Child's Nervous System (2023) 39:1791–1804
https://doi.org/10.1007/s00381-023-05938-9

ORIGINAL ARTICLE

What has changed in pediatric neurosurgical care in spina bifida? A 30-year UAB/Children's of Alabama observational overview

Jeffrey P. Blount[1,2] · Betsy D. Hopson[1,2] · James M. Johnston[1,2] · Brandon G. Rocque[1,2] · Curtis J. Rozzelle[1,2] · Jerry W. Oakes[1,2]

Received: 28 February 2023 / Accepted: 23 March 2023 / Published online: 26 May 2023

Abstract

Spina bifida (SB) remains the most serious and most common congenital anomaly of the human nervous system that is compatible with life. The open myelomeningocele on the back is perhaps the most obvious initial problem, but the collective impact of dysraphism upon the entirety of the nervous system and innervated organs is an equal or greater longitudinal threat. As such, patients with myelomeningocele (MMC) are best managed in a multi-disciplinary clinic that brings together experienced medical, nursing, and therapy teams that provide high standards of care while studying outcomes and sharing insights and experiences. Since its inception 30 years ago, the spina bifida program at UAB/Children's of Alabama has remained dedicated to providing exemplary multi-disciplinary care for affected children and their families. During this time, there has been great change in the care landscape, but many of the neurosurgical principles and primary issues have remained the same. In utero myelomeningocele closure (IUMC) has revolutionized initial care and has favorable impact on several important co-morbidities of SB including hydrocephalus, the Chiari II malformation, and the functional level of the neurologic deficit. Hydrocephalus however is not solved by IUMC, and hydrocephalus management remains at the center of neurosurgical care in SB. Ventricular shunts were long the cornerstone of treatment for hydrocephalus, but we came to assess and incorporate endoscopic third ventriculostomy with choroid plexus coagulation (ETV-CPC). Educated and nurtured by an experienced senior mentor, we dedicated ourselves to fundamental concepts but persistently evaluated our care outcomes and evolved our protocols and paradigms for improvement. Active conversations amidst networks of treasured colleagues were central to this development and growth. While hydrocephalus support and treatment of tethered spinal cord remained our principal neurosurgical charges, we evolved to embrace a holistic perspective and approach that is reflected and captured in the Lifetime Care Plan. Our team engaged actively in important workshops and guideline initiatives and was central to the development and support of the National Spina Bifida Patient Registry. We started and developed an adult SB clinic to support our patients who aged out of pediatric care. Lessons there taught us the importance of a model of transition that emphasized personal responsibility and awareness of health and the crucial role of dedicated support over time. Support for sleep, bowel health, and personal intimate cares are important contributors to overall health and care. This paper details our growth, learning, and evolution of care provision over the past 30 years.

Keywords Spina bifida · Hydrocephalus · Multi-disciplinary clinic · Myelomeningocele · Neural tube defects

Introduction

There has been a great expansion of knowledge in care for children with spina bifida (SB) over the last 30 years. This paper provides an overview of the experience at Children's of Alabama/UAB during this time and the evolution of institutional practice preferences in pediatric neurosurgery for children born with spina bifida.

The current era of pediatric neurosurgical care for patients with SB in Birmingham began in 1993 when Dr.

✉ Jeffrey P. Blount
Jeffrey.blount@childrensal.org

[1] Division of Pediatric Neurosurgery, Department of Neurosurgery, University of Alabama at Birmingham, Birmingham, USA

[2] Children's of Alabama, Lowder 400, 1600 Seventh Avenue South, Birmingham, AL 35233, USA

W. Jerry Oakes came to UAB from Duke University. Neural tube defects (NTDs) were central to Dr. Oakes' academic interests and expertise. Consequently, new paradigms and protocols were initiated in the Pediatric Spina Bifida Clinic (PSBC) that have been followed, evaluated, and adapted over the intervening three decades. Fundamentally, we have evolved to a lifetime care model that has broadly impacted virtually all dimensions of care. Members of our team organized and participated in impactful conferences, workshops, and initiatives that further changed clinical practice. Our clinical team contributed to several sets of practice guidelines which modified several paradigms. In 2011, we initiated an adult/transitional spina bifida clinic which has subsequently grown to provide ongoing support to more than 200 adults with SB. Insights from those patients and that experience re-shaped our entire approach to practice and gave rise to the Individualized Transition Program and Lifetime Care Plan [1].

Two technical advances also fundamentally changed the practice landscape of SB care in children during this time. First, the report of the management of myelomeningocele study (MOMS) trial in 2011 presented class 1 evidence strongly supporting benefits of in utero myelomeningocele closure (IUMC) [2]. Second, the development and popularization of endoscopic third ventriculostomy with choroid plexus coagulation (ETV-CPC) provided a promising means to treat hydrocephalus that spared much of the morbidity of ventricular shunts [3–5]. The impact, evolution, and current role of these approaches and events are outlined in this overview.

Background and clinic/program organization

Children's of Alabama (COA) is a tertiary pediatric health center in Birmingham, Alabama, USA, that features 15 unique facilities including the flagship 275 bed Benjamin Russell Hospital for Children. It operates in partnership with, but independent from, the University of Alabama at Birmingham (UAB) and as such is the operational center for all UAB Pediatric programs and activities. The entire campus facility occupies more than 2 million square feet making it the third largest pediatric facility in the USA. In 2022, children from Alabama as well as 46 other states and 7 foreign countries received sub-specialty care at COA. The institutional mission is to provide exceptional quality medical care for the children of Alabama regardless of their family's ability to pay. As such, COA serves as the pediatric referral center for the state of Alabama (2022 population 4.8 million persons) which, in 2021, resulted in more than 670,000 outpatient evaluations, 15,000 inpatient admissions, and 3000 surgical operations. Since opening in 2012, more than 5.8 million visits, 240,000 surgeries, 250 organ transplants, and 650,000 emergency room visits have occurred at the Benjamin Russell Children's Hospital. It was ranked by US News and World Report in the top 50 pediatric programs in the USA in 10 specialties and was overall ranked the #1 pediatric medical facility in the Southeastern USA in 2021.

The Pediatric Spina Bifida Clinic (PSBC) at COA has grown over the past decades and is one of the most robust programs in North America. Currently, more than 600 children/families with dysraphism are actively followed and treated. The PSBC at UAB/COA was one of the inaugural members of the National Spina Bifida Patient Registry (NSBPR) and has consistently led enrollment amongst all NSBPR centers. Experience from the PSBC at UAB/COA was also instrumental in the development of the Spina Bifida Association's (SBA) Clinical Care pathway and significantly impacted multiple sets of spina bifida clinical practice guidelines. An adult/transitional program was initiated in 2011 and now supports more than 200 adults with dysraphism. Insights from this experience have impacted multiple areas of care (detailed below).

Patients with both open (myelomeningocele) and closed/occult/variant forms (spinal lipoma, split cord malformations, myelocystocele) are actively followed. Patients with simpler forms such as filum lipomas and dermal sinus tracts typically require less longitudinal care once surgical intervention has occurred [6]. As such, they are followed acutely postoperatively in pediatric neurosurgery clinic only. A limited number of patients (< 10% clinic volume) without dysraphism, due to a medically complex syndrome (e.g., VACTERL, cloacal exstrophy, anorectal malformations (ARM)) with similar longitudinal needs, are also followed in PSBC.

The clinic is multi-disciplinary, encompassing 5 medical disciplines (neurosurgery, urology, orthopedic surgery, rehabilitation medicine, gastrointestinal pediatrics), 3 therapy domains, social work, nurse practitioners, nurse clinicians, and administrative coordinators. The clinic is coordinated by the PSBC coordinator who assures that multiple parts of the multi-disciplinary clinic function as a cohesive whole. This involves scheduling appointments, coordinating needed investigations (radiographic examinations, diagnostic procedures, laboratory tests, and consultations), and facilitating patient flow and inter-service communication. Clinic nursing and administrative staff assure a smooth and safe flow of patients, collect demographic information and clinical metrics, survey new signs/symptoms, and collect vital signs in addition to assuring the clinic remains safe, clean, and hygienic.

At present, intra-uterine myelomeningocele closure (IUMC) is not provided at UAB/COA. Eligible maternal–fetal couplets are identified and referred by members of the Maternal–Fetal Medicine Division of the UAB

Department of Obstetrics and Gynecology to a regional center that offers endoscopic IUMC. The direct but not indirect costs of care are supported by public insurance. Following IUMC patients return to the UAB/COA PSBC for ongoing care is conducted in cooperation with the IUMC that performed closure. The number of annual new cases of open MMC evaluated and closed has diminished since IUMC has become widely available. Historically, we have managed approximately 16.2 new cases of open MMC/year. Since 2020, we have averaged 12.1 new cases of open MMC annually, and all have had public insurance.

