Pediatric Neurosurgical Disorders: from head to tail

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Children's Neurosurgical Associates
Cities of 2011 Surgery Patients
**INPATIENT BRAIN TUMOR CASES, 2009**

Children’s performed nearly a quarter of all of northern California’s pediatric inpatient brain tumor surgeries in 2009.

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**INPATIENT CRANIOSYNOSTOSIS CASES, 2008-09**

Children’s treated over 39% of all of northern California’s pediatric inpatient craniosynostosis cases in 2008-09.

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**INPATIENT SPASTICITY CASES, 2008-09**

Children’s treated nearly half of all of northern California’s pediatric spasticity cases in 2008-09.
Surgical volume 2009-2011
OR Minutes, 2011

Neurosurgery

Surgeon: Peter P. Sun, MD
Case Count: 307
Minutes: 74,591 = 73%
Minutes + Set Up/Tear Down: 83,291

Surgeon: Kurtis I. Auguste, MD
Case Count: 107
Minutes: 26,050
Minutes + Set Up/Tear Down: 29,800

Total Case Minutes: 101,592
(including Dr. Gupta and Dr. Sheinberg)
Total Minutes + Set Up/Tear Down: 114,252

Top 5 Services Ranked by Minutes

GPS
Minutes: 166,653 = 32%

ORT
Minutes: 120,626 = 23%

NES
Minutes: 101,592 = 19%

OTO
Minutes: 90,670 = 17%

PLS
Minutes: 44,078 = 8%

Total OR Minutes (not including Set Up/Tear Down): 523,619
Skull Biology

- Sutures form at borders of advancing skull ossification centers with inductive signals from underlying dura
- New bone is laid down perpendicular to the suture line
- Bone deposition at the suture is driven by brain growth
- 87% of adult head size by age 2
Abnormal Head Shape

• Craniosynostosis: premature closure or absence of calvarial suture
  - creates characteristic skull deformities at birth
• External forces
• Compensatory changes from brain disorders
Abnormal Head Shape

- Plagiocephaly
- Scaphocephaly/Dolichocephaly
- Turricephaly
- Brachycephaly
- Trigonocephaly
Diagnosis

**History:** present at or apparent shortly after birth

**Exam:** shape
  - palpate suture separation
  - ridge over fused *midline* sutures

**Radiographs:** confirmatory
  - CT: for surgical planning
Computer reconstruction can give false positive!
Frontal plagiocephaly

Positional frontal plagiocephaly
- Primary, or secondary to occipital positional plagiocephaly
Frontal plagiocephaly

Unilateral coronal synostosis:
-1/10,000
-frontal plagiocephaly, ipsilateral enlarged orbit,
Contralateral nasal deviation, strabismus
- Not familial, 15% with FGR mutations
Occipital plagiocephaly

Positional plagiocephaly vs lambdoid synostosis

1/300 – 48% vs 3/100,000
Occipital plagiocephaly

**Lambdoid synostosis**
- 3/100,000
- some ipsilateral occipital flattening
  - compensatory parietal and mastoid bulges
- trapazoid vertex view
Occipital plagiocephaly

Positional plagiocephaly (deformational plagiocephaly, flat head)
- common
- occipital only, parallelegram with compensatory frontal plagiocephaly, bi-occipital
- Helmet for severe cases
Scaphocephaly

Sagittal synostosis
Scaphocephaly

• Sagittal synostosis

- 2/10,000
- Frontal bossing, occipital keel, scaphocephaly
- Early dx offers option of less invasive correction
- 15% of uncorrected pts develop elevated ICP (only single suture synostosis to have proven ICP complication)
- Possible higher incidence of developmental delays unrelated to the timing of correction
- 6% familial in an autosomal dominant pattern
Shunted hydrocephalus
Premi head shape
Trigonocephaly

**Metopic synsotosis**
- 10-20% of synostosis
- Wide spectrum of manifestations: triangular forehead to minor midline ridge
- Surgery based on degree of deformity
- 15% associated with other anomalies: heart, GU, brain
- 5.6% familial
Turricephaly/Brachycephaly

