

Guidelines for the Care of People with Spina Bifida

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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