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Spina Bifida Guideline

Neurosurgery guidelines for the care of people with spina bifida

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Abstract. Myelomeningocele (MMC) arises from an early neural developmental anomaly and results in a variety of structural abnormalities and associated functional neurologic deficits. As such, neurologic issues are central to virtually all clinical problems. Neurosurgical intervention strives to correct or improve these defects and prevent secondary complications. These interventions include closure of the open myelomeningocele and management (across the life span) of hydrocephalus, the Chiari II malformation (C2M) and tethered spinal cord (TSC). The development of pre-natal closure techniques and reports of improved outcome with in-utero closure (IUMC) have revolutionized the neurosurgical approach to myelomeningocele. Controversies remain surrounding patient selection, maternal risks, technique of IUMC (endoscopic vs. open) and long-term outcomes. However, real gains include reduced rates of hydrocephalus, modestly improved motor capabilities and reduction in C2M morbidity. For many decades, the cornerstone of treatment of hydrocephalus for many decades has been the placement and support of ventricular shunts. Endoscopic third ventriculostomy (ETV) with or without choroid plexus coagulation (ETV/CPC) is an appealing alternate strategy that avoids the morbidity and complications associated with shunts. The exact criteria for ETV-CPC candidacy and best metrics for outcome analysis remain active areas of debate and controversy. Similarly, neurosurgical management C2M, has centered upon the indications and clinical thresholds for performing posterior fossa surgical decompression. Tethered spinal cord management incorporates the diagnosis and surgical management of adhesions formed at the initial closure site, the consequent longitudinal traction related stress on the cord and the resulting neurologic signs and symptoms.

Keywords: Spina bifida, myelomeningocele, intra-uterine myelomeningocele closure (IUMC), hydrocephalus, ventricular shunt, endoscopic third ventriculostomy (etv), endoscopic third ventriculostomy with choroid plexus coagulation (ETV-CPC), chiari II malformation (C2M), tethered spinal cord (TSC), health care guidelines, neural tube defects

1. Introduction

The genesis of all problems in myelomeningocele (MMC) arises from a disorder in development of the nervous system. The most clinically obvious problem involves the caudal spinal cord but the entire ner-

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vous system is affected. Many other body systems become involved and require management but all dysraphism associated morbidities result from impaired neurologic control of those systems. Consequently, neurosurgical issues are central in the care of patients with myelomeningocele. Despite the central nature of neurologic problems, the fundamental anomalies cannot be surgically reversed and virtually all interventions are palliative or serve to prevent further complications [1–5]. Much progress has been made in improving neurosurgical care for patients with Spina Bifida but many important questions remain [1–5].

Five topics predominate in contemporary neurosurgical care for patients with Spina Bifida:

- In utero closure of MMC (IUMC)
- Optimal management of hydrocephalus/closure
- Treatment of the Chiari II malformation (C2M)
- Management of the symptomatically tethered spinal cord (TSC)
- Transitional care to adulthood and neurosurgical care for adults with spina bifida

1.1. In utero closure of myelomeningocele

The 2012 publication of the Management of Myelomeningocele study (MOMs) trial galvanized the clinical landscape of neurosurgical care in MMC and brought prenatal neurosurgical issues to the forefront [6]. This prospective, randomized, multi-center trial demonstrated improved outcomes in multiple neurological domains associated with prenatal closure including [6]:

- A reduction (82% conventional closure compared to 40% pre-natal closure) in the need for ventricular shunts.
- A reduction in both radiographic and symptomatic Chiari II malformations (C2M).
- Improved lower extremity motor function scores that exceeded those predicted from the anatomical lesion level (on average by a single level).
- A significant improvement in the composite score of neurodevelopmental outcomes. This was a secondary outcome measure and was a composite score for which the primary scores did not show significant improvement.

