Gaps and Opportunities: An Agenda for Further Research, Services, and Program Development in Spina Bifida

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• Spina bifida • Research • Services • Program development

GAPS AND OPPORTUNITIES IN SPINA BIFIDA RESEARCH AND PRACTICE

Both the current project to develop the Life Course Model for spina bifida, undertaken with the support of Centers for Disease Control and Prevention (CDC), and the activities of professionals in the spina bifida (SB) community[1–4] have highlighted gaps in the knowledge and programs available to individuals with SB. In 2003 CDC, the Agency for Healthcare Research and Quality (AHRQ), the National Institutes for Health, and the Department of Education sponsored a state-of-the-science consensus conference at which invited professionals presented reviews of the evidence in 16 areas and generated a research agenda (see **Table 1** for research priorities relative to the Life Course Model project.)[1] Evidenced-based guidelines for health care providers (HCPs) were developed in 2006.[4] In addition, syntheses of major focus areas in the First World Congress on Spina Bifida Research and Care further identified evidence available for practice in 2009.[2] In

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Table 1
2003 research priorities[1] relative to Life Course Model Project

Domain of this Transition Project	Category from 2003 Priorities	Priorities
Self-management/health	Self-care[a]	Factors that affect the teaching and learning of self-management
		Assessment of ways to measure self-management
		Optimizing use of assistive devices
	Urology	Optional proactive therapy for urinary function
	Mobility	Optimizing mobility changes during adolescence
	Orthopedics	Prevention and management of osteoporosis
	Latex allergies	Optimal prevention management
		Latex allergy in adults
	Integument	Optimizing preventive skin care
		Optimizing treatment of skin breakdown
	Gastroenterology	Optimizing bowel management
Personal/social	Socialization	Determination of the prevalence and nature of social challenges
		Determination of risk factors (and protective factors) for impaired socialization
		Optimizing socialization
	Sexuality	Optimizing psychosocial development
	Family	Optimizing parenting
	Behavioral/mental health	Development of trajectories of mental health/behavioral health issues
		Optimizing mental health
Education/employment/income support	Education/employment	Factors that predict performance in school
		Participation in the labor force
		Models of transition from school to work
	Neuropsychology and learning	Evaluation of the core processing deficits
		Determination of the earliest indicators of learning difficulties
		Determination of institutional and developmental interventions that are most effective in facilitating learning
	Independence	Assessment of current functioning of adults
		Secondary conditions in adolescents and adults
		Optimizing self-determination and independence

[a] Referred to as self-management in this article.

the past 3 years, 20 professionals have also participated in the Life Course Model project described elsewhere in this issue of Pediatric Clinics of North America. The purpose of this article is to build on past work by the SB professional community[1,2,4] by discussing the gaps in the knowledge, resources, or programs available across the life course for individuals with SB identified during the development of the Life Course Model Web site.

Functioning and disability are the interaction among the health conditions and personal and environmental factors according to the International Classification of Functioning, Disability, and Health.[5] Minimizing disability-related challenges for people with SB is as important as the goal of prevention of conditions leading to SB. This analysis focuses on the 3 broad functional domains of the Life Course Model project aimed at minimizing these disability-related challenges (self-management/health, personal and social relationships, and employment/income support) and the common issues that cross these areas.

As the Life Course Model Web site was developed, strategies supported by empiric data were identified and integrated into its development. It became evident during this development process that there were gaps in the research to support the selection of empirically based clinical strategies for this resource. Most of the evidence in each of the broad functional domains is descriptive in nature, with few clinical trials establishing new interventions. This finding is particularly true in the personal/social relationships category, in which only one pilot project (n = 10) on family interventions has been reported,[6] even although this gap was identified several years ago.[7] Some progress has been made in identifying learning challenges in young people with SB, especially in core processing challenges and strengths.[8] However, interventions to address these challenges and build on the strengths have yet to be tested.

