Modern Management of Hydrocephalus in Children: An Update for Pediatricians

David I. Sandberg, MD, FAANS, FACS, FAAP
Director of Pediatric Neurosurgery & Associate Professor, UT-Houston, Children’s Memorial Hermann Hospital, & Mischer Neuroscience Institute
Associate Professor, University of Texas MD Anderson Cancer Center
I have nothing to disclose
**Hydrocephalus**

**Definition:**
Imbalance between the production relative to the absorption of cerebrospinal fluid (CSF) causes relative ventricular enlargement and elevated intracranial pressure.
“Noncommunicating” Hydrocephalus

- Intraventricular-obstructive hydrocephalus
“Communicating” Hydrocephalus

- Extraventricular-obstructive hydrocephalus
Hydrocephalus

• 0.2 to 0.8/1000 live births in the USA
• Over 38,000 discharges each year with the diagnosis of hydrocephalus in children
• Total hospital charges of $1.4 to $2.0 billion dollars per year in children

(Chi et al, J Neurosurgery 103:113-118, 2005
Etiologies of Hydrocephalus

• Etiology is varied; ½ is pre-term (mostly intraventricular hemorrhage), ½ is full-term

• Usually sporadic; X-linked and autosomal recessive patterns are rare

• Associated with myelomeningocele, Dandy-Walker, and other congenital conditions
Clinical Features of Hydrocephalus

**Infants**
- Full anterior fontanelle
- “Splayed” sutures
- Head circumference crossing percentile lines (correct for prematurity)
- “Sunsetting” eyes
- Apnea/bradycardia

**Older Children/Adults**
- Headache/vomiting
- Lethargy/mental status changes
Radiographic Features Suggesting Hydrocephalus Rather than Atrophy

- Dilatation and rounding of frontal horns and temporal horns
- Effacement of Sulci
- Transependymal CSF absorption

Periventricular hypodensity (CT)

Periventricular hyperintensity (FLAIR)
Imaging

- *Head ultrasound*: exam of choice - accurate in infants, safe, portable, fast
- CT: for emergencies
- MRI
  - “Quick Brain MRI”
Treatment of Hydrocephalus

• Reduce CSF production
  – Metabolic (carbonic anhydrase inhibitors; i.e. acetazolamide)
  – Choroid plexectomy/coagulation
Treatment of Hydrocephalus

• Increase CSF absorption
  – Restore normal anatomy (i.e. remove obstructing lesion)
  – CSF Diversion
    • Ventricular shunting
    • Endoscopic procedures
Temporizing Measures in Premature Infants

- Repeated Lumbar punctures
- Repeated Ventricular Taps
- External ventricular drainage
- Ventricular Access Device/Reservoir
- Ventriculosubgaleal shunt
Ventricular taps

- Effective but...
- Risk of porencephaly and hemorrhage
Intraventricular Fibrinolysis (TPA)

• Randomized clinical trial of rTPA and artificial csf flush in and out in 70 premature infants
• no decreased shunt need; increased risk of secondary intraventricular hemorrhage

Whitelaw 2007, Pediatrics; 119; 1071-8
Ventriculostomy: Treatment of Choice for Deteriorating Patient with Hydrocephalus
Example of Ventriculostomy for Acute Hydrocephalus

- 5 month old boy presented with intermittent twitching, arching of the neck
- Sent to pediatric neurologist, EEG ordered (negative)
- Age 11 months – vomited several times over a few days then became lethargic, stopped breathing
Gross Total Surgical Resection for Definitive Treatment of Hydrocephalus
CSF Diversionary Shunts

- Proximal system
- Valve/Regulatory Mechanism
- Distal System
  - Peritoneal
  - Atrial
  - Pleural
  - Other
Shunt Complications

- 30-45% failure in 1st year
- 4-5% failure per year after 1st year
- Shunt revision is the most common surgical procedure of the pediatric neurosurgeon
Shunt Infection

• Average infection rate = 5 -10% within first year after shunt revision; vast majority within 1st 3 months postoperatively

• Highest risk in younger patients; 46% in one study of premature infants

  (Bruinsma, et. Al., Clin Microbiol Infect 2000)

• Risk of intellectual impairment, loculated CSF compartments, and even death
CSF Shunting

“The development of the valve-regulated shunt has led to the saving of more lives and the protection of function for more patient-years than any other procedure done by neurosurgeons.”

- Hal Rekate, 1999

“Everybody hates shunts. They become blocked and infected; they wander, ulcerate and perforate. . . they must be taken out, put in again, replaced or removed”

- Thomas Morley, 1976
Neuroendoscopy

• The best way to avoid shunt problems is to not put in a shunt!