Clinical principles—clinic and outpatient care

Logistics

The multi-disciplinary clinic at COA/UAB meets every other week. Typically, about 2 dozen patients are seen and reviewed. Acute, symptomatic problems are referred to the emergency department to expedite assessment and are not evaluated in Pediatric Spina Bifida Clinic (PSBC). Children are seen every 3 months for the first year of life and then every 6 months until 5 years of age at which time they move to annual follow-up. Routine brain imaging (fast MRI) is performed intermittently for patients with ventricular shunts. Routine spine MRIs are obtained when there is clinical concern for tethered cord or syringomyelia. Brain imaging assures a comparison in the event of symptomatic presentation with shunt failure symptoms while occasional spine MRIs allow observation for the development of a syrinx. This practice has evolved toward less frequent imaging over 3 decades. Initially, scans were performed every other year, and intervention was undertaken for isolated ventricular enlargement. The prevailing concept was that proactive imaging directed decision which prevented or reduced urgent presentation of shunt failure. However, we observed an elevated rate of infection and no quantifiable reduction in acute shunt presentation [7]. As such, the rate of imaging was decreased, and patients with isolated enlarged ventricles (without clinical signs) were reviewed in clinic more frequently but not operated upon.

At clinic intake, basic demographic data is captured, reviewed, and updated. Vital signs, weight, and head circumference measurements are taken. Imaging studies are often arranged in the morning for the afternoon clinic. Following check-in, research-based surveys are often distributed/completed if consent/assent has been obtained. During clinic, each clinical team rotates between exam rooms, and patients and families remain in the rooms until they see each team. Care is discussed with the families and with other care team members from different disciplines. A master whiteboard captures progress, notes, and follow-up to facilitate interdisciplinary interaction and ensure proper follow up. A post-clinic review of each patient further facilitates interdisciplinary care and fosters coordinated evaluation and surgical decision-making. Clinic logistics are further detailed elsewhere [8].

Outpatient care—infancy and childhood

Hydrocephalus

Key age-specific neurosurgical metrics are obtained at each clinic to assess the progress and status of each child. Across the lifespan, the adequacy of hydrocephalus management is the predominant concern because of its capacity to adversely impact other neurologic problems [9–19]. Consequently, assessments of head size, evidence for stridor or other brain stem dysfunction, and wound closure predominate the newborn and infant evaluation. Head size assessments (graphs), developmental milestones, and ventricular imaging predominate for young children. School aged children need ongoing assessments for headaches and worsening back pain or lower extremity dysfunction that may suggest tethered spinal cord (TSC). Each of these age-specific components of care is further detailed below, and the lessons from the impact of experience in adult-transitional care have impacted our approach to many issues.

Tethered spinal cord—recognition, diagnosis, and intervention

Tethered spinal cord is a functional diagnosis. All patients who had MMC closure appear tethered on MRI, yet traditionally, only about 40% require subsequent untethering [16, 20, 21]. In our clinic, patients with MMC are screened for symptoms of increasing pain in the low back, hips, and buttocks. Sensory impairment is common in the lower extremities in MMC, so back pain tends to predominate. Clinical signs including motor changes such as reduced walking endurance, tripping, or falling may represent signs of tethering. We emphasize the "3 P's" rubric to families: symptoms that are persistent, progressive, or profound are the most important to distinguish from background musculoskeletal pain that frequently affects patients with SB.

Urologic indices including reduced volitional control, more urinary accidents (for those with retained volitional control), or increasing urinary tract infections are all important markers for tethered cord [22, 23]. As such, we have evolved toward screening clinically and verifying with urodynamic studies. MRI plays a lesser role in making the diagnosis of tether except for the presence of a new or enlarging

syrinx. Otherwise, MRI imaging contributes an important "road map" for surgery but plays a lesser role in the diagnosis of TSC.

We and other centers have observed signs of TSC to be initial manifestations of shunt failure [24–27]. Ventricular enlargement is an insufficiently sensitive metric for detecting shunt problems in patients with SB. As a result, we consider shunt exploration before surgical untethering in most cases of symptomatic TSC. Each case is individually considered (some/many patients have had recent shunt revision), but shunt revision is an emphasized, standardized part of our approach to TSC evaluation and care.

Urologic observations are evaluated in consultation with urology colleagues utilizing renal ultrasonography and urodynamic studies (UDS) [28, 29]. While UDS are subjective and require experience and consistency to interpret, they can play a critical role in the assessment of adequate lower urinary tract function deterioration over time. Bladder cystometry (CMG) demonstrates the reactivity of the bladder when it is filled while sphincteric EMG monitors pelvic flood musculature [22, 28]. Detrusor hyperactivity is the finding most consistently observed with TSC [22]. However, it may also be seen in unaffected children with delayed but otherwise normal micturition [28–31]. Bladder function is screened clinically by the frequency of urinary tract infections or decline in volitional control. The primary objective of all urologic management is the preservation and protection of the upper urinary tract and control of infection [32]. Close multi-disciplinary care between urology and neurosurgery has been and remains emphasized at every clinic. Urinary incontinence and the threats to renal function with progressive and long-standing lower tract disease have long been a prominent concern for patients with SB.

Outpatient care—adolescent/teen/young adult years

Adolescents and young adults require initiation into independence, transition concepts, headache, and back pain assessment as well as self-care and judicious introduction of personal and intimate cares.

Preparation for transition

The critical objective for well care in SB in adolescence is to foster self-care and awareness of medical need. The development and adherence to the individual transition plan (ITP) are the cornerstone of our program [1]. This holistic approach was built from the "Got Transition" program which defines six core elements of transition [32]. At its core, the ITP involves the creation of specific individualized objectives and goals for each patient that arise from the patient, the family, and the care team. Prior findings from our adult program suggested that QOL is highest for those adult patients without bowel incontinence and those who have an external pursuit such as a simple job or volunteer activity that gets them out of the house and interacting with other people [33, 34]. As such, these are central components of the ITP that are emphasized at each visit.

Sexual and personal care

Recent advances in medical treatments and therapies have made it possible for those with SB to experience a wider breadth of life experiences and lead a more fulfilling QOL. Individuals with SB are presented with unique challenges related to social opportunities, dating, and sexual health and function. Increasing awareness of the importance of reproductive/sexual health education has been noted within the SB community. Studies show that women with SB report a poor understanding of their current reproductive health and pregnancy potential as well as an overall lack of sexual education. Likewise, men with SB overwhelmingly report a lack of sexual education. As many as 75% of men with SB have erectile dysfunction which further highlights the importance of providing men with adequate information regarding their sexual health, function, and fertility issues [35]. Not only does there exists a gap in knowledge from the patient's perspective, but physicians also report inadequate training and a lack of confidence in providing reproductive health education to patients with SB [36, 37]. This gap in knowledge and physician support leaves these individuals particularly vulnerable for sexual misconduct, coercion, and abuse.

Clinical principles—inpatient/procedural care

Newborn care in myelomeningocele (all post-natal closure)

Peri-natal care-preferred route of delivery

The preferred route of delivery for a baby harboring a MMC is controversial. However, the preponderance of evidence suggests that there is no discernable adverse impact to the child regardless of means of delivery [38]. As such, our group has no expressed preference. However, most deliveries at our center with known MMC occur via cesarean section. These pre-natal decisions are often made by maternal–fetal medicine physicians with minimal neurosurgical input. Cases where the MMC was not known prenatally (usually in women without pre-natal care) often deliver via vaginal delivery, and we have not noticed any difference in outcome in this group.

Peri-natal care management of the exposed placode

We prefer that the exposed neural placode is covered in damp, non-adherent gauze and kept moist and free from physical or chemical injury. As such, we position newborns in the lateral or prone position and keep fresh (changed every 6 h or if stained) and moist dressings in place until closure is complete. After closure, we continue lateral/prone positioning for 3 days, then utilize a soft foam donut for another week to minimize physical pressure on the recently closed back.

Assess for a lethal lesion

MMC closure is contra-indicated if a lethal lesion (e.g., anephria) is encountered. Traditionally, in our program, this has required a renal ultrasound and cardiac echocardiogram. Practical observation evolved such that a child observed to make urine and sustain normal perfusion color (pink extremities, digits) can be safely assumed to have sufficient cardiac and renal status to plan operative closure of the MMC. Closure is not typically undertaken until the renal US and echo confirm normality, but no child has been refused closure in 30 years based upon these study results.