Bicoronal synostosis

- Mostly with syndromic craniosynostosis: Apert’s, Crouzon’s, Pfeiffer’s
Craniosynostosis Treatment

- Correct deformity: psychosocial benefits of deformity correction outweighs risk of surgery
- Reduce the risk of elevated ICP in sagittal synostosis
- Traditional reconstruction 6-9 months of age
- Endoscopic assisted craniectomy + helmet < 3 months of age

- Craniofacial Team approach for syndromic patients: neurosurgeon, plastic surgeon, ENT, orthodontist, oral surgeon, neuropsychologist, social worker etc...
Endoscopically Assisted
Craniosynostosis Treatment

- Endoscopic repair:
  - not yet widely accepted
  - needs post op helmet for optimal correction
  - 2 small scars = minimally invasive..

- EARLY DX!!
**Hydrocephalus:** Mismatch of CSF production and absorption

**Ventriculomegaly:** Large ventricles
Etiology of Hydrocephalus

Congenital
  Chiari II (myelomeningocele) 85%
  Aqueductal Stenosis
  Dandy Walker malformation

Acquired
  infectious
  post-hemorrhagic - IVH: 20-50%
  tumor
  post-operative
## Clinical Characteristics

<table>
<thead>
<tr>
<th>Newborns</th>
<th>Infants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Full Fontanelle</td>
<td>Enlarging HC</td>
</tr>
<tr>
<td>Sunsetting eyes</td>
<td>IV nerve palsy</td>
</tr>
<tr>
<td>Split Sutures</td>
<td>Papilledema</td>
</tr>
<tr>
<td>A&amp;Bs</td>
<td>Irritability</td>
</tr>
<tr>
<td>Macrocephaly</td>
<td>Emesis</td>
</tr>
</tbody>
</table>
# Outcome with shunts

## Natural history of untreated hydrocephalus

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mortality</td>
<td>70%</td>
</tr>
<tr>
<td>Disability</td>
<td>75%</td>
</tr>
<tr>
<td>Mental Retardation</td>
<td>&gt;50%</td>
</tr>
</tbody>
</table>

Foltz EL, J Neurosurg 1963  
Laurence KH, Arch Dis Chil 1965

## Outcome of shunted hydrocephalus

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mortality</td>
<td>&lt;15%</td>
</tr>
<tr>
<td>IQ&gt;80</td>
<td>50%</td>
</tr>
<tr>
<td>(verbal IQ &gt; performance IQ)</td>
<td></td>
</tr>
</tbody>
</table>

Palliative

Hirsch JF, Child’s Nerv Syst 1994
Prognostic variables of shunted hydrocephalus

• Etiology
• Age at Diagnosis
• Degree of Ventriculomegaly
• Age at Treatment
## Outcome of shunted hydrocephalus

<table>
<thead>
<tr>
<th>Etiology</th>
<th>IQ &gt; 80</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chiari II</td>
<td>70%(^1-3)</td>
</tr>
<tr>
<td>Aqueductal Stenosis</td>
<td>50-65%(^4,5)</td>
</tr>
<tr>
<td>Dandy-Walker</td>
<td>30-50%(^6,7)</td>
</tr>
<tr>
<td>IVH</td>
<td>~Grade &amp; lesion</td>
</tr>
</tbody>
</table>

1. Hemmer R, Arch Psych Nerv (German) 1981
2. McCullough DC, J Neurosurg 1982
Fetal hydrocephalus: AW > 20mm for greater than 2 months leads to uniformly poor outcome

- Rapidly progressing fetal hydrocephalus, early delivery and shunting may be considered after 32 weeks (C-section)
For infants with overt hydrocephalus, clinical experience points to a critical limit of age 2-5 month for shunting to achieve a good outcome.
1. No traffic
2. IV abx
3. IT Vanco
4. Abx irrigation (Baci/Kefzol)
5. Triple prep

2010: Infection rate 1.3%!
Implications of a shunt

• Lifetime dependence
Shunt Malfunction

- 30-40% failure rate in 1st year\(^1\)
- @ 4-6 revisions / 20yrs\(^1\)
- independent of shunt type, 50% in 2 years\(^2\)
Proximal Shunt Malfunction

- choroid plexus / ventricular surface: OVERDRAINAGE - siphoning, one way valve
- too short
- poor positioning
(Left) Do not remove the valve from the sterile packaging. Turn sterile package face down so that the translucent tray and valve are facing up. Position the Locator Tool above the valve so that the Locator Tool’s blue flow direction arrow is aligned and centered with the direction of CSF flow through the valve.