In this relative vacuum, important clinical programs have emerged, including a technology-based transition program that has engaged young adults by focusing on building skills in transition-age young people with SB and mentoring programs,[9] the purpose of which is to enhance the development of socialization skills and those related to community integration.[10] Despite the lack of evaluation data published on these programs, they provide important options for individuals with SB when available. Each of these Life Course Model domain pathways can provide relevant information for individuals with SB and their families and the HCPs who work with them. Knowledge gaps can be addressed directly with new research studies that then provide the community with evidence for practice. Simultaneously, programs can be evaluated and practice-based evidence generated that can provide useful information to the SB community and also inform future research projects. Gaps remain in both clinical programs and research; these gaps are the focus of this article.

GAPS IN RESEARCH, PROGRAMS, AND SERVICES

Health/Self-management

An abiding appreciation of and respect for the challenges families face led to the identification of the need for the Life Course Model project. The research on health-related outcomes of adults with SB shows the importance of acquiring the knowledge and self-management skills necessary for health maintenance and SB condition stability. Problems in self-management can result in serious, even fatal, complications, as shown by shunt malfunctioning, impaired kidney functioning, renal failure, and pressure sores.[11–15] The serious and long-term consequences of these complications can be life limiting. In turn, the individual's lifestyle can be seriously affected, as shown by higher rates of unemployment and diminished ability to live independently.[13,15,16]

The evidence to inform and enlarge understanding of strategies to foster self-management/health of children with SB was reviewed and guided the development of the Life Course Model Web site to provide families with life course guidance in raising and caring for their children. Many of the recommendations provided in the self-management/health domain of the Life Course Model Web site (assessment tools, interventions, tips, resources, and referrals) were based on recommendations of clinical experts, promising practices, and theoretic rationale.

The literature on the health and self-management of children and adolescents with SB examines the phenomena from several perspectives, with noticeable gaps. The literature is characterized in part by the use of more global areas of inquiry such as adherence and independence in contrast to examining more discrete questions pertaining to central self-management tasks such as continuous intermittent catheterization (CIC). Levels of independence and adherence have been used as proxies for examining the child's self-management competencies. The studies of adherence and independence do not directly provide the evidence needed to help providers and families find the most effective instructional strategies and long-term approaches to use nor do they identify factors that facilitate or impede the acquisition of knowledge and skills needed to competently engage in SB self-management. However, these studies do provide some direction and insights to guide assessment and intervention, as presented later.

Preschool-aged and school-aged children

Having a child with SB immediately thrusts families into a new world order whose challenges seem overwhelming. These new challenges require families not only to cope with the psychosocial and economic ramifications associated with raising a child with a significant disabling condition but also to learn the surveillance and medical management skills necessary to ensure their child's physical and psychological well-being. As parents become clinically competent in managing their child's care, the process of transmitting their knowledge and skills of SB and transfer of management responsibilities to their child begins as well. For long-term management of SB, families, and eventually the child, must acquire central and peripheral sets of knowledge and skills. The central set includes the pathophysiology of SB, shunt care, CIC, bowel management, skin care, medications, and the use and maintenance of assistive devices. The peripheral set of knowledge and skills includes access to supports and resources needed for care and obtaining the health-related accommodations needed for school, work, and community living. Although empiric studies have reported findings on the medically and surgically related care for individuals with SB across the life span, there is a lack of evidence on effective instructional programs and services for parents whose children are in this age group.

During early childhood, parents are instructed by the members of the interdisciplinary team on raising and caring for their infant with SB. The professional context within which instruction takes place can positively or negatively affect parental attitudes on raising a child with SB. Parents are likely to feel overwhelmed with the new and unexpected care requirements for their child. Empathetic support coupled with patient responses to the parents' need for information about SB and its management increases parents' comfort levels, so that they ask the questions they need answered to fill in the gaps in their knowledge and skills. Connecting parents of newly diagnosed children with other more experienced parents who can serve as mentors and information experts is a cherished resource valued by the less experienced parents. Experienced parents of children with SB are viewed uniquely as having the lived experience that adds to their credibility as resource experts. Referrals to

community-based parent resource centers provide parents and their families with an array of services that include service coordination, the provision of agency referrals, access to their resource clearinghouse, and advocacy training and services. Studies examining parental needs for supports and services during this stage of their child's development are needed for the development and implementation of evidence-based approaches responsive to these needs.

