SPINA BIFIDA ASSOCIATION

Guidelines for the Care of People with Spina Bifida

Neurosurgery

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Introduction

Myelomeningocele (MMC) is the most common and most serious congenital anomaly of the human nervous system that is compatible with long-term survival.[1–5] It arises from an error in neural development early in embryonic life and results in a variety of structural abnormalities and associated functional neurologic deficits. In open MMC, the caudal spinal cord is open and exposed and distal neurologic function is often lost. As such, neurologic issues last the lifespan of the individual and are central to virtually all clinical problems.[2–4] Other variant forms of dysraphism are frequently less severe, due to the absence of associated brain anomalies, and result in skin-covered anomalies that are collectively referred to as occult spinal dysraphism.[5] Spina Bifida properly refers to the full spectrum of dysraphic conditions, but by convention has evolved to refer primarily to open MMC.

The publication of the Management of Myelomeningocele study (MOMs) trial galvanized the clinical landscape of neurosurgical care in MMC and cast 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 [6,7]:

1. A pronounced reduction (82% compared to 40%) in the need for ventricular shunts.
2. A reduction in the radiographic indicators of the Chiari II malformation (C2M).
3. Improved lower extremity motor function scores that exceeded those predicted from the anatomical lesion level (> 2 levels better than expected).
4. A significant improvement in the composite score of neurodevelopmental outcomes.[8] This was a secondary outcome measure and was a composite score for which the primary scores did not show significant improvement.

These 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.[7–12] Subsequent research has focused on refinement of surgical technique and protocols to reduce and minimize these complications.[2,9,13–16] There has been an associated increase in the number of centers offering Intrauterine Myelomeningocele Closure (IUMC), with a trend toward a fetoscopic approach.[9,14,15] This less invasive uterine surgical procedure has been shown to allow mothers to deliver vaginally after the in-utero procedure and in subsequent pregnancies. The rate of premature rupture of membranes and preterm birth, however, has not improved with this less invasive procedure. However, there remain issues and challenges that:

- mandate that these results are interpreted with caution, and
- limit the widespread availability and utility of IUMC techniques.

These issues and challenges include, but are not limited to, the following:

- The procedure is costly and as such is of limited contribution in environments of resource constraint (where the incidence of dysraphism is highest). Despite recent expansion in centers performing prenatal closure, there is still limited availability of centers and access remains limited and potentially subject to disparities.
- Longitudinal outcome studies are not yet available to assess whether the favorable results are durable, lasting, and not offset by evolving new problems related to IUMC. Best available, current studies on the original MOMS cohort suggest that improvements in hydrocephalus, Chiari II malformation/brainstem dysfunction, motor function and learning are persistent.[9,13,17,18] The incidence of tethered cord in infants who undergo IUMC appears higher than those closed by conventional techniques.[19] Neurologic loss from tethered cord has some 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 decrease in need for clean intermittent catheterization in the most recent follow up from the MOMs cohort.[19–21]
- The original maternal cohort was homogeneous and dissimilar to many of the demographics of mothers that typify mothers and families with a pregnancy with Spina Bifida.[22]
- Prematurity has been reduced but not eliminated.[17,23,24]
- Maternal factors remain significant. Uterine closure remains a difficult challenge and infers some risk to subsequent pregnancies and ensures that delivery by cesarean section will be required for this and all subsequent pregnancies.[24–26]
- As experience with and the number of centers offering IUMC has increased there has been a simultaneous evolution of techniques such that several surgical approaches (i.e., open vs. endoscopic repair, and dural or skin patching techniques) now exist.[24] It is unclear which techniques will result in the best long-term outcomes with lowest complications, morbidity and mortality. As center number increases, each center is likely to see fewer cases and thereby reduce sample size associated with a given technique which may challenge studies that assess outcome and guide technical evolution of IUMC.

