Devices for Neurological Disorders in Children & Infants

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One Size Does NOT Fit All… >>
Disorders

- Hydrocephalus & other abnormalities of fluid dynamics
- Movement disorders
- Epilepsy
- Neuromuscular disorders
- Cerebrovascular disorders
- Deformational plagiocephaly
Issues

• Neurological disorders are often lifelong challenges
• Many children are multiply affected
  – 10 yr old former preterm infants with shunted hydrocephalus
    • 10% have severe spasticity
    • 25% have autism spectrum disorder
    • 25% have other behavioral abnormalities (ADHD)
    • 30-50% have epilepsy, often multifocal
    • 50% have significant cognitive delay
  – Cannot exclude cognitively impaired children
• Some of the disorders are life-threatening
• Most have other medical problems other stressors

• Potentially vulnerable population
  – Multiple treatment options
  – Professional conflicts
Devices

– Hydrocephalus & other abnormalities of fluid dynamics
  – A shunt diverts fluid
    » Start in ventricles, cyst, subdural fluid, lumbar thecal sac
    » End in peritoneum, pleural cavity, cardiac atrium, gallbladder,…

– Movement disorders
– Epilepsy
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– Cerebrovascular disorders
– Deformational plagiocephaly
Shunts divert fluid

Before Shunt

After Shunt
Shunt Components

- **Catheter**
  - Tube in brain with tiny holes within 2 cm of the tip
- **Valve**
  - Multiple types – none is superior by multiple RTC
- **Tubing**
  - Length to accommodate growth
  - Rely on cavity to absorb ~ 200 ml per day
Shunts - Issues

- **Longevity**
  - Lifelong dependence from early infancy
  - Throughout life

- **Efficacy**
  - Basic plumbing – high malfunction rate
    - Catheter becomes obstructed
    - Tubing breaks or disconnects
    - Malfunction can cause transient or permanent neurological deficits or death
  - Repair requires surgery

- **Infection**
  - Excellent rates are 3-5%
  - Often requires at least 2 surgeries & 10-14 day hospital stay
  - Associated with cognitive decline and malfunction
Shunts – Clinical Trials

- Multicenter randomized controlled trials
  - Relatively small group of surgeons
  - Treatment is rarely optional
- Definable objective specific criteria
  - Population
    - Initial procedure or revision for specific diagnoses
    - Lifelong follow-up is required
  - Generally agreed upon specific clinical criteria for treatment
  - Specific outcome parameters
- Poor current efficacy allows differences to be detected
  - Infant shunts – 50% fail in first year
  - Can use relatively short follow-up period (6 mo to 1 year)
  - Treatment is required
  - Few other effective treatment options
Shunts – Other Treatment Option

• Endoscopic third ventriculostomy
  – Suitable for limited specific population
  – Use in infants is controversial
• Efficacy varies by population
  – Teen with late-onset aqueductal stenosis
    • Initial treatment with ETV
    • Expect 95% long term success
  – Teen with myelomeningocele and current shunt infection
    • ETV to try to avoid replacing shunt
    • Expect 50% long term success with ETV if anatomy suitable
Devices

• Hydrocephalus & other abnormalities of fluid dynamics
• Movement disorders
  – Intrathecal baclofen pump – spasticity & dystonia
  – Deep brain stimulator - dystonia
• Epilepsy
• Neuromuscular disorders
• Cerebrovascular disorders
• Deformational plagiocephaly
Intrathecal Baclofen Pump

- **Components**
  - Catheter within thecal sac
  - Tissue pocket in abdominal wall
  - holds pump
- **Intrathecal baclofen**
  - GABA (g-amino butyric acid) agonist
  - Intrathecal baclofen 1000x
  - more potent than oral
Advantages - Intrathecal Baclofen Pump

• Any etiology of spasticity or dystonia
  – Prematurity
  – Meningoencephalitis
  – Inflicted trauma
  – Optimal candidate is typically non-ambulatory
• Adjustable dose
• Treats combination of spasticity and dystonia
• No risk of direct intracranial injury
• Track record over decades
Issues - Intrathecal Baclofen Pump

- Longevity
  - Pump requires replacement every 7 years
- Size
  - Difficult to use in child under 15 kg
- Malfunction
  - Physiological dependence on baclofen
  - Withdrawal or overdose requires emergent expert care
  - Most malfunctions require surgery
    - Catheter breaks, clogs
    - Pump malfunction rare
- Infection
  - Excellent rate is 5-8%
  - Often requires removal (surgery)
    - Prolonged intravenous antibiotics
    - Treatment for withdrawal
Deep Brain Stimulation

• Primarily for dystonia
  – Indicated for primary dystonia
  – Populations for secondary dystonia being defined

• Components
  – Electrode inserted into a specific brain location
  – Generator inserted in tissue pocket in upper chest

• Advantages
  – Less bulky – easier on children with poor weight gain
  – Much lower physiological dependency
  – Frequent use in adults provides information
Issues  Deep Brain Stimulation

• Longevity
  – Generator requires replacement

• Malfunction

• Infection
  – Often requires removal (surgery)
    • Prolonged intravenous antibiotics
Devices

- Hydrocephalus & other abnormalities of fluid dynamics
- Movement disorders
- Epilepsy
  - Vagal nerve stimulator
  - Deep brain stimulator
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Vagal Nerve Stimulator

- Spectrum of epilepsy patients
  - Much more effective than new medicine
  - Much less effective than intracranial resection of seizure focus

- Advantages
  - Relatively low surgical morbidity

- Issues
  - Longevity
    - Generator replacement every 3-5 years
  - Malfunction
    - Lead breakage
  - Infection
    - Re-operation has much higher risks
  - Not MRI compatible
    - Relatively contra-indicated in tumor patients
Devices

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**Neuromuscular disorders**
- Spine Instrumentation
- Bladder stimulation
  - Multiple surgical options – none are ideal
  - Diaphragm pacing

- Cerebrovascular disorders
- Deformational plagiocephaly
Devices

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  - Interventional neuroradiology
    - Coils
    - Stents
    - Glue
    - Too new and too few patients to test in trials
- Deformational plagiocephaly
Devices

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  - Cranial orthotic devices are currently regulated as Class II devices despite no data to support safety concerns
  - No trial currently planned
    - No associated neurological deficits or risks
    - No agreed upon rating scale to define patient population
    - Five-year follow-up required – huge expense

Conclusions

• Each disorder and device had unique concerns
  – Specific criteria
    • Diagnosis
    • Treatment with device
    • Comparison to other treatment options
    • Efficacy
    • Complications
    • Follow-up

• Children are not little adults with adult disorders
  – Different disease processes (even with same name)
  – Longevity, malfunction, and infection

One size does NOT fit all!