Neurosurgery in the NICU:

Spina Bifida and Hydrocephalus

Daniel Fulkerson, M.D.

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Disclosures:

- None

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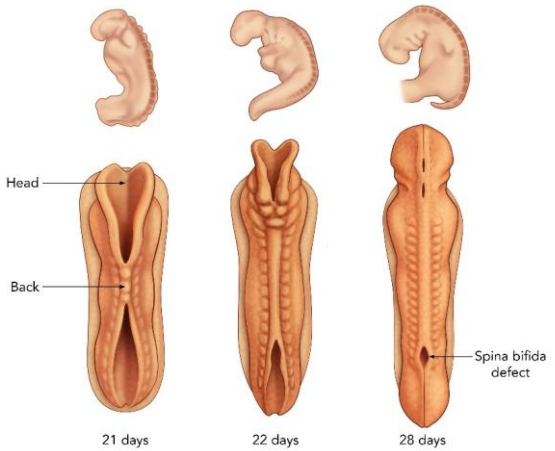
What is Spina Bifida?



- The neural tube is a structure made up of special cells that eventually come together to form the babies' brain, spinal cord and the structure that surrounds them
- Neural tube closure is usually complete by the 28th day of pregnancy, before a woman may even know she is pregnant
- In Spina Bifida, the neural tube fails to properly form and encapsulate the neural structures. This can occur anywhere along the spinal column. This is called a **NEURAL TUBE DEFECT (NTD)**

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Epidemiology:

- Spina Bifida occurs in about 7 of 10,000 live births in the United States
- Risk has declined because of folate supplementation and prenatal screening
- Risks:
  - Anticonvulsants (VPA, carbamazepine)
  - Moms with Type 1 Diabetes
  - Obesity
  - Siblings:
    - After 1 child, risk is 2.8%
    - After 2 children with spina bifida, risk is 4.8%

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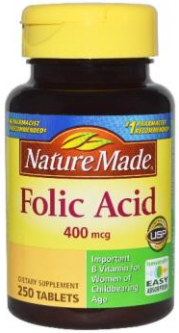
What causes Spina Bifida?

- Both genetic and environmental factors
- One well-known risk factor for Spina Bifida is ***folate deficiency***. That is why it's recommended that all women of child-bearing age take a daily multivitamin (that contains appropriate amounts of folic acid)
- Other risk factors that may increase the risk of having a child with Spina Bifida include; maternal obesity, use of certain seizure medications during pregnancy, history of diabetes and exposure to heat early during pregnancy
- 1st degree relatives of people with Spina Bifida also have a slightly increased risk of having a child with Spina Bifida (compared to the general population)



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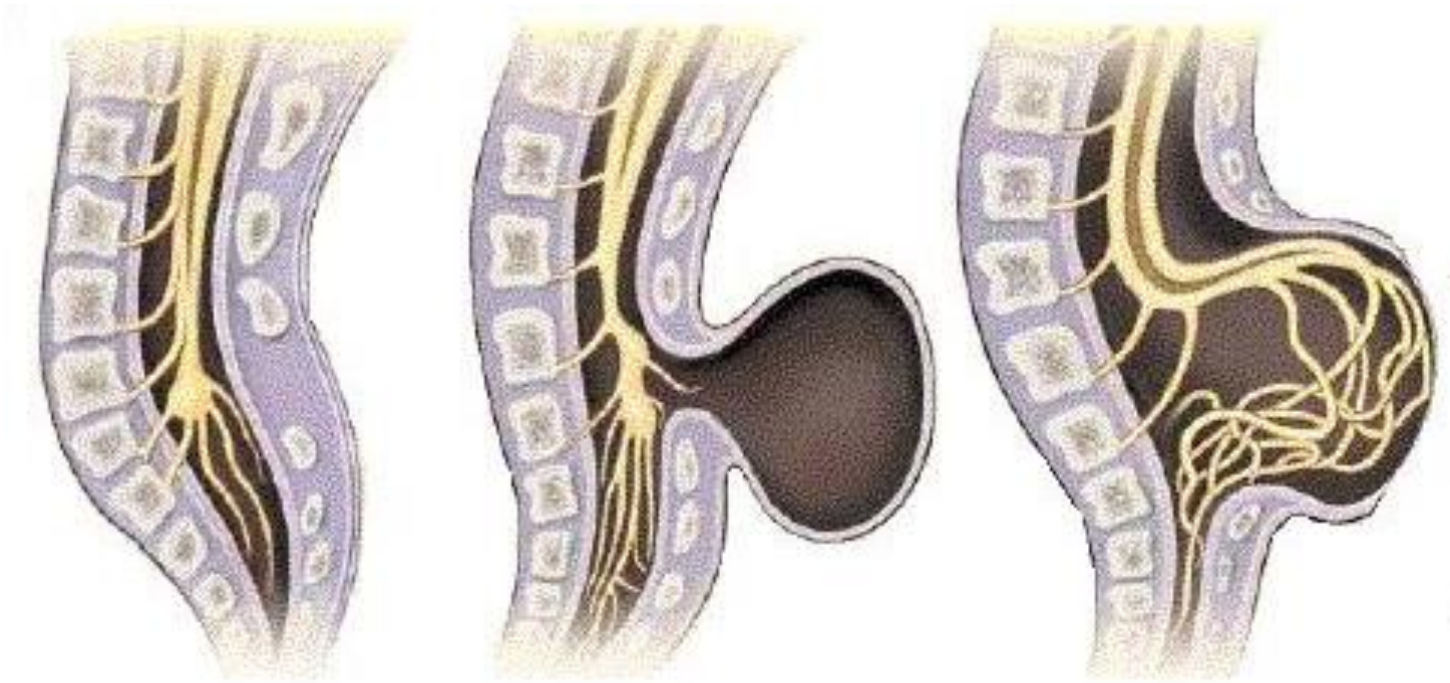
Main types of Spina Bifida



Spina bifida occulta

Meningocele

Myelomeningocele

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What are the complications of myelomeningocele?

- Children born with a myelomeningocele are usually more profoundly affected when their defect is higher up on the spine (towards the head)
- Skin issues
- Bowel and bladder issues
- Orthopedic Issues
- Chiari malformation
- Hydrocephalus

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Initial management:

- Exam
  - Measure the head size
  - Monitor respirations
- Cover the defect with a wet, non-stick pad
- Place child prone
- Initiate antibiotics: ampicillin and gentamycin
- Head Ultrasound

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Skin issues:

- We close the back within 48 hours
- The wound may be very near the diaper area – it needs meticulous care!
- Baby is kept prone for 5-7 days
- Daily measurement of OFC and weekly ultrasound
- Antibiotics are continued for 48 hours after closure

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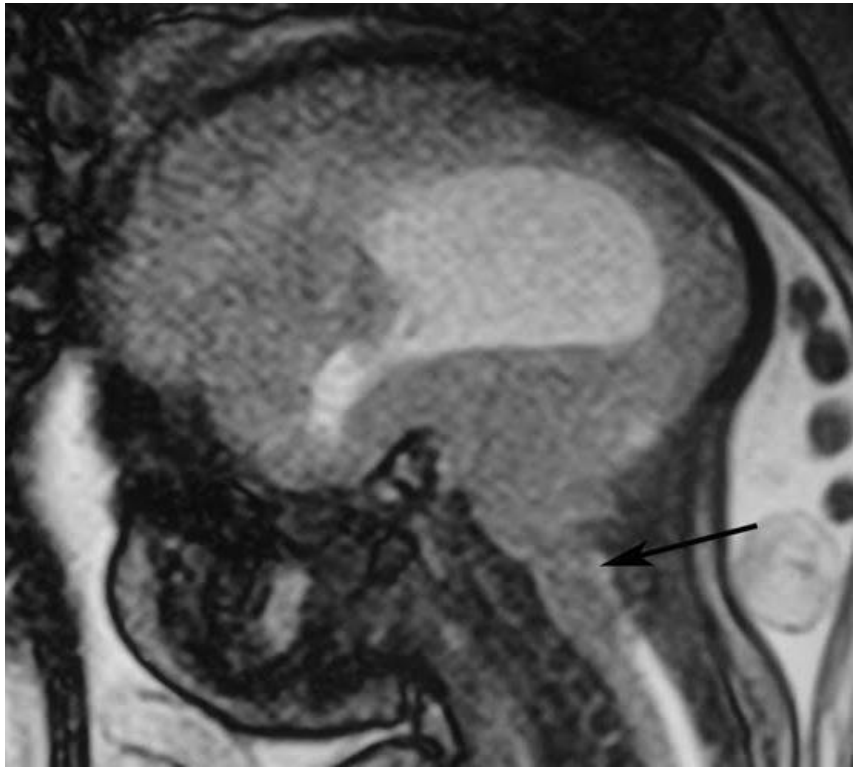
Chiari Malformation

- Herniation of the brainstem and part of the cerebellum through the base of the skull
- 95% of children born with a myelomeningocele defect have a Chiari II malformation
- Symptoms: progressive swallowing difficulties and disordered breathing patterns
- The best surgical option to manage Chiari symptoms is to *treat hydrocephalus*
- Newborns with apnea, bilateral vocal cord palsies, absent gag or swallowing function and hypotonia usually have a poor outcome despite treatment



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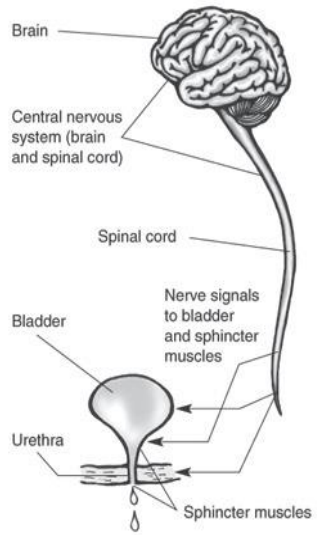
Urinary issues in Spina Bifida

