

## Commentary

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Spina bifida, myelomeningocele, was recognized by the ancients. Few survived. The mortality rate in the 1950s approached 90%. Today, between 70,000 and 100,000 individuals with spina bifida live in the United States.

Spina bifida is a birth defect, the result of the failure of the neural tube to close early in embryonic life. Failure of the neural tube to close also causes the Chiari II malformation and it is this malformation that makes spina bifida different from traumatic paraplegia. The Chiari II malformation is the principle cause of early mortality. This cranial malformation inflicts hydrocephalus, with all its complications; the learning disabilities, attention deficits, and hindbrain neurological abnormalities that occur as a result of hydrocephalus and the Chiari II malformation are now believed to contribute to some of the challenges individuals face to acquire the skills needed to live independently.

In the late 1950s, a son was born with spina bifida to the Holter family. The father, John, an engineer invented a small implantable valve that allowed the diversion of CSF to body cavities outside the central nervous system. This device, the shunt, dramatically changed the outlook for these infants. With the shunt in hand during the 1960s there was great enthusiasm for aggressive treatment of the newborn and the survival rate of the infant approached 90%.

By the 1970s concerns about the disabilities of these children began to mount. Many, led by John Lorber, a neurologist in the UK, felt that these infants were a burden to their parents, society and themselves. This led to selective treatment of the newborn based on criteria felt to be predictive of the child's outcome. As high as 70% of the newborns were selected for non-treatment. A failure of this management was the survival of a child who had been selected for non-treatment. Success was insured by denying food and water to the selected infants and most died within weeks. Today, in most centers, viable newborns with spina bifida are aggressively treated. However, the debate continues. Recently in the Netherlands 22 newborn infants with spina bifida were given lethal injections by pediatric intensivists. As adults, especially physicians, making life and death

decisions based on what they feel is a life of quality should disturb everyone.

By the 1980s it became apparent that the problems did not end with survival of the infant. Loss of function from a variety of causes led to the majority of teenagers being much more impaired than they were as newborns. Complications of the management of hydrocephalus, renal failure, tethered cord, and scoliosis are a few of the causes. During the 1980s the miracle of CT and MRI scans gave new insight into the causes and treatment of deterioration. Today we know that most deterioration in function has a treatable cause and function can be preserved into adulthood.

Continence of bladder and bowel is a major concern for the family, and for some, outweigh concerns for intellectual development or ambulation. Clean Intermittent Catheterization (CIC) is a major contribution to their quality of life. Deterioration in renal function has almost disappeared. It may even be the most important advance that has allowed children with spina bifida to enter mainstream education.

The single most important contribution in regards to care of the child, infant through adolescent, has been the multidisciplinary team. Pediatric specialists, pediatricians, surgeons, nurses, therapists, social workers, are all in the same place at the same time, at regular intervals with good data, and are monitoring the child's progress.

The cause of spina bifida is "multi-factorial", which means that we are not sure but it appears to be some mix of nutritional and genetic factors. Recurrence within families and animal models support the genetic cause. The amazing discovery that a vitamin, folic acid, can reduce the recurrence rate by 70%, supports the nutritional cause. It is interesting that in laboratory animals, folate blocks the genetic cause. The evidence for folate is so compelling, the Centers for Disease Control (CDC) now recommends that all women of childbearing age take a multi-vitamin that contains folic acid. Folic acid, general improvement in nutrition, and women having the right to terminate their pregnancy has resulted in a significant decline in the incidence of spina bifida.

As we look back on almost 35 years of treatment new challenges are apparent. Our clinic started in 1975 with about 65 families. Today there are 1500. The adults in their 30s reveal some of the challenges for the future. Seventy-five percent (75%) of the infants have survived into their 30s. Complications of hydrocephalus continue to inflict mortality on this population. The good news: 80% have graduated high school and 50% have gone to college. Several have degrees. The not so good news: only 15% have full time employment and 80% are living with their parents, sitting in the living rooms of ageing parents. Where will they go when their parents die?

Each year thousands of young adults graduate from children's hospitals around the world with chronic illness and significant disabilities. This population that did not exist 40 years ago presents a new problem. In almost every city that I have visited around the world, the transition to adult medical care and independent living for this population is a major problem. Many of the adults, for a variety of reasons, have failed to acquire the skills needed to live independently and the medical community that treats adults is unprepared to deal with the numbers and the medical problems these young adults present. What is the answer? The transition process needs to begin much earlier in adolescence. Pediatric specialists must assist their adult counterparts in gaining the experience needed to become comfortable with this population. We need to encourage adult specialists, especially in the surgical specialties, to become interested in this difficult problem. Working with parents, independence can be acquired for most. The literature shows that camping is an excellent method for teaching independence skills to children with disabilities.

What about the future? Will the problem of hydrocephalus be solved or will regeneration of the defective nervous system be possible? Like Yogi said, "making predictions is hard, especially about the future".

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David McLone is a pioneering pediatric neurosurgeon who began performing ventricular shunt placements in the early 1970s. In 1977, he established the Children's Memorial Spina Bifida Clinic in Chicago and served as the director for 30 years along with a dedicated group of surgeons who subsequently joined him. During his tenure, he became concerned when he saw many adults with spina bifida who were inactive, unemployed, and still very dependent on their parents. Consequently, he established the Village Foundation, gained the support of the Chicago community, and built a 14 unit fully accessible apartment building which also houses a program to help young adults with spina bifida live independently. Moreover, his work with the Village Foundation has established Camp Independence, teaching life skills in a camp setting for children and youth with Spina Bifida. His commitment to this population has led to a loyal following of patients, families, and trainees across America and the world.