No studies examining self-management/health have been conducted with parents of preschool-aged children with SB. Knowledge of the child's cognitive, gross motor, fine motor, and visual-perceptual skills serves as the framework for determining the child's level of developmental functioning. Considerable research has been conducted with diverse groups of children to assess their developmental status, as is found in the early intervention literature and follow-up studies of children enrolled in preschool programs such as First Steps and Head Start. However, these studies do not include samples of children with SB. The few studies conducted to assess parental reactions to having a child with SB revealed altered and negative parental perceptions of their child with SB. These perceptions can have detrimental effects on the child's psychosocial and emotional development.[17–19]

A meta-analysis of studies was conducted on parents' adjustment to having a child with SB.[19] The analysis revealed the child's SB had a moderate to large effect on parents' psychological adjustment. The researchers surmised that parents had lower performance expectations for their children with SB compared with typically developing children, thereby expecting the child to perform at a lower level of proficiency compared with typical children of their age. Findings from another study revealed a relationship between parental perception of the child's ability and their reluctance to establish behavioral expectations for their children that fostered self-reliance and self-management.[20]

These findings show the importance of providing parents with anticipatory guidance in fostering the acquisition of self-management skills during this period of development. The initial steps in learning self-management skills are the prerequisite tasks that the child needs to learn to become as independent as possible with their own SB self-management. Clinical programs are based on this assumption of the importance of learning self-management skills. No evaluations of differing approaches to teaching parents management of health have been conducted. The Life Course Model Web site is a tool that can be used both by professionals for self-management instructions for parents and by parents themselves. Evaluation of the strengths and weaknesses of the Web site may add to the literature for younger children.

One study addressed adherence of school-aged children (8–9 years old) with SB.[21] Parents and the child's teacher and physician assessed the child's treatment adherence, referring to their ability to engage in self-management pertaining to the 5 central tasks of SB management: catheterization, bowel care, skin care, medication, and ambulation. Findings revealed that parents reported higher levels of nonadherence, compared with physician and teacher ratings. Children were rated as highest on adherence related to ambulation and lowest on bowel care by parents, physicians, and teachers. The correlations between the rater groups were low and nonsignificant. The lack of agreement was attributed to the variability of the child's behavior across settings as well as the situational differences in the raters' opportunities to observe the children. Researchers found that parents offered several attributions for their children's treatment adherence difficulties. Some parents believed their children were capable but lazy or not motivated to perform self-management tasks; others believed that their children's physical limitations impeded their abilities to adhere to the self-management tasks.

Little is known about HCP expectations for achievement of self-management. Using a list of 25 self-management behaviors of different levels of complexity, Greenley[22] generally found HCPs expected competence in self-management skills for those with moderately severe SB (lumbar lesion, normal intelligence, needing a bowel and bladder program) to be achieved at exit from elementary school or middle school. Children with more severe SB (borderline intelligence, thoracic lesion, using diapers for bowel/bladder program) were expected to achieve skills in high school. No differences were found by HCP type but HCPs seeing more children with SB annually expected tasks to be achieved at an earlier age.

These findings show the importance of assessing parents' and HCPs' perceptions of children's self-management behaviors. Unlike in the earlier years, parents begin to ascribe motivational reasons for the child's self-management behaviors. Parents can be encouraged to use positive reinforcement and refrain from using negative remarks as their children engage in self-management to foster treatment goals. Parents can be encouraged to integrate the child into the daily SB management tasks by using the strategies listed on the Life Course Model Web site.

Adolescents and young adults

Although self-management has been explored in its relationship to level of independence, self-concept, and effect on parents, few studies have been conducted examining self-management in adolescents and young adults with SB. A limited number of studies have examined the relationship of continence and mobility to selected constructs such as self-concept, quality of life, and protective factors. Continence and mobility are indicators of optimal functioning, which are dependent on the individual's ability to competently self-manage their condition.

