Does spine surgery impact any of these variables?
7. In wheelchair users, are there signs of early shoulder or wrist wear and tear? Does early wheeling adversely or protectively affect upper extremity and trunk development?
8. What factors positively encourage independent mobility?

Guidelines

1. Assess neurologic level and strength changes using standardized assessment tools at each clinic visit. Monitor for changes in gait, sensation, bowel and bladder function, and musculoskeletal changes.⁹ (clinical consensus)
2. Discuss with families the benefits of the different types of mobility devices including ambulation aides and wheelchairs based on predicted mobility potential.¹²
3. Monitor walking or wheeling ability with standardized outcome measures. Consider gait studies if ambulation is changing or information is needed on optimizing bracing.¹³
4. Continue flexibility, range of motion (ROM) and strengthening exercises to maintain mobility goals, whether using ambulation devices or a wheelchair.¹⁴
5. Teach independence in putting on and taking off orthoses. (clinical consensus)
6. Educate child about importance of physical activity to maintain flexibility, strength and health, especially during growth years and explore adapted physical education opportunities or recreational sports options with the family.¹⁵ (Physical Activity Guidelines)
7. Start teaching children to be involved in their own care by educating them to watch for signs and symptoms of pressure injuries, fracture, and neurologic changes. (clinical consensus) (Self-Management and Independence Guidelines)
8. Ensure proper wheelchair fit, posture, and technique in children who use wheelchairs, in order to reduce energy expenditure and promote long-term function.¹⁶
9. Encourage weight-bearing activities every day to promote bone health. (clinical consensus)
10. Collaborate with orthopedic specialists to monitor for age-specific musculoskeletal problems. (Orthopedic Guidelines)

13-17 years 11 months

Clinical Questions

1. What is the role of gait analysis to monitor gait and recommend interventions?
2. Should forearm crutches or KAFOs be used to protect the knee when torque has been identified? When should they be instituted? Does early use prevent damage to the knee joint and prevent pain from developing?
3. What is the impact of scoliosis on gait, transfers, and wheeled mobility? Does spine surgery impact any of these variables?
4. What is the impact of linear growth on walking ability?

5. What factors influence the child's preference of wheelchair mobility over walking (for instance, energy efficiency, balance, and speed)?
6. What is the rate and pattern of loss of ambulation for community and household ambulators by neurologic level? Are there other main causes for loss of mobility besides pain, progressive weakness, growth, and obesity?
7. Are there benefits to using standing devices on ROM, bone health, and quality of life?

Guidelines

1. Assess neurologic level and strength changes using standardized assessment tools at each clinic visit. Monitor for changes in gait, sensation, bowel and bladder function, and musculoskeletal changes.⁹ (clinical consensus)
2. Monitor ambulation or wheelchair mobility. If ambulation is declining, suggest alternate mobility options. (clinical consensus)
3. Continue therapy or home programs to maintain mobility goals, emphasizing flexibility, range of motion, and overall strengthening. (clinical consensus)
4. Verify that the teenager knows how to check insensate skin, especially after activity, and how to ameliorate friction and pressure. (clinical consensus) (Integument (Skin) Guidelines)
5. Optimize gait with supportive orthoses or devices for balance. Monitor for torque forces on the joints or excessive forces in the upper body.¹¹
6. Explore the best mobility option with the teenager and have a frank discussion about the risks and benefits of all systems. (clinical consensus)
7. Monitor for a secondary injury and, if identified implement a prevention program. Areas at risk of secondary injuries for children who walk are the knees and ankles and the shoulders and wrists in those who use a wheelchair. (Orthopedic Guidelines)
8. Recommend therapy interventions to maintain mobility, if there is a change in functional status.¹⁷ (clinical consensus)
9. Collaborate with orthopedic specialists to monitor for age specific musculoskeletal problems. (Orthopedic Guidelines)

18+ years

Clinical Questions

1. What is the rate and pattern of loss of ambulation, ability to effect transfers and wheeled mobility? What causes loss of mobility function (for instance, pain, obesity, aging, and fitness)?
2. Is there a role for gait analysis to monitor gait and optimize function (for instance, to assess joint torque and shear forces)?
3. What is the role of forearm crutches or KAFOs to protect the knee when valgus forces at the knee may cause long term knee pain?
4. Are there benefits to standing devices and therapy walking as an adult?
5. What is the role of physical therapy and fitness programs in maintaining mobility?
6. What factors impact mobility long-term (i.e., improving technique, shoulder strengthening, engaging in fitness programs, and other activities)?

Guidelines

1. Assess neurologic level and strength changes using standardized assessment tools at each clinic visit. Monitor for changes in gait, sensation, bowel and bladder function, and musculoskeletal changes.⁹ (clinical consensus)
2. Monitor walking or wheeling ability and check for factors that may negatively impact mobility.^{18,19}

3. Continue to discuss the benefits of being involved in physical activities. (clinical consensus)
4. Continue with home programs to maintain flexibility, range of motion, and strengthening as this will impact mobility. (clinical consensus)
5. Optimize gait with supportive orthoses or devices for balance, monitor for torque forces at the knee or excessive forces in the upper body. (clinical consensus)
6. Teach adults with Spina Bifida about the systems of care related to mobility equipment and orthoses. Adults need to know how to identify who to call when they experience problems with their mobility devices, and the extent of their health insurance coverage and benefits. (clinical consensus)
7. Educate adults on the importance of preventing loss of mobility (both ambulation and wheelchair) through the use of appropriate technique and maintaining a healthy weight and level of strength. (clinical consensus)
8. Collaborate with orthopedic specialists to monitor for age specific musculoskeletal problems. (Orthopedic Guidelines)

Research Gaps

1. What therapies can be used in infants and children to optimize future mobility?
2. Can we develop a method to track developmental milestones based on neurologic levels, and are existing measures appropriate to use? Will this allow the medical community to develop anticipatory milestones specific for Spina Bifida?
3. What is the best method of mobility based on neurologic level/orthopedic complications and when should it be introduced?
4. What are the physiologic benefits from passive standing and how long should it be continued?
5. What are the best orthotic options for any given neurologic level?
6. What are the factors that improve energy cost for gait and mobility especially with aging?
7. How do physical changes such as growth, weight, and orthopedic changes impact mobility?
8. At what point does the frequency of joint pain at the knees, wrists, and shoulders outweigh the benefits of continued ambulation?
9. Are there better ways to protect joints from overuse injuries?
10. What impact do all the mobility options have on quality of life?
11. What are the best standardized outcome measures to monitor mobility for different age groups?

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Orthopedics

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Introduction

Orthopedic problems and musculoskeletal deformities are common in patients with Spina Bifida. The prevalence of spinal deformities including scoliosis and kyphosis is proportionate to the severity of the neurologic lesion. Spinal deformities can be challenging to treat. Body casting that can be used in idiopathic early onset scoliosis may cause skin breakdown due to insensate skin. Bracing can be used, but there is limited data to support it. Surgical treatment carries high risks including infection, pseudarthrosis, and loss of mobility. Recent studies have questioned the effect of surgery on overall function.¹⁻⁵ Nevertheless, patients with early onset scoliosis or gibbus (kyphus) deformity present special challenges due to concerns about pulmonary function. Growing rod strategies such as spine to spine or rib to pelvis distraction are increasingly used for these patients, but the small size of the patient relative to the construct and poor soft tissue envelope remain challenges.⁶⁻⁸ Further studies are needed to understand the risks and benefits of spinal surgery versus acceptance of the natural history of the spinal deformity.

Lower limb deformities can also be problematic and can affect function and gait. These can include contractures of the hip or knee or rotational deformities. Correction of rotational deformities, particularly external tibial torsion, has been shown to improve gait parameters on computerized gait analysis testing.⁹ Release of contractures can also render the limb more amenable to bracing.

Hip subluxation and dislocation are common due to muscle imbalance, particularly in the patient with mid-lumbar lesion. In previous times, hip reduction surgery, including bone procedures and muscle transfers were commonly performed. A recent study suggests that hip reduction surgery is of questionable benefit in myelomeningocele while computerized gait analyses show that contracture, not subluxation, has a deleterious effect on ambulation.¹⁰ Therefore, the use of hip reduction surgery has waned in recent decades, with the possible exception of individuals with low lumbar or sacral neurologic levels. For that reason, the present guidelines do not recommend routine surveillance of the hip or surgical treatment of hip subluxation/dislocation, although patients with a low lumbar or sacral lesion with unilateral dislocation could be considered an exception and must be treated on an individual basis.¹¹⁻¹²

The non-plantigrade foot is a frequent problem in myelomeningocele. The feet typically manifest sensory impairments and consequently, skin breakdown can occur. Orthoses such as ankle-foot orthoses (AFOs) can support the foot in patients with motor impairments, but some foot deformities may preclude bracing. A variety of orthopedic strategies, both operative and non-operative, can be used to treat foot deformities. These include stretching, bracing, serial casting, and surgery. Surgery can include tendon releases and resections, tendon transfers, joint capsular releases, osteotomies, and fusions. In general, the younger, less rigid foot may respond to soft tissue procedures while the older or more rigid foot may also require osteotomy. A classic dictum is that fusions should be avoided when possible as they render the foot more rigid which can increase the risk of skin breakdown (clinical consensus). Nevertheless, some deformities may be sufficiently severe as to require salvage procedures such as talectomy,

subtalar arthrodesis¹⁴ or triple arthrodesis.¹⁵

The proper timing for foot surgery is debatable, but a foot deformity that has become so severe as to be unbraceable in someone who still wants to pursue ambulation is certainly an appropriate indication. However, some surgeons may take a more proactive approach, performing tendon balancing surgery earlier in life to prevent bony deformity later. For example, a patient with an L4-level lesion with an unopposed anterior tibial tendon function will generally develop a calcaneus deformity. With time, weight bearing will be only on the calcaneus with no weight bearing through the forefoot. This can lead to calcaneal skin breakdown. Anterior tibial tendon transfer to the Achilles or merely anterior tibial tendon release done at an early age can improve or prevent this. If done later in life, a calcaneal osteotomy may be necessary.^{13,16} Ultimately, the approach that is taken will be at the discretion of the surgeon and the family.

Outcomes

Primary

1. Maintain a stable and balanced spine.
2. Optimize pulmonary function and avoid restrictive pulmonary disease.
3. Optimize spinal growth.
4. Avoid or facilitate healing of sacral/ischial decubiti.

Secondary

1. Maintain plantigrade feet.
2. Prevent skin breakdown.

Tertiary

1. Preserve or improve gait efficiency.
2. Early identification and stabilization, or correction, of lower limb deformities.

0–11 months

Clinical Questions

1. What are the consequences of early onset scoliosis, kyphosis and pulmonary insufficiency syndrome in patients with Spina Bifida?
2. Which foot deformities merit correction in the child 0-11 months old, and what is appropriate treatment?

Guidelines

1. Perform neonatal kyphectomy, if required to facilitate skin closure.⁶
2. Orthopedic evaluations are recommended every three months in the first year of life. (clinical consensus)
3. Consider hip imaging using ultrasound in the infant and anteroposterior pelvis radiographs after 6 months in patients with low lumbar or sacral lesions. Consider using a rigid abduction orthosis to treat hip instability, but only in children with low lumbar and sacral deformities. (clinical consensus)
4. Ponseti casting or release is recommended for clubfoot or congenital vertical talus deformities.¹⁷⁻¹⁹
5. Perform spine evaluations by conducting a physical exam. Obtain scoliosis radiographs if a spinal deformity is suspected and monitor the spine for the progression of the deformity. In children who have not achieved sitting balance, perform radiographs in a supine position. Once sitting balance is achieved, perform spinal radiographs in a sitting position. (clinical consensus)
6. Consider bracing or casting when there is a documented progression of scoliosis.²⁰

1-2 years 11 months

Clinical Questions

1. What is the proper timing for correction of rotational deformities of the femur and/or tibia?
2. Are twister cables useful for rotational deformities?
3. What is the role of bracing or Mehta casting for early onset scoliosis?
4. Should gibbus deformity be treated surgically?
5. Is rib to pelvis distraction rather than kyphectomy the optimum treatment for gibbus deformity?

Guidelines

1. Monitor the spine for development or progression of a deformity that may be due to a tethered cord or syrinx. Obtain anteroposterior and lateral scoliosis radiographs if a deformity is suspected on clinical exam. Perform radiographs in a sitting position if the patient is able to sit but not able to stand or in a standing position if the patient is able to stand. Repeat radiographs every one to two years if the deformity is present, depending on rate of progression. (clinical consensus)
2. Evaluate for neurologic changes or progression of scoliosis and discuss with neurosurgery specialists. (clinical consensus) (Neurosurgery Guidelines)
3. Initiate treatment for progressive early onset scoliosis that may involve casting²¹ or bracing.
4. Consider tendon releases/transfers for unbalanced foot deformities such as the calcaneus foot²² or equinovarus foot, if the foot is unbraceable, to facilitate orthotic management.
5. Consider twister cables for significant rotational deformities to facilitate ambulation until such time as surgical correction is appropriate.²³
6. Surgical correction of rotational deformities of the tibia or femur is recommended only if they are limiting further motor development and causing difficulty with bracing. (clinical consensus)
7. Teach families about fractures and related precautions. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. Is bracing effective for early onset, non-congenital scoliosis?
2. Is rib-pelvis distraction versus the spine-based growing rod construct the optimal treatment for progressive scoliosis?

Guidelines

1. Evaluate gait with careful attention to orthopedic deformities that render gait inefficient and preclude orthotic management. (clinical consensus)
2. Consider derotational osteotomy when rotational abnormality adversely impacts ambulation.²⁴
3. Consider treating foot deformities with stretching, casting, bracing, soft tissue release or tendon transfers to facilitate orthotic management. (clinical consensus)
4. Evaluate the spine clinically and obtain scoliosis radiographs every one to two years if a progressive spinal deformity is suspected. Perform radiographs in a sitting position in children who can sit but not stand and in a standing position in children who can stand. (clinical consensus)
5. Work with neurosurgery specialists to determine whether a neurogenic cause of scoliosis progression is present. (clinical consensus) (Neurosurgery Guidelines)
6. Consider bracing for progressive, non-congenital scoliosis in the 25 to 50-degree range. (clinical consensus)

7. It is recommended that surgical treatment of scoliosis be reserved for a progressive deformity that is unresponsive to non-operative management. For example, when there is progression of the scoliosis in spite of bracing and after a neurosurgical cause, such as a tethered cord, has been ruled out. It is also recommended that management with growing rod surgery and fusionless technique should include spinal cord monitoring in patients with distal neurologic function.²⁵
8. Consider surgical treatment of gibbus deformity for intractable skin breakdown or to free up the upper limbs for independent sitting.²⁶
9. Teach children and families about fractures and related deformities. (clinical consensus)

6-12 years 11 months

Clinical Questions

1. Is bracing effective for early onset, non-congenital scoliosis?
2. Is rib-pelvis distraction versus the spine-based growing rod construct the optimal treatment for progressive scoliosis?

Guidelines

1. Monitor gait, rotational deformities and foot position. (clinical consensus)
2. Consider correction of foot deformities to facilitate orthotic management with soft tissue release, tendon transfer and osteotomy, if necessary. It is recommended that fusion be avoided if possible. (clinical consensus)
3. Consider correction of tibial and femoral rotational deformities when they are interfering with gait and precluding orthotic management.²⁴
4. Consider conducting computerized gait analysis, when available, in children with low lumbar or sacral level lesions who have atypical gait abnormalities. This information will be helpful when making decisions regarding surgery or bracing.^{3,16}
5. Monitor for the development of scoliosis/kyphosis. (clinical consensus)
6. Obtain anteroposterior and lateral scoliosis radiographs every one to two years if deformity is suspected clinically. Do so more frequently in patients with progressive spinal deformity. Perform radiographs in a sitting position in those who can sit but not stand and in a standing position in patients who can stand. (clinical consensus)
7. It is recommended that surgical treatment of scoliosis be reserved for a progressive deformity that is unresponsive to non-operative management. An example is when the scoliosis has progressed in spite of bracing and after a neurosurgical cause, such as a tethered cord, has been ruled out. It is also recommended that management with growing rod surgery and fusionless technique should include spinal cord monitoring in children with distal neurologic function.¹⁴ Growing rod surgery with sacral-pelvic fixation is effective in correcting deformity and achieving growth.¹⁷
8. Consider surgical treatment of gibbus deformity for intractable skin breakdown or to free up the upper limbs for independent sitting.¹⁵ The current literature describes multiple techniques.^{26,28-32}
9. Teach children and families about fractures and related precautions. (clinical consensus)

11-17 years 11 months

Clinical Questions

1. What is the impact of scoliosis/kyphosis on gait, sitting balance, and upper limb function?
2. What is the relationship between spinal deformity and skin breakdown?

3. Which patients benefit from spinal deformity surgery?
4. How can spinal deformity surgery be safely accomplished?
5. In lumbar scoliosis, how high must the fusion extend?

Guidelines

1. Monitor for the development or progression of scoliosis clinically, with radiographs as necessary, if indicated by the physical exam. Perform radiographs in a sitting position in those who can sit but not stand and in a standing position in those who can stand. If the curve has progressed to an operative magnitude (50 degrees), discuss the risks and benefits of surgical treatment with the family. (clinical consensus)
2. Monitor for deterioration of gait and consider treatment of orthopedic deformities leading to deterioration such as hip and knee contracture or rotational deformities. Computerized gait analysis may be useful for decision-making in the case of children with low lumbar and sacral level lesions.^{11,16}
3. Conduct a history and physical examination (with radiographs, if indicated) on an annual basis, unless greater frequency is indicated. (clinical consensus)

18+ years

Clinical Questions

1. What is the optimal orthopedic transition plan?
2. What degenerative issues can be expected for specific levels of function (e.g., knee arthrosis for mid-lumbar lesions with valgus thrust gait pattern) and what treatments can mitigate against these problems (e.g. knee, ankle, and foot orthosis (KAFO) or crutches for the above example)?

Guidelines

1. Develop an orthopedic transition plan. (clinical consensus) (Mobility Guidelines).
2. Counsel the patient about potential orthopedic degenerative problems. Consider bracing across the knee, such as the use of a KAFO, for patients with coronal plane valgus knee stress, or adding forearm crutches to decrease coronal and transverse plane trunk motion.¹¹
3. Counsel the patient about fractures and related precautions. (clinical consensus)

Research Gaps

1. What are the consequences of early onset scoliosis, kyphosis, and pulmonary insufficiency syndrome in patients with Spina Bifida?
2. Is bracing effective in treating developmental (non-congenital) scoliosis in individuals with myelomeningocele?
3. Is Mehta-casting effective in non-congenital early onset scoliosis in myelomeningocele?
4. What is the impact of scoliosis/kyphosis on gait, sitting balance, or upper limb function?
5. Should gibbus deformity be treated surgically? What is the optimal age and surgical procedure for repairing gibbus deformities?
6. What is the relationship between spinal deformity and skin breakdown?
7. Which patients benefit from spinal deformity surgery?
8. Which patients with scoliosis will benefit from sacral-pelvic instrumentation, or one-stage versus two-stage operation? An evidence-based review concluded that spine surgery, if performed, should be through both an anterior and posterior approach.²
9. How high must the fusion extend in patients with lumbar scoliosis?

10. What is the proper timing for correction of rotational deformities of the femur and/or tibia?
11. What is the relationship between specific foot deformities and the development of skin breakdown? Does foot deformity surgery alter the risk of skin breakdown?
12. Is there an ideal, specific orthopedic transition plan?
13. Are twister cables useful for rotational deformities?
14. Which foot deformities merit correction in the child 0-11 months old, and what is appropriate treatment? What degenerative issues can be expected for specific levels of function (e.g., knee arthrosis for mid-lumbar lesions with valgus thrust gait pattern) and what treatments can mitigate against these problems (e.g. knee, ankle, and foot orthosis (KAFO) or crutches for the above example)?

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Physical Activity

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Introduction

The benefits of physical activity are well documented¹⁻⁴ and include lower rates of heart disease, depression, type II diabetes, and stroke.⁵ It has been suggested that physical activity should be used as a vital sign⁶ because it is a leading indicator of health and lowers mortality and morbidity.⁵ Unfortunately the majority of Americans fail to meet the recommended guidelines for physical activity.⁷ Additionally, it is recognized that both children and adults with Spina Bifida, as well as groups of individuals with other disabilities, are even less active when compared to the general population.⁸⁻¹⁰

Increasing physical activity for individuals with Spina Bifida may be critical since a loss of strength or fitness may lead to less independence and function in carrying out activities of daily living.¹¹ In addition to physical and functional benefits of physical activity, there are many mental health benefits such as perceived improvement in activities of daily living¹² higher athletic competence, better perceived physical appearance, greater self-worth, and higher quality of life.¹³

The National Physical Activity Guidelines are for everybody.¹⁴ These guidelines are incorporated into the Spina Bifida Guidelines for Physical Activity and should be followed as closely as possible by children and adults with Spina Bifida unless deemed medically unsafe by a health care provider. Physical activity is the key to health and wellness. Endorsement rather than caution is recommended for the vast majority of individuals with Spina Bifida.

The physical activity guidelines for children ages 6-17 state:¹⁵

- Children should engage in 60 minutes or more of physical activity each day.
- Aerobic activity should make up most of the youth's activity each day; vigorous intensity aerobic activity should be done at least 3 days/week.
- Muscle strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.
- Bone-strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.

The physical activity guidelines for adults state:¹⁵

- Adults should avoid inactivity. Some physical activity is better than none, and adults who participate in any amount of physical activity gain some health benefits.
- For substantial health benefits, adults should do at least 150 minutes (2 hours and 30 minutes) a week of moderate-intensity, or 75 minutes (1 hour and 15 minutes) a week of vigorous-intensity aerobic physical activity, or an equivalent combination of moderate- and vigorous-intensity aerobic activity. Aerobic activity should be performed in episodes of at least 10 minutes, and preferably, it should be spread throughout the week.
- For additional and more extensive health benefits, adults should increase their aerobic physical activity to 300 minutes (5 hours) a week of moderate-intensity, or 150 minutes a week of vigorous-intensity aerobic physical activity, or an equivalent

combination of moderate- and vigorous-intensity activity. Additional health benefits are gained by engaging in physical activity beyond this amount.

- Adults should also include muscle-strengthening activities that involve all major muscle groups on two or more days a week.

Please see Appendix for physical activity and disability resources.

Outcomes

Primary

1. Increase (or maintain) the minutes per day of daily physical activity at the different age levels as established by the National Physical Activity Guidelines.

Secondary

1. Increase knowledge and awareness of physical activity (i.e., benefits, safety, what/how to do it).

Tertiary

1. Improve health outcomes through physical activity participation across the lifespan (holistically, to maintain function, prevent secondary conditions, and for mental health and wellness).

0-11 months

Clinical Questions

1. Is there evidence that early motor skill intervention increases physical activity across the lifespan?
2. How early should doctors and therapists talk to parents/caregivers about physical activity for infants with Spina Bifida?
3. Are physical activity goals included in the Individual Family Service Plan (IFSP) for children beyond physical and occupational therapy?

Guidelines

1. Conduct infant motor development assessment to evaluate motor function in children with Spina Bifida to identify the most appropriate therapeutic intervention to enhance motor development outcomes.¹⁶ (Mobility Guidelines)
2. Provide guidance to parents and caregivers and include physical therapists in discussions about how to encourage movement and activity in their child.¹⁷ (Health Promotion and Preventive Health Care Service Guidelines)
3. Inform parents and caregivers of their child's right to early intervention services that include adapted physical education/activity (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans). Encourage parents/caregivers to request that physical activity goals be added to the IFSP, if eligible for IFSP.¹⁸

1-2 years 11 months

Clinical Questions

1. What strategies work to educate parents/caregivers about the importance of physical activity and ways to get their child involved?
2. How early should parent/caregiver education about physical activity start? What resources are available?
3. What is the most successful way to encourage parents/caregivers to invest time and money in physical activities for their child with Spina Bifida?

