Neurosurgery

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Primary Outcomes

• Protect neurocognitive development by optimizing CSF dynamics throughout the life span.

• Optimize metrics for the management of CSF anomalies to protect/optimize neurocognitive development:
  • Ventricular size/morphology
  • Standardized normal growth curves
  • Correlation (and lack thereof) between clinical symptoms and changes in ventricular size in shunt malfunction; indications for shunt exploration/revision

• Preserve and sustain spinal level of function
  • Optimize metrics for assessment of spinal cord function
  • Determine/optimize long-term effectiveness of surgical intervention (or lack of) to protect spinal cord function from tethered spinal cord

• Maintain stability of brain stem, lower cranial nerve function
Outcomes

Secondary Outcomes

• Minimize occurrence of shunt obstruction and infection
  • Reduce overall dependence upon ventricular shunts to manage hydrocephalus
• Determine effectiveness of intra-uterine closure strategies to prevent recognized morbidities and mortality; define and disseminate quality metrics among established IUMC programs
• Identify optimal strategies to prevent, diagnose and arrest symptomatic tethered cord syndrome.
• Timing-frequency and proper role of studies - Brain/spine imaging, urodynamics, sleep/swallow studies. Maximize and protect neurologic outcome while minimizing expense and risk of diagnostic studies. Establish a life time care model program that allows for successful transition to independent health decision making in adulthood.
Clinical Questions

1. How can IUMC strategies evolve to minimize maternal risks and reduce premature delivery? What is the role for IUMC of MMC?

2. What is the role for operative C2MD for symptomatic C2M in neonatal period?

3. Does surgical neurulation of the placode reduce the risk for Tethered Cord Syndrome (TCS)?

4. Does concomitant or staged closure and CSF diversion produce fewer complications and less cost?

5. What are appropriate criteria for CSF diversion in infancy?

6. In what economic situations is IUMC a cost-effective strategy?
Prenatal/Infancy (through age 1 year)

Guidelines

• Patient/Family
  1. Support/encourage dietary consumption of MVI/folate to minimize incidence NTDs
  2. Learn/know of regional options with regard to IUMC programs to provide this service upon pre-natal diagnosis of NTD
Prenatal/Infancy (through age 1 year)

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  1. Know of regional IUMC closure centers and refer appropriately after discussion of options with family
  2. IUMC Centers-utilize best available techniques to minimize premature delivery and other risks of IUMC
  3. Define and disseminate quality metrics for IUMC
  4. Protect newborn MMC patient placode with clean, moist dressings
  5. Close new MMC within 48 hours of birth
  6. Surgically neurulate the placode and close in sequential layers
  7. Manage CSF dynamics and acute hydrocephalus utilizing principles of head growth, back wound leakage/failure and brain stem failure signs (e.g. stridor, opisthotonus, silent cry, poor oral secretion control) as guiding criteria for CSF diversion or ETV/CPC
  8. Consider C2MD for neonates in setting of brainstem crisis
Toddler (1-3 years)

Clinical Questions

1. Are there surgical techniques that optimize shunt performance?
2. Are there optimal metrics to assure stable brain stem function? (swallow, sleep studies)
3. What are the optimal metrics to assure optimized CSF dynamics? (head growth, imaging modalities / frequency)
4. Does ventricular size/morphology correlate with neurocognitive outcomes?
5. Are outcomes following ETV (with or without CPC) of overall value in protecting neurocognitive outcomes and minimizing risks of hydrocephalus?
6. What is the optimal follow-up frequency of clinic visits and neuroimaging during the first three years?
Guidelines

- Patient/Family
  1. Learn and observe child for clinical signs of shunt failure, brainstem stress and tethered spinal cord (TSC)
  2. Foster/develop working relationship with SB providers/team
Toddler (1-3 years)

