Spina Bifida: What We Learned from Consumers

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In the professional literature, much is written about the causative factors associated with spina bifida, its prevention, and the interdisciplinary approaches to addressing the biopsychosocial needs for treatment.[1–4] From the consumer perspective, for those who have spina bifida, the operative word about the knowledge and science of spina bifida is "about." That is, the literature to inform constituent groups, whether they are interdisciplinary professionals, policymakers, advocates, parents, or individuals with spina bifida themselves, often "lacks the voice" of those who live with spina bifida every moment of their lives.

The advances in medical treatment and interdisciplinary management of spina bifida have led to impressive improvements in the life expectancy of individuals with spina bifida. Issues of survival recede as quality of life issues and concerns about adulthood potential draw greater attention.[5] As has been discussed throughout this issue, comprehensive preparation for adulthood, beginning in early childhood, is of vital importance to foster the attainment of the lifetime goals that individuals with spina bifida dream and seek to achieve.

Best and evidence-based practices are predicated on data gathered through empiric means, expert practice, and the wisdom and insights of others who have had the lived experience, whether the consumers themselves or their family members. This article provides the perspectives and insights of several individuals with spina bifida who have experienced the transition experience themselves. Their unvarnished

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stories will open the door, metaphorically speaking, to their own candid reflections of this important period of time in their lives. These personal commentaries, each one unique, reveal the commonalities of this shared life experience. Next, preliminary findings of a longitudinal study entitled, “Examining the Trajectory of Transition in Adolescents and Young Adults with Spina Bifida,” conducted at the Gillette Lifetime Specialty Healthcare Clinic, is presented. The compilation of personal impressions serves to inform those involved in the provision of care, services, policymaking, and advocacy services to youth and young adults with spina bifida, their families, and their circle of support of what is of foremost importance during this period of transition. The article concludes with recommendations, many of which can be located in the new transition Web-based resource, for the provision of youth- and adult-centered services and resources.

AMY’S STORY

Amy’s reflections about living and growing up as a child with spina bifida and later as an adolescent and young adult, reveal a number of significant influences and challenges she faced. As Amy tells her story, she recounts the very important role her parents had in her life—a role of support and guidance, which at times she described as “overprotective.” Her description of her school experiences in high school and college reveal the challenges that she encountered without the assistance of comprehensive transition planning, including for health care planning. Today, experts and advocates alike recognize the importance of supporting families and children with complex medical conditions, including spina bifida, to access services and/or programs that will assist children to develop self-advocacy and self-determination skills as exemplified with the development of the life span Web-based resource tool.[6,7] Amy’s brief autobiography reveals the power of her positive and accepting attitude toward living her life to its fullest. The following is Amy’s story.

There are many factors that affect how we handle life transitions, but to me the most significant one is the family. Mine has always been supportive. My parents made sure I got the care I needed. But I was also overly protected. Their concentration, understandably, was on doing all they could to help me survive and walk rather than encouraging independence. Yet I’ve struggled with confidence issues throughout my life, which I believe stem in part from that protectiveness. It is vital, in my opinion, for children with spina bifida to be encouraged to explore their world and be independent.

I was excited to enter the new world of school that my older siblings got to experience every day. I was mainstreamed, which I think was a good idea, but I became aware that I was different from the other kids, some of whom stared and made fun of my wobble walk. School took me out of my comfort zone. My self-consciousness grew. I believe now that counseling could have helped but, to my knowledge, it wasn’t suggested by my medical team.

For health reasons, I was frequently absent from class and found it increasingly difficult to keep up with my work. The teachers and staff, although generally caring, didn’t seem to know how to handle my situation. Early on, I went to physical education classes but spent most of the time on the sidelines. No arrangements were made for activities specific to my capabilities, and eventually I was exempt from physical education classes altogether. And I was held back in second grade because of frequent absenteeism more than an inability to do the work. After a rough start, the school provided more tutoring to help me keep up. I preferred this one-on-one learning experience over being in a classroom full of other kids.

With the help of family, friends, understanding teachers, and counseling, I blossomed in high school. My health improved, as did my confidence. I joined clubs, attended school dances, and did volunteer work. But I was still exempt from physical education classes. The school nurse tried to arrange something for me, but by then, unfortunately, I wasn't interested. I had too often gotten out of doing what I didn't want to, and I remained stubborn.