Guidelines

1. Discuss with parents and caregivers the benefits of involving their child with Spina Bifida in recreation, physical activity, and social programs and services, and provide information and/or resources about adapted and inclusive activities.¹⁵ (Health Promotion and Preventive Health Care Services Guidelines)
2. Collaborate with parents/caregivers to identify physical activities they can do in everyday life to model the importance of physical activity as part of a healthy lifestyle. (clinical consensus)
3. Use motivational interviewing techniques with parents/caregivers to talk about physical activity goals for their child with Spina Bifida and work through barriers.¹⁹
4. Inform parents/caregivers of the rights of their child to adapted physical education/activity and encourage parents/caregivers to advocate for physical activity goals to be added to their IFSP* or Section 504 plan (if they are eligible for an IFSP or Section 504 plan) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans).¹⁸

3-5 years 11 months

Clinical Questions

1. Given that some children with Spina Bifida will be limited in their physical function, what activities should they be exposed to, and when? Typically, children start playing baseball, soccer and other team sports at age 4 or 5. Should children with Spina Bifida start similar activities at the same age?
2. What training will coaches need to welcome children to meaningful play?
3. How does proper/improper mobility equipment affect the child's ability to participate in physical activity (e.g., a wheelchair that fits properly, proper seating and back setup, ankle foot orthotics, crutches, etc.)?
4. Who can parents and caregivers and doctors consult when they have questions related to physical activity and exercises that help maintain upper and lower body function (e.g., physical therapists (PTs), occupational therapists (OTs), recreation therapists, fitness staff, the National Center on Health, Physical Activity and Disability)?

Guidelines

1. Discuss with parents and caregivers the importance of involving the child in recreation, physical activity, limiting sedentary behaviors and engaging in social programs/services where they can be actively engaged with peers who have and those who don't have a disability.²⁰ Also, give parents/caregivers information about the life-long benefits of physical activity (e.g. active adults with Spina Bifida report more functional independence and a higher quality of life compared to those with Spina Bifida who are inactive).¹¹ (Health Promotion and Preventive Health Care Services Guidelines)
2. Discuss strategies with parents/caregivers that balance parental involvement in their child's physical activities and the child's need for autonomy to increase independence.²¹
3. Identify and provide additional support and information on precautions that children with shunts and ambulatory limitations should take when being physically active.²²
4. Use a team approach and include PTs/OTs to work with parents/caregivers to ensure children have proper fitting mobility equipment to maximize participation in physical activity. (clinical consensus)
5. Educate parents/caregivers of their child's right to adapted physical education/activity in preschool and encourage parents/caregivers to advocate for physical activity goals

to be added to their Individualized Education Plan (IEP) or Section 504 plan (if eligible for IEP or Section 504 plan).^{18,23}

6-12 years 11 months

Clinical Questions

1. What are some strategies to continue physical activity or physical education while in the hospital or after a long hospital stay (e.g., exercise band/tubing stretches or increasing knowledge of physical activity through reading/lecture- style learning if no physical activity is allowed)?
2. Is there evidence about the benefits/effects on the physical activity levels of children with Spina Bifida who advocate for themselves on their physical education goals in their IEP meetings?
3. Does the evidence support the least restrictive environment? Is this support related to health, socialization, something else, or a combination of factors?
4. What training do schools need to include children with Spina Bifida in meaningful play throughout the day (at recess, physical education class, on-site after-school programs)?
5. What are some ways that physical education teachers can be more inclusive of children with Spina Bifida? How do we start this process as early as possible?
6. Does getting children engaged in sports at a young age improve the likelihood that they will remain engaged in activity throughout their lifespan?

Guidelines

1. Discuss the benefits of participating in physical activity, recreation, and sports with children with Spina Bifida.^{24,29} Discuss with parents/caregivers the importance of limiting sedentary behaviors²⁰. Encourage parents to give their child choices about where they can be actively engaged with peers who have and those who don't have a disability.²⁵ (Health Promotion and Preventive Health Care Services Guidelines)
2. Recommend that parents/caregivers follow the National Physical Activity Guidelines for their child with Spina Bifida as closely as possible unless a health care provider advises that they are medically unsafe. (clinical consensus)
 - Engage in 60 minutes of physical activity or more each day.¹⁵
 - Aerobic activity should make up most of the child's activity each day; vigorous intensity aerobic activity should be done at least 3 days/week.
 - Muscle strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.
 - Bone strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.
3. Use health care encounters and follow-up meetings to develop physical activity goals and monitor progress (e.g. minutes of physical activity per day). Use motivational interviewing techniques with the child and parents/caregivers to talk about physical activity goals and work through barriers.¹⁹ Support parents/caregivers to develop an action plan with strategies to support their child's participation in physical activity in their community and school.²⁸ Prescribe, using a prescription pad, physical activity based on goals identified by the child.¹⁴ (clinical consensus)
4. Perform pre-participation evaluations for children with Spina Bifida in collaboration with the child and family, pediatric specialists, therapists, coaches, and others to identify medical risks and modifications that can be made to ensure participation.²⁴
5. Identify strategies to minimize risk of illness and injury related to participation through activity adaptations and safety precautions.²⁴ Identify and provide additional support and information for parents/caregivers on precautions to take when children with

- shunts and ambulatory limitations are being active.²² Discuss strategies that balance the parents'/caregivers' involvement with their child's need for independence when they participate in physical activity.²¹
6. Use a team approach and include PTs/OTs to work with parents/caregivers to ensure their child has proper fitting mobility equipment to maximize physical activity participation. (clinical consensus)
 7. Work with children with Spina Bifida and their family to address personal barriers such as bowel/bladder care, medical events, assistive devices, as well as environmental factors that may affect participation.³⁰
 8. Advocate for and address barriers to participation of children with Spina Bifida in physical activity, recreation, and sports.²⁴
 9. Inform parents/caregivers of their child's right to adapted physical education/activity and encourage parents/caregivers to advocate for physical activity goals to be added to their child's IEP or Section 504 plan (if eligible for IEP or Section 504 plan).^{18,23}
 10. Advocate for the participation of children with Spina Bifida in both unified and adapted sports, recreation, and physical activity programs.²⁴
 11. Provide families with a local/regional therapeutic recreation and adapted sport resource guide.²²

13-17 years 11 months

Clinical Questions

1. Do adapted physical education programs in schools adequately prepare (via a transition plan) children with Spina Bifida to lead physically active lifestyles?
2. What are the physical activity contraindications for children with Spina Bifida?
3. What are the types of physical activities used or recommended in the literature specific to children with Spina Bifida (resistance, cardio, incidental activity vs. planned physical activity/exercise)?
4. In what setting are children the most comfortable and likely to continue participation in a physical activity (e.g., in a group, at home, etc.)?
5. What are the doses of physical activity used or recommended in the literature for children with Spina Bifida? Are they effective for health changes?
6. Is physical activity included in the Individualized Transition Plan for children? (Transition Guidelines)

Guidelines

1. Discuss with children the benefits of participating in physical activity, recreation, and sports.^{24,29} Discuss with children and parents/caregivers the importance of limiting sedentary behaviors.²⁰ Encourage children and parents/caregivers to consider choices about where they can be actively engaged with peers who have and those who don't have a disability.²⁵ (Health Promotion and Preventive Health Care Services Guidelines)
2. Recommend that children and parents/caregivers follow the National Physical Activity Guidelines as closely as possible, unless a health care provider advises that they are medically unsafe. (clinical consensus)
 - Engage in 60 minutes of physical activity or more each day.¹⁵
 - Aerobic activity should make up most of the child's activity each day; vigorous intensity aerobic activity should be done at least 3 days/week.
 - Muscle strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.
 - Bone strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.

3. Use health care encounters and follow-up meetings to develop physical activity goals and monitor progress (e.g. minutes of physical activity per day). Use motivational interviewing techniques with children and parents/caregivers to talk about physical activity goals and work through barriers.¹⁹ Support children and parents/caregivers to develop an action plan with strategies to support the participation of children age 13-17 years in physical activity in their community and school.²⁸ Prescribe, using a prescription pad, physical activity based on goals identified by children with Spina Bifida.¹⁴ (clinical consensus)
4. Perform pre-participation evaluations for children with Spina Bifida in collaboration with parent/caregivers, pediatric specialists, therapists, coaches, and others to identify medical risks and modifications that can be made to ensure participation.²⁴
5. Identify strategies to minimize risks of illness and injury related to participation through activity adaptations and safety precautions.²⁴ Identify and provide additional support and information for children age and parents/caregivers on precautions to take when children with shunts and ambulatory limitations are being active.²² Discuss strategies that balance the parents'/caregivers' involvement with their child's need for independence when they participate in physical activity.²¹
5. Use a team approach and include PTs/OTs to work with parents/caregivers to ensure their child has proper fitting mobility equipment to maximize physical activity participation. (clinical consensus)
6. Work with children with Spina Bifida and their family to address personal barriers such as bowel/bladder care, medical events, assistive devices, as well as environmental factors that may affect participation.³⁰
7. Advocate for and address barriers to participation of children with Spina Bifida in physical activity, recreation, and sports.²⁴
8. Inform parents/caregivers of their child's rights to adapted physical education/activity and encourage children and parents/caregivers to advocate for physical activity goals to be added to their IEP or Section 504 plan (if eligible for IEP or Section 504 plan).^{18,23}
9. Assist students who are considering post-secondary education to assess supports for physical activity in the educational institutions they are considering and include these in their individual transition plans.
10. Advocate for the participation of children with Spina Bifida in both unified and adapted sports, recreation, and physical activity programs.²⁴
11. Provide families with a local/regional therapeutic recreation and adapted sport resource guide.²²
12. Discuss with children where they feel most comfortable being physically active and about their options (e.g., in the community, at school, with peers with/without disabilities).²⁵
13. Highlight that ages 13-17 years is a critical period to build physical activity into a daily routine that will preserve overall lifelong satisfaction and community participation among persons with Spina Bifida.²⁹

18+ years

Clinical Questions

1. What are the physical activity contraindications for adults with Spina Bifida?
2. What are the types of physical activities used or recommended in the literature specific to adults with Spina Bifida (resistance, cardio, incidental activity vs. planned physical activity/exercise)?

3. What is the best setting for physical activity for adults (e.g., in a group, at home, etc.)? Where are they most comfortable and likely to continue participation?
4. What are the doses of physical activity used or recommended in the literature for adults with Spina Bifida? Are they effective for health changes?

Guidelines

1. Discuss the National Physical Activity Guidelines with adults with Spina Bifida.¹⁵ (Health Promotion and Preventive Health Care Services Guidelines)
2. Follow the guidelines for adults with Spina Bifida as closely as possible, unless a health care provider advises that they are medically unsafe. (clinical consensus)
3. For substantial health benefits, it is recommended that adults should do at least 150 minutes (2 hours and 30 minutes) a week of moderate-intensity, or 75 minutes (1 hour and 15 minutes) a week of vigorous-intensity aerobic physical activity, or an equivalent combination of moderate- and vigorous-intensity aerobic activity. It is recommended that aerobic activity should be performed in episodes of at least 10 minutes, and preferably, it should be spread throughout the week. (clinical consensus)
4. For additional and more extensive health benefits, it is recommended that adults should increase their physical activity to 300 minutes (5 hours) a week of moderate-intensity aerobic exercise, or 150 minutes (2 hours and 30 minutes) a week of vigorous-intensity aerobic physical activity, or an equivalent combination of moderate- and vigorous-intensity activity. Additional health benefits are gained by engaging in physical activity beyond this amount. (clinical consensus)
5. It is recommended that adults should include muscle-strengthening activities that involve all major muscle groups on 2 or more days a week. (clinical consensus)
6. It is recommended that all adults should avoid inactivity. Some physical activity is better than none, and adults who participate in any amount of physical activity gain some health benefits. (clinical consensus)
7. Identify and provide additional support and information on physical activity precautions for adults with shunts and ambulatory limitations.²²
8. Use health care encounters and follow-up meetings to develop physical activity goals and monitor progress (e.g. minutes of physical activity per day). Employ motivational interviewing techniques to discuss and set physical activity goals and strategies to overcome barriers to achieving those goals.¹⁹ Discuss the importance of physical activity and physical activity options with adults with Spina Bifida.¹¹
9. Prescribe, using a prescription pad, physical activity based off on goals discussed with adults with Spina Bifida.²⁷
10. Assist students who are considering post-secondary education to assess supports for physical activity in the educational institutions they are considering.
11. Use a team approach and include PTs/OTs to work with the adult with Spina Bifida to make sure that their mobility equipment fits properly in order to maximize their participation in physical activities. (clinical consensus)
12. Emphasize that any movement is beneficial.¹⁵

Research Gaps

1. What are the strategies/solutions needed to intrinsically motivate individuals with Spina Bifida to be physically active at different ages across the lifespan (e.g., peer support, increased knowledge, self-efficacy, and other approaches)?
2. How can we best educate and train parents and individuals with Spina Bifida to be advocates for their inclusion in physical activity in their communities at large?

3. What physical activity resources are available for doctors nationwide? Locally? What resources need to be created?
4. Is there evidence that physical activity prevents secondary conditions? If so, how strong is this evidence?
5. What are the social/health benefits of participating in physical activity for individuals with Spina Bifida at different ages across the lifespan?
6. What are some strategies to continue physical activity or physical education while in the hospital or after a long hospital stay (e.g., exercise band/tubing stretches or increasing knowledge of physical activity through reading/lecture- style learning if no physical activity is allowed)?
7. Is there evidence that the early motor skill intervention increases physical activity across the lifespan?
8. What strategies work to educate parents/caregivers about the importance of physical activity and ways to get their child involved?
9. How early should parent/caregiver education about physical activity start? What resources are available?
10. What is the most successful way to encourage parents/caregivers to invest time and money in physical activities for their child with Spina Bifida?
11. Given that some children with Spina Bifida will be limited in their physical function, what activities should they be exposed to, and when? Typically, kids start playing baseball, soccer and other team sports at age 4-5 years. Should children with Spina Bifida start similar activities at the same age?
12. What training will coaches need to welcome children to meaningful play?
13. How does proper/improper mobility equipment affect the child's ability to participate in physical activity (e.g., a wheelchair that fits properly, proper seating and back set up, ankle foot orthotics, crutches, etc.)?
14. Who can parents/caregivers and doctors consult when they have questions related to physical activity/exercises that help maintain upper and lower body function (e.g., physical therapists (PTs), occupational therapists (OTs), recreation therapists, fitness staff, National Center on Health, Physical Activity and Disability)?
15. Is there evidence about the benefits/effects on the physical activity levels of children with Spina Bifida who advocate for themselves on their physical education goals in their IEP meetings?
16. Does the evidence support the least restrictive environment? Is this support related to health, socialization, something else, or a combination of factors?
17. What training do schools need to include children with Spina Bifida in meaningful play throughout the day (at recess, physical education class, on-site after-school programs)?
18. What are some ways that physical education teachers can be more inclusive of children with Spina Bifida? How do we start this process as early as possible?
19. Does getting children engaged in sports at a young age improve the likelihood that they will remain engaged in activity throughout their lifespan?
20. What is the best setting for physical activity for adults (e.g., in a group, at home, etc.)? Where are they most comfortable and likely to continue participation?
21. What are the doses of physical activity used or recommended in the literature for adults with Spina Bifida? Are they effective for health changes?

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Appendix

1. National Physical Activity Guidelines (can be read online, or downloaded as a PDF):

- <https://health.gov/paguidelines/guidelines/>
2. The Centers for Disease Control and Prevention's disability & physical activity resources for doctors and other health professionals:
<https://www.cdc.gov/ncbddd/disabilityandhealth/pa.html>
 3. The National Center on Health, Physical Activity, and Disability (NCHPAD):
<http://www.nchpad.org/>
 4. NCHPAD's health professionals resources (includes DocTalk and other videos for doctors and other health professionals):
<http://www.nchpad.org/Health~Care~Providers>
 5. NCHPAD's Physician's Toolkit:
<http://www.nchpad.org/1195/5822/Physician~s~Toolkit>
 6. Foundation for PM&R RX for Exercise (resources for doctors to prescribe exercise):
<http://foundationforpmr.org/rx-for-exercise/>



Men's Health

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Introduction

Until recently, adult sexual function in men and women with Spina Bifida had not been widely considered, as many born with this condition did not live to adulthood. Even after the advent of modern medical breakthroughs like ventriculoperitoneal shunting, intermittent catheterization, and urinary diversion increased quality of life and longevity, many adults with Spina Bifida continue to be cared for by pediatric specialists well into adulthood. Similarly, urologic issues that affect adults are often ignored.

It is clear that sexual function is altered in a majority of men with Spina Bifida, as male sexual organs are innervated by the distal spinal cord which is often impaired by Spina Bifida. Evidence suggests that young adults with Spina Bifida generally feel under informed about sexual health, with nearly one third of respondents stating that they were not provided appropriate information related to how Spina Bifida can affect sexual function)¹⁻³ Additionally, traditional points of emphasis in men's health care, such as prostatic hypertrophy and cancer, have not been addressed in this population. The health care community now widely accepts the need for a better understanding of the specific issues that men and women with Spina Bifida face regarding sexuality, fertility, and aging reproductive organs.

This document will review the following men's health topics:

- Male sexual function
- Male fertility considerations
- Prostate cancer screening and treatment

The purpose of these guidelines is to: 1) highlight the existing evidence regarding the male sexual health in Spina Bifida, 2) make recommendations based on existing data and expert opinion, and 3) emphasize research gaps and areas for additional opportunities to improve the health of men with Spina Bifida.

Sexual Function: Outcomes

Primary

1. Optimize sexual function and fertility in men with Spina Bifida.

Secondary

1. Evaluate and characterize penile and genital sensation.
2. Evaluate and characterize erectile function.
3. Evaluate and characterize orgasmic and ejaculatory function.
4. Maximize fertility potential of men with Spina Bifida, if desired.
5. Ensure sexual education and safe practices (Sexual Health and Education Guidelines).
6. Determine the sexual activity and interest in men with Spina Bifida.

Tertiary

1. Describe known therapies for decreased genital sensation, erectile/orgasmic/ejaculatory dysfunction, and infertility.
2. Assess the impact of fertility and sexual function on the quality of life in men with

Spina Bifida.

Men's Health Guidelines begin at age 6-12 years 11 months

6-12 years 11 months

Clinical Questions

1. Are there strategies to promote healthy self-identity and avoid sexual abuse?
2. When should a testicular exam be conducted for boys with Spina Bifida?

Guidelines

1. Provide anticipatory guidance regarding sexual function and its potential challenges. (clinical consensus)
2. Conduct an annual scrotal exam that documents testicular position, size, consistency, symmetry, and presence or absence of masses. (clinical consensus)
3. Access and document genital sensation (penile, scrotal) and Tanner staging annually.⁴⁻⁶
4. Offer human papillomavirus (HPV) vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, if appropriate.⁷⁻⁸

13-17 years 11 months

Clinical Questions

1. When should a testicular exam be conducted?
2. What is the prevalence of decreased penile/genital sensation?
3. What is the prevalence and nature of erectile dysfunction?
4. What is the best way to inquire about and assess sexual dysfunction?
5. What is the understanding of normal sexual function as well as Spina Bifida-related alterations in boys with Spina Bifida?
6. At what age or developmental level should sexual function and fertility evaluation be offered?
7. Are boys with Spina Bifida in this age group aware of contraceptive techniques, specifically the availability of latex-free condoms? Are latex-free condoms as effective as latex-containing condoms? Are there alternative methods of barrier contraception for this population?

Guidelines

1. Conduct an annual scrotal exam that documents testicular position, size, consistency, symmetry, and presence or absence of masses. (clinical consensus)
2. Access and document genital sensation (penile, scrotal) and Tanner staging annually.⁴⁻⁶
3. Instruct men about monthly testicular self-examinations (TSE). (clinical consensus)
4. Initiate open-ended conversations with boys age 13-17 with Spina Bifida about their knowledge of normal sexual function when the provider deems that the boy is developmentally ready, or when there is evidence of sexual curiosity and experimentation in their medical history.⁹⁻¹² (clinical consensus) (Sexual Health and Education Guidelines)
5. Educate patients that sexual function may be altered as a sequela of Spina Bifida.^{10-11,13} (clinical consensus)
6. Provide information about safe sexual practices and genetic risk factors.¹⁴ (clinical consensus) (Sexual Health and Education Guidelines)
7. Offer human papillomavirus (HPV) vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, as appropriate.⁷⁻⁸

8. Characterize and document erectile function when it becomes developmentally appropriate. Providers may use the International Index of Erectile Function (IIEF) or Sexual Health Inventory for Men (SHIM)^{9,15} (clinical consensus)
9. When relevant, characterize and record orgasmic and ejaculatory function.^{10,13,16} (clinical consensus)

18 + years

Clinical Questions

1. What is the prevalence of hypogonadism (abnormal testes and/or testosterone levels) in men with Spina Bifida?
2. What is the prevalence of decreased penile/genital sensation in men with Spina Bifida?
3. What is the prevalence and nature of erectile dysfunction in men with Spina Bifida?
4. What is the best way to inquire about and assess sexual activity in men with Spina Bifida?
5. What is the understanding of normal sexual function as well as Spina Bifida-related alterations in men with Spina Bifida?
6. What is the best way to inquire about sexual function, including nocturnal emissions, non-genital stimulation, masturbation, and oral and genital contact?
7. How much does sexual function influence the quality of life in men with Spina Bifida?
8. Are men with Spina Bifida aware of contraceptive techniques, specifically the availability of latex-free condoms? Are they as effective as latex-containing condoms? Are there alternative methods of barrier contraception for this population?
9. What are the paternity goals and outcomes in men with Spina Bifida?
10. What is the optimal approach to men with Spina Bifida desiring an infertility evaluation?
11. How much does fertility and paternity influence the quality of life in men with Spina Bifida?

Guidelines

1. Conduct annual scrotal exam that documents testicular position, size, consistency, symmetry, and presence/absence of masses.⁴⁻⁶
2. Assess and document genital sensation (penile, scrotal) yearly⁴⁻⁶
3. Instruct patients on monthly testicular self-examination (TSE) to age 40 (clinical consensus)¹⁷⁻¹⁸
4. Inform patients that sexual function and reproductive capacity may be altered as a sequela of Spina Bifida.^{10-11,13,16,19}(clinical consensus)
5. Provide information about safe sexual practices and genetic risk factors.¹⁴ (clinical consensus) (Sexual Health and Education Guidelines)
6. Refer the man to a urologist with expertise in male sexual function if he expresses concern regarding sexual dysfunction or an exam suggests impaired sensation or function of the genitalia. (clinical consensus) Similarly, it is recommended to make an appropriate referral to a specialist in male sexual function and/ or male infertility if he expresses concern related to orgasmic or ejaculatory function. (clinical consensus)
7. Characterize and record erectile function, orgasmic and ejaculatory function when relevant. (clinical consensus)
8. Explain to men with Spina Bifida that phosphodiesterase inhibitors are first-line pharmacologic treatments for erectile dysfunction. Men should be offered these treatments and instructed on their use if they do not have contraindications. (clinical consensus)

9. Employ open-ended questions to explore the man's interest in paternity and concerns about the heritability of Spina Bifida. Offer genetic counseling and infertility evaluation when questions about these topics arise.¹¹ (clinical consensus)
10. Educate men about the risk of heritability of Spina Bifida for their offspring and offer their female partners additional supplementation with folic acid to reduce the risk. (clinical consensus) (Women's Health Guidelines)

Research Gaps

1. There is a need to characterize sexual function and interest among men with Spina Bifida.
2. There is a lack of understanding about the impact of sexual dysfunction on quality of life among men with Spina Bifida.
3. There is a need to characterize the incidence and etiology of hypogonadism in men with Spina Bifida.
4. Mechanisms should be developed and standardized to assess and monitor penile/genital sensation in men with Spina Bifida.
5. The prevalence and nature of penile/genital sensation based on the type and level of lesion in men with Spina Bifida needs to be characterized.
6. There is a need to understand the prevalence and nature of erectile dysfunction in men with Spina Bifida.
7. Validated questionnaires for erectile, ejaculatory, and orgasmic dysfunction specific to men with Spina Bifida or other congenital neuropathies are needed.
8. The extent of the effect of sexual dysfunction (erectile, ejaculatory, and orgasmic), decreased genital sensation, and fertility concerns on quality of life in adult men with Spina Bifida remains uncharacterized.
9. There is a lack of mechanisms and tools to assess young men's and men's developmental readiness to discuss sexual function and interest.
10. There is a lack of information on the prevalence of infertility, and mechanisms to treat infertility in men with Spina Bifida are undefined.
11. The impact of infertility and paternity on the overall quality of life in men with Spina Bifida is unknown.
12. Information is needed on the use, safety, and need of latex-free condoms in men with Spina Bifida.
13. Research is needed to determine whether early sensation is predictive of future male sexual function.
14. Information is needed to determine the best strategies to promote anatomical awareness and a healthy self-identity, and to avoid sexual abuse.
15. There is a need to improve the characterization of paternity goals and outcomes in men with Spina Bifida.