Beyond prenatal closure decisions, neurosurgical prenatal counseling 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 (Prenatal Counseling Guidelines). Route of delivery remains a controversial issue in open MMC but significant evidence for one route of delivery over another, such as cesarean versus vaginal delivery, remains lacking.[27,28]

Neurosurgical care for most infants who are born with MMC begins with closure of the spinal defect and subsequent evaluation for the need to treat associated hydrocephalus.[16,29–32] Ventricular shunts remain the cornerstone of treatment for hydrocephalus in Spina Bifida but there are active controversies and research surrounding:

- the thresholds for initiating treatment, and
- the evolving role of endoscopic third ventriculostomy with choroid plexus coagulation (ETV/CPC).[3,4,33–35]

Traditionally, about 80% of patients with open MMC require treatment of hydrocephalus with a shunt, but the frequently problematic and troublesome natural history of shunts has fostered several experienced centers to challenge conventional thresholds for treatment.[3,4] By tolerating larger ventricles and performing more local wound care, several experienced centers have reduced shunt rates to 55-65%.[5] A study using the National Inpatient Sample database (NIS) confirmed a significant increase in the use of delayed treatment of hydrocephalus in the study period 1998-2014.[36] Long-term follow up studies of the neuro-cognitive impact of these changes are unknown but appear limited in short-term evaluation. Most importantly, these patients are spared the morbidity of repeated shunt operations and infections.

Endoscopic third ventriculostomy with choroid plexus coagulation (ETV/CPC) is a recently developed, promising alternative to shunts for treating hydrocephalus. Warf and colleagues refined traditional techniques of endoscopic third ventriculostomy (ETV) by adding choroid plexus coagulation (CPC) and reported initial high efficacy in a cohort of East African children with hydrocephalus from a variety of etiologies.[37] Both in the original cohort and subsequent work by Warf's team in the United States, cohorts of children with hydrocephalus from Spina Bifida did the best of all etiologies with success rates of 70-75%.[33,34] This led to enthusiasm and rapid expansion of the number of centers performing and offering ETV/CPC. A grading scale for success has been developed and is widely utilized to predict success of ETV/CPC.[35] Extensive research is underway to assess ETV/CPC but other centers appear to be struggling to attain the high rates of effectiveness observed and reported by Warf and colleagues.[33,34,37]

C2M remains an important issue for children with open MMC.[38–43] By definition, every child with open MMC has a C2M, which properly refers to the entire abnormality of the brainstem and posterior fossa which is characterized by anatomic distortion with elongation and caudal displacement of the medulla and cerebellar vermis into the cervical spinal canal. This distortion imparts or is associated with brainstem dysfunction that can range widely in its clinical severity. Controversy regarding surgical management prevails but there has been a decline in the frequency with which Chiari II malformation surgical decompression (C2MD) of the posterior fossa for the C2M is performed. The decline was quantified in the report by Kim et al.[41] which used data from the National Spina Bifida Patient Registry. This study found that frequency of Chiari decompression was significantly lower for those registry subjects born before 2005 than those born in 2005 or later (10.05% vs 7.63%, p=0.0068, OR 1.32, 95% CI 1.08-1.61). This decline has been in part due to:

- growing awareness of the inconsistent impact of posterior fossa decompression upon symptomatic C2M,
- the frequency with which a symptomatic C2M is precipitated by hydrocephalus or shunt failure,[38] and
- the recognition that some children have underlying irreversible brainstem pathology.[39–43]

Tethered Spinal Cord (TSC) is another important neurosurgical issue in Spina Bifida. Ongoing research efforts have focused on understanding the optimal thresholds and triggers for intervention, and improving technical aspects of untethering procedures to reduce re-tethering. This problem will require particular attention as children undergoing IUMC mature due to the potential for increased risk of TSC from IUMC.[44–47]

There is increasing interest in transitional and adult care for patients with Spina Bifida.[48,49] With increased survival, there are more adults than children alive with Spina Bifida, and there is a growing need for ongoing research to define optimum protocols and paradigms to maintain quality care.[48–50] Early results suggest that there is a wide spectrum of quality of life for adults with Spina Bifida and that issues such as bowel management and the pursuit of personal, volunteer or job activities outside the home are associated with higher quality of life.[44–48] More centers in North America are developing transition protocols and programs but much work in this domain remains.