A study of 60 adolescents with SB aged 12 to 21 years found that their level of lesion was significantly related to their functional status, their level of self-management, and social competence. Although this group of adolescents were found to be fairly independent in functional status, they were less so in SB self-management,[23] especially in advanced skills needed for independent living (eg, ordering supplies, making appointments, performing household chores, managing money, transportation management). In addition, in this same group of adolescents, higher levels of functional status and self-management were significantly associated with adolescents who had household responsibilities compared with those who did not. Neuropsychological status also was a major predictor of functional status[24] and self-management outcomes. Parental expectations for condition-related skills generally were at 17.3 years of age, 2 to 3 years less than independent living skills.[25] These investigators concluded that these independent living skills, especially money management, may be critical precursors to success in employment. Lack of congruency between parent expectations and HCP expectations for select self-management skills suggests a need for more study and intervention.

There are inconsistent findings when examining the relationships of continence and mobility to self-concept and mental health.[26] A study of 24 girls and 26 boys aged 6 to 19 years revealed young people who were continent had significantly higher levels of self-concept, and incontinent girls scored significantly lower in self-concept measures.[27] The findings of this study suggest that incontinence is a socially more sensitive issue for girls in contrast to boys.[27] Other studies found that incontinence was not related to the child's self-concept.[23]

In addition, a synthesis of quality-of-life studies in SB revealed similar inconsistencies.[26] Some investigators found no relationship between continence and health-related quality of life (HRQOL).[28–31] Other investigators, using parent reports and

younger children, did find bladder program success related to SB-specific HRQOL.[32] Further, those using an investigator-created HRQOL measure that had continence-specific items found HRQOL increased following bladder surgery.

Researchers studying the emotional effect of CIC on the family (child aged 1–20 years; n = 40) found that raising a child with SB led to strain on the parent-child relationship. This strain adversely affected the emotional component of the parent-child relationship and CIC management.[33] Other studies found that the child's and young person's self-esteem is not adversely affected by CIC issues and may increase the child's level of self-esteem.[34] The effect of continence on mental health and HRQOL needs further study.

Mobility is an important component of self-management and presents a major challenge for many young people with SB. Achievement of optimal mobility enhances multiple outcomes. Low intelligence, hypotonia above the level of the lesion, using a wheelchair for mobility, and poor executive function were noted as significant risk factors for poor self-management skills.[35] However, the role of mobility on self-concept, mental health, and quality of life remains complex. When only the characteristics of SB were evaluated, mobility was found to be the most important determinant for HRQOL for individuals with SB.[35] However, when protective factors such as attitudes, hope, coping, and self-efficacy were included, these psychosocial protective factors had more effect on mental health and quality of life than mobility itself.[28,36]

The other central self-management tasks of SB have not been investigated. No studies were located that examined the factors that enhance or impede shunt management, bowel management, managing skin integrity, weight management, and physical activities.

More study is needed to better understand the self-management needs of children and young people with SB. Researchers have only begun to explore the factors that facilitate or hinder children's/young people's acquisition of self-management skills. Little is known about the most effective strategies to foster the acquisition of the SB self-management skills needed to promote optimal healthy outcomes and prevent and/or minimize the risk of complications and secondary conditions during the period of adolescence. Also needed are studies that delineate the effect of dealing with the hormonal changes of puberty and the developmental tension that exists between independence and continued dependency. The clinical recommendations related to self-management are based on the experience and expertise of clinicians, parents, and individuals with SB. Evidence is needed to inform relevant constituencies of clinical interventions to improve the care for successful acquisition of these skills.

The evidence base related to SB self-management is incomplete, partly because the methodologies used are not comparable. Studies were also limited by use of small convenient sample sizes with unknown biases. Another limitation has been the use of tools with insufficient psychometric properties. In addition, findings about self-management that were associated with specific tasks, such as CIC, were not generalizable to other tasks of SB self-management. More research is needed to better understand the factors that support or inhibit SB self-management. This research should include the use of psychometrically sound instruments with sufficient sample sizes.

Personal/Social Relationships

Gaps in research, programs and services were identified across stages of individual and family development as it pertained to personal/social relationships. The skills needed for effective transition later in life are based on skills developed in early childhood, and the lack of these developmentally appropriate skills puts children with SB at risk. Parents are central to these interventions.