• Minimally invasive means of treating primarily non-communicating hydrocephalus
Endoscopic Third Ventriculostomy
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Endoscopic Third Ventriculostomy

- 7 y/o boy, presented with headache, n/v, lethargy, and upgaze palsy

3 months Post-op
Endoscopic Third Ventriculostomy

- 9 y/o boy presented with acute h/a, n/v, papilledema
- ETV performed 10/4/2005
- Asymptomatic post-op with improvement in school grades (straight A’s) and concentration
ETV for Shunt Malfunction

- 15 year old, multiple shunt revisions, presents with headache and vomiting
- ETV/Septostomy performed, no further shunt revisions to date (9 month follow-up)
Neuroendoscopy

- Septostomy/Fenestration
  - Eliminates or reduces shunt burden
  - Compartmentalized or multiloculated hydrocephalus
Endoscopic Septostomy – Illustrative Patient

• Former 25 wk premature infant with Grade 4 IVH
• Reservoir placed for progressive macrocephaly and hydrocephalus
• VP shunt placed when weight adequate, discharged home with soft fontanelle
• Presents two months later with progressively full fontanelle; head circumference crossing percentile lines

Pre-VP shunt

Post- VP Shunt
F/u: Head circumference stabilized, no further interventions over 6 month follow-up
Endoscopic Fenestration of Intraventricular/Suprasellar Arachnoid Cysts

7 month old girl, presented with macrocephaly (>98th percentile) and full fontanelle
Endoscopic Fenestration of Intraventricular/Suprasellar Arachnoid Cysts
Endoscopic Fenestration of Intraventricular/Suprasellar Arachnoid Cysts

- Clinical result: now 5 years old, normal neurologically
Endoscopic Biopsy and Simultaneous Third Ventriculostomy

- 15 year-old boy, presented with headache and vomiting
- ETV and tumor biopsy performed: germinoma
- Patient received radiation therapy and chemotherapy with complete response
Endoscopic Aqueductoplasty

- For patients with aqueductal stenosis and isolated 4th ventricle
- With or without catheter placement

Endoscopic Aqueductoplasty: Illustrative Patient

- 25 week premature infant with Grade IV IVH
- Reservoir placed for progressive ventriculomegaly and increased head circumference
- Patient now 3 months of age, MRI shows supratentorial hydrocephalus and isolated 4th ventricle
Endoscopic Aqueductoplasty: Illustrative Patient

Pre-op

6 weeks post-op

- ETV performed in 550 children
- 81% < 1 year of age; 58% post-infectious hydrocephalus
- 284 = ETV only, 266 ETV and choroid plexus coagulation
- 47% success for ETV alone, 66% for combined ETV and CPC
Illustrative Case: ETV/Septostomy/CPC as Alternative to VP shunt in Infants

• 3 month old former 25 week premature infant with IVH, hydrocephalus s/p reservoir, now needs CSF diversion
• Previous laparotomy for necrotizing enterocolitis
Choroid Plexus Coagulation: Illustrative
Illustrative Patient: Choroid Plexus Coagulation for Hydranencephaly

- 9 month old boy
- 4 previous surgeries for giant abdominal hernia; doctors at outside institution told family nothing could be done for this child’s brain and that he would die shortly
Choroid Plexus Coagulation: Illustrative Patient

6 weeks post-op

- Patient died 2 years later of pneumonia, no further CSF diversion required
Endoscopic Tumor Biopsy and Third Ventriculostomy: Illustrative Patient

- 15 year old girl, presented with partial cranial nerve VII palsy
Combined Open and Endoscopic Approaches For Hydrocephalus and Brain Tumors

- 6 year old from Venezuela with massive craniopharyngioma and hydrocephalus
Combined Open and Endoscopic Approaches for Hydrocephalus and Brain Tumors
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Pre-op  
After Endoscopic Fenestration  
After Craniotomy
Hydrocephalus: Outcomes

Natural History of Untreated Hydrocephalus: In a study of 182 patients prior to advent of VP shunting. . .

- 64% mortality in infancy
- 20% survival to adulthood
- 60% intellectual impairment among survivors
- 70% motor impairment among survivors

(Laurence K, Coates S; Arch Dis Child, 1962)
Hydrocephalus: Outcomes

In Recent Long-Term Outcome Studies...

• 3 to 10% mortality rates for infants shunted with nontumoral hydrocephalus

• 60% attend normal school classes

• 70% can be expected to achieve social independence

• Intellectual outcomes related to etiology of hydrocephalus
  - Myelomeningoceles: < 20% require special ed.
  - Postinfectious and posthemorrhagic: > 50% require special ed

Concluding Thoughts

- Multiple etiologies, variable outcomes
- Shunts are highly effective but have many potential complications
- Technological advances in neuro-imaging and neuro-endoscopy are expanding treatment options for patients with hydrocephalus
- Many patients with hydrocephalus can have normal lives with appropriate therapy
Thank you!