Outcomes

Primary

1. Protect neurologic function and neurocognitive development by optimizing cerebrospinal fluid (CSF) dynamics throughout the lifespan, and by using the following parameters to balance the risks of ongoing hydrocephalus against the risks of treatment:
    - presence or absence of neurological symptoms or signs (including those referable to C2M such as stridor and poor secretion management) or tethering as manifestations of hydrocephalus and/or shunt malfunction;
    - ventricular size/morphology (particularly changes in ventricular size on serial imaging studies), yet retain the crucial awareness that important and threatening clinical changes can occur from shunt malfunction in the absence of demonstrable changes in ventricular size;
    - head size for age as compared with normal head growth curves, and status of fontanelle(s) when applicable.
2. Perform or order 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.
3. Preserve and sustain spinal cord function using the following interventions:
    - perform regular and ongoing assessments of spinal cord function,

- refer to and collaborate with urology colleagues for urodynamic studies to support assessment for possible TSC,
- recognize and diagnose tethered cord syndrome (clinically with consideration for supporting evidence from urodynamic function studies) and perform surgical tethered cord release to preserve spinal cord function and minimize recurrent spinal cord tethering,
- optimize surveillance and treatment for symptomatic syringomyelia
- maintain stability of brain stem and lower cranial nerve function,
- recognize the importance of hydrocephalus and shunt failure in promoting symptomatic C2M, and
- optimize hydrocephalus before considering C2MD operations.

4. Improve overall mortality and morbidity of open Spina Bifida by increasing attentiveness of patient/family/medical providers to the broad clinical spectrum of neurologic decline.
5. Educate the medical community regarding the full spectrum of signs and symptoms of ventricular shunt failure.

Secondary

1. Determine short- and long-term efficacy of intra-uterine closure to prevent recognized morbidities and mortality.
2. Define and disseminate the following quality metrics among established IUMC programs:
   - fetal morbidity metrics,
   - maternal metrics, and
   - neurological outcome metrics.
3. Minimize occurrence of shunt obstruction and infection by taking steps to:
   - reduce overall dependence upon ventricular shunts to manage hydrocephalus,
   - define and refine optimal thresholds for initial treatment of hydrocephalus, and
   - refine and optimize candidacy criteria for ETV/CPC.
4. Identify optimal strategies to prevent, diagnose, and treat symptomatic tethered cord.
5. Determine the optimum timing, frequency, and role of adjunctive studies both for surveillance and in evaluating neurologic deterioration. Maximize and protect neurologic outcomes while minimizing expense and risk of diagnostic studies.
6. Establish a lifetime care model program that allows for successful transition to independent health decision-making in adulthood.

0-11 months

Clinical Questions

1. How can IUMC strategies evolve to minimize maternal risks and reduce premature delivery? What is the role for IUMC of MMC and what are its short- and long-term benefits and risks?
2. In what economic situations is IUMC a cost-effective strategy?

3. Does surgical pia-to-pia re-approximation of the neural placode (surgical “neurulation”) reduce the risk for Tethered Cord Syndrome (TCS)?
4. Does concomitant or staged closure and shunt placement reduce complications and cost?
5. What are appropriate criteria for shunt placement in infancy?
6. Are there surgical techniques that optimize shunt performance?
7. Are there optimal metrics to evaluate brainstem function?
8. What are the optimal metrics to assure optimized CSF dynamics (head growth, frequency of follow-up imaging studies and adjunctive testing)?
9. What is the appropriate role for ETV/CPC in infants with MMC?
10. What is the role for operative decompression of the posterior fossa (C2MD) for symptomatic C2M in the neonatal period?
11. 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?

Guidelines

Patient/Family

1. Consult with a multi-disciplinary team prior to birth to establish a joint delivery plan and a plan of care. (clinical consensus)
2. Learn about regional centers that could provide evaluations for the suitability of IUMC upon prenatal diagnosis of NTD if desired. (clinical consensus)
3. Support and encourage periconceptional dietary consumption of folate to minimize the incidence of folate-related Spina Bifida.[38] (Women’s Health Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

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. Review options with regard to continuation versus termination of pregnancy and IUMC and provide information on newborn care management. Provide prognosis for neurologic capabilities and limitations and explain the need for long-term multidisciplinary care. (clinical consensus) (Prenatal Counseling Guidelines)
2. 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)
3. Define and disseminate quality outcomes for IUMC. (clinical consensus)
4. Encourage IUMC centers to seek, use, and continue to refine best available techniques to minimize premature delivery and other risks of IUMC.
5. 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.[36,51]

