Neurosurgical Options to Treat Epilepsy in Children

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Pediatric Neuroscience Symposium 5/14/2016

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Financial disclosures

• None
Overview

• Pediatric Epilepsy Team at CMHH
• What is Intractable Epilepsy?
• How does an evaluation work?
• What treatments do we offer for pediatric patients?
• Contact info
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Pediatric Epilepsy in the U.S. and Texas

- Epilepsy and seizures affect almost 3 million in USA
  - 6,895,969 children in Texas (2011), Approx. 69,000 (1%) with Epilepsy
- 1/3 of children with epilepsy will be refractory to medications (failure of 2-3 AEDs to control seizures)
  - Aprox. 21,000 children in Texas with Intractable Epilepsy
- Epilepsy surgery is a very effective treatment
  - Average length of time from seizure onset to surgery is 17 to 23 years
  - Children with uncontrolled convulsive seizures who are surgical candidates are in highest risk group for SUDEP

What is refractory (intractable) epilepsy?

- 2/3 of children with epilepsy respond well to our current medications (Anticonvulsant drugs)
  - Many of these children can “outgrow” their seizures and need for medications over time
- But once a child fails 2 or 3 different drugs (i.e. continues to have seizures) with the proper type of medicine and dosing, their chances of becoming seizure free with subsequent drugs is less than 2%.
  - This category of epilepsy is called “intractable”
Intractable Epilepsy

• Persistent seizures, especially in EARLY INTRACTABLE EPILEPSY, have a detrimental effect on cognitive and social development, and quality of life

• Goal of epilepsy treatment is to control seizures as soon as possible, and thus optimize cognitive development and improve behavior and quality of life
Children are not little adults...

RATIONALE FOR PEDIATRIC EPILEPSY
SURGICAL SERVICES

General medical agreement was reached (with evidence) that children are best cared for in clinical units with experience in providing pediatric care (1,2). The committee agreed that neurobiologic aspects of epilepsy are unique to children, especially the young, and as such require specific pediatric epilepsy expertise. Collectively, these features justify the unique approach necessary for dedicated pediatric epilepsy surgery centers.
Challenges of Epilepsy in Children

Effects of errors in brain development combined with early brain functional plasticity result in:

- **Complex or unpredictable reorganization of functional representation and connections**
  - atypical anatomical representation and widespread inefficiencies of cortical networks for cognitive and language functions

- **Complex epileptogenic networks**
  - Focal lesions can be associated with epileptogenesis and seizure onset at remote sites
  - MRI rarely shows full functional extent of epileptogenic zone (EZ)
Challenges of Epilepsy in Children

• Unlike adults, extratemporal epilepsy is more common than temporal lobe epilepsy
• The most common pathological finding is Cortical Dysplasia (50-70% of surgical cases)
• High prevalence of “nonlesional” or “multilesional” MRI findings
• Hemispheric or unilateral focal etiologies can manifest generalized seizures and EEG patterns, rapid evolution of electroclinical features, and progressive neurologic findings
• Developmental arrest, or progressive cognitive, behavioral and psychiatric problems are common, making early surgical intervention critical in preventing further developmental regression
Pediatric Epilepsy Surgery Evaluation at UT/CMHH

• Noninvasive Presurgical Tools:
  • Scalp Video EEG monitoring
  • 3T MRI - structural
  • Developmentally targeted Neuropsychological testing
  • PET, SPECT, fMRI, DTI, MEG

• Invasive Pre-resection Tools:
  • Stereo-EEG (SEEG) placement and monitoring
  • Subdural grid and strip electrode placement and monitoring
Role of Pediatric Epilepsy Surgery

- **Goals of presurgical evaluation:**
  - Is patient a “good candidate” for resection?
  - Localize or at least lateralize the Epileptogenic Zone (EZ)
  - Identify functional areas and proximity to EZ
  - Determine need/location of invasive monitoring (iEEG, SEEG)

- **Goals of resection:**
  - Seizure freedom or reduction in seizure burden
  - Spare eloquent cortex as much as possible
  - Disrupt/reverse developmental arrest or regression, to improve long-term developmental outcome
Surgical Treatments

• Vagal Nerve Stimulation
• Temporal Lobectomy
• Corpus Callosotomy
• Hemispherotomy
• Invasive Monitoring
• Focal Resection
Vagal Nerve Stimulation Patients

• Patients with multiple regions of onset for their epilepsy (usually both sides of their brains)

• **Benefits**
  – Short outpatient procedure
  – Reversible
  – Can reduce seizure frequency in certain patients

• **Risks**
  – Does not always work
  – Infection (1-2%)
  – Voice Changes
Vagal Nerve Stimulator

Stein, AG. “Alternative Treatments for Intractable Epilepsy,” BNI Quarterly, 1999
Temporal Lobectomy Patients

• Patients have confirmed epilepsy originating from one temporal lobe
  – Electroencephalogram (EEG)
  – High resolution structural Brain MRI
    • Mesial Temporal Sclerosis (scarring of the hippocampus)
  – Neuropsychological Testing
  – Adjunct method
    • Magnetoencephalogram (MEG)
    • Positron Emission Tomography (PET)
    • Single Photon Emission Computed Tomography (SPECT)

• Typically will spend 2-3 days after surgery in the hospital

• Benefits
  – 70% likelihood of seizure freedom

• Risks
  – Potential for limited Vision loss
  – Limited Language processing/speech deficits (if on dominant side)
Temporal Lobectomy