Prostate Health: Outcomes in Men 18+

Primary

1. Address urologic cancer screening criteria specifically for adults.

Secondary:

1. Achieve optimal use of prostate-specific antigen (PSA) testing.
2. Follow advanced screening considerations of adult males with Spina Bifida.

3. Follow treatment considerations for adults with prostate cancer (PCA) and Spina Bifida.

18+ years

Clinical Questions

1. How should men with Spina Bifida be screened for PCA?
2. What additional testing could be offered to men to appropriately screen them for PCA, such as genomic testing and MRIs? When are these appropriate?
3. Are there specific recommendations for antibiotic prophylaxis for transrectal ultrasonography (TRUS) biopsy in a man with chronic bacteriuria?
4. Are PSA norms established for men who perform intermittent self-catheterization?
5. How should men be evaluated and counseled for treatment after a diagnosis of PCA is established?

Guidelines

1. Do not offer PSA testing to men with a life expectancy of less than 10-15 years or to men who are <55 and >69 years of age unless they are at elevated risk for prostate cancer based on family history.²⁰
2. For men between the ages of 55-69 with neuropathic bladder and chronic bacteriuria with at least 10-15 years life expectancy, the value of PSA alone as a screening tool is low. Discuss and offer PSA and digital rectal exam (DRE) testing as appropriate.²⁰
3. If a biopsy is recommended, consider using MRI-guidance, transperineal technique, and pre-treat men with culture-specific antibiotics prior to biopsy.²¹⁻²³
4. Consider waiting for fPSA normalization and tPSA nadir, typically about 12 weeks, before determining whether a biopsy should be performed based on elevated PSA in men with congenital neuropathic bladder on ISC who had a recent urinary tract infection.²⁵⁻³⁰
5. Adequately assess pre-treatment bowel, urinary, and sexual function to guide counseling about treatment options for prostate cancer.³¹ (clinical consensus)
6. Prior to decision-making for treatment of prostate cancer, men with Spina Bifida may benefit from adjunct testing to fully characterize the risks of various treatments (e.g., cystourethroscopy to evaluate the external sphincter or urodynamics to evaluate bladder storage function).³¹ (clinical consensus)

Research Gaps

1. The question of PSA cutoff for biopsy has not been clearly elucidated in any population.
2. Incidence of screening in men with Spina Bifida is unknown.
3. No studies have been conducted to investigate outcomes after treatment for prostate cancer in men with Spina Bifida.
4. Determine the effect of intermittent self-catheterization on prostate-specific antigen testing.
5. Define the role of digital rectal exams on cancer screening in men with Spina Bifida.

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Sexual Health and Education

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Introduction

As stated by the World Health Organization, “Sexual health is a state of physical, emotional, mental and social well-being in relation to sexuality; it is not merely the absence of disease, dysfunction or infirmity. Sexual health requires a positive and respectful approach to sexuality and sexual relationships, as well as to the possibility of having pleasurable and safe sexual experiences, free of coercion, discrimination, and violence.”¹⁻²

Sexual Health

Peer-reviewed health literature indicates that people with Spina Bifida have varying levels of satisfaction with their sex lives, with approximately half reporting dissatisfaction with their sex lives.^{3,4} Sexual satisfaction and intimacy are directly related to quality of life,⁵ but they are rarely studied. Sexual activity in people with Spina Bifida is delayed.⁶ People with the lowest lesion levels had the highest chance of finding a partner and engaging in sexual activity.^{6,7} Lower lesion levels are associated with sexual satisfaction.⁸ In general, having hydrocephalus was predictive of having more problems with sexual function and relationships.⁴ Urinary incontinence was associated with altered sexual functioning in multiple studies,^{7,9-10} but not all.⁶ Bowel and bladder incontinence has been demonstrated to interfere with sexual activity,¹¹ such that continence enhances sexual functioning.¹² As may be expected, restored penile sensation is associated with improved sexual health and satisfaction.¹³

Sexual Education

The receipt of sexual education, especially as it relates to Spina Bifida, has been found to be inadequate in multiple studies.^{7,10-11} Adolescents with Spina Bifida are less knowledgeable about sex than their peers.¹² Sex education specifically concerning Spina Bifida was rarely provided by health care professionals.¹¹ Fewer than a quarter of people reported that their sexual education was specific to Spina Bifida.⁴ In a study by von Linstow, less than half of the subjects reported that their sexual education at puberty was useful and one-third lacked knowledge about how their sexual functioning was impacted by their disability.³

Both young men and young women wanted more information from their health care providers especially about sexual education specific to living with Spina Bifida.¹⁴ Young women with Spina Bifida want increased knowledge of fertility, birth control and heredity of Spina Bifida.¹⁵ In a qualitative study, the questions and concerns that youth with Spina Bifida reported fell into four themes: romantic relationships, sexuality, fertility/parenthood, and the need for more sexual education.¹⁶ Inadequate sexual education may explain why compared to healthy controls, people with Spina Bifida were less likely to use birth control when sexually active.¹⁷

There also needs to be more sexual counseling for people with Spina Bifida in order to increase their sexual satisfaction and quality of life.³ The lack of access to information about sexual health, training, and skill-building that is specific to people with Spina Bifida over their life span contributes to these knowledge gaps and suboptimal outcomes. It is important to provide people with Spina Bifida with opportunities to acquire relevant and accurate knowledge about sexual health, and to develop and implement skills to negotiate sexual desire, intimacy, and activity. Doing so can support healthy sexuality while limiting negative outcomes of sexual activity related to sexually transmitted infections, HIV transmission, unintended pregnancy, or sexual

exploitation. Sexual education and health promotion has proven to specifically benefit youth by combining education with skill-development training.¹⁸

Outcomes

Primary

1. Optimization of sexual health outcomes for people with Spina Bifida, leading to:
 - Satisfaction with sexuality and sexual relationships.
 - Knowledge of sexual health specific to Spina Bifida.

Secondary

1. Maximization of the ability of adults with Spina Bifida to participate as desired in meaningful and fulfilling sexual relationships through the provision of accurate sexual health education across the life span.

Tertiary

1. Empowerment of people with Spina Bifida to seek knowledge and skill-building regarding sexual relationships by way of the advancement of knowledge and comfort of health care professionals to provide them with sexual health education.

0-11 months

Clinical Questions

1. Is there evidence that prenatal closure impacts sexual function?
2. Is there evidence that discussing the neurologic sequelae of Spina Bifida improves parent's understanding of sexual health for their infant as they become an adult?

Guideline

1. Educate parents and caregivers about the anticipated neurologic sequelae of Spina Bifida including how sexual functioning may be impacted and that sexuality is a part of life for everyone including people with disabilities. (clinical consensus)

1-2 years 11 months

Clinical Questions

1. Should the timing of parental sexual education for children with Spina Bifida differ from that of typically-developing children?
2. Does early sexual education improve sexual health outcomes or social adjustment for children with Spina Bifida?

Guidelines

1. Educate parents and caregivers about the anticipated neurologic sequelae of Spina Bifida including how sexual functioning may be impacted. (clinical consensus)
2. Educate parents and caregivers that sexuality is a part of life for everyone including people with disabilities. (clinical consensus)
3. Provide factual information to parents and caregivers and encourage them to provide developmentally-appropriate sexual education to their children.²⁰
4. Explore the parent's expectations regarding their child's sexual development.²¹
5. Explain that sexual exploration is a normal and healthy part of early childhood development.²²
6. Explain the importance of minimizing the child's risk of sexual abuse through teaching children about their body parts, privacy, who may touch their bodies and what do to if inappropriate touching occurs.²³

3-5 years 11 months

Clinical Questions

1. What preschool activities promote healthy sexual development for children with Spina Bifida?
2. How should health care professionals promote developmentally appropriate sexual education for young children with Spina Bifida?

Guidelines

1. Provide factual information to parents and caregivers and encourage them to provide developmentally-appropriate sexual education to their children, including information about appropriate versus inappropriate touching.
2. Explore the parent's expectations regarding their child's sexual development.²⁵
3. Explain that sexual exploration is a normal and healthy part of early childhood development.²⁴
4. Underscore the goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships in the future. (clinical consensus)
5. Review relevant literature that addresses sexual health and education, such as "Bright Futures" and other reports prepared by the American Academy of Pediatrics.^{24,25,27}
6. Provide education about pubertal development, evaluate concerns or abnormal physical findings, and explain the risks of precocious puberty. (Endocrine: Puberty and Precocious Puberty)

6-12 years 11 months

Clinical Questions

1. What should be taught to children with Spina Bifida regarding sexual health?

Guidelines

1. Provide factual information to parents and caregivers and encourage them to provide developmentally-appropriate sexual education to their children.^{20,25}
2. Review relevant literature that addresses sexual health and education, such as "Bright Futures" and other reports prepared by the American Academy of Pediatrics.^{20,24-25}
3. Allow the child to ask questions about sexual development and sexuality.²⁴
4. Serve as a resource to schools²⁴ to ensure that children with Spina Bifida participate in sexual education.
5. Encourage parents to discuss information that their children are receiving about healthy relationships from school, their peers, the media, and social media.²⁴
6. Promote skill-building to identify dangerous situations, refuse or break off a sexual attack, and summon help.²⁶
7. Promote socially-appropriate behaviors and social skills.²⁵
8. Underscore the goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships in the future (clinical consensus)
9. Provide education about sexuality, pubertal development, evaluate concerns or abnormal physical findings, and explain the risks of precocious puberty (Endocrine: Puberty and Precocious Puberty Guidelines).²⁰

13-17 years 11 months

Clinical Questions

1. What should teens with Spina Bifida be taught about sexuality?

2. How can healthy relationships be promoted for teens with Spina Bifida?

Guidelines

1. Acknowledge that sexual health is an important part of life. (clinical consensus)
2. Discuss healthy relationships in gender-neutral language as the teen years are the time when many achieve self-awareness about sexual orientation.²⁴
3. Educate teens about intimate partner violence and sexual assault.²⁰
4. Discuss safe-sex practices including the use of non-latex condoms to prevent sexually transmitted infections and unwanted pregnancies.^{4,24-25}
5. Refer to a women's health provider such as a gynecologist, adolescent medicine specialist, or family medicine practitioner if the teen with Spina Bifida intends to become sexually active. Refer young men to a sexual function clinic if desired. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
6. Ensure that the Guidelines for Adolescent Preventive Services are implemented.²⁷
7. Create an environment in which the teen feels comfortable and safe discussing sexual health, including being able to speak to them alone and confidentially.²⁴
8. Educate parents by presenting them with factual information and encourage them to provide developmentally appropriate sexual education to their children.^{20,25}
9. Encourage parents to discuss information that their children are receiving about healthy relationships from school, their peers, the media, and social media.²⁴
10. Discuss sexuality routinely and openly during health care visits, and acknowledge the fluidity of sexuality and gender.²⁴
11. Allow the teen to ask questions about sexual development and sexuality.²⁴
12. Serve as a resource to schools²⁴ to ensure that children with Spina Bifida participate in sexual education.
13. Underscore the goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships in the future. (clinical consensus)
14. Provide education about pubertal development and evaluate pubertal development and any abnormal physical findings (Endocrine: Puberty and Precocious Puberty Guidelines).²⁴
15. Educate teens and parents regarding birth control options, pregnancy, genetic risk, and sexually transmitted infection risk associated with sexual activity including the use of non-latex barrier methods.^{15,24-25,28} (Men's Health Guidelines, Women's Health Guidelines)

18+ years

Clinical Questions

1. How can the ability of adults with Spina Bifida to engage in meaningful and satisfying sexual relationships be maximized?

Guidelines

1. Acknowledge that sexual health is an important part of adult life.¹
2. Take a history of sexual interest, functioning, experience and problems. (Men's Health Guidelines, Women's Health Guidelines – specifically information about fertility, reproduction, and anatomic functioning.)
3. Use factual information to educate adults about sexual health including intimate partner violence and sexual assault. (clinical consensus)
4. Provide guidance about safe sex practices including non-latex condoms to prevent sexually transmitted infections and unwanted pregnancies.^{20,24-25}
5. Refer to a women's health provider such as a gynecologist, adolescent medicine physician or family medicine practitioner. Refer men to a sexual function clinic if desired. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)

6. Educate about heritability of Spina Bifida. (Men's Health Guidelines, Women's Health Guidelines)
7. Create an environment in which the adult feels comfortable and safe discussing sexuality and sexual health routinely and openly during health care visits. (clinical consensus)
8. Refer to support groups and general audience literature regarding disability and sexuality. (clinical consensus)
9. Provide visual samples of items to facilitate discussions, such as female and male condoms, relevant websites, and other online resources. (clinical consensus)
10. Educate about the role of self-examination and routine health maintenance visits. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
11. Underscore goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships. (clinical consensus)

Research Gaps

The research gaps regarding sexual health and functioning for people with Spina Bifida are extensive. Research is warranted to study:

1. The impact of prenatal closure on sexual functioning.
2. The best methods of providing sexual education.
3. Strategies to promote sexual health and well-being.
4. Whether sexual education improves safe sex practices.
5. The barriers and factors that enhance sexual performance and satisfaction.
6. Interventions that are geared toward improving the sexual health of people with Spina Bifida.
7. The relationship of self-esteem, self-image, and self-worth with sexual health outcomes.
8. The relationship between sexual health and quality of life.

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Urology

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Introduction

The goals of urologic management and care of individuals with Spina Bifida focus on maintaining normal renal function during all ages, transitioning through stages of urinary continence, and achieving independence with personal care as aging continues through adulthood. Significant advancements in other specialties, particularly neurosurgery, have prolonged life and unmasked the importance of maintaining normal renal function and a healthy bladder. Over 90% of children at birth have a normal upper urinary tract (kidney and ureter). Historically, we know that if left unattended, 50% of those children will suffer upper urinary tract damage due to lower urinary tract (bladder and urethra) hostility.¹

During the first several years of life, the urologic focus on a child's health is based on maintaining normal kidney function at a time when the kidneys are most susceptible to kidney damage. As the child begins to approach school age, greater interest is directed toward gaining urinary continence. As a teenager, there is structured transition of care. Each of these urologic management milestones builds upon the last, potentially affecting their status in a positive or negative fashion.

Institutions create protocols based on their program's philosophy and available resources. Two general philosophies prevail: a proactive approach that attempts to identify children at risk for upper urinary tract deterioration and treat them before a problem occurs; and a reactive approach that follows a child closely and begins management at the first sign of any adverse change.²⁻⁴

Advocates of a proactive approach favor early identification of "at risk" children by assessing bladder function and managing hostile bladder parameters. This is done in an attempt to prevent adverse upper urinary tract changes and preserve normal renal function, thus limiting exposure to possible irreversible upper tract deterioration.

Institutions favoring a reactive approach rely on close evaluation of the upper urinary tract, renal function, and documentation of urinary infections. It is felt that adverse upper urinary tract changes can be detected early with minimally invasive assessment using ultrasonography. Renal function is typically assessed and followed with a serum creatinine. Adverse changes are assumed to be reversed with medical, pharmacologic, and operative management. Treating children reactively "as needed" allows for precise selective management limiting the stress and potential side effects of invasive procedures, medications, catheterization, and surgery.

The importance for maintaining normal renal function within this guideline cannot be overstated. It is also appreciated that while creatinine is a good screening tool of renal function, it is limited in the non-ambulatory child and adult with Spina Bifida with low muscle mass and thus provides a false sense of normality.⁵ Renal function may be more accurately measured with serum cystatin C or with a nuclear medicine glomerular filtration rate test (GFR).⁶ Currently, the best measure of renal function in children and adults with Spina Bifida is unknown and will require

ongoing investigation.

This guideline merges aspects of proactive and reactive philosophies based on a best practice methodology. The Centers for Disease Control and Prevention (CDC) are undertaking a prospective management protocol for newborns through age five developed by a team of pediatric specialists.⁷ It is anticipated that the outcome will positively impact the urologic care of children as well as the kidney health for individuals with Spina Bifida across the lifespan. Providing a strong foundation for pediatric care directly impacts the lifetime goals related to continence, self-management, and renal health. It is appreciated that urologic care is a dynamic, ever changing process.

Outcomes

Primary

1. Maintain normal renal function throughout the lifespan.
2. Achieve urinary continence as early as socially acceptable.
3. Maximize urologic independence.

Secondary

1. Eliminate hostile bladder dynamics through medical management.
2. Reduce or eliminate operative reconstruction of the bladder.
3. Maximize renal outcome while minimizing expense of studies, keeping watch over the timing and frequency of studies such as urodynamic testing, upper tract imaging, and lab studies.
4. Reduce impact of urinary tract infections (UTIs) and antibiotic overuse.
5. Establish a care program that allows for urologic independence, such as through clean intermittent self-catheterization (self-CIC).

Tertiary

1. Determine the best measure of renal function.
2. Minimize occurrence of urolithiasis.
3. Determine whether surgical interventions are effective in the long-term.

0-11 months

Clinical Questions

1. How do you define a symptomatic urinary tract infection and what is its long-term sequela?
2. Can diagnostic studies of the lower urinary tract (urodynamic) or upper urinary tract (ultrasonography) predict and prevent an adverse change in kidney function?
3. What is proactive management?
4. Is proactive management better than reactive to maintain normal upper tract?

Guidelines

1. Obtain the following baseline studies within three months of birth:
 - Renal/bladder ultrasound and repeat in six months
 - Urodynamic testing
 - Serum creatinine³ (clinical consensus)
2. Initiate CIC and antimuscarinic therapy for the treatment of mixed incontinence when indicated based on the above results.³ (clinical consensus)
3. Consider the presence of a UTI when there is a fever (100.4 F / 38.0 C). In neonates less than one month of age with failure to thrive and dehydration.

Define a UTI by:

4. a positive UA, and
5. a positive urine culture (UC) on a catheterized specimen, and
6. fever (100.4 F / 38.0 C).

Define a positive Urine Analysis (+ UA) as:

7. >trace nitrite or leukocyte esterase on dip UA, and
8. >10 white blood cells/high power field (WBCs/hpf), uncentrifuged specimen, or
9. >5 WBCs/hpf, centrifuged specimen.

Define a positive UC (+UC) as:

10. >50,000 colony forming units/milliliter (CFUs/mL) (sterile specimen obtained by catheter or suprapubic catheter aspirate).
11. >100,000 CFUs/mL in a clean voided specimen.⁸

1-2 years 11 months

Clinical Questions

1. How can providers account for neurologic bladder changes due to growth and/or tethering?
2. What diagnostic tools are reliable to assess renal function?
3. Are upper tract changes reversible once they occur?
4. How should symptomatic UTIs be defined? What is the sequela of symptomatic UTIs? What is the optimal upper and lower urinary tract surveillance?
5. Does the use of proactive CIC and antimuscarinic medication help to maintain a normal upper tract?

Guidelines

1. Obtain renal/bladder ultrasound every six months when the child is under the age of two. After that, obtain an ultrasound yearly if the child is stable, without UTIs or imaging changes. (clinical consensus)
2. Obtain a renal/bladder ultrasound, as needed if the child has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing yearly through age three. Repeat as needed if the following are noted:^{1,2,7} (clinical consensus)
 - bladder hostility
 - upper urinary tract changes
 - recurrent symptomatic UTIs
4. Obtain a serum creatinine test if there is a change in the upper urinary tract. (clinical consensus)
5. Assess suspected UTIs with a urine specimen obtained by sterile catheterization technique. Repeat a positive bag urine specimen with a catheterized specimen. (clinical consensus)

Define a UTI by:

- a positive UA, and
- a positive urine culture (UC) on a catheterized specimen, and
- fever (100.4 F / 38.0 C).

Define a positive Urine Analysis (+ UA) as:

- >trace nitrite or leukocyte esterase on dip UA, and
- >10 white blood cells/high power field (WBCs/hpf), uncentrifuged specimen, or
- >5 WBCs/hpf, centrifuged specimen.

Define a positive UC (+UC) as:

- >50,000 colony forming units/milliliter (CFUs/mL) (sterile specimen obtained by catheter or suprapubic aspirate).
- >100,000 CFUs/mL in a clean voided specimen. ⁸

6. Initiate CIC and antimuscarinic therapy for the treatment of mixed incontinence when indicated by upper urinary tract changes, recurrent symptomatic UTIs, or bladder hostility noted on urodynamic testing.²⁻⁴ (clinical consensus)

3-5 years 11 months

Clinical Questions

1. How can providers account for neurologic bladder changes due to growth and/or tethering?
2. What diagnostic tools are reliable to assess renal function?
3. Are upper tract changes reversible once they occur?
4. How should symptomatic UTIs be defined? What is the sequela of symptomatic UTIs? What is the optimal upper and lower urinary tract surveillance?
5. Does the use of proactive CIC and antimuscarinic medication help to maintain a normal upper tract?
6. Are the caregivers compliant with CIC? Who is performing CIC – the caregivers and/or the child?

Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed, if the child has recurrent symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing only if the following are present: (clinical consensus)
 - upper tract changes
 - recurring UTIs
 - interest in beginning a urinary continence program
4. If the child is on CIC, begin to involve the child in the process of self-catheterization. ⁹ (clinical consensus) (Self-Management and Independence Guidelines)
5. Obtain a serum creatinine test if there is a change in imaging of the upper urinary tract. (clinical consensus)
6. Obtain serum chemistries (includes serum creatinine) at age 5. Assess suspected UTIs with a catheterized urine specimen. Repeat a positive bag urine specimen with a catheterized specimen. (clinical consensus)

Define a UTI by:

- a positive UA, and
- a positive urine culture (UC) on a catheterized specimen, and
- leakage between CIC, and
- onset of pelvic or back pain, and
- fever (100.4 F / 38.0 C).

Define a positive UA (+ UA) as:

- >trace nitrite or leukocyte esterase on dip UA, and
- >10 white blood cells/high power field (WBCs/hpf), uncentrifuged specimen, or
- >5 WBCs/hpf, centrifuged specimen.

Define a positive UC (+UC) as:

- >50,000 colony forming units/milliliter (CFUs/mL) (sterile specimen obtained by catheter or suprapubic aspirate).
 - >100,000 CFUs/mL in a clean voided specimen.⁸
7. Initiate CIC and antimuscarinic therapy when indicated by upper urinary tract changes, recurring symptomatic UTIs, or bladder hostility noted on urodynamic testing.²⁻⁴ (clinical consensus)
 8. Introduce urinary continence and discuss interest in beginning the program and options at each visit.⁹⁻¹⁰ (clinical consensus) (Self-Management and Independence Guidelines)
 9. Introduce bowel management and discuss interest and options at each visit. (clinical consensus) (Bowel Function and Care Guidelines)

6-12 years 11 months

Clinical Questions

1. What is the best way to measure renal function in the child that is non-ambulatory?
2. What social, environmental, and economic limitations or hurdles are encountered when working to achieve urinary continence?
3. What is worse: stool or urinary incontinence?
4. How we define urologic continence? Is the definition of continence congruent with the perspective of the patient, family, and physician?

Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed if the child has recurrent symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing when initiating a urinary continence program, if the following are present: (clinical consensus)
 - upper urinary tract changes such as hydronephrosis or renal scarring
 - recurring symptomatic UTIs
 - changes in urinary continence status
4. Obtain a serum creatinine test yearly. If the child has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus)
5. Obtain serum chemistries yearly on any child who has had urinary reconstruction.
6. Obtain a serum B12 level test every year beginning two years after urinary reconstruction.¹¹⁻¹³ (clinical consensus)
7. Discuss a urinary continence program and interest in beginning the program and options at each visit.⁹⁻¹⁰ (clinical consensus) (Self-Management and Independence Guidelines)
8. Discuss a bowel management program and the interest and options at each visit. (clinical consensus) (Bowel Function and Care Guidelines)

13-17 years 11 months

Clinical Questions

1. How is continence affected by a shift in responsibility to self-care?
2. How is a normal upper urinary tract affected by a shift in responsibility to self-care?
3. What is optimal surveillance of the upper and lower urinary tract?
4. If reconstructive continent bladder surgery was undertaken, would you do it again?
5. If no reconstructive surgery was undertaken do you wish it had been?

Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed, if the child has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain a serum creatinine test yearly. If the child has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus)
4. Obtain serum chemistries including B12 yearly on any child who has had urinary reconstruction.¹¹⁻¹³ (clinical consensus)
5. Transition urologic care to self-management, if doing so is developmentally appropriate for the child.¹⁴⁻¹⁵ (clinical consensus) (Self-Management and Independence Guidelines)
6. Transition bowel program to self-management, if doing so is developmentally appropriate for the child. (clinical consensus) (Bowel Function and Care Guidelines)

18+ years

Clinical Questions

1. What is optimal surveillance of the upper and lower urinary tract? What cancer screening is needed?
2. How do we define UTI in the adult and when do we treat?
3. How do we minimize sequelae of secondary incontinence in adulthood?

Guidelines

1. Obtain a renal/bladder ultrasound yearly. (clinical consensus)
2. Obtain a renal/bladder ultrasound, as needed if the adult has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain a serum creatinine test yearly. If the adult has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus) (Self-Management and Independence Guidelines)
4. Obtain serum chemistries including B12 on anyone who has had urinary reconstruction.¹¹⁻¹³ (clinical consensus)
5. Undertake cystoscopy and appropriate upper tract imaging in adults who have had a bladder augmentation when the following are present:¹⁶⁻¹⁸ (clinical consensus)
 - clinically-noted change in upper or lower urinary tract status
 - gross hematuria
 - recurrent symptomatic UTIs
 - increasing incontinence
 - pelvic pain
 - the adult has had a renal transplant with the presence of BK/polyomavirus
6. Evaluate patterns of continence/incontinence and address issues collaboratively with the individual and family. Include assessment of amount (volume) of incontinence as the amount in adults may be more bothersome than frequency.¹⁹
7. Continue to support self-management and independent living. (Self-Management and Independence Guidelines)

Research Gaps

Proactive treatment: The foundation of management is based on the ability to predict individuals at risk for kidney deterioration and then influence management prior to an adverse event.

1. What is the ability of urodynamic testing to identify individuals at risk?

2. Does early medical (e.g. intermittent catheterization) and pharmacologic (antimuscarinic) management based on urodynamic testing prevent upper tract deterioration?

Renal Function: Renal function is assessed through serum studies and imaging. However, it is not known what the best assessment is in the population with Spina Bifida.

1. How is creatinine influenced by height, weight and mobility status of a patient with Spina Bifida?
2. Is cystatin C a more accurate indicator of renal function in the population with Spina Bifida?
3. What degree of renal dysfunction has occurred by the time changes are noted on imaging (i.e., renal scarring in ultrasonography or DMSA)?
4. Are changes on imaging reversible?
5. Is yearly serum and upper tract testing necessary?

Urinary Infections: Chronic bacteriuria is suspected to have less of an impact on adverse health and renal deterioration than symptomatic UTIs.

1. What is the definition of a symptomatic UTI?
2. Does the definition of symptomatic UTI change with aging?
3. Do symptomatic UTIs in children under the age of five have greater morbidity?

Continence: Continence of the bowel and bladder plays an important role in socialization. The following only relates to urinary continence. Continence from a medical perspective is absolute (i.e. dry or wet).

1. Does the medical definition of absolute continence translate into a patient's and their family's quality of life?
2. Is the perception of continence from the perspective of the medical care provider and patient and family congruent?
3. Does achieving "some" degree of continence become beneficial?
4. Is there a threshold of "social" continence that is critical?
5. Is the cost (e.g. change in patient and family lifestyle, need for increased supervision, risk of intermittent catheterization, risk of medicines, and both short- and long-term surgical risk) worth the benefit?
6. What are the long-term challenges of patients who have undergone surgical intervention?
7. Would patients who have chosen surgery as a management option, make the same decision if they had the opportunity?

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Women's Health

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Introduction

Women and girls with Spina Bifida have specific needs and concerns, especially in the context of sexuality, pregnancy, and childbirth. Every woman's and girl's body undergoes multiple changes throughout her lifetime, and these changes are further impacted by Spina Bifida.

More information is still needed regarding sexual response in women with Spina Bifida. Many factors may affect a woman's ability to position herself during sexual activity, including level of mobility, history of osteoporosis and fracture risk, and respiratory function.¹ Innervation also impacts the body's physiologic response to stimulation including vaginal engorgement, lubrication and the ability to achieve orgasm.

Women with Spina Bifida may have unique health concerns regarding their reproductive health, such as structural anomalies of the reproductive tract (such as a bicornuate uterus which may be found on exam or ultrasound). Changes to the hips and spine may require special attention to positioning during pelvic examinations and birth.¹ Many women with physical disabilities, including those with Spina Bifida, choose to become pregnant,²⁻³ and with pregnancy come specific concerns, such as preterm birth and changes in a woman's bowel, bladder and mobility. Understanding how pregnancy will affect a woman with Spina Bifida, and how her current state of health and quality of life might be affected by pregnancy is critical to her ongoing health. Menopause can cause vasomotor symptoms and changes in vaginal and bladder health, and women may benefit from both lifestyle and medical management.⁴

This guideline aims to address many of the main health concerns specific to women with Spina Bifida.

Outcomes

Primary

1. Provide accurate information to women with Spina Bifida about the impact of Spina Bifida on pregnancy and the impact of pregnancy on women with Spina Bifida.

Secondary

1. Help women with Spina Bifida maximize sexual functioning.

Tertiary

1. Understand menopause management options for women with Spina Bifida.

Women's Health Guidelines begin at age 6-12 years 11 months

6-12 years 11 months

Clinical Question

1. When do pubertal changes happen to girls with Spina Bifida?

Guidelines

1. Puberty occurs earlier in girls with Spina Bifida than in the general population. It is recommended that, along with Tanner staging, care providers discuss the possibility

- of early puberty with girls and their families and create an atmosphere of open communication.¹ (Endocrine: Puberty and Precocious Puberty Guidelines, Sexual Health and Education Guidelines)
2. Offer human papillomavirus (HPV) vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, if appropriate.⁵⁻⁶

13-17 years 11 months

Clinical Questions

1. How can pelvic organ prolapse be managed?
2. What are the sexual education needs of girls with Spina Bifida?
3. What information and screening do girls need regarding contraception and sexually transmitted infections?

Guideline

1. Manage pelvic organ prolapse, which can occur at any stage of life in women with Spina Bifida, in consultation with an urogynecologist. Take into account the possibility of decreased pelvic sensation.⁷
2. Provide guidance on reproduction, sexual health and education. (Sexual Health and Education Guidelines)
3. Contraception options should be made available and discussed to in a non-judgmental manner, taking into account health concerns such as decreased mobility, risk of decreased bone mineral density, latex allergy and use of antiepileptic medications and genetic risk factors.^{8,9} (clinical consensus) Consider consulting a gynecologist in a complex scenario.
4. Offer HPV vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, if appropriate.⁵⁻⁶

18+ years

Clinical Questions

1. How can we manage pelvic organ prolapse in adults with Spina Bifida?
2. What gynecological care should women with Spina Bifida have?
3. How can we maximize the physical sexual functioning of women with Spina Bifida, including orgasm and lubrication?
4. What is the impact of pregnancy on the global physiology of a woman with Spina Bifida?

Guidelines

1. Manage pelvic organ prolapse, which can occur at any stage of life in women with Spina Bifida, in consultation with an urogynecologist. Take into account the possibility of decreased pelvic sensation.⁷
2. Encourage women with Spina Bifida to have routine gynecological care, including Pap smears and mammograms.¹⁰⁻¹¹
3. Provide guidance on sexual health, education and birth control. (Sexual Health and Education Guidelines) Contraception options should be made available and discussed in a non-judgmental manner, taking into account health concerns such as decreased mobility, risk of decreased bone mineral density, latex allergy and use of antiepileptic medications and genetic risk factors.^{8,10} (clinical consensus) Consider consulting a gynecologist in a complex scenario.
4. Raise awareness of availability of screening and treatment for sexually transmitted infections, and offered Pap smears as per guidelines.¹⁰
5. Provide gynecology exam rooms and tables that are accessible for women with physical disabilities. (clinical consensus)

6. Clinicians should initiate a discussion of sexual function with women with Spina Bifida in a sensitive manner to facilitate problem problem-solving and acknowledge common concerns such as inability to orgasm, prolonged time to achieve an orgasm, and decreased lubrication. In some cases, an experienced sexologist may be helpful.¹²
7. Encourage women with decreased pelvic sensation to explore other parts of the body with themselves or a partner, especially the lips, nipples, arms, and earlobes, or other areas of the skin, as they may find them to be more sensitive.¹³
8. Inform women that they can use commercially available sexual lubricants to improve lubrication.¹³
9. Women with urinary incontinence should be encouraged to catheterize or void before having sex to prevent incontinence during sex.¹³
10. Inform women that their choice of sexual positions may need to take into account their level of mobility, osteoporosis and fracture risk, and respiratory function.¹
11. Motivate women with Spina Bifida to communicate with their sexual partners about what they enjoy and do not enjoy during sex.¹³
12. Recommend preconception consultation with an obstetrician who specializes in high-risk pregnancies. Depending on the woman's medical history, she may also benefit from preconception consultation with her neurosurgeon, urologist, physiatrists, and other health care providers to discuss the potential impact of pregnancy on health. (clinical consensus) (Prenatal Counseling Guidelines)
13. Recommend that women with Spina Bifida, who are at increased risk of having a baby with a neural tube defect, decrease their risk by taking a daily oral supplement of 4mg of folic acid starting at least 1 month but preferably 3 months prior to conception and continuing until 12 weeks of gestational age.^{14,15} (Prenatal Counseling Guidelines)

Targeted Guidelines: Pregnancy

General Considerations

Clinical Question

1. What is the impact of Spina Bifida on pregnancy?

Guidelines

1. Discuss the increased risk of preterm birth and review the signs and symptoms of preterm birth in the context of the woman's sensory abilities.¹⁶
2. Inform women that maternal Spina Bifida increases a woman's chance of having a baby with Spina Bifida. (Prenatal Counseling Guidelines)

Musculoskeletal and Respiratory Considerations during Pregnancy

Clinical Questions

1. How should respiratory function be monitored during pregnancy?
2. How should changes in mobility during pregnancy be managed?

Guidelines

1. Conduct pulmonary function testing at least once during pregnancy in the case of kyphoscoliosis. This is because dyspnea can occur during pregnancy when there is an associated kyphoscoliosis deformity. (clinical consensus)
2. Ask about symptoms of shortness of breath at each antenatal visit, and undertake pulmonary function testing or assess for pulmonary embolism as indicated.^{17,19}

3. Consider temporary wheelchair use in women and girls who use braces and crutches to ambulate, to reduce the risk of falls and subsequently trauma to maternal joints and the fetus.¹⁸
4. Follow for back and leg pain and consider temporary wheelchair use (clinical consensus), modified bedrest, and massage and physical therapy if back and leg pain is severe.²⁰
5. Consider referral to orthopaedics and physical medicine and rehabilitation as needed when there are significant or concerning changes in mobility. (clinical consensus)
6. Consider referral to occupational therapy and physical therapy early in pregnancy to discuss the impact of pregnancy on self-management ability as well as to discuss plans for after-delivery care and care of her baby.²¹⁻²² (Self-Management and Independence Guidelines)

Bowel Care Considerations during Pregnancy

Clinical Questions

1. How should bowel concerns be managed during pregnancy?

Guidelines

1. Discuss bowel care early in the pregnancy, as pregnancy can worsen constipation. A diet high in fibre, increased fluid intake and exercise can alleviate constipation, however are not always sufficient. Bulk-forming agents such as psyllium, stool softeners such as docusate sodium, lubricant laxatives, osmotic laxatives and stimulant laxatives are considered safe in pregnancy. However, osmotic and stimulant laxatives may cause significant abdominal cramping and bloating and therefore should not be used for a prolonged period of time.²³
2. Consider having a consultation with a gastroenterologist or expert in neurogenic bowel management to maximize the methods to alleviate constipation. (clinical consensus)
3. Manage a suspected bowel obstruction with a team consisting of a general surgeon, neurosurgeon, and high-risk obstetrician. (clinical consensus)

Shunt Management Considerations during Pregnancy

Clinical Questions

1. How can shunt complications be assessed during pregnancy and how should they be managed?

Guidelines

1. Review signs of increased pressure, headache, nausea, and vomiting at each prenatal visit because the enlarging uterus can cause a shunt malfunction by increasing intra-abdominal pressure.²⁴
2. Manage signs of shunt malfunction with a team consisting of a neurosurgeon, obstetrician and anesthesiologist. Other specialties may be needed depending on the clinical scenario.²⁴
3. Conduct a thorough workup for both preeclampsia and shunt obstruction if a pregnant woman with a shunt has nausea, vomiting, headache, or neurological symptoms. A preeclampsia workup consists of assessing for the following: fetal well-being; blood pressure; proteinuria; and blood work to test for elevated aspartate aminotransferase (AST), and alanine transaminase (ALT), and thrombocytopenia.^{24,25}

Seizure Considerations during Pregnancy

Clinical Question

1. How should seizure risk be managed during pregnancy?

Guideline

1. Optimize medical management of seizures prior to conception. Women who have a history of seizures have a higher risk of seizure during pregnancy and labor. If possible, avoid anticonvulsant medications that have a greater risk of teratogenicity while still providing good control.¹

Bladder and Renal Function Considerations during Pregnancy

Clinical Question

1. What considerations should be made for bladder and kidney health during pregnancy?

Guidelines

1. Perform regular urinalysis and urine culture tests throughout the pregnancy and treat infections promptly, as urinary tract infections are common during pregnancy in mothers with Spina Bifida.^{19,26-27}
2. Make a baseline renal assessment, ideally prior to pregnancy or early in the pregnancy, in order to make appropriate referrals to nephrology care.²⁸
3. Coordinate with a nephrologist to manage women with Spina Bifida who already have evidence of renal disease and a risk of decreased renal function in pregnancy.²⁸⁻²⁹
4. Perform intensified maternal and fetal monitoring with women who have renal disease in pregnancy and are at increased risk of preeclampsia and intrauterine growth restriction.^{25,27}
5. Ask women at each visit about their ability to catheterize, and refer them to a urologist if there are concerns because urostomies can develop poor conduit drainage as the uterus grows.^{27,30}
6. Urgently consult with urology specialists if women with continent urinary diversions develop increased incontinence or difficulties in intermittent self-catheterization.³⁰

Targeted Considerations: Childbirth

General Considerations during Childbirth

Clinical Question

1. What are considerations for birth for a woman with Spina Bifida?

Guidelines

1. Consult a high-risk obstetrician when planning the mode of delivery. Although vaginal births are possible for women with Spina Bifida, severe spinal and pelvic skeletal deformities may prevent vaginal birth.¹⁷
2. Consider facilitating vaginal deliveries in women with ventriculoperitoneal (VP) shunts by means of a shortened pushing stage, possibly aided by a vacuum or forceps to decrease elevation of intracranial pressure.³¹
3. Teach women who may be unaware of labor contractions to palpate for hardening of the belly and observe for rupture of membranes, and watch for signs of autonomic dysreflexia.^{19,32}
4. Watch for autonomic dysreflexia triggered by labor among women who have a lesion above T6. Autonomic dysreflexia can be life-threatening and women experiencing any signs or symptoms should seek emergency care and transportation to the hospital. As well, there is significant clinical overlap between autonomic dysreflexia and preeclampsia, and therefore the woman should be evaluated for both.^{19,32}

5. Make the decision between a planned caesarean birth (with available urology back-up if needed) versus a planned trial of a vaginal birth (with the associated risks of having an emergency caesarean birth) in conjunction with a team consisting of an anesthesiologist, urologist, and obstetrician, and acknowledge the woman's goals and preferences. Keep in mind that a caesarean birth in a woman with previous lower urinary tract surgery may be complex. Intestinal and omental adhesions to the lower uterine segment may necessitate a classic upper segment section.³⁰
6. Recommend a caesarean birth to protect continence for women with vesical neck reconstruction or artificial sphincter placement.²⁷
7. Take into account that pregnancy itself can exacerbate an existing pelvic organ prolapse and that a vaginal birth will likely exacerbate it. Consider the impact of a worsening pelvic organ prolapse, and the possible need for subsequent surgery, in consultation with an obstetrician and urogynecologist, and taking into account the woman's preferences. The plan for the mode of birth should take into account the impact of this worsening and the possible need for subsequent surgery in consultation with an obstetrician and urogynecologist, and acknowledge the woman's preferences.²⁷
8. Consider that Spina Bifida can be associated with congenital renal malformations such as horseshoe kidney and pelvic kidney.³³ If a caesarean birth is required, the surgeon should be aware of unique renal anatomy prior to conducting the surgery if needed.
9. Ensure that a consultant urologist be available for the caesarean birth in women who have had a previous lower urinary tract surgery.³⁴
10. Keep in mind that Spina Bifida is not a contraindication to epidural anesthesia. As such, ensure that each woman has an anesthesia consultation prior to delivery to discuss the risks and benefits of regional versus general anesthesia.³⁶⁻³⁷

Targeted Considerations: Breastfeeding

General Considerations for Breastfeeding

Clinical Question

1. Is breastfeeding impacted by Spina Bifida? If so, how?

Guidelines

1. Encourage mothers who wish to breastfeed to do so and provide them with support from a lactation consultant. Keep in mind that there is no literature specifically about breastfeeding in the context of Spina Bifida. (clinical consensus) (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
2. Be aware that while anti-epileptic medications are for the most part considered compatible with breastfeeding, some require close monitoring of the baby for side effects and a reduction in the baby's exposure. Consider informing mothers of any possible side effects associated with an anti-epileptic medication they are taking while breastfeeding.³⁸

Targeted Considerations: Menopause

General Considerations during Menopause

Clinical Questions

1. How should vasomotor symptoms of menopause be managed by women with Spina Bifida?
2. How should the urogenital changes of menopause be managed by women with Spina Bifida?
3. How should women be screened for breast and gynecological cancers?

Guidelines

1. Inform women that vasomotor symptoms such as hot flashes can sometimes be managed by lifestyle changes such as avoiding alcohol, cigarette smoking and warm drinks, as well as maintaining a normal body mass index.⁴
2. Take into account that medical management of vasomotor symptoms includes both hormonal and non-hormonal prescription medication. Decisions on which medication to take should be made in conjunction with a physician experienced in managing menopausal symptoms, and take into consideration the severity of the woman's symptoms, bone mineral density, risk for blood clots, and behavioral or emotional symptoms such as depression.⁴
3. Inform women with vaginal dryness that they may benefit from topical vaginal lubricants.⁴
4. Consider treating vaginal atrophy with vaginal estrogen by a physician experienced in managing the symptoms of menopause. This may also help with urinary urge incontinence and may prevent some urinary tract infections.⁴
5. Women should be made aware of their breast anatomy, and should be encouraged to bring any changes to the attention of their physician.³⁸
6. Women should participate in breast cancer screening programs, which for many women will begin at age 45. This may be initiated sooner if there is a family history or other risk factors for breast cancers.³⁹
7. Women should continue to participate in cervical cancer screening programs in accordance with local guidelines.¹⁰
8. Women should be made aware that abnormal vaginal perimenopausal bleeding and post-menopausal bleeding can be a sign of endometrial cancer. Strongly encourage women to tell their physician if they experience abnormal perimenopausal bleeding or any spotting or bleeding after menopause.⁴⁰

Targeted Considerations: Deep Vein Thrombosis

General Considerations for Deep Vein Thrombosis

Clinical Question

1. When do girls and women with Spina Bifida need thromboprophylaxis?

Guideline

1. Consider thromboprophylaxis on a case-by-case basis for girls and women with limited mobility and those who use wheelchairs. Girls and women with decreased mobility may have an increased risk of deep vein thrombosis and pulmonary embolism in pregnancy. Girls and women with thrombophilia; BMI>30; those who smoke; those with pelvic girdle pain that restricts mobility; those undergoing caesarean section or prolonged labor; those with preeclampsia; and those with a preterm birth are at further risk. Consider consultation with hematology to assist with risk assessment and thromboprophylaxis duration.⁴¹

Research Gaps

1. There is no information available on the effect of pregnancy on continence.
2. There is no literature specifically on breastfeeding in the context of Spina Bifida.
3. More research is required to understand the incidence and cause of preterm birth in women with Spina Bifida.
4. More research is required to understand the incidence of gestational diabetes, preeclampsia, malpresentation and postpartum complications in women with Spina Bifida.
5. There is limited information on maximizing the physical sexual functioning of women with Spina Bifida.
6. There is limited information on the management of menopause symptoms specifically for women with Spina Bifida.

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Bowel Function and Care

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Introduction

Managing bowel function can be one of the biggest and most important challenges accompanying the diagnosis of Spina Bifida. Spina Bifida results in the loss of normal motor and sensory control in the gastrointestinal tract and anorectal dysfunction. This is defined as a neurogenic bowel dysfunction (NBD). Nervous system lesions above the conus medullaris result in upper motor NBD leading to failure to evacuate the bowel, resulting in constipation or impaction. Lesions at or below the level of the conus medullaris result in lower motor NBD, resulting in failure to contain stool and thus fecal incontinence. Attention to these pathophysiologic causes of neurogenic bowel will lead to a better understanding, better program adherence, and increase the probability of attaining the goals of continence without constipation.^{3,10,24}

In the first 10 clinics in the National Spina Bifida Patient Registry (NSBPR), it was found that less than 30% were continent of stool. Further defining the problem, select NSBPR clinics identified males, non-Hispanic blacks, those with higher lesion levels, and those with public insurance were less likely to have bowel continence. The secondary complications from NBD in Spina Bifida extend beyond constipation and incontinence as these contribute to urinary incontinence, urinary tract infections, shunt malfunction, potential for skin breakdown, hemorrhoids, anal fissures, loss of social and work opportunities, and decreased quality of life.^{4,5,11,17-18,26}

Proactive, systematic, and rational approaches can lead to continence and a more functional lifestyle.^{7,12-24} The following step-by-step guidelines were developed to emphasize management leading to the specific goal of bowel continence without constipation. The guidelines should be considered from least invasive to most invasive.

Tailoring to the individual, considering upper or lower motor bowel dysfunction, is important in the success of the bowel program.^{7,24} In working with school-age children, consider use of school staff to aid in tracking. The school nurse plays a vital role in assisting the child to reach educational goals and manage health concerns.⁹ These guidelines should be followed with the guidance of a health care professional with expertise in bowel management in Spina Bifida.

Outcomes

Primary

1. Maintenance of social continence as appropriate for age level.

Secondary

1. Maximization of independence with managing bowel program.
2. Maximized knowledge and compliance with diet and bowel program.

Tertiary

1. Minimization of constipation.

0-11 months

Clinical Questions

1. What evidence exists that prevention of constipation in the first year of life improves the outcome of bowel management in later childhood?

Guidelines

1. Monitor stool frequency, consistency, and amounts.^{7,17}
2. Use dietary management, in particular breastfeeding if possible, as it is easier to digest and offers better restoration of the microbiome after surgery.²⁵ (clinical consensus)
3. Consider dietary management (fiber and fluids) before pharmacologic adjuncts (sennoside), and/or rectal stimulants (glycerin suppositories) to manage constipation.^{7,24,26}
4. Use barrier creams to protect perineal area from breakdown as needed.² (Integument (Skin) Guidelines).

1-2 years 11 months

Clinical Questions

1. Is there evidence to support the benefit of toilet training at the same developmental stage as peers without dysfunction?

