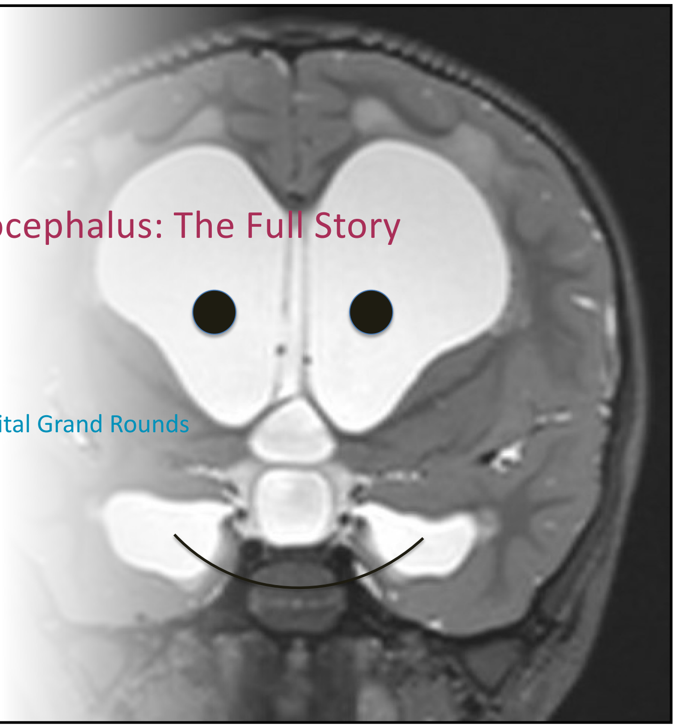
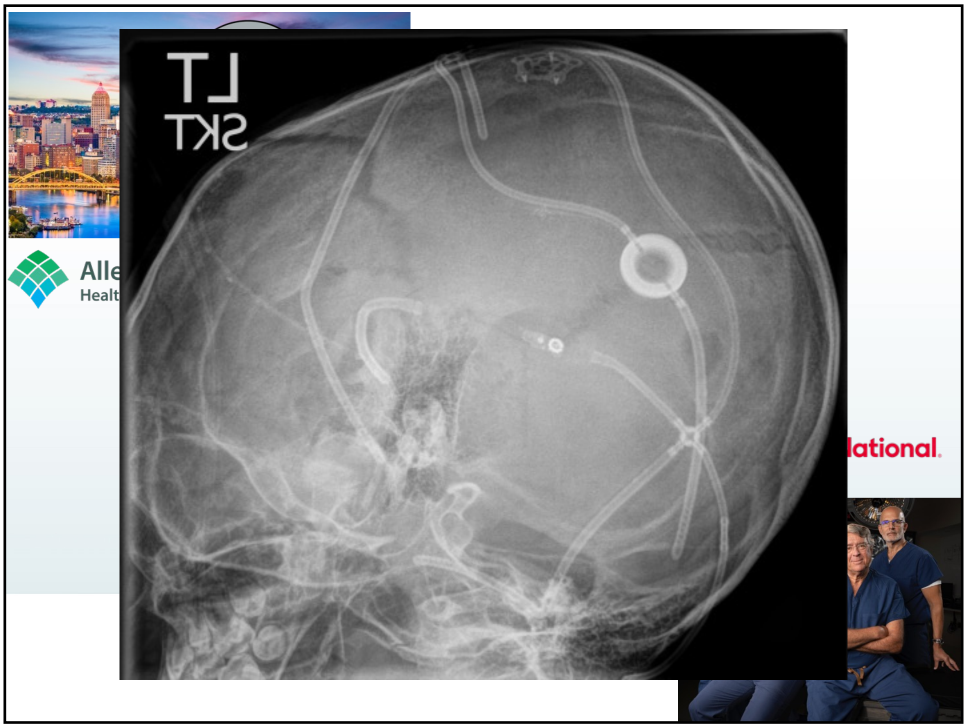
