Youth, adults and transitional patients with spina bifida: a multidisciplinary approach to facilitating the passage from paediatric to adult care.

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Children and adolescents with spina bifida – a unique population

Spina bifida (SB), meaning “split spine” in Latin, is the most common neural tube defect and the most severe congenital disorder compatible with life. During the third to fourth week of embryonic development, the central nervous system begins as a neural plate and fuses into a neural tube. SB is the failure of this neural tube to close and results in three different categories of defects: occulta, meningocoele and myelomeningocele, the latter being the most severe and the focus of this article. Fifty years ago, the prognosis of an infant with myelomeningocele was grim as only ten percent were expected to survive their first year. Today, survival into adulthood exceeds 85% due to advances in the management of complications. While this is certainly a success in neurosurgical medicine, deterioration in adulthood is common in these survivors. One of the greatest challenges remaining is the continuity of care between paediatric and adult services, which requires a functional multidisciplinary team.

Morbidity in the aging SB population is a complex combination of physical, emotional and developmental obstacles. The long term physical effects of SB include muscle weakness or paralysis below the level of the lesion, loss of sensation, and loss of bowel and bladder control. Following repair of the myelomeningocele, 70-80% of infants develop some degree of hydrocephalus, which is treated by implanting a shunt shortly after birth. If hydrocephalus is untreated, or if the shunt later malfunctions, excess fluid may result in permanent brain injury, seizures, or blindness. Unrecognized shunt malfunction is the leading cause of death in adults with SB. This is a topic that adult-centered physicians must be educated in when caring for patients with SB.

Concerning the emotional development of children with SB, independent mobility is an important factor in determining quality of life. The level of the lesion corresponds to prospective independent mobility. For lesions above L2, loss of quadriceps and ilioosas muscle function is typical and wheel chair dependence is expected. However, lower lumbar and sacral lesions are usually compatible with ambulation, for which early physiotherapy and orthopedics consultations are crucial. Ambulation may mitigate the decline in activity through adulthood resulting from obesity, spinal and foot deformities including scoliosis and clubfoot, and respiratory compromise. In a 2004 cohort study of 117 patients, 30% were ambulatory at 30 years follow-up, of which 88% had lesions at L5 or below. This highlights the importance of early intervention and appropriate rehabilitation in response to changes that occur over time.

Another important area of treatment for people living with SB is the lifelong management of urinary incontinence and impaired sexual function. Sexual function may be reduced due to impaired neurological innervation, or the psychological effects of negative self-image and incontinence anxiety. However, there are two common misconceptions that are held more by medical students and doctors than by the young adults themselves: that people with SB cannot be sexually active, and that they cannot have babies. Both are untrue. Healthcare professionals should provide the proper education and counseling of safe sex and contraceptive services and should address the increased risk of latex allergies. Family planning services and pap smears should also be readily available to patients with SB, and must be sensitive and specific to the emotional and physical needs of this unique population.

As recently as twenty-five years ago, young children with developmental problems, including SB, faced an isolated homebound existence or were placed in often inadequate custodial facilities. A literature review also reveals that children with SB commonly have language deficits that often go unrecognized despite average intelligence. In response, early interventions focusing on psychosocial and educational models have been emphasized. Failure in independent self-care has been found to be a hindering factor for employment in young adults with SB. It is therefore of vital interest to understand the factors leading to this failure in self care, whether they be physical, emotional, or developmental in origin, and to facilitate functional transition to adult life.

Issues surrounding transition

Knowledge and communication by paediatricians

A study by Binks et al. outlines several barriers that prevent successful health care transition from paediatric to adult medicine for young adults with SB. One key point is that it is often difficult to sever the relationship between the paediatrician and the patient. The paediatrician is seen as a trusted person, and builds trust and relationships with the entire family; this is the nature of paediatric care. Thus, there is a lack of incentive for all involved to discuss the process of leaving the paediatric system to transition to adult care. This lack of communication inevitably results in a delay of the transition process until paediatric services are no longer applicable or available – in Ontario, when the patient turns 19. This is contradictory to the literature, which suggests that transition should begin as early as possible – by the ages of 14-16.
We believe there are several components necessary to facilitate an effective earlier transition. First, it is essential that the paediatrician must have a realistic expectation of the duration of their patient's care, since their expertise does not always translate to effective management of adults. It is in the best interest of the patient for the paediatrician to relinquish the patient's care.