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  1. Follow at 6 month intervals in SB clinic
  2. Teach families the signs of acute shunt failure (headache, vomiting, lethargy/sleepiness), chronic shunt failure (accelerated head growth, loss of developmental milestones) and follow clinically observing for these signs
  3. Teach families of signs of brain stem stress/failure in toddlerhood (poor secretion control, impaired language acquisition) and follow clinically observing for these signs
  4. Teach families of signs of tethered spinal cord (back pain, declining motor milestone development) and follow clinically observing for these signs
  5. Judiciously utilize adjunctive studies (imaging- MRI/CT, urodynamics, sleep studies, swallow studies) to augment clinical decision making
Clinical Questions

1. Inclusive of all of the above
   a) Optimal metrics to assure optimized CSF dynamics? (head growth trajectory no longer contributory)

2. What are the clinical presentations, surgical indications, and optimal surgical management for syringomyelia?
   a) Holocord syrinx
   b) Cervical syrinx/Chiari-like symptoms
   c) Thoracolumbar syrinx/TCS symptoms

3. What is the optimum frequency of clinic follow-up and neuroimaging in this age group?

Preschool (3-5 years)
Guidelines

• Patient/Family
  1. Learn and observe child for clinical signs of shunt failure, brainstem stress, tethered spinal cord (TSC) and syringomyelia.
  2. Foster/develop working relationship with SB providers/team
Preschool (3-5 years)

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  1. Follow at 6-12 month intervals in SB clinic
  2. Teach/review signs of acute shunt failure (headache, vomiting, lethargy/sleepiness), chronic shunt failure (low grade recurring headache and neck pain, loss of developmental milestones) and follow clinically observing for these signs
  3. Teach families of signs of brain stem stress/failure in toddlerhood (poor secretion control, declining language) and follow clinically observing for these signs
  4. Teach families of signs of tethered spinal cord (back pain, declining motor milestone development) and follow clinically observing for these signs
  5. Teach families of signs of syringomyelia (back pain, sensory changes in hands) and follow clinically observing for these signs
  6. Judiciously utilize adjunctive studies (imaging- MRI/CT, urodynamics, sleep studies, swallow studies) to augment clinical decision making
Clinical Questions

1. Does an aggressive approach to diagnosis and surgical intervention reduce morbidity from symptomatic TSC?

2. How do you account for neurologic bladder changes due to growth and/or tethering?

3. What are the clinical presentations, indications, and optimal surgical management for syringomyelia?
   a) Holocord syrinx
   b) Cervical syrinx/Chiari-like symptoms
   c) Thoracolumbar syrinx/TCS symptoms
School Age

Guidelines

• Patient/Family
  1. Observe child for clinical signs of shunt failure, brainstem stress, tethered spinal cord (TSC) and syringomyelia.
  2. Foster/develop working relationship with SB providers/team
  3. Establish working relationships with educational system and other providers to optimize resources in the setting of potential neurocognitive dysfunction, identify neurocognitive changes and relay to medical team
School Age

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  1. Follow at 12 month intervals in SB clinic
  2. Review signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), chronic shunt failure (recurring low grade headache/neck pain, loss of developmental milestones, neurological, orthopedic, or urological regression) and follow clinically observing for these signs
  3. Review signs of brain stem stress/failure in childhood (poor secretion control, declining language) and follow clinically observing for these signs
  4. Teach/review signs of tethered spinal cord (back pain, declining motor milestone development) and follow clinically observing for these signs
  5. Teach/review signs of syringomyelia (back pain, sensory changes in hands) and follow clinically observing for these signs
  6. Judiciously utilize adjunctive studies (imaging- MRI/CT, sleep studies, swallow studies) to augment clinical decision making
Teenage

Clinical Questions

• Inclusive of all of the above

1. What is cause of bump in shunt failure incidence in adolescence?
2. Does aggressive approach to diagnosis and surgical intervention reduce morbidity from symptomatic TSC?
3. What are barriers to beginning of transition process?
Guidelines

• Patient/Family
  1. Observe teen for clinical signs of shunt failure, brainstem stress, tethered spinal cord (TSC) and syringomyelia.
  2. Foster/develop working relationship with SB providers/team
  3. Begin/Learn Concepts of Transition to Adult Care
Teenage

