Need for the Life Course Model for Spina Bifida

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KEYWORDS

• Spina bifida • Transition • Self-management • ICF

In previous generations, people with conditions such as spina bifida (SB), sickle cell disease, cystic fibrosis, and muscular dystrophy were not expected to live very far, if at all, into adulthood. If these people did live, health care professionals and other providers were pleased that they had even lived that long. Because children with chronic conditions have grown up into adults in increasing numbers,[1–4] they and their families have increasingly questioned whether they have reached their full potential and maximized their participation in adult activities. When standards of care and relevant outcomes are expressed in traditional medical parameters (mortality, morbidity, occurrence of secondary health conditions involving other organ systems), scant attention was paid to the functional outcomes that are the norm for typical young adults, including whether the young adult had a job or sufficient self-directed income, had friendships and relationships, lived independently, and engaged in recreation and other health-promoting activities. Expectations were often low to nonexistent about this group's potential to function well in these domains of adulthood.

Pediatric chronic disease specialists are increasingly recognizing the shortcomings in the quality of life of their patients as they reach adulthood. As patients leave pediatric care, their doctors are moving beyond the issue of a simple transfer of health care to adult doctors to feeling a responsibility and an interest in functional outcomes, in terms of full participation in adult living.[5]

The disability movement of the 1990s, reinforced by the Americans with Disabilities Act and other legislations and policies, has empowered people with disabilities to view themselves differently. These people now aspire to the full quality of life as demonstrated by their typical peers and portrayed in the media. With aspirations, comes an expectation that the systems of services and support will prepare them for these enhanced adult roles.[6]

The findings and conclusions in this article are those of the author and do not necessarily represent the official position of the Centers for Disease Control and Prevention.

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Pediatr Clin N Am 57 (2010) 893–901
doi:10.1016/j.pcl.2010.08.001 **pediatric.theclinics.com**
0031-3955/10/$ – see front matter

TRANSFER OF HEALTH SERVICES

Because of the lack of adult health care providers with expertise in SB, many young people struggle to find health services. Many patients stay with their pediatric provider well into their adulthood, reflecting the trend seen in other childhood-onset lifelong conditions. Indeed, Boston Children's Hospital now allows patients up to 35 years of age to attend certain clinics run by their pediatric staff. Still, as children with SB reach adulthood, most of them lose access to health care providers who have knowledge about SB. This situation places an increasing responsibility on the patients to understand their own condition, the pattern of symptoms unique to them, the self-management of symptoms that works, and the threshold for seeking medical attention.[7] Shunt function typifies this situation. Young adult patients have a long experience with possibly shunt-related symptoms, such as headache and fever. Ideally, these patients would be able to make measured decisions on what intensity and duration of symptoms can be self-managed and when to seek care for possible shunt malfunction. Management of urinary symptoms is a similar situation. This enhanced self-management would be desirable if the patient has access to experienced adult health care providers but is even more critical when the access is limited. Covering these eventualities requires preparation of the young patient with SB, starting in early childhood.

The Life Course Model merges several concepts and principles related to children with disabilities and provides a framework for services and research to achieve the desired adult outcomes (**Fig. 1**).

NORMAL CHILD DEVELOPMENT

Children are dynamic in their progress from infancy to adulthood. Their development follows a series of predictable stages or milestones in each of their domains of development. An important principle is that stages in development generally have to follow a sequence, that is, important stages cannot be skipped to move on to future stages. One cannot walk until one learns to stand upright. This principle applies to motor milestones as well as language, cognitive, emotional, and behavioral milestones. The sequence of development may be tighter and more predictable in the preschool years but still needs to be followed in school years and adolescence.[8]

If a critical milestone is missed, it may be difficult to go back and relearn that stage. For instance, children who fail to resolve their struggle for autonomy at ages 2 to 3 years may struggle with autonomy in their teen years and may possibly have trouble with authority and self-control throughout their lives.

INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY AND HEALTH

The International Classification of Functioning, Disability and Health (ICF)[9] was developed as the latest addition to the family of international classification systems. Thinking that the previous International Classification of Diseases codes were heavily medical and deficit-oriented, experts proposed a new system of classification oriented to function and participation rather than disease and limitations. The ICF, proposed in 1992 and finalized in 2001, is based on the principles that (1) activities and participation are the desired outcomes that should be monitored and measured and (2) these outcomes are determined by an interaction between impairments (of a body part or system) and other factors (environmental and personal). These principles suggest that a responsive accommodative environment will lead to a higher degree of function and participation than a less accommodating environment. Environment

