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REVIEW

Multidisciplinary spina bifida clinic: the Chicago experience

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Abstract

Open spina bifida (open SB) is the most complex congenital abnormality of the central nervous system compatible with long-term survival. Multidisciplinary care is required to address the effect of this disease on the neurological, musculoskeletal, genitourinary, and gastrointestinal systems, as well as the complex psychosocial impact on the developing child. Individuals with SB benefit from the involvement of neurosurgeons, orthopedic surgeons, urologists, physical medicine and rehabilitation specialists, pediatricians, psychologists, physical/occupational/speech therapists, social workers, nurse coordinators, and other personnel. Multidisciplinary clinics are the gold standard for coordinated, optimal medical and surgical care. Ann and Robert H. Lurie Children's Hospital, formerly known as Children's Memorial Hospital, was one of the first hospitals in the USA to manage patients with this complex disease in a multidisciplinary manner. We describe the longitudinal experience of the multidisciplinary Spina Bifida Center at our institution and highlight the advances that have arisen from this care model over time. This clinic serves as an exemplar of organized, effective, and patient-centered approach to the comprehensive care of people living with open SB.

Keywords Birth defect · Fetal repair · Folate fortification · Myelomeningocele · Neural tube defects

Introduction

Spina bifida (SB) is a complex neural tube defect with an incidence of approximately 1 in 1000 and a prevalence of 39 infants per 100,000 live births [1–3]. The causes of SB are heterogeneous, including chromosome abnormalities, single-gene disorders, and teratogenic exposures, though the cause is usually unknown [4]. An estimated majority of 70–80% of cases result from inadequate folate (vitamin B9) consumption before neural tube closure at 25–28 days of gestation [5, 6]. The recommended folate consumption is 400 mcg per day in the periconceptional period [7, 8]. Folate is generally provided via folate supplements or fortification of staple foods such as wheat and maize [9, 10]. By 2019, only 56 countries had implemented folate fortification, such that only 23% of folate-preventable SB was averted [5]. Early prenatal diagnosis of SB through high-resolution ultrasound allows women to prenatally seek information regarding the long-term impact of this disease on the patient. They are counseled regarding treatment options, including fetal surgery to close the open defect before birth versus postnatal back closure or termination of the pregnancy [11]. Access to early prenatal care is limited in certain communities, predominantly those that are low-income and with racial/ethnic minority groups [12, 13].

Although the US-implemented folate fortification of cereal grains in 1998 and voluntary fortification of corn masa flour in 2016[14], the volume of children with SB at specialized children's hospitals in major cities, such as Chicago, remains high. Survival has been reported at 87% at 1 year and 78% at 17 years [15]. Of the many types of SB, open SB, also known as myelomeningocele, is the most severe form, involving potential spinal cord dysfunction with concomitant brain abnormalities. Individuals with open SB often have Chiari II malformations, which may predispose

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them to the development of hydrocephalus [4]. Children with a thoracic level lesion experience greater lower extremity dysfunction relative to those with a lumbosacral lesion, as exposure of neural elements superiorly promotes the greater potential for injury [16]. Children often frequently experience bowel and bladder dysfunction given deficits of the sacrococcygeal nerves [4]. Medical and surgical needs are extensive and may change as the patient ages and comorbidities develop. Moreover, individuals with open SB have diverse psychosocial needs involving education, work, independent living, social support, and anxiety and depression [17]. Due to these myriad factors, open SB is generally associated with decreased health-related quality of life [17].

No single medical provider is trained in all aspects of care required by a patient living with open SB [18]. Individuals with SB benefit from the involvement of neurosurgeons, orthopedic surgeons, urologists, physical medicine and rehabilitation specialists, and pediatricians [19]. The average child with myelomeningocele undergoes 0.62 surgeries per year, while those over 18 years old undergo 0.19 surgeries per patient per year [20]. Moreover, specialists in physical, occupational, and speech therapy; nutrition; psychology; and social work are often involved in the care of children with SB [19]. Multidisciplinary clinics are the gold standard for improving care coordination and cohesiveness across the varied and complex needs of these patients. At present, there are more than 100 multidisciplinary SB clinics in the USA [21]. Our institution, Ann and Robert H. Lurie Children's Hospital (Lurie Children's), formerly known as Children's Memorial Hospital (CMH), was one of the first hospitals in the US to create a multidisciplinary SB clinic. In this manuscript, we describe the longitudinal experience of our multidisciplinary SB Center and highlight the lessons learned.