Layered closure of MMC within 48 h

A layered closure that reconstitutes the normal anatomic layers is the preferred surgical objective [39]. Prep consists of debridement and saline or lactated Ringers irrigation over the placode and iodine-based preparation of the surrounding skin.

Dissection commences at the edges of placode and surgically identifies pial and dural layers. The placode is typically imbricated, and rostral cord is inspected ventrally and dorsally for tethering bands (meningocele manque). The dura is then isolated and closed in watertight fashion. Fascia is closed if possible, and then, the skin is undermined and closed in two layers with careful attention to minimizing tension. Wounds that leak are carefully inspected for technical closure issues, but leaks are typically considered to be due to under treated hydrocephalus. Children with severe hydrocephalus at birth are typically managed with an external ventricular drain or shunt placement to optimize wound closure and decrease risk of CSF leak. In general, techniques for closure have changed little over the past 30 years. There has been recent introduction of the microscope to improve visualization and comfort, but this is not uniformly embraced, and most neurosurgeons at our center use loupe magnification.

Acute hydrocephalus management

Key thresholds for hydrocephalus treatment in our program are as follows: (a) back wound leak following good closure; (b) stridor, poor secretion management, or other signs of brainstem dysfunction; or (c) progressive macrocephaly/accelerated head growth trajectory on head growth measurement curves.

Shunts have traditionally been the cornerstone of hydrocephalus treatment, but there has been a steady increase in number/percent of cases, managed with ETV-CPC [15, 16]. For children undergoing shunt placement, a medium-fixed pressure small/infant valve with flow regulation is preferred. Timing of intervention depends upon size of the head but is generally deferred and performed as a second procedure if indicated. Occasionally, a significantly macrocephalic infant will undergo acute shunt placement or EVD placement at the time of closure. Stridor in a baby with MMC indicates brain stem stress and dysfunction and warrants urgent shunt placement/revision or EVD. Endoscopic third ventriculostomy with choroid plexus coagulation has steadily increased its role in management of hydrocephalus.

Early evaluation/management of the C2M

C2M can present acutely in the newborn period with stridor, poor oral secretion management, or opisthotonus. Acute perinatal C2M crisis is considered a primary neurodevelopmental problem that is acutely precipitated by a decline in control of hydrocephalus [40–42]. As such symptomatic C2M crisis is best managed by urgent, aggressive control of hydrocephalus [15]. Surgical decompression is nearly never done because of historic inefficacy and risk of significant complications. Children who do not respond to acute hydrocephalus intervention and continue to demonstrate brain stem symptoms have an ominous prognosis. Only in these rare and desperate cases have C2MD been undertaken at our center. When (rarely) performed, C2MD centers on decompression of the descended brainstem via cervical laminectomy and emphasizes meticulous attention to the descended location of the caudally displaced torcula. Failure to be aware that the torcula is typically caudally displaced to the cranio-cervical junction can lead to inadvertent intrusion into the torcula at the time of dural opening with catastrophic results.

Lifetime care plan

During the initial hospitalization, the clinic coordinator visits with the family on several occasions to detail the Lifetime Care Plan and logistics of how support is provided in the PSBC. Care principles are reviewed by clinic coordinator, neurosurgery nurses, and some attendings. A notebook of

information and checklists is provided. Enrollment in the NSBPR is offered and initiated. Availability of the treating team is emphasized.

Spina bifida clinic follow-up (newborns)

Well, infants are seen 3 weeks after discharge from hospital and then routinely at 3 month intervals until 1 year of age. Cranial imaging is performed via fsMRI before the 1st 3 month visit. Head size is measured/plotted with each visit. Assurance is made that urologic care is established prior to discharge to home. Nursing contact information is assured with each family. Every effort is made to avoid ionizing radiation across the lifespan.

Hydrocephalus management/shunt failure across life span

Five principles have evolved that underlie our approach to the management of hydrocephalus in SB. These are based upon observations about (a) the central and crucial role that hydrocephalus has in the overall well-being of patients and (b) the variance in presentation and acuity of decline/risk in patients whose hydrocephalus arises from SB.

1. A high index of suspicion for decompensated hydrocephalus as contributor for other pathologies
    - Hydrocephalus stresses the entire central nervous system. When there is an inherent "weak link" (like brain stem function in C2M), hydrocephalus can precipitate neurologic decline/collapse.
    - As such, early symptoms of brain stem compromise (stridor, poor secretion control) are first addressed as signs of hydrocephalus.
    - Similar concepts support the presentation of symptomatic tethered cord later in life. Shunt exploration often precedes TSCR in our program particularly if any signs or symptoms of shunt failure are present.
2. Shunt failure presentation varies in SB/explore the shunt early
    - Ventricular enlargement is a variable finding in shunt failure in SB.
    - Headache patterns that include neck pain or craniocervical junction pain are highly associated with shunt failure in SB.
    - Symptoms of low back pain (TSC) may be and often are related to shunt insufficiency/failure and warrant approach.
    - Patients with hydrocephalus from SB may decline quickly and severely with brain stem/respiratory embarrassment/collapse. As a result, a low threshold for operative exploration of shunts in SB has evolved in our program.
3. Evolution of ETV-CPC as a meaningful alternative to care
    - Hydrocephalus arising from SB has consistently demonstrated responsiveness to successful management via ETV-CPC [3, 4].
    - Successful ETV-CPC requires coagulation of at least 90% of the choroid plexus. For this reason, there is a strong preference for use of flexible endoscopy for ETV-CPC.
    - Successful ETV-CPC does not require ventricular size reduction. Rather, a growth curve that parallels the normal growth curve with evidence of good wound healing and normal neurologic development are the current preferred metrics of a successful ETV-CPC. Ongoing prospective studies of neurodevelopmental outcomes in children after shunt and ETV-CPC will more definitively answer this important question.
4. Nursing support/availability of care is crucial
    - Immediate availability of support for hydrocephalus evaluation and treatment is the cornerstone of ongoing neurosurgical support for patients with SB.
    - Our program incorporates skilled, dedicated nurse clinicians who maintain availability to patients via multiple modes of communication.
    - After hours, weekend and holiday coverage is provided by house officers and on-call attendings.
    - The continuous availability of support for questions and concerns about shunt/hydrocephalus treatment is emphasized throughout all communications with patients across the lifespan.
5. Annual visit
    - The annual visit is emphasized in our clinic. While short and focused, the annual visit allows ongoing parent education and supports the concept that care needs may arise promptly and unpredictably and that surgical support remains available.
    - Symptoms and signs of shunt failure and TSC are reviewed, and communication with the SB team is emphasized.
    - New concepts and opportunities for social connection via support organizations are shared and encouraged.
    - Updated patient is tracked, recorded, and logged in central clinical repositories (e.g., the NSBPR)

Milestone events in the UAB/COA evolution of multi-disciplinary care

Several conceptual milestones have profoundly impacted the growth, approach, and direction of our spina bifida program.

Conferences/meetings/ task forces

Conferences, workshops, and task forces have played an important role in directing practice standards and preferences.

Evidence-based practice in spina bifida conference ("Green Book Conference")

In 2003, the Spina Bifida Association of America (SBA), the Agency for Healthcare Quality and Research (AHRQ), and the National Center for Birth Defects/Office of Rare Diseases of the Centers for Disease Control (CDC) organized/sponsored an invitational meeting In Washington D.C that sought to summarize standards of care and establish a prioritized agenda for research and care in spina bifida. Participants included members of the SBA, the AHRQ, and more than 100 invited multi-disciplinary providers from many working clinics across North America. The Professional Advisory Council of the SBA (SBA-PAC) had previously met to establish a prioritized list of active clinical issues in spina bifida care. Working groups from each multidisciplinary SB clinical discipline were formed to discuss key gaps in knowledge in the clinical provision of service for patients with SB. Following conversations and debate this list of gaps evolved into a prioritized agenda for SB clinical research [43].

A key presentation at the meeting was from the AHRQ which demonstrated the critical contribution that a registry can make. Examples were provided from the Cystic Fibrosis Society that maintained detailed registries of clinics and patients. By identifying outcome objectives and prioritizing and sharing key metrics, clinics could compare outcomes between clinics. Protocols and paradigms of the highest performing clinics could then be imitated/followed to result in broad ranged improvements across clinics. Attendees found this approach compelling and sought to initiate both patient and clinic registries. These conversations created much of the impetus for the National Spina Bifida Patient Registry (NSBPR). UAB members were active in these discussions and in the organization and subsequent implementation of the NSBPR. The enthusiasm and sense of purpose that characterized this meeting set the stage for the World Congress meetings that were initiated in 2009.

