TRANSITION TO ADULT HEALTH CARE FOR ADOLESCENTS WITH SPINA BIFIDA: RESEARCH ISSUES

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The increasing survival of children and young people with congenital disabilities such as spina bifida (SB) provides a challenge to health care systems globally about how best to respond to the multitude of health, developmental, and psychosocial needs of those affected by this complex disorder across the lifespan, not just in childhood and adolescence. The goal of transition to adult health care is to maximize lifelong functioning through the provision of quality, developmentally appropriate health care that continues uninterrupted as the individual moves from adolescence to adulthood. The objective of this article is to outline the type of evidence we have around transition to adult health care in young people with SB, and to identify what additional research evidence would help inform the development of models of clinical care for young adults.

Key Words: health transition; adult health care; developmental health care; spina bifida; preventive health care; satisfaction with care; health services

INTRODUCTION

The increasing survival of children and young people with congenital disabilities such as spina bifida (SB) provides a challenge to health care systems globally about how best to respond to the multitude of health, developmental, and psychosocial needs of those affected by this complex disorder across the lifespan, not just in childhood and adolescence [Dicianno et al., 2008; McLone, 2008; Sawyer, 2010].

Historically, as with most disorders of congenital or childhood onset, the clinical and academic leadership that has been responsible for the multitude of advances associated with improvements in survival and quality of life has been based within the realm of specialist pediatrics [McLone, 2008]. This expertise, whether embodied in neurosurgeons or nurses, in technical innovations in radiology, pharmacology, or surgery, or within the clinical and community networks that have been developed by pediatricians, is the result of the extent of clinical focus, coordination of care and academic commitment that has accompanied a critical mass of children and adolescents with SB within pediatric services. Since the 1960s, the dominant model for the delivery of health care to children with SB and other complex developmental conditions has been the subspecialty multidisciplinary clinic [Binks et al., 2007]. Whether judged by improvements in health outcomes as measured by survival or through measures of quality including aspects of family centered-care, this model of care has been rated as highly successful [Dicianno et al., 2008].

As children with SB mature into adolescents and young adults, they must graduate beyond pediatric services to the adult health care system. Specialist services are now well developed for adults with conditions of congenital or childhood onset such as cystic fibrosis (CF) and diabetes mellitus where there has been a sufficient critical mass of adults to warrant specialist services for many decades [Kennedy and Sawyer, 2008]. For other conditions such as chronic renal disease and haemophilia, services have been somewhat slower to develop [Geerts et al., 2008; Chaturvedi et al., 2009]. The development of models of adult health care for young people with disabilities such as SB has lagged behind service developments for adults with chronic medical conditions, despite the additional complexities of more pronounced functional disability that has been argued to render the need for comprehensive specialist health services even more critical [Sawyer, 2010].

For the developmental milestone known as transition to adult health care, there is a wealth of empirical evidence about the generic barriers and challenges facing individuals with congenital or childhood-onset chronic health conditions, their families and health care providers as they negotiate the move from child oriented to adult focused health care. This has been distilled into a series of policy statements that reinforce the importance of ongoing access to quality health care across the lifespan [American Academy of Pediatrics et al., 2002; Rosen et al., 2003].

Although there is some evidence about the barriers and challenges facing young adults with SB and other disabling
conditions as they transition to adult health services [Binks et al., 2007], there is a distinct lack of evidence about other important aspects which, if available, could inform the development of clinical care standards for young adults with SB. For example, there is little evidence about the extent of co-morbidities at different developmental stages and how this changes with age, about patterns of health care utilization in older adolescents and young adults with SB, or about health costs across the lifespan. In addition, there is a need for better understanding of variation in educational and workforce participation and social inclusion and whether this can be affected by different models of health care delivery, and about the relative value of different models of health care delivery across the lifespan. A further challenge for clinicians and researchers alike is to better delineate the similarities and differences facing young people with SB as they transfer to adult services, whether in comparison to other young people with developmental disability such as cerebral palsy (CP) or acquired brain injury or more generally when compared to those with chronic medical conditions such as diabetes mellitus or cystic fibrosis.