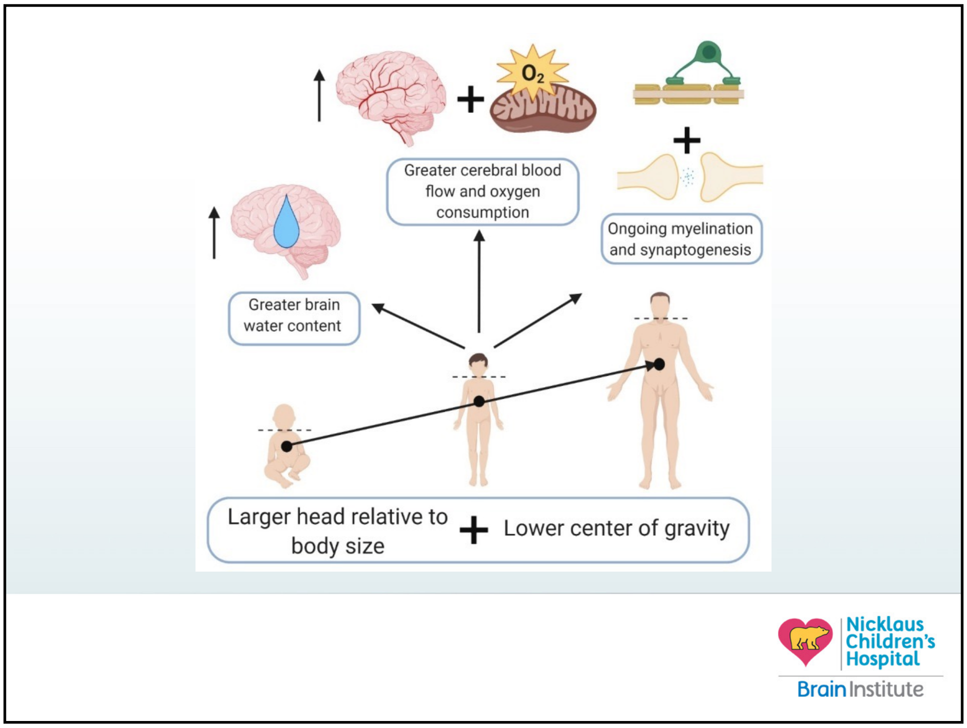