The National Spina Bifida Patient Registry (NSBPR)

The National Spina Bifida Patient Registry (NSBPR) is sponsored by the National Center on Birth Defects and Developmental Disabilities of the Centers for Disease Control and Prevention (CDC) and enrolls patients with SB in the USA from participating clinics. The data collection is done longitudinally in conjunction with annual visits to multi-disciplinary SB clinics. The data are de-identified and stored/managed in a repository at the CDC. When a clinical topic of interest is identified, a data access proposal (DAP) is forwarded to the CDC. Within the NSBPR, a smaller committee, Committee for Science and Publication (CSP) comprised of various NSBPR Principal Investigators (PIs), reviews the DAP, and plans for statistical analysis. After approval is granted from the CSP, CDC sends aggregate data to the site requesting the data. All NSBPR PIs are given the opportunity to participate in the approved projects. CDC contributes organizational infrastructure and statistical evaluation and maintains regulatory oversight of projects arising from the NSBPR. Since inception, the NSBPR has been responsible for more than 40 multi-institutional studies on SB. From its initial inception at the Research Agenda Conference UAB, team members were contributory and central to the design and implementation of the NSBPR. UAB/COA has been the leading center for patient enrollment and has directed more than 7 DAP-based NSBPR investigations. UAB DAP-related NSBPR project topics have included shunt survival and hydrocephalus management, Chiari II malformation, sleep-disordered breathing, loss of ambulation, and a couple on urologic management. NSBPR participation has fundamentally impacted care for patients with SB at UAB. Arguably, the single greatest impact of the NSBPR has been the network that has been developed and the energizing and brainstorming of new projects and ideas that has come from the frequent meetings and sub-specialty working groups. Insights gained and questions raised have shaped investigations and clinical programs that have improved care [43–46].

UAB/Children's of Alabama multi-disciplinary invitational conference on spina bifida care

Despite the development of a registry, there were initially no clinical guidelines to standardize care for patients with SB. Clinical guidelines had emerged in many other areas of neurosurgery, and there was a growing awareness of the contribution of guidelines to improve care. In October 2011, we convened a workshop conference on SB guidelines at UAB/Children's of Alabama and invited 38 leaders in the pediatric

neurosurgical care of SB to attend. Prior to arrival, papers on key neurosurgical topics were shared and reviewed. At the meeting, an organized approach was taken to reviewing best evidence for best current practice. These formed a basis for forthcoming UAB and other contributions to guidelines.

Guidelines of clinical care for patients with spina bifida

Guidelines for care for patients with SB are critically important as they can standardize care according to best evidence-based medicine. Consequently, there are now more than 40 articles that provide guidelines on specific aspects of care in spina bifida. These are built on the foundational efforts in SB guidelines that were simultaneously pursued by the SBA and the Congress of Neurologic Surgeons Joint Guidelines Committee [47]. These initial (2015–2018) efforts surveyed and prioritized all published world literature and embraced the central role of stratification by the strength of medical evidence. However, important differences in priorities and emphases occurred.

- SBA guidelines: The SBA sought a practical work product that would provide a useful resource in SB clinics. Practical approaches to common and difficult problems were emphasized, and some guidelines were approved based upon agreement of expert assembled faculty. SBA guidelines addressed each step of care sequentially and temporally across the lifespan [48]. These guidelines have been adapted and served as the basis for subsequent refinements that define most current widely used guidelines.
- Congress of Neurologic Surgeons (CNS) guidelines: The objectives of MMC CNS guidelines were to create recommendations for best practice based upon evidence-based medicine principles, to obtain multi-disciplinary endorsement, and to share the educational content to optimize care [47]. The salient SB literature was reviewed with assistance of medical librarians. The CNS adhered strictly to EBM dictates and consequently produced a more narrowly focused guideline product. Five PICO questions were generated, reviewed, and answered with best available evidence:

    1. Is there a difference in the proportion of patients who develop shunt-dependent hydrocephalus between fetuses who underwent prenatal MM closure compared to infants who underwent postnatal MM repair?

        Recommendation: IUMC reduces rates of need of treatment for hydrocephalus when maternal/fetal MOM inclusion criteria are met (level I) [49].

    2. In patients with MM, does prenatal or postnatal closure improve the ability to ambulate?

        Recommendation: IUMC resulted in better ambulatory capability in the short term (30 month) but long-term impact unknown (level III) [50].

    3. In patients born with a MM, does closure of the defect within 48 h reduce the rate of infection?

        Recommendation: There is insufficient evidence to conclude that closure within 48 h of birth reduces infection (level III) [51].

    4. In MM patients with HC, does persistent enlargement of the ventricles adversely impact neurocognitive development?

        Recommendation: There is currently insufficient data to conclude that ventricle size correlates with neurocognitive development [52].

    5. Is there a difference in the rate of development of tethered cord syndrome in infants who had prenatal MM closure compared to infants who had MM closure after birth?

        Recommendation: There is evidence that inclusion cysts and other means of tether exist at higher rate for infants undergoing IUMC. Close observation for signs of TSC are recommended (level III) [53].

UAB team members participated actively in the development and implementation of both initial guideline initiatives. Each set of guidelines impacted our practice by emphasizing the evidence quality for clinical decisions being made in the PSBC, but impact varied. The UAB Maternal–Fetal Medicine team recognized candidates for IUMC and made regional referrals. Patients returning from IUMC are followed in clinic for signs and symptoms of TSC, but protocols for care do not differ. We have maintained a preference to close MMC promptly, and nearly all are closed within 48 h. The lack of evidence to support impaired neurocognition with larger ventricles supported broader use of ETV-CPC which typically results in less ventricular reduction in size than shunt placement. While most evidence for current interventions are class III or IV, the opportunity to develop guidelines enhanced this awareness, raised opportunities for new inquiry, and emphasized the importance of an inquiring, humble clinical approach.

World congresses of spina bifida

The first World Congress on Spina Bifida Research and Care was the natural extension of the May 2003 Evidence Based Practice (EBP) meeting and occurred in March, 2009

in Orlando, Florida. Following the EBP meeting, the SBA PAC (chaired by Dr. Jeffrey Blount from UAB) and CDC formed the Program Planning Committee (PPC) for the World Congress meeting. Under their direction, the first World Congress meeting was organized. Dr. Blount and Dr. Oakes served on the PPC for neurosurgery. The first World Congress occurred in March 2009 and featured more than 350 participants (including researchers, clinical providers, patients, and families) from over 30 countries who participated in lectures, breakout sessions, workshops, and plenary sessions. Key themes included transitional care, epidemiology, prevention, transition, nursing/allied health, and sexual health in SB. The cross communication between these groups allowed a unique opportunity for communication, inter-disciplinary sharing, and prioritization of issues by the patients and families.

There have been 3 World Congress meetings since 2009. Each has served specific and broad objectives. The Third World Congress (San Diego, March 2017) had a preliminary meeting to help foster guideline development. We are awaiting the 4th World Congress meeting as this paper is being prepared.

While guidelines brought the best available evidence to guide clinical practice, the World Congress meetings "fence-posted" our assessments of new approaches/interventions and represented an optimized opportunity to bring new ideas. Dr. Brandon Rocque directed/coordinated multiple sessions and currently serves on the SBA PAC. Betsy Hopson has been central in coordination and organization of World Congress events since its inception. Abstract and study workshop plans started long in advance and remained as prioritized initiatives until completed. Our approach evolved such that the first event upon return to UAB from a WC was an organizational meeting to appropriately incorporate important observations presented at WC into our active practice.

Evolution of 2 technical advances

Two technical advances over the last 30 years have revolutionized neurosurgical care for patients with SB.

Endoscopic third ventriculostomy with choroid plexus coagulation (ETV-CPC)

Endoscopic third ventriculostomy with choroid plexus coagulation (ETV-CPC) presents an important alternate hydrocephalus treatment to ventricular shunts. Shunts are effective but are often plagued with significant complications and morbidity. Patients with hydrocephalus from SB have been demonstrated to show more favorable response and better outcomes than patients with other etiologies of hydrocephalus in virtually all large clinical ETV-CPC series. A consistent success rate of 50–60% has been reported by large individual centers as well as the Hydrocephalus Research Network (HCRN) [54, 55]. The HCRN is a network of clinical hydrocephalus research centers that agree to standardize protocols, metrics, and analysis upon shared data. As such, data from the HCRN and the NSBPR represent the largest accumulated experience in treatment for hydrocephalus. These outcomes remain slightly lower than those attained by Warf and colleagues who developed and perfected techniques for ETV-CPC [3, 4]. UAB pediatric neurosurgeons embraced ETV-CPC early and trained with Dr. Warf to optimize our technical capabilities. ETV-CPC has remained an active treatment in our approach to hydrocephalus in SB, and all patients with hydrocephalus are evaluated and considered for ETV-CPC as an initial form of treatment.