The improvements in fetal/infantile outcomes were offset by higher maternal morbidity, a higher incidence of premature delivery and increased risk for invasive care and obstetrical complications in subsequent pregnancies [6]. Subsequent research by the MOMs centers has centered on refinement of surgical technique and protocols to reduce and minimize these complications [7–13]. These efforts have been fruitful and recent outcome studies suggest reductions in prematurity and maternal morbidity [13–15]. There has been an associated increase in the number of centers offering intrauterine myelomeningocele closure (IUMC). Some centers use purely endoscopic/fetoscopic technique while others perform open hysterotomy and direct repair of fetal tissue [14–16]. These appear to be practice and center preferences and a clearly superior technique has not emerged.

However, issues remain that mandate that these results are interpreted with caution. The MOMs maternal cohort was homogeneous and demographically dissimilar to many of the characteristics of groups at highest risk for Spina Bifida [22]. There is limited availability of IUMC centers and access remains limited and potentially subject to disparities. The procedure is costly and as such is of limited value in resource constrained environments where the incidence of dysraphism is highest.

There are still limited numbers of longitudinal studies that assess whether the favorable results are durable, and not offset by new problems related to IUMC. Maternal factors remain significant. Uterine closure remains a challenge after open procedures and confers some risk to subsequent pregnancies. An open approach ensures that cesarean section delivery will be required for the affected pregnancy and all subsequent pregnancies [12–16].

The frequency of premature birth has been reduced but not eliminated [14,15]. Urologic dysfunction appears higher in infants who undergo IUMC than those closed by conventional techniques [17–19]. Neurologic loss from tethered cord has the potential to reduce and offset gains seen in lower extremity motor function and bladder control observed in the original MOMs cohort. IUMC did not result in a decreased in need for clean intermittent catheterization in the most recent follow-up from the MOMs cohort [18,19]. However, the best available, current studies on the original MOMs cohort suggest that improvements in hydrocephalus, C2M/brainstem dysfunction, motor function and learning are persistent [20].

Neurosurgical prenatal counselling of parents with a fetus with spina bifida is important for all families. Neurosurgeons experienced with and dedicated to caring for patients with neural tube defects (NTDs) are uniquely qualified to discuss with families the realistic long-term expectations and challenges facing a child born with open Spina Bifida (see Prenatal Counselling

Guidelines). Route of delivery remains a controversial issue in open MMC but strong evidence that clearly demonstrates superiority of one route of delivery, (cesarean or vaginal) is lacking [21–24].

1.2. Optimal management of hydrocephalus – Initial closure

Neurosurgical care for most infants who are born with MMC begins immediately after birth with closure of the spinal defect and evaluation of the need to treat hydrocephalus [29,30]. Closure techniques have changed little over several generations and surgically defining normal layers that have become fused in the embryonal defect. Once established, these layers are used to close the spinal defect. Ventricular shunts remain the cornerstone of treatment for hydrocephalus in spina bifida but there are active controversies and research surrounding the thresholds for initiating hydrocephalus treatment and the evolving role of endoscopic third ventriculostomy with choroid plexus coagulation (ETV/CPC) [2–4,30,33,34].

Traditionally about 80% of patients with open MMC require treatment of hydrocephalus with a shunt, but the frequently problematic natural history of shunts has fostered several experienced centers to challenge conventional thresholds for treatment [5]. By tolerating larger ventricles and performing more local wound care, several experienced centers have reduced shunt rates to 55–65% [5]. The long-term neuro-cognitive impact of allowing larger ventricles is unknown but appears limited in short term evaluation. Most importantly, these patients are spared the morbidity of repeated shunt operations and infections [5].

Warf and colleagues refined traditional techniques of endoscopic third ventriculostomy (ETV) by adding choroid plexus coagulation (ETV/CPC) and reported initial high efficacy in a cohort of East African children with hydrocephalus from a variety of etiologies [32]. Further studies by Warf and colleagues in the United States as well as initial work by the Hydrocephalus Research Network (HCRN) suggest that children with spina bifida associated hydrocephalus had among the greatest success rates of 70–75% [33,34]. This led to enthusiasm and rapid expansion of the number of centers performing and offering ETV-CPC. Extensive research is underway to assess ETV-CPC but other centers (including the HCRN) appear to be struggling to attain the high rates of effectiveness observed and reported by Warf and colleagues [34].

