

Recent advances in spina bifida care extend life and improve quality of life

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Spina bifida (myelomeningocele) is the most common, permanently disabling birth defect compatible with life. In a collection of articles, published in the *Journal of Pediatric Rehabilitation Medicine*, experts describe important advances made in the care of spina bifida patients that extend life and improve quality of life.

Globally, it is estimated that more than 300,000 babies are born each year with neural tube defects. However, there is substantial global variation in birth prevalence rates. In the United States, for example, birth prevalence is seven cases per 10,000 live births, whereas in certain regions of the developing world, it has been reported to be as high as 125 cases per 10,000. Individuals born with spina bifida historically had high mortality in childhood, but advances in neurosurgery, genitourinary surgery, gastroenterology, and physical medicine and rehabilitation have led to marked increases in survival. Seventy-five percent to 85% of individuals with spina bifida now survive into adulthood.

While immigration from Latin America is often a result of social determinants, immigration itself can be understood as a social determinant of health (SDH). The Centers for Disease Control and Prevention (CDC) has stated that when attempting to mitigate racial/ethnic health disparities, it should be remembered that SDHs effectively determine longevity and quality of life (QOL). Subsequently, investigators have begun to recognize existing disparities through the use of the CDC National Spina Bifida Patient Registry (NSBPR). In the face of these documented health disparities among [minority populations](#) with

spina bifida, the need for timely and culturally-competent studies among Hispanics/Latinos, who have the highest spina bifida prevalence, is self-evident.

Furthermore, social variables have been linked with worse outcomes in other national patient registries, illustrating the importance of accounting for SDH in multicenter comparative analysis. Therefore, accounting for these individual level differences becomes even more critical when making comparisons within spina bifida care – a condition with a known ethnic/racial gradient in incidence.

"For some minority populations, precursors to poor spina bifida-related outcomes may be best conceptualized through a 'three-hit' model. Whereas the first two 'hits' are experienced prenatally (i.e., the neural tube lesion itself and its environmental exposure to amniotic fluid), the 'third hit' may involve combinations of additional genetic and/or non-genetic exposures, such as unfavorable SDH," explained Jonathan Castillo, MD, MPH, Director of the Spina Bifida Program at Texas Children's Hospital, who served as guest editor for the issue. "As we face an increasingly [global community](#), with growing travel and immigration, fresh approaches will be required, such as community-based participatory research and transnational learning collaboratives, in order to address the emerging global challenges."

With so many children with spina bifida living longer, it has become increasingly important to manage their transition from childhood to adulthood. In an article that addresses this situation, lead author Betsy Hopson, MSHA, of the Spina Bifida Program, Children's of Alabama (COA), University of Alabama at Birmingham, and colleagues describe the development and implementation of the Children's of Alabama Spina Bifida Lifetime-Care-Model. They critically evaluated the team's initial ten-year experience in the provision of care for spina bifida-affected youths transitioning into an adult care environment. This article

describes early failures that led to an evolution in approach and implementation of a Lifetime-Care-Model that result in a smooth transition between all phases of life.

"Early in our process of developing a lifetime model, we found that [patients](#) with spina bifida were not ready for adult healthcare or adult life in general," explained Ms. Hopson. "The majority of our patients reached adult age but were not engaging in typical adult behaviors such as finding a job, developing relationships, or even leaving their homes very often. This research highlights our efforts to prepare patients for adult life through specific goal setting and interventions at an early age."

In 2010, members of the pediatric team at COA began to evaluate limitations in access to care for patients with spina bifida at various stages of life. Through clinic surveys, observations, and caregiver reports, a Lifetime-Care-Model was developed and implemented. Partnerships were made with adult medicine colleagues to create an interdisciplinary model at each stage. The program has now evolved to include standardized care protocols.

The experience at COA suggests that patients with spina bifida benefit from continuity of care throughout their lifetime. This research describes the importance of pediatric providers and adult providers working together for patients as they age, patient and family preparation and readiness, a functioning bowel management program, and an approach that emphasizes capability and individual responsibility rather than anything related to specific medical procedures or interventions.

"Transition has become a 'hot topic' in medical care for patients with lifelong complex conditions," added Jeffrey P. Blount, MD, Chief of Pediatric Neurosurgery and Professor of Neurosurgery, Department of Neurosurgery, University of Alabama at Birmingham, who led the investigation. "We found that the transition encompasses more than a

change in the location of healthcare delivery, but more importantly involves a process for ensuring the patient and their family are ready for that change. We hope that other institutions may adapt and build upon it to create programs unique to their specific patient needs."

Patients with spina bifida often have hydrocephalus (excess cerebrospinal fluid), and shunting is frequently used to treat the condition. But how does this affect the development of children with spina bifida? Amy J. Houtrow, MD, MPH, Ph.D., Associate Professor and Vice Chair, Department of Physical Medicine and Rehabilitation, University of Pittsburgh, and colleagues evaluated the impact of hydrocephalus and shunt placement for differences in developmental outcomes at 30 months of age for children with spina bifida enrolled in the Management of Myelomeningocele Study (MOMS).

Children with no hydrocephalus (27), shunted hydrocephalus (108), and unshunted hydrocephalus (36) were compared at 30 months of age on several developmental scales. Generalized linear models were used to adjust for factors significantly different between the groups at baseline. Additional analyses were conducted to evaluate the impact of the severity of hydrocephalus.

Investigators initially noted statistically significant differences between the three groups. After controlling for factors such as premature birth or large ventricles (a sign of hydrocephalus) during prenatal screening, no statistically significant differences were identified. However, in sub-analyses, children with more severe hydrocephalus fared worse. The investigators concluded that there were no neurodevelopmental differences with children enrolled in MOMS across the three groups after adjustment for characteristics not attributable to shunting that differed between the groups. Severity of hydrocephalus was associated with poorer neurodevelopmental outcomes.

"This is important to neurosurgeons who are treating babies with [spina bifida](#) who have hydrocephalus because it helps them recognize that putting in a shunt doesn't mean that the child's developmental outcomes will be poor," noted Dr. Houtrow. "It is the severity of the hydrocephalus, not the shunt itself, that predicts worse developmental outcomes. This study sets the stage for additional research to inform decision-making and can aid neurosurgeons in guiding families regarding shunt placement for hydrocephalus, an important medical and surgical decision."

More information: Jonathan Castillo et al. Social determinants of health and spina bifida care: Immigrant and minority health in an era of quality of life and multicenter comparative analysis, *Journal of Pediatric Rehabilitation Medicine* (2018). [DOI: 10.3233/PRM-180484](https://doi.org/10.3233/PRM-180484)

Betsy Hopson et al. The development of a lifetime care model in comprehensive spina bifida care, *Journal of Pediatric Rehabilitation Medicine* (2018). [DOI: 10.3233/PRM-180548](https://doi.org/10.3233/PRM-180548)

Amy J. Houtrow et al. Comparing neurodevelopmental outcomes at 30 months by presence of hydrocephalus and shunt status among children enrolled in the MOMS trial, *Journal of Pediatric Rehabilitation Medicine* (2018). [DOI: 10.3233/PRM-170481](https://doi.org/10.3233/PRM-170481)

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