An important development in the practical use of ETV-CPC was the development of a set of predictors for success of ETV-CPC [56]. This scale is used actively at UAB to determine candidacy and contribute to the informed consent process.

Some controversy remains regarding the impact of persistently larger ventricles observed with ETV-CPC. Large ventricles with macrocephaly are accepted clinical outcomes for success in ETV-CPC provided that any pathologically rapid trajectory of growth is mitigated, and the head growth curve parallels the normal curve. The best available current neuro-cognitive outcome assessments based upon Bayley III scores suggest similar outcomes with ETV-CPC and ventricular shunts [56–64].

Publication of MOM trial/growth of IUMC programs

No event broadly impacted care for patients with SB in the last 30 years more than the publication of the MOMS trial in 2012 [2]. This widely heralded, randomized, prospective trial demonstrated superior outcomes in several important domains for fetuses treated with intra-uterine MMC closure (IUMC) to post-natal closure cohorts. These included a lower need for a shunt, reduced hindbrain herniation on imaging, and improvements in sensorimotor capabilities. Since then, there has been an explosion of activity surrounding IUMC and a rapid expansion in the number of centers offering the procedure. Two alternate techniques of IUMC have evolved in this fast moving, highly competitive environment. Open IUMC was the technique for MOMS and involved exposure of the gravid uterus, hysterotomy, and open closure of the fetal MMC under direct observation. Endoscopic closure involved similar objectives, but all intrauterine portions were accomplished endoscopically. Each technique appears to have advantages and disadvantages, and the rapid expansion of centers offering IUMC can be broadly

divided into endoscopic and open procedure-based centers. Class I data has only been presented for open techniques, but outcomes appear broadly similar. Follow-up studies of patients treated in early cohorts is generally favorable with most advantages persisting over time [28, 65–70]. An important exception is that patients treated with IUMC (especially endoscopic IUMC) may have an increased incidence of acute tethered cord that adversely impacts urologic and renal health. The prospective UMPIRE trial of the SBA NSBPR is closely studying this concerning finding, and results are anticipated soon [21, 28, 71, 72].

The UAB spina bifida program does not currently offer IUMC services. Maternal–fetal medicine physicians support a full range of complex OB services but have opted to prefer to invest in other clinical initiatives. As a result, potential candidates at our center are screened by UAB MFM providers, and suitable maternal–fetal pairs are referred to a regional (endoscopic) center of excellence. Following IUMC, patients return to and deliver at UAB and receive subsequent care through the UAB/COA PSBC.

We have observed a decrease in the number of open postnatal closures since IUMC has become available. Further, the socio-economic profile has evolved to nearly exclusive public insurance/Medicaid patients. While there is coverage for medical services for regional IUMC, important gaps such as lost time from jobs, travel, accommodation, and food costs abroad are not offset in current models. These represent important public health and policy opportunities for improvement [73].

Impact of adult spina bifida clinic

No single event impacted overall thinking and understanding of spina bifida more than the development of our adult SB clinic (ASBC). The ASBC at UAB started in 2011 when we grew increasingly dissatisfied with the quality and consistency of care available for patients transitioning from pediatric care. After initiation of the ASBC, observations were made that significantly impacted the way care is delivered in both our pediatric and adult SB programs. Ultimately, we embraced a lifetime model of care model which represented a fundamental change in our approach to patients. Only when care is approached and planned across the broad expanse of a person's life can fundamental and critical objectives such as autonomy, self-actualization, self-esteem, and fulfillment begin to be accomplished. Each component of care impacts the overall individual and, as such, is essential if outcomes are to be optimized. These approaches are perhaps less immediate surgically but so substantially impact outcomes, satisfaction, and overall quality of life as to deserve central attention in all measures of care. Social determinants of care are central to a lifetime model and must be incorporated as rigorously as any medical or surgical components [74, 75].

The following clinical observations from the UAB ASBC led to peer-reviewed studies which impacted approaches and practice preferences and protocols.

1. Shunt needs decrease but do not resolve in adulthood: we reviewed our experience in UAB/COA PSBC and UAB ASBC with regard to the number of shunt revisions in each year of life. For 417 patients within the cohort, the rate of shunt revision per patient-year diminished except for an increase in early teen years but continued as late as the 4th decade. From this data, we have been encouraging with our patients about the natural history of shunts failure and have used these data in counseling and consent [25].

We extended these investigations into the adult community when we anecdotally noted that an increasingly large percent of patients in the ASBC appeared to have their original shunt or have had only one or two shunt revisions. This led us to study shunt clusters and shunt survival [76]. We studied a 10-year (2008–2017) prospective sample of 465 patients who underwent shunt placement and had all shunt-related care at COA to identify shunt failure clusters and failure characteristics. This review demonstrated that 6% of the overall cohort accounted for more than 30% of all shunt revision surgeries. This suggested that a small percent of patients have highly complex courses and that a larger group of patients had less complicated shunt histories. These observations were extended with a follow-up study that further detailed the time to shunt failure.

To obtain best available data, we turned to the NSBPR and searched for patients enrolled in the first 5 years of their life who had all shunt procedures recorded in the registry [77]. Inclusion criteria were satisfied by 1691 patients, and the median follow-up time was 5 years. Overall, 55% had at least one shunt failure, 26% had two or more, and 14% had 3 or more shunt failures. More importantly, 45% of patients in this cohort never had a shunt failure. This review provided important data to support a more complete understanding of the natural history of shunts in SB. We have incorporated these findings into consent for treatment options for hydrocephalus.

2. Sleep is a critically important to understand and optimize in SB: two phenomena predominantly informed and motivated this inquiry and study. First, one of the most serious issues in adult care was sudden death during sleep [78–80]. This rare but catastrophic condition is estimated to occur with a frequency of 1/10,000 cases of adult SB. Understandably, it tops the list of concerns of many adult patients. The etiology of the respiratory embarrassment in sleep is unknown but hypothesized to arise from brain stem dysfunction, perhaps precipi-

tated by suboptimal shunt function. Second, many adults with SB also carry a diagnosis of sleep apnea. From these observations, we sought initially to look at the prevalence of sleep disordered breathing (SDB) in our patients who had undergone sleep study [81]. We retrospectively reviewed 52 patients from our PSBC who had a sleep study and observed an 81% incidence of sleep disordered breathing. Sleep apnea was mild in 12%, moderate in 19%, and severe in 50%. Amongst those with a sleep study before and after neurosurgical intervention, we sought to determine whether neurosurgical intervention impacted the degree of obstructive sleep apnea. While a trend was observed, we had insufficient sample size to attain significance. We recognized that the obvious limitation of this study was that only people with symptoms underwent a sleep study. After reporting this, we changed our practices to do baseline sleep studies on all patients based upon the high prevalence of SDB that was observed.

This change in our practice paradigm allowed us to perform a more vigorous analysis. All patients seen in PSBC between 2016 and 2020 were evaluated via polysomnography [82]. Cross sectional analysis of this cohort demonstrated that 49/117 (42%) patients had an apnea–hypopnea index of 2.5 or greater (significant SDB). Other centers are now beginning to look at this association and finding similar elevated prevalence rates of asymptomatic SDB in SB.

3. Quality of life in SB varies widely between patients and across time. These metrics can direct an effective model for care provision: we observed substantial variance in overall happiness, depression, and subjective quality of life in routine interactions in both the PSBC and ASBC. To further investigate this, we prospectively studied patients in our PSBC with surveys that addressed health related quality of life (HRQOL) with a validated scale. In a cohort of 159 patients, we found that patients with myelomeningocele had lower QOL scores than patients with occult dysraphism and that younger patients had higher HRQOL scores than older patients. While demographics did not correlate with outcome, the presence of a shunt or need for C2MD predicted lower HRQOL significantly.

