Guidelines for the Care of People with Spina Bifida 1-2 years 11 Months

Care Coordination 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.
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.
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.
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 11 records from the medical home, such as immunizations, growth charts, developmental screenings, and other materials.
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)

Health Promotion and Preventive Health Care Services Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations, vision and hearing screens).
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. (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, chronic urinary issues, problems with shunts, achieving an inclusive environment, overweight/obesity risk, importance of physical and recreational activity,1 managing unexpected changes in function, keeping regular medical appointments, and pain. (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. (Family Functioning Guidelines)

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

Family Functioning Guidelines

1. Provide support and ongoing counseling as needed to parents, the child, and siblings. (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).
3. Promote effective parenting techniques or provide referral for such services.
4. Provide anticipatory guidance for parents regarding possible behavioral challenges and autonomy needs in children with Spina Bifida and their siblings. (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.

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.

Mental Health Guidelines

1. Address developmental concerns and optimize typical child development by building on resilience, resources, and supports. (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.
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.
5. Provide additional age-appropriate information about Spina Bifida as the child grows.
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/)

Quality of Life 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)

Self Management and 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)

Neuropsychology 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. (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.

Neurosurgery 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.
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.(clinical consensus)

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. (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. Make sure parents are aware that all children who have the potential to walk may have some delay in achieving this milestone.
4. Use appropriate bracing to assist weak muscles and protect the lower limbs from torque and shear forces.
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)

Orthopedics 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 casting or bracing.
4. Consider tendon releases/transfers for unbalanced foot deformities such as the calcaneus foot 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.
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)

Physical Activity 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. (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.
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).

Sexual Health and Education 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.
4. Explore the parent's expectations regarding their child's sexual development.
5. Explain that sexual exploration is a normal and healthy part of early childhood development.
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.

Urology 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: (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.

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. (clinical consensus)

Bowel Function and Care Guidelines

1. Discuss toilet training and habit training with parents.
2. Establish goal of working toward bowel continence.
3. Focus on fiber, fluids, exercise, and timed bowel movements after meals.
4. Consider two-pronged approach of oral and rectal interventions to meet the goal of bowel continence without constipation.
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.
6. Use barrier creams to protect perineal area from breakdown as needed.
7. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus

Endocrine: Puberty and Precocious 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. (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. (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)

Endocrine: Short Stature and the Effects of the Human Growth Hormone Guidelines

1. Take frequent and accurate weight, length, and occipital frontal circumference measurements during infancy and early childhood.
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. (Nutrition Guidelines)
4. Discuss issues surrounding growth of children with Spina Bifida with the family. (clinical consensus)

Integument (Skin) Guidelines

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

Latex and Latex Allergy 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. (Appendix 1)

3. Encourage careful parental observation of latex avoidance.
4. Encourage the child with a history of latex allergy to wear a medical identification bracelet showing allergy to latex. (clinical consensus)

Nutrition, Metabolic Syndrome, and Obesity Guidelines

1. Assess weight and height at every clinical encounter. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. Measure occipital frontal head circumference until two years of age. (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.
    - Recognize that children with Chiari malformation may have sensitivity to different food textures.
    - Consider that adults and families with lower incomes may experience food insecurity.
    - 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.
    - 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. 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. 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.
    - 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).
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. A trusting therapeutic relationship can greatly facilitate an honest and open discussion.
    - Partner with parents to identify and address specific challenges that the family is facing.
    - Discuss that poor eating habits and reduced activity may lead to obesity, constipation, skin breakdown, osteoporosis, anemia, and other problems. 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).

● 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.

● Highlight the importance of parents modeling healthy behaviors themselves to their children from an early age. Encourage the whole family to get involved in healthy living activities, not just the child with Spina Bifida.

● 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.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.

● Recommend the same guidelines for daily fiber intake that are recommended for all children:

- 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:

- 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.

Sleep Related Breathing Disorders 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.

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. 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.
4. Conduct periodic, comprehensive cardiac evaluations on children with documented SRBD and hypoxemia to assess for pulmonary hypertension and cor pulmonale.
5. Discuss sleep hygiene (expectations, normal variations, and interventions) with parents and caregivers to promote healthy sleep. (clinical consensus)