As the Life Course Model Web site was in development, several gaps emerged related to the personal/social domain. First, evidence for interventions that provided parents with effective strategies for developing autonomy and social skills in children with SB was lacking. A second gap pertained to interventions aimed at enhancing resilience in families so that they can provide enhanced support to both their child with SB and other children in the family. These interventions would encompass developing realistic expectations for the child with SB and strategies to facilitate their child's full participation in society.[37–39] Holmbeck and Devine[38] recommended investigating a resilience-disruption theoretic perspective. The third gap involved studies of interventions that increased family understanding of the effect of structural and functional neurologic changes that occur with SB. Specifically, early and ongoing interventions that facilitate identification and remediation of neurologically based executive functioning challenges and the effect of these challenges on peer and family relationships need to be evaluated. The fourth gap was related to parent support programs that focused on building skills of authoritative parenting (ie, providing a mix of structure, affection, and increasing choice to adolescents).

Preschool and school age

Children as young as 3 years of age with SB may lack age-appropriate initiation skills. Clinical programs are needed to support parents in developing a more active approach to facilitate the development of autonomy skills that are like those in typically developing preschool children. Evaluation of programs aimed at increasing choice, increasing age-appropriate responsibility for household chores, and self-care is needed. In addition, although good functional assessment measures exist, practical measures of progressive autonomy need to be developed. The relationship of the level of autonomy skills to socialization skills and peer relationships also needs to be investigated. Evaluating interventions that facilitate development of early socialization skills is also important.

Young people determine their own values and sense of self from those experienced within their family, by peers, or perceived from society's messages as often seen through various media. Along with self-identity, young people are also determining their fit or feelings of belonging to a peer group. Understanding the factors associated with development of self-concept and resilience-based interventions is a priority recommendation.

Adolescence and young adults

The social relationships of young adolescents were studied by Levitt and colleagues,[40] using person-oriented analysis to differentiate patterns of support for children in fourth and sixth grade. Significant shifts in the child's social milieu occurred with the transition from an elementary to a middle-school environment. Self-demands and demands imposed by others for autonomous functioning increased at this time, along with a push/pull toward involvement and identification with a peer group. These researchers reported that children with multiple sources of support were better adjusted than those whose support came primarily from close family members. They reasoned that the availability of alternative sources compensated when close family was not available to meet needs or during times of family conflict. Programs that help young people develop multiple sources of support could provide important sources of protection for young people with SB.

In addition to the normal trials of growing up in an evolving world with parental and peer pressures, children with disabilities must also deal with their disability and the social biases or ignorance that accompanies it. These children are differently able

and in some cases markedly so. Late adolescence and emerging adulthood is a time of exploration and finding one's competencies for occupation, relationships, and both independent and interdependent living. It is normally a time for some risk taking. For young people with disabilities, accomplishing the tasks of adolescence and young adulthood is more challenging than for those without disability. Their margin for error may be smaller, the effect of failure perhaps more consequential, and their opportunities for exploration more limited. Few clinical or intervention programs have been conducted to help understand factors that enhance resilience and social skills during this vulnerable time.

For young people with SB, the Life Course Model project emphasized the need for interventions to facilitate social interaction, especially the ability to begin to build reciprocal relationships. Activities that teach the child and parent how to read subtle social cues and provide extensive rehearsal for social interactions are also needed. In addition, interventions/programs to provide children and adolescents with skills necessary for subtle communication and reciprocal relationships with peers are needed.

Also, interventions are needed to teach compensatory strategies for executive functioning challenges because they have an effect on peer and family relationships. Evaluation of programs that (1) facilitate joint problem solving, (2) provide access to new resources, and (3) promote full participation in society is needed. Of special interest are technology-based opportunities to connect with others, as discussed later in the section on employment.

Important programs for school-aged children, adolescents, and young adults exist in limited numbers. Examples are (1) wheelchair sports programs, (2) organizations that promote inclusive recreation and activities, and (3) camps and mentorship programs. Evidence is needed on the effect of these recreational, mentorship, and sports programs in developing social skills, effective self-care skills, and organizational skills.

