The SBA's Healthcare Guidelines for People with Spina Bifida

How it Came to Be Use to Families, Youth with Spina Bifida, and Programs



Richard Adams, MD
Division Director,
Developmental Behavioral Pediatrics
UT Southwestern Medical Center
Texas Scottish Rite Hospital

UTSouthwesternMedical Center

SBANT Education Day February 2020 Frisco, TX



The Challenges Before Us All

Quality Healthcare Throughout the Lifespan for People With Spina Bifida

*Access to Healthcare

*Knowledge of the Person with Spina Bifida & Family

*Clinicians and Health
Systems Knowledgeable of
Issues Related to Quality
Care





Healthcare for All with Spina Bifida

Across Ages

Across Cultural Backgrounds





Differing Levels

Differing Needs

Healthcare for All



GUIDELINES FOR THE CARE OF PEOPLE WITH

SPINA BIFIDA

4th Edition 2018



Spina Bifida is the most commonlyoccurring complex congenital birth condition associated with longterm survival.

The Guidelines for Spina Bifida Health Care Services Throughout the Lifespan was published by the SBA in 1990 in its 1st Edition. This was the work of the SBA's Professional Advisory Council (PAC). A 2nd edition came in 1995. The suggestions then were mostly based on contemporary knowledge and consensus opinion.

Guides for Future Research into Spina Bifida

Questions Identified by Consensus Conference

May 9-10, 2003

Washington, D.C.



NIH AHRQ CDC U.S. Dept Ed SBAA

Meeting of about 30 clinicians from about 20 programs / agencies around the country

Purpose: Identify the current scientific evidence related to Spina Bifida Identify research gaps and priorities for future research To foster new directions & funding for research

Guides for Future Research into Spina Bifida

Questions Identified by Consensus Conference

May 9-10, 2003

Washington, D.C.

NIH AHRQ CDC U.S. Dept Ed SBAA

Outcome:

- Much of the published information about Spina Bifida was based on low levels of research quality.
- Research related to adults with Spina Bifida was nearly non-existent
- Directions for research began to emerge.
- A text of our work was published in 2003 summarizing the research review

Evidence-Based Practice in Spina Bifida: a Research Agenda

2003 Greg Liptak, MD Editor

CDC / NIH / AHRQ / U.S. Dept Education SBAA

- 1. Orthopedics
- 2. Neuro-Development
- 3. Integument
- 4. Neurosurgery
- 5. Mobility
- 6. Work and School
- 7. Urology
- 8. Self-Care
- 9. Nutrition
- 10. Behavior

- 11. Women's Issues
- 12. Socialization
- 13. Learning
- 14. Sexuality
- 15. Family
- 16. Independence
- 17. Fetal Surgery

Evidence-Based Practice in Spina Bifida: a Research Agenda

2003 Greg Liptak, MD Editor

CDC / NIH / AHRQ / U.S. Dept Education SBAA

- 1. Orthopedics
- 2. Neuro-Development
- 3. Skin
- 4. Neurosurgery
- 5. Mobility
- 6. Work and School
- 7. Urology
- 8. Self-Care
- 9. Nutrition
- 10. Behavior

- 11. Women's Issues
- 12. Socialization
- 13. Learning
- 14. Sexuality
- 15. Family
- 16. Independence
- 17. Fetal Surgery

GUIDELINES FOR THE CARE OF PEOPLE WITH

SPINA BIFIDA



2018: 4th Edition of the Guidelines for the Care of People with Spina Bifida

Result of 3 years of research, workgroups, writing, editing by over 100 experts in the field across the United States.

Increased focus on the adult with Spina Bifida

Workgroups were assigned topics that became the separate chapters of the ~250 page document that is the latest set of Guidelines.

GUIDELINES FOR THE CARE OF PEOPLE WITH

SPINA BIFIDA

- The recommendations will continually be updated
- These are *guidelines*, not standards of care
- As *guidelines*, these are not legal requirements
- Rather, aides to the clinician for assessment and intervention
- Hopefully will guide patients and families in their journeys



Outline of Sections in the Guidelines

System of Care	
Care Coordination	7
 Health Promotion and Preventive Health Care Services 	19
Prenatal Counseling	27
Transition	33
Psychosocial Context for Self-Management	
Family Functioning	41
Mental Health	50
 Self-Management and Independence 	78
Quality of Life	60
Neuropsychology and Neurosurgery	
Neuropsychology	95
Neurosurgery	107
Mobility, Orthopedics, and Physical Activity	
Mobility	121
Orthopedics	128
Physical Activity	136
Urology and Sexual Health	
Men's Health	148
 Sexual Health and Education 	155
Urology	162
Women's Health	176
Specific Health Issues	
Bowel Function and Care	186
 Endocrine: Puberty and Precocious Puberty 	192
 Endocrine and the Use of Human Growth Hormone 	196
Integument (Skin)	201
 Latex and Latex Allergy in Spina Bifida 	206
 Nutrition, Metabolic Syndrome, and Obesity 	215
 Sleep-Related Breathing Disorders 	235
<u>Appendix</u>	
Early Intervention Services, Individualized Educational Plans (IEP)	
and 504 Plans	243
Contributors	245