Guidelines

1. Discuss toilet training and habit training with parents.^{7,17,24,26}
2. Establish goal of working toward bowel continence.^{7,17,24,26}
3. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
4. Consider two-pronged approach of oral and rectal interventions to meet the goal of bowel continence without constipation.^{3,6-7,14,24,26}
5. Use dietary management (fiber and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation and fecal incontinence.^{3,6-7,14,24,26}
6. Use barrier creams to protect perineal area from breakdown as needed.²
7. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. Is there evidence that “habit training,” or forced evacuation with stimulants such as suppositories or enemas, increases social continence?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary tract infections (UTIs), skin breakdown, social isolation.^{4,7,11,18-20,24,26})
2. Establish the goal of bowel continence and institute the bowel continence program using guidelines below.^{6-7,13,17, 24,26}
3. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
4. Consider two-pronged approach of oral and rectal interventions to meet the goal of bowel continence without constipation or fecal incontinence.^{3,6-7,14,24,26}
5. Use dietary management (fiber and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation and fecal incontinence.^{3,6-7,14,24,26}

6. Use barrier creams to protect perineal area from breakdown as needed.²
7. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

6-12 years 11 months

Clinical Questions

1. What is the evidence that the Malone Antegrade Continence Enema (MACE) procedure or continent cecostomy is an effective form of bowel management in children with refractory incontinence?
2. What are the most effective protocols for MACE?
3. What is the evidence that electrical stimulation (trans-rectal or intravesicular) provides benefit for increased bowel continence?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation)^{4,7,11,18-20,24,26} and focus on developing independent management skills. (Self-Management and Independence Guidelines)
2. Establish the goal of bowel continence and institute the bowel continence program using the guidelines below.^{7,17,24,26}
3. Assist the child with learning how to minimize and manage bowel accidents.^{9,26}
4. Use barrier creams to protect perineal area from breakdown as needed.²
5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.^{7,9,17,24,26}
6. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
7. Consider twofold attack of oral and rectal interventions to meet the goal of bowel continence without constipation or fecal incontinence.^{3,6-7,14,24,26}
8. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.^{3,6-7,14,24,26}
9. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or MACE.^{1,3,5,14-15,21,24,26}
10. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

13-17 years 11 months

Clinical Questions

1. What support is needed by teens with Spina Bifida to be successful in maintaining their bowel program?
2. Is there evidence that hormonal fluctuations impact continence?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation)^{4,7,11,18-19,20,24,26} and focus on developing independent management skills. (Self-Management and Independence Guidelines)
2. Establish or maintain the goal of bowel continence and institute or maintain the bowel continence program using the guidelines below.^{7,17,24,26}
3. Assist the child with learning how to minimize and manage bowel accidents.^{7,9-10,17,24,26}
4. Use barrier creams to protect perineal area from breakdown as needed.²

5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.^{7,9,17,24,26}
6. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
7. Consider a twofold attack of oral and rectal interventions to meet the goal of bowel continence without constipation.^{3,6-7,14,24,26}
8. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.^{3,6-7,14,24,26}
9. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or antegrade continence enema (Malone).^{1,3,6,14-15,21,24,26}
10. Refer to Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)
11. Access support services for personal care, if needed.^{9,12,26}

18 + years

Clinical Questions

1. What impact does pregnancy have on bowel management or on use of a cecostomy or MACE? (Women's Health Guidelines)
2. Does early chronic constipation impact management of constipation in adult years?
3. Is there a change in bowel function later in life that should be addressed with a more aggressive bowel program? Does menopause result in changes?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation).^{4,7,11,18-19,20,24,26}
2. Establish the goal of bowel continence and institute the bowel continence program using timing, suppositories, pharmacologic agents or enemas as needed.^{7,17,24,26}
3. Assist the adult with learning how to minimize and manage bowel accidents.^{7,9-10,17,24,26}
4. Use barrier creams to protect perineal area from breakdown as needed.²
5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.^{7,9,17,24,26}
6. Discuss management of bowel program as it may impact sexual relations.^{4,11,26}
7. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
8. Consider a twofold attack of oral and rectal interventions to meet goal of bowel continence without constipation or fecal incontinence.^{3,6-7,14,24,26}
9. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol, lubiprostone, or other prescription), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.^{3,6-7,14,24,26}
10. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or MACE.^{1,3,6,14-15,21,24,26}
11. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)
12. Access support services for personal care if needed.^{9,12,26}

Research Gaps

1. Are there benefits to probiotic use in the population with Spina Bifida?

2. What is the evidence that electrical stimulation (trans-rectal or intravesicular) provides benefit for bowel continence in Spina Bifida?
3. Does an individualized, stepped approach to bowel management in patients with Spina Bifida lead to less constipation and incontinence?
4. What factors contribute to a successful bowel program in the population with Spina Bifida?
5. What challenges do pregnancy or menopause create for an established bowel program?
6. What are the relative efficacies of interventions for management of incontinence and constipation in the setting of neurogenic bowel in Spina Bifida by age group?
7. What evidence exists that prevention of constipation in the first year of life improves the outcome of bowel management in later childhood?
8. Is there evidence to support the benefit of toilet training a child with Spina Bifida at the same developmental stage as peers without dysfunction?
9. Is there evidence that "habit training," or forced evacuation with stimulants such as suppositories or enemas, increases social continence?
10. What is the evidence that the MACE procedure or continent cecostomy is an effective form of bowel management in children with refractory incontinence?
11. What are the most effective protocols for cecostomies?
12. What support is needed by teens with Spina Bifida to be successful in maintaining their bowel program?
13. Is there evidence that hormonal fluctuations impact continence?
14. Does early chronic constipation impact management of constipation in adult years?
15. Is there a change in bowel function later in life that should be addressed with a more aggressive bowel program?

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Endocrine: Puberty and Precocious Puberty

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Introduction

Puberty is defined as the presence of secondary sexual characteristics. For girls, it involves breast development and pubic hair, and for boys, pubic hair, enlargement of the penis and testicles. The onset of breast and pubic hair development for girls and penile enlargement and pubic hair in boys is often a concern to parents and caregivers. The question parents often ask is if the changes are a normal variant or need medical evaluation. During the health supervision visit, especially starting at age 7, breasts and genitalia should be examined using Tanner Staging. The Tanner Staging results should then be compared to societal norms for sexual maturity ratings.^{3,4} In general, precocious puberty is defined as the onset of puberty before the age of 8 years in girls and 9 years in boys.¹

Children with Spina Bifida and hydrocephalus are at an especially higher risk of precocious puberty compared to typically-developing children, most likely because hydrocephalus removes inhibitory signals that allow the hypothalamus and pituitary gland to signal the testes and ovaries to release testosterone and estradiol, respectively.² If a child enters puberty early there could be issues with actual versus expected height, and psychological and psychosocial problems. Although parents and caregivers are often the ones to bring the assessment of puberty to the attention of medical providers, examining the breasts and genitalia should be a routine part of the health supervision visit. If noted, the signs of early onset of puberty should be discussed with parents or caregivers and patient referrals should be provided to a pediatric endocrinologist if concerns arise.

Outcomes

Primary

1. Timely assessment of the onset of puberty, identification and counseling for normal variants of puberty, and appropriate referral and management of precocious puberty.

Secondary

1. Decrease risk of unwanted consequences of precocious puberty among children with Spina Bifida.

0-11 months

Clinical Questions

1. How often should weight and length be measured in ages 0-11 months?

Guidelines

1. Monitor and document weight and length closely at every health supervision visit. Length should be measured with a length measuring board.² (clinical consensus)
2. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by a provider of the same sex if the child and/or family is more comfortable with a same-sex provider.² (clinical consensus)
3. Document all positive and negative findings of the physical exam. (clinical consensus)
4. Discuss the outcomes of the evaluation with the parents or caregivers and ask them if they have any concerns. (clinical consensus)

5. Refer the child to a pediatric endocrinologist if abnormal signs of puberty are observed. (clinical consensus)

1-2 years 11 months

Clinical Question

1. What effect does hydrocephalus have on early onset of puberty?

Guidelines

1. Monitor and document weight and height velocity closely at every health supervision visit. Length should be measured using a length measuring board.² (clinical consensus)
2. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by a provider of the same sex if the child and/or family is more comfortable with a same-sex provider.² (clinical consensus)
3. Document all positive and negative findings of the physical exam. (clinical consensus)
4. Discuss the outcomes of the evaluation with the parents or caregivers and ask them if they have any concerns. (clinical consensus)
5. Refer the child to a pediatric endocrinologist if abnormal signs of puberty are observed. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. What is the best practice for detection and management of early puberty?
2. Should every child with early signs of puberty be referred to an endocrinologist?

Guidelines

1. Monitor and document weight and height velocity closely at every health supervision visit. Height should be measured (if possible) using a stadiometer. Often there may be difficulty assessing height due to inability to stand, scoliosis or contractures. In these cases, arm span or another appropriate parameter may be used. Care should be taken to use the same parameter at subsequent visits.² (clinical consensus)
2. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by a provider of the same sex if the child and/or family is more comfortable with a same-sex provider.² (clinical consensus)
3. Document all positive and negative findings of the physical exam. (clinical consensus)
4. Discuss the outcomes of the evaluation with the parents or caregivers and ask them if they have any concerns. (clinical consensus)
5. Refer the child to a pediatric endocrinologist if abnormal signs of puberty are observed. Abnormal signs could include progressive breast development over a 4- to 6-month period of observation or progressive penis and testicular enlargement, especially if accompanied by rapid linear growth. Children exhibiting these true indicators of early puberty need prompt evaluation by an appropriate pediatric endocrinologist.¹

6-12 years 11 months

Clinical Questions

1. Does intervening in pubertal development affect a child's self-perception?

2. What is the psychological impact on the initiation of puberty on the parent/caregiver versus the child?

Guidelines

1. Monitor and document weight and height velocity closely at every health supervision visit. Height should be measured (if possible) using a stadiometer. Often there may be difficulty assessing height due to inability to stand, scoliosis or contractures. In these cases, arm span or another appropriate parameter may be used. Care should be taken to use the same parameter at subsequent visits.² (clinical consensus)
2. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by same provider of the same sex if the child and/or family is more comfortable with a same-sex provider.² (clinical consensus)
3. Document all positive and negative findings of the physical exam. (clinical consensus)
4. Discuss the outcomes of the evaluation with the parents or caregivers and ask them if they have any concerns. (clinical consensus)
5. Refer the child to a pediatric endocrinologist if abnormal signs of puberty are observed. Abnormal signs could include progressive breast development over a 4- to 6-month period of observation or progressive penis and testicular enlargement, especially if accompanied by rapid linear growth. Children exhibiting these true indicators of early puberty need prompt evaluation by an appropriate pediatric endocrinologist.¹
6. Consider a referral to a mental health professional if the child is having psychosocial issues with his or her growth or development. (clinical consensus)

13-17 years 11 months

Clinical Question

1. How does puberty affect the self-perception of the 13-17-year-old with Spina Bifida?

Guidelines

1. Monitor and document weight and height velocity closely at every health supervision visit. Height should be measured (if possible) using a stadiometer. Often there may be difficulty assessing height due to inability to stand, scoliosis or contractures. In these cases, arm span or another appropriate parameter may be used. Care should be taken to use the same parameter at subsequent visits.² (clinical consensus)
2. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by a provider of the same sex if the child and/or family is more comfortable with a same-sex provider.² (clinical consensus)
3. Document all positive and negative findings of the physical exam. (clinical consensus)
4. Discuss the outcomes of the evaluation with the parents or caregivers and ask them if they have any concerns. (clinical consensus)
5. Refer the child to a pediatric endocrinologist if there is clear evidence of abnormal timing, tempo, or sequence of pubertal development. (clinical consensus)
6. Consider a referral to a mental health professional if the child is having psychosocial issues with his or her growth or development. (clinical consensus)

18+ years

Clinical Questions

1. How has completing puberty affected the individual's relationships with others?
2. Has the individual's self-perception changed as a result of completing puberty?

Guidelines

1. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by a provider of the same sex if the adult is more comfortable with a same-sex provider.² (clinical consensus)
2. Document all positive and negative findings of the physical exam.² (clinical consensus)
3. Discuss the outcomes of the evaluation with the patient and also with parents or caregivers, if appropriate, asking them if they have any concerns. (clinical consensus)
4. Consider a referral to a mental health professional if the individual is having psychosocial issues with their growth or development. (clinical consensus)
5. Discuss sexual health issues and make appropriate referrals to urologists, gynecologists or other sub-specialists such as endocrinology, adolescent medicine, genetics or others, as clinically appropriate. (clinical consensus) (Men's Health Guidelines, Sexual Health and Education Guidelines, Women's Health Guidelines)

Research Gaps

1. What effect does puberty and precocious puberty have on self-perception and psychological development of an individual with Spina Bifida?
2. How often should weight and length be measured in the 0-11-month age group?
3. What effect does hydrocephalus have on early-onset of puberty?
4. Is there an optimal age to intervene when precocious puberty is identified?
5. What is the best practice for counseling children and parents/caregivers during puberty or precocious puberty?
6. What is the best practice for detection and management of early puberty?
7. What is the psychological impact on the initiation of puberty on the parent/caregiver versus the child?
8. Does intervening in pubertal development affect a child's self-perception?
9. How has completing puberty affected the individual's relationships with others?
10. Has the individual's self-perception changed as a result of completing puberty?

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Endocrine: Short Stature & the Effect of Human Growth Hormone (hGH)

Workgroup Members: Joseph O'Neil, MD, MPH, FAAP (Chair); John S. Fuqua, MD

Introduction

The incidence of short stature caused by disproportionate growth of the lower body segments among children with Spina Bifida is well documented.¹⁻⁴ Hydrocephalus affects the hypothalamic-pituitary axis and the secretion of pituitary hormones responsible for growth and pubertal development.³ It is estimated that at least 30% of children with Spina Bifida have human growth hormone (hGH) deficiency.⁴ While the effect of short stature on quality of life has not been directly assessed; obesity and body image have been evaluated. Stature and subsequent body habitus does factor into perception of self-image and quality of life. However, body image ranks lower than other quality of life parameters compared to other factors such as pain, incontinence, and independence.⁵

There have been multiple studies regarding the use of hGH and its use among children with Spina Bifida. Most of these studies have demonstrated increased linear growth. The linear growth appears to have the greatest impact on the supine length and arm span. In some patients treated hGH, there was an increase in the frequency of symptomatic spinal cord tethering and progression of scoliosis. The overall consensus was that hGH treatment did improve both growth rate and length.⁶⁻¹⁰ While short stature has potentially negative consequences, treating with hGH in the face of normal labs is not recommended, if deficiency is suspected, referral is recommended, and the decision to treat for deficiency should be made using shared decision making within the context of the family centered care model.

Outcomes

Primary

1. Identify individuals with Spina Bifida who are growth hormone deficient.

Secondary

1. Improve quality of life by improving strength, mobility, body image, and health.
2. Reduce morbidity and mortality secondary to obesity.

0-11 months

Clinical Questions

1. At what post-conceptual age do pituitary-hypothalamic hormones become affected by Chiari malformation, hydrocephalus, or placement of shunts?
2. Could growth during infancy and first three years be improved by use of hGH?
3. Does the use of hGH worsen other comorbidities associated with Spina Bifida, such as scoliosis, tethered cord, or spasticity?
4. What and when are the appropriate evaluations for use of hGH?

Guidelines

1. Take frequent and accurate weight, length, and occipital frontal circumference measurements during infancy and early childhood.¹¹
2. Make referrals to physical therapy to maximize range of motion, strength, and functional mobility as appropriate for the developmental age. (clinical consensus) (Mobility Guidelines)

3. Encourage breastfeeding and appropriate nutrition.¹¹ (Nutrition Guidelines)
4. Discuss issues surrounding growth of children with Spina Bifida with the family. (clinical consensus)

1-2 years 11 months

Clinical Questions

1. At what post-conceptual age do pituitary-hypothalamic hormones become affected by Chiari malformation, hydrocephalus, or placement of shunts?
2. Could growth during infancy and first three years be improved by use of hGH?
3. Does the use of hGH worsen other comorbidities associated with Spina Bifida, such as scoliosis, tethered cord, or spasticity?
4. What and when are the appropriate evaluations for use of hGH?

Guidelines

1. Take frequent and accurate weight, length, and occipital frontal circumference measurements during infancy and early childhood.¹¹
2. Make referrals to physical therapy to maximize range of motion, strength, and functional mobility as appropriate for the developmental age. (clinical consensus) (Mobility Guidelines)
3. Encourage breastfeeding and appropriate nutrition.¹¹ (Nutrition Guidelines)
4. Discuss issues surrounding growth of children with Spina Bifida with the family. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. While linear growth is impacted by the effects of the myelomeningocele, at which age does the length become most affected (pre-pubertal years, pubertal growth spurt, and puberty)?
2. At what age is the short stature evaluation best initiated?
3. Who should do the evaluation and where should the evaluation be conducted?
4. Which parameters best predict a positive response to hGH?
5. Is hGH only indicated where growth hormone deficiency is identified?
6. Who should cover the cost of hGH?
7. Are there eligibility limitations to hGH treatment, such as: normal development, shortened arm span, minimal skeletal deformities, level of spinal lesion, amount of paresis, syringomyelia, tethered cord, scoliosis, vertebral anomalies, contractures or advanced pubertal development, with or without documented growth hormone deficiency?
8. Does hGH improve lipid or bone metabolism?
9. Does hGH result in enough of a positive change in adult height to see improved self-esteem, reduced obesity, better muscle strength and bone density, and rehabilitation potential?

Guidelines

1. Assess weight, height at each health supervision visit. ¹¹ If height is not able to be measured using a stadiometer, it is recommended that a consistent parameter (such as arm span) should be measured and recorded. (clinical consensus)
2. Have a discussion with the family about the expected height of the child, based on the limitations due to myelomeningocele and the parents' height. (clinical consensus)
3. Discuss the risks and benefits of hGH therapy with the parents. (clinical consensus)

4. If concerns about growth arise, a referral to a pediatric endocrinologist is recommended for growth assessment, IGF-1, IGF Binding Protein-3, and GH stimulation tests. (clinical consensus)
5. If hGH treatment is initiated, monitor pituitary function, scoliosis, tethering of spinal cord, growth velocity, and pubertal development. This may be done in collaboration with a pediatric endocrinologist. (clinical consensus)

6-12 years 11 months

Clinical Questions

1. While linear growth is impacted by the effects of the myelomeningocele, at which age does the length become most affected (pre-pubertal years, pubertal growth spurt, and puberty)?
2. At what age is the short stature evaluation best initiated?
3. Who should do the evaluation and where should the evaluation be conducted?
4. Which parameters best predict a positive response to hGH?
5. Is hGH only indicated where growth hormone deficiency is identified?
6. Who should cover the cost of hGH?
7. Are there eligibility limitations to hGH treatment, such as: normal development, shortened arm span, minimal skeletal deformities, level of spinal lesion, amount of paresis, syringomyelia, tethered cord, scoliosis, vertebral anomalies, contractures or advanced pubertal development, with or without documented growth hormone deficiency?
8. Does hGH improve lipid or bone metabolism?
9. Does hGH result in enough of a positive change in adult height to see improved self-esteem, reduced obesity, better muscle strength and bone density, and rehabilitation potential?

Guidelines

1. Assess weight, height at each health supervision visit.¹¹ If height is not able to be measured using a stadiometer, it is recommended that a consistent parameter (such as arm span) should be measured and recorded. (clinical consensus)
2. Have a discussion with the family about the expected height of the child, based on the limitations due to myelomeningocele and the parents' height. (clinical consensus)
3. Discuss the risks and benefits of hGH therapy with the parents. (clinical consensus)
4. If concerns about growth arise, a referral to a pediatric endocrinologist is recommended for growth assessment, IGF-1, IGF Binding Protein-3, and GH stimulation tests. (clinical consensus)
5. If hGH treatment is initiated, monitor pituitary function, scoliosis, tethering of spinal cord, growth velocity, and pubertal development. This may be done in collaboration with a pediatric endocrinologist. (clinical consensus)

13-17 years 11 months

There are no relevant clinical questions or guidelines for this age group. Please refer to younger age groups if needed.

18+ years

There are no relevant clinical questions or guidelines for this age group. Please refer to younger age groups if needed.

Research Gaps

1. Since linear growth is impacted by the effects of Spina Bifida, at which age does the length become most affected?
2. At what age should short stature evaluation be initiated?
3. Does hGH improve lipid or bone metabolism?
4. Does hGH result in enough of a positive change in adult height to see improved self-esteem, reduced obesity, better muscle strength and bone density, and rehabilitation potential?
5. What are the condition-specific risks of hGH treatment in the population with Spina Bifida?
6. Does fetal surgery improve linear growth or reduce the rates of hGH deficiency or precocious puberty?
7. Who should cover the cost of hGH?
8. At what post-conceptual age do pituitary-hypothalamic hormones become affected by Chiari malformation, hydrocephalus, or placement of shunts?
9. Which parameters best predict a positive response to hGH?
10. Are there eligibility limitations to treatment with hGH?

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Integument (Skin)

Workgroup Members: Patricia Beierwaltes, DNP, CPNP (Chair); Sharon Munoz, RN, MS, BC-CNS, CWOCN; Jennifer Wilhelmy, CWCN, APRN, CNP

Introduction

Skin-related issues have a significant impact on health, activities of daily living, and quality of life among people with Spina Bifida.^{1,2}

Data presented by select clinics that participate in the National Spina Bifida Patient Registry reported that 26% of individuals had a history of pressure injuries and 19% reported having had one in the past year.³ Complications related to wounds were reported to be the second most common primary diagnosis in Spina Bifida clinics.⁴ The literature on this topic indicates that the cost to care for an individual patient with a pressure injury ranges from \$20,900 to \$151,700 per pressure injury.⁵ A multi-clinic study from the NSBPR identified seven factors associated with pressure injuries that included the level of lesion, wheelchair use, urinary incontinence, shunt presence, above the knee orthopedic surgery, recent surgery and male sex.⁶ Though skin issues are not confined to pressure injuries, pressure injury prevention programs have shown as much as a 67% reduction in incidence with a substantial reduction in the cost of care.¹ With that goal in mind, the information campaign to improve skin care awareness and wound prevention, “*Did You Look?*” is being evaluated as a prevention program.⁷ Elements from this campaign are included in these guidelines.

The following integument (skin) guidelines focus on prevention, not treatment, of existing problems. Though Spina Bifida-specific evidence is limited, practices related to wound prevention can be applied to the Spina Bifida population. These guidelines are built on that evidence as well as clinical expertise.

Outcomes

Primary

1. Maximize healthy skin. Minimize disruptions in skin integrity.

Secondary

1. Increase awareness of skin issues, risks, self-assessment, and prevention measures.

Tertiary

1. Improve health outcomes with minimal skin integrity issues across the lifespan.

0-11 months

Clinical Questions

1. What is the evidence for the pathogenesis of skin breakdown (pressure injury) in infants with Spina Bifida?
2. Is there evidence that insensate skin in infants with Spina Bifida can be protected from breakdown?