History and origins

Hydrocephalus was the predominant etiology of morbidity and mortality for individuals with open SB until the 1950s [22, 23]. The introduction of the valve for cerebrospinal fluid (CSF) shunt by John Holter, an engineer who had a son with open SB [24], allowed effective treatment of hydrocephalus, which significantly improved the long-term survival of this complex disease [22]. Some individuals, such as John Lorber of Sheffield, England, advocated for selective treatment of infants with open SB under the assumption that many burdened themselves, caregivers, and society [25–27]. Lorber and similarly minded colleagues indicated that active treatment should only be utilized for infants who were thought to have a reasonable chance of a favorable neurologic outcome, accounting for only 30% of infants with open SB [26, 28–30]. The rationale behind this selective treatment approach was to avoid treating patients who would survive with severe physical disability[31, 32]. Infants with paraplegia with no innervation below L3, spinal curvature, macrocephaly, perinatal cerebral injury, or other congenital abnormalities were proposed to be selected for nontreatment [26, 33, 34]. Children who developed meningitis or ventriculitis after closure and children with mental and neurological deficits who experience any life-threatening episode were also categorized in the nontreatment group [26, 29]. Nontreatment involved withholding of antibiotics and artificial feedings while providing supportive nursing care and pain management, as survival of these children was not a goal [26, 28]. Nearly all children receiving nontreatment died within the first year [25, 27, 30, 35]. In rare cases, children who survived to 6 months of age had treatment initiated [32].

In opposition to the notion of nontreatment by Lorber and colleagues, many health professionals, including Drs. Anthony Raimondi and David McLone at Children's Memorial Hospital of Chicago, advocated for active treatment in all children with SB [32]. McLone and colleagues recognized the complex needs of individuals with SB and their caregivers and the evolution of these needs over time. Multidisciplinary clinics aligned with outpatient care for children with SB were established in most states in the 1970s and 1980s [36]. These clinics sought to integrate care for SB by housing various surgical and medical specialties, allied health professionals such as nurses and therapists, and social workers [37]. Anthony J. Raimondi, the chief of neurosurgery at Northwestern University and head of pediatric neurosurgery at Children's Memorial Hospital, established the multidisciplinary Spina Bifida Clinic in 1972, one of the earliest such clinics in the USA [38]. This clinic included specialist doctors and nurses in neurosurgery, orthopedics, and urology. After Raimondi stepped down in 1977, David G. McLone was appointed head of pediatric neurosurgery at CMH in 1979 and became the medical director for the SB Center [38]. Following McLone's retirement in 2004, Dr. Robin Bowman became medical director of the Multidisciplinary Spina Bifida Center at Lurie Children's Hospital. Currently, Dr. Bowman is assisted by a team of 9 surgical and medical physicians with Dr. Vineeta Swaroop leading SB Orthopedics and Dr. Elizabeth Yerkes as director of SB Urology. The Lurie Children's SB Center remains a robust multidisciplinary clinic treating around 800 patients with all forms of spina bifida annually and are proud inaugural members of the national Spina Bifida Clinical Care Network, established in 2020.

The multidisciplinary clinic

Structure of the multidisciplinary clinic

Children born with open SB at our institution are evaluated at the multidisciplinary SB clinic at least every 3 months

during the first year of life, every 6 months until school age, and then annually [39]. All evaluations are conducted by the entire multidisciplinary team, involving specialists in neurosurgery; orthopedic surgery; urology; pediatrics; physical medicine and rehabilitation; psychology; physical, occupational, and speech therapy; nursing and social work [39]. Our management goal for all children with open SB is to ensure that they experience stability or improvement in function throughout childhood [39]. Specialists determine their individual assessment of the patient. Subsequently, the providers and parents/caregivers make important treatment or surgical decisions together as a team. For example, the decision to offer an untethering procedure to a child with an unexpected reduction in function is made by the multidisciplinary team in conjunction with the family and patient [39]. Caregivers, and eventually the maturing patient, are instructed to contact the SB team if they notice any neurological, orthopedic, or urological change between clinic visits [39]. The clinic has managed patients prospectively since its inception and actively maintains a database of all patients [39].