That stubbornness led me to go away to college. But it backfired. I was living 6 hours from home, confronted with responsibilities I was ill-prepared to face on my own—most notably health maintenance. I quit after 2 weeks and entered the local community college the next year, where I thrived. There, I was required to take an activity class, so I chose Personal Fitness. The teacher was understanding toward my limitations and adjusted workouts accordingly. It was one of my favorite classes. I also worked as a tutor in my spare time.

Throughout my adult life, I've fought depression, confidence issues, and preconceived notions of others. I let others think for me quite often. But through growth and maturity, that attitude has slowly changed. I've explored different career paths. When one didn't work out, I tried something else. Through my experience with spina bifida, I've learned the importance of patience, health maintenance, and having empathy for others.

Transitions are a part of life. With a good base of support and a positive attitude, those with spina bifida can more smoothly navigate life transitions.

CARLEY'S STORY

As Carley relates in her report of the transition from adolescence to emerging adulthood, she acknowledges the helpful support she received from educators and other professionals to prepare for the transition to college and the world of work. Yet, this same level of assistance was not available from the team of health care professionals who managed her care for many years. Carley described her health care transition planning and transfer to adult health care as a "non-story," meaning that she did not receive the assistance she obviously needed.

Carley's disappointment with the lack of acknowledgment of the important milestone of the termination of her pediatric care is apparent as she describes "there was no fanfare," a ceremonial oversight by her pediatric providers. Curiously, this "rite of passage," an important one for both the recipient and provider of care is rarely, if ever, acknowledged. As one expert recommended, "Celebrate transitions as they occur with graduation ceremonies, certificates of completion, and other rites of passage."[8(p1313)] The lack of assistance with the transfer of her care from pediatric to adult providers is obvious as well. Instead, without the helpful guidance from a member of her longstanding specialized pediatric team, Carley forged ahead to locate her own adult physician. Carley's story follows.

In reflecting on my transition from pediatric to adult health care, I have trouble putting the experience into words. I realize that this is because, while I can clearly recall much preparation going into other transitions in my life, from high school to college and from college into the work force, I see my health care transition as a non-story, with few details to recall. In my educational and career transitions, I remember a lot of preparation, with teams of professionals coming together with me and possibly my parents, and discussing in detail what the next steps would be to help me reach my goals. For my health care transition, there was no fanfare, no official meetings where the next steps would be neatly spelled out to carry me on my way to my goal of optimal health care. I remember mention of final appointments with my pediatric doctors; however, there was no clear answer as to where I would go next for care. I was left to search through multiple provider lists from my mother's insurance carrier, hoping to find doctors who had at least a basic knowledge of spina bifida. Luckily, I have found doctors that I am confident will help me reach my health care goals; however, I know not everyone has the access to or knowledge of the health care system to navigate it as successfully as I have, which is why comprehensive health care transition planning is crucial for every adolescent with a chronic illness.

JAMIE'S STORY

As with Amy, Jamie's parents had an important role in her life. She credits her parents with not only supporting her but actively encouraging her to learn to become more self-determined and to learn to advocate for herself. She acknowledges her parents' efforts to foster self-management of her spina bifida in all aspects of her special health care needs from doing her own self-care to making her own decisions regarding her medical care. Her parents started early, apparently recognizing that learning to become independent involves a life span approach. In most schools, formalized transition begins between 14 and 16 years of age. Waiting until then to address transition issues can hinder development of these maturity milestones.[7,9,10] This same philosophic approach guided the development efforts of the Web-based resource. The following is Jamie's story.

When I consider transitioning into adult health care in regard to managing spina bifida, it's hard to pinpoint just when that happened. I think it's because, really, there were multiple steps to that transition.

I was blessed to have parents who knew I would be most successful if I was taught to be independent. I wasn't treated as their "disabled child," I was simply taught to manage my disability. What I mean is, I was treated like my able-bodied brother was with regard to expectations of responsibility, and so forth.

Another blessing was that my parents included me in on decisions about my medical care from an early age. This included treatment, but mostly on choosing doctors. I remember meeting a new neurosurgeon and I didn't like how he treated me (he spoke to my parents—not me, etc) so we never went back. I also was allowed to stop seeing my (wonderful) male pediatrician when I was 9 years old because I had started to develop "lady issues" that I felt a woman doctor should hear. So, I started seeing a "grown-up doctor" for my "grown-up issues." Through these experiences, I was being trained to gauge what kind of doctors I liked and thought were good for me.