1.3. Chiari II malformation management

C2M remains a critically important issue for children with open MMC [35–39]. Virtually all children with open MMC have a C2M, defined as caudal migration of the cerebellar vermis, the brainstem and/or fourth ventricle and distortion of the posterior fossa anatomy. This distortion is associated with brainstem dysfunction that can range widely in the severity of its clinical impact. There is ongoing controversy regarding surgical management of C2M, but there has been a decline in the frequency with which surgical decompression is performed. This decline has been in part due to growing awareness of a.) the inconsistent clinical impact of posterior fossa decompression upon symptomatic C2M, b.) the central role of decompensated hydrocephalus in symptomatic C2M, and c.) the recognition that some children have underlying irreversible brainstem pathology [36–39].

1.4. Tethered spinal cord

Tethered Spinal Cord (TSC) is thought to arise from longitudinal traction on the spinal cord that arises from the child's natural growth when scar tissue that fixates the distal cord and placode at the closure site. The normal process of conus ascension is impaired and longitudinal traction over time imparts injury and painful progressive neurologic dysfunction of the lower extremities and sphincters. While all patients with spina bifida are at risk of TSC, only about a third of patients with open MMC develop symptoms of TSC. Ongoing research efforts have focused on understanding the optimal thresholds and triggers for intervention along with improving technical aspects of untethering procedures to reduce acute morbidity and the risk for re-tethering. This problem will require particular attention as children undergoing IUMC mature due to the potential for increased risk of TSC from IUMC [40–43].

1.5. Transitional care and care for adults with spina bifida

There is increasing interest in transitional and adult care for patients with Spina Bifida [44–49]. With increased survival, there are more adults than children alive with Spina Bifida, thus there is a growing need for ongoing research to define optimum protocols and strategies to maintain quality care [44–46]. Early results suggest that there is wide variability in the quality of life for adults with Spina Bifida, and that issues such as bowel management and the pursuit of activities outside the home are associated with higher quality of life [47,48]. More centers in North America are developing transition protocols and programs however much work in this domain remains.

2. Guidelines goals and outcomes

Several guidelines were chosen by the neurosurgery working group. The goals of the guidelines were both practical and aspirational.

Primary

- Protect neurologic function and neurocognitive development by optimizing CSF dynamics throughout the lifespan.
- Preserve and sustain brain stem and spinal cord function.
- Determine short and long-term efficacy and safety of IUMC.
- Maintain and foster awareness that important clinical changes can occur from shunt malfunction in the absence of changes in ventricular imaging.

Secondary

- Reduce overall dependence upon ventricular shunts to manage hydrocephalus.
- Define and refine optimal thresholds for initial treatment of hydrocephalus.
- Refine and optimize candidacy criteria for ETV/CPC.
- Define, standardize and disseminate quality fetal and maternal morbidity and neurological outcome metrics among established IUMC programs.
- Identify optimal strategies to prevent, diagnose, and treat symptomatic TSC.
- Increase attentiveness of patient/family/medical providers to the broad clinical spectrum of neurologic decline.
- Determine the optimum timing, frequency, and role of adjunctive studies.
- Establish a lifetime care model program that allows for successful transition to independent health decision-making in adulthood.