(Right) Keeping the Locator Tool in position, set the Indicator Tool into the Locator Tool while aligning the red bands on the tools and rotating the Indicator Tool as necessary until it settles into place. Record current performance level setting.

NOTE: All valves are preset to Performance Level 0.5.
Adjustable Shunts + Intraoperative Imaging + Computer Navigation

Shunt Revision:
2001: N=128 (240)

2010: N=47 (400+)
No more CT scans!!!
“quick MRI”: no sedation, no radiation!!
Endoscopic fenestration:
- large instrumentation
  not ideally suited for neonatal brain

Endoscopic 3rd ventriculostomy
- obstructive hydrocephalus
- relies on well developed CSF circulation,
  high failure rate <2yo
Benign External Hydrocephalus* 

Presentation: Macrocephaly
Imaging: Prominent SA spaces with mild ventriculomegaly
Resolves by 12-24 mo
Developmentally normal (40% interim motor delay)
Associated with positional plagiocephaly
88% had family history of macrocephaly

Do not Shunt

*Alvarez L, Pediatrics 1986
Parietal bossing
Primary Neurolation - (C1- S2)
Abnormal Dorsal Midline Neural Development: conceptual classification

Abnormal Primary Neurulation

Failure of Neural Tube Closure - Neural Tube Defects: spina bifida aperta / myelomeningocele
Tethered spinal cord

- Defective dorsal midline formation
  (*spina bifida, myelodysplasia, spinal dysraphism*)

- Allows inelastic mesenchymal elements to fuse with the spinal cord at the caudal level of malformation

- Tethers the spinal cord during rapid spinal column growth
Tethered spinal cord

Stretched spinal cord, conus below L2

Altered blood flow, stretched neuronal membrane, neuronal and interneuron degeneration

Progressive neurological deficits
Ascent of the Conus Medullaris

Tip of conus at birth: L2-3 interspace
Tip of conus >2 months of age: **L1-2** interspace

- **Tip of the conus at or below** L2-3** suggests spinal cord is tethered**
Subcutaneous tract
Tethered spinal cord
Diagnosis

80-90% have cutaneous manifestations

MRI screening for high risk patients with cutaneous manifestations and those patients with associated caudal abnormalities
Tethered spinal cord
Cutaneous Manifestations

1. Dimple above the crease
2. Capillary Hemangioma / dystrophic skin
3. Subcutaneous Lipoma
4. Hair Tuft
5. Asymmetrical Crease
6. Midline Appendage
Tethered spinal cord
Cutaneous Manifestations

1. Dimples that represent tethered spinal cord from a dermal sinus tract is above the intergluteal cleft (S2)
Dimples

- Shallow or deep
- Frequently associated with dystrophic skin, hemangioma
Subcutaneous tract
Dermal sinus tract: tethering lesion
mass lesion
source of infection
(epidural abscess/meningitis)
Dimple in a straight crease = normal
Sacral dimple:
- in the crease
- near the tip of the coccyx
- does not extend into the spine
- 2% of infants
Dimples above the crease = not normal
Not normal
Lumbar lesions = not normal
Smooth capillary hemangiomas may be normal
Not just a bump
Like wise……
lipoma

Dura

Tethered spinal cord
This hair is too long
Minor crease deviation @15% tethered cords, needs screening
Not just a skin tag…
Tethered spinal cord
Associated caudal lesions

MRI recommended
- Sacral agenesis
- Imperforate anus (54%)
- Cloacal extrophy, VATER
- Urinary tract anomalies
- Genital duct anomalies
Tethered spinal cord
Management