I was also taught to make my own doctors' appointments from the time I was a young teenager. This is a vital skill that I don't understand waiting until adulthood to learn. Being able to know what doctors to call and when, as well as being able to articulate the reason for the visit is really not as hard as some people with spina bifida make it out to be, in all honesty. It's a simple phone call that is no more difficult or complicated than ordering a pizza.

All of these skills were incredibly important (and put to the test) when I left home to attend college 2 states away. I was dealing with doctors in a small town that didn't always fully understand the issues related to being an adult with spina bifida. I learned even better self-advocacy skills that I continue to practice more than a decade later.

What I continue to find challenging is knowing what issues I need to be aware of now that I'm an adult with spina bifida. I have the health care guide for adults, but there's been several times that I've been told that some of the tests mentioned in that book are not necessary. As there is only a growing amount of knowledge of adults with spina bifida, I never know if I should trust information from the Spina Bifida Association, or from doctors who are sitting right in front of me. This is where my decision skills come into play. I'm learning to gauge when I should fight for certain things or trust the doctor's judgment. It's a fine line that seems to continue moving.

One thing about this journey into adulthood is that it's not over yet. And that's not just because I have spina bifida. As I get older, there will be more and better treatments, but there will also be more issues. I believe I've been equipped with the latent skills to handle those as they arise.

JILL'S STORY

Jill's recollection of her childhood experiences in dealing with health care professionals demonstrate the absence of her "voice" in dealing with her medical condition. This lack of self-advocacy and self-determination carried over into the school setting as well, leaving Jill feeling uncomfortable in offering her preferences about what she wanted. As a result, Jill did not receive the instruction needed to thoroughly manage her spina bifida. Confidence in important skills such as recognizing the triggers for possible complications and illnesses and learning to become an informed consumer by adopting the practice of compiling her health care records for later use were undeveloped.[7,11] The lack of transition preparation is the same theme echoed in the recalled memories of Amy and Carley. The following is Jill's story.

When you are young, you are told to be seen and not heard. These are words that ring in my ears today. Although I believe in respecting your elders, I wish I would have listened to myself a little more. When doctors would talk they would always talk to my mom. I felt as if I weren't even in the room. The same thing happened when I was at school and had an IEP (Individual Educational Plan) meeting. The teacher would talk to my mom. I previously wondered, "Is this my health and education they are talking about?" This has came back to haunt me, not knowing or understanding what was going on.

Doctors would make predictions on my life expectancy; they actually said I would die at the age of 9. I'm here today to tell you I am now 44 years young with 2 beautiful children. Having children is something else doctors said would probably not happen. Most of these doctors have passed on or have retired; I wish they could see me today. I graduated from high school, attended college, and have been working since the age of 13. When I turned 18, I could no longer go to the Myelo Clinic because I was too old. I searched high and low for an adult clinic, but no such luck. After all, doctors didn't think individuals with spina bifida would reach the age of 18.

I then got wrapped up in my own life and forgot that I had a disability that may need medical attention and ongoing assessments. I waited until my body would hurt or my skin would break down before I sought out any medical help. Over and over they would say, "Why have you waited so long to get medical attention?" In my eyes, if it weren't broken, you didn't need to fix it. I feel now that if I had been more involved in my care as a teenager, I would be more proactive with my health care needs today.

In closing, if I could give any advice, it would be that you are your best advocate. Let your voice be heard. Please get involved in your medical care and your education. After all, it is your life and you only get one chance at it. Ask for copies of your medical records because I've been told that records are destroyed after 7 years. I can tell you this is true from my experience. I called the hospital where I had undergone several surgeries as a child, and I was told there are no records of any surgeries or treatments.

This is crucial because other physicians will not want to treat you if they don't know what procedures have been done in the past.

Each of these personal recollections reflects the need for ongoing support and assistance in learning the skills and knowledge needed to function independently and enjoy an improved quality of life. The reflections also reveal the importance of a sustained and ongoing life course approach to fostering the development of developmental competencies for individuals with spina bifida to realize their lifetime goals and dreams for the future.[7]

The next portion of this article reports insights learned from youth and young adults about their lifestyle experiences in living with spina bifida. Their candid sharing of information about their lives provides greater understanding of the challenges they have and continue to face as well as the achievements they have attained. These insights provide a view of their life experiences that serve to provide health care professionals with a unique form of evidence for practice purposes.