We sought to further understand these observations in the ASBC because of similar thoughts about variance in QOL. Despite modest shunt morbidity in adulthood, adults demonstrated seemingly greater morbidity, depression, and less overall satisfaction as they aged. We surveyed 188 sequential patients in ASBC with a standardized NSBPR survey and utilized univariate and multivariate logistic regression to identify variables associated with disability. Overall, 56% of patients characterized themselves as permanently disabled. Education attainment, a pursuit (hobby, job, or volunteer activity) outside the home, and presence of a functional bowel management program were the strongest predictors of disability. The findings precipitated a fundamental change in our approach in the PSBC. Education and a functioning bowel program are both modifiable factors, so they became increasingly prioritized. These observations were combined with core elements of the American Academy of Pediatrics "Got Transition" program to give rise to the concept of an Individualized Transition Plan (ITP). Development and pursuit of the ITP became the focal point of our transition program and were an integral component in the lifetime care model that is central to the UAB program. This model has now been published and presented and is increasingly widely being utilized to direct lifetime care in many different SB clinics [1, 83].

Conclusions

The UAB/COA team has been privileged to have a robust, evolving SB program for nearly three decades. Core principles of care were imparted by an experienced mentor that guided and shaped our initial approach. From these, we developed a multi-disciplinary holistic plan that emphasizes interdisciplinary care across the life span and incorporates many social determinants that impact care. Key principles are unchanged, like the centrality of hydrocephalus in the neurosurgical support of the patient with SB. Yet, much has changed. IUMC has indelibly changed the landscape, and hydrocephalus treatment now includes both shunts and ETV-CPC. Sleep, transitional care, awareness of intimacy needs, and registries are all new and important domains for growth and future research. Continued improvements in care will arise from refinements of existing surgical techniques, more comprehensive understanding of disease mechanisms, specification and optimization of specific inclusion criteria for adjunct studies, and more widespread incorporation of lifelong models of care that recognize and embrace medical and social determinants of disease.

Author contribution Concept/idea: Blount and Hopson; draft original article: Blount; revise drafts: all authors; oversight/support: Oakes.

Availability of data and materials Not applicable.

Declarations

Ethical approval Not applicable.

Conflict of interest The authors declare no competing interests.

References

1. Hopson B, Alford EN, Zimmerman K, Blount JP, Rocque BG (2019) Development ofan evidence-based individualized transition plan for spina bifida. Neurosurg Focus 47(4):E17
2. Adzick NS, Thom EA, Spong CY, Brock JW 3rd, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Dabrowiak ME, Sutton LN, Gupta N, Tulipan NB, D'Alton ME, Farmer DL, Investigators MOMS (2011) A randomized trial of prenatal versus postnatal repair of myelomeningocele. N Engl J Med 364:993–1004
3. Warf BC, Campbell JW (2008) Combined endoscopic third ventriculostomy and choroid plexus cauterization as primary treatment of hydrocephalus for infants with myelomeningocele: long-term results of a prospective intent-to-treat study in 115 East African infants. J Neurosurg Pediatr 2:310–316
4. Warf BC (2005) Comparison of endoscopic third ventriculostomy alone and combined with choroid plexus cauterization in infants younger than 1 year of age: a prospective study in 550 African children. J Neurosurg 103(6 Suppl):475–481
5. Kulkarni AV, Sgouros S, Leitner Y, Constantini S (2018) International Infant Hydrocephalus Study Investigators. International infant hydrocephalus study (IIHS): 5-year health outcome results of a prospective, multicenter comparison of endoscopic third ventriculostomy (ETV) and shunt for infant hydrocephalus. Childs Nerv Syst 34:2391–2397
6. Blount JP, Elton S (2001) Spinal lipomas. Neurosurg Focus 10:e3
7. Chern JJ, Muhleman M, Tubbs RS, Miller JH, Johnston JM, Wellons JC 3rd, Blount JP, Oakes WJ, Rozzelle CJ (2012) Clinical evaluation and surveillance imaging in children with spina bifida aperta and shunt-treated hydrocephalus. J Neurosurg Pediatr 9:621–626
8. Hopson B, Rocque BG, Joseph DB, Powell D, McLain ABJ, Davis RD, Wilson TS, Conklin MJ, Blount JP (2018) The development of a lifetime care model in comprehensive spina bifida care. J Pediatr Rehabil Med 11:323–334
9. Hopson B, Shamblin I, Zimmerman K, Rocque B, Salehani A, Blount JP (2021) Scope of care in the first four years of life for individuals born with myelomeningocele: a single institution experience1. J Pediatr Rehabil Med 14:667–673
10. Norkett W, McLone DG, Bowman R (2016) Current management strategies of hydrocephalus in the child with open spina bifida. Top Spinal Cord Inj Rehabil Fall 22:241–246
11. Ware AL, Kulesz PA, Orkisz JS, Arrington CN, Bowman RM, Fletcher JM (2020) Long-term intellectual and fine motor outcomes in spina bifida are related to myelomeningocele repair and shunt intervention history. J Int Neuropsychol Soc 26:364–371
12. McCarthy DJ, Sheinberg DL, Luther E, McCrea HJ (2019) Myelomeningocele-associated hydrocephalus: nationwide analysis and systematic review. Neurosurg Focus 47:E5
13. Wetzel JS, Heaner DP, Gabel BC, Tubbs RS, Chern JJ (2018) Clinical evaluation and surveillance imaging of children with myelomeningocele and shunted hydrocephalus: a follow-up study. J Neurosurg Pediatr 23:153–158
14. Kestle JR, Kulkarni AV, Warf BC (2016) Introduction: Pediatric hydrocephalus: a continuing evolution in our understanding and management. Neurosurg Focus 41:E1
15. Blount JP, Maleknia P, Hopson BD, Rocque BG, Oakes WJ (2021) Hydrocephalus in spina bifida. Neurol India 69(Supplement):S367–S371
16. Rocque BG, Hopson BD, Blount JP (2021) Caring for the child with spina bifida. Pediatr Clin North Am 68:915–927
17. Kim I, Hopson B, Aban I, Rizk EB, Dias MS, Bowman R, Ackerman LL, Partington MD, Castillo H, Castillo J, Peterson PR, Blount JP, Rocque BG (2018) Treated hydrocephalus in individuals with myelomeningocele in the National Spina Bifida Patient Registry. J Neurosurg Pediatr 22:646–651
18. Tuli S, Drake J, Lamberti-Pasculli M (2003) Long-term outcome of hydrocephalus management in myelomeningoceles. Childs Nerv Syst 19:286–291
19. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA (2001) Spina bifida outcome: a 25-year prospective. Pediatr Neurosurg 34:114–120
20. Dias MS, Wang M, Rizk EB, Bowman R, Partington MD, Blount JP, Rocque BG, Hopson B, Ettinger D, Lee A, Walker WO (2021) National Spina Bifida Patient Registry group- tethered spinal cord among individuals with myelomeningocele: an analysis of the National Spina Bifida Patient Registry. J Neurosurg Pediatr 7:1–7
21. Shlobin NA, Yerkes EB, Swaroop VT, Lam S, McLone DG, Bowman RM (2022) Multidisciplinary spina bifida clinic: the Chicago experience. Childs Nerv Syst 38:1675–1681
22. Tuite GF, Thompson DNP, Austin PF, Bauer SB (2017) Evaluation and management of tethered spinal cord syndrome in occult spinal dysraphism: recommendations from the International Children's Continence Society. Neurourol Urodyn 1:1–4
23. Joseph DB, Baum MA, Tanaka ST, Frimberger DC, Misseri R, Khavari R, Baillie S, Yerkes EB, Wood H (2020) Urologic guidelines for the care and management of people with spina bifida. J Pediatr Rehabil Med 13:479–489
24. Liptak GS, Masiulis BS, McDonald JV (1985) Ventricular shunt survival in children with neural tube defects. Acta Neurochir (Wien) 74:113–117
25. Dupepe EB, Hopson B, Johnston JM, Rozzelle CJ, Jerry Oakes W, Blount JP, Rocque BG (2016) Rate of shunt revision as a function of age in patients with shunted hydrocephalus due to myelomeningocele. Neurosurg Focus 41:E6
26. Alford EN, Hopson BD, Safyanov F, Arynchyna A, Bollo RJ, Hankinson TC, Rocque BG, Blount JP (2019) Care management and contemporary challenges in spina bifida: a practice preference survey of the American Society of Pediatric Neurosurgeons. J Neurosurg Pediatr 30:1–10
27. Shannon R, Meyer T, Saldano DD, Beilke L, Snow-Lisy DC, Patel SJ, Rosoklija I, Johnson EK, Yerkes EB (2022) Prospective evaluation of a pediatric urodynamics protocol before and after limiting urine cultures. J Pediatr Urol 16(S1):477–513
28. Yerkes EB, Cheng EY, Wiener JS, Austin JC, Tu DD, Joseph DB, Routh JC, Tanaka ST (2021) Translating pediatric urodynamics from clinic into collaborative research: lessons and recommendations from the UMPIRE study group. J Pediatr Urol 17:716–725
29. Snow-Lisy DC, Yerkes EB, Cheng EY (2015) Update on urological management of spina bifida from prenatal diagnosis to adulthood. J Urol 4(2):288–96
30. Atchley TJ, Dangle PP, Hopson BD, Graham A, Arynchyna AA, Rocque BG, Joseph DB, Wilson TS (2018) Age and factors associated with self-clean intermittent catheterization in patients with spina bifida. J Pediatr Rehabil Med 11:283–291
31. Zimmerman K, Williams M, Arynchyna A, Rocque BG, Blount JP, Graham A, Hopson B (2019) Program evaluation of Camp V.I.P: promoting self-confidence and independence for patients with spina bifida. J Pediatr Nurs 47:30–35
32. www.gottransition.org
33. Davis MC, Hopson BD, Blount JP, Carroll R, Wilson TS, Powell DK, Jackson McLain AB, Rocque BG (2017) Predictors of permanent disability among adults with spinal dysraphism. J Neurosurg Spine 27(2):169–177
34. Rocque BG, Bishop ER, Scogin MA, Hopson BD, Arynchyna AA, Boddiford CJ, Shannon CN, Blount JP (2015) Assessing health-related quality of life in children with spina bifida. J Neurosurg Pediatr 15:144–149