Guidelines

1. Discuss insensate skin with parents/caregivers.⁸
2. Discuss the risk factors that may contribute to impaired skin integrity.^{5-6,9-14}
3. Teach to inspect the skin for changes in color, texture, and temperature.^{5-6,9-14}

4. Discuss the need to check water temperature and encourage the use of a bath water thermometer.^{6,9-14}
5. Tell parents and caregivers to check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.^{6,9-14}
6. Recommend the use of barrier creams to protect the skin from damage as a result of bowel and bladder incontinence.^{8,12}

1-2 years 11 months

Clinical Questions

1. What is the evidence that early intervention and education will reduce skin injury?

Guidelines

1. Teach parents and caregivers to inspect the skin (especially weight bearing or insensate areas) for changes in color, texture, and temperature.^{6,9-14}
2. Recommend the use of barrier creams to protect the skin from damage as a result of bowel and bladder incontinence.^{8,12}
3. Discuss the need to check water temperature and encourage the use of a bath water thermometer.^{6,9-14}
4. Tell parents and caregivers to check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.^{6,9-14}
5. Teach parents/caregivers how to inspect for well-fitting orthoses.¹⁵
6. Teach parents and caregivers that the child should wear protective clothing and footwear over insensate areas.¹²
7. Tell parents and caregivers to seek treatment if the child's skin is compromised.¹²

3-5 years 11 months

Clinical Questions

1. What activities promote self-awareness and self-inspection in children with Spina Bifida?

Guidelines

1. Teach parents and caregivers to inspect the skin daily (especially weight bearing or insensate areas) for changes in color, texture, and temperature.^{6,9-14}
2. Encourage the child's involvement in skin inspection.¹²
3. Teach child to develop awareness of insensate areas.^{6,9-14}
4. Review with parents and caregivers the consequences of heat, moisture, or pressure related to insensate areas.^{6,9-14,20}
5. Recommend the use of barrier creams to protect the skin from damage as a result of bowel and bladder incontinence.^{8,12}
6. Discuss the need to check water temperature and encourage the use of a bath water thermometer.^{6,9-14}
7. Tell parents and caregivers to check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.^{6,9-14}
8. Teach parents/caregivers how to inspect for well-fitting orthoses.¹⁵
9. Teach parents and caregivers that the child should wear protective clothing and footwear (including water shoes in a pool or on pool deck) over insensate areas.¹²
10. Tell parents and caregivers to seek treatment if the child's skin is compromised.¹²

6-12 years 11 months

Clinical Questions

1. What evidence is there that coaching independence will reduce skin breakdown?

Guidelines

1. Teach parents and caregivers to inspect the skin daily (especially weight bearing or insensate areas) for changes in color, texture, and temperature.^{6,9-14}
2. Encourage the child's involvement in skin inspection.¹²
3. Teach child to develop awareness of insensate areas.^{6,9-14}
4. Review with parents and caregivers the consequences of heat, moisture, or pressure related to insensate areas.^{6,9-1,20}
5. Teach parents/caregivers how to inspect for well-fitting orthoses and other equipment that may cause injury to skin.¹⁵
6. Teach parents and caregivers that the child should wear protective clothing and footwear over insensate areas.¹²
7. Discuss the need to check water temperature and encourage the use of a bath water thermometer.^{6,9-14}
8. Tell parents and caregivers to check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.^{6,9-14}
9. Promote adequate hydration and proper nutrition for healthy skin.^{3,12,17} (Nutrition, Metabolic Syndrome, and Obesity Guidelines)¹⁹
10. Encourage parents, caregivers, and the child to keep skin clean and dry.^{6,9-14}
11. Suggest wearing seamless socks that are clean and dry.¹²
12. Suggest the use of antiperspirant on areas with perspiration, such as the feet and intertriginous areas.¹²⁻¹³
13. Tell parents and caregivers to seek treatment if the child's skin is compromised.¹²
14. Advise parents and caregivers to engage non-ambulatory children in pressure-relieving activities every 15 minutes.^{1,5,13}

13-17 years 11 months, 18+ years

Clinical Questions

1. What are the key factors associated with skin breakdown?
2. Does the incidence of skin breakdown relate to the level of Spina Bifida?
3. What evidence is there that specific prevention measures will reduce the chance of skin breakdown?

Guidelines

1. Inspect skin daily. Explore the teen perceptions of self-efficacy for skin checks and barriers to skin checks. Develop plans to increase self-efficacy, if needed.^{6,9-14}
2. Suggest children and adults who use wheelchairs to use a pressure-relieving cushion and check it daily.^{1,13-14}
3. Identify and discuss risk factors that specifically increase the risk of pressure injuries in children and adults with Spina Bifida, such as using a wheelchair, having had surgery above the knee, shunts, a higher level of lesion, recent surgery, bladder incontinence, and being of the male gender.^{11,15-16,18}
4. Review with the caregiver, child, or adult the consequences of heat, moisture, or pressure related to insensate areas.^{6,9-14,2}
5. Teach parents/caregivers/child/adult how to inspect for well-fitting orthoses.¹⁵
6. Discuss the need to check water temperature and encourage the use of a bath water thermometer.^{6,9-14}
7. Tell children/adults to check for hot surfaces that have been exposed to the sun such as car seats.^{6,9-14}
8. Promote adequate hydration and proper nutrition for healthy skin.^{12,17} (Nutrition, Metabolic Syndrome, and Obesity Guidelines)¹⁹
9. Encourage parents, caregivers, children, and adults to keep skin clean and dry.^{6,9-14}

10. Suggest wearing seamless socks that are clean and dry.¹²
11. Suggest the use of antiperspirant on areas with perspiration, such as the feet and intertriginous areas.¹²⁻¹³
12. Encourage seeking treatment if the skin is compromised.¹²
13. Advise children and adults who are non-ambulatory and use a wheelchair to engage in pressure-relieving activities every 15 minutes.^{1,13-14}
14. Teach safe transfer skills to non-ambulatory patients.^{12,15-16}
15. Seek treatment immediately for any pressure injury. Refer to wound clinic for any pressure injury at stage three or greater.¹²

Research Gaps

1. Does consistent implementation of a defined bundle of prevention strategies reduce the incidence of skin breakdown in individuals with Spina Bifida?
2. What evidence is there that coaching independence will reduce skin breakdown?
3. What is the evidence for the pathogenesis of skin breakdown (pressure injury) in infants with Spina Bifida?
4. Is there evidence that insensate skin in infants with Spina Bifida can be protected from breakdown?

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Latex and Latex Allergy in Spina Bifida

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Introduction

(In these guidelines, "latex allergy" refers to type I hypersensitivity to *Hevea brasiliensis*, also known as natural rubber latex.)

The history of latex allergy and its intersection with people with Spina Bifida dates back to the late 1980's in the United States with the advent of Universal Precautions and no regulation of latex in gloves. As clinical reports of severe allergic reactions, including anaphylaxis and a 500-fold increase of life-threatening events in surgery for Spina Bifida, efforts were made to better understand the patterns of these episodes. In doing so, latex allergy was subsequently found to be the associated trigger in surgical procedures in patients with Spina Bifida and other conditions, particularly those with congenital neurogenic bladder conditions.¹⁻²

As a result, by the early 1990's there were efforts to systematically avoid exposing infants and children with Spina Bifida to natural rubber products such as red rubber catheters and surgical gloves, or latex products used in various settings such as in neonatal intensive care units or newborn nurseries. As the importance of these measures became more widely accepted, there followed an extension to other areas of children's hospitals, emergency rooms, and to many pediatric offices where the children were subsequently seen.

Despite these efforts, exposure to latex remains relatively prevalent in the different environments frequented by people of all ages with Spina Bifida — hospitals, clinics, schools, homes, and community facilities. Exposure to latex could take place by direct contact or inhalation. Symptoms of latex allergy may initially be considered mild, such as skin irritations, rash, hives, flushed cheeks, itchy eyes, or sneezing. However, they can immediately progress or evolve related to subsequent exposures to more dramatic systemic responses such as generalized urticaria, wheezing, coughing, periorbital erythema and swelling, and even nausea and vomiting.³

Until better scientific explanations are available to specifically drive prevention and intervention, people with Spina Bifida should continue to avoid skin contact with latex protein in the environment including foods with similar proteins, and avoid inhalation of powder that contains latex.⁴⁻⁷ Avoidance of latex should extend to latex-containing products used for personal care, medical care, dental care, and community participation.⁸⁻⁹

Children, families and adults should be aware that caution should be taken regarding what has come to be labeled "latex fruit syndrome." This remains incompletely understood and likely related to epigenetic factors.¹⁰ The protein allergen (example, *Hev b 6* hevein) in some latex products makes up a considerable amount of the total protein. This has been shown to have significant cross-reactivity to certain proteins (chitinases) in banana, avocado and chestnuts, for example. While these fruits have been the most commonly described, there are at least 25 other fruits that may have some level of cross-reactivity with latex. For example, potatoes, eggplant, and kiwi have been described as potential concerns.

At this time, it is helpful to understand that not all patients with true latex allergy have clinical reactions to fruit (~50%) and that few (~ 10%) of individuals with known allergy to a latex-cross-reacting fruit develop latex allergy symptoms.¹⁰ Parents and patients should be aware of potential “latex-fruit syndrome” reactions, but should also be aware of its relative risk. (Appendix 2)

For additional details and resources on latex allergy, please review Appendix 1: Latex Allergy Fact Sheets and Other Materials and Appendix 2: Latex Allergy and Foods.

Outcomes

Primary

1. Avoidance of all direct contact to natural rubber latex.
2. Awareness and understanding that latex allergy remains a relatively high-risk condition for this group.
3. Avoidance of skin contact with latex protein in the environment and inhalation of powder that contains latex (i.e. gloves).
4. Avoidance of latex-containing products used for personal care, medical care, and community participation (e.g., adhesive bandages, latex gloves, surgeries in medical setting that may not be latex-free.)
5. Awareness of signs and symptoms of latex allergy.

Secondary

1. Persons with known latex allergy routinely wear medical-alert identification.
2. Persons with known latex allergy and their family know the signs of life-threatening anaphylaxis.

Tertiary

1. Persons with known latex allergy and their family have a pre-arranged plan for action in the event of a severe, life-threatening anaphylaxis.

0-11 months

Clinical Questions

1. Are health care providers becoming complacent about latex risks in hospital and office settings?
2. How do we recognize infants who are most at risk or those that may turn positive?

Guidelines

1. Inform parents and caregivers about latex allergy and ways to provide safe infant care while avoiding exposure to latex products.^{3-4,11-19}
2. Avoid using health care products that contain latex when caring for infants with Spina Bifida^{3-4,11-19}
3. Inform staff and families of any latex-containing products such as bottle nipples, pacifiers, teething rings, toys, and other items such as urinary catheters.^{3-4,11-19} (Appendix 1)

1-2 years 11 months

Clinical Questions

1. Is there consensus on how to conduct preventive screening, or should investigation begin when a patient has a reaction and needs specific testing?
2. How do we recognize those most at risk or those who may turn positive?
3. When are diagnostic studies being done, and is there unified consensus on the process or timing?

4. Are health care providers becoming complacent about latex risks?
5. Has the problem of airborne latex been solved by not having powder in latex gloves?

Guidelines

1. Develop awareness that increased mobility puts the child at greater risk for exposure to latex products. (clinical consensus)
2. Avoid toys and other items such as urinary catheters with latex. All toys should be latex-free.^{1,20-23} (Appendix 1)
3. Encourage careful parental observation of latex avoidance.^{1,20-23}
4. Encourage the child with a history of latex allergy to wear a medical identification bracelet showing allergy to latex. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. Is there consensus of how preventive screening should be done, or should investigation begin when a patient has a reaction and needs specific testing?
2. How do we recognize those most at risk or those who may turn positive?
3. When are diagnostic studies being done, and is there unified consensus on the process or timing?
4. Has the problem of airborne latex been solved by not having powder in latex gloves?
5. Are health care providers becoming complacent about latex risks?
6. Given that there is potential for cross-reactivity in numerous foods, how should families prepare their children?

Guidelines

1. Screen toys and the environment of preschoolers as they start to interact with their peer group more regularly. Keep children away from toys and other products that contain latex such as latex-containing urinary catheters. (clinical consensus) (Appendix 1)
2. Discuss avoidance of rubber balloons at parties, school activities, restaurants, and other gathering places for events.^{14-16,18, 21-30}
3. Teach children to ask questions about items that may contain latex.^{24,26}
4. Teach children, at a very basic level, to avoid latex products.^{24,26}
5. Help children and parents identify latex-free substitute products, such as Mylar balloons, for celebrations. (clinical consensus) (Appendix 1)
6. Instruct families to check that food made in public venues has been prepared with latex-free gloves. (clinical consensus)
7. Refer to an allergist when the child is allergic to latex but does not know if he or she is allergic to cross-reacting foods; this is particularly crucial in those who have had a systemic or anaphylactic episode. (Appendix 2) If a positive test is found, then a food challenge would be indicated in the case where there is no history of food-related clinical reaction. Many of the positive tests may be due to laboratory cross-reactivity, but a clinical response of allergy will not be provoked.³²

6-12 years 11 months

Clinical Questions

1. Is there consensus of how preventive screening should be done, or should investigation begin when a patient has a reaction and needs specific testing?
2. How do we recognize those most at risk or those who may turn positive?
3. When are diagnostic studies being done, and is there unified consensus on the diagnostic process or timing?
4. Has the problem of airborne latex been solved by not having powder in latex gloves?

5. Are health care providers becoming complacent about latex risks?
6. Given that there is potential for cross-reactivity in numerous foods, how should families prepare their children?

Guidelines

1. Educate school-age children about their avoidance of latex products such as latex-containing urinary catheters and inform them about safe, latex-free alternatives. (clinical consensus) (Appendix 1)
2. Discuss avoidance of rubber balloons at parties, school activities, restaurants, and other gathering places for events.^{14-16,18, 21-30}
3. Tell parents and caregivers of children identified as having a latex allergy, and the children themselves, to have diphenhydramine and self-administered epinephrine available at all times. (clinical consensus)
4. Instruct families to check that food made in public venues has been prepared with latex-free gloves. (clinical consensus)
5. Urge children to continue following latex precautions because risk-taking during the teen years is common. (clinical consensus)
6. Review the principles of latex precaution with the child during a clinic visit and answer any questions. (clinical consensus)
7. Refer to an allergist when the child is allergic to latex but does not know if he or she is allergic to cross-reacting foods; this is particularly crucial in those who have had a systemic or anaphylactic episode. (Appendix 2) If a positive test is found, then a food challenge would be indicated in the case where there is no history of food-related clinical reaction. Many of the positive tests may be due to laboratory cross-reactivity, but a clinical response of allergy will not be provoked.³²

13-17 years 11 months

Clinical Questions

1. Is there consensus of how preventive screening should be done, or should investigation begin when a patient has a reaction and needs specific testing?
2. How do we recognize those most at risk or those who may turn positive?
3. When are diagnostic studies being done; any unified consensus on the process or timing?
4. Has the problem of airborne latex been solved by not having powder in latex gloves?
5. Are health care providers becoming complacent about latex risks?
6. Given that there is potential for cross-reactivity in numerous foods, how should families prepare their children?

Guidelines

1. Educate teens directly about avoidance of latex products including latex-containing urinary catheters and educate them to know about safe latex-free alternatives. (clinical consensus) (Appendix 1)
2. Discuss avoidance of rubber balloons at parties, school activities, restaurants, and other gathering places for events.^{14-16,18, 21-30}
3. Teens identified as having a latex allergy should have diphenhydramine and self-administered epinephrine available at all times. (clinical consensus)
4. Instruct families to check food preparation in public venues as it should be prepared with latex-free gloves. (clinical consensus)
5. Educate teens about latex-safe contraceptive products before they decide to become sexually active. (clinical consensus) (Sexual Health and Education Guidelines) (Appendix 1)

6. Urge children to continue following latex precautions because risk-taking during the teen years is common. (clinical consensus)
7. Review principles of latex precaution with the teen during a clinic visit and answer any questions. (clinical consensus)
8. If a latex allergic patient does not know if he or she is allergic to cross-reacting foods and has had anaphylaxis to latex exposure, it may be prudent for an allergist to test the patient. If a positive test is found, then a food challenge would be indicated in the case where there is no history of food related clinical reaction. (Appendix 2) Many of the positive tests may be due to laboratory cross-reactivity, but a clinical response of allergy will not be provoked.³²

18+ years

Clinical Questions

1. Is there consensus of how preventive screening should be done, or should investigation begin when a patient has a reaction and needs specific testing?
2. How do we recognize those most at risk or those who may turn positive?
3. When are diagnostic studies being done, and is there unified consensus on the process or timing?
4. Are health care providers becoming complacent about latex risks?
5. What research endeavors, clinical practices, and/or education is needed to best assure a latex-free medical environment for women with Spina Bifida who are being seen in obstetric/gynecologic medical environments?
6. Has the problem of airborne latex been solved by not having powder in latex gloves?
7. Given that there is potential for cross-reactivity in numerous foods, how should families and adults prepare for cross-reactivity in numerous foods?

Guidelines

1. Urge adults with Spina Bifida to continue following latex precautions, even if they have not experienced an adverse response to latex products (for example, latex-free condoms), until better scientific explanations are available to specifically drive prevention and intervention.^{1-2,12-15,22,24,31-33} (Sexual Health and Education Guidelines)
2. Educate adults directly about avoidance of latex products including latex-containing urinary catheters and educate them to know about safe latex-free alternatives. (clinical consensus) (Appendix 1)
3. Discuss avoidance of natural rubber products in the home and work environments.^{14-16,18, 21-30}
4. Adults identified as having a latex allergy should have diphenhydramine and self-administered epinephrine available at all times. (clinical consensus)
5. Instruct adults to check food preparation in public venues as it should be prepared with latex-free gloves. (clinical consensus)
6. Educate adults about latex safe contraceptive products before they decide to become sexually active. (clinical consensus) (Sexual Health and Education Guidelines) (Appendix 1)
7. Review principles of latex precaution with the adult during a clinic visit and answer any questions. (clinical consensus)
8. If a person that is allergic to latex does not know if he or she is allergic to cross-reacting foods and has had anaphylaxis to latex exposure, it may be prudent for an allergist to test the patient. If a positive test is found, then a food challenge would be indicated in the case where there is no history of food related clinical reaction. Many of the positive tests may be due to laboratory cross-reactivity, but a clinical response of allergy will not be provoked.³² (Appendix 2)

Research Gaps

1. Updated measures are needed on the true incidence and prevalence among people with Spina Bifida and a comparison to other potentially high-risk populations (i.e. nurses, environmental services workers, and others who routinely make use of latex products).
2. Determine if there is an impact of antenatal repair on latex allergy.
3. Further investigation is needed into patients with Spina Bifida who “turn positive,” including questions about immunity, genetic differences, differences in exposure, and other factors.
4. Since latex gloves are used less frequently in children’s hospitals now than in the past, and measures of true levels of exposure in other settings such as adult hospitals or dental offices is likely to be inexact, consider carrying out animal model studies to better answer questions of clinical impact of exposure to powder in latex gloves.
5. For cohorts of people with Spina Bifida who become positive to latex (clinically or based on screening labs), in-depth epidemiology studies need to be constructed and implemented.
6. Are health care providers becoming complacent about latex risks in hospital and office settings?
7. How do we recognize infants, children, teens, or adults who are most at risk or those that may turn positive? Is there consensus on how to conduct preventive screening, or should investigation begin when a patient has a reaction and needs specific testing?
8. When are diagnostic studies being done, and is there unified consensus on the process or timing?
9. Has the problem of airborne latex been solved by not having powder in latex gloves?
10. Among individuals with latex sensitivity, there is potential for cross-reactivity in numerous foods; what precautions might be shared with families in these instances?
11. What research endeavors, clinical practices, and/or educational initiatives are needed to best assure a latex-free medical environment specifically for women with Spina Bifida who are being seen in obstetric/gynecologic medical environments?

Appendix 1: Latex Allergy Fact Sheets and Other Materials

Permission to use the materials found in the [Latex Allergy Toolbox](http://www.allergyasthmanetwork.org/education/allergies/latex-allergy/) [links to <http://www.allergyasthmanetwork.org/education/allergies/latex-allergy/>] has been granted by the Allergy & Asthma Network. The toolkit features variety of fact sheets and other materials for health care providers, parents, students, school staff, and others, including:

- Parent and practitioner resources
- Latex and vaccines
- School resources
- Allergy & Asthma Network webinars

Appendix 2: Latex Allergy and Foods

Parents and patients should be aware that caution should be taken regarding what has come to be labeled “latex fruit syndrome.” Research has shown that some foods have proteins that are like those in rubber tree sap. Sometimes people with latex allergies experience a reaction to “latex reactive foods.” This may be referred to as latex-food syndrome or latex-fruit allergy. Latex reactive foods include primarily nuts and fruit, but also some vegetables.^{10,34-36}

Foods with a high degree of latex allergy association or prevalence:

- Avocado
- Banana
- Chestnut
- Kiwi

Foods with a moderate degree of latex allergy association or prevalence:

- Apple
- Carrot
- Celery
- Melons
- Papaya
- Potatoes
- Tomatoes

Foods with low or undetermined latex allergy association:

Apricot, Buckwheat, Castor Bean, Cayenne Pepper, Cherry, Chick Peas, Citrus Fruits, Coconut, Dill, Fig, Grape, Hazelnut, Lychee, Mango, Nectarine, Oregano, Passion Fruit, Peach, Peanut, Pear, Persimmon, Pineapple, Plum, Rye, Sage, Shellfish, Soybean, Strawberry, Sunflower Seed, Sweet Pepper, Walnut, Wheat, Zucchini.

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Nutrition, Metabolic Syndrome, and Obesity

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Introduction

Healthy nutrition is important for everyone. Individuals with Spina Bifida experience unique challenges related to healthy dietary intake across the lifespan.¹ During infancy, some babies may experience slow weight gain and inadequate nutrition. This is typically due to a complex interplay of medical and social factors, such as brain stem dysfunction, shunt obstruction, silent aspiration, sleep apnea, recurrent infections, and altered feeding dynamics related to frequent hospitalization and caregiver stress. However, most children with Spina Bifida follow typical growth patterns until they are four years of age. After that, increased fat mass (versus lean mass) has been found when compared with children without Spina Bifida.² Children with Spina Bifida who have a latex allergy may limit their intake of fresh fruits and vegetables due to cross-reacting foods (Latex and Latex Allergy in Spina Bifida Guidelines). Those with the Chiari II malformation may have an aversion to textured foods because of brainstem dysfunction.³ For many individuals with Spina Bifida, concern about bowel and bladder accidents may be prioritized over hydration and optimal nutrition.⁴ Poor eating habits can lead to constipation, skin breakdown, osteoporosis, anemia, obesity, metabolic syndrome, and other preventable secondary conditions.⁵ It is therefore critical to work with children, families and adults with Spina Bifida to emphasize the importance of healthy nutrition and a balanced diet on overall health and wellness.⁶

Children and adults with Spina Bifida have higher rates of overweight and obesity compared to the general population,⁷ which may lead to negative health outcomes later in life such as metabolic syndrome, cardiovascular disease and type II diabetes.⁸ Girls with Spina Bifida diagnosed with premature puberty may experience weight gain,⁹ especially if pharmacological treatments are used. Polycystic Ovary Syndrome can also lead to weight gain in women with Spina Bifida.¹⁰ Sleep apnea can result from weight gain or result in changes in weight over time.¹¹ Overweight among people with Spina Bifida may also impact mobility, independence, and quality of life.¹² It is important for people with Spina Bifida to understand the possible risks associated with both poor nutrition and obesity and for health care professionals to discuss these topics using a collaborative, strengths-based approach. (Health Promotion and Preventive Health Care Services Guidelines, Physical Activity Guidelines)

It is recommended that body mass index (BMI) be calculated at clinical encounters in order to track an individual’s BMI trajectory and guide discussion of appropriate nutrition and weight management strategies. The appendix accompanying these guidelines summarizes anthropometric measurement techniques that can be used to accurately calculate BMI in people with Spina Bifida.

These guidelines aim to provide the best available evidence for promoting healthy nutrition in people with Spina Bifida across the lifespan. The guidelines will also suggest how health care professionals can have positive weight-related conversations with their patients, and offer strategies to prevent and manage obesity.

Outcomes

Primary

1. Maximize and support wellness through the lifespan.

Secondary

1. Reduce and prevent other secondary conditions related to poor nutrition and overweight/obesity, including metabolic syndrome.

Tertiary

1. Support the development of client/caregiver knowledge, self-management skills, and self-efficacy related to nutrition and dietary habits.

0-11 months

Clinical Questions

1. What nutritional support should be provided when infants with Spina Bifida first go home from the hospital?
2. What guidance on breastfeeding and/or use of breast milk should be given to parents of infants with Spina Bifida?