INSIGHTS FROM THE TRANSITION EXPERIENCES OF MINNESOTA YOUTH

Researchers at Gillette Lifetime Specialty Healthcare Clinic in St Paul, Minnesota, have been conducting the first and second of 3 annual interviews with youth in transition for a research study entitled, “Examining the Trajectory of Transition in Adolescents and Young Adults with Spina Bifida” (University of Minnesota IRB 0802S273241). In this recent series of interviews, youth described their transition journeys. Most could describe their disability in general terms, could give a basic health history, and tell what their medications were for, even if they were unable to name them. Several young adults described being independent with their own physical self-care but they continued to be dependent on parents for all other aspects of their special needs management; however, they indicated that they were generally satisfied with these circumstances.

When youth were asked to describe the nature of their social relationships, they revealed a variety of different types. The continuum of their personal relationships ranged from social isolation or relationships limited to family members or caregivers, to active electronic communication, but scant face-to-face interactions with a few close friends to the rare experience of actually socializing with small groups of friends in the community.

The section that follows presents excerpts from the interviews that illustrate some of the common themes from this research investigation. These themes are

- Challenges in preparation for self-management
- Limited social relationships
- Awareness of cognitive challenges
- Cost of independence.

The themes gathered from this study mirror many of the issues raised in the personal reflections written earlier.

Challenges in Preparation for Self-Management

When asked what percentage of their own care the youth preformed and what percentage their parents performed, one young woman replied, “I do 100% of my self-care and my mother does the administrative stuff.” When pressed for detail, the young woman explained that she took care of her personal hygiene, dressed and fed herself, and had assigned household chores like doing the laundry, emptying the dishwasher, or caring for a pet. The caregiving activities performed by her mother, which she referred to as “administrative stuff” were dealing with paperwork, tasks that related to dealing with doctors, addressing insurance issues, ordering supplies, paying bills, and managing aspects of the youth’s health condition. Following this clarification, the youth was once again asked what percentage she felt she was responsible for and what percentage her mother did, she replied, “I do 100% of what I do and she does 100% of what she does.”

Some youth indicated an awareness of a need to adapt to new circumstances as they matured into adulthood. Most showed a general or vague concern about needing to learn more about managing their own affairs. For example, when discussing a young adult’s understanding of his or her health insurance plan, their responses typified the attitude of not seriously attending to it, as evidenced by the statement, “I just worry about it when I have to.” Instead, youth expressed a general awareness of the name of their health insurance plan but were unfamiliar with the details of their insurance plan coverage such as benefits available, copayment requirements, and the use of the appeal process in the event of a denial of services.

Some youth express a “stubbornness” about trying to do as much as they can for themselves, whereas others indicated an acceptance or desire to have someone else manage or comanage their lives. As one youth stated, “I’m not able to remember all my medications, so I’m always going to have to have somebody set them up for me.”

Strategies for money management were limited. These strategies included asking their parents for funds when out of money and performing household jobs for neighbors like watering plants or pet sitting to earn a little cash. Others indicated that they were learning about using an ATM or managing a checking account. A few young people indicated that they were unable to balance their bank accounts; instead, they relied on calling the bank to obtain information about their balance. For them, it was a convenient strategy to determine the amount of money available to spend.

In contrast, one young man reported his money management had improved over the previous year. He said he used to forget to check his account balance via his bank’s automated system before going out. However, negative consequences from that experience helped him learn to remember. As he remarked, “I’m a hands on learner. I can’t learn by demonstration of others.”

The study participants also reported having a range of household responsibilities. A few youth remarked that they were not required to perform household chores, whereas others reported having clear expectations of doing their “share” within the family and were treated no differently than their siblings. In addition, some indicated that beginning in their teens, they were responsible for ordering their medical supplies and making medical or transportation appointments.

Limited Social Relationships

Youth expressed feelings of social isolation from peers in postsecondary life with a dependence on family members for both planning and accompaniment in social activities. One young woman revealed the following insight about “fitting into” different social groups.

“I did not participate in diversity at my high school, and I do not plan to in college. In the past, I have had experiences where it appears to me that diversity starts working backward; [diversity groups] start creating more discrimination, although the result is positive. What ends up happening is that people start saying, ‘Hey, I am going to become your friend because you’re different’ versus the equality that it is suppose to be promoting, which is you’re my friend because you are another human being and we connect on this.”

One young man indicated that he had never been alone by himself. At school, he typically had a friend accompany him on the elevator in the event it got stuck between floors. His explanation was, “Cause she’s got a cell phone and I do too in case her cell phone dies, I can call for help.” By age 18, he could still not think of a situation where he might be comfortable being alone out in the community if he had the opportunity to do so.