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  1. Follow at 12 month intervals in SB clinic
  2. Initiate transition preparation early in teen years to promote self knowledge and functional independence for transition
  3. Review signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), chronic shunt failure (recurring low grade headache/neck pain, loss of developmental milestones) and follow clinically observing for these signs
Teenage

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  4. Teach/review signs of brain stem stress/failure in teens (poor secretion control, declining language) and follow clinically observing for these signs
  5. Teach/review signs of tethered spinal cord (back pain, declining motor milestone development) and follow clinically observing for these signs
  6. Teach/review signs of syringomyelia (back pain, sensory changes in hands) and follow clinically observing for these signs
  7. Judiciously utilize adjunctive studies (imaging- MRI/CT, sleep studies, swallow studies) to augment clinical decision making
Clinical Questions

1. Does the incidence of symptomatic shunt failure change/decline in adulthood? Are the metrics for observing for shunt failure impacted?

2. What are the variables associated with highest quality of life for adults living with SB?

3. What are the clinical presentations and optimal management of TCS in adulthood? How does it differ from TCS in childhood?

4. What is the strength of evidence that multidisciplinary care in adulthood improves overall outcomes? Should all adults with spina bifida be treated in a multidisciplinary clinic? How best to use precious resources in this population?
Adult

Guidelines

• Patient/Family
  1. Observe patient for clinical signs of shunt failure, brainstem stress, tethered spinal cord (TSC) and syringomyelia.
  2. Foster/develop working relationship with SB providers/team
  3. Complete Transition to Adult Care
     1. Knowledge-autonomy for personal health decisions
     2. Awareness of own body symptoms/signs
     3. Knowledge of predictors of good QOL in adulthood
Adult

Guidelines

• Providers/ Neurosurgeons/ SB Clinic
  1. Follow at 12 month intervals in adult-SB clinic
  2. Facilitate and support completion of transitional care
  3. Review signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), chronic shunt failure (recurring low grade headache/neck pain, loss of neurologic milestones) and follow clinically observing for these signs
Adult

Guidelines

• Providers/ Neurosurgeons/ SB Clinic

4. Teach/review signs of brain stem stress/failure in adults (poor secretion control, declining language) and follow clinically observing for these signs

5. Teach/review signs of tethered spinal cord (back pain, declining motor milestone development) and follow clinically observing for these signs

6. Teach/review signs of syringomyelia (back pain, sensory changes in hands) and follow clinically observing for these signs

7. Judiciously utilize adjunctive studies (imaging- MRI/CT, sleep studies, swallow studies) to augment clinical decision making
Research Gaps

• Will the long term results and continued evolution of surgical technique in IUMC support broadening the use of this modality of treatment? How might the results differ when IUMC is broadened to include a much larger number of institutions and providers? How do we monitor quality indicators and disseminate the results of these metrics?

• Can we reduce morbidity and improve QOL in MMC by reducing the use of ventricular shunts in managing hydrocephalus in Spina Bifida without compromising neurocognitive development? What, is the relationship, if any exists, between ventricular size/volume and clinical outcomes? What thresholds should we be using in assessing the need for CSF diversionary procedures in this population?
Research Gaps

• What are the most meaningful and cost effective studies and tests to obtain on MMC patients throughout the life span to support normalization and optimization of neurologic function?

• How frequently in this population does shunt malfunction occur without a demonstrable change in neuroimaging, and how does the presentation in this population differ from those in whom ventricular enlargement is seen?

• Does surgical exploration of the shunt for radiographic change alone protect from more acute, threatening, emergent shunt failure presentation later or does it cause more morbidity than it prevents?
Research Gaps

• What is the optimum threshold events for spinal cord untethering to protect and support spinal cord function throughout the life span.

• What is the role of posterior fossa decompressive surgery for symptomatic Chiari II malformation in infancy? In childhood or later life?

• What are the optimal strategies to initiate and sustain transitional care modalities to facilitate and enable mature independent medical decision making?
References

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