Guidelines

1. Assess weight, height and occipital frontal head circumference at every clinical encounter.¹³⁻¹⁴ (Appendix: BMI and Body Composition Measurements)
2. Ensure that the family's nutrition plan is followed closely by a primary care provider:
 - Refer the family to community nursing and other support groups to ensure close monitoring of the child's growth and whether there are issues with feeding and elimination. (clinical consensus)
 - Connect the family with the Spina Bifida specialist clinic nearest them. (clinical consensus)
3. Provide parents and caregivers with pre- and post-natal guidance and support on breastfeeding.
 - Discuss with them that ideally, infants with Spina Bifida should breastfeed or be given breast milk exclusively for the first six months. Infants should continue to have breast milk for a year or more, as with all neonates.¹⁵
 - Inform the mother that if the spinal surgery precludes immediate breast feeding, she will need to pump breastmilk to feed her baby until it is possible to transfer her baby to her breast.¹⁶
 - Urge the mother to begin pumping breast milk within six to 12 hours of delivery.
 - Emphasize the need to pump frequently (eight to 10 pumping sessions per 24 hours for the first seven-10 days) to ensure enough will be available once the infant has surgery.¹⁶
 - Advocate for babies to be hospitalized in close proximity to their mothers to facilitate breastfeeding.¹⁷
 - Provide mothers with information about accessing breast milk banks and to plan for situations where she cannot provide the breast milk herself.¹⁸
 - Encourage mothers to nurse their child in a flat position for five days following surgery to reduce pressure on the wound and avoid a cerebral spinal fluid leak.¹⁹
 - Provide the mother with information about breastfeeding equipment options that can help meet the individual needs of the child with Spina Bifida (e.g., different types of propping pillows, nipples, bottles, pumps, latex-free equipment, and supplemental nursing systems).¹⁹
 - Mention that severe Chiari malformation may affect successful latching and coordination of sucking, swallowing, and breathing. A referral to a lactation

- consultant should be made if mothers continue to experience challenges.¹⁶
- Support mothers to thicken their breast milk to prevent aspiration.²⁰
- Suggest breastfeeding or non-nutritive sucking (finger or pacifier) as ways to comfort their baby and assist them with pain management for acute procedures such as injections.²¹
- Highlight that the baby's transition from drinking breastmilk to eating solid food can cause constipation.²²
- Close multi-disciplinary follow up is indicated for infants with slow weight gain and failure to thrive. (clinical consensus)

1-2 years 11 months

Clinical Questions

1. What evidence-based information can be provided to parents on nutrition and obesity prevention and management?
2. What is the best way to manage constipation with diet for this age group?
3. How can providers communicate with parents about the benefits of a healthy diet in an understandable manner?

Guidelines

1. Assess weight and height at every clinical encounter.¹⁴ (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. Measure occipital frontal head circumference until two years of age.¹³ (Appendix: BMI and Body Composition Measurements)
3. Support families as they work to establish a healthy relationship and behavior towards food with their child. Tailor the advice to the family as appropriate.
 - Start introducing healthy foods as early as possible to get them integrated into food preferences. It may be harder to do so later, when food preferences are more entrenched.²³
 - Recognize that children with Chiari malformation may have sensitivity to different food textures.³
 - Consider that adults and families with lower incomes may experience food insecurity.¹
 - Caution the parents that habitually providing unhealthy foods can lead to a cycle of more requests and greater consumption of unhealthy foods. In contrast, healthy feeding practices early on can help avoid that cycle.²⁴
 - Discuss with parents and caregivers that overly restricting food, especially energy-dense foods that are high in fat and have a low water content such as cookies, chips, and nuts, can lead their child to overeat those foods when they become available.²³ Therefore, balance is needed.
 - Caution parents against using food as a reward or positive reinforcement, which can create an unhealthy relationship with food that is hard to break later on and that may lead to undesirable eating behaviors.^{23,25} Provide parents with other strategies for positive reinforcement rewards such as praise, stickers, and small toys. (clinical consensus)
 - Educate families on the importance of consuming a balanced diet and how it affects the whole body.⁶
 - Discuss that some fluid and food options used to help ensure hydration and bladder/bowel function are not necessarily the right choices for weight management (e.g. chocolate milk, juice, and sports beverages). Instead, encourage them to hydrate by drinking non-caloric fluids (e.g. water, club soda, sugar-free flavored drinks).²⁶

4. Speak with parents about nutrition in terms of their child's health and growth.
 - Provide regular opportunities for parents to discuss any concerns about their child's weight, growth, and/or eating behaviors.²⁷ A trusting therapeutic relationship can greatly facilitate an honest and open discussion.²⁸
 - Partner with parents to identify and address specific challenges that the family is facing.²⁸
 - Discuss that poor eating habits and reduced activity may lead to obesity, constipation, skin breakdown, osteoporosis, anemia, and other problems.⁵ Additionally, mention that children with Spina Bifida have a high risk of obesity because they have less calorie-burning tissue (lean body mass) and a lower rate of burning calories (metabolic rate).^{7,29}
 - Show parents the trajectory of a child's weight and height (or other measures of growth and adiposity), if appropriate. Use a growth chart as a visual aid, without referring to growth cut-offs developed for typically developing children.¹ A steeply-increasing trajectory would indicate that overweight or obesity may be a concern and warrant proactive discussions of preventative strategies.²⁸
 - Highlight the importance of parents modeling healthy behaviors themselves to their children from an early age.^{23,30-31} Encourage the whole family to get involved in healthy living activities, not just the child with Spina Bifida.³²
 - Discuss that children with Spina Bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and those with higher body fat levels or contractures, are at increased risk for bone fractures.³³ Encourage physical activity and healthy lifestyles. (Physical Activity Guidelines)
5. Provide guidance on maintaining good bowel health.
 - Explain that increased fiber in the child's diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals.⁵
 - Recommend the same guidelines for daily fiber intake that are recommended for all children:³⁴
 - 1-3 years: 19g
 - 4-8 years: 25g
 - 9-13 years: female–26g, male–31g
 - 14-18 years: female–26g, male–38g
 - Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. (clinical consensus)
 - Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the 24-hour period daily maintenance fluid requirements calculation³⁵:
 - 100 mL/kg for the first 10 kg body weight
 - + 50 mL/kg for the next 10 kg body weight
 - + 20 mL for every kilogram of body weight over 20 kg
 - Further guidance can be found in the Bowel Function and Care Guidelines.
6. Screening for dyslipidemia (fasting lipid profile) is recommended every two years from two years of age if the child's BMI is above 95th percentile or a family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives is present.³⁶

3-5 years 11 months

Clinical Questions

1. What evidence-based information on nutrition and obesity prevention and management can be provided to parents?
2. What is the best way to manage constipation with diet for this age group?
3. How can providers communicate with parents about the benefits of a healthy diet in an understandable manner?

Guidelines

1. Conduct annual assessments of weight, height or arm span, and calculate BMI. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. Discuss general weight-management principles with all families of children with Spina Bifida, and highlight the importance of healthy behaviors for the entire household.
 - Provide regular opportunities for parents to discuss concerns with their child's weight, growth and/or eating behaviors.²⁷ A trusting therapeutic relationship can greatly facilitate an honest and open discussion.³⁷
 - Emphasize the broad benefits of healthy eating and physical activity, offering strategies to enable the child to incorporate healthy lifestyle behaviors appropriate to their abilities.⁴
 - Consider that adults and families with lower incomes may experience food insecurity.¹
 - Highlight that early eating patterns and relationships with food are critical for ongoing good nutrition through the lifespan.²⁷
 - Discuss that some fluid and food choices to help ensure hydration and bladder/bowel function are not necessarily the right choices for weight management (e.g. chocolate milk, juice, and sports beverages).²⁶
 - Show parents the trajectory of a child's weight and height (or other measures of growth and adiposity). Use a growth chart as a visual aid, without referring to growth cut-offs developed for typically-developing children.¹ A steeply-increasing trajectory would indicate that overweight or obesity may be a concern and warrant proactive discussions of preventative strategies.²⁸
 - Discuss with parents, if relevant, that the Body Mass Index (BMI) is an imperfect indicator of health in all young people and especially in children with Spina Bifida due to difficulties measuring height and body composition.²⁹
 - Consider monitoring other measures of adiposity, such as waist circumference.³⁸ (Appendix: BMI and Body Composition Measurements)
 - Explain that most children with Spina Bifida follow typical growth patterns until they are four years of age. After that, increased fat mass (versus lean mass) has been found when compared with children without Spina Bifida.²
 - Discuss that linear growth or height will also be slower than peers without Spina Bifida due to paresis or paralysis of lower limbs,³⁹ which also reduces calorie requirement.
 - Highlight that children with Spina Bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and who have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D.³³
3. Provide guidance on maintaining good bowel health.
 - Explain that increased fiber in the child's diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals.⁵
 - Recommend the same guidelines for daily fiber intake that are recommended for

all children³⁴:

- 1-3 years: 19g
 - 4-8 years: 25g
 - 9-13 years: female–26g, male–31g
 - 14-18 years: female–26g, male–38g
- Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. (clinical consensus)
 - Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the 24-hour period daily maintenance fluid requirements calculation:³⁵
 - 100 mL/kg for the first 10 kg body weight
 - + 50 mL/kg for the next 10 kg body weight
 - + 20 mL for every kilogram of body weight over 20 kg
 - Further guidance can be found in the Bowel Function and Care Guidelines.
4. Screening for dyslipidemia (fasting lipid profile) is recommended every two years if the child's BMI is above the 95th percentile or a family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives is present.³⁶

6-12 years 11 months

Clinical Questions

1. What information do schools and communities need to know about the special dietary and adapted equipment needs of students with Spina Bifida in order to help children eat and access food independently at school or in the community?
2. What parenting strategies can encourage a balanced and healthy diet for the whole family?
3. What is the best way to manage constipation with diet for this age group?
4. Are children with Spina Bifida and obesity at higher risk for metabolic syndrome?
5. Should screening for metabolic complications of obesity be performed in children aged 6-12 years with Spina Bifida?
6. Is there evidence to support the role of weight management intervention in the prevention of metabolic syndrome?

Guidelines

1. Conduct annual assessment of weight, height or arm span, and BMI. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. Consider monitoring other measures of adiposity, such as waist circumference.³⁸ (Appendix: BMI and Body Composition Measurements)
3. Conduct annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension. (clinical consensus)
4. Highlight dietary needs specific to living with Spina Bifida.
 - Discuss the importance of consuming fiber and water to manage bowel and bladder health. Sources of fiber include fruit, vegetables, whole wheat or whole grain bread and cereals. A mix of each along with regular fluids will help avoid constipation.⁵
 - Advise limiting sugary drinks such as juice, chocolate milk, and sports beverages.⁴⁰⁻⁴¹
 - Discuss that children with Chiari malformation may have a sensitivity to different food textures.³

- Recommend that the child have access to food purchasing and preparation spaces. (clinical consensus)
 - Recommend a diet with adequate calcium and vitamin D for children with Spina Bifida, in order to avoid fractures due to osteoporosis.³³
 - Consider that adults and families with lower incomes may experience food insecurity.¹
 - Refer clients to National Center on Health, Physical Activity, and Disability (<http://www.nchpad.org>), which provides advice on nutrition and physical activity for persons with disabilities, including Spina Bifida.⁴²
5. Provide families with nutritional information tailored to their circumstances.
- Take into account a family's geographical location, ethnicity, access to food, and other related factors when providing dietary education.⁴³
 - Consider that adults and families with lower incomes may experience food insecurity.¹
 - Encourage parents to include their children from an early age to participate in grocery shopping and food preparation, as appropriate to their age and ability.⁴⁴
 - Suggest parents to let their children choose a new healthy food to try. Involving children in choices in food selection can lead them to increased independence and interest in their foods and to learn about making healthy choices. Repeating their exposure to healthy food options can increase children's acceptance and enjoyment of these foods.⁴⁵
 - Involve children in discussions about healthy lifestyles in order to explore their understanding, perceptions, and priorities regarding nutrition.²⁸ If appropriate, ask parents to identify one or two small, healthy nutrition changes that they feel they could integrate into their daily life.⁴⁶
 - Consider making a referral to a healthy lifestyle program and/or a smartphone application, while recognizing that few such programs are tailored to individuals with disabilities. (clinical consensus)
 - Celebrate any successes, such as drinking more water, introducing a new fruit or vegetable, cutting back on sugary drinks, and having regular meal times. Focus upon the strengths of the family.²⁸
 - Highlight the importance of parents modeling healthy behaviors themselves to their children from an early age.^{23,30-31} Encourage the whole family to get involved in healthy living activities not just the child with Spina Bifida.³²
 - Understand that experiencing food insecurity may lead to a poor-quality diet, and have developmental consequences on the child.⁴⁷
 - Highlight that children with Spina Bifida, especially those who are non-ambulatory, undertake low levels of physical activity, and have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D.³³
6. Screening for diabetes (fasting glucose, HbA1c or oral glucose tolerance test) is recommended every two years in children 10 years of age or older (or at the onset of puberty if it occurs at a younger age), and for all children with a Body Mass Index (BMI) over the ≥85th percentile and who have two or more additional risk factors including:³⁶
- family history of type 2 diabetes mellitus (T2DM) in a first- or second-degree relative
 - high-risk ethnicity
 - acanthosis nigricans
 - hypertension
 - dyslipidemia

- polycystic ovary syndrome (PCOS)
- 7. Screening for dyslipidemia (fasting lipid profile) is recommended every two years for children up to 8 years of age with a Body Mass Index (BMI) over the 95th percentile or other risk factors for cardiovascular disease, such as:³⁶
 - family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives
 - history of diabetes, hypertension, or smoking in childhood
- 8. Screening for dyslipidemia (fasting lipid profile) is recommended once for all children ages 9-11 years.³⁶
- 9. Provide guidance on maintaining good bowel health.
 - Explain that increased fiber in the child's diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals.⁵
 - Recommend the same guidelines for daily fiber intake that are recommended for all children:³⁴
 - 1-3 years: 19g
 - 4-8 years: 25g
 - 9-13 years: female—26g, male—31g
 - 14-18 years: female—26g, male—38g
 - Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. (clinical consensus)
 - Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the following 24-hour period daily maintenance fluid requirements calculation:³⁵
 - 100 mL/kg for the first 10 kg body weight
 - + 50 mL/kg for the next 10 kg body weight
 - + 20 mL for every kilogram of body weight over 20 kg
 - Further guidance can be found in the Bowel Function and Care Guidelines.

13-17 years 11 months

Clinical Questions

1. What is the most effective protocol to approach diet and nutrition goals in annual Spina Bifida clinic visits?
2. What are biggest barriers to healthy nutrition for children with Spina Bifida?
3. What self-management skills and resources related to healthy nutrition should be provided for children with Spina Bifida?
4. What is the best way to manage constipation with diet?
5. Are children with Spina Bifida and obesity at higher risk for metabolic syndrome?
6. Should screening for metabolic complications of obesity be performed?
7. Is there evidence to support the role of weight management intervention in the prevention of metabolic syndrome?

Guidelines

1. Conduct annual assessments of weight, height or arm span, and BMI. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. Consider monitoring other measures of adiposity, such as waist circumference.³⁸ (Appendix: BMI and Body Composition Measurements)

3. Conduct annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension. (clinical consensus)
4. Provide opportunities for teens and parents to talk about their priorities and concerns regarding nutrition and weight.
 - Discuss how nutrition can play an important role in helping individuals with Spina Bifida maintain a healthy weight; minimize skin breakdown, and increase activity and endurance.⁷
 - Discuss that children with Spina Bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and who have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D.³³
 - Provide regular opportunities for teens to discuss any concerns with their weight, growth and diet.²⁷ A trusting therapeutic relationship can greatly facilitate an honest and open discussion.²⁸
 - Identify the teen's priorities and negotiate goals that meet those priorities as well as the parent's and clinician's goals.⁴⁸
 - Use a strengths-based approach that highlights their nutritional achievements and celebrates successes.²⁸
 - Discuss with parents, if relevant, that the Body Mass Index (BMI) is an imperfect indicator of health in all young people and especially in children with Spina Bifida due to difficulties measuring height and body composition.²⁹ Instead, show the child and parents the trajectory of the child's weight and height (or other measures of growth and adiposity) on a growth chart as a visual aid. Do not refer to growth cut-offs developed for typically developing children.¹ A steeply increasing trajectory would indicate that overweight or obesity may be a concern and warrant preventative strategies.²⁸
 - Avoid using scare tactics in older children with Spina Bifida. Instead, discuss the following potential negative consequences of gaining excessive weight, as it relates to their individual circumstances:
 - Moving and transferring may become more difficult, which may also reduce independence and self-care activities.⁷
 - Increased pressure on the skin when seated for long periods of time (such as when using a wheelchair) may result in skin breakdown⁴⁹
 - Weight gain alongside existing scoliosis or kyphosis may result in additional breathing problems.⁵
 - Refer clients to National Center on Health, Physical Activity, and Disability (<http://www.nchpad.org>), which provides advice on nutrition and physical activity for persons with disabilities, including Spina Bifida.⁴²
 - Consider referral to a "Healthy Lifestyle" program and/or use a mobile application, while recognizing that few such programs are tailored to individuals with disabilities (clinical consensus).
5. Consider the broader literature for all older children, given that there is little evidence that specifically refers teens with Spina Bifida. For instance:
 - Understand that eating habits generally worsen as children move into the teen years and become more autonomous.⁵⁰
 - Emphasize the positive health benefits of breakfast and eating fruits and vegetables.^{31,51} Skipping breakfast and low fruit and vegetable consumption is common in teens.⁵²
 - Consider that food insecurity and lower socioeconomic status can be related to poorer diets.⁵³
 - Emphasize that the family setting remains important for teens. Parental

modelling, dietary intake, and encouragement are all associated with fruit and vegetable consumption among teens.³¹

6. Discuss opportunities for the older child to participate in nutrition-related activities, such as:
 - Identify the teen's knowledge level about healthy eating habits. (clinical consensus)
 - Encourage the family to identify roles that the older child can play as part of daily life, such as in meal planning, shopping, and food preparation.⁵⁴
 - Encourage older children to select a new healthy food to try, which can encourage broader food preferences.⁵⁵
 - Identify the older child's existing strengths and resources regarding nutrition and how they can be built upon to reach their goals.⁵⁶
7. Screening for diabetes (fasting glucose, HbA1c or oral glucose tolerance test) every two years with a Body Mass Index (BMI) over the $\geq 85^{\text{th}}$ percentile and have two or more additional risk factors including:³⁶
 - family history of type 2 diabetes mellitus (T2DM) in a first- or second-degree relative
 - high-risk ethnicity
 - acanthosis nigricans
 - hypertension
 - dyslipidemia
 - polycystic ovary syndrome (PCOS)
8. Screening for dyslipidemia (fasting lipid profile) is recommended every two years for children with a Body Mass Index (BMI) in the $\geq 85^{\text{th}}$ percentile or other risk factors for cardiovascular disease (family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives, history of diabetes, hypertension, or smoking in childhood).³⁶
9. Provide guidance on maintaining good bowel health.
 - Explain that increased fiber in the child's diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals.⁵
 - Recommend the same guidelines for daily fiber intake that are recommended for all children:³⁴
 - 1-3 years: 19g
 - 4-8 years: 25g
 - 9-13 years: female–26g, male–31g
 - 14-18 years: female–26g, male–38g
 - Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. (clinical consensus)
 - Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the following daily maintenance fluid requirements (24-hour period) calculation:³⁵
 - 100 mL/kg for the first 10 kg body weight
 - + 50 mL/kg for the next 10 kg body weight
 - + 20 mL for every kilogram of body weight over 20 kg
 - Further guidance can be found in the Bowel Function and Care Guidelines.

18 + years

Clinical Questions

1. How do nutrition issues vary by different demographics (e.g., age, geography, level of lesion, economic status, race and ethnicity, gender, and other characteristics) among adults with Spina Bifida?
2. What considerations should be given to nutritional intake when adults with Spina Bifida are taking medications to address other health concerns?
3. What is the best way to manage constipation with diet?
4. Are adults with Spina Bifida who have obesity at higher risk for metabolic syndrome?
5. Should screening for metabolic complications of obesity be performed in adults with Spina Bifida?
6. Is there evidence to support the role of weight management intervention in the prevention of metabolic syndrome?

Guidelines

1. Conduct annual assessments of weight, height or arm span, and calculate BMI. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. However, explain that BMI is not accurate for people with paralysis, who have lowered ratios of fat to lean muscle tissue and that looking at the trajectory over time may be more useful.⁵⁷
3. Consider monitoring other measures of adiposity, such as waist circumference.³⁸ (Appendix: BMI and Body Composition Measurements)
4. Conduct an annual assessment of blood pressure or blood pressure percentiles to monitor for pre-hypertension and hypertension. (clinical consensus)
5. Tailor the discussion around healthy nutrition to the adult's context. Consider that adults and families with lower incomes may experience food insecurity.¹
 - Refer clients to National Center on Health, Physical Activity, and Disability (<http://www.nchpad.org>), which provides advice on nutrition and physical activity for persons with disabilities, including Spina Bifida.⁴²
 - Identify who requires the information about healthy food (i.e. the adult with Spina Bifida, the caregiver, the attendant, the family member, or others).¹
 - Discuss the adult's existing access to cooking options and food preparation areas. (clinical consensus)
 - Involve a social worker or disability organization representative who can speak to adults about available local, state, and federal nutritional benefits such as the Supplemental Nutrition Assistance Program (SNAP), farmer's market vouchers or coupons, and other sorts of food vouchers that are available for eligible individuals.⁵⁸⁻⁵⁹
 - Consider referral to a "Healthy Lifestyle" program and/or use a smartphone application, while recognizing that few such programs are tailored to individuals with disabilities (clinical consensus).
6. Provide information about potential interactions between nutrition in foods and medications.
 - Highlight that some medications, such as corticosteroids, have side-effects including weight gain, increased appetite, high blood pressure and a higher risk of developing osteoporosis or diabetes.⁶⁰
 - Provide information about specific foods and beverages that may interact with medications, such as antihypertensive, anticoagulant, or corticosteroid medications.⁶¹
 - Encourage adults to disclose any prescribed, over-the-counter or complementary and alternative medications they are taking to all of their health care professionals, including pharmacists. (clinical consensus)

- Emphasize the importance of reading medication labels to identify any dietary contraindications.¹ If this is difficult, discuss other ways that the adults could find out about potential contraindications, such as making the medication labels available in a larger font or asking the pharmacists for assistance.⁶²
7. Screening for abnormal blood glucose is indicated as part of assessing cardiovascular risk assessment in adults aged 40 to 70 years who have a BMI > 25 kg/m². Persons who have a family history of diabetes, have a history of gestational diabetes or polycystic ovarian syndrome, or are members of high risk racial/ethnic groups may be at increased risk for diabetes at a younger age or at a lower body mass index. Clinicians should consider screening earlier in persons with one or more of these characteristics.⁶³
 8. Screening for dyslipidemia (fasting plasma profile) is recommended for men ≥ 40 years of age, and women ≥ 50 years of age or postmenopausal. Adults with the following risk factors should be screened at any age: current cigarette smoking, diabetes, arterial hypertension, family history of premature coronary heart disease, family history of hyperlipidemia, high risk ethnicity (individuals of First Nations or of South Asian ancestry), or the presence of rheumatoid arthritis, systemic lupus erythematosus, psoriatic arthritis, ankylosing spondylitis, inflammatory bowel disease, chronic obstructive pulmonary disease, chronic HIV infection, chronic kidney disease, abdominal aneurysm, or erectile dysfunction.⁶⁴

Research Gaps

1. Evidence-based guidelines for weight-management, obesity prevention, and obesity treatment specific to children and adults with Spina Bifida.
2. Accurate assessment of body composition in a standardized and accessible manner.
3. Growth curves and weight classification cut-offs specifically for children and adults with Spina Bifida.
4. Evidence about the energy needs of people with Spina Bifida across the lifespan that is based on their mobility methods.
5. Whether children and adults with Spina Bifida are at higher risk for metabolic syndrome.
6. Whether screening for metabolic complications of obesity should routinely be performed in children and adolescents with Spina Bifida.
7. Evidence for the role of weight management interventions in the prevention of metabolic syndrome.

Appendix: BMI and Body Composition Measurements

Detailed instructions on anthropometric procedures can be found in the National Health and Nutrition Examination Survey (NHANES) [Anthropometry Procedures Manual](https://wwwn.cdc.gov/nchs/data/nhanes/2017-2018/manuals/2017_Anthropometry_Procedures_Manual.pdf), located on the Centers for Disease Control and Prevention website [INSERT LINK:

[https://wwwn.cdc.gov/nchs/data/nhanes/2017-](https://wwwn.cdc.gov/nchs/data/nhanes/2017-2018/manuals/2017_Anthropometry_Procedures_Manual.pdf)

[2018/manuals/2017_Anthropometry_Procedures_Manual.pdf](https://wwwn.cdc.gov/nchs/data/nhanes/2017-2018/manuals/2017_Anthropometry_Procedures_Manual.pdf)]. The following are selected excerpts from the manual.