Some youth reported changing their self-catheterization schedule as well as altering or disbanding their taking of medications so as not to call attention to their differences when with others. In one situation, both the young woman being interviewed and her brother had a disability. She revealed that when they infrequently visited people, they “didn’t feel comfortable” to have them see her and her brother with medications.

A sensitivity to appearing different from their peers or even extended family members resulted in neglect of self-care for some. This sensitivity to how others perceive them may explain the reluctance to use assistive technology or techniques that set them apart. That said, many in this group demonstrated a quick and ready adoption of technologies used to socialize. Perhaps because of the challenges of physically getting together with others, electronic communication networking through Facebook, Twitter, texting, and cell phones was popular. Although some used the technology for socializing, others had mastered the use of alarms, calendars, directions for finding their way, and preprogrammed phone books to augment their memory.

Awareness of Cognitive Challenges

Some youth with spina bifida were aware that they might hold a slightly different world view. They reported a reliance on verbal communication skills for navigating through situations, as they experienced difficulties with executive functioning, math, and reading comprehension. As one respondent stated, “I know how to be assertive. I think a weakness would be trying to verbalize kind of complex ideas. Sometimes I think I just see things a little bit different from some people.” Another challenge identified by youth was associated with time. As one young man stated, “I will think that 2 hours has gone by and it is like 4 or 5 hours has gone by.” When asked if the young man had adopted a strategy for tracking time, he said that people just reminded him of the time.

This young man reported he was aware of “cognitive problems such as having trouble getting thoughts out, learning disabilities, and difficulty with problem solving. He stated that he knows he is good “with verbal” and “not good with subtle hints.” He summarized his abilities by revealing, “I have a hard time communicating my needs. I am not in tune with my own body and my own mind.”

Way finding is another challenge encountered. Several youth talked about their concern for traveling alone. They were reluctant going alone to some place new or far away requiring additional efforts for travel. They were comfortable traveling alone to familiar places or those that were nearby.

Cost of Independence

Youth talked of their ambivalence in attempting to balance between their need for independence and their family. One young woman who had lived outside of the family home successfully, returned home primarily for financial reasons. Although she could make her living expenses, she did not have funds for discretionary spending. The young woman noted how her father had resumed “taking care” of her after she had moved back home. He shopped, cooked, cleaned, laundered the clothes, and basically did everything for her. He drove her to all of her medical appointments even though she had a car and driver's license. Although she wished for her lost independence, she was reluctant to negotiate with her father because she didn't want to hurt his feelings.

Another young woman who lived at home and whose supplemental security income contributed to the household expenses and grocery purchases, indicated that it was a good experience to know there was someone for her if she needed her parents. However, she has remained dependent on her parents and in effect has become reluctant to venture into the community and get engaged with outside activities. Her recognition of this problem, prompted her to say, "There are days where I've started to wonder what life would be if I lived on my own and had friends."

One young man in the study indicated that he did not want to be in charge of managing his health care saying it required too much effort. In effect, he allows others to decide when he should see doctors; although, when he has a health problem or need for medical supplies, he will inform the staff at the group home. His dependence on others for medically related needs is in stark contrast to other areas of his life as he stated, "They [group home staff] are responsible for probably 97% of my medical life. I'm in charge of 98% of my social and everything else." Just a year later, he indicated he was less satisfied with this arrangement.

In the course of these interviews it became evident that youth with spina bifida had opinions and experiences that were powerfully informative about how they perceive their place in the world. As this study has evolved, the respondents have become more self-disclosing with their responses. Our ability to probe more deeply has provided rich data for analysis.

RECOMMENDATIONS

As the "voices" of youth and adults with spina bifida have illustrated, living with a medically complex disability is difficult. However, this life journey can be made more manageable and meaningful with providing families from the very beginning with the access to services and supports to foster the development of their children's skill building in all life domains. The Life Course Model Web site described in other articles in this issue was designed to provide families with the resources to facilitate their efforts in this regard in these life domains: health self-management, personal-social relationships, employment, and income support.

This discussion raises the issue of how to accept reliance on others while maintaining a comfortable amount of independence.[12] Given the consumers' remarks about how much effort it takes for them to perform tasks independently, consumers, families, and care providers should strive to strike a balance between the benefits associated with independent task performance and the costs in time, effort, and personal stress.

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