Systems of Care

Psychosocial Context for Self-Management

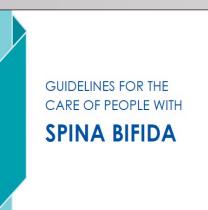
Neuropsychology and Neurosurgery

Mobility, Orthopedics, and Physical Activity

Urology and Sexual Health

Specific Health Issues

Early Intervention and Special Education





Systems of Care

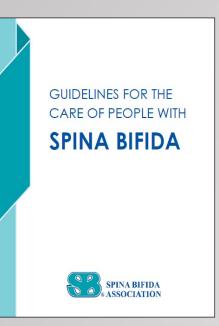
- Care Coordination
- Health Promotion & Preventive Health Care
- Prenatal Counseling
- Transition

Psychosocial Context for Self-Management

- Family Functioning
- Mental Health
- Self-Management & Independence
- Quality of Life

Neuropsychology and Neurosurgery

Mobility, Orthopedics, and Physical Activity



Urology and Sexual Health

- Men's Health
- Women's Health
- Sexual Health & Education
- Urology

Specific Health Issues

- Bowel Function and Care
- Endocrine: Puberty and Precocious Puberty
- Skin
- · Latex and Latex Allergy in Spina Bifida
- Nutrition, Metabolic Syndrome, & Obesity
- Sleep-Related Breathing Disorders

Early Intervention, Special Education, 504 Plans





Concepts supporting the Guidelines and Their Use

- 1. <u>Care Coordination</u> is an essential component of health care delivery
- 2. <u>Patient- and family-centered care</u>: provided via a medical neighborhood with coordinated teambased care.
- 3. <u>Partnership</u> between families, people with spina bifida, health professionals, health systems, and community services.
- 4. Guidelines should serve people with spina bifida through application of the best and most scientifically-based care and treatment throughout their lives.

General Outline for Each Chapter

- **➤** Workgroup Members
- > Introduction
 - ➤ Background history
 - ➤ Review of studies since last Guidelines Edition
 - State of present understanding of research and clinical practices

Latex and Latex Allergy in Spina Bifida

Workgroup Members: Richard Adams, MD (Chair); Kevin Kelly, MD; Sue Lockwood, Allergy and Asthma Network; Suzanne McKee, RN BSN; Candice Walker, PhD

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

General Outline for Each Chapter

- ➤ Workgroup Members
- > Introduction
- **Outcomes**
 - ➤ Maximize & support wellness throughout the lifespan
 - ➤ Reduce & prevent other conditions related to poor nutrition and overweight/obesity,
 - Support development of client/caregiver knowledge and self-direction related management skills and
 - ➤ Self-concept related to nutrition

Nutrition, Metabolic Syndrome, and Obesity

Workgroup Members: Amy C McPherson, PhD, (Chair); Lorry Chen, RD; Joseph O'Neil, MD, MPH; Kerri A Vanderbom, PhD

General Outline for Each Chapter

- ➤ Workgroup Members
- > Introduction
- ➤ Outcomes

roup Members: David B. Joseph. MD. FAC

Workgroup Members: David B. Joseph, MD, FACS, FAAP (Chair); Sharon Baillie, RN, CNC, MN; Michelle A. Baum, MD; Dominic C. Frimberger, MD; Rose Khavari, MD; Rosalia Misseri, MD, FAAP; Stacey T. Tanaka, MD, MS; Hadley Wood, MD; Elizabeth B. Yerkes, MD

Urology

> Research Gaps

- ➤ What is the ability of urodynamic testing to identify individuals at risk?
- ➤ Does early medical (e.g. intermittent catheterization) and medication management based on urodynamic testing prevent upper tract deterioration?
- ➤ How is creatinine influenced by height, weight and mobility status of a patient with Spina Bifida?
- ➤ What degree of renal dysfunction has occurred by the time changes are noted on imaging (i.e., renal scarring in ultrasonography or DMSA)?

