GUIDELINE STATEMENT
This guideline outlines the management of patients with spina bifida as required by the Children’s Rehabilitative Services Program, Arizona Health Care Cost Containment System, State of Arizona. Clinical guidelines are not used within UnitedHealthcare to decide benefit coverage. Benefit coverage decisions are based upon language in the consumer-specific benefit document.

PURPOSE
Clinical Practice Guidelines represent the minimum requirements for providing care for individuals with Spina Bifida. Care and treatment should be provided in a manner that includes adherence to and consistency with the following Guideline.

DEFINITIONS:
Children’s Rehabilitative Services (CRS): An AHCCCS program for children with certain diagnoses which provides services using an integrated family-centered, culturally competent, multi-specialty, interdisciplinary approach.

Spina Bifida (SB): is a neural tube defect that develops within the first trimester of pregnancy. It refers to any defect where the backbone and/or spinal cord do not close entirely.

Mylomeningocele (MM): The most common type of neural tube defect. It is a birth defect in which the backbone and spinal canal do not close before birth. It is another name for spina bifida.
Chiari II malformation: the downward displacement of the cerebellum, elongation and upward displacement of the medulla and fourth ventricle, dysgenesis of the corpus callosum, a small posterior fossa.

Hydrocephalus: a condition in which the primary characteristic is excessive accumulation of CSF in the brain. The excessive accumulation of CSF results in an abnormal widening of the ventricles. This widening creates potentially harmful pressure on the tissues of the brain.

Multi Specialty Interdisciplinary Clinic (MSIC): The Specialty Medical Home for the members with diagnoses as designated by the Arizona Administrative Code (AAC) R9-7-202 (R9-22-1303, 10-1-2013).

I. PROCEDURAL GUIDELINES for POLICY COMPLIANCE

A. CRS Enrollment:
Children diagnosed with Spina Bifida must be seen at a CRS Multispecialty Interdisciplinary Clinics (MSIC) site with a Spina Bifida Clinic.

B. Interdisciplinary Team Membership:
The following Team Members must be present during MSIC sites and team conferences to review the patient information and determine the need to see the patient at a MSIC site. The Team Members must be available for inpatient consultation or coordination of care with inpatient staff:

- Child Psychologist and/or Neuropsychologist
- CRS member / Caregiver
- Genetic Counselor/Nurse
- Neurosurgeon
- Nurse with expertise in Bowel and Bladder Care
- Nutritionist
- Occupational Therapist
- Orthopedist
- Pediatrician or nurse practitioner
- Physical Therapist
- Primary Care Physician (Invited)
- Registered Nurse Coordinator
- Social Worker
- Urologist

C. Available Personnel:
The following personnel must be available to the member at the MSIC Spina Bifida Clinic:

- Advocate
- Child Life Specialist
- Educator
- Orthotist
- Skin / Wound Specialist (may be RN/LPN with additional expertise in Wound Care)
- Translator

D. Consultative Personnel:
The MSIC site must have access for consultation to specialists including, but not limited to the following:
• Audiologist
• Cardiologist
• Endocrinologist
• Gastroenterologist
• Geneticist
• Nephrologist
• Ophthalmologist
• Otolaryngologist
• Pediatric Neurologist
• Physiatrist
• Plastic Surgeon
• Psychiatrist
• Pulmonologist

E. Spina Bifida Team/Staff Meetings:
Team and staff meetings will be held based on the age of the patient and their diagnosis. At a minimum the following will occur:

1. Interdisciplinary Team Meetings: review and planning meetings (member specific meetings) are to be held at least once a year. Based on the individual impairment, the Team may determine that once every two years is appropriate.

2. MSIC Spina Bifida Team meetings will occur annually to focus on issues of team management and improvement in member experience and outcome. The team should show that there has been continuing education regarding the care and treatment for persons with myelomeningocele. Education may occur in off site meetings, in conjunction with other nationally accredited training. The goal of the requirement is to have the MSIC Spina Bifida Interdisciplinary Team to show continued education for their subject matter expertise for Spina Bifida.

F. Lead Physician Specialists:
Qualifications: The Lead Physician Specialist should be a pediatrician with experience and expertise in Spina Bifida

G. Outreach Clinics:
Outreach Clinics are designed to provide a limited specific set of services including evaluation, monitoring and treatment in settings closer to the family than a MSIC site. Major treatment plan changes must be communicated to the MSIC site. Members with Spina Bifida may be seen in other specialty clinics including Orthopedic, Urology, Neurosurgery and Neurology outreach clinics. Outreach clinic records must be provided to the MSIC site serving the member for effective coordination of care to aid in team recommendations and treatment.