Pediatric Hydrocephalus: The Full Story (kind of…)

Kristen Stabingas, MD
Pediatric Neurosurgeon
Nicklaus Children's Hospital Grand Rounds
February 2, 2024





Greater cerebral blood flow and oxygen consumption

Ongoing myelination and synaptogenesis

Greater brain water content

Larger head relative to body size + Lower center of gravity

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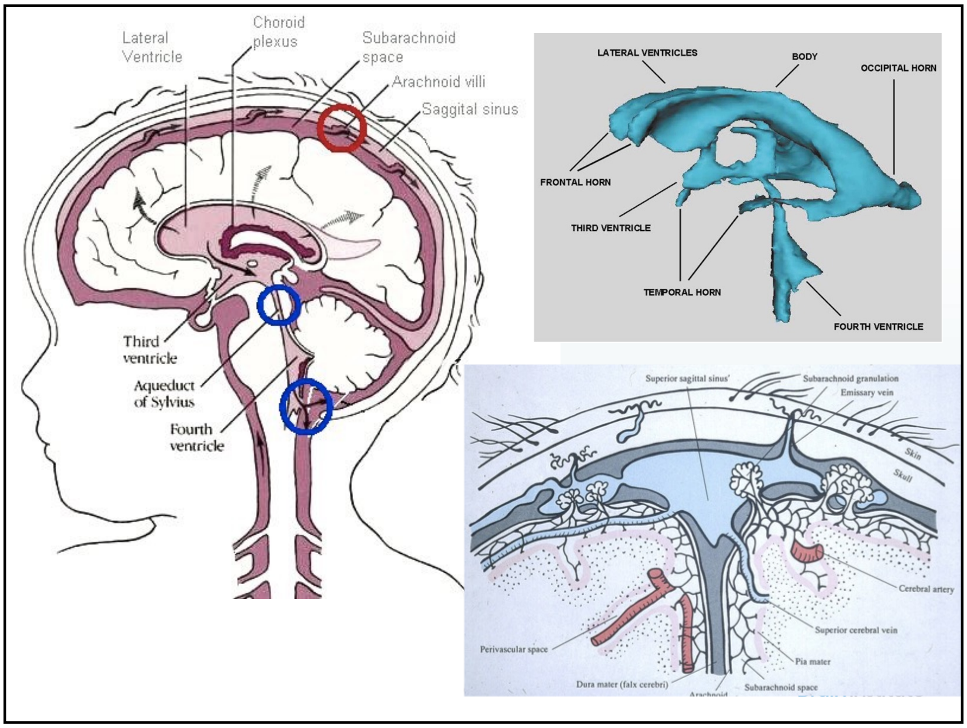

Intracranial Pressure

- ICP= SSP ~ formation of CSF~ R to CSF outflow
- Newborn ICP = 2-6 mmHg
- Child ICP = < 15 mmHg

- CSF Production= 0.15-0.30 cc/min ~ 450 cc/day
- Adult = 150 cc
- Neonate = 50 cc

Khasawneh et al., 2018

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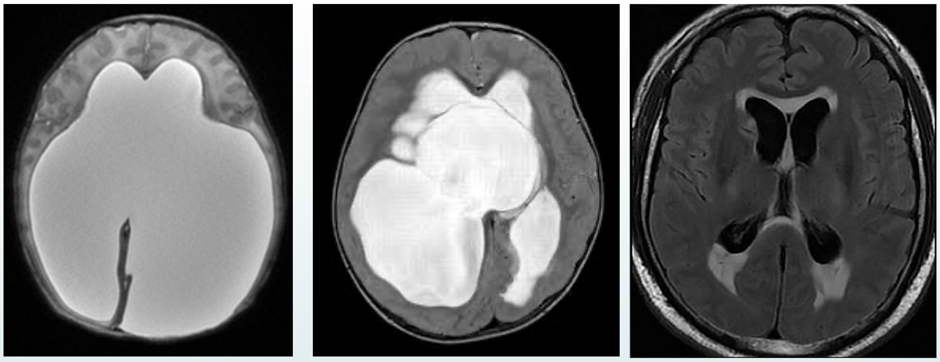
Brain Institute

Hydrocephalus

An imbalance of CSF production and absorption



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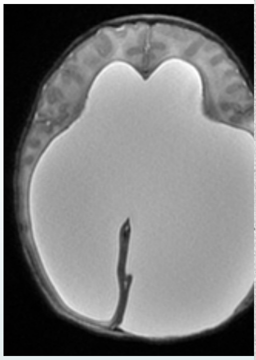
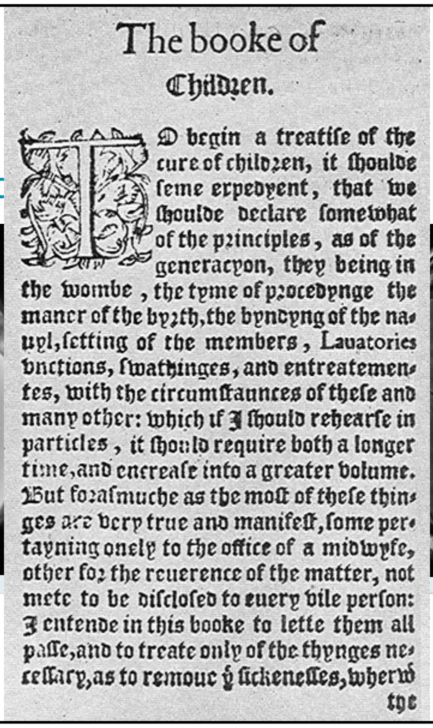
Brain Institute

7

An imbalanc





The booke of

Children.