- Open the skin
- Remove a window of bone
- Open the covering around the brain (dura)
- Approach the temporal horn
- Identify the amygdala and hippocampus
- Resection

Starr et al., Neurosurgical Operative Atlas: Functional, 2009
Corpus Callosotomy Patients

• Patients with drop attacks (atonic seizures)
• Patients with epileptic encephalopathy
• Typically will spent 2-3 days in the hospital after surgery

• Benefits
  – 60-70% effective at stopping drop attacks
  – Can halt encephalopathy from progressing

• Risks
  – Difficulty coordinating tasks with both hands
  – Can have trouble with visuospatial tasks (if posterior 1/3 also disconnected)
  – Transient weakness (supplementary motor area syndrome)
Corpus Callosotomy Procedure

This is the bundle of fibers that we divide

Concha et al., Neuroimage, 2006

Albright et al., Principles and Practice of Pediatric Neurosurgery, 2014
Hemispherotomy Patients

- Patients with weakness (hemiparesis) who have seizures coming from one side of the brain
  - Stroke
  - Rasmussen’s encephalitis
  - Malformations of Cortical development
    - Hemimegalencephaly
    - Cortical dysplasia
    - Polymicrogyria
  - Sturge-Weber Syndrome
- Typically will spend 6-7 days in hospital after surgery

**Benefits**
- Around 70% seizure freedom rate after surgery

**Risks**
- Increased weakness on the other side of the body (hemiparesis)
- Vision loss
Hemispherotomy Procedure

- Resection of operculum
- Occipitobasal Disconnection
- Amygdalohippocampectomy
- Transventricular Corpus Callosotony
- Frontobasal Disconnection
- Insular Resection
- Ventricular Drain
Hemispherotomy

• One of the most challenging procedures in epilepsy surgery
• Very few centers offer this in the youngest children due to surgical risks
• Illustrative case: 3 mo boy with 50-100 seizures a day, all coming from right hemisphere
• 6 hour surgery
• 7 days in hospital
• Seizure-free
From 50-100 seizures a day to Cure
Focal Resective Surgery Outcome

– Outcome studies vary, but **seizure freedom** after surgery ranges from 50 to 92%
– Outcome is best with:
  • Shorter duration of epilepsy before surgery
  • Concordance of MEG and invasive or non-invasive EEG abnormality, and abnormality on MRI (if detectable)
  • Complete removal of abnormal brain tissue based on MRI and epileptogenic zone based on MEG and/or invasive EEG
  • Certain findings on post-operative pathology
Invasive Monitoring

• When more than one hypothesis in anatomically distant areas
• Subdural Grids
• Stereotactic ElectroEncephaloGraphy (SEEG)
Stereo-ElectroEncephaloGraphy vs. Grids?

Optimal Indications for SDE
- Pediatric Patients due to Diffuse Epilepsy Etiology
- Quicker to remove epilepsy focus
- Superficial lesions that need delineation
- Mapping of language

Optimal Indications for SEEG
- Lesional cases
  - Deep sulcal focal cortical dysplasia
  - Insular/cingulate/medial parietal epilepsy
  - Where it is essential to preserve stereotactic laser ablation ability
  - Better 3D definition of the epileptogenic zone
- Non-lesional cases
  - ? Prior craniotomy/prior failed epilepsy surgery
  - Bi-lateral implants – e.g. mesial frontal epilepsy
  - Bi temporal implants
Subdural Grid Electrode Implantation
SDE

• Benefits
  • Step I – 3.5 hours, Step 2 – 2.5 hours
  • Children seize very often
  • Usually both stages can be done within 1 week and patient is home within 10 days
  • Only one skin incision
• Risks
  • Literature reports infection risk from prolonged grid implantation
Traditional vs mini-SEEG electrodes

1.2 mm

0.8 mm
SEEG: 3D Localization (interictal)
Robotic SEEG speeds up placement of electrodes and allows for greater variation in trajectories.
Low complication rates of SEEG

- Tanriverdi et al. 2009 – Montreal series >2500 electrodes
- Rate of infectious complications 1.8%
- Rate of hemorrhagic complications 0.8%

- UT experience with SEEG
- 45 patients with 624 electrodes
- 0 infections and 0 hemorrhages
- Robotic placement in 41, frame based in 4
- Skull fiducials in robotic cases to optimize registration
- Errors monitored and minimized.
- Obsessive planning and precise placement are crucial
Application of laser ablation approach to a complex case of Periventricular nodular heterotopia (PVNH)

Left temporo-occipital spikes and onsets by scalp

Case # 2
Ablation Verification
T1+Contrast
Visualase Images
Optimal targets for Laser Ablation

- Peri-ventricular nodular heterotopia
- Hypothalamic hamartoma – sessile
- Deep seated cortical dysplasia

Risks:
- Hemorrhage
- Poor seizure control if used for wrong etiology
Epilepsy Surgery Summary

• Multi-disciplinary approach!!!
• Vagal Nerve Stimulators
• Resective Surgery
• Disconnective Surgery (Callosotomoy)
• Hemispherotomoy
• Invasive Monitoring
• Stereotactatic Laser Ablation
Thanks!!

- Alisse Pratt, Anne Crocker, Monica Tso
- All Y’all
- Pediatric Neurology
  - Gretchen Von Allmen, MD
  - Jeremy Lankford, MD
  - Michael Watkins, MD
  - Michael Funke, MD
  - PEMU Staff
  - Epilepsy Clinic Staff
- Pediatric Neurosurgery
  - David Sandberg, MD
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- Residents and Fellows
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