General Outline for Each Chapter

- ➤ Workgroup Members
- > Introduction
- ➤ Outcomes
- > Research Gaps
- > References
 - > Literature Review
 - > Selection of best scientific papers that inform clinical practice
 - > Historical of importance
 - > New since last Guideline Edition
 - ➤ Listed Last --- but this was 1st step and very time and labor intensive

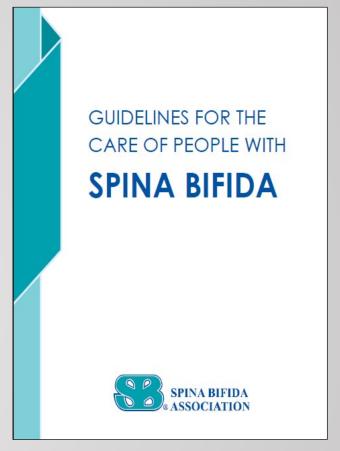
Neurosurgery

Workgroup Members: Jeffrey P. Blount, MD (Chair); Robin Bowman, MD; Mark Dias, MD; Betsy Hopson, MHSA; Michael Partington, MD; Brandon Rocque, MD

- Laurence KM. The natural history of Spina Bifida cystica: detailed analysis of 407 cases. Arch Dis Child 1964; 39: 41-57.
- Bowman RM, McLone DG, Grant JA McLone DG, Grant JA, Tomita T, Ito JA. Spina Bifida Outcome: a 25 year prospective. Pediatr Neurosurg 2001; 34: 114-120.
- Talamonti G, D'Aliberti G and Collice M. Myelomeningocele: long term neurosurgical treatment and follow up in 202 patients. J Neurosurg. 2007; (5 Supple Peds) 107: 368-386.
- Steinbok P, Irvine B, Cochrane DD and Irwin BJ. Long-term outcome and complications of children born with myelomeningocele. Childs Nervous System 1992; 8:92-96.
- Thompson DNP. Postnatal management and outcome for neural tube defects including Spina Bifida and encephaloceles. Prenat Diagn 2009;29: 412-419. Adzick NS, Thom EA, Spong CY et.al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. NEJM. 2011; 364:993-1004.
- 6. Adzick NS, Fetal myelomeningocele: natural history, pathophysiology and in-utero intervention. Semin Fetal Med 2010; 15: 9-14.
- 7. Gupta N, Farrell JA, Rand L, Cauldwell CB, Farmer D. Open fetal surgery for myelomeningocele; a review. J Neurosurg Pediatr 2012; 9:265-273.
- 32. Warf BC and Campbell JW. Combined endoscopic third ventriculostomy and choroid plexus cauterization as primary treatment of hydrocephalus for infants with myelomeningocele: long term results of a prospective intent to treat study in 115 East African infants. J Neurosurg Pediatr .2008; 2: 310-316.
- 33. Stone SSD and Warf BC. Combined endoscopic third ventriculostomy and choroid plexus cauterization as primary treatment for infant hydrocephalus: a prospective North American series. J Neurosurg Pediatr. 2014; 14: 439-446.
- 34. Kulkarni AV, Riva-Cambrin J, Browd SR et.al Endoscopic third ventriculostomy and choroid plexus cauterization in infants with hydrocephalus: a retrospective Hydrocephalus Clinical Research Network study. J Neurosurg Pediatr 2014; 14: 224-229.

General Process Toward Guidelines Completion

- Workgroup Members
- > Introduction
- Outcomes
- > Research Gaps
- > References
- > Workgroup Organization
 - Workgroup Steps
 - > Research
 - ➤ Desired Outcomes
 - > Recommendations (Clinical)
 - ➤ New Research suggestions to meet gaps in scientific knowledge



General Process Toward Guidelines

GUIDELINES FOR THE

SPINA BIFIDA

SPINA BIFIDA

Completion

- Workgroup Members
- > Introduction
- Outcomes
- Research Gaps
- > References
- > Workgroup Organization
 - > Workgroup work on Research
 - ➤ Workgroup work on desired Outcomes, Recommendations, New Research to meet gaps in scientific knowledge
- ➤ Guideline Consensus Among the Multiple Workgroups
 - > Meetings with the CDC
 - > Multiple Phone Conferences



So ... Now that we have it, how do we make use of it?

- Individual with spina bifida & family
- Spina Bifida Centers of Excellence with Sub-Specialty Inter-disciplinary Clinics
- Other clinics where individuals with spina bifida are seen
- Primary care pediatricians, family practitioners, internal medicine, OB-GYN, etc.
- Schools and school nurses
- Universities and university student health clinics
- Academic Centers involved in research

GUIDELINES FOR THE CARE OF PEOPLE WITH SPINA BIFIDA



GUIDELINES FOR THE CARE OF PEOPLE WITH SPINA BIFIDA



Thank You! Questions?

- 1. Systems of Care
- 2. Psychosocial Context for Self-Management
- 3. Neuropsychology and Neurosurgery
- 4. Mobility, Orthopedics, and Physical Activity
- 5. Urology and Sexual Health
- 6. Specific Health Issues
- 7. Early Intervention and Special Education

https://www.cdc.gov/ncbddd/spinabifida/clinical-care-guidance.html

UTSouthwestern Medical Center SBANT Education Day February 2020 Frisco, TX



Guidelines Mobile App

- A new interactive mobile app has been developed by SBA and is currently in the testing phase.
- This app will allow you to search the guidelines to assist you with care
- It will also contain a symptom tracker for you to better track your health
- It will hopefully be launched in the next month
- Take a look at what is coming! https://invis.io/W4W3UPBUC3H