H. Facilities & Services:
1. Quality and consistent Radiology services will be available to effectively monitor changes in spinal curve—Full service neuroradiology and urodynamic labs will be available.
2. Equipment and expertise to measure height and weight.
3. Access to the pharmacy.
4. Latex-safe environment.

I. Radiation Exposure:
Care should be taken when ordering radiology studies to consider cumulative radiation exposure to the child. The technique that produces the best result with the least radiation exposure should be utilized.
G. Community Based Services not provided by CRS:
Community based services means all local services including provider agencies, schools, private
physician offices, hospitals, and/or any other local setting.
The following community based services may be provided from a community based setting:
- Lab Services
- Nutrition Services
- Occupational Therapy
- Pharmacy Services
- Physical Therapy
- Radiology Services

II. GUIDELINES FOR MANAGEMENT AND TREATMENT OF SPINA BIFIDA

The purpose of this guideline is to promote a uniform level of care at CRS MSIC sites for members
with Spina Bifida and to provide a general framework for good patient care. The relevance to specific
situations will depend on individual variations in clinical course and professional judgment. In
addition, this document should serve as a tool to assess programs, secure resources needed to
enhance patient care and education, and guide the future development and treatment of Spina Bifida.

A. Goals:
1. To maintain maximum individual functioning and member ability to monitor for signs of
deterioration in motor functioning, urinary and bowel continence, central nervous system
functioning and skeletal alignment.
2. To improve quality of life for members with SB

B. Objectives:
1. To maintain a network with specialists able to anticipate and treat SB related physical and
psychosocial problems for effective management and outcomes. This implies easy
accessibility to all team members experienced with the comorbidities and educational needs
of Spina Bifida. Specialists must be able to provide accurate and timely diagnosis as well as
be knowledgeable of appropriate treatments for these conditions. This will allow appropriate
community/social integration including transition to adulthood. Nurses at the MSIC must also
be experienced or have additional training to be competent in supporting the members
treatment and educational needs.
2. Services will be provided in a multispecialty, interdisciplinary clinic in a member centric, family
friendly, culturally sensitive manner.

See Guidelines for Spina Bifida Health Care Services Throughout the Lifespan; Spina Bifida
Association of America, Professional Advisory Council, June 2006, or updates as available for
monitoring/clinic scheduling (www.spinabifidaassociation.org)

III. Orthopedic Management and Treatment

A. Selecting Surgical Candidates for Children With Spina Bifida with Hip Dislocation
Nationally, the trend is to not be aggressive in the treatment of dislocated hips in patients with
Spina Bifida. The Spina Bifida Association stated "No evidence has documented that surgery is
advantageous for function or comfort. Pain is apparently more common in children who have had
surgery. Only if unilateral dislocation in low level Spina Bifida (L5-S1) does reduction of dislocated
hip potentially offer improved gait and/or pain resolution."
1. Children with low level SB (L5-S1) have fairly normal hip function. Children with this level of SB should be treated in a similar fashion as children without Spina Bifida with regards to hip dislocation. If a child with low level SB has a unilateral hip dislocation it will lead to a functional impairment and needs to be treated. The prognosis for treatment is close to an otherwise normal child with hip dislocation.

2. There is controversy for treatment in children with mid-level SB (L3-L4). At this level of spinal defect there is not an intact gluteus medius or hip extensors and as a result the hip tends to dislocate. It is unclear in the medical literature as to whether these should be reduced. Some orthopedists advocate tendon transfers to keep the hip reduced, but this has fallen out of favor. Current orthopedist preference is to not perform this procedure. If the dislocation is bilateral it should probably not be treated. If it is unilateral and if it interferes with the child’s functional ambulation then some would consider treating it with surgical intervention. Children with mid-level SB should be treated for the occasional unilateral dislocation that has a reasonable prognosis without treatment.

3. Another controversy is in the children who have different levels of SB, such as an L5 on one side and an L2 on the other. The treatment of the hip dislocation with the side L5 SB should be reduced. The hip on the side with L2 level SB will be dislocated but functional. It is very difficult to get the hip on the side with L2 level SB to stay reduced. In this scenario, surgery would not be indicated for this side.

4. In general, children with high level SB (thoracic or L2) should rarely have an indication for reduction of the hip. It generally does not help the child and frequently fails. As a general guideline, children with a high-level SB should not be treated surgically.

5. Pain: In the rare instance where there is pain, then consideration for surgical intervention can be made. Sometimes the intervention is to reduce the hip, sometimes it is to do a valgus osteotomy to allow it to dislocate further. In a non-ambulatory child a resection of the proximal femur may be an option.

