Complex Hydrocephalus

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Difficult Problems Often Requiring Multiple Shunts

- Slit Ventricle Syndrome
- Dandy-Walker Syndrome
- Multiloculated Hydrocephalus
- Septal Fenestration
- Avoiding an additional shunt
Slit Ventricle Syndrome

Intermittent symptoms of shunt malfunction in a child who appears otherwise healthy

- Headaches
- Varying degrees of lethargy
- +/- nausea/vomiting
Pathophysiology of Slit Ventricle Syndrome

- Shunt over drainage of CSF
- Growing brain fills the intracranial space
  - a fixed skull filled with brain parenchyma, blood, meninges, vasculature and only small amounts of CSF
- Loss of normal intracranial compensatory mechanisms
SVS is a phenomenon occurring in children.

Adults do not get SVS.
Treatment of the Slit Ventricle Syndrome

- Observation
- Limit over drainage
- Antimigrainous therapy
- ICP monitoring & EVD placement
- Shunt revision
- Subtemporal decompression
- Cranial morcellation
- Third ventriculostomy

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Slit Ventricle Syndrome
Signs & Symptoms

- Overdrainage
- Positional headaches
- Relief by lying down
- Poor tolerance of school
- May be asymptomatic for prolonged periods
Slit Ventricles are relatively common in patients shunted in childhood.

SVS is not as common and occurs only in patients shunted in childhood.

Treatment options include observation, medication, shunt revision and third ventriculostomy.

No specific treatment or shunt type has been shown to be the best option.
Dandy-Walker Syndrome (DWS) is determined by three findings:

1. Cystic dilatation of the fourth ventricle
2. Total or partial aplasia of the cerebellar vermis
3. Supratentorial hydrocephalus
Dandy Walker Syndrome

Dandy-Walker Variant (DWV):
1. the floor & lateral walls of the 4th ventricle are visible
2. the vermis is hypoplastic
Differential Diagnosis

DWS must be differentiated from an arachnoid cyst of the posterior fossa
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Pathogenesis

- DWS develops at about the fourth week of gestation

- Associated with other anomalies:
  - partial or total agenesis of the corpus callosum
  - many genetic disorders
    - facial
    - cardiac
Incidence of Dandy Walker Syndrome

- 1:25,000 - 35,000 births
- Female:Male 1.3:1
- 80% diagnosed before 1 year of age
- Dandy Walker Variant may be an incidental finding
Incidence of Hydrocephalus

- DWS occurs in 2–4% of cases of hydrocephalus
- 90% of cases of DWS have enlarged ventricles and/or hydrocephalus
Associated Anomalies

- CNS anomalies occur approximately 70%
  - agenesis of the corpus callosum (40%)
  - encephaloceles (17%)
  - heterotopias
  - aqueductal stenosis
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Associated Anomalies

- Non-CNS anomalies include:
  - Cardiac
    - ventricular septal defects
    - patent ductus arteriosus
    - arterial septal defects
  - External defects often associated with genetic disorders
    - trisomies 18, 21, and 13
    - <2% recurrence risk
Signs & Symptoms

- Macrosomia (C-section rate 26%)
- Delayed motor development
- Cognitive dysfunction 40–70%
- Focal motor findings very low
- Almost no cerebellar dysfunction
Imaging Studies

CT Scan

MRI
Surgical Treatment

- Cyst removal
  - failure rate >75%
- Shunting for hydrocephalus
  - cyst?
  - ventricle?
  - both?
Multiloculated Hydrocephalus
Some form of guidance is necessary
- ultrasound
- stereotaxic frame
- frameless
Multiloculated Hydrocephalus

- Often seen in patients with post-hemorrhagic or post-infectious hydrocephalus.
- Often have required multiple shunts unless endoscopic techniques are successful.
Multiloculated Hydrocephalus

- The most difficult cases!
- Many technical difficulties
- The poorest outcomes
- 45% successful shunt avoidance
Frameless Stereotaxy
Maintain Intracranial Orientation
Complex Hydrocephalus

Maintain Intracranial Orientation

- Use guidance techniques liberally
  - Ultrasound
  - Stereotaxis
Frameless Stereotaxy
Isolated Lateral Ventricular Hydrocephalus

- **Etiology:**
  - Neoplastic: e.g. hypothalamic glioma
  - Congenital: e.g. atresia of Foramen of Monroe
  - Benign lesions: choroid plexus cyst or hypertrophy
  - Post infectious or hemorrhagic scarring
  - Iatrogenic: unilateral shunt overdrainage
Isolated Lateral Ventricular Hydrocephalus

- Clinical signs of increased ICP
- Radiographic: Ventricular asymmetry +
  - Non-communication of intraventricular contrast
  - Progressive monoventricular enlargement
  - Known mass obstructing Foramen of Monroe
Treatment Outcome

Success:
- On last follow-up: Absence of ILVH

Failure:
- Recurrence of ILVH symptoms & radiographic evidence
First septostomy

Success 17

Patients re-operated 10

2nd septostomy

Success 8

Patients re-operated 1

Failure 2

Patients not re-operated 5

3rd septostomy

Success 1

Total Successes: 26

n=32

Total Failures: 6
Success Rates

- All patients: 81.2% (27/32)
- All septostomies: 60.5% (26/43)
- First septostomies: 53.1% (17/32)
- Redo patients: 90% (9/10)
Septostomy: Survival Curve

*No failures occurred after 6 months*
Septostomy vs. VP Shunt

Conclusions

• Endoscopic septostomy provided long term ILVH relief in 81% of patients

• After 6 months post-op, septostomy failures not observed

• 2 or more prior shunt procedures negatively affects outcome

• Good results possible in redo septostomies

• A reasonable alternative to CSF shunting in cases of isolated lateral ventricle hydrocephalus
Avoid That Second Shunt!

- Communicate intraventricular cysts
- Communicate through the septum pellucidum for isolated ventricles or unilateral hydrocephalus
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Conclusions

- Management decisions for difficult shunt problems are best approached in a systematic manner.

- Successful treatment of the slit ventricle syndrome is often accomplished by conservative management.

- Several surgical attempts may be necessary in order to avoid additional shunts in patients with loculated ventricles.