TRANSITION
SPINA BIFIDA HEALTHCARE GUIDELINES
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Overall Outcomes:
**Primary:** Maximize and support independence in self-care, daily living responsibilities, education/vocation, and social engagement for individuals with spina bifida throughout life.

**Secondary:** Promote uninterrupted access to comprehensive healthcare services, educational/vocational opportunities, and community supports to promote health, development, and independence throughout life.

**Tertiary:** Provide patient-centered and coordinated chronic condition management throughout the process of transitioning from pediatric to adult care, including orientation to adult healthcare services, preventative health strategies, self-management support, and adaptive resources.

OUTCOMES
- Infancy
  - Clinical Questions
    1) How can a patient’s probable trajectory regarding future independence be identified?
    2) What strategies have been successful in enhancing self-care skills?
  - Guidelines
    1) During routine infant visits, healthcare providers should discuss realistic expectations for optimizing independence throughout the lifespan with families (1).
      - Survival to adulthood for individuals with spina bifida now exceeds 85% due to improvements in medical and surgical management (2).
      - Individuals with higher lesions, above L2, and hydrocephalus are more dependent regarding sphincter control, mobility, self-care, transfers, social cognition, communication, employment, and independent living as adults (3,4). However, compromised motor function does not prevent positive attitude towards well-being for adults with spina bifida. There is a relationship between everyday math and reading ability and quality of life and thus maximizing math and reading skills should be encouraged beginning in early childhood (4).
2) Families should be connected to parent groups and if possible, adults with spina bifida for encourage and mentorship.

3) Ensure regular follow up for spina bifida specific and general health and developmental screenings. Provide referrals to appropriate services as needed to support patient developmental progress.
   - Access to SB outpatient multidisciplinary care can lead to more positive health outcomes (6).
   - Assessing for age-appropriate skills throughout childhood and providing intervention and support for unmet developmental skills is crucial to promoting the best outcomes for adult independence (7).
   - Individuals with SB tend to be 2–5 years delayed in developing autonomy skills compared with their typically developing peers so individualized goals should be emphasized (8).
   - A framework for clinical care to address functionality includes the following domains: (1) comprehensive in addressing physical function and activity participation, (2) coordination with in a multidisciplinary team to coordinate goals/treatment plan, (3) longitudinally-anticipatory guidance, prevention, and transition planning (9).

☐ Toddler
  - Clinical Questions
    1) How can a patient’s probable trajectory regarding future independence be identified?
    2) What strategies have been successful in enhancing self-care skills?
  - Guidelines
    1) Provide anticipatory guidance regarding developmental needs for toddlers (such as exploration of environment, participation in routines, and age-appropriate choices).
    2) Promote engagement in age-appropriate activities to optimize functional and social development. If children are developmentally delayed, refer to appropriate therapies and early intervention.
    3) Refer to community resources that promote independence and peer inclusion such as early education programs and play groups involving children with and without special needs.
    4) Encourage families to expect participation in daily life activities, including picking up toys, cleaning up, and imitative housework.
    5) Teach families to regularly offer age-appropriate choices such as choosing between two articles of clothing, two books to read, etc.
    6) Families should be connected to parent groups and if possible, adults with spina bifida for encouragement and mentorship.
    7) Ensure regular follow up for spina bifida specific and general health and developmental screenings. Provide referrals to appropriate services as needed to support patient developmental progress (6,7,9).

☐ Preschool
 Clinical Questions
1) How can a patient’s probable trajectory regarding future independence be identified?
2) What strategies have been successful in enhancing self-care skills?