The objective of this article is to describe the evidence we have around some of these issues to make it clearer why greater development of clinical services is important for young adults with SB and what research would help inform this agenda.

TRANSITION VERSUS TRANSFER

The importance of transition from pediatric to adult healthcare settings for young people with special health care needs has been formally recognized for at least two decades, with policy statements dating back to the early 1990s [Blum et al., 1993]. While much of the transition literature has focused on chronic disease, there is growing appreciation of the importance of transition to adult health care for young people with disabilities such as SB [Binks et al., 2007]. The notion of transition to adult health care is one that generally involves a change in place from specialist pediatric to adult services and a change in orientation from family-centered to more individually focused services [Kennedy and Sawyer, 2008]. Typically, the term transition describes the period of preparation prior to and after the event of transfer, a term that describes the actual relocation from pediatric to adult health care including the transfer of health information [Sawyer et al., 1997]. This difference in terminology is not commonly differentiated in the literature or understood by clinicians, which risks downplaying the importance of the wider aspects embodied within the process of transition to adult health care and the more specific aspects of what is required to support engagement in adult health care services following transfer of healthcare. The implicit goal of transition to adult health care is to maximize lifelong functioning and potential through the provision of high quality, developmentally appropriate health care that continues uninterrupted as the individual moves from adolescence to adulthood.

HEALTH AND LIFE TRANSITIONS

SB has the capacity to affect every aspect of adolescent development, whether in terms of altered trajectories of physical growth and maturation [Liptak and El Samra, 2010], its effect on cognitive maturation and functioning [Dennis and Barnes, 2010], or in terms of psychosocial development [Holmbeck and Devine, 2010]. Thus, in addition to the usual developmental challenges faced by adolescents, young people with SB face an additional set of challenges that relate to the presence of the developmental disorder and its associated co-morbidities, whether in terms of limitations of mobility, education and employment [Buran et al., 2002; Olsson et al., 2007] or in relation to wider aspects of enacted stigma associated with disability, lack of social inclusion, poor social support, and low self-esteem [Kinavey, 2007; Van Naarden et al., 2006].

At its broadest, transition can be viewed as encompassing far more than simply ensuring that young people develop sufficient skills to manage their health within the adult health care system. Indeed, it can be seen to encompass a holistic set of goals that support the transition to adult life and that ensure access to a range of services to maximize life long functioning [Stewart, 2009]. Specifically in relation to health, transition can be considered to embody a set of related and overlapping constructs that include the principles underpinning the practice of adolescent medicine, the acquisition of self management skills, and knowledge about the health care system [Kennedy and Sawyer, 2008], as outlined in Figure 1. This schema somewhat echoes the

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**Fig. 1.** A schematic representation that shows transition (T) to adult health care placed at the intersection of the overlapping knowledge, skill sets and resources that constitute adolescent medicine, self-management of chronic disease and health care systems. Reprinted with permission from Kennedy A, Sawyer SM. 2008. Transition from pediatric to adult services: are we getting it right? Current Opinion in Pediatrics 20:403–309. © 2008 Lippincott, Williams & Wilkins.
equally complex landscape reported in a study that explored correlates of functional status, self-management, and developmental competence in adolescents with SB [Sawin et al., 2003].

While advocacy to ensure the breadth of services that support the continued functional and emotional well-being of young adults with SB is important, a critical task for health care professionals is to ensure that sufficient priority is given to making sure the health care system works well for these young people and their families across the lifespan.

HEALTH STATUS OF YOUNG ADULTS
In addition to detailed description of the specific health issues affecting children with SB, a number of recent publications have described the range of health issues affecting adolescents [Olson et al., 2007] and adults [Dicianno et al., 2008; Roebroeck et al., 2009; Webb, 2010]. Together, these papers provide a comprehensive review of the medical issues and psychosocial concerns experienced by adults, together with an account of the impact of SB on education, employment, and quality of life.