Tertiary

- Perform, order, interpret adjunctive tests as necessary including ventricular imaging studies (MRI or CT), shunt taps, shunt X-rays, shunt settings (for programmable shunts), radionuclide studies, manual muscle testing, swallowing evaluations, direct laryngoscopy, sleep studies and neuropsychological testing.
- Use important clinical (head size, stridor/secretion control, symptoms of TSC) and imaging (changes on serial ventricular imaging parameters) to assess the contribution of hydrocephalus to other clinical problems including C2M and TSC decompensation and decline.
- Retain the crucial awareness that important and threatening changes may be present in the absence of ventricular change in imaging [10].
- Preserve and sustain spinal cord function using
  - serial clinical assessments including pain tracking and lower extremity manual muscle testing
  - urodynamic studies in collaboration with Urology colleagues
  - clinical observation for sensory and other changes typical of syringomyelia

3. Methods

Methods developed by Dicianno and the steering committee for Spina Bifida Association guidelines were followed [48]. Initially, the prior guidelines were reviewed and discussed [50]. Standardized methods were developed by the organizing committee and were disseminated amongst all working groups and carefully followed in the development of these guidelines [48]. The proposed methods centered on the development of a hierarchy of important and timely age-appropriate clinical questions. Medical librarians were consulted to perform comprehensive literature review of publications related to neurosurgery in spina bifida. Papers were stratified according to study design, cohort size, quality of the analysis and overall level of evidence present within the manuscript. These determinations were then used to designate a level of evidence for a particular guideline or consensus statement. The group met at the Spina Bifida World Congress March 16–19, 2017 and other times to discuss, debate and come to consensus on the level of evidence to support and inform the guidelines. If available literature could not specifically address or support a consensus opinion of the group then "best practice" was defined as consensus of the working group after discussion and debate. Any such determinations were further vetted by the methodology utilized for the entire guideline project. These guidelines were constructed as part of a larger overall project of comprehensive clinical guidelines for care in spina bifida.

4. Discussion

Guidelines should be directed by goals of care. Neurosurgical goals center upon the preservation of neurologic function across the lifespan. Fundamentally, spina bifida is a neurologic disorder that arises from an

Table 1
Clinical questions

Age group	Clinical questions
Pre-natal	1. What is the role for IUMC of MMC and what are its short- and long-term benefits and risks? 2. How can IUMC strategies evolve to minimize maternal risks and reduce premature delivery? 3. In what economic situations is IUMC a cost-effective strategy?
0–11 months	1. How does surgical pia-to-pia re-approximation of the neural placode (surgical "neurulation") during myelomeningocele closure reduce the risk for Tethered Cord Syndrome (TSC)? 2. Does concomitant or staged closure and shunt placement reduce complications and cost? 3. What are appropriate criteria for shunt placement in infancy? 4. Are there surgical techniques for initial shunt placement that optimize shunt performance? 5. What are the optimal metrics to evaluate brain stem function in infancy? 6. What are the optimal metrics (e.g. head growth, frequency of follow-up imaging studies and adjunctive testing) to assure optimized CSF dynamics in the newborn period? 7. What is the appropriate role for ETV-CPC in infants with MMC? 8. What is the role for operative decompression of the posterior fossa (C2MD) for symptomatic C2M in the neonatal period? 9. What is the appropriate role, timing, and frequency of ventricular imaging in the assessment of the child from 0–11 months with open spina bifida?
1 yr–2 yrs, 11 months	1. Are there surgical techniques that optimize shunt performance in infancy? 2. Are there optimal metrics to assure stable brain stem function such as swallow and sleep studies in infancy? 3. What are the optimal metrics to assure optimized CSF dynamics (head growth, frequency of follow-up imaging studies and adjunctive testing) in infancy? 4. How do ventricular size and morphology correlate with neurocognitive outcomes? 5. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes? 6. What is the optimal frequency of clinic visits and neuroimaging during infancy (ages 1–2 years 11 months)?
3–5 yrs, 11 months	1. Are there surgical techniques that optimize shunt performance? 2. Are there optimal metrics to assure stable brain stem function, such as swallow and sleep studies? 3. How does ventricular size and morphology correlate with neurocognitive outcomes? 4. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes? 5. What is the optimal frequency of clinic follow-up and neuroimaging? 6. What are the optimal metrics to assure optimized CSF dynamics (head growth trajectory no longer contributory)? 7. What are the clinical presentations, surgical indications, and optimal surgical management for syringomyelia?
6–12 yrs, 11 months	1. Are there surgical techniques that optimize shunt performance in childhood? 2. Are there optimal metrics in childhood to assure stable brain stem function, such as swallow and sleep studies? 3. How does ventricular size and morphology correlate with neurocognitive outcomes? 4. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes? 5. What is the optimal frequency of clinic visits and neuroimaging during childhood (ages 6–12 years 11 months)? 6. What are the optimal metrics to assure optimized CSF dynamics in childhood? 7. What are the clinical presentations, surgical indications, and optimal surgical management for syringomyelia in childhood? 8. Does a more aggressive approach to diagnosis and surgical intervention reduce morbidity from symptomatic TSC? 9. What is the best algorithm for assessing bladder function and interpreting changes in response to somatic growth and/or tethering?
13–17 yrs, 11 months	1. Are there surgical techniques that optimize shunt performance during teen years? 2. Are there optimal metrics to assure stable brain stem function, such as swallow and sleep studies? 3. How does ventricular size and morphology correlate with neurocognitive outcomes? 4. Are outcomes following ETV (with or without CPC) effective over time in preserving neurologic well-being and protecting neurocognitive outcomes? 5. What is the optimal frequency of clinic visits and neuroimaging during ages 13–17 years 11 months? 6. What are the optimal metrics to assure optimized CSF dynamics (head growth trajectory no longer contributory)? 7. What are the clinical presentations, surgical indications, and optimal surgical management for various forms of syringomyelia? – Holocord syrinx – Cervical syrinx – Thoracolumbar syrinx