- Many children with Spina Bifida have a neurogenic bladder. This occurs when the nerves that control the bladder do not work
- In addition, damage of the nerves can also affect the ability of the bladder to empty completely. The bladder's sphincter muscle also may be affected (it may stay relaxed or not relax when the bladder is contracting)
- Most children will require anticholinergic medications (like Ditropan) and clean intermittent catheterization to prevent problems
- Many children with Spina Bifida also take daily prophylactic antibiotics to prevent a urinary tract infection



Urinary issues:

- Only a very small percentage of children with S.B. (~ 5%) can properly empty their bladders without help (catheterization or surgical diversion)
- All children with Spina Bifida will be followed throughout their life by a urologist, since a neurogenic bladder is a life-long issue

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Bowel issues in Spina Bifida

- Bowel elimination issues are another common problem that people with Spina Bifida may experience. Constipation, diarrhea, incontinence and fecal impaction can all occur if not managed properly
- Children are often placed on aggressive bowel regimens to prevent constipation (stool softeners or enemas may be used)
- Patients should have established toilet times each day to train their bowels
- Children should eat diets high in fiber, be encouraged to exercise as much as possible and to take in adequate fluids (especially water)
- Some children who are unable to achieve fecal continence with the above mentioned strategies may require placement of a M.A.C.E

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Orthopedic Issues:

- Foot and ankle deformities are present in 80 to 95 percent of patients
- Some children will have hip dislocation or subluxation
- can be impaired by this abnormality)
- Spine issues: scoliosis

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Other services:

- **Nutrition**: Recommend baby be placed on daily multivitamin/ zinc supplement to promote wound healing
- **Speech therapy**: Baby should have a speech therapy evaluation with the first oral feed. Some infants will need a formal swallow study if there is concern for aspiration
- **Physical Therapy/Occupational therapy**

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Hydrocephalus:

70-85% of children with MMC will develop hydrocephalus

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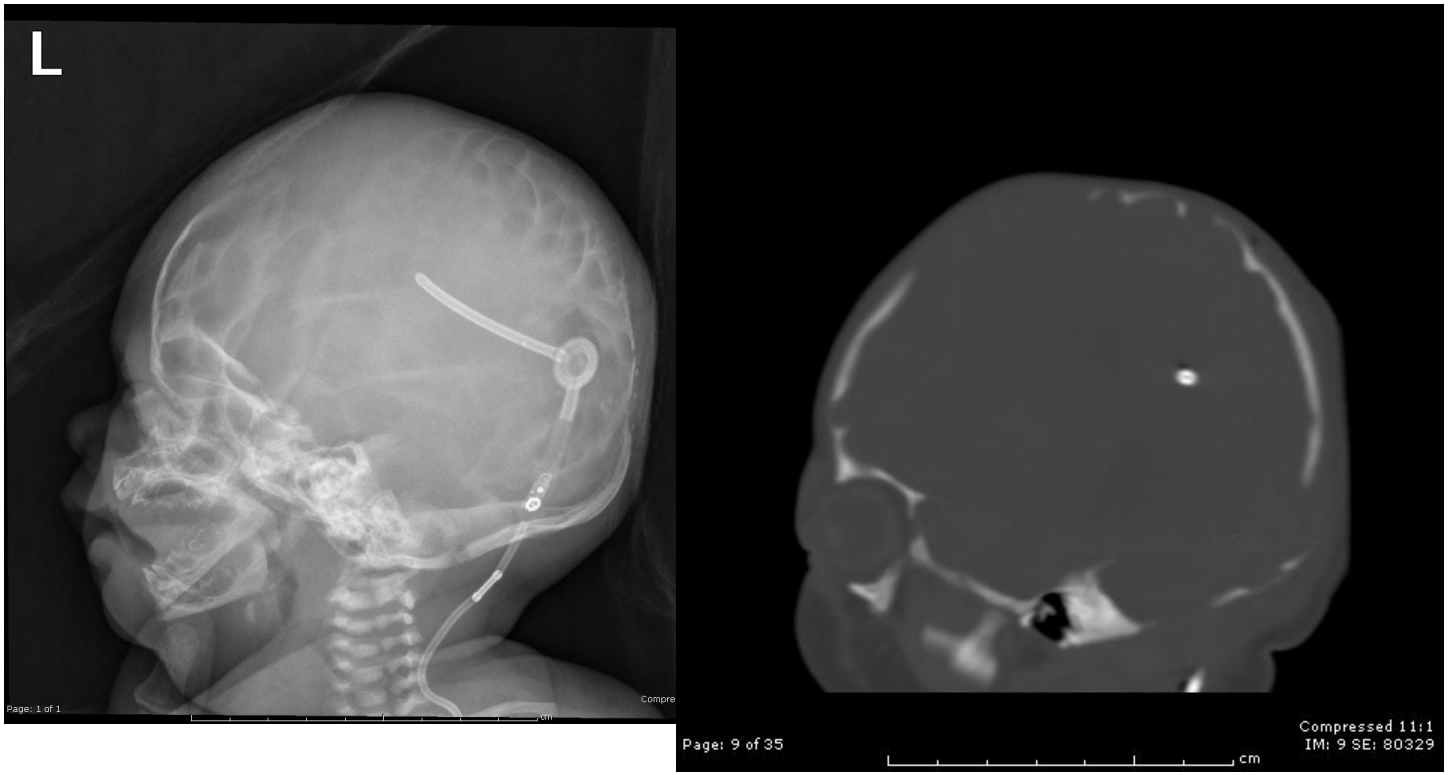

X-ray and sagittal CT showing severe thinned, copper-beaten skull consistent with the description of "Lukenschladel"

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Signs of Hydrocephalus:

- Excessive irritability
- Lethargy
- Frequent vomiting
- Splaying of the cranial sutures
- A bulging/tense fontanelle
- Prominence of the veins on the scalp
- Changes in eye movements (persistent downgaze)
- Progressive enlargement of the head (out of proportion to the rest of the child's growth)

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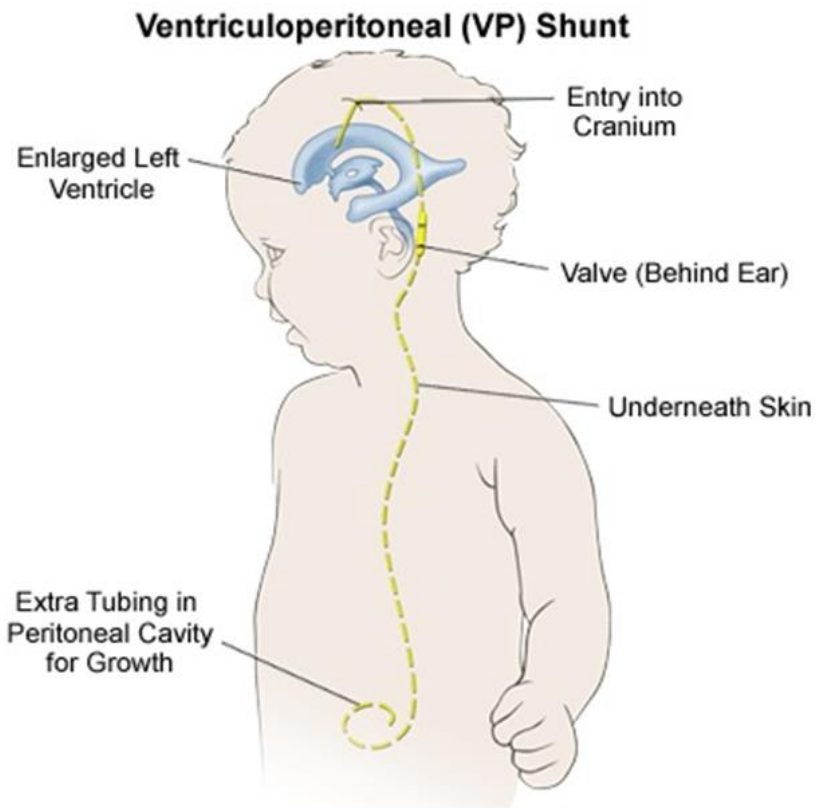
Shunting:

- Proximal catheter (into CSF space)
- Valve
- Distal catheter
  - Peritoneum
  - Right Atrium
  - Pleural space
  - Bone marrow
  - Gallbladder
  - Transverse sinus



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"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."

Harold Rekate M.D.

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Cerebrospinal Fluid Shunt

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Which shunt valve is the best?