Advances in neurosurgical care

The Lurie Children's multidisciplinary SB clinic advocates for innovative, prospective surgical management of patients afflicted with any form of spina bifida [40]. From its inception, Raimondi and McLone promoted aggressively treating all patients with open SB in a nonselective, prospective manner, in staunch opposition to the views of Lorber [41]. McLone described a technique for closure of the open spina bifida involving the closure of the placode and suspension of the neural tissue in a CSF compartment lined with pia and arachnoid to facilitate a physiologically normal microenvironment [42]. All patients, regardless of level, were treated in a prospective manner, with each complication being addressed in a timely manner by the multidisciplinary team.

In 1981, McLone et al. published an outcome study of the initial 100 patients treated aggressively and nonselectively following birth. They reported a mortality rate of 6% from birth to 4 years, with 15% of patients sustaining a shunt infection [43]. A subsequent publication in 2001 detailed the 25-year outcome for children with open SB treated by the Chicago team. The mortality rate during childhood was 24% with a majority of patients requiring CSF diversion (84%). Approximately half of the cohort developed scoliosis, with 43% requiring a spinal fusion. Social bladder continence was achieved by a majority, and most attended secondary education or college [44].

A pivotal change in open spina bifida care occurred in 2011 with the publication of the randomized control trial of prenatal versus postnatal closure for the open fetal defect (MOMS trial) [45]. In 2017, Lurie Children's began offering in utero fetal SB surgery for mothers/fetuses who met the MOMS inclusion criteria. Our fetal team–consisting of pediatric neurosurgery, fetal surgery, maternal–fetal medicine, and neonatology—counsels the expectant parents regarding the spina bifida diagnosis and long-term impact of the disease as well as the prenatal and postnatal treatment options. Our primary objective is to educate expectant parents that prenatal fetal back closure may improve the child's outcome [37] but does not cure the disease. There are potential complications for the mother and child in fetal surgery that they would not otherwise incur in postnatal repair.

Since 2017, the Chicago Institute for Fetal Health has counseled more than 150 expectant parents, with approximately a third of mothers electing to proceed with prenatal fetal surgical repair. Our initial 25 cases utilized the open uterine approach as detailed in the MOMS trial [45]. However, given the concern of uterine rupture with possible fetal loss in subsequent pregnancies[46], our fetal team has moved toward a three-port, fetoscopic uterine approach to the open fetal defect. Regardless of the timing of back closure—pre- or postnatal—all open spinal cord defects are closed as detailed by McLone [42]. The placode is imbricated with pial-to-pial closure, the dural sac is closed primarily, and the skin is reapproximated in the midsagittal plane. If there is a paucity of dura or skin, a patch may be utilized.

Spina bifida is a disease that is rarely static and requires continual management throughout the patient's lifetime to maintain stability in neurologic, urologic, and orthopedic functioning. Optimal care is provided by a multidisciplinary team who establishes the infant's baseline function and subsequently works jointly with the parents, and eventually with the maturing patient in adolescence, to address each health issue as they arise. Any decline in function is addressed immediately to determine the cause and provide medical/surgical interventions to reestablish or improve the child's baseline function.

The most common cause of the decline and biggest masquerader in spina bifida care is a shunt malfunction. In the 1980s, our team published a case series emphasizing the association between shunt malfunction and acute respiratory arrest, with the Chiari II malformation, or baseline hindbrain abnormalities, presumed to be the inciting factor increasing the risk of shunt dependency [47, 48]. With this knowledge and the long-term outcome data published in 2001, the Lurie Children's neurosurgical team focused on modifiable factors to decrease the mortality associated with open spina bifida [44]. With acquired longitudinal clinical experience as well as recognition of universal challenges in patient populations with longstanding shunted hydrocephalus, we came to adopt the practice of permissive ventriculomegaly. Consequently, we developed a more conservative approach

toward ventriculoperitoneal shunt placement in open SB infants with asymptomatic, stable ventriculomegaly. With changing our management strategy, our shunt placement rate decreased from 86 to 55% [49, 50]. A similar management strategy and decline in shunt dependency were reported by Dr. Dominic Thompson's team at Great Ormond Street, London in 2008 [51]. The shunt placement rate continues to decline with the increasing adoption of prenatal closure and the adoption of endoscopic third ventriculostomy for hydrocephalus management [50].