Publications such as these implicitly acknowledge the changing impact with age of the various health and developmental issues related to SB. While the impact of some of these issues will lessen with growing age, the impact of other issues will become appreciably greater in adult life, such as the extent of pain, the impact of SB on intimacy and sexual wellbeing, or the long-term consequences of obesity [Webb, 2010]. As with other health conditions of congenital or childhood onset, such changing patterns of morbidity and mortality across the lifespan are not well described in SB. Better understanding of how morbidity changes with age has important ramifications for the orientation of adult services which, like child focused services, must have expertise in aspects of both disease-specific and generic elements of health care [Liptak and El Samra, 2010]. For example, growing understanding of the extent that cardiac disease now contributes to mortality in adults with SB [Singhal and Matthew, 1999] should inform the development of clinical practice guidelines around cardiac assessment. Similarly, greater understanding of the impact of pregnancy in contemporary cohorts of women is indicated [Dicianno et al., 2008], given the expectation of greater numbers of pregnancy as more women survive to childbearing age.

Within the SB literature, there has been relatively little description of the common generic comorbidities accompanying chronic conditions in adolescence and young adulthood, such as mental disorder (e.g., depression, anxiety, and eating disorders) and health risk behaviors (e.g., tobacco and other substance use, risky sexual behaviors). Assessment of these developmentally significant factors, including sexual abuse, is encouraged, as there is evidence that older adolescents with special health care needs are at increased rather than decreased risk of such behaviors and mental health states [Siris et al., 2008]. The available evidence suggests that young people with SB have higher rates of mental disorder and issues with psychosocial adjustment than age matched controls [Appleton et al., 1997; Holmbeck and Devine, 2010].

UNMET HEALTH CARE NEEDS
There have been decades of research that show substantial unmet health care needs in children and youth with special health care needs [Homer et al., 2008], which is also apparent for SB [Liptak and El Samra, 2010]. In relation to transition to adult health care, a Dutch study showed that 15% of 18–22 year olds with CP were continuing to see their pediatric team, while 30% were attending an adult rehabilitation team [Roebroeck et al., 2009]. Fifty percent of these young people had not visited a rehabilitation team in the past year and three in four young people with CP with normal intelligence reported one or more unmet needs (a mean of four) for activities and participation, most commonly in relation to functional mobility [Roebroeck et al., 2009]. Perhaps surprisingly, 50% identified they needed more disease-specific information and information about the consequences of the disorder in adulthood. This may reflect a lack of orientation around the delivery of health information to young people through their parents. While there are few studies of patient satisfaction in families of children with developmental disabilities [Liptak et al., 2006], King et al. [1999] found that the receipt of health information was associated with parent satisfaction which was in turn associated with lower parent stress. Whether this association holds true for young adults with SB is unknown.

Data from Canada reporting the health status of adolescents and adults with SB, CP and acquired brain injury is particularly concerning [Young et al., 2006]. While the health status of adolescents with SB aged 13–17 years was similar to same-aged Canadians, the health status of young adults aged 23–32 years with SB was worse. The health status of all three disability groups was lower in young adults than in adolescents, which is contrary to the pattern in the general population, where self-rated health status is higher in young adults than in adolescents. That young adults with SB had the worst health status of all three groups was of added concern. One explanation is that young adults are less likely than adolescents to access health or other support services. This is also the case for young adults with chronic medical conditions such as diabetes and chronic renal disease, even when specialist services are available [Kipp et al., 2002; Chaturvedi et al., 2009]. While Young et al. [2006] showed that 22% of these Canadian young people with SB had been hospitalized in the previous year, it is not clear what proportion of these hospitalisations were for potentially preventable conditions (i.e., that might have reflected less than adequate health surveillance). Nor were differences in admission rates reported between adolescents and young adults [Young et al., 2006]. Interestingly, however, adolescents with SB were more likely to receive services from a greater number of multidisciplinary health professionals than adults. Most adolescents saw five different types of health professionals in comparison to adults, who most commonly saw two [Young et al., 2006].