6. 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)
7. Protect newborn MMC patient placode with clean, moist dressings.[14,15,23]
8. Close new MMC within 48 hours of birth in viable newborns.[17,25]
9. Surgically re-approximate the pial edges of the neural placode ("surgical neurulation") and close the wound in sequential layers.[14,15,23]
10. Manage CSF dynamics and acute hydrocephalus. Consider the following signs and symptoms as criteria for shunt placement or ETV/CPC:
    - increasing intracranial pressure (accelerating head growth, bulging fontanelle(s),
    - splitting sutures,
    - increasing irritability,
    - declining oral intake and/or vomiting,
    - extraocular palsies or sun setting eyes,
    - alteration in mental status,
    - brainstem signs (stridor, opisthotonus, silent cry, poor control of oral secretions, hypopnea/apnea), and
    - CSF leak from the back wound.[2,13,14]
11. 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.[16,26–28]
12. Encourage and help families to develop a relationship with a multidisciplinary Spina Bifida clinic.[23]
13. Follow infants younger than 12 months in clinic, at three-to-four-month intervals. (clinical consensus)

1-2 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brainstem function, such as swallow and sleep studies?
3. What are the optimal metrics to assure optimized CSF dynamics (head growth, frequency of follow-up imaging studies and adjunctive testing)?
4. How does 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 ages 1-2 years 11 months?

Guidelines

Patient/Family

1. Learn about and observe the child for clinical signs of brainstem dysfunction (stridor, silent cry, failure to control secretions), shunt failure, and TSC. (clinical consensus)
2. Foster and develop working relationships with the team of Spina Bifida providers. (clinical consensus)

Providers/Neurosurgeons/Spina Bifida Clinic

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 brainstem 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.[31,32]
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.[34] (clinical consensus)

3-5 years 11 months

Clinical Questions

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 during ages 3-5 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 syringomyelia?
    - Holocord syrinx
    - Cervical syrinx
    - Thoracolumbar syrinx

Guidelines

Patient/Family

1. Teach the family to learn about and observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)
2. Foster and develop working relationship with the team of Spina Bifida providers.[1,23] (clinical consensus)

Providers/Neurosurgeons/Spina Bifida clinic

1. Follow children aged 3-5 years 11 months at intervals of 6-12 months in the Spina Bifida clinic. (clinical consensus)
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 brainstem 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.[30–32,51,52]
5. Teach families the 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.[34] (clinical consensus)

6-12 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brainstem 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 6-12 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 syringomyelia?
    - Holocord syrinx
    - Cervical syrinx

- Thoracolumbar syrinx

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?

Guidelines

Patient/Family

1. Continue to encourage the family to observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)
2. Foster and develop working relationship with the team of Spina Bifida providers.[1,23] (clinical consensus)
3. Motivate the family to establish working relationships with their child's educational system including teachers and other educational professionals. (clinical consensus)
4. Urge the family to collaborate with the clinic coordinator and/or social worker to optimize resources in the setting of potential neurocognitive dysfunction, and to identify and relay neurocognitive changes to the medical team. (clinical consensus) (Neuropsychology Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow children ages 6-12 years 11 months at 12-month intervals in the Spina Bifida clinic. (clinical consensus)
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.[2,4,14]
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.[29–32,51,52]
4. Teach or review with the family and urge them to observe for 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 brainstem dysfunction that might occur in this age range (poor control of secretions, swallowing dysfunction, stridor, and declining language function) with the family. Follow clinically to observe for these signs. (clinical consensus)
6. To augment clinical decision-making, use adjunctive studies during routine visits with the well child (for example, imaging such as MRI/CT and urodynamic and sleep and swallow studies), doing so judiciously and according to experience, preference, and best clinical judgment.[34] (clinical consensus)

13-17 years 11 months

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brainstem 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 syringomyelia?
    - Holocord syrinx
    - Cervical syrinx
    - Thoracolumbar syrinx
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 aged 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?