After shunt malfunction, tethering of the spinal cord at the level of the placode is the second most common neurosurgical cause of decline in children with open SB, occurring in 23% of our cohort, with 29% requiring multiple untetherings [39]. In 1990, investigators at our institution identified tethering as a cause of scoliosis in children with open SB [52], emphasizing the importance of a tethered cord release in those with function at L3 and below to stabilize or improve their scoliosis when the curve is below 45°, decrease spasticity, improve motor function, and resolve back pain [53]. These data were confirmed in a longitudinal study reported in 2008 where a majority of patients experienced improved lower extremity strength, spasticity, gait, and bladder urodynamics postoperatively [39]. Although pain was the least common indicator of tethering, all patients experienced improvement after surgical intervention. The most common associated pathology was dermoid tumor in 6% of this cohort who underwent postnatal back closure. Neurological worsening was a rare adverse event in 4 of the 116 patients (2.5%). Wound dehiscence (7%) and CSF leak (4%) was the most common postoperative complication. If a decline in function is noted by our team, the patient undergoes a complete evaluation, including cystometrogram, renal/bladder ultrasound, and manual muscle test with physical therapy. The patient is then evaluated by orthopedics, urology, and neurosurgery to determine if the changing function is related to tethering.

Most children with open SB have a Chiari II malformation, either in structure, as demonstrated on MRI, and/or function. Previously our team demonstrated the link between swallowing derangement [54], breathing [55], and the Chiari II. It is important to assess shunt function prior to performing posterior fossa decompression to facilitate less extensive procedures and to prevent herniation syndromes [56]. Given the diminished intervention in those with stable, moderate cerebral ventriculomegaly, demonstration of stable hindbrain function is essential prior to discharge of a newborn with open SB. Currently, all newborns undergo a swallow study by speech therapy and a sleep study to establish brainstem function and provide any necessary support. Should a newborn experience decline in hindbrain function, the initial response is CSF diversion either via ventriculoperitoneal shunt placement or endoscopic third ventriculostomy [32].

Transition of care

Additional studies at our Center highlight the changing landscape of spina bifida care over the lifespan. A 25-year prospective, outcome study of a cohort closed postnatally reported greater than 75% survival into adulthood, 86% with CSF diversion, 33% underwent a tethered cord release, and 28% required a spinal fusion [44]. A total of 85% of adults attended or graduated from high school and/or college, while 80% attained social bladder continence [44]. As survival has increased and the management of SB has improved, the primary challenge facing young adults with spina bifida is transitioning medical care to adult practitioners [44, 49].

The principal aim for the transition of care for those living with spina bifida is the establishment of a medical home with adult providers who will partner with the patient/family in the preservation of function while promoting independence and overall health [57]. Importantly, the needs of people with spina bifida change during adulthood. The proportion of individuals requiring tethered cord release or scoliosis intervention decreases, while the risk of mortality related to shunt issues, hindbrain dysfunction, and sleep-disordered breathing increases, necessitating specific education regarding how to recognize worsening symptoms and emergencies [44, 58]. Mobility becomes a greater concern, influenced by the level of the defect [57], and impacting decubitus formation [36]. Continence and renal health remain a continual challenge. Moreover, sexual and reproductive health become increasingly important [57]. Many individuals would like to become sexually active but lack formal knowledge regarding sexuality, fertility, and pregnancy [59]. Male patients with open SB also often have erectile dysfunction, azoospermia, retrograde ejaculation, and anorgasmia [60]. Females with open SB enter puberty and menarche at different time points than typically developing females and must be made aware of the possible effects of pregnancy on aspects of open SB such as hydrocephalus or continence [60].