The establishment of a new clinic for young adults with SB in the UK revealed that one of two young adults had unmet medical needs when first assessed, reinforcing the importance of access to an appropriate level of health care [Morgan et al., 1993]. Unmet medical needs are costly to the community as well as to individuals with SB. Kinsman and Doehring [1996] undertook a detailed study of preventable admissions because of secondary complications in a cohort of 98 adults with SB who had a mean of 3.6 admissions per patient (range 1–25) over the 11 year period of study. 47% of these admissions were considered to be due to secondary complications that generally resulted from common health concerns such as urinary tract infection and skin ulcers. While it is debatable what proportion of these secondary complications are truly preventable, prevention strategies exist for each nomi-
nally preventable condition which underscores the importance of self-management and regular health care monitoring. The extent of financial costs accrued from these preventable admissions [Kinsman and Doehring, 1996] provides yet further support for the need to focus more on reducing preventable morbidity in young adults with SB.

There are few studies of satisfaction with health care, and remarkably few in relationship to transition to adult health care. Young et al. [2006] reported that 92% of adolescents with disability who were continuing to receive care within a pediatric setting were satisfied with the health care they received in childhood, which was not dissimilar to young adults who had already transferred to adult, in whom 88% reported they were satisfied with childhood services. While a high level of satisfaction was reported in adolescents about current services (90%), only 74% of adults were satisfied with their current adult health care. Importantly however, adults who continued to receive care within a pediatric setting were the least satisfied with their current care (61%).

ACCESS TO QUALITY HEALTH CARE FOR ADULTS

The attributes of quality primary health care for children and youth with special health care needs are that it is accessible, family-centered, continuous, comprehensive, coordinated, compassionate, and culturally effective (www.medicalhomeinfo.org). A systematic review of the attributes of such quality primary care (that in the US is commonly referred to as a "medical home") suggests that children and young people with special health care needs have better health outcomes when they have access to this type of primary care [Homer et al., 2008]. In particular, they are about half as likely to experience delayed or forgone care and to have unmet health needs [Strickland et al., 2004]. Ensuring that there is adequate health insurance to pay for required services is more challenging in the US than in many other high-income countries, but is equally problematic in many low-income countries. Ensuring access to appropriate transportation and physical access (from parking lot to examination table) are equally critical. Ensuring that there are accessible community-based services to make an appropriate transition to all aspects of adult life including health care, work, and independence is as important for adults as it is for children and youth [Buran et al., 2002; Walker, 2008]. As health insurance in the US comes from either employment or disability benefits, help with obtaining one or both of these is an essential aspect of transition to adult health care.

The complexity and severity of health issues experienced by children and adolescents with SB means there is little doubt about the importance of access to specialist health services, with evidence of improved health outcomes when coordinated specialist health care is provided [Kauffman et al., 1994]. Given the extent of ongoing health issues experienced by adults [Webb, 2010], it should be no surprise that there is growing evidence to support a model of comprehensive coordinated specialist services for young adults as much as with children and adolescents with SB [Morgan et al., 1993; Bent et al., 2002]. However, it is apparent that in most high-income countries, while there are outstanding examples of comprehensive coordinated, specialist services for adults with SB, there is difficulty in ensuring that the same high quality of health care is widely available to all adults [Olsson et al., 2007; Roesbroek et al., 2009; Sawyer, 2010]. Given the importance of limiting health care costs to medical administrators and governments alike, researchers are encouraged to use health economic modeling to enable the costs of comprehensive coordinated specialist services to be compared to the savings that might be accrued were even a minority of secondary complications leading to hospitalization able to be prevented.