**MRI:** 3 months of age

**Surgery:**
- Generally, tethered cords should be untethered 6-12 months of age: relatively low risk, no transfusions, 3 days hospitalization
- Routine neurophysiological monitoring
Pediatric Brain Tumors:

- Almost never present with headaches alone
- Tend to be midline, and large
- Biology determined by both histology *and* location developing molecular characterization
- Most are curable; in many others long term control achieved through combination of surgery, chemotherapy and radiation
- Long term sequelae requiring requiring comprehensive pediatric subspecialty care
Pediatric Brain Tumors:

Incidence rate: 3.9 cases per 100,000 children (0-19)

@2500 new cases in the United States 2000

1. Central Brain Tumor Registry of the United States (CBTRUS)
Site of Origin
(1038 Pediatric CNS Tumors)

Cerebrum (30%)
Posterior Fossa (37%)
Ventricular (10%)
Optic Nerve/ Chiasm (9%)
Sellar/Parasellar/Pituitary (7%)
Spinal Cord (4%)
Pineal (2%)
Presentation: Symptoms*
(supratentorial / infratentorial)

Headache  +1: 99.1% / 99.4%
  +2: 87.7% / 93.1%
  +3: 64.8% / 71.3%

nausea/vomiting (71% / 86%)
seizures
speech/personality/academic performance change <6 (87% / 80%)
visual symptoms/diplopia (90% / 80%)
difficulty walking/balance (65% / 92%)
motor weakness
bladder symptoms
neck pain
back pain
failure to thrive
Presentation: Symptoms*
(supratentorial / infratentorial)

Headache  +1: 97.7 / 99.0%
+2: 90.8 / 97.2%
+3: 82.6 / 92.5 %

confusion/stupor                             coma
head tilt                                     irritability
lethargy                                      stiff neck
paresis                                       abnormal reflexes
ataxia                                         hypesthesia >3
papilledema (64%/80%)                        decreased visual acuity
visual field defect >4 (55%/9%)               CN palsy: VI, VII
tense fontanel

Low Grade Gliomas

**Good**

- JPA (may have well defined margins)
- cerebellum
- cerebral hemisphere
- tectum
- midbrain
- spinal cord

**Bad**

- Fibrillary (infiltrative)
- thalamus
- hypothalamus
- basal ganglion
- pons
- optic pathway/chiasmatic
Low Grade Gliomas

Treatment:

Surgery for tissue diagnosis and remove as much tumor safely as possible.

- Total resection offers long term DFS and is potentially curative (95-100% 10yr DFS for cerebellar astrocytomas)
- Resection limited by certain locations and infiltrative margins
- Radical resection >90-95% improves EFS (84% vs. 65% 2yr EFS Wisoff, CCG9891, 726 pts 1991-1996)
- Small residual disease may be followed
- Mindful that adjuvant therapy can offer long term control
All gone
Not all gone, rest intertwined with optic nerve treated with chemo
PNET (medulloblastoma)

Treatment: Surgery (<1.5cm²), chemotherapy, craniospinal XRT + local Xrt

Prognosis
1. Staging, amount of post operative residual tumor
2. Age < 3yo
3. Emerging evidence for histology, molecular markers
Intraoperative ultrasound:
PNET (medulloblastoma)

Survival: 65-85 % standard risk
40-50% high risk

Relapses (2-4 yrs, locally and CSF pathways) are very difficult to cure

Current directions:
HEAD START 3 and 4- chemo for high risk pts, less Xrt (1800cGy) for standard risk pts to reduce neurocognitive sequelae, high dose chemo and bone marrow transplant with no Xrt for young children
CHO is the only Head Start institution in Northern California
Ependymoma

4th ventricle, fingers projecting out of foramens

Treatment:
Surgical resection major determinant of outcome
Complete Resection + Xrt: 60-70% 5 yr EFS

Disseminated Disease (11%), subtotal resection: 20-30%
K. A.
MEG 5/4/07
MRI 11/28/05

Right hemisphere spike
Surgical removal of seizure focus