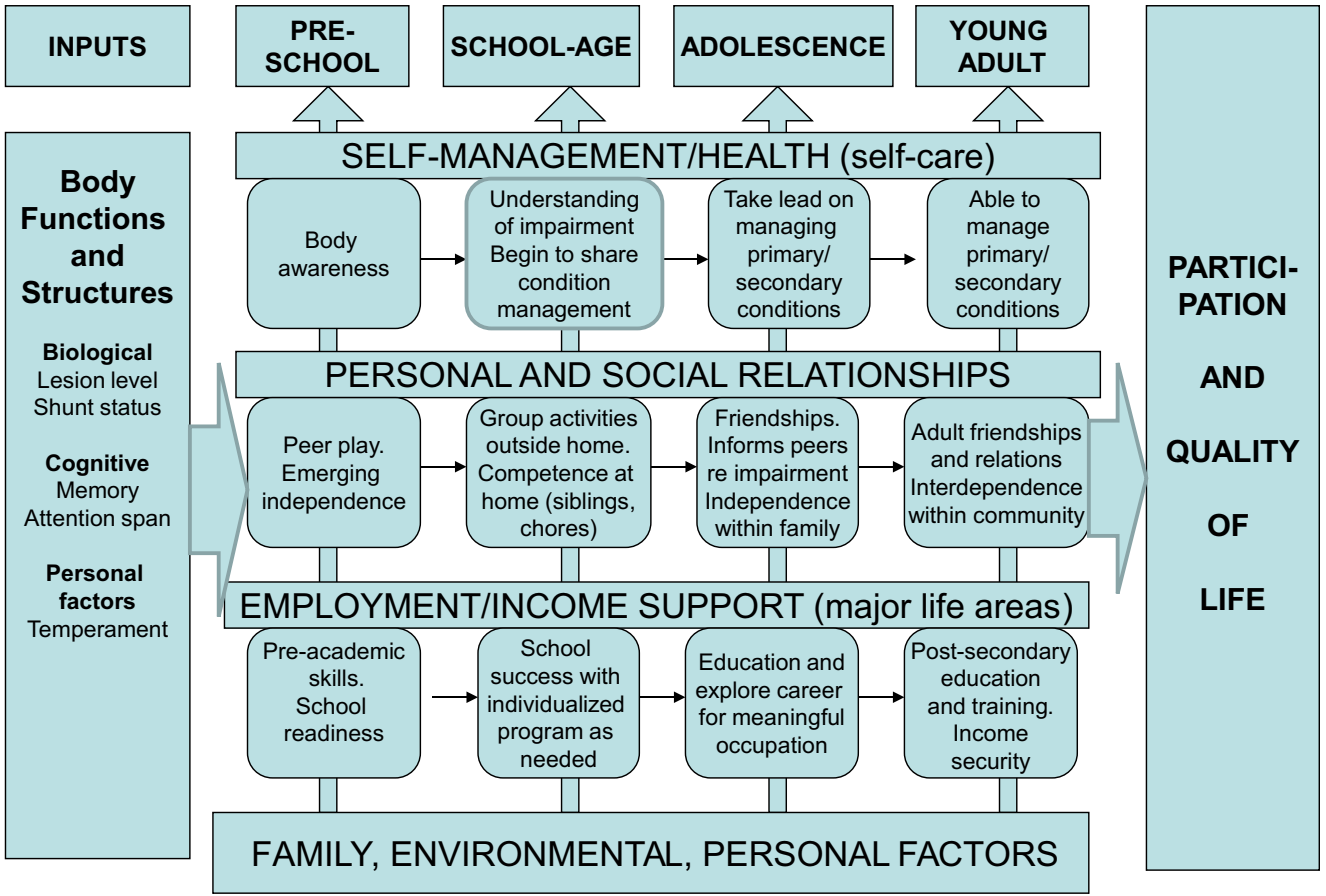

Fig. 1. The life course model for SB.

includes family, community, school, and service systems and encompasses attitudes as well as direct services. Although the details used in the ICF to measure environmental factors and participation outcomes can be cumbersome, the ICF is especially useful in its broad categorization of outcomes, specifically its participation domains or categories. The ICF is a world-recognized classification system that now shapes almost all current discussions of disability-related services and science.

ICF	System for Classifying Impairments, Activities, and Participation
Impairment	Affected body structure or function (eg, SB, paralysis, cognitive limitation, primary emotional disturbance, sickle cell disease)
Activity	Execution of a task or action by an individual (often in a controlled situation, such as home)
Participation	Involvement in a life situation (or performing activities in the real world)

What is most appealing about the ICF in relation to SB is the recognition that a biosocial model is needed to explain the differences in outcomes for persons with SB. A biologic model would suggest that the initial lesion and birth anatomy would greatly determine the outcomes in children with SB. This suggestion may hold true for some of the biologic outcomes (motor function, cognition) that are heavily influenced by lesion level, anatomic brain differences, and shunt malfunctions, which are always followed in a medical setting.