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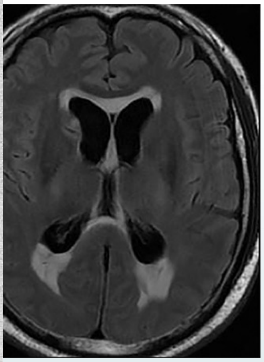
the

absorption



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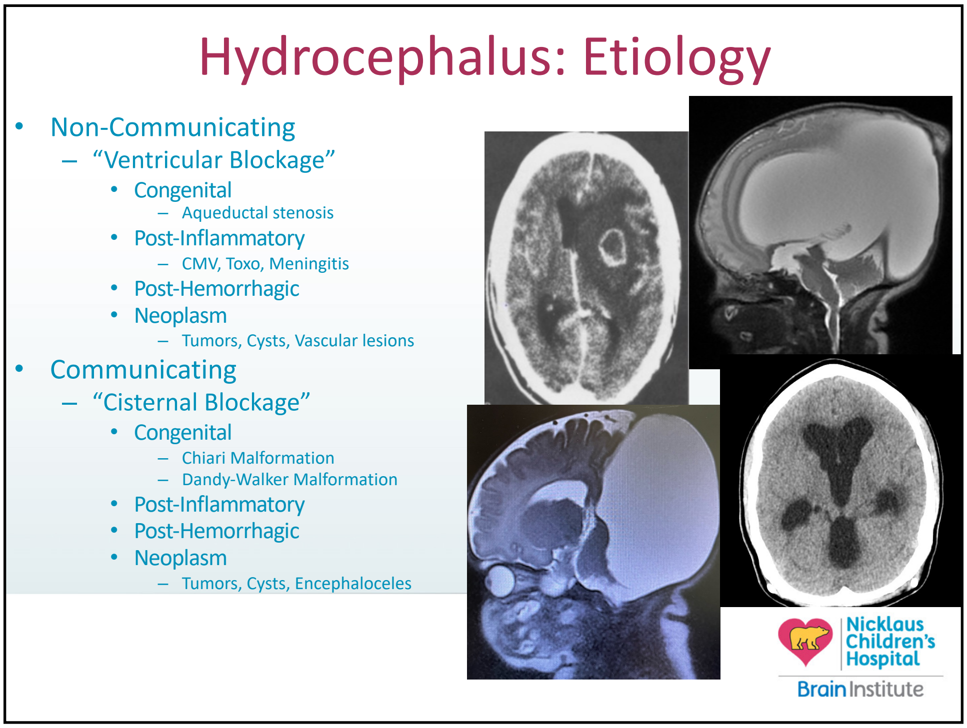
8



Hydrocephalus: Etiology

- Non-Communicating
  - “Ventricular Blockage”
    - Congenital
      - Aqueductal stenosis
    - Post-Inflammatory
      - CMV, Toxo, Meningitis
    - Post-Hemorrhagic
    - Neoplasm
      - Tumors, Cysts, Vascular lesions
- Communicating
  - “Cisternal Blockage”
    - Congenital
      - Chiari Malformation
      - Dandy-Walker Malformation
    - Post-Inflammatory
    - Post-Hemorrhagic
    - Neoplasm
      - Tumors, Cysts, Encephaloceles