 Guidelines
1) Provide anticipatory guidance regarding general developmental needs for preschoolers (such as play with peers, routines, and discipline expectations).
2) Refer to community resources that promote independence and peer inclusion such as early education programs and play groups involving children with and without special needs.
3) Encourage family educational opportunities (counting and colors at the grocery store and reading to children nightly).
   - Family emphasis on intellectual and cultural activities promotes language performance (10).
4) Encourage families to expect participation in daily life activities, including such things as picking up toys and age/ability appropriate housework.
   - Family influences on children’s functions, activities, and participation have become increasingly recognized as salient modifiers on developmental trajectories (10).
5) Involve the child in daily self-care routines such as bladder catheterization, bowel regimens and skin checks. Normalize daily routines and start to teach the steps using pictures and songs (for example with cathing, “This is the way we wash our hands...This is the way we get out supplies...”).
6) Review expectations for appropriate behavior with families. Children with spina bifida may have delayed functional and social skills, but should be expected to maximize their age-appropriate developmental milestones (11). Praise appropriate behavior such as being helpful or kind (12). Follow through on stated consequences for inappropriate behavior and encourage the child until the appropriate behavior is achieved.
7) Discuss diagnosis with the child using age-appropriate language and pictures. Teach scripted responses to peer questions regarding diagnosis.
8) Families should be connected to parent groups and if possible, adults with spina bifida for encouragement and mentorship.
9) Ensure regular follow up for spina bifida specific and general health and developmental screenings. Provide referrals to appropriate services as needed to support patient developmental progress (6,7,9).

 School age
 Clinical Questions
1) How can a patient’s probable trajectory regarding future independence be identified?

2) What strategies have been successful in enhancing self-care skills?
   o Guidelines
   1) Assess for self-care ability and have individualized, step-wise goals to increase independence in the bowel, bladder, skin, and mobility care. Parents need to be supported as self-management coaches.
      o There should be a comprehensive assessment of self-management knowledge, skills, strengths, goals, and areas that need support as well as an individual or group approach to addressing self-management needs via tailored interventions (13).
      o Children with spina bifida need more parental guidance than their peers who are typically developing. They need particular guidance to overcome process deficits—to learn not only how to do things but also how to get things done (14).
      o Children may view themselves as more responsible for medical management than their parents. Medical adherence is supported by low family conflict and when parents are responsible for medical tasks. However, efforts should be made to help parents transition their management role to their children as developmentally appropriate (15,16).

2) Children should be expected to catheterize, perform bowel routines, check skin, manage adaptive equipment (wheelchairs, orthotics, crutches, and walkers), and do self-hygiene independently by middle school to promote age-appropriate development. Families should begin with hand over hand techniques and shared responsibility for bladder, bowel, and skin in early elementary school. By middle school, children with spina bifida should be the primary managers of completing routine tasks and remembering to do them on time (phone or watch alarms and posted reminders may be necessary). By middle school, parents should take a more supervisory role to ensure that these essential tasks are being completed (for example, have children check of their completed daily tasks and review with parents and have parents check techniques approximately once per week).

3) Encourage family educational opportunities (math facts and reading with children nightly).
   o Family influences on children’s functions, activities, and participation have become increasingly recognized as salient modifiers on developmental trajectories (10).

4) Children should participate in age and ability appropriate household chores.

5) Discuss diagnosis with the child using age-appropriate language and pictures. Teach scripted responses to peer questions regarding diagnosis.

6) Routinely assess for learning difficulties in school and with self-care, refer for neurocognitive assessment and supportive therapies for identified skill deficits including school evaluations for individualized education plans (IEP) for educational
modifications and 504 for physical modifications.
  o Generally, children with spina bifida demonstrate less socially competent behaviors (e.g., less involvement in the activity, more off-task behavior, explanation and clarity of thought), less social dominance, less promotion of dialogue, and less collaboration as compared with their peers who are typically developing. Relative strengths included eye contact, openness to the other’s thoughts, and listening skills. Language fluency and attention support social functioning (17).

7) Assess peer relationships and socialization (17,18). Encourage involvement in community activities including spina bifida groups, camp, school activities, adaptive sports, and volunteering.

8) Emphasize positive attitudes and self-empowerment while setting realistic goals to increase skills and responsibilities (13,14). Praise small achievements and demonstrate patience and encouragement when learning new skills.

Teenage

o Clinical Questions

Primary Outcome
1) How can a patient’s probable trajectory regarding future independence be identified?
2) What are barriers and facilitators to participating in emerging adult milestones for individuals with SB?
3) What strategies have been successful in enhancing self-care skills?