Table 1, continued

Age group	Clinical questions
	8. Does a more aggressive approach to diagnosis and surgical intervention reduce morbidity from symptomatic TSC? 9. What is the best algorithm for assessing bladder function and interpreting changes in response to somatic growth and/or tethering? 10. What is the cause of the observed temporal increase in shunt failure rates in children during their teens? (13–17 years 11 months?) 11. What are the neurosurgical barriers to beginning the transition process? What are the optimal strategies to assure successful transition to adult care?
18+ years	1. Does the incidence of symptomatic shunt failure change or decline in adulthood? Does a lower risk for shunt malfunction impact algorithms for monitoring shunt function? 2. What variables are associated with the highest quality of life for adults living with spina bifida? 3. What are the clinical presentations and optimal management of TCS in adulthood? How do these differ from TCS during childhood? 4. What is the evidence that multidisciplinary care in adulthood improves overall outcomes? Do all adults with spina bifida need to be followed in a multidisciplinary clinic? What is the most judicious use of neurosurgical resources in this population?

Table 2
Neurosurgery guidelines

Age group	Guidelines	Evidence
Pre-natal	1. Meet with the parents of patients with fetal spina bifida soon after the diagnosis to discuss the impact of the spina bifida on the child and family.	Clinical consensus
	2. Review options with regard to continuation versus termination of pregnancy and IUMC and provide information on newborn care management.	Clinical consensus
	3. Provide prognosis for neurologic capabilities and limitations and explain the need for long-term multidisciplinary care.	Clinical consensus
	4. Recognize indications for IUMC when infants are prenatally diagnosed with MMC. Discuss this with families and refer them to regional centers that provide IUMC.	Clinical consensus
	5. Define and disseminate quality outcomes for IUMC.	Clinical consensus
	6. Encourage IUMC centers to seek, use, and continue to refine best available techniques to minimize premature delivery and other risks of IUMC.	Clinical consensus
0–11 months	1. Deliver babies with MMC who are being carried to term via cesarean OR vaginal delivery. Babies undergoing IUMC are uniformly delivered via cesarean delivery. Despite the lack of consistent evidence of superiority there appears a clinical preference toward cesarean delivery.	[22–24]
	2. Coordinate care with local and regional medical centers to optimize delivery, immediate care, transfer to centers with subspecialty availability and optimize early care for infant and mother.	Clinical consensus
	3. Protect newborn MMC patient placode with clean, moist dressings.	[26–29]
	4. Close new MMC within 48 hours of birth in viable newborns.	[5, 25–26]
	5. Surgically re-approximate the pial edges of the neural placode (“surgical neurulation”) and close the wound in sequential layers.	[26–29]
	6. Manage CSF dynamics and acute hydrocephalus. Consider the following signs and symptoms as criteria for shunt placement or ETV/CPC: a. increasing intracranial pressure (accelerating head growth, bulging fontanelle(s) b. splitting sutures c. increasing irritability d. declining oral intake and/or vomiting e. extraocular palsies or sun setting eyes f. alteration in mental status g. brainstem signs (stridor, opisthotonus, silent cry, poor control of oral secretions, hypopnea/apnea) h. CSF leak from the back wound	[2, 3, 5, 30, 32, 34]
	7. Consider C2MD for neonates in setting of brainstem crisis and only after operatively confirming the presence of functioning shunt or other adequate CSF diversion technique.	[2, 35–38]
	8. Encourage and help families to develop a relationship with a multidisciplinary spina bifida clinic.	[2, 5, 47]
	9. Follow infants younger than 12 months in clinic, at three to four month intervals.	Clinical consensus