35. Deng N, Thirumavalavan N, Beilan JA, Tatem AJ, Hockenberry MS, Pastuszak AW, Lipshultz LI (2018) Sexual dysfunction and infertility in the male spina bifida patient. Transl Androl Urol 7:941–949
36. Spinoit AF, Waterschoot M, Sinatti C, Abbas T, Callens N, Cools M, Hamid R, Hanna MK, Joshi P, Misseri R, Salle JLP, Roth J, Tack LJW, De Win G (2021) Fertility and sexuality issues in congenital lifelong urology patients: male aspects. World J Urol 39:1013–1019
37. De Win G, Dautricourt S, Deans R, Hamid R, Hanna MK, Khavari R, Misseri R, Mueller MG, Roth J, Spinoit AF (2021) Fertility and sexuality issues in congenital lifelong urology patients: female aspects. World J Urol 39:1021–1027
38. Thompson DNP (2009) Postnatal management and outcomes for neural tube defects including spina bifida and encephaloceles Prenat Diag 29:412–419
39. Blount JP, George TM, Koueik J, Iskandar BJ (2019) Concepts in the neurosurgical are of patients with spinal neural tube defects: an embryologic approach. Birth Defects Res 1564–1576
40. Kim I, Hopson B, Aban I, Rizk EB, Dias MS, Bowman R, Ackerman LL, Partington MD, Castillo H, Castillo J (2018) Decompression for Chiari malformation type II in individuals with myelomeningocele in the National Spina Bifida Patient Registry. J Neurosurg Pediatr 22:652–658
41. Pindrik J, Riva-Cambrin J, Kulkarni AV, Alvey JS, Reeder RW, Pollack IF, Wellons JC, Jackson EM, Rozzelle CJ, Whitehead WE, Limbrick DD, Naftel RP, Shannon C, McDonald PJ, Tamber MS, Hankinson TC, Hauptman JS, Simon TD, Krieger MD, Holubkov R, Kestle JRW, Hydrocephalus Clinical Research Network (2020) Surgical resource utilization after initial treatment of infant hydrocephalus: comparing ETV, early experience of ETV with choroid plexus cauterization, and shunt insertion in the Hydrocephalus Clinical Research Network. J Neurosurg Pediatr 26(4):337–345. https://doi.org/10.3171/2020.4.PEDS19632. PMID: 32559741
42. Hankinson TC, Hauptman JS, Simon TD, Krieger MD, Holubkov R, Kestle JRW (2020) Hydrocephalus clinical research network. surgical resource utilization after initial treatment of infant hydrocephalus: comparing ETV, early experience of ETV with choroid plexus cauterization, and shunt insertion in the hydrocephalus clinical research network. J Neurosurg Pediatr 26:337–345
43. Evidence based practice in spina bifida: developing a research agenda. Liptak GS (ed) Spina Bifida Association (Washington) 2003
44. Thibadeau JK, Alriksson-Schmidt AI, Zabel TA (2010) The National Spina Bifida Program transition initiative: the people, the plan, and the process. Pediatr Clin North Am 57(4):903–10
45. Thibadeau JK, Ward EA, Soe MM, Liu T, Swanson M, Sawin KJ, Freeman KA, Castillo H, Rauen K, Schechter MS (2013) Testing the feasibility of a national spina bifida patient registry. Birth Defects Res A Clin Mol Teratol 97:36–41
46. Sawin KJ, Liu T, Ward E, Thibadeau J, Schechter MS, Soe MM, Walker W (2015) NSBPR Coordinating Committee. The national spina bifida patient registry: profile of a large cohort of participants from the first 10 clinics. J Pediatr 166:444–450
47. Shamblin IC, Corwin I, Blount JP, Hopson BD, Davis D, Swanson-Kimani E, Rocque BG (2022) Functional level of lesion scale: validating fourteen years of research with the national spina bifida patient registry. J Pediatr Rehabil Med 15:587–591
48. Mazzola CA, Assassi N, Baird LC, Bauer DF, Beier AD, Blount JP, Durham SR, Flannery AM, Klimo P, McClung-Smith C, Nikas DC, Rehring P, Tamber MS, Tyagi R (2019) Congress of neurological surgeons systematic review and evidence-based guidelines for pediatric myelomeningocele: executive summary. Neurosurgery 85:299–301
49. Blount JP, Bowman R, Dias MS, Hopson B, Partington MD, Rocque BG (2020) Neurosurgery guidelines for the care of people with spina bifida. J Pediatr Rehabil Med 13:467–477
50. Tamber MS, Flannery AM, McClung-Smith C, Assassi N, Bauer DF, Beier AD, Blount JP, Durham SR, Klimo P Jr, Nikas DC, Rehring P, Tyagi R, Mazzola CA (2019) Congress of neurological surgeons systematic review and evidence-based guideline on the incidence of shunt-dependent hydrocephalus in infants with myelomeningocele after prenatal versus postnatal repair. Neurosurgery 85:E405–E408
51. Bauer DF, Beier AD, Nikas DC, Assassi N, Blount J, Durham SR, Flannery AM, Klimo P Jr, McClung-Smith C, Rehring P, Tamber MS, Tyagi R, Mazzola CA (2019) Congress of neurological surgeons systematic review and evidence-based guideline on the management of patients with myelomeningocele: whether prenatal or postnatal closure affects future ambulatory status. Neurosurgery 85:E409–E411
52. Beier AD, Nikas DC, Assassi N, Bauer DF, Blount JP, Durham SR, Flannery AM, Klimo P, McClung-Smith C, Rehring P, Tamber MS, Tyagi R, Mazzola CA (2019) Congress of neurological surgeons systematic review and evidence-based guideline on closure of myelomeningocele within 48 hours to decrease infection risk. Neurosurgery 85:E412–E413
53. Blount JP, Durham SR, Klimo P Jr, Assassi N, Bauer DF, Beier AD, Flannery AM, McClung-Smith C, Nikas DC, Rehring P, Tamber MS, Tyagi R, Mazzola CA (2019) Congress of neurological surgeons systematic review and evidence-based guideline on the management of patients with myelomeningocele: whether persistent ventriculomegaly adversely impacts neurocognitive development. Neurosurgery 85:E414–E416
54. Mazzola CA, Tyagi R, Assassi N, Bauer DF, Beier AD, Blount JP, Durham SR, Flannery AM, Klimo P Jr, McClung-Smith C, Nikas DC, Rehring P, Tamber MS (2019) Congress of neurological surgeons systematic review and evidence-based guideline on the incidence of tethered cord syndrome in infants with myelomeningocele with prenatal versus postnatal repair. Neurosurgery 85:E417–E419
55. Kulkarni AV, Riva-Cambrin J, Browd SR, Drake JM, Holubkov R, Kestle JR, Limbrick DD, Rozzelle CJ, Simon TD, Tamber MS, Wellons JC 3rd, Whitehead WE (2014) Hydrocephalus Clinical Research Network. Endoscopic third ventriculostomy and choroid plexus cauterization in infants with hydrocephalus: a retrospective Hydrocephalus Clinical Research Network study. J Neurosurg Pediatr 14:224–229
56. Kulkarni AV, Riva-Cambrin J, Rozzelle CJ, Naftel RP, Alvey JS, Reeder RW, Holubkov R, Browd SR, Cochrane DD, Limbrick DD, Simon TD, Tamber M, Wellons JC, Whitehead WE, Kestle JRW (2018) Endoscopic third ventriculostomy and choroid plexus cauterization in infant hydrocephalus: a prospective study by the Hydrocephalus Clinical Research Network. J Neurosurg Pediatr 21:214–223
57. Riva-Cambrin J, Kestle JRW, Rozzelle CJ, Naftel RP, Alvey JS, Reeder RW, Holubkov R, Browd SR, Cochrane DD, Limbrick DD, Shannon CN, Simon TD, Tamber MS, Wellons JC, Whitehead WE, Kulkarni AV (2019) Predictors of success for combined endoscopic third ventriculostomy and choroid plexus cauterization in a North American setting: a Hydrocephalus Clinical Research Network study. J Neurosurg Pediatr 24(2):128–138
58. Warf B, Ondoma S, Kulkarni A, Donnelly R, Ampeire M, Akona J, Kabachelor CR, Mulondo R, Nsubuga BK (2009) Neurocognitive outcome and ventricular volume in children with myelomeningocele treated for hydrocephalus in Uganda. J Neurosurg Pediatr 4:564–570