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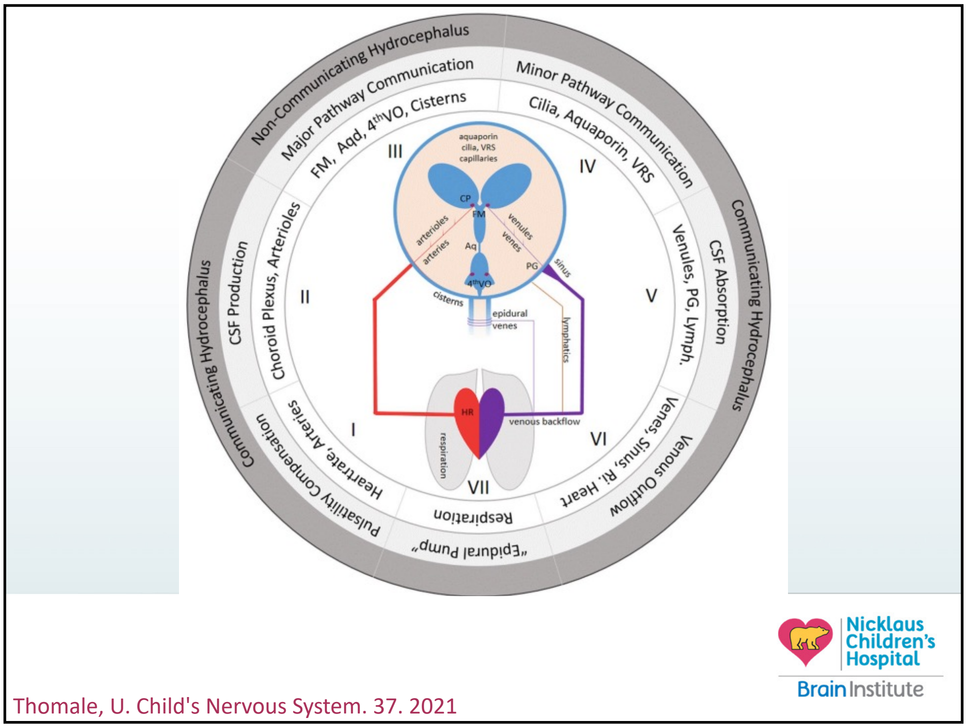