But the differences in functional outcomes of employment, relationships, and emotional well-being are not explained by these biologic inputs. Other factors must be identified to explain the variation in functional outcomes. So the ICF offers 2 concepts:

- Participation is the desired outcome.
- Participation results from an interaction between the impairment and environmental response.

There are 8 areas of participation defined by the ICF:

1. Interpersonal interactions and relationships
2. Major life areas
3. Self-care (management)
4. Learning and applying knowledge
5. Communication
6. Domestic life
7. Community, social, and civic life
8. General tasks and demands.

The authors' Transition Working Group focused on adaptations of the first 3 areas of participation in creating the Life Course Model. These areas were thought to be most compatible with a participation theme and a developmental model that would track progress from early childhood to adulthood.

The authors have operationalized the ICF to define optimal adult function as successful participation in the first 3 areas. Certain elements of the unchosen 5 areas were incorporated into the 3 themes chosen for this project.

Definitions

Interpersonal interactions and relationships[9]:
Informal social relationships

Entering into relationships with others, such as casual relationships with people living in the same community or residence; with coworkers, students, playmates; or with people from similar backgrounds or professions.

Formal relationships

Creating and maintaining specific relationships in formal settings, such as with employers, professionals, or service providers.

Intimate relationships

Creating and maintaining close or romantic relationships between individuals, such as husband and wife, lovers, or sexual partners.

Major life areas[9]

Education

Work

Economic life

Self-care[9]

Looking after one's health

Ensuring physical comfort, health, and physical and mental well-being by maintaining a balanced diet and an appropriate level of physical activity, keeping warm or cool, avoiding harms to health, following safe sex practices, including using condoms, and getting immunizations and regular physical examinations.

Toileting

Planning and performing the elimination of human waste (menstruation, urination, and defecation), and cleaning oneself afterwards.

Impairments are the underlying conditions that are inputs into the Life Course Model. People with impairments are not unhealthy or limited in participation. The goal of persons with impairments should be assumed to be the same as for those who are nonimpaired, namely full participation.

INTEGRATION OF THE ICF AND DEVELOPMENTAL APPROACH ASSUMPTIONS/VALUES

1. Children with disabilities and their families have the same aspirations (ICF domains) for successful adult living as those without disabilities.
2. Successful adult living follows effective preparation, which is demonstrated by making steady progress in life domains, starting in the early childhood.
3. Impairment and its subsequent interventions (treatment) and environmental response can adversely affect the trajectory of development in children with chronic health conditions, such as SB, and lead to disability when environmental factors are insufficiently accommodative to allow participation.
4. The natural history of many impairments is unknown for 2 reasons:
   - Only recently have children lived well into adulthood.
   - Physical health and, especially, functional outcomes have not been documented.
5. Lack of knowledgeable adult medical providers and longitudinal data about natural history places more responsibility on individuals and their family for self-care of the impairment.

Because child development is a naturally dynamic process, it is subject to environmental factors that can interrupt the process. Even in children with no body impairment, environmental factors such as a chaotic home, overly critical or lax parenting, unresponsive teachers, unstructured classrooms or dangerous communities can threaten development. Typical children tend to have a greater reserve and capacity

to deal with these environmental challenges than a child with a constant impairment.[10] The ability to adjust or accommodate to an environmental stressor is less with a cognitive, motor, or emotional impairment. Critical developmental milestones may be delayed or missed between a permanent impairment and an intermittently unsupportive environment.

By systematically measuring development (proximal outcomes) in children who are at risk for altered development, parents and clinicians can track progress in life domains.[11–13] If a child begins to fall off the normal trajectory in a given domain, there is an opportunity to intervene and bolster progress in that track.

SELF-MANAGEMENT OF CHRONIC ILLNESS

The chronic care model exemplifies many of the principles of self-management of chronic illness. Patients and their families, in essence, are the primary caregivers in chronic illness. Self-management has the goals of promoting health and preventing complications. These goals are accomplished through monitoring of physical and emotional statuses and making appropriate decisions based on self-monitoring.[14] The added benefit to this process is empowerment. Positive results reinforce the process. Self-management includes how to develop and maintain exercise and nutrition programs, manage symptoms, determine when to seek medical help, work effectively with doctors, properly use medications and minimize side effects, find community resources, and discuss the illness with family and friends.

Young people with SB face the challenge of living with a chronic condition that affects multiple organ systems and can require immediate, if not emergency, attention. Because most people have ventricular shunts, they have to monitor symptoms that could be related to shunt malfunction, such as headache, malaise, fever, and weakness. Urinary symptoms have to be interpreted in the light of decreased sensation. Pressure sores also need visual diligence to augment limited sensation. Lower extremity function is dealt with in the face of motor and sensory deficits.