Given the complexity and severity of health issues faced by adults with SB, a critical challenge for the health care system for those affected is that adults can access both specialist services and primary health care, and that there is effective communication between the two levels of care to ensure that there is neither duplication nor omission of key services. There are various models of specialist services for both children and adults with SB [Sawyer, 2010]. For example, while in pediatric services, many clinics are coordinated by specialist surgeons, rehabilitation specialists are more likely to take the clinical lead in adult services. In Victoria, Australia, advocacy from the pediatric disability sector resulted in a recommendation to develop multidisciplinary clinics within adult health services to assist with the transition of young adults with complex medical needs from pediatric to adult health services. Four clinics were set up in both urban and rural centers in 2004 and 2005. In 2008, an independent review of these clinics [Ipsos-Eureka Social Research Institute, 2008] showed the major achievements included: clearer pathways into the adult sector; consolidation and further development of expertise in complex disabilities within the adult sector; a holistic model of care for young people; stronger relationships and information sharing between pediatric and adult health services; and better care and positive health outcomes for the majority of young people and their carers. However, multiple gaps and issues were also apparent. Some of these resulted from lack of preparation of young people and families, that might be expected to reduce with time as confidence in the service grows. Other issues reflected more challenging aspects such as long waiting lists and lack of retaining suitably qualified staff that may reflect chronic underfunding. The lack of access to particular specialist services and the lack of emotional support described by patients and carers alike may also reflect funding shortfalls, but may equally reflect a genuine lack of specialist services within the adult sector together with a lack of training of sufficiently qualified staff within the adult health care sector.

A major feature that differentiates SB from other complex congenital conditions is the extent of surgical involvement that is required by specialties including neurosurgery, urology, and orthopedics [Liptak and El Sunra, 2010], with this issue providing updated information on evidence-based standards of care within each of these disciplines. Some of the surgical procedures performed in SB are highly complex, surgical procedures such as the urological procedures of cecostomy, bladder augmentation, and appendix-vesicostomy; Clayton et al., 2010] are specific to pediatric surgery. The lack of experience of such techniques by adult surgical specialists means that the care, maintenance, and surveillance of these altered tissues can be extremely challenging for adult services. Ensuring that adult services develop an appropriate level of specialist surgical expertise is a major challenge for the SB field [Webb, 2010]. Another major feature that differentiates SB from other chronic medical conditions such as cystic fibrosis, diabetes, or hemophilia is the complex cognitive deficit which can subtly (or not so subtly) impact on self-management skills and autonomy and can highly frustrate adult providers who may be less experi-

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enced in managing cognitive limitations in young adults [Dennis and Barnes, 2010]. Some of these difficulties could be overcome by stronger continuities between specialist pediatric and adult programs. Similarly, stronger academic linkages could help promote a culture of innovation, evaluation, and quality improvement.

As yet, there is no evidence that supports one model of specialist care over another. Rather, evidence supports access to comprehensive, coordinated and developmentally appropriate specialist services across the lifespan. While there is no evidence to support transferring young people at any particular age, there is increasing consensus that maintaining young people within adolescent oriented services is appropriate while they are still at school [Sawyer et al., in press].

TRANSITION TO ADULT HEALTH CARE

There is more evidence around the type and extent of generic barriers and the various challenges faced by individuals, families, and indeed health care providers in managing adolescents with complex chronic conditions and in ensuring a seamless transfer to adult services [Binks et al., 2007; Kennedy and Sawyer, 2008]. While many of these barriers relate to young people and parents not being sufficiently prepared to transfer to adult oriented services, other barriers reflect a lack of trust between pediatric and adult providers that historically, in the absence of adult services, might be one explanation for the difficulty described by pediatricians in ‘letting go’ of their patients [Binks et al., 2007]. While it may be expected that the development of more comprehensive, coordinated and developmentally appropriate services for adults with SB will in time help build relationships and trust between pediatric and adult providers, more explicit mechanisms such as regular meetings and shared protocols are also recommended [Duguépoux et al., 2008].