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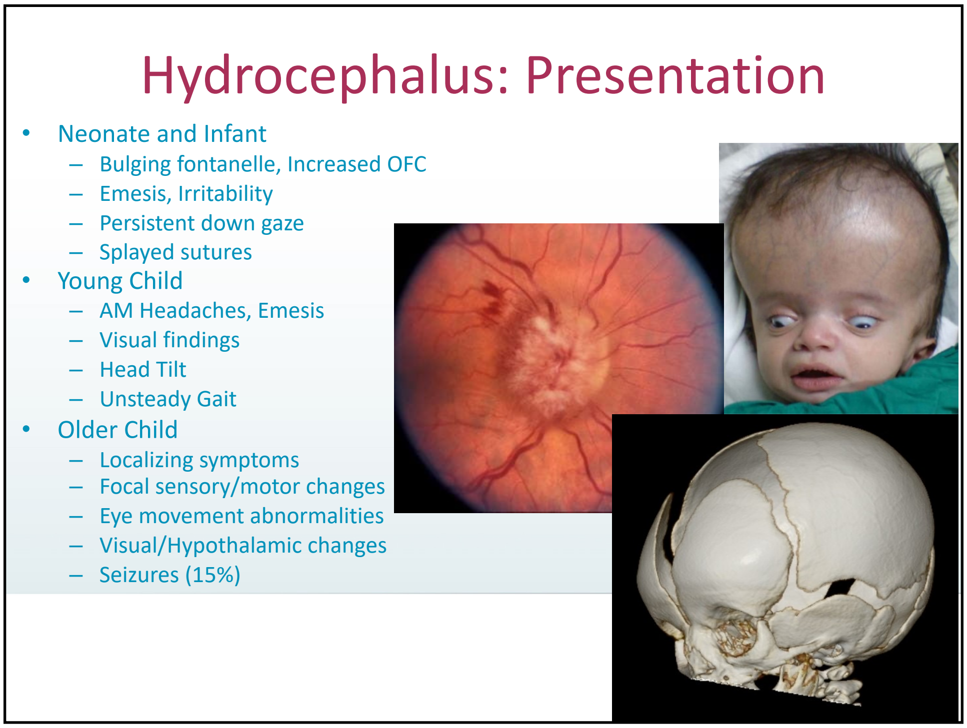
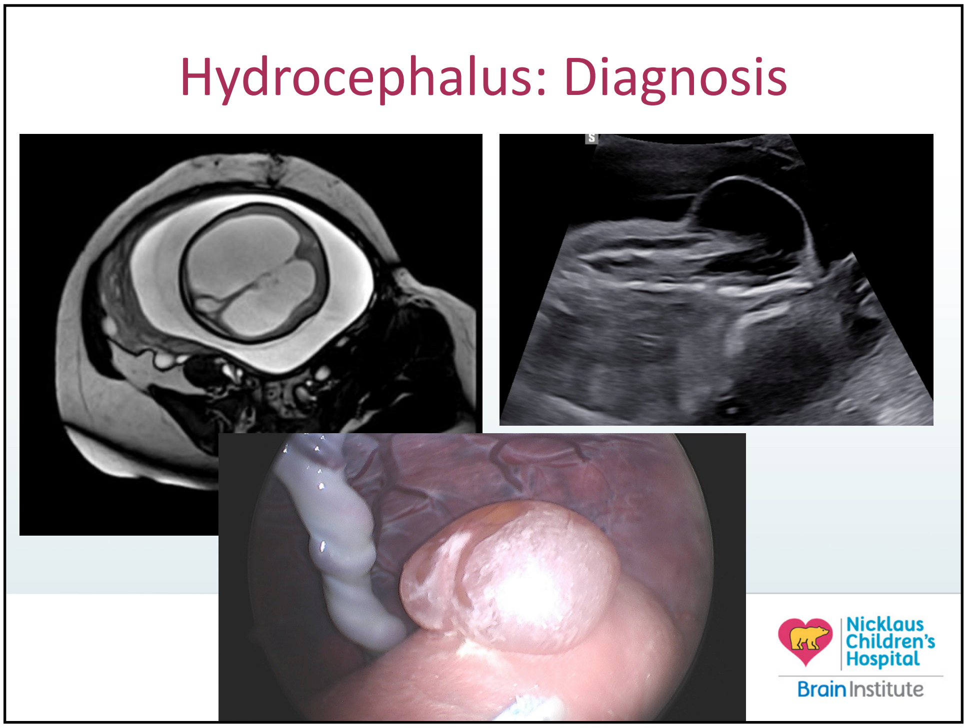
Thomale, U. Child's Nervous System. 37. 2021