Studies report that adolescents with SB are at risk for depression,[38,41] with the incidence of anxiety and depression in young adulthood even higher.[42] The limited data indicate that the transition period to adulthood is particular challenging. However, little is known about the risk and protective factors that play an important role in depression or the application of effective interventions to prevent or treat depression in young people with SB. Data from adolescents and young adults suggest that beliefs (attitude, hope, positive coping) are related to the adolescent's report of mental health and that the family influences the development of these resilience beliefs.[23,28,41] Programs that enhance resilience skills need to be developed and tested. These interventions may be especially important for young people whose family has limited financial or family resources.

Another programmatic and knowledge gap identified in the Life Course Model project was relationships with siblings. We need to better understand the needs of both the young people with SB and their siblings and provide services to both. Research has shown both the positive effect and the challenges for siblings of young people with SB.[43,44] However, little is known about the effect of sibling programs (eg, Sibshops) on the sibling and family.

The personal and social relationship gaps identified in the development of this Life Course Model project are consistent with a recent review of the psychosocial research in SB by Holmbeck and Devine.[38] These investigators suggest that future research should address 3 overall priorities: (1) evaluate longitudinal models of psychosocial outcomes and particularly investigate mediators of outcomes; (2) identify individual, family, and parenting factors that explain success in emerging adulthood for young

people with SB; and (3) identify resilience factors associated with successful outcomes in young people with SB and their families.

Employment/Income Support

Characteristics of SB and academic achievement are both precursors to employment success. Despite some descriptive studies, the relationship of spinal lesion level, brain, and adaptive behavioral differences to outcome is not well understood.[23,29,45,46] The efficacy of interventions designed to promote independence and productivity has yet to be clearly shown.

Characteristics of SB

The lives of individuals with SB are complex and the challenges multifactorial. An understanding of the natural history of individuals with SB across the life course is only now emerging. Technological and health care developments in recent years have offered potential for longer, fuller, healthier lives. This potential is yet to be fully realized at each developmental stage across the life course. Assessment of the individual and environmental factors and the implementation of interventions at critical periods, particularly at key transition points such as preschool to school, junior high to high school, graduation, transition from pediatric to adult health care, and moving from home to less supervised living arrangements, offer opportunities for reconsideration and implementation of new strategies.[23,47,48]

Performance patterns across the lesion-level subgroups revealed that the lesion level had a greater effect on motor than cognitive function, although both domains were affected. The high rates of learning and attention disabilities may lead to more adverse outcomes in terms of social communication and community living.[45] Future studies on effective strategies to advance functional performance within and across the domains of health, education, employment, and community participation should incorporate a longitudinal view to determine if acquisition of discrete skills, such as balancing a checkbook, monitoring the condition of skin, transferring independently, or driving, translates into an improved life trajectory as measured by academic achievement, independence, employment, health, and personal/social relationships.

Although mobility is a predictor of employment, little has been published regarding its effect on independence and employment. Assistive technology use among adolescents and young adults with SB may influence achievement of independence, academic achievement, and employment, yet little research has been carried out to determine the frequency of assistive technology use and its effectiveness on outcomes for individuals with SB. The more traditional technology applications for mobility, such as crutches, walkers, and wheelchairs, have been found to increase the risk of upper-extremity damage. This musculoskeletal stress and strain could have an effect on longer-term self-care and mobility. So there is a gap in our understanding of the long-term implications of the various modalities of mobility. In a focus group sponsored by the Spina Bifida Association in 2010, working adults with SB regretted pushing themselves so hard physically because they experienced later pain and fatigue. Assistive technologies such as smart phones, personal data assistants, and miniaturized recorders are experiencing a higher level of social acceptance. Maintenance, cost, and other factors, such as the need for setup and training, may be barriers to use of assistive technologies for individuals with SB. Johnson and colleagues[49] stated that using assistive technology can result in significantly enhanced independence, employment, and life satisfaction. Study is needed to determine which assistive technologies can contribute across the lifespan, across a broader

range of performance domains, and to determine what protections are needed to mitigate negative secondary effects from their use.[49]