Table 2, continued

Age group	Guidelines	Evidence
1 yr–2 yrs 11 months	1. Follow children of 1–2 years 11 months at 6-month intervals for routine care in the spina bifida clinic and remain available in event of clinical change.	Clinical Consensus
	2. Teach families the signs of acute shunt failure (headache, vomiting, and lethargy/sleepiness) and chronic shunt failure (accelerated head growth, loss of developmental milestones or neurological deterioration). Follow the child clinically to observe for these signs.	Clinical Consensus
	3. Teach families the signs of brain stem failure that might occur in this age range (poor control of oral secretions, swallowing dysfunction, stridor, and impaired language acquisition). Follow the child clinically to observe for these signs.	Clinical Consensus
	4. Teach families the signs of TSC (back pain, declining lower extremity sensorimotor function). Follow the child clinically to observe for these signs.	[42–43]
	5. Use adjunctive studies judiciously (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) to augment clinical decision-making according to clinical experience and judgment.	Clinical consensus
3–5 yrs	1. Follow children aged 3–5 years 11 months at intervals of 6–12 months in the spina bifida clinic.	Clinical consensus
11 months	2. Teach families about and review the signs of acute shunt failure (headache, vomiting, and lethargy/sleepiness), and chronic shunt failure (low grade recurring headache and neck pain, loss of developmental milestones). Follow the child clinically to observe for these signs.	Clinical consensus
	3. Teach families the signs of brain stem dysfunction that might occur in this age range (poor control of oral secretions, swallowing dysfunction, stridor, and impaired language acquisition). Follow the child clinically observing for these signs.	Clinical consensus
	4. Teach families the signs of TSC (back pain, declining lower extremity sensorimotor function) and urologic dysfunction. Follow the child clinically to observe for these signs.	[40–43]
	5. Teach families of signs of syringomyelia (back pain, sensory changes in hands). Follow the child clinically to observe for these signs.	Clinical consensus
	6. Use adjunctive studies judiciously (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well child, according to experience, preference and best clinical judgment, to augment clinical decision-making.	Clinical consensus
6–12 yrs	1. Follow children ages 6–12 years 11 months at 12-month intervals in the spina bifida clinic.	Clinical consensus
11 months	2. Review the signs of acute shunt failure (headache, neck pain, vomiting, and lethargy/sleepiness), and chronic shunt failure (recurring low-grade headache and neck pain; loss of developmental milestones; cognitive, behavioral, or neurological decline; and orthopedic or urological regression) with the family. Follow the child clinically to observe for these signs.	[2, 3, 5, 30, 32, 34]
	3. Teach or review with the family and urge them to observe for the signs of TSC (back pain, declining lower extremity sensorimotor function, bladder or bowel control decline and progressive orthopedic deformities and/or scoliosis). Follow the child clinically to observe for these signs.	[42–43]
	4. Teach or review with the signs of syringomyelia (neck or back pain and sensorimotor changes in arms and hands). Follow clinically to observe for these signs.	Clinical consensus
	5. Review the signs of brain stem dysfunction that might occur in this age range with the family. Follow clinically to observe for these signs.	Clinical consensus
	6. Use adjunctive studies to augment clinical decision-making, during routine visits with the well child judiciously and according to experience, preference, and best clinical judgment.	Clinical consensus
13–17 yrs	1. Follow children ages 13–17 years 11 months at 12-month intervals in a spina bifida clinic.	Clinical consensus
11 months	2. Begin to address transition to adult neurosurgical provider early in teen years.	Transition Guidelines
	3. Review and observe for signs of acute shunt failure and chronic shunt failure.	[2, 3, 5, 30, 32]
	4. Review with the family and child the signs of brain stem dysfunction. Follow the child clinically to observe for these signs.	Clinical consensus
	5. Teach/review with the family and child signs of TSC. Follow the child clinically to observe for these signs.	Clinical consensus
	6. Teach/review with the family and child signs of syringomyelia. Follow them clinically to observe for these signs.	Clinical consensus
	7. Use adjunctive studies judiciously during routine visits with the well child, according to experience, preference and best clinical judgment, to augment clinical decision-making.	Clinical consensus
18+ years	1. Follow adults of 18+ years at 12-month intervals in an adult Spina Bifida clinic setting.	Clinical consensus
	2. Assist the patient/family in identifying an adult neurosurgery provider.	Clinical consensus
	3. Facilitate and support completion of transitional care.	Transition Guidelines
	4. Review with the adult and family the signs of acute shunt failure and chronic shunt failure. Follow clinically to observe for these signs.	Clinical consensus