Secondary Outcome
1) What are patient centered perceptions of a successful transition experience?
2) What are the systems level barriers to successful transition and strategies that have effectively mitigated them?
3) What are the best strategies to find and engage adult providers?
4) What are the essential elements for transition preparation and hand-off to promote uninterrupted access to care for individuals with SB?

Tertiary
1) Do individuals with spina bifida prioritize learning self-management skills, becoming independent, and setting their own goals?
2) What are the key readiness parameters for patients with spina bifida that can be measured over time?
3) What are the preventative and chronic condition management considerations in the transition age group?
4) What are examples of successful transition models of SB?

o Guidelines
1) Assess for self-care ability and have individualized, step-wise goals to become more independent in the bowel, bladder, skin, and mobility
Becoming independent in the monitoring and management of spina bifida care is a salient milestone and a necessary precursor to achieving the more “typical” milestones of emerging adulthood. Once a young adult is proficient in medication management, prevention of secondary conditions (pressure ulcers and urinary tract infections), and addressing accessibility challenges in the community, he or she may be better positioned to live independently and obtain employment (18,19).

2) Emphasize positive attitudes and self-empowerment while setting realistic goals to increase responsibilities. Praise small achievements and demonstrate patience and encouragement when learning new skills. Encourage teens to ask questions and be their own advocate.
   ○ Youth with spina bifida did not achieve as many emerging adult milestones as their typically developing peers by age 18-19 years. Emerging adults with spina bifida are less likely to leave home, attend college, maintain employment, and have romantic relationship experience (20).

3) Teens should be expected to be the primary managers of their self-care routines including bladder, bowel, and skin checks. Teens should be able to explain their diagnosis to healthcare providers and peers. Teens should learn to identify signs and symptoms of health problems such as urinary tract infections, constipation, and skin ulcers. Teens should meet with their health care providers independently for some portion of the visit around age 14. Parents/caregivers should still have a supervisory roll to ensure that essential tasks are completed and that teens can identify problems and take appropriate action steps.

4) Encourage involvement in community activities including spina bifida groups, camp, school activities, adaptive sports, and volunteering.
   ○ Teens engaged in required helpfulness were more resilient and felt more empowered and confident about themselves (12).

5) Assess for learning difficulties in school and with self-care, refer for neurocognitive assessment and supportive therapies for identified skill deficits.
   ○ Executive functioning (attention, working memory, problem solving and planning), rather than intelligence, memory and word production, is significantly associated with quality of life for young adults with spina bifida (21).

6) Encourage the teen to participate in IEP planning that addresses transition skills (mandated at age 14). Refer for vocational counseling through school or the Department of Vocational Rehabilitation.
   ○ Enablers for the school-work transition include professional support, social support and school accommodations. Barriers include lack of education and information on finances, housing and transportation, discrimination and stigma, and challenges coping with spina bifida at school and work (15).

7) Assess individual and system barriers to access to care and chronic condition management during the transition from
pediatric to adult health care (responsibility for health management, advocacy, assertiveness, insufficient adult services, and insurance).

- In general, health of individuals with spina bifida declines during adolescence and early adulthood due to multiple factors: lack of coordinated health care, decreased formal social involvement as individuals leave school—with lower levels of fitness and possible depression from isolation—and deterioration from neurologic complications (like tethered spinal cord) that are not being addressed in a timely fashion (12). Annual hospitalizations for adults with spina bifida are 12-20 time higher than their peers (22).

- Chronic condition sequelae need to be considered throughout transition: including monitoring for neurosurgical changes, bladder care, reproductive and sexual health, musculoskeletal/orthopedic concerns, mobility, bowel care, cognitive concerns, and social/vocational concerns (23).

- Decreased mobility is associated with a trend toward decreased general health and vitality. Living alone trended toward decreased emotional well-being and mental health. Medical outcomes may not correlate with quality of life issues (24).

- Barriers to transition identified by patients: not having early planning with flexibility around the individual's developmental level, not addressing financial/employment issues, finding access to health care providers who take care of adults with spina bifida, and parents not giving up control of care/patients not able to make decisions for themselves (23,24).

- Five key elements for transition have been identified: early preparation, flexible timing based on developmental level and health status, coordination of care, transition clinic visits, and identifying health care providers interested in taking care of adults with disabilities (25).