Many multidisciplinary clinics like ours have developed transitional programs. For example, Children's Hospital of Alabama developed the "Lifetime Care Model" [57, 61]. In this model, Transition Readiness Assessment Questionnaire assess readiness to transition to adult care at age 14, teaching begins at age 19, and increasing responsibility is given until age 21 [57, 61]. At age 21, an individualized transition plan is utilized to transition patients to the adult clinic [57, 61].

At our institution, all patients aged 12 years or older complete the Transition Readiness Assessment Questionnaire to guide the transition process. A recent study from our institution reported that patient-reported transition readiness is associated with health literacy after adjustment for educational level and other factors [62], emphasizing the importance of structuring the transition plan to the level of health literacy of the individual patient. In the early teen

years, all providers initiate medical discussions primarily with the patient to promote independence and education about their disease. Currently, we have an Adult Transition Spina Bifida Clinic in which young adults are seen by our neurosurgeon, urologist, nurse coordinator, and social work. This 2-year transition clinic encourages patients to establish a medical home with the following adult providers: (1) a primary care provider, who will help coordinate all of their future medical needs; (2) a neurosurgeon; (3) a urologist; (4) a physical medicine/rehabilitation specialist. For those with shunted hydrocephalus, baseline imaging is obtained. Given the increased risk of sleep-disordered breathing in patients living with open SB [28], a sleep study with sleep medicine consult is obtained. A comprehensive medical document summarizing their prior medical and surgical history, bracing and assistive device(s) needs, and bowel/bladder management/supplies is provided to the patient to aid in a smooth, medically safe transition to adult care.

Present and future directions

It is beyond the scope of this piece on multidisciplinary care of open SB to dive into our interdisciplinary research findings. It is important to note that with multidisciplinary care, interdisciplinary research with psychology and health services research perspectives has yielded knowledge in the neurocognitive outcomes and challenges in this patient population, as well as in optimizing patient-centered healthcare delivery [41, 47, 63–74].

The multidisciplinary SB clinic at Lurie Children's Hospital remains one of the largest multidisciplinary SB clinics worldwide, with over 3016 patient visits in 2018 [75]. The lessons learned over nearly 50 years have facilitated large-scale collaborations. Recently, Bowman worked collaboratively with neurosurgeons from across the country to develop neurosurgical guidelines for the care of people living with SB [28]. Lurie Children's Hospital has the second largest patient enrollment of the 21 hospitals in the National Spina Bifida Patient Registry, a clinic-based longitudinal registry established by the Centers for Disease Control that seeks to determine the treatments and services that promote the greatest improvements in health outcomes for those living with any form of spina bifida [21, 75]. This registry has yielded insights regarding the multi-systemic effects of open SB and associated treatments, including practice patterns in the management of hydrocephalus, Chiari II malformation, and tethered cord syndrome; factors associated with pressure ulcers; and bowel and bladder management and continence paradigms [48, 76–81]. In the future, the Spina Bifida Multidisciplinary Clinic will remain a leader in clinical care, research, and education, and will continue to seek ways to improve care of those affected by spina bifida.

Conclusion

Open SB is a neural tube defect associated with anomalies of the central nervous, skeletal, gastrointestinal, and genitourinary systems. An increasing proportion of individuals with open SB are living well into adulthood due to advances in perinatal care, management of hydrocephalus and the Chiari II malformation, and bowel and bladder care. Accordingly, children and adults with open SB require multidisciplinary care from neurosurgeons, urologists, orthopedic surgeons, physical medicine and rehabilitation physicians, pediatricians, psychologists, therapists, nurses, social workers, and others. The multidisciplinary SB Center at Ann and Robert H. Lurie Children's Hospital, formerly known as Children's Memorial Hospital, serves as an exemplar of a well-organized, effective, patient-centered approach to the comprehensive care of people living with open SB.

Author contribution NAS conceptualized the idea and wrote the manuscript draft. EBY, VTS, SL, and DGM assisted in conceptualizing and revising the manuscript. RB conceptualized the idea, assisted in revising the manuscript, and provided project supervision. All authors reviewed and approved the submitted manuscript.

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Declarations

Competing interests The authors declare no competing interests.

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