Second, it is essential that care is taken by the paediatrician, with help of a multidisciplinary team, to prepare the child as best as possible for the physical and emotional challenges of adulthood. As mentioned, they include (but are not limited to), sexual health, social behaviours, alcohol and drug use, body image, mobility, employment strategies, anxiety and depression. Here, it is crucial for the information to be tailored to the patient's age and stage of mental and physical development, which highlights the importance of expertise from multiple disciplines.

Third, it has been shown that young adults with SB rarely feel comfortable navigating the adult health care system. Therefore, they are less able to advocate for themselves and are hindered in seeking the best possible care. Often, the paediatrician will continue to care for the patient. This has resulted in poorer health statuses of young adults with SB as compared to age-matched Canadians, whereas children with SB have the same health status as their Canadian counterparts. This inequality is due in part to a lack of preparation of the patient by their paediatrician and health care team. Therefore, early in the transition process, the patient must be made aware of the differences between paediatric and adult medicine, as well as the structure and function of the adult medical system.

Training and abilities of adult care providers

The second barrier outlined by Binks addresses the perceived lack of knowledge of adult care providers surrounding the needs of young adults with SB. They may have little to no exposure to childhood diseases or their sequelae in adulthood. Young et al. notes that paediatricians are often concerned that their adult counterparts do not fully understand or appreciate the nature and extent of SB. Additionally, Sawyer et al. found that paediatricians specializing in SB felt that adult care providers did not fully grasp the needs of this population.

An ideal approach to transitioning would allow for several meetings between the paediatrician and future adult care provider, patient and patient's family before the transition begins. These meetings would ensure several things. First, the family physician would receive all relevant information about the patient from three different sources: the paediatrician, the family and the patient. They would be more aware of the patient's ability to manage their condition, as well as the role the family plays in the patient's care. In such a way, the family physician is educated and may prepare to face the specific challenges of dealing with a childhood disease progressing into adulthood.

Transition organization – multidisciplinary approach

The ongoing goal is to ensure that quality health services are available to the patient with SB, given their physical, mental and emotional needs. Therefore, an organized, cohesive and coordinated system must be in place during and after their transition to adult care. Overall health of young with SB is reported to be much higher than their adult counterparts. This indicates that adults with SB are not receiving adequate care. Professionals seen by patients with SB include nurses, speech therapists, occupational therapists, physical therapists, dentists and many others, but the variety of care received is much richer for children with SB than adults.

Furthermore, in a study by Sawyer et al., several specific issues with the transition process itself were highlighted: the time gap between receiving paediatric and adult care, the sentiment that the adult health care practitioners' skills were insufficient (especially at the beginning), and the lack of reassured permanence of their care. Coordinated multidisciplinary centers could help to address these issues.

Studies have outlined different transition programs which include transition clinics. A study by Westwood, et al. showed that over 90% of children and adults with cystic fibrosis, another disease in which it is common to transition from paediatric to adult care, felt that a transition clinic would be helpful. These clinics unite family doctors, paediatricians, families and patients, and can provide links to additional resources such as counsellors and psychologists. With the current technology in electronic media that is available, the feasibility of meetings between multiple individuals and disciplines becomes possible with conference calling and other internet-based solutions. The scope of practice may no longer be strictly limited by location, as these online networks become available to all professional communities.

The ultimate benefit of such multidisciplinary clinics or teams would be to all patients with chronic paediatric conditions, who, thanks to advancing medical technology, are growing into their adult years.

Conclusion

Shunt problems and spinal cord-related symptoms typically account for a substantial amount of morbidity affecting young adults with SB and may be avoided if recognized early in their course. There are, however, also many important health education, vocational, and psychological issues that are specific to this age group. The transition from paediatric to adult care presents a huge challenge to the health care system itself. It must involve adequate resources and educated health care professionals to enable these paediatric patients to take the appropriate steps towards independence, maturity, and adult fulfillment.
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References