8) Use electronic health care records and internet resources to create medical summaries and care plans, communicate patient-centered health education, and support goals and interventions to increase self-management skills (16,26). This planning should begin in early adolescence, ages 12-14 (27).

9) Assess access and ability in using public and private transportation. Provide resources for adaptive public transportation. If the teen cognitively able drive, encourage enrollment in driver's education and referring for adaptive driving as needed based on mobility.

10) Discuss sexual health including sexuality, contraception (including latex allergy precautions), relationships, fertility and childbearing, genetic counseling, and folic acid supplementation before conception. Refer for subspecialty care as needed.

11) Assess mental health and family/peer relationships. Refer for mental health care as needed.

- Anxiety and depressive symptoms are common for adolescents and young adults with spina bifida. Family support can decrease anxiety. Increased self-management ability can decrease depression (28).
Adult

- Clinical Questions

Primary Outcome
1) What are barriers and facilitators to participating in emerging adult milestones for individuals with SB?
2) What strategies have been successful in enhancing self-care skills?

Secondary Outcome
1) What are patient centered perceptions of a successful transition experience?
2) What are the best strategies to find and engage adult providers?
3) What are the essential elements for transition preparation and hand-off to promote uninterrupted access to care for individuals with SB?

Tertiary
1) Do individuals with spina bifida prioritize learning self-management skills, becoming independent, and setting their own goals?
2) What are the preventative and chronic condition management considerations in the transition age group?
3) What are examples of successful transition models of SB?

- Guidelines

1) Assess for self-care ability and chronic condition management including ability to detect health problems and take appropriate action. Create individualized, step-wise goals and action steps to optimize self-management independent for bowel, bladder, skin, and mobility care. Refer for health education and home health care services to promote independence as needed.

2) Regularly evaluate and monitor cognitive function as it underpins decision-making, self-management, employment, independent living, and socialization (4,19). A change in cognitive functioning may also indicate shunt malfunction or sleep apnea.

3) Encourage social and community engagement including involvement in spina bifida groups, mentoring, volunteering, adaptive sports, and employment. Support employment placement efforts through higher education institutions and vocational rehabilitation (18,23,24,29).

4) Assure continuity of primary health care and continued access to comprehensive and coordinated specialty care. Pediatric providers should ensure that patients have appropriate providers prior to discharging them from the pediatric setting.

- Many young adults with spina bifida experience health-related problems such as deterioration in walking, development of secondary impairments, chronic pain or fatigue, and a low level of physical fitness and inactive lifestyle. A significant number are restricted in their participation in life especially in paid work, housing, intimate relationships, and sexual
experience (1).

- Patient identified adult healthcare concerns: pain, pressure ulcers, bowel/bladder, depression, sleep disturbance, limited social/community participation regardless of age, gender, level of lesion (30).

5) Assess for independently living needs including personal care attendant needs, accessible ramps, bathing/toileting equipment, cooking/cleaning support, and transportation. Provide information about accessible housing, financing, home health care, adaptive equipment, and physical/occupational therapy.

- Most individuals with spina bifida have low social integration and economic self-sufficiency scores, regardless of whether they can ambulate or use wheelchairs (31).

6) Discuss sexual health including sexuality, contraception (including latex allergy precautions), relationships, fertility and childbearing, genetic counseling, and folic acid supplementation before conception. Refer for subspecialty care as needed.

7) Assess mental health and family/peer relationships. Refer for mental health care as needed.

- Anxiety and depressive symptoms are common for adolescents and young adults with spina bifida. Family support can decrease anxiety. Increased self-management ability can decrease depression (28).

### RESEARCH GAPS

1) Studies to identify best practices for comprehensive care for adolescents and adults with spina bifida are needed. These studies should address preventative health in general and related to spina bifida, assessment of secondary conditions related to spina bifida and aging, access to care, and self-management support throughout transition (18,23,32).

2) Educational programs for health care professionals are needed to improve their awareness and knowledge of the medical and social issues of transition and adult life for individuals with spina bifida (1,32).

3) Best practice interventions to support self-management development, developmental milestone achievement, and maximal adult independence are needed.

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