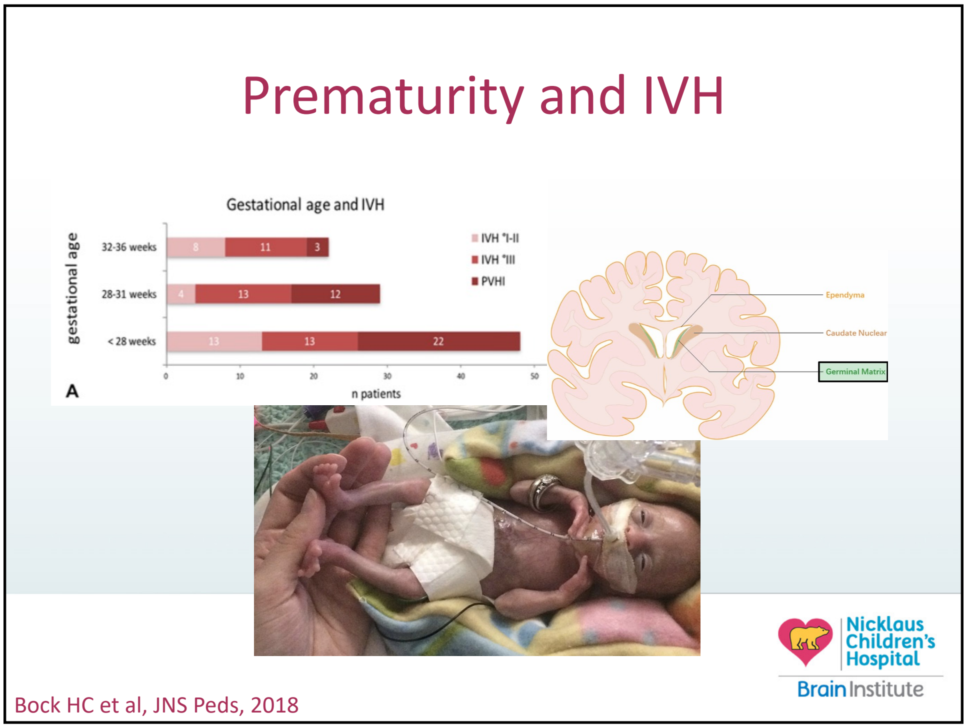


Hydrocephalus: Presentation

- Neonate and Infant
  - Bulging fontanelle, Increased OFC
  - Emesis, Irritability
  - Persistent down gaze
  - Splayed sutures
- Young Child
  - AM Headaches, Emesis
  - Visual findings
  - Head Tilt
  - Unsteady Gait
- Older Child
  - Localizing symptoms
  - Focal sensory/motor changes
  - Eye movement abnormalities
  - Visual/Hypothalamic changes
  - Seizures (15%)