B. Scoliosis

1. CRS Spina Bifida Scoliosis Clinic:
   a. Members diagnosed with scoliosis, kyphosis, or other spinal related conditions must be enrolled in a MSIC site scoliosis clinic. Based on the recommendations of the interdisciplinary team, the member may be seen in MSIC site orthopedic clinics and orthopedic outreach clinics. Information from the MSIC site or outreach orthopedic clinics must be provided to the MSIC site spinal deformities clinic. For members with multiple CRS diagnoses involved with multiple teams, the appropriate members of each interdisciplinary team shall review the patient status and refer to the Scoliosis or Orthopedic Team when appropriate for treatment related to the spinal deformity is needed.

2. Members with myelomeningocele and scoliosis should attend an MSIC Scoliosis or Orthopedic clinic at least once every 6 months unless there is a curve of less than 15 degrees and the member is skeletally mature and then the MM scoliosis clinic should be attended at least once every two years.

3. Special Equipment:
   Quality and consistent Radiology services will be available to effectively monitor changes in spinal curve

IV. Neurosurgical Management and Treatment
A. Chiari Malformation and hydrocephalus. Many children with spina bifida develop hydrocephalus after closure of the myelomeningocele in conjunction with their Chiari malformation. Newborns need to be aggressively monitored for evidence of progressive hydrocephalus and need of a VP shunt. Members and families must be taught signs and symptoms of shunt malfunction or infection, as well as for worsening of Chiari malformation and the urgency for being evaluated by a neurosurgeon.

B. Tethered Cord Deterioration in lower extremity function, changes in urinary tract function, progressive scoliosis, or pain suggest the possibility of a tethered cord, which may occur at any time, but with growth spurts may have increased frequency. Members and families should be taught these signs of worsening function to bring to the attention of the appropriate specialists for evaluation.

V. Urological Management and Treatment
A. Neurogenic bladder: Children with SB frequently have trouble with storage or emptying bladder and sexual dysfunction. The level of the spinal lesion, and the neurologic examination cannot adequately predict the type or severity of urinary tract dysfunction. This is because the spinal cord level at which the bladder is controlled is below the spinal roots directing lower extremity function. For this reason routine renal ultrasound, voiding cystourethrogram (VCUG), cystometrogram, and electromyogram are recommended at birth, ages three to four months, one year, around two to three years of age (because this is a typical time for toilet training), and every two years thereafter.

A. Goals for urologic management of SB are:
   a. preserving renal function and
   b. maintaining independent continence of bowel and bladder at a developmentally appropriate age.

B. Bowel and Bladder Training
Bowel and bladder training are an integral part of management for members with spina bifida. Early clean intermittent catheterization is renal protective. Bowel programs allow improved socialization and member control over their condition. The MSIC has a nurse with expertise in bowel and bladder training.

VI. Dermatologic Management
A. Wound Prevention and Management
Given the variable sensation depending on the level of the SB, daily skin checks and providing information on how to prevent skin wounds is an important function of the SB team.

VII. Other
A. Osteoporosis and fracture prevention/awareness.
Education of members and family regarding risks for osteoporosis and early fracture identification beneath the level of SB impairment aids in improved treatment.

B. Obesity prevention
Excessive weight has been associated with poor outcomes in function and mobility for members with SB. Education in early childhood for the importance of healthy diet and appropriate food portions in combination with exercise plays an important role in maintaining function.

C. Latex Allergy:
   1. Persons with Spina Bifida are at high risk of latex allergy. Latex allergy can be life threatening. The Spina Bifida Association(SBA) states “all people with Spina Bifida
should be considered at high risk for having an allergic reaction to natural latex. They should avoid latex products right from birth. Things made of silicone, plastic, nitrile or vinyl can be used instead.

2. The MSICs will provide Spina provide latex free equipment at the clinics. The team will educate the members and their families about the risks for latex allergy. Families should be taught to advise schools, day cares, camps, visitors and anyone else who is involved with the person who is allergic about latex allergies.

3. According to the SBA, “Those who had a reaction to latex should:
   a. wear a medic-alert bracelet or necklace;
   b. carry auto-injectable epinephrine; and
   c. carry sterile non-latex gloves and other non-latex medical items for emergencies.”

Bibliography:


Foster, Mark R. MD, PhD, Emedicine from WEBMD, November 11, 2004.

Kolaski, Kat MD, Myelomeningocele, November 6, 2004.


Sgouros, Spyros MD, FRCS (SN) (Glasg), Management of Spina Bifida, Hydrocephalus and Shunts, November 6, 2004.

Spina Bifida Association, © 2006, Guidelines for Spina Bifida Health Care Services Throughout the Lifespan.


Children with Spina Bifida: Key Clinical Issues; Adrian D. Sandler MD; Pediatric Clinics of NA, 57 (2010) 879-892. doi:10.1016/j.pcl.2010.07.009

Swaroop, Vineeta T, MD, Orthopedic issues in myelomeningocele (spina bifida), UpToDate, literature through Octobor 1, 2012.