Delta Valve

Orbis-Sigma Valve

Differential Pressure Valve:
(Hakim)

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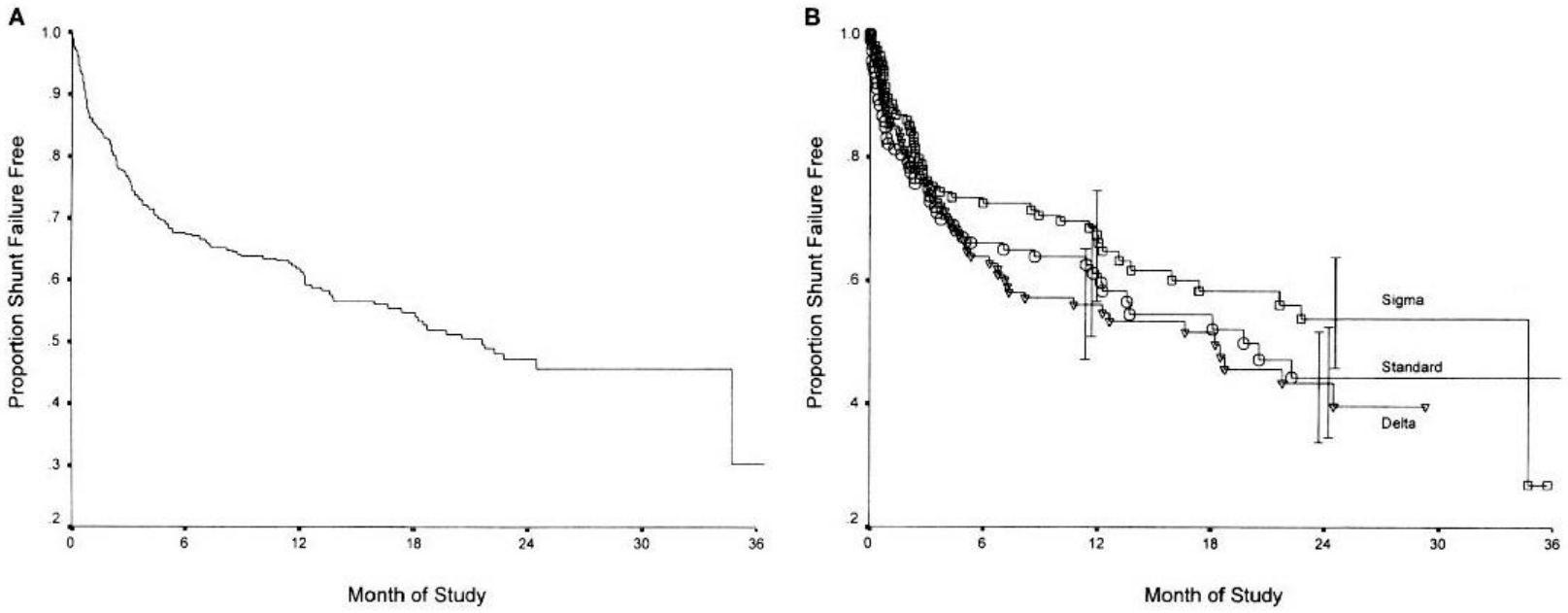
Results:



- 1 year failure rate: 39%
- 2 year failure rate: 53%
- Infection rate: 8.1%
- There was no difference between valves

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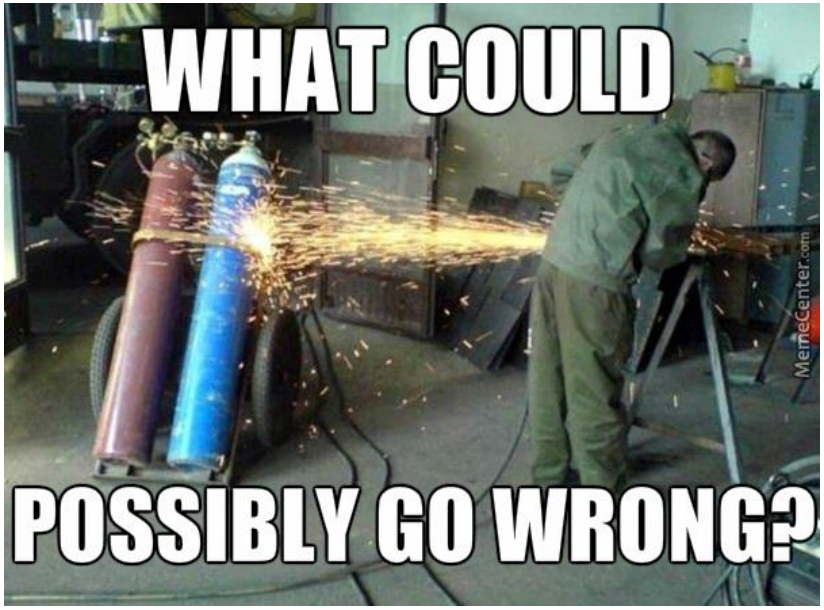
50% revision rate in 2 years!



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Intelligence:

- 80% of children with spina bifida will have normal intelligence (older data)
  - Some correlation to extent of ventriculomegaly on prenatal studies
- However, if the CNS gets infected, only 31% will have normal intelligence

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Original Paper

Pediatric Neurosurgery

Pediatr Neurosurg 2000;33:230–236

Received: July 31, 2000
Accepted: September 13, 2000

Long-Term Follow-Up Data from the Shunt Design Trial

J. Kestle[a] J. Drake[b] R. Milner[h] C. Sainte-Rose[c] G. Cinalli[c] F. Boop[d] J. Piatt[e] S. Haines[f] S. Schiff[g] D. Cochrane[h] P. Steinbok[h] N. MacNeil[h] for the collaborators

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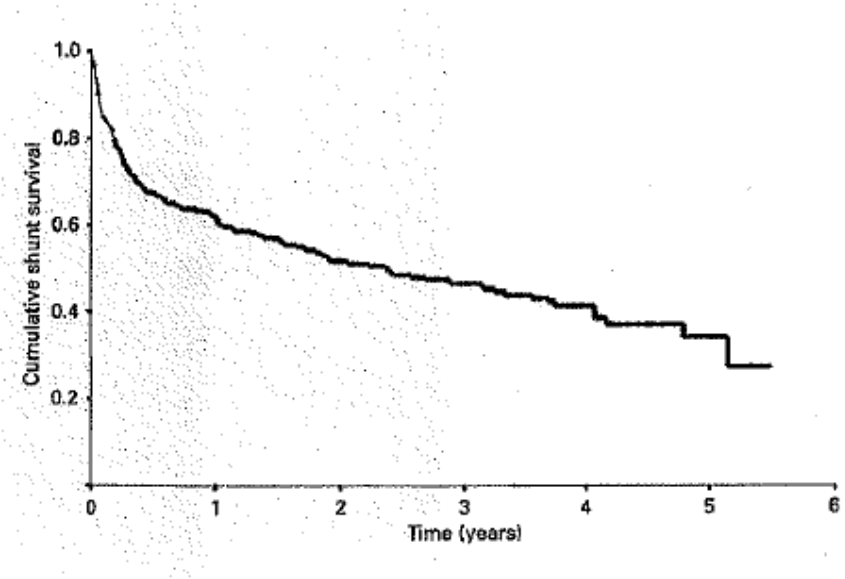

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38 y.o. spina bifida patient with severe headaches and frequent ER visits

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Childs Nerv Syst. 2013 Sep;29(9):1569-79. doi: 10.1007/s00381-013-2179-4. Epub 2013 Sep 7.

Myelomeningocele: the management of the associated hydrocephalus.

Tamburrini G[1], Frassanito P, Iakovaki K, Pignotti F, Rendeli C, Murolo D, Di Rocco C.

- Review of strategies and options
- Interesting points:
  - Overall rate of shunting is dropping
  - Etiology of hydrocephalus is complex – components of both "communicating" and "obstructive"
  - Complications/death are higher in spina bifida patients than other shunted children
  - Argued for delay in shunt placement after MMC closure

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Are there options other than shunting?

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Endoscopic Third Ventriculostomy

- Minimally invasive way to connect the ventricular system to the cisternal subarachnoid space
- Avoids the problems of shunt infection/malfunction
- First described by Dandy in 1922

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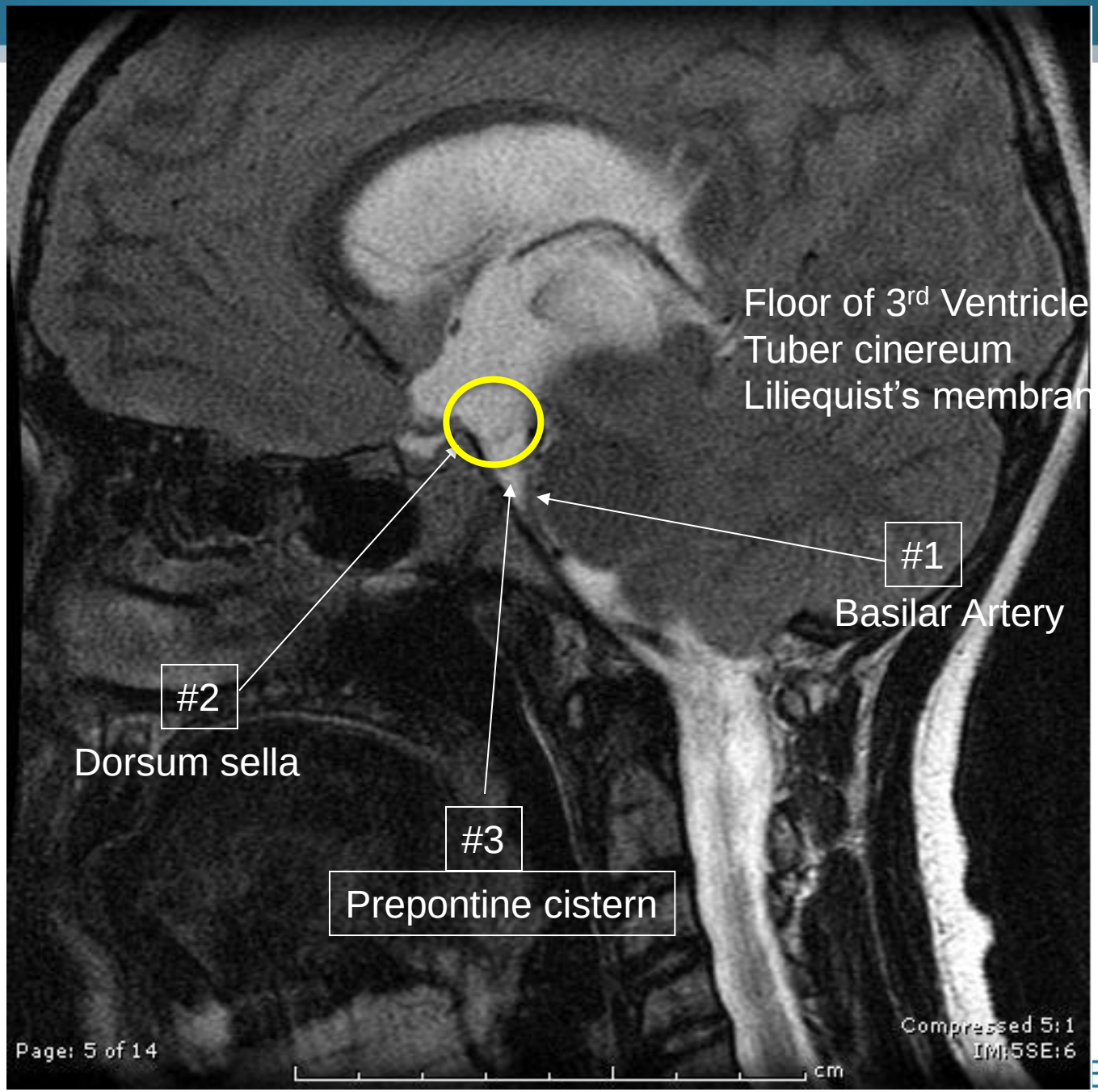