Table 2, continued

Age group	Guidelines	Evidence
	5. Review with the adult and family the signs of brain stem dysfunction in adults. Follow them clinically to observe for these signs.	Clinical consensus
	6. Teach/review with the adult and family to observe for signs of TSC Follow clinically to observe for these signs.	Clinical consensus
	7. Teach/review with the adult and family to observe for signs of syringomyelia. Follow the adult clinically to observe for these signs.	[28–33]
	8. Use adjunctive studies judiciously during routine visits with the well child, according to experience, preference, and best clinical judgment, to augment clinical decision-making.	Clinical consensus
	9. Encourage pediatric neurosurgeons to be available for education and teaching opportunities from the adult spina bifida team in order to learn how to provide care to those with spina bifida.	Clinical consensus

anomaly that occurs very early in embryonic development. Most, if not all other clinical problems arise from impaired neurologic control of other body systems. No neurosurgical procedure can repair or ameliorate the fundamental problem, therefore at some level all procedures are palliative. Despite these limitations it is clear that support of neurologic function by closing the initial defect, optimizing CSF dynamics and preventing complications related to impaired brainstem function (C2M) and tethered cord is critically important in care for patients with spina bifida. These guidelines identify important steps in supporting this objective.

The original guidelines arose from a 2003 meeting entitled Evidence Based Practice in Spina Bifida: Developing a Research Agenda. The meeting was jointly sponsored by several government agencies including the Centers for Diseases Control and Prevention, the National Institute of Child Health and Human Development, the Agency for Healthcare Research and Quality, the National Institute of Health Office of Rare Disease, as well as the Spina Bifida Association. The objective of the symposium was to bring together a multi-disciplinary panel of experts and develop an evidence based prioritized agenda for research. The neurosurgery research priorities included management of hydrocephalus, the role of tethered spinal cord, scoliosis, and development of metrics to identify neurologic deterioration. Many current guidelines strive to address challenges in exactly these domains of care. In some domains progress has been slow and many of the same questions remain unanswered. Fundamental questions persist surrounding the optimal treatment of hydrocephalus, tethered cord and C2M. Each of these problems is complex and outcomes are impacted by multiple variables. As a result, observational clinical studies are challenging and limited in number. Therefore, many of the guidelines therefore arise from expert consensus. While not achieving the highest standards of evidence-based practice, these guidelines provide a framework that reflects current practice in a significant number of contemporary centers of excellence.

