Preface

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 the Care of People with Spina Bifida 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 included people with Spina Bifida. Research related to adults with Spina Bifida was nearly nonexistent. 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.

Following the publication of the third edition of the Guidelines, there were advances in health care service delivery concepts related to improving the care of children with medical complexity, including Spina Bifida. These concepts are important in ensuring the full implementation of the fourth edition of the Guidelines for the Care of People with Spina Bifida. The first of the delivery concept advances was 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 neighborhood 8 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 the 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.

The fourth edition of the Guidelines for the Care of People with Spina Bifida was the result of three years of planning, literature review and content development by nearly 100 volunteers. These 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 Guideline versions did not have robust coverage of the care needs of adults. Additionally, the fourth version featured 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 of the 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, the fourth edition featured a number of new topics, including Transition and Quality of Life, important to the health and well-being of all people living with Spina Bifida.

The extensive literature review done for the fourth edition of the Guidelines continued to identify that research in Spina Bifida remains limited. Where evidence exists, it was included in the fourth edition. 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 robust. 1-5

As such, the Guidelines 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.

As we move forward, the Guidelines continue to be updated and as needed will continue to feature new topics such as the 2023 addition of the Osteoporosis and Bone Health Guideline.

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 should be supported with additional health care navigation services, along with oral and written information provided in their preferred language.11

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