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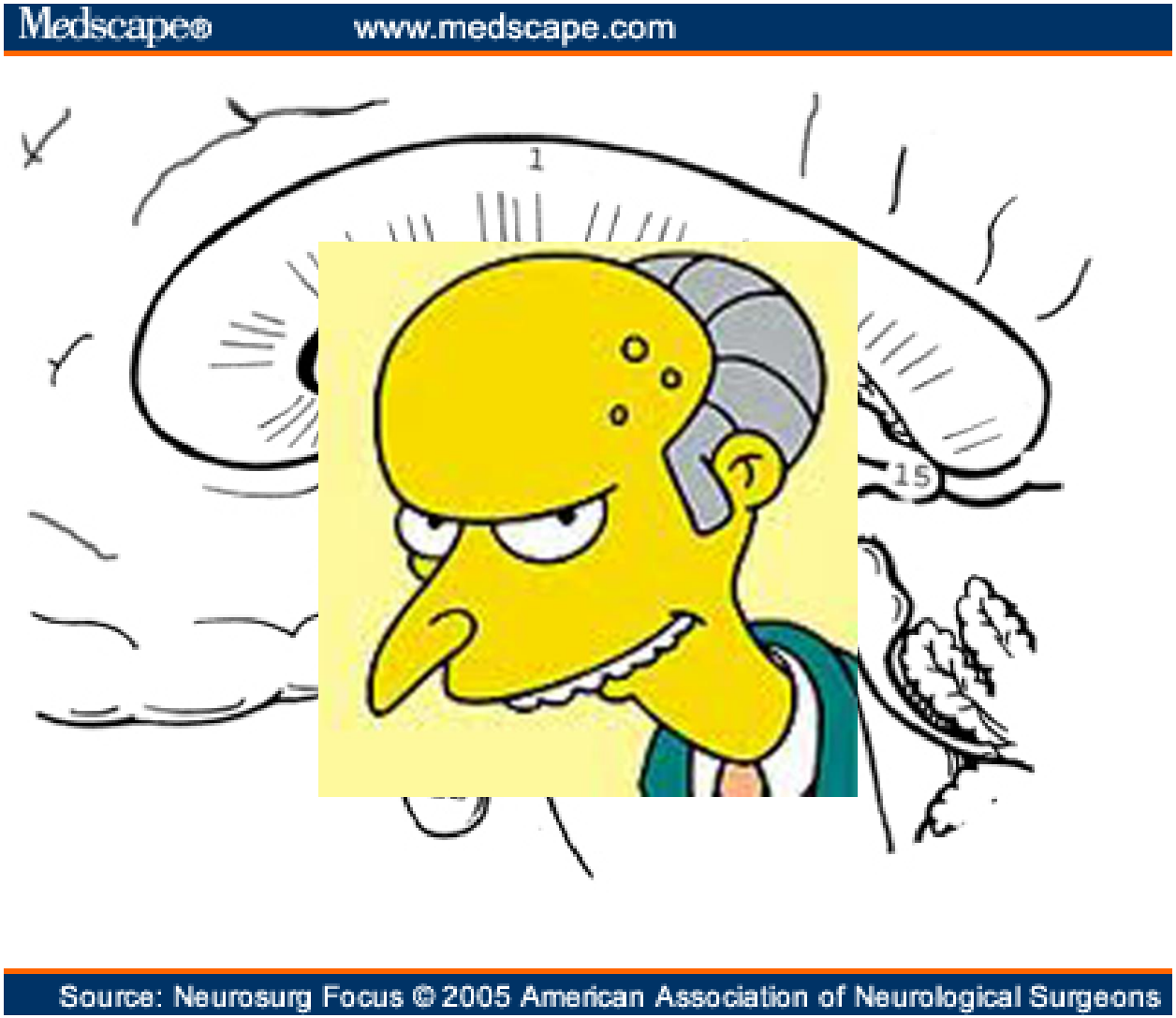

1.) Corpus Callosum

2.) Septum Pellucidum

3.) Fornix

4.) Foramen of Monro

5.) Anterior commisure

6.) Massa intermedia

7.) Choroid plexus

8.) Lamina terminalis

9.) Optic chiasm

10.) Optic recess

11.) Infundibular recess

12.) Pituitary

13.) Tuber cinereum

14.) Mamillary body

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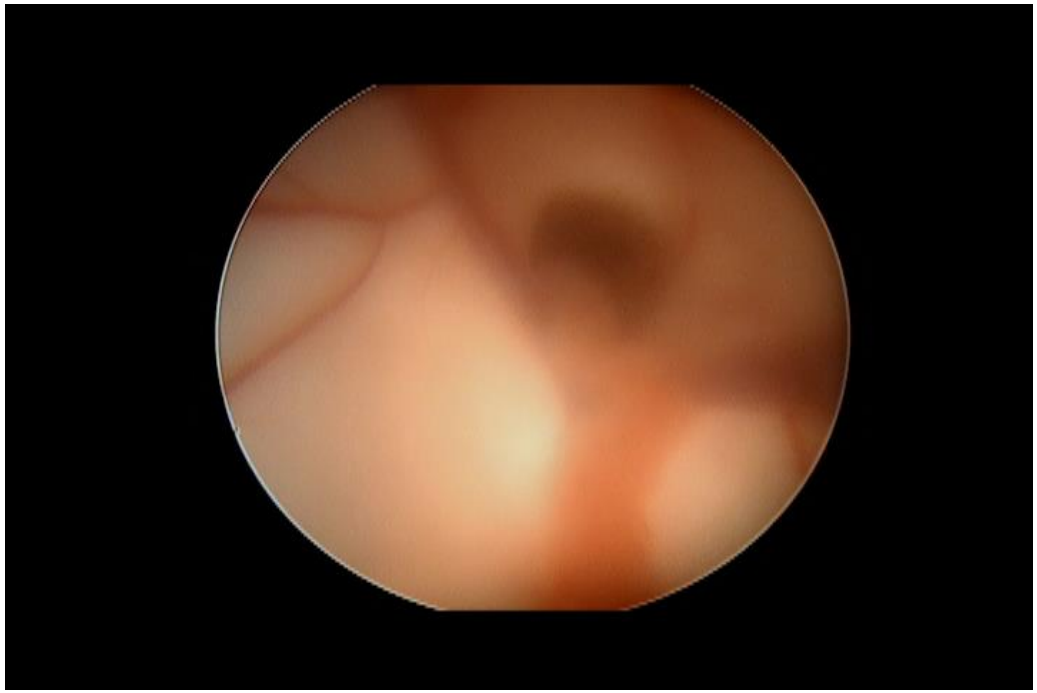
ETV:



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ETV SUCCESS SCORE

= Age Score + Etiology Score + Previous Shunt Score
≈ percentage probability of ETV success

SCORE	AGE ↓	ETIOLOGY ↓	PREVIOUS SHUNT ↓
0	<1 MONTH	POST-INFECTIOUS	PREVIOUS SHUNT
10	1 MONTH TO <6 MONTHS		NO PREVIOUS SHUNT
20		MYELOMENINGOCELE INTRA-VENTRICULAR HEMORRHAGE NON-TECTAL BRAIN TUMOR	
30	6 MONTHS TO <1 YEAR	AQUEDUCTAL STENOSIS TECTAL TUMOR OTHER ETIOLOGY	
40	1 YEAR TO <10 YEARS		
50	≥10 YEARS		

Endoscopic Third Ventriculostomy in the Treatment of Childhood Hydrocephalus

Abhaya V. Kulkarni, PhD, James M. Drake, FRCSC, Conor L. Mallucci, FRCS(SN), Spyros Sgouros, FRCS(SN), Jonathan Roth, MD, and Shlomi Constantini, MD, MSc, for the Canadian Pediatric Neurosurgery Study Group*

(J Pediatr 2009;155:254-9).