The combination of chronic and acute symptoms places considerable responsibility on the family when the child is young. With independence as an adult goal, a family's health monitoring needs to be a shared responsibility during childhood, with the child steadily taking on more and more of the responsibility for self-management of his or her health.[7] Collaborative management with health care professionals is desirable. Such management is more often available for children with SB because they have access to pediatricians and pediatric-oriented specialists health care professionals who have knowledge of SB and are aware of the key role played by families in providing care.

Management includes the self-care taken on by all children and also the special features of SB. All children learn to manage their own acute symptoms (pain, malaise, fever) and make decisions on when to seek help (from parents or the health care provider). Monitoring of the development of this self-care skill is especially important for children with SB, given the projected need to manage their own health in adulthood and to educate adult health care providers about management of the secondary conditions associated with SB. The chronic care model offers useful guidance for the life course model. Although originally targeted for chronic disease management in adults, the chronic care model offers useful principles and techniques that can be adapted for chronic disease management in childhood. Again, in keeping with a developmental model, one must account for and recognize that children are acquiring cognitive skills across the life course and such skills allow them to take on increasing self-management over time. The final goal of adult self-management must be kept in mind. The shared approach has driven the service system developed in Toronto for youth in transition

to adult care. The model (originally presented by Kieckhefer and Trahms[15]) depicts how the roles of health care providers, parents, and individuals change over time. The child moves from passively receiving care to active engagement and finally to management of care (as an adult). Service providers' roles change from the major care provision to consultation and finally becoming a resource. Parents' role also diminishes over time, ending as a consultant to the family member with impairment.[7]

The scarcity of adult health care providers who are knowledgeable in SB puts more responsibility on the individuals with SB and their families to prepare for adult management of SB. The adult urologist and neurosurgeon need general information about SB as well as particular information about the unique aspects of the condition in that individual. Thus, the Life Course Model has a strong emphasis on the adult goal of strong patient-directed self-management and outlines steps to prepare for such an occurrence, which is consistent with the prevailing belief of empowerment that undergirds the adult disability movement. Admittedly, this self-management pushes the limits by encouraging what might be viewed by some as an unwelcomed intrusion into medical care by patients. But this tension is welcomed by others as a reflection of changing paradigms, needed to accompany the longer life and higher aspirations of people with disabilities. The driving force behind the Life Course Model is echoed in the statement on self-determination:

> *Self-Determination is both person-centered and person-directed. It acknowledges the rights of people with disabilities to take charge of and responsibility for their lives. In Self-Determination, the individual, not the service system, decides where he or she will live, and with whom; what type of services he or she requires, and who will provide them; how he or she will spend his or her time, which may include the type of vocational or educational opportunities he or she wishes to engage in, and how he or she will relate to the community, which may include joining in community events, taking part in civic groups, and developing and maintaining relationships with others in the community.*[6]

SUMMARY

Empowerment and self-determination have become dominant themes for adults with disabilities. Parents and their children with disabilities can see examples of how adults are doing to guide the management of their lives, including health self-management, and to motivate them for full participation in adulthood. Examples of typical childhood development serve as a guide to preparing for adult life.

The trajectory to full participation may be lower and slower, but one needs to be established. A developmental approach is needed to map trajectory to successful participation.

Self-management of health conditions is especially important when adult health care providers lack knowledge of adult patients with pediatric onset chronic conditions and disabilities.

CHALLENGES AND OPPORTUNITIES POSED BY THE LIFE COURSE MODEL

For many health care providers, the Life Course Model is a new paradigm, whereby functional adult outcomes become the goal of traditional management of body systems. Tracking and measuring progress in these domains may seem subjective. Better measures of participation need to be developed at all stages of development. Intentionally sharing condition management with parents and ultimately with the child may be a shift in approach to some health care providers.

For individuals and families, the Life Course Model forces a focus on long-term outcomes: what will happen when the child becomes an adult. Initially, families often focus on their own loss and grief. When the child is born (even earlier, if the diagnosis is known before delivery), the family faces the reality of having a child with lifelong impairment and its possible consequences. Just getting through the first few years of medical issues, such as hydrocephalus/shunting, is demanding. When the child stabilizes medically, the family can start looking forward to the more distant events, preschool and school. Providers and support persons can help inch the family toward the long view of adult life and reinforce the steps that can be taken in the present to prepare for the future. Showing a pathway to adult life through the Life Course Model can help families see that there is a step-by-step process that can be broken down into manageable steps across the life course. Families can learn that each milestone builds on a previous one and that participation in adulthood is attainable.

Although this article and issue focus on SB as a sentinel condition conducive to the Life Course Model, the model can be easily applied to all chronic conditions of childhood onset. Regardless of how the impairments are clustered in a specific condition or in an individual, the principles of preparation for adult participation through the use of a Life Course Model can be adopted and implemented by all affected families and their health care providers.

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