The increasing survival of adolescents with complex childhood onset disorders who are transferring to various adult oriented services in significant numbers suggests the importance of educating both adult and pediatric providers about the generic principles of practice of adolescent health and medicine [Sawyer et al., 2007; Kennedy and Sawyer, 2008; Feddock et al., 2009] at both undergraduate and postgraduate levels.

It is also apparent that patients and families require more explicit prepara-

tion than many are currently receiving. In the US, a population-based study of parents of adolescents with special health care needs described that only half reported that their child’s doctor had talked about changing needs in adulthood, and that only one in five had ever discussed transferring their child’s care to an adult provider [Lotstein et al., 2005].

The development of transition clinics is one approach to more explicitly prepare young people and families to transfer to adult services. In general, these provide more opportunity for young people to engage more autonomously in their own health care while also providing opportunities to meet various members of the adult health care team [Binks et al., 2007; Chaturvedi et al., 2009].

Yet, even in the presence of specific transition clinics, young people report inadequate preparation [Chaturvedi et al., 2009]. Thus, repeated discussion with the young person and family is recommended in order to build expectations and confidence about the timing of transfer to adult services, just as parents would be expected to repeatedly discuss various educational transitions with their children. Introducing families to the concept of transition to adult health care should arguably begin at the time of diagnosis or in infancy [Peterson et al., 1994]. More specifically, a stronger focus on supporting young people and families to acquire the self-management skills that will enable them to function more independently as they mature is required [Buran et al., 2002; Binks et al., 2007]. Young people with disabilities and their parents may also benefit from learning from each other, whether by participating in peer support groups, through social networking sites, or through recreational activities such as camps.

CLINICAL LEADERSHIP

If the goal of transition to adult health care is to maximize lifelong functioning and potential through the continued provision of high-quality developmentally appropriate health care across the lifespan, one approach to determine how well this is being achieved is to measure the extent of variation in both health outcomes and quality indicators. The example of cystic fibrosis is interesting to consider [see also Liptak and El Samra, 2010] as careful exploration of the putative explanations of better health outcomes in the US has led to major efforts in generating a culture that supports continuous quality improvement around efforts to standardize care delivery. The advocacy and support of this agenda by the Cystic Fibrosis Foundation has resulted in an explicit commitment to increasing the quality of care delivered by CF centers in the US, increasing the standardization of CF care across the US, together with the promotion of patient and family centred care. The result of this work has been significant improvements in patient outcomes [Britton et al., 2008; Kraynack and McBride, 2009]. Accreditation of CF centers by the CF Foundation requires a transition program to be in place, which includes the presence of an adult CF program. This approach is garnering significant interest beyond cystic fibrosis. The general emphasis on quality improvement and more standardized care and the specific emphasis on transition to adult health care would be equally appropriate for clinical leaders and advocacy organizations that support people with SB to consider.

FUTURE DIRECTIONS

Such approaches require both organizational and management support as well as clinical and academic leadership. Promoting the development of specialist adult services for young people with SB and other developmental disabilities is a strategy that should be pursued in order to ensure access to comprehensive specialist services for individuals with SB. This is critically needed now by young adults with SB, but it is equally a strategy that will help develop a critical mass of patients around which future research agendas must be based to better delineate the changing nature of health issues experienced across the lifespan. There is an urgent need for research on the multiple issues that involve the transition of people with childhood disabilities into adulthood. Because the population of people with SB is large and because their needs involve multiple disciplines, evaluating programs that facilitate the transition of people with SB into the adult health care system may provide a model for effective approaches to this problem. This research needs to consider rehabilitative approaches and issues pertaining to the continuity of care. A research agenda focused on transition issues is required to understand how best to remediate these complex congenital disorders across the life span.

REFERENCES

American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians – American Society of Internal Medicine.