Academic achievement

Current theory, as articulated by Jaffari-Bimmel and colleagues[50] in 2006, concurs with Erikson's step-wise or building-block approach. "Development is the interplay between a changing environment and a changing individual, but early experiences and adaptations do not fade away. Instead, they indirectly shape future adaptation through their influences on intermediary developmental steps."(p1150) Bronstein and colleagues'[51] study provided evidence that parental behavior may affect children's motivation during the transition to middle school by fostering academic performance and sense of scholastic competence. These investigators found academic success led to children having more positive perceptions of their scholastic competence, leading to the development of an intrinsic motivational orientation. Poorer academic performance led to self-perception that was more negative and to the development of a more extrinsic motivational orientation. Autonomy-supporting parental behaviors such as allowing children to express their ideas and opinions and to participate in family decisions may have helped develop the children's capacity for independent thinking and problem solving[51] (see also the section on personal/social relationships). This encouragement for independent thinking, problem solving, and self-efficacy–promoted achievement may stimulate inquiry to better understand factors that influence the child's level of functioning.

Employment

Previous research on work participation generally showed overall rates of employment between 19% and 38%. Predictors of having paid work (at least 1 hour a week) were level of education, level of lesion, hydrocephalus, IQ, self-care independence, and ambulation.

In the study by van Mechelen and colleagues,[52] bowel and bladder continence made it 2.5 times more likely for the individual with SB to be employed. A higher level of education was an important and positive indicator of employment. Another study found lower levels of post-secondary education, with 41%–49% of individuals with SB attending college vs 66% of typically developing young people[38] which may have a major effect on underemployment.

Van Mechelen and colleagues[52] also found that the most effective time to help young people find a job was immediately after they finished their education, noting that motivation to work would gradually decrease once they accepted welfare benefits. Young adults with SB who had experienced problems in finding suitable employment reported difficulties, with reluctant attitudes among employers (57%), work offered that was physically (30%) or mentally (27%) too demanding, transportation (32%), accessibility of buildings (23%), and toilet space (23%). To date most studies have focused on either demographic or condition severity factors as predictors of employment success. Understanding individual, family, environmental, and financial factors associated with successful completion of higher education should also be studied. These factors might include lack of autonomy-related socialization,[53,54] accessibility and transportation difficulties, insurance complexities, stigma, and lack of job training.[38,55,56]

Reiss and colleagues'[57] research identified 3 stages of transition: envisioning the future, age of responsibility, and age of transition. Children with disabilities need encouragement to envision their future. Assessment and intervention approaches to

facilitate independence and readiness for employment should be appropriate for the individual's age and stage of development.

Preschool and school-aged children

Early intervention programs for infants and toddlers exist throughout the United States and almost universally children with SB qualify for these services. Evidence is lacking about the interventions in these programs that improve mobility, overall development, and efficient transition to school-based programs.

Difficulties in language, learning, memory, and attention emerge during this time. There is limited evidence regarding when to perform neuropsychological assessment of these skills. Individuals with SB and their parents or caregivers have expressed a lack of awareness with not only the terminology but also the concepts of executive function and nonlearning disabilities. A few programs for preschool children have developed mechanisms to systematically evaluate executive, attention, and learning functions.[58] These programs, aimed at children with various disabilities[59] including SB, focus on developing compensatory skills at an early age. Because these programs offer the opportunity for early intervention, their evaluation should be a high priority.

Programs that institute early testing for core neuropsychological processing skills are needed in the early elementary years. Studies evaluating age-appropriate interventions and accommodation strategies (especially for mathematics) are needed to optimize early school success. Autonomy skills may be limited by these core neuropsychological skills (see section on personal/social relationships). Evidence of the most effective, age-appropriate, reality-based interventions to increase function in areas such as managing money, planning and organizing, and solving real life problems should be evaluated. Evaluation of technology to organize, support, and prompt behavior in both school and self-care activities is needed.

Adolescents

Evidence is lacking of the optimal structure and components of an effective transition plan that addresses the critical areas for students who have an Individualized Education Plan. In 1999 to 2000, the standard diploma graduation rate for students with disabilities aged 14 years and older was 56.2%. During the same period, the dropout rate declined from 34.1% to 29.4%.[60] Graduation from high school and/or postsecondary education programs leading to employment are key factors in attaining the highest level of autonomy.