The most striking exception to this is the publication of the MOMs trial in 2012. The design of this prospective, randomized trial provided solid Class I evidence that demonstrated reductions in hydrocephalus, improvements in brainstem function, and possibly improved motor function. This has led to a rapid expansion in the number of centers offering IUMC. No clearly superior surgical technique has emerged and the techniques of prenatal closure remain active areas of investigation. Other research efforts have been directed toward quantifying and reducing maternal/obstetric complications and defining long-term outcomes. Concern has arisen over an increased rate of TSC and progressively worsened bladder function in patients treated with IUMC.

Pre-natal guidelines center on the long-term prognosis for neurological, orthopaedic, and urological function in children with myelomeningocele and aspects of lifetime care. Dedicated neurosurgeons with training and experience treating patients across the lifespan are uniquely qualified to advise families on these important decisions. IUMC is a critical part of the current treatment paradigm and neurosurgeons must be prepared and informed to discuss and refer patients and families for this important procedure.

Hydrocephalus is the ground zero of neurosurgery treatment in myelomeningocele. It has traditionally warranted treatment in about 80% of patients with MMC. Shunts have been the cornerstone of care and have a mixed history. While effective shunts have saved thousands of lives the morbidity remains significant for a large number of patients. New options for hydrocephalus treatment include the performance of ETV/CPC or increasing the threshold criteria for shunt placement. Early studies from both of these alternatives are promising in subsets of patients with hydrocephalus but they are not effective in all patients with MMC. The neurosurgical guidelines center on the diagnosis of

hydrocephalus and shunt failure across the lifespan and, in particular, the central concept that ventricular size on CT and MRI images is sometimes an insufficient metric for assessing hydrocephalus or shunt function. Patient symptoms and clinical deterioration may be more meaningful indications of shunt failure than any imaging finding. In infancy hydrocephalus can stress an already malformed and poorly myelinated brainstem and lead to deterioration from the C2M. In later years shunt failure may precipitate signs of tethered cord or C2M in addition to traditional symptoms of headache, emesis, neckache, and lethargy. These guidelines across the patient lifespan emphasize recognition and proactive surveillance for hydrocephalus and shunt malfunction.

Additionally, there has similarly been debate about the appropriate indications for treatment of the C2M. This has centered around the extent to which symptoms of brainstem dysfunction are caused by compression from a small posterior fossa as opposed to an intrinsic brainstem disorganization. Although urgent surgical decompression was advocated initially, poor clinical results and occasional serious complications contributed to a reduced enthusiasm for surgical decompression. These guidelines address C2M across the lifespan focusing on its protean and often age-specific manifestations.

The tethered spinal cord is an important cause of loss of neurologic function. These guidelines establish important measures to assess for clinically symptomatic TSC and educate patients and family members about identifying deterioration. Judicious use of adjunctive studies including urodynamics is an important component of the guidelines that helps support the diagnosis.

While there has been mixed progress in the neurosurgery care for the patient with spina bifida over the past 20 years the growth of the National Spina Bifida Patient Registry offers real hope for better studies to inform future protocols and guidelines. There are now $>$ 10,000 patients enrolled in the NSBPR and the number and complexity of captured variables continues to grow. This rich source of clinical data for patients with spina bifida offers the promise of larger cohorts from which to identify best practices and improve care.

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The Spina Bifida Association has already embarked on a systematic process for reviewing and updating the guidelines. Future guidelines updates will be made available as they are completed.

Executive Committee

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Conflict of interest

None of the authors has any conflict of interest to report. Each author has submitted a separate standardized form from the publisher further attesting to this.

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