Weight: Weigh clients with light indoor clothing but without shoes, socks, and/or slippers using a digital chair or wheelchair scale. Ask the person to remove any heavy or excess clothing, braces, or other heavy items. If using a wheelchair scale, ask the person to remove any heavy items from the wheelchair.

Height: Measure to the closest millimeter with a portable stadiometer or length-measuring board. For infants, use an infantometer with a fixed head piece and horizontal backboard, and an adjustable foot piece.

Children under four years of age:

1. Ask the parent or guardian to remove the child's clothes except for diapers or underpants.
2. Ask an assistant to support the child's head while you position the feet. Ensure that the head lies in the Frankfort horizontal plane.
3. Apply gentle traction to bring the top of the head in contact with the fixed head piece.
4. Secure the child's head in the proper alignment by lightly cupping the palms of your hands over the ears.
5. Align the child's legs by placing one hand gently but with mild pressure over the knees.
6. With the other hand, slide the foot piece to rest firmly at the child's heels. The toes must point directly upward with both soles of the feet flexed perpendicular against the acrylic foot piece. To encourage the child to flex the feet, run the tip of your finger down the inside of the foot.

If a person can stand unaided:

1. Assist person to stand with his/her back against a wall mounted height scale (stadiometer) with heels together and eyes looking straight ahead (Frankfort plane).
2. Adjust the horizontal arm of the scale until it sits on top of the person's head.
3. The person's height is indicated by the position of the scale arm.
4. Record measurement in centimeters.

If a person cannot stand, measure the person's length (recumbent):

1. Ask the person to lie on a measuring board, face or front upward.
2. Position the person so feet are touching the footboard together, shoulders are relaxed and touching baseboard, arms at sides, legs straight and knees together, and crown of head is touching headboard.

Where a measuring board is not available and/or for people with severe contractures, measure segmental length:

1. Ask the person to lie on the measuring board (or examination table), face or front upward.
2. Measure from head to neck (just above shoulder).

3. Measure from shoulder to hip.
4. Measure from hip to knee.
5. Measure from knee to ankle bone.
6. And measure from ankle bone to bottom of foot.
7. Add measurements together and record in centimeters.
8. For people with scoliosis, measure both sides of the body.

Other methods to assess height:

Where height/length is challenging to assess, alternative methods have been shown to be useful, including arm span³⁹ and ulna length.⁶⁵

Arm span

1. Extend both arms outward (each arm abducted to 90 degrees).
2. Measure from fingertip to fingertip using a metal rod across the area of the Adam's apple.
3. Record measurement in centimeters.
4. To calculate BMI using arm span length, multiply by 0.95 for those with mid-lumbar lesions (i.e. those who lack gluteus medius and maximus function and/or those who lack foot dorsiflexion) and 0.90 for those with high lumbar/thoracic functional motor levels (i.e. those who lack quadriceps function).⁷ Note that this refers to the functional level of lesion, not the anatomic level.

Ulna length measurement (Segmometer)

1. First, determine which forearm to measure. Measure the forearm that has been least affected by trauma, injury, or progression of weakness when compared to the opposite side. Measure the non-dominant arm if both arms are not affected.
2. Support and position the arm in pronation with 90–110 degrees of elbow flexion.
3. Palpate the distal tip of the ulnar styloid process (the prominent bone of the wrist) and mark lightly with a pen. Make sure to palpate superiorly on the wrist to avoid mistaking the tendon of the extensor carpi ulnaris for the distal tip of the ulna.
4. Palpate the tip of the olecranon (the tip point of the elbow) and place one arm of the segmometer on the olecranon.
5. Place the other end of the segmometer at the tip of the ulnar styloid.
6. Measure in centimeters, to the nearest millimeter (e.g. 19.7 cm) to obtain ulnar length in centimeters.
7. Complete the following calculation for height:⁶⁵
 - Male: height (cm) = (4.605 x ulnar length in cm) + (1.308 x age in years) + 28.003
 - Female: height (cm) = (4.605 x ulnar length in cm) + (1.315 x age in years) + 31.485

Body Mass Index (BMI):

BMI should be calculated using both height and length as kilograms per meter squared (kg/m²) and classified using Centers for Disease Control and Prevention cut-offs (85th–95th percentile=overweight, above 95th percentile=obese).⁶⁶

Occipital head circumference (up to two years of age)¹³

1. Ask the parent/caregiver to hold the baby over their shoulder or sit with the baby in their lap
2. Place the head circumference tape around the child's head so that the tape lies across the frontal bones of the skull; slightly above the eyebrows; perpendicular to

- the long axis of the face; above the ears; and over the occipital prominence at the back of the head.
3. Move the tape up and down over the back of the head to locate the maximal circumference. Tighten the insertion tape so that it fits snugly around the head and compresses the hair and underlying soft tissues.
 4. Measure the circumference to the nearest 0.1 cm.

Waist circumference:

1. Ask the patient to place him/herself in the following manner:
 - Clear the abdominal region.
 - Feet shoulder-width apart.
 - Arms crossed over the chest.
2. It is suggested to kneel down to the right of the patient in order to measure waist girth.
 - Palpate the patient's hips to locate the top of the iliac crest.
 - Draw a horizontal line halfway between the patient's back and abdomen.
3. Place the measuring tape horizontally around the patient's abdomen (*to work comfortably, it is suggested to wrap the tape around the patient's legs and then move up).
4. Align the bottom edge of the tape with your marked point.
5. It is recommended to use a measuring tape with a spring handle, such as the Gulick measuring tape, in order to control the pressure exerted on the patient's abdomen.
 - Gently tighten the tape around the patient's abdomen without depressing the skin.
6. It is suggested to request the patient to relax and breathe NORMALLY (abdominal muscles should not be contracted).
 - Ask the patient to take two or three NORMAL breaths.
 - Measure from the zero line of the tape to the nearest millimeter) at the end of a NORMAL expiration.
7. Note the lesion level and/or any bulky masses, liposuction incision marks or spinal curvature.

Waist circumference in supine

1. Ask the patient to lie down and place him/herself in the following manner:
 - Clear the abdominal region.
 - Arms crossed over the chest.
2. Palpate the patient's hips to locate the top of the iliac crest. If iliac crest cannot be located, measure smallest part of abdomen.
 - Draw a horizontal line halfway between the patient's back and abdomen.
3. Complete steps 3-6 as indicated above.

Skinfold thickness

1. Ask the patient to remove their shirt if comfortable and clear the abdominal region.
2. Begin with right arm placed at a 90-degree angle and request that the patient places his/her arm across the abdomen, with the palm facing inward.

Triceps:

- Along the midline on the back of the triceps of the right arm, determine the midpoint located between the top of the acromial process (top of shoulder) to the bottom of the olecranon process of the ulna (elbow).

- Pinch the skin to create a vertical skinfold with the thumb and forefinger about 0.5 inches from the measurement site.
- Release the calipers on the skinfold three times for 1 second each and record the measurements.

Subscapular:

- Ask patient to place arm behind his/her back
- The skinfold should angle 45 degrees from horizontal, in the same direction as the inner border of the scapula
- Release the calipers on the skinfold three times for 1 second each and record the measurements
- Use the Slaughter equation to calculate the estimated body fat.⁶⁷

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Sleep Related Breathing Disorders

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Introduction

The assessment and management of Sleep Related Breathing Disorders (SRBD) are important clinical and research priorities in individuals with Neural Tube Defects (NTD), specifically in those with Myelomeningocele (MMC).¹

SRBD is common in the general pediatric population with a prevalence of up to 5% in children and 11% in adolescents.²⁻³ In the general adult population, the prevalence of Obstructive Sleep Apneas (OSA) defined by ≥ 5 apnea and hypopnea events per hour of sleep associated with excessive sleepiness is approximately 3-7% in men and 2-5% in women.⁴ Untreated and unrecognized SRBD are associated with significant neurocognitive, psychological, metabolic, immunologic, cardiovascular consequences and even death. Chronic SRBD have been identified as the cause of death in 12.8-16.3% of patients with MMC independent of adjustments for sensory level, motor level and birth head circumference.⁵⁻⁷ Sudden unexplained death during sleep is also described, especially between birth and 19 months of age.⁸

The types of SRBD described in patients with NTDs include central apnea, periodic breathing, obstructive apnea, and central hypoventilation.^{4,11,13,25-28} Central apnea refers to pauses in respiratory effort; periodic breathing refers to series of at least 3 central apneas separated by breaths of no more than 20 seconds; obstructive apneas and hypopneas refer to partial or complete airway obstruction, and central hypoventilation refers to persistent low tidal-volume breathing or bradypnea causing hypercarbia and hypoxemia (central hypoventilation syndrome/ventilatory dysfunction). Patients with MMC also have absent arousal responses to hypoxia and hypercapnia and absent ventilatory responses to hypoxia and hypercapnia.^{6,13-15,27-28} Unfortunately, symptoms alone do not predict treatment outcome.¹⁶

The prevalence of SRBD in MMC has been reported between 62-81%; the prevalence of moderate to severe OSA in this population has been reported to be as high as 20-31%.^{2,13,17} Obstructive events are more likely to occur during rapid eye movement (REM) sleep state. Efforts to quantify the incidence of central apneas are based on small series and case reports and likely represent under recognition / reporting. In children with MMC, risk factors for SRBD include higher spinal cord lesions, brain stem dysfunction resulting from decreased cervicomedullary subarachnoid space, very abnormal Chiari malformations, pulmonary function abnormalities (obstructive lung disease and restrictive lung disease), and disorders of upper airway maintenance.^{7,13-14} Obesity is an independent risk factor for SRBD in all children with an estimated prevalence of 13-78%.³ The combination of higher obesity rates in individuals with MMC and an abnormal brainstem respiratory brainstem control center secondary to their Chiari malformation place this population at increased risk for SRBD.

The clinical course of infants and children with MMC and central respiratory control abnormalities is variable. In infants up to a year, the most common presentations include stridor, apnea, and feeding difficulties. These symptoms may not be present perinatally but can present before 3 months of age.^{4,18} When present in young infants, neurological deterioration may progress rapidly and can result in cardiorespiratory arrest and death.^{18,30} In infancy, central apneas, periodic breathing, hypoventilation, breath holding spells, and cyanosis have been described even after relief of upper airways obstruction with decompression, shunt revision,

and/or tracheostomy tube placement.⁹⁻¹¹ In older children, the most frequently reported symptoms include apnea, blue spells, shortness of breath, snoring, choking, and irritability.¹⁷ Excessive daytime sleepiness is less common in children than in adults.³⁰ Symptoms of SRBD in adults include snoring, witnessed cessation of breathing, gasping or choking at night, excessive daytime sleepiness, impaired cognition, and mood changes.^{1,19} Because adults with NTD are a recognized high-risk population for SRBD, the recent US Preventive Services Task Force (USPSTF) recommendations to not routinely screen for SRBD in adults within the general population do not apply.¹⁹

Little is known about the effect of neurosurgical intervention (hydrocephalus intervention, cervical decompression) on sleep hygiene/SRBD in individuals with NTD. Although cervical decompression of symptomatic Chiari malformation may be effective, this treatment does not always resolve the apneic symptoms in infants.⁴ Persistent central apneas, periodic breathing, hypoventilation, breath holding spells, and cyanosis have also been described after relief of upper airway obstruction with decompression, shunt revision, and tracheostomy tube placement.⁹⁻¹¹

Identification/Tools:

To screen for OSA, the American Academy of Pediatrics recommends that each child be questioned regarding snoring and other signs and symptoms of OSA.³ Several questionnaires are available to screen for sleep disorders such as the Children's Sleep Habits Questionnaire and the Pediatric Sleep Questionnaire (PSQ). These instruments are not specific to NTDs. The PSQ is a valid and reliable instrument that can be used to identify SRBD or associated symptom-constructs (habitual snoring, insomnia, excessive daytime sleepiness, inattentive/hyperactivity, sleep terrors, sleepwalking, nocturnal bruxism).⁶ In adults, validated questionnaires to predict the presence of sleep apnea include the STOP-BANG (Snoring, Tiredness, Observed apnea, High blood Pressure-Body Mass Index (BMI), Age, Neck Circumference, Gender), Berlin, Epworth Sleepiness Scale (ESS), and OSA 50 (Obesity-Snoring-Apnea-Age>50).^{8,20}

In children, clinical evaluation alone (history, physical exam, audio or visual recordings, standardized questionnaires) does not have sufficient sensitivity or specificity to establish a diagnosis of SRBD.^{11,16,20} Specifically, Waters et al. report 83% accuracy and 65% sensitivity in predicting the presence of moderate/severely abnormal SRBD in patients with MMC based on a history of stridor and dysphagia in infancy, a history of apnea or cyanosis, or a high level of spinal lesion. Overnight observed polysomnography (PSG) that includes the measurement of respiratory variables (carbon-dioxide levels, oxygen saturation, sleep state and electromyogram recordings) remains the "gold standard" to identify SRBD but is not readily available in all settings.³ Nocturnal pulse oximetry has been used as an abbreviated testing method to detect moderate to severe SRBD with positive predictive value of 44% and a negative predictive value of 100%.¹³

Because of the low sensitivity of clinical evaluation alone, the high morbidity associated with untreated OSA, and the high incidence of sleep apnea in patients experiencing sudden death, it is recommended that all patients with NTD, whether they are symptomatic or asymptomatic, undergo polysomnography that evaluates for central apneas, hypoventilation, as well as obstructive sleep apneas.

Outcomes

Primary

1. Improve recognition of signs and symptoms of sleep related breathing disorders (SRBD) across the lifespan, recognizing that symptoms important for its recognition in infants will be different than in adults.

Secondary

1. Implement a strategy to identify SRBD in the clinical setting through reliable screening methods that improve timely referral for additional appropriate assessment (polysomnography).

Tertiary

1. Minimize the adverse impact of unrecognized SRBD on physical well-being (including sudden, unexplained death) and neurocognitive function.

0-11 months

Clinical Questions

1. Is there any predictable sequence to cranial nerve dysfunction (is eating affected before facial weakness and/or respiratory regulation) or is each child different?
2. Is there any anatomic (imaging) or physiologic marker that identifies children at greatest risk for SRBD?
3. Do any observed signs/symptoms predict a greater need for a specific intervention (ventricular shunting, foramen decompressions, oxygen supplementation)?

Guidelines

1. Screen for SRBD signs and symptoms in all infants with NTD at each health care maintenance visit using available standardized questionnaires.^{3,12,16,24}
2. Encourage that all symptomatic infants or those with additional risk factor for OSA (high spinal lesion, small cervicomedullary arachnoid space, or severe Chiari malformation) undergo a formal evaluation for SRBD with overnight polysomnography or be referred to a specialist with expertise in sleep-related breathing disorders.^{3,13,16,20,29}
3. Refer all infants with documented SRBD referred to appropriate specialists with expertise in SRBD (pediatric pulmonologist or sleep specialist), neurosurgeon, and/or otolaryngologist) for ongoing management.^{14,17-18}
4. Conduct periodic cardiac evaluations on infants with documented SRBD and hypoxemia to assess for pulmonary hypertension and cor pulmonale.³

1-2 years 11 months

Clinical Questions

1. Is there any predictable sequence to cranial nerve dysfunction (is eating affected before facial weakness and/or respiratory regulation) or is each child different?
2. Is there any anatomic (imaging) or physiologic marker that identifies children at greatest risk for SRBD?
3. Do any observed signs/symptoms predict a greater need for specific interventions (ventricular shunting, foramen decompressions, oxygen supplementation)?

Guidelines

1. Screen for OSA and other SRBD signs and symptoms in all children with NTD at each health care maintenance visit using available standardized questionnaires.^{3,12,16,24}
2. Encourage that all symptomatic children or those with additional risk factors for OSA (high spinal lesion, small cervicomedullary arachnoid space, or severe Chiari

- malformation) undergo a formal evaluation for SRBD with overnight polysomnography or be referred to a specialist with expertise in sleep-related breathing disorders.^{3,11,13,16,20,22}
3. Refer all children with documented SRBD to appropriate specialists with expertise in SRBD (pediatric pulmonologist or sleep specialist), neurosurgeon, and/or otolaryngologist for ongoing management.^{14,17-18}
 4. Conduct periodic, comprehensive cardiac evaluations on children with documented SRBD and hypoxemia to assess for pulmonary hypertension and cor pulmonale.³
 5. Discuss sleep hygiene (expectations, normal variations, and interventions) with parents and caregivers to promote healthy sleep. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. Is there a sufficiently sensitive and specific method (questionnaire, screening test) prior to polysomnography that would support routine screening of children with NTD for SRDB? Is there a clinical profile (signs, symptoms, other risk factors like obesity, hypertension) that would warrant a higher priority referral?
2. Are asymptomatic individuals with NTD really asymptomatic or are they only unrecognized?

Guidelines

1. Recognize that the symptoms of SRBD in children (mouth breathing, a history of delayed growth, features of inattention and hyperactivity) are different compared to adults (snoring and excessive daytime sleepiness are less frequent).^{2,7,9-11,13,16,22}
2. Ask questions related to sleep quality, quantity and other possible symptoms at every visit (at least annually). Standardized screening questionnaires for SRBD in children are useful in clinical settings.^{3,12,24}
3. Further evaluate changes in respiratory status/function.^{3,14,17}
4. Discuss sleep disordered breathing with parents and care providers so they can better observe for early symptoms or changes.⁸

6-12 years 11 months

Clinical Questions

1. Is there a sufficiently sensitive and specific method (questionnaire; screening test) prior to polysomnography that would support routine screening of children with NTDs for SRDB?
2. Is there a clinical profile (signs, symptoms, other risk factors like obesity, hypertension) that would warrant a higher priority referral?
3. Are asymptomatic individuals with NTD really asymptomatic or are they only unrecognized?

Guidelines

1. Recognize that the symptoms of SRBD in children (mouth breathing, a history of delayed growth, features of inattention and hyperactivity) are different compared to adults (snoring and excessive daytime sleepiness are less frequent).^{2,4,7,9-11,13,16}
2. Ask questions related to sleep quality, quantity and other possible symptoms at every visit (at least annually). Standardized screening questionnaires for SRBD in children are useful in clinical settings.^{3,6,12}
3. Further evaluate changes in respiratory status/function.^{3,14,17}
4. Discuss sleep disordered breathing with parents and care providers so they can better observe for early symptoms or changes.⁸

13-17 years 11 months

Clinical Questions

1. Are asymptomatic individuals with NTDs really asymptomatic or are they only unrecognized?
2. What is the effect of SRBD on morbidity and mortality?

Guidelines

1. Use a standardized sleep questionnaire to query patients at each visit (at least annually) because patients are unlikely to discuss sleep-related symptoms spontaneously with a primary care or specialty provider.^{3,15-16,24,26}
2. Recognize clinical findings that may either contribute to or be the result of sleep disordered breathing: hypertension, obesity, and scoliosis.^{1,3,23}
3. Improve patients' awareness of SRBD, its presentation and its adverse impact on quality of life.^{1,6,8,23}

18+ years

Clinical Questions

1. Are asymptomatic individuals with NTD really asymptomatic or are they only unrecognized?
2. Does SRBD increase or evolve with adulthood?
3. What is the effect of SRBD on morbidity and mortality?

Guidelines

1. Use a standardized sleep questionnaire to query patients at each visit (at least annually) because patients are unlikely to discuss sleep-related symptoms spontaneously with a primary care provider.^{3,19-20,24,26}
2. Recognize clinical findings that may either contribute to or be the result of sleep disordered breathing: hypertension, obesity, and scoliosis.^{1,3,23}
3. Improve patients' awareness of SRBD, its presentation and its adverse impact on quality of life.^{1,6,8,23}

Research Gaps

1. Is the frequency of or reasons for sleep disorders in the NTD population truly greater/different than the general population or is this the result of referral/assessment bias?
2. Are these frequency differences related to the presence or degree of Chiari malformation and/or brainstem dysfunction?
3. How do unrecognized sleep disordered breathing disorders contribute to the neurocognitive profile/decline in individuals with a NTD?
4. Is there any anatomic (imaging) or physiologic marker that identifies children at greatest risk for SRBD?
5. Do any observed signs/symptoms predict a greater need for a specific intervention (ventricular shunting, foramen decompressions, oxygen supplementation)?
6. Is there a sufficiently sensitive and specific method (questionnaire, screening test) prior to polysomnography that would support routine screening of children with NTD for SRDB? Is there a clinical profile (signs, symptoms, other risk factors like obesity, hypertension) that would warrant a higher priority referral?
7. Are asymptomatic individuals with NTD really asymptomatic or are they only unrecognized?
8. Does SRBD increase or evolve with adulthood?

9. What is the effect of SRBD on morbidity and mortality?

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Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans

Understanding what services exist for children and adolescents with disabilities can be very confusing; this is particularly true for school-age children and adolescents. This appendix will provide some basic information about these services.

Early intervention services (sometimes called birth to three-year services) are available to infants and toddlers up to the age of 3. Federal law mandates these services under Part C of the Individuals with Disabilities Act (IDEA) of 2004. Infants and toddlers with disabilities may qualify for services if they are experiencing delays in one or more areas of development as measured by appropriate diagnostic instruments and assessments or have a diagnosed physical or mental condition that is likely to result in a developmental delay. If it is determined that a child is appropriate to receive early intervention services, the next step is the development of an Individualized Family Services Plan (IFSP). The IFSP serves as the document and plan for a family's involvement with early intervention. The IFSP includes information about a child's current functioning and needs, but also what the needs of the family may be to best support the child's development. The IFSP is reviewed every 6 months and must be updated with the family's active involvement at least yearly.

Services for preschool age children with disabilities are typically provided by public schools through the development of an Individualized Educational Plan (IEP). Students with disabilities from kindergarten through grade 12 can receive assistance from IEPs or a Section 504 Plan, also known as a 504 Plan.

A 504 Plan refers to legislation of Section 504 of the Rehabilitation Act of 1973, under the supervision of the Office for Civil Rights of the U.S. Department of Education. The Act is a broad federal civil rights law that protects individuals with disabilities against discrimination, including in the school setting. A 504 Plan describes how a child with disabilities will have access to services, accommodations, and modifications to the learning environment to access the general education as adequately as students without disabilities. Disability is described broadly in Section 504. To qualify for a 504 Plan, a student must have (1) a physical or mental impairment that substantially limits one or more major life activities; or (2) have a record of such an impairment; or (3) be regarded as having such an impairment [Section 504 regulatory provision at 34 C.F.R. 104.35]. The definitions of impairment and what is meant by substantially limiting major life activities are quite broad compared to requirements to qualify for an IEP. An evaluation by the school is necessary before it can be determined if a child is eligible for a 504 Plan. However, that evaluation can be informal, with information gathered from a variety of sources, instead of the school performing a formal evaluation as required for an IEP.

The current version of the Individuals with Disabilities Act (IDEA) allows children and adolescents with disabilities to receive special services in school through an IEP. To qualify for an IEP, a child must have one or more of 13 specific disabilities listed in IDEA. Most children with Spina Bifida will qualify under one or more of these, including "multiple disabilities," "orthopedic impairment," "other health impairment," or "specific learning disability." (Note: non-verbal learning disabilities are NOT recognized as one of the specific learning disabilities. Thus, a category like "other health impairment" is preferred.) In addition, the disability must affect the child's educational performance or ability to learn and benefit from the general educational

curriculum, requiring a need for specially-designed instruction, not just accommodations or modifications.

A student cannot have both an IEP and a 504 Plan. If a student qualifies for an IEP, all the support accommodations and modifications which might be provided by a 504 Plan are incorporated into an IEP.

For more information, please review the following materials:

- What is the difference between an IFSP and an IEP?
<http://www.pacer.org/parent/php/PHP-c59.pdf>
- Early Intervention Program for Infants and Toddlers with Disabilities
<https://www2.ed.gov/programs/osepeip/index.html>
- A Guide to the Individualized Education Program
<https://www2.ed.gov/parents/needs/speced/iepguide/index.html>
- Frequently Asked Questions About Section 504 and the Education of Children with Disabilities
<https://www2.ed.gov/about/offices/list/ocr/504faq.html>

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