Attaining meaningful employment that reflects the preferences and interests of the individual with disabilities is too important to be left to chance. Careful planning, dedication of resources, and educational programming are essential to achieving this goal. Because of the marked disadvantages for young people with disabilities when competing for employment, opportunities for career exploration and skill building should occur before the young person is placed in a specific program. Assessment of aptitudes, special needs, learning style, personal and social skills, values, and attitudes toward work as well as work tolerance can assist the student to learn about himself or herself and inform their career choices.[61] The effectiveness of work-related programs offered in high schools to build self-management, social skills, and work behaviors needs to be evaluated. Access to and the effectiveness of vocational rehabilitation/career counseling interventions needs to be assessed in terms of assisting students with SB to obtain postsecondary employment.

Young adults

Material consequences, pay for performance, and other social rewards and privileges are common motivators. Individuals regulate their own behavior by self-evaluative and

other self-produced consequences.[62] If an individual is unable to earn material rewards or experience positive recognition and acknowledgment of accomplishment, their sense of self may suffer. The long-term implications of diminished self-confidence and self-esteem need more study.

Programs that provide coordination for posthigh-school skills, supportive employment, or transition to postsecondary education need to be developed and evaluated. Especially important is delineating the components of these programs that are the most effective in predicting employment, quality of life, and community participation outcomes.

Across domains and life course

Executive function challenges and nonverbal learning differences are 2 conditions that can greatly affect how an individual with SB does in life. A person's executive function system is responsible for self-awareness and the ability to plan and carry out tasks in daily life. It modulates both emotions and behavior. Learning disorders are caused by a difference in the brain that affects how information is received, processed, or communicated. The expressive verbal skills and social nature of people with SB may conceal nonverbal challenges.

A learning disability is not a problem with intelligence. Individuals with learning disabilities have difficulty processing sensory information because they see, hear, and understand things differently. Advancements in the science of how the brain adapts, through a process called neuroplasticity, suggests a natural, lifelong ability to form new connections and generate new brain cells in response to experience and learning. Opportunities to affect learning and executive function by retraining the brain may hold promise for reorganizing neuronal connections and facilitate skill-building capabilities for individuals with SB. Application of the science of neuroplasticity to individuals with SB, particularly those with hydrocephalus, is an exciting area for future research. Training (repetitive practice for skill building) offers a different way of facilitating learning, compared with teaching (telling once or twice), a method commonly used in academic, employment, and health care settings.

SUMMARY AND PRIORITY RECOMMENDATIONS

The Life Course Model project has identified that little progress has been made on the research agenda priorities generated after the 2003 consensus conference on SB (see **Table 1**). Gaps persist in the knowledge, programs, and services across individual and family developmental stages that need to be addressed to facilitate transition in self-management/health, personal and social relationships, and employment/income support. Research is needed that is theory-driven (model testing), longitudinal, addresses critical developmental periods or transition points, is inclusive of ethnic and socioeconomic diversity, is multisite and with sufficient sample size to address multiple variables, and identifies the trajectory of SB across the life course.

Furthermore, studies are needed that move from identifying differences between those chronic health conditions, like SB, and typically developing peers to addressing why the differences occur. Studies that identify personal, family, and environmental factors associated with successful achievement of self-management/health, personal and social relationships, and employment and interventions based on these factors are immediate priorities for optimizing the successful transition to adulthood. The effect of successful employment experiences on social and quality-of-life outcomes needs to be investigated.

The 3 domains defined in the Life Course Model have natural overlaps that need more study. For instance, how do cognitive skills affect self-management and personal/social relationships? We also need to determine if there are critical time

points at which intervention might make a difference in the trajectory and outcomes. Valid and reliable measurement of the milestones depicted in the Life Course Model is also a challenge. The commonly used and available instruments may not be appropriate for measurement of important constructs. With so many more adults living with SB, they should be used to identify important proximal and distal outcomes, develop research questions, and validate measures. A collaborative partnership with all stakeholders offers the most promise of enhancing the science and improving the lives of all living with SB.

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