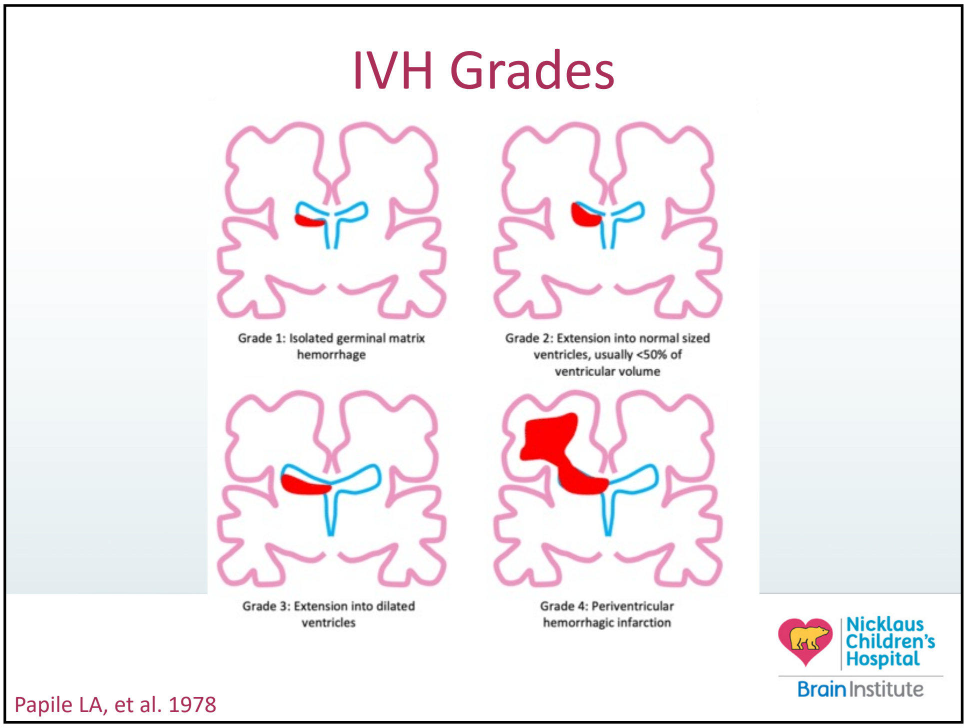


Hydrocephalus: Diagnosis



Prematurity and IVH

Bock HC et al, JNS Peds, 2018



IVH Grades

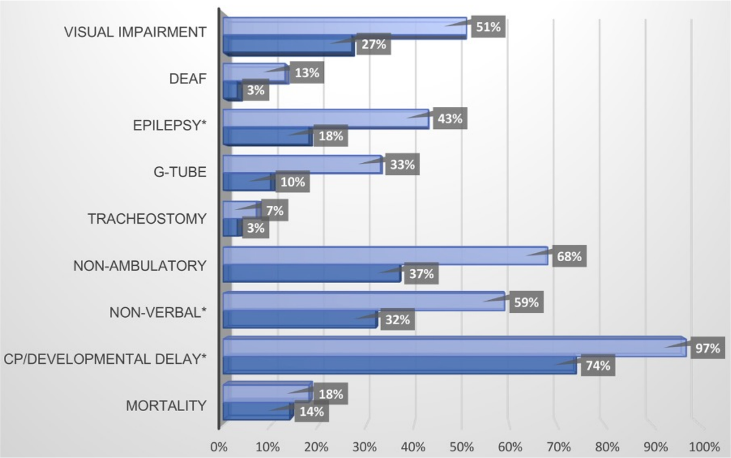
Grade 1: Isolated germinal matrix hemorrhage

Grade 2: Extension into normal sized ventricles, usually <50% of ventricular volume

Grade 3: Extension into dilated ventricles

Grade 4: Periventricular hemorrhagic infarction

Papile LA, et al. 1978



Comparison of functional outcomes and mortality in grade III (*dark blue bars*) versus grade IV (*light blue bars*) IVH patients. The x-axis represents the percentage of patients.

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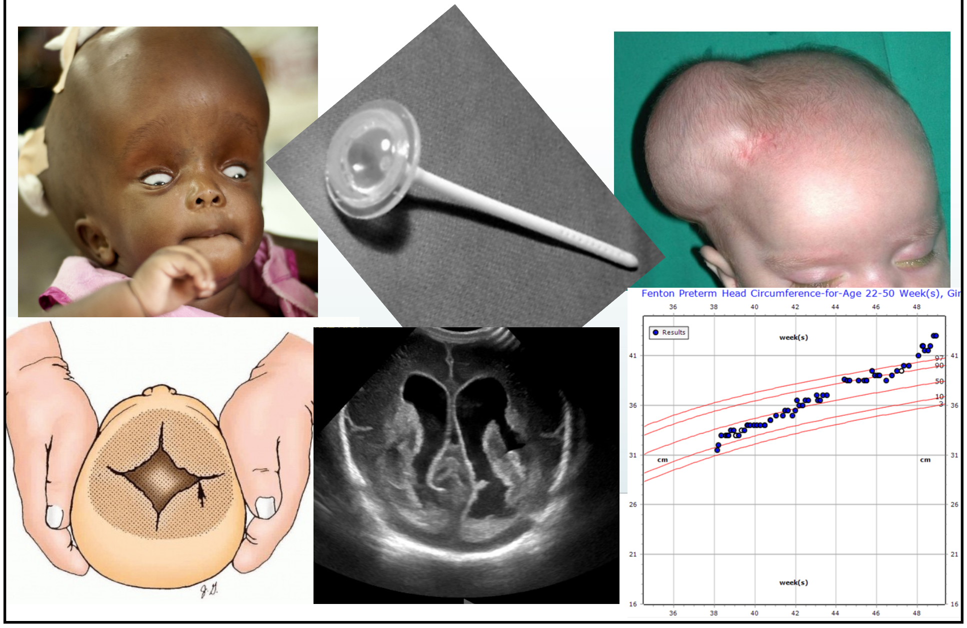