59. Schiff SJ, Kulkarni AV, Mbabazi-Kabachelor E, Mugamba J, Ssenyonga P, Donnelly R, Levenbach J, Monga V, Peterson M, Cherukuri V, Warf BC (2021) Brain growth after surgical treatment for infant postinfectious hydrocephalus in Sub-Saharan Africa: 2-year results of a randomized trial. J Neurosurg Pediatr 9:1–9
60. Kulkarni AV, Schiff SJ, Mbabazi-Kabachelor E, Mugamba J, Ssenyonga P, Donnelly R, Levenbach J, Monga V, Peterson M, MacDonald M, Cherukuri V, Warf BC (2017) Endoscopic treatment versus shunting for infant hydrocephalus in Uganda. N Engl J Med 377:2456–2464
61. Piatt JH Jr (2010) Treatment of myelomeningocele: a review of outcomes and continuing neurosurgical considerations among adults. J Neurosurg Pediatr 6:515–525
62. Williams VJ, Juranek J, Stuebing KK, Cirino PT, Dennis M, Bowman RM, Blaser S, Kramer LA, Fletcher JM (2015) Postshunt lateral ventricular volume, white matter integrity, and intellectual outcomes in spina bifida and hydrocephalus. J Neurosurg Pediatr 15:410–419
63. Riva-Cambrin J, Kulkarni AV, Burr R, Rozzelle CJ, Oakes WJ, Drake JM, Alvey JS, Reeder RW, Holubkov R, Browd SR, Cochrane DD, Limbrick DD, Naftel R, Shannon CN, Simon TD, Tamber MS, McDonald PJ, Wellons JC, Luerssen TG, Whitehead WE, Kestle JRW (2021) Impact of ventricle size on neuropsychological outcomes in treated pediatric hydrocephalus: an HCRN prospective cohort study. J Neurosurg Pediatr 12:1–12
64. Dewan MC, Wellons JC (2019) Fetal surgery for spina bifida J Neurosurg Pediatr 24:105–114
65. Farmer DL, Thom EA, Brock JW 3rd, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Gupta N, Adzick NS (2018) Management of Myelomeningocele Study Investigators. The management of myelomeningocele study: full cohort 30-month pediatric outcomes. Am J Obstet Gynecol 218:256–274
66. Houtrow AJ, Thom EA, Fletcher JM, Burrows PK, Adzick NS, Thomas NH, Brock JW 3rd, Cooper T, Lee H, Bilaniuk L, Glenn OA, Pruthi S, MacPherson C, Farmer DL, Johnson MP, Howell LJ, Gupta N, Walker WO (2020) Prenatal repair of myelomeningocele and school-age functional outcomes. Pediatrics 145:e20191544
67. Moldenhauer JS, Adzick NS (2017) Fetal surgery for myelomeningocele: after the management of myelomeningocele study (MOMS). Semin Fetal Neonatal Med 22:360–366
68. Kiehna EN, Blount JP, McClung Smith C, Ocal E, Chatterjee S (2019) Introduction. Advancing the care of children with spina bifida, prenatally and postnatally. Neurosurg Focus 47(4):E1
69. Worley G, Greenberg RG, Rocque BG, Liu T, Dicianno BE, Castillo JP, Ward EA, Williams TR, Blount JP, Wiener JS (2021) Neurosurgical procedures for children with myelomeningocele after fetal or postnatal surgery: a comparative effectiveness study. Dev Med Child Neurol 63:1294–1301
70. Laskay NMB, Arynchyna AA, McClugage SG 3rd, Hopson B, Shannon C, Ditty B, Wellons JC 3rd, Blount JP, Rocque BG (2017) A comparison of the MOMS trial results to a contemporaneous, single-institution, postnatal closure cohort. Childs Nerv Syst 33:639–646
71. Tanaka ST, Yerkes EB, Routh JC, Tu DD, Austin JC, Wiener JS, Vasquez E, Joseph DB, Ahn JJ, Wallis MC, Williams T, Rose C, Baum MA, Cheng EY (2021) Urodynamic characteristics of neurogenic bladder in newborns with myelomeningocele and refinement of the definition of bladder hostility: findings from the UMPIRE multi-center study. J Pediatr Urol 17:726–732
72. Tanaka ST, Paramsothy P, Thibadeau J, Wiener JS, Joseph DB, Cheng EY, Tu D, Austin C, Koh CJ, Wallis MC, Walker WO, Smith KA, Routh JC, Baum MA (2019) Baseline urinary tract imaging in infants enrolled in the UMPIRE Protocol for children with spina bifida. J Urol 201:1193–1198
73. Blount JP, Rocque BG, Hopson BD (2021) Editorial. Social determinants in care for dysraphism. J Neurosurg Pediatr 29(4):363–364
74. Liu T, Ouyang L, Walker WO, Wiener JS, Woodward J, Castillo J, Wood HM, Tanaka ST, Adams R, Smith KA, O'Neil J, Williams TR, Ward EA, Bowman RM, Riley C (2022) Education and employment as young adults living with spina bifida transition to adulthood in the USA: a study of the National Spina Bifida Patient Registry. Dev Med Child Neurol 16:154–156
75. Karmur BS, Kulkarni AV (2018) Medical and socioeconomic predictors of quality of life in myelomeningocele patients with shunted hydrocephalus. Childs Nerv Syst 34:741–747
76. Rocque BG, Waldrop RP, Shamblin I, Arynchyna AA, Hopson B, Kerr T, Johnston JM, Rozzelle CJ, Blount JP (2020) Shunt failure clusters: an analysis of multiple, frequent shunt failures. J Neurosurg Pediatr 27:287–293
77. Rocque BG, Hopson B, Shamblin I, Liu T, Ward E, Bowman R, Foy AB, Dias M, Heuer GG, Smith K, Blount JP (2022) Time to shunt failure in children with myelomeningocele: an analysis of the National Spina Bifida Patient Registry. J Neurosurg Pediatr 19:1–6
78. Staal MJ, Meihuizen-de Regt MJ, Hess J (1987) Sudden death in hydrocephalic spina bifida aperta patients. Pediatr Neurosci 13:13–18
79. McDonnell GV, McCann JP (2000) Why do adults with spina bifida and hydrocephalus die? A clinic-based study. Eur J Pediatr Surg 10(Suppl 1):31–32
80. Oakeshott P, Hunt GM, Poulton A, Reid F (2010) Expectation of life and unexpected death in open spina bifida: a 40-year complete, non-selective, longitudinal cohort study. Dev Med Child Neurol 52:749–753
81. Patel DM, Rocque BG, Hopson B, Arynchyna A, Bishop ER, Lozano D, Blount JP (2015) Sleep-disordered breathing in patients with myelomeningocele. J Neurosurg Pediatr 16:30–35
82. Rocque BG, Maddox MH, Hopson BD, Shamblin IC, Aban I, Arynchyna AA, Blount JP (2021) Prevalence of sleep disordered breathing in children with myelomeningocele. Neurosurgery 88:785–790
83. Rocque BG, Weprin BE, Blount JP, Hopson BD, Drake JM, Hamilton MG, Williams MA, White PH, Orrico KO, Martin JE (2020) Health care transition in pediatric neurosurgery: a consensus statement from the American Society of Pediatric Neurosurgeons. J Neurosurg Pediatr 14:1–9