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Patients:

- Ideal candidate: older patient (> 2 y.o.) with stenosis or obstructing lesion around the aqueduct
  - Success rate: 90%
- Spina bifida with favorable anatomy
  - Success: 50%
- Chiari and hydrocephalus
- Tumor

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Unfavorable characteristics:

- Previous intraventricular hemorrhage
- Hydrocephalus from CNS infection
- Young age
  - 1 year?
  - 4 months?
  - 2 months?
- Anatomy:
  - Small lateral/third ventricle
  - Small prepontine cistern
  - Large massa intermedia

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Complications:

- Mortality: 1%
  - Basilar artery injury
- Neurological deficits:
  - Confusion
  - Oculomotor palsy
  - Diabetes insipidus
  - Seizure
  - Hypothalamic injury
- Infection
- CSF leak
- Failure: return of hydrocephalus

Personal opinion based on experience:

The complication rate is under-reported

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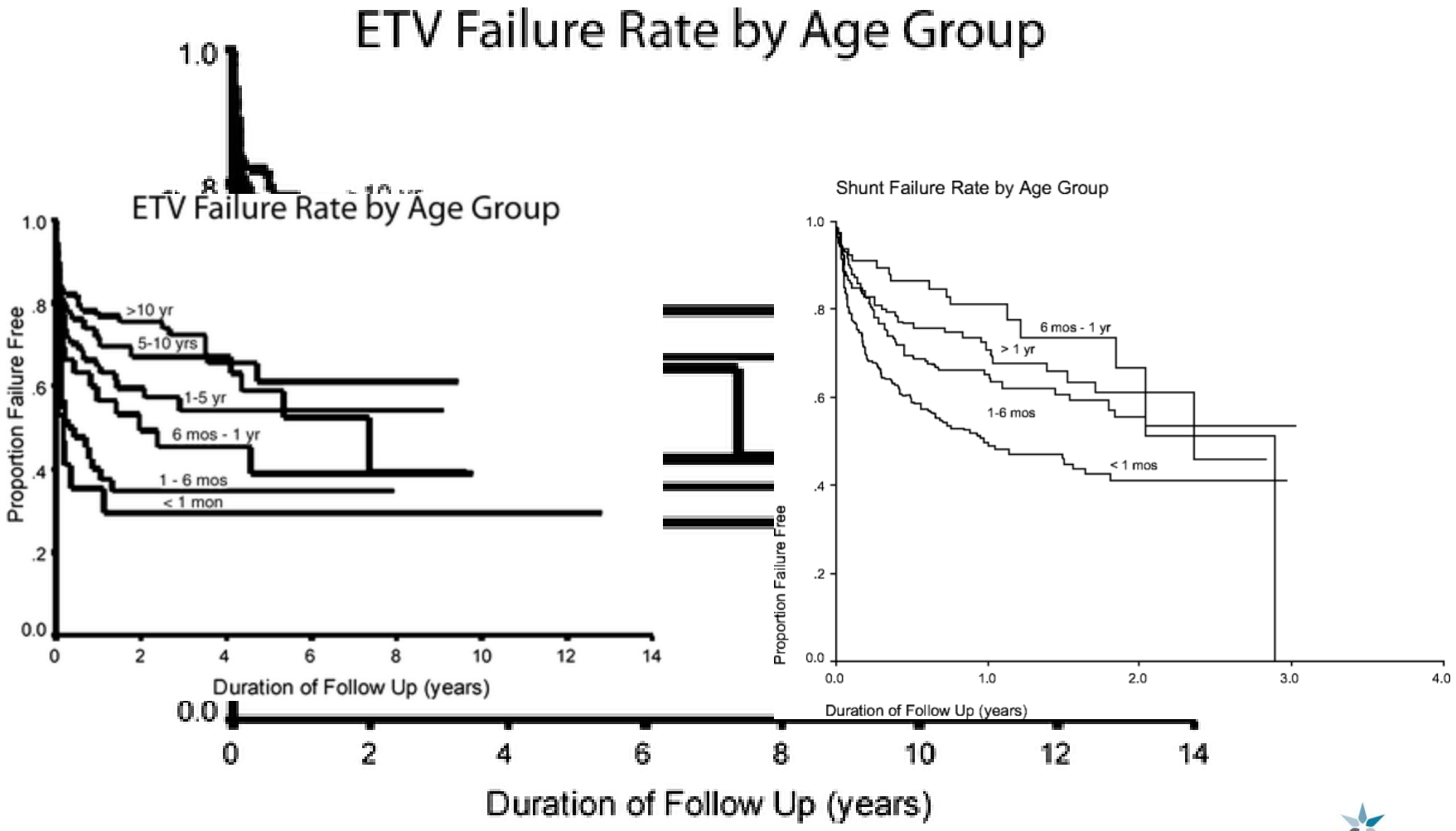
Endoscopic third ventriculostomy versus ventriculoperitoneal shunt in pediatric patients: a decision analysis

James M. Drake[1, 3] , Abhaya V. Kulkarni[1] and John Kestle[2]



Child's Nervous System© Springer-Verlag 200810.1007/s00381-008-0761-y

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ETV with choroid plexus cauterization:

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Cool! Does it work?

- Prospective study of 118 patients treated with ETV/CPC in Hydrocephalus Clinical Research Network centers
- Compared to their historical data: ETV or shunt
- 6 month "success" rate was 36%
  - Lower than shunting
  - ETV+CPC results were basically equivalent to ETV alone

Endoscopic third ventriculostomy and choroid plexus cauterization in infant hydrocephalus: a prospective study by the Hydrocephalus Clinical Research Network

Abhaya V. Kulkarni, MD, PhD,[1] Jay Riva-Cambrin, MD, MSc,[2] Curtis J. Rozzelle, MD,[3] Robert P. Naftel, MD,[4] Jessica S. Alvey, MSc,[5] Ron W. Reeder, PhD,[5] Richard Holubkov, PhD,[5] Samuel R. Browd, MD, PhD,[6] D. Douglas Cochrane, MD,[1] David D. Limbrick Jr., MD, PhD,[7] Tamara D. Simon, MD, MSPH,[8] Mandeep Tamber, MD, PhD,[9] John C. Wellons III, MD, MSPH,[4] William E. Whitehead, MD,[10] and John R. W. Kestle, MD, MSc,[11] for the Hydrocephalus Clinical Research Network

J Neurosurg Pediatr December 15, 2017

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Data from Uganda: (Dr. Warf)

- Addition of CPC to ETV increased the success rate from 47% to 66%
- With this procedure, shunt placement was avoided in:
  - Post-infectious hydrocephalus: 63%
  - Spina bifida: 76%
  - Dandy-Walker: 69%
  - Aqueductal Stenosis: 82%
  - Idiopathic: 72%

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Discussion Questions:

- What is an acceptable "failure rate"?
- What is an acceptable complication rate?
- Can the results from Africa really be applied to the United States?
  - What factors may cause a difference?

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A Randomized Trial of Prenatal versus Postnatal Repair of Myelomeningocele

N. Scott Adzick, M.D., Elizabeth A. Thom, Ph.D., Catherine Y. Spong, M.D., John W. Brock, III, M.D., Pamela K. Burrows, M.S., Mark P. Johnson, M.D., Lori J. Howell, R.N., M.S., Jody A. Farrell, R.N., M.S.N., Mary E. Dabrowiak, R.N., M.S.N., Leslie N. Sutton, M.D., Nalin Gupta, M.D., Ph.D., Noel B. Tulipan, M.D., Mary E. D'Alton, M.D., and Diana L. Farmer, M.D. for the MOMS Investigators*

March 17, 2011
N Engl J Med 2011; 364:993-1004
DOI: 10.1056/NEJMoa1014379

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MOMs Trial

- Stopped for efficacy!
- 183 patients
  - Decreased rate of shunt placement (40% versus 82%)
  - Decreased the appearance of Chiari II malformation
  - Increased risk of prematurity

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Further study of MOMs patients:

- Degree of prenatal hydrocephalus correlates to need for shunt
  - Ventricle size < 10 mm: 20% needed shunt
  - Ventricle size 10-15 mm: 45%
  - Ventricle size > 15 mm: 79%

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Discussion questions:

- Inclusion criteria
- Should everybody do this?

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Other causes of hydrocephalus:

- Congenital
- Premature babies with intraventricular hemorrhage

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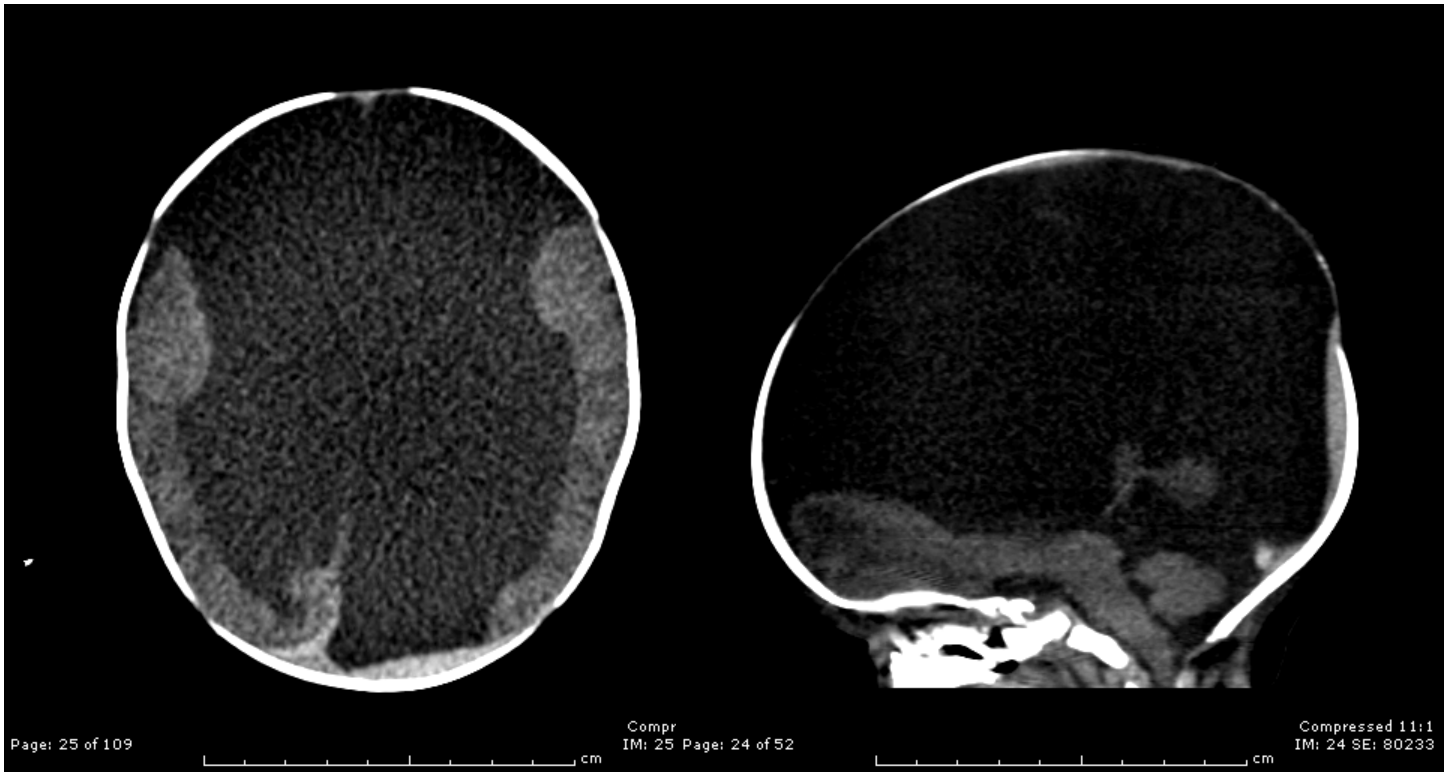

CT scans in axial and sagittal view. Note the severe lack of cerebral cortex with a relatively well developed brainstem and intact posterior fossa structures. The etiology of the condition is unknown, but the distribution suggests early vascular injury of the internal carotid distribution bilaterally.

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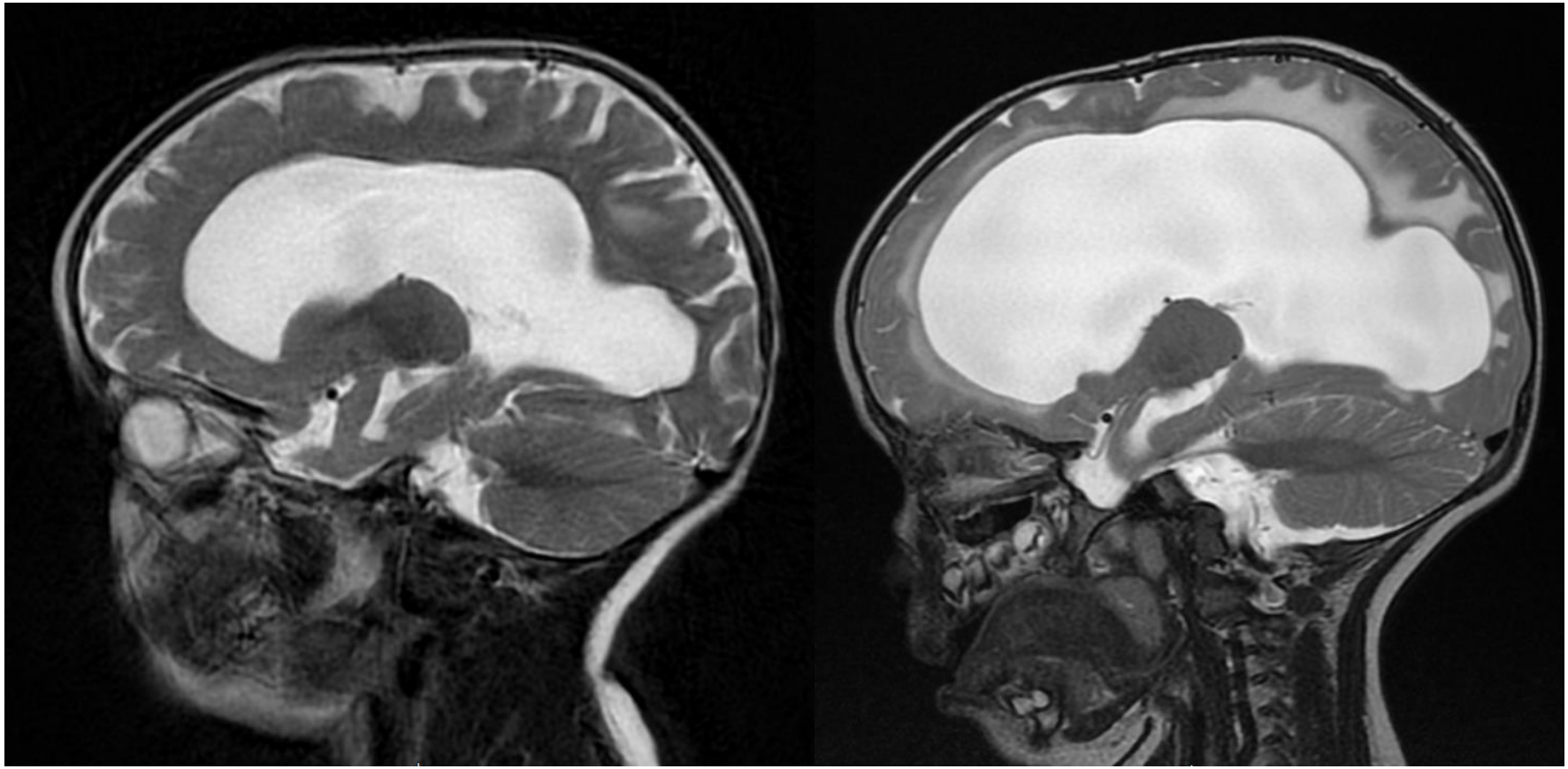
Our first ETV patient in South Bend (and TV star!)

After:

Before:



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Premature babies with intraventricular hemorrhage and hydrocephalus

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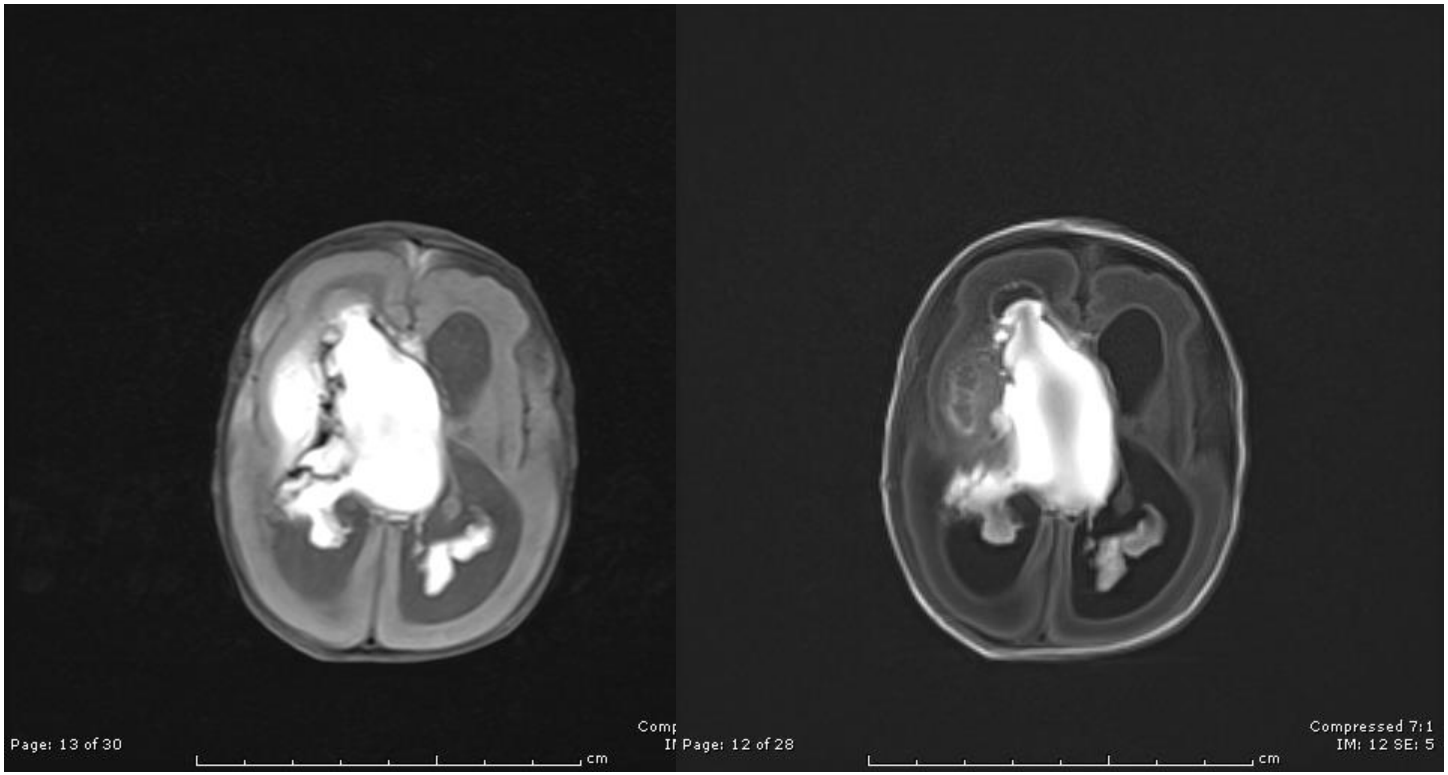

Premature infant with Grade IV intraventricular hemorrhage. Left: FLAIR MRI. Right: T1 MRI. Note the lack of sulci related to age. Large clot extending throughout right lateral ventricle and into the right basal ganglia.