Guidelines

Patient/Family

1. Observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC, and/or syringomyelia. (clinical consensus)
2. Continue to foster a working relationship with the team of Spina Bifida providers. (clinical consensus)
3. Neurosurgery should assist child and family in learning the concept of transition to adult care and in identifying an adult neurosurgery provider. (clinical consensus) (Transition Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow children ages 13-17 years 11 months at 12-month intervals in a Spina Bifida clinic. (clinical consensus)

2. Begin to address transition to adult neurosurgical provider early in teen years to promote self-knowledge and functional independence and encourage teen self-monitoring.[32,35] (Transition Guidelines, Self-Management and Independence Guidelines)
3. Review and observe for signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), and chronic shunt failure (recurring low-grade headache and neck pain, behavioral and/or cognitive changes, neurological decline, urological changes, and increasing orthopedic deformities and/or progressive scoliosis). Follow the child clinically to observe for these signs.[2,4,14]
4. Review with the family and child the signs of brainstem dysfunction that might occur in this age range (poor control of secretions, swallowing dysfunction, stridor, and declining language function). Follow the child clinically to observe for these signs. (clinical consensus)
5. Teach or review with the family and child and urge them to observe for signs of TSC (back pain, declining sensorimotor function, urological changes, and progressive orthopedic deformities and/or scoliosis). Follow the child clinically to observe for these signs.[29–32,51,52]
6. Teach or review with the family and child and urge them to observe for signs of syringomyelia (back pain and sensorimotor changes in arms and hands). Follow the child clinically to observe for these signs. (clinical consensus)
7. 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.[34] (clinical consensus)

18+ years

Clinical Questions

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?

Guidelines

Patient/Family

1. Observe the adult for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)

2. Continue fostering a working relationship with the team of Spina Bifida providers. (clinical consensus)
3. Adult and family should be encouraged to review information about transitioning to adult care, including 34,37:[35,37] (Transition Guidelines, Self-Management and Independence Guidelines)
    - Knowledge and autonomy for personal health decisions.
    - Awareness of own body symptoms/signs.
    - Knowledge about predictors of good quality of life in adulthood.

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow adults of 18+ years at 12-month intervals in an adult Spina Bifida clinic setting. (clinical consensus)
2. Neurosurgery should assist the patient and family in identifying an adult neurosurgery provider and facilitate and support completion of transitional care. (clinical consensus) (Transition Guidelines)
3. Review with the adult and family the signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), and chronic shunt failure (recurring low-grade headache/neck pain and changes in behavioral or cognitive function). Follow clinically to observe for these signs. (clinical consensus)
4. Review with the adult and family the signs of brainstem dysfunction in adults (poor control of secretions, swallowing dysfunction, stridor, and declining language function). Follow the adult clinically to observe for these signs. (clinical consensus)
5. Teach or review with the adult and family and urge them to observe for signs of TSC (back pain, declining sensorimotor function, and urologic dysfunction). Follow the adult clinically to observe for these signs.[29–32,51,52]
6. Teach or review with the adult and family and urge them to observe for signs of syringomyelia (back pain and sensorimotor changes in arms and hands). Follow the adult clinically to observe for these signs. (clinical consensus)
7. Use adjunctive studies judiciously to augment clinical decision-making (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well adult, according to experience, preference, and best clinical judgment.[34] (clinical consensus)
8. 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 adults with Spina Bifida.

Research Gaps

1. Will the long-term results and continued evolution of surgical technique in IUMC support broadening the use of this treatment? How will the results differ when IUMC is performed by a larger number of institutions and providers? How will quality be monitored, and with what indicators? How will quality metrics be disseminated to providers and families?

2. What clinical and/or radiological parameters should be used in deciding the need to treat hydrocephalus? What is the relationship between ventricular size and volume and long-term neurocognitive outcomes? Can morbidity and mortality be reduced - and quality of life improved - by reducing the use of ventricular shunts to manage hydrocephalus, without compromising long-term neurocognitive development?
3. What is the appropriate role for ETV/CPC?
4. What are the most meaningful and cost-effective studies to surveil and evaluate neurological decline, and how should these be used throughout the lifespan to optimize neurologic function?
5. How frequently does shunt malfunction occur without a demonstrable change in neuroimaging, and how does this population differ from those having ventricular enlargement?
6. Does shunt revision for radiographic change alone improve outcomes and prevent morbidity or mortality from emergent shunt failure later, or does the increased morbidity of such a strategy outweigh the benefits?
7. What is the optimum strategy to untether the spinal cord to protect and support spinal cord function throughout the lifespan?
8. What is the role of posterior fossa decompressive surgery for symptomatic C2M in infancy, childhood, or adulthood?

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