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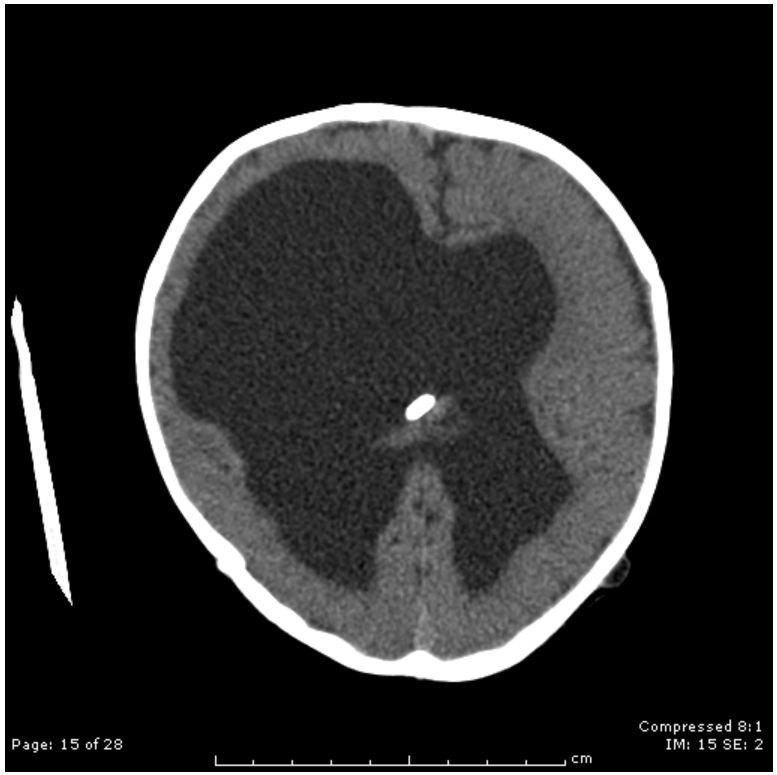

CT scan at 17 months of age showing reduced ventricle size, expansion of the left cortex, with continued loss of right frontal cortex. Shunt catheter seen at midline.

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JNS PEDIATRICS

CLINICAL ARTICLE

J Neurosurg Pediatr 20:19–29, 2017

Shunting outcomes in posthemorrhagic hydrocephalus: results of a Hydrocephalus Clinical Research Network prospective cohort study

John C. Wellons III, MD, MSPH,[1] Chevis N. Shannon, MPH, MBA, DrPH,[1] Richard Holubkov, PhD,[2] Jay Riva-Cambrin, MD, MSc,[3] Abhaya V. Kulkarni, MD, PhD,[4] David D. Limbrick Jr., MD, PhD,[5] William Whitehead, MD, MPH,[6] Samuel Browd, MD, PhD,[7] Curtis Rozzelle, MD,[8] Tamara D. Simon, MD, MSPH,[9] Mandeep S. Tamber, MD, PhD,[10] W. Jerry Oakes, MD,[8] James Drake, MBBCh, MSc,[4] Thomas G. Luerssen, MD,[6] and John Kestle, MD, MSc,[11] for the Hydrocephalus Clinical Research Network

J Neurosurg Pediatrics (Suppl) 14:8–23, 2014
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Pediatric hydrocephalus: systematic literature review and evidence-based guidelines. Part 2: Management of posthemorrhagic hydrocephalus in premature infants

Catherine A. Mazzola, M.D.,[1] Asim F. Choudhri, M.D.,[2,3] Kurtis I. Auguste, M.D.,[4] David D. Limbrick Jr., M.D., Ph.D.,[5] Marta Rogido, M.D.,[6] Laura Mitchell, M.A.,[7] and Ann Marie Flannery, M.D.[8]

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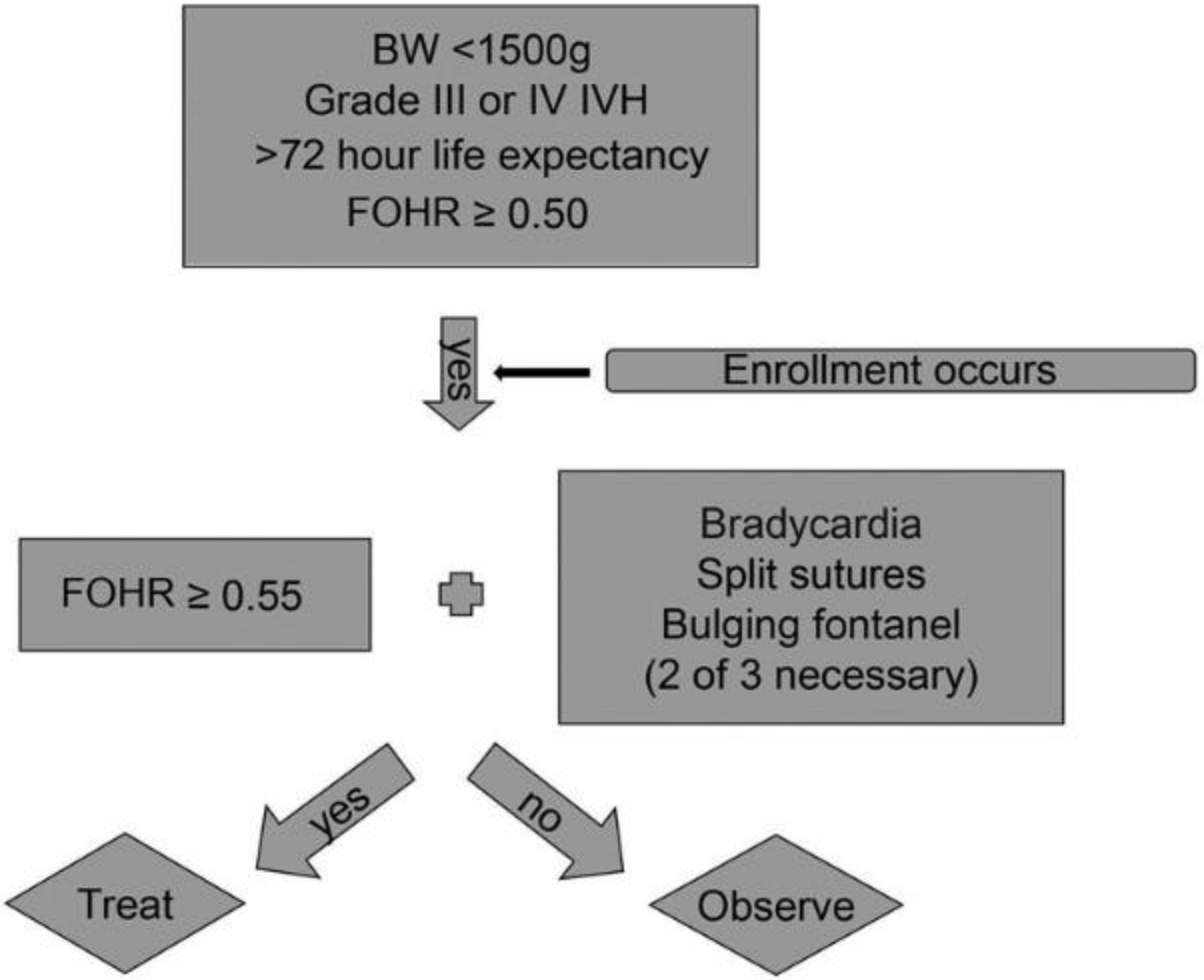

FIG. 1. Rubric illustrating the decision to treat. BW = body weight.

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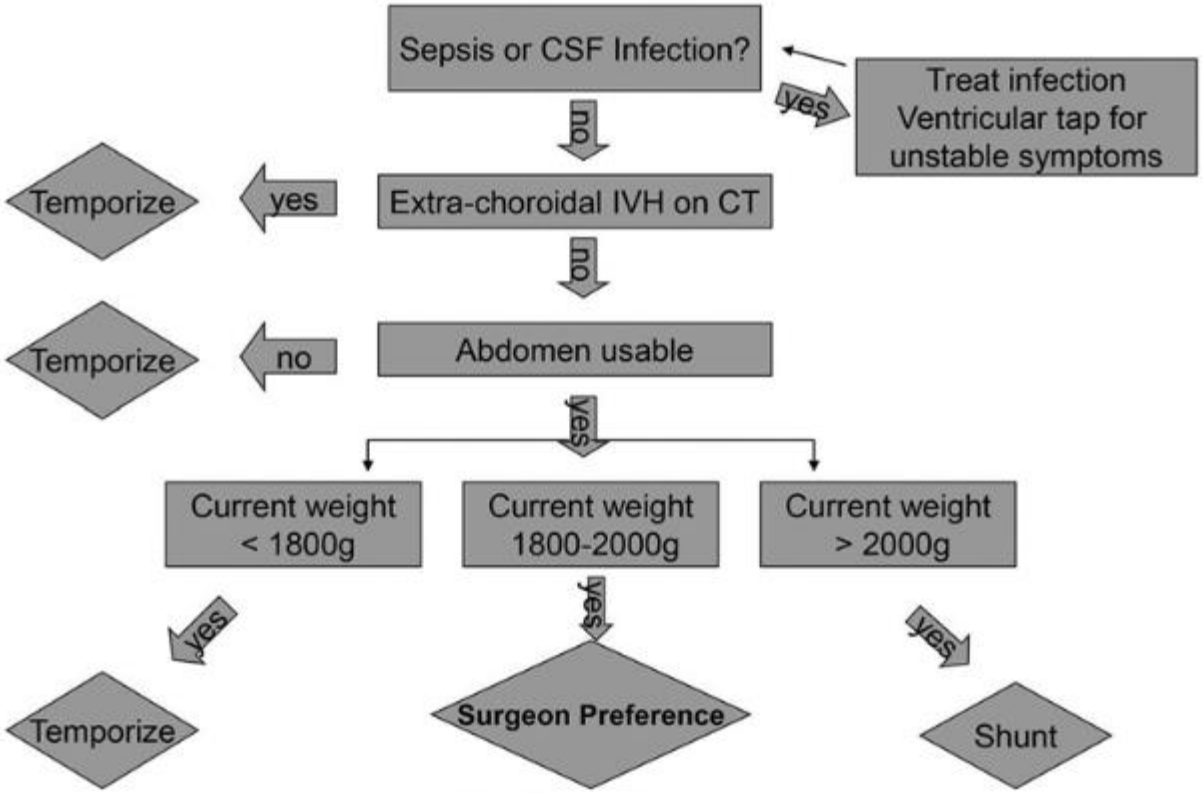

FIG. 2. Decision rubric regarding whether to temporize or place a shunt.

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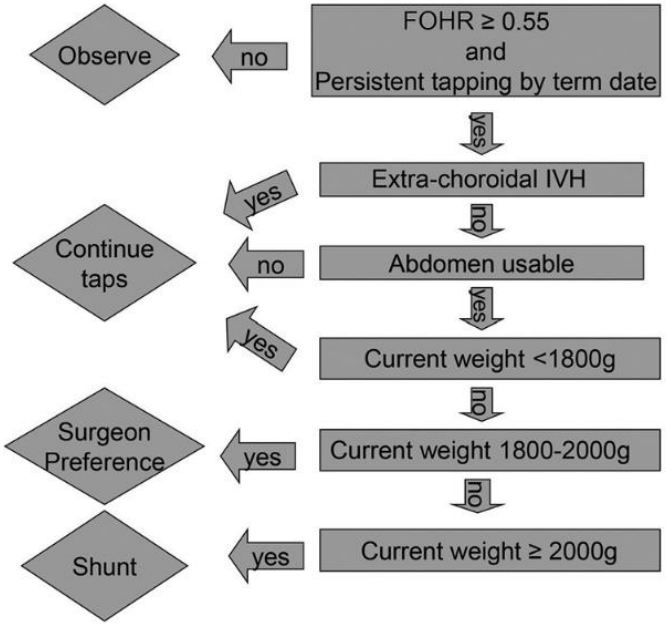

FIG. 3. Decision rubric illustrating whether to convert a VR.

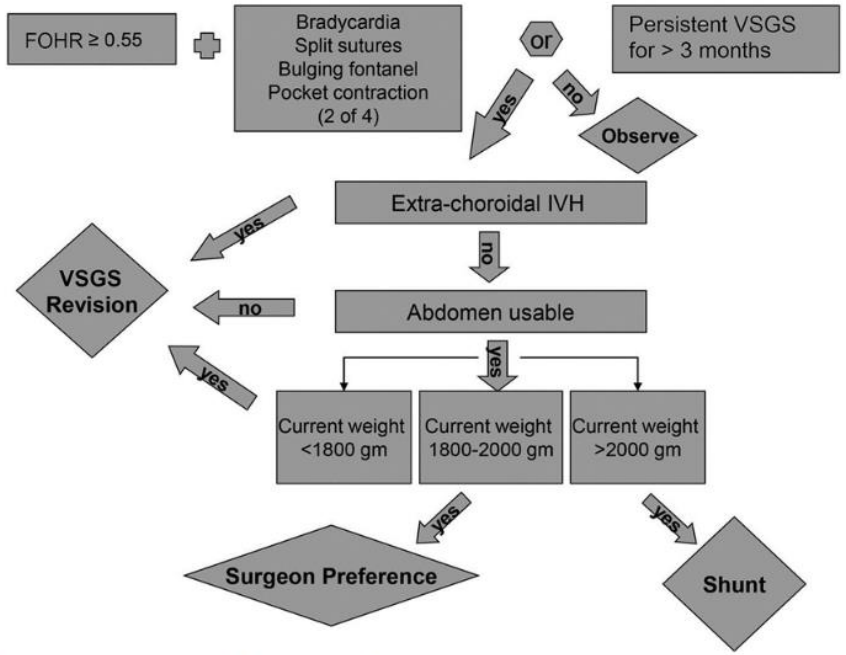

FIG. 4. Decision rubric regarding whether to convert a VSGS. gm = grams.

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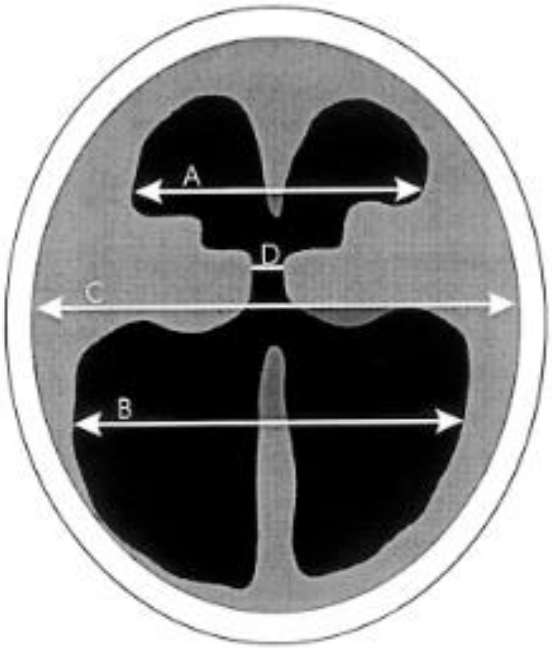

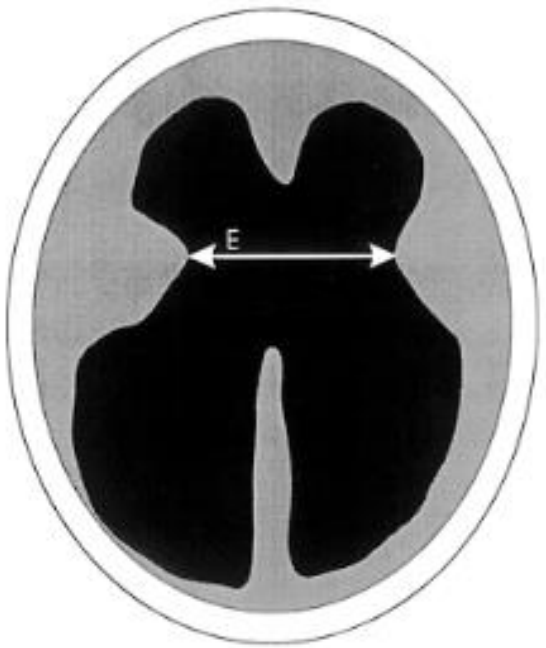

Evans' ratio = A/C
Frontal + occipital horn ratio = (A + B)/2C
Minimal lateral ventricular width = D
Huckman's measurement = A + D

Ventricular span at the body of the lateral ventricle = E
Ventricular brain ratio = area (■)/Area (▨+■)

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Literature review Summary:

- Level 1:
  - Thrombolytic agents are not recommended
  - Serial LP is not recommended to avoid progression of hydrocephalus
  - Acetazolamide and furosemide are not recommended
- Level 2:
  - Multiple options exist (VAD, VSG, LP, EVD)
  - VSG reduces need for tapping (DUH)
- Level 3:
  - No specific weight recommended
  - Role of ETV is uncertain

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Other notes:

- After a reservoir tap, replace volume cc/cc with NS (IV or enteral)
- Check daily OFC
- Check qweek head ultrasound
  - This stops once a definitive shunt is placed

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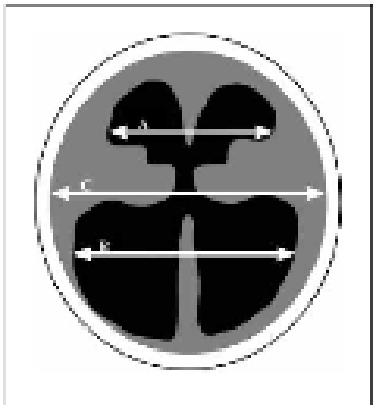
SOPHH Appendix B:
Protocol Specific Definitions:

1. Intraventricular Hemorrhage of Prematurity [10]

Grade I: Germinal Matrix Hemorrhage
Grade II: Germinal Matrix Hemorrhage with extension into the ventricular system without dilatation
Grade III: Germinal Matrix Hemorrhage with extension into the ventricular system with dilatation
Grade IV: Germinal Matrix Hemorrhage with extension into the ventricular system with dilatation and extension into the adjacent brain parenchyma

2. Frontal Occipital Horn Ratio (FOR) [36]



$$FOR = (A + B)/2C$$

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Prenatal Evaluation:

- Prenatal:
  - Counselling should be considered if hydrocephalus detected on ultrasound
  - Potential options for spina bifida patients if diagnosed early enough

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Conclusions:

- Spina bifida patients are complex and have many issues
- Preventing infection is paramount!
- There are options for treating hydrocephalus:
  - Shunt
  - ETV
  - ETV with CPC
- Prenatal closure provides definite benefit in some patients

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Conclusions (cont):

- Congenital hydrocephalus
- Premature babies
  - Generally temporize treatment until the child is old/big enough for a full shunt
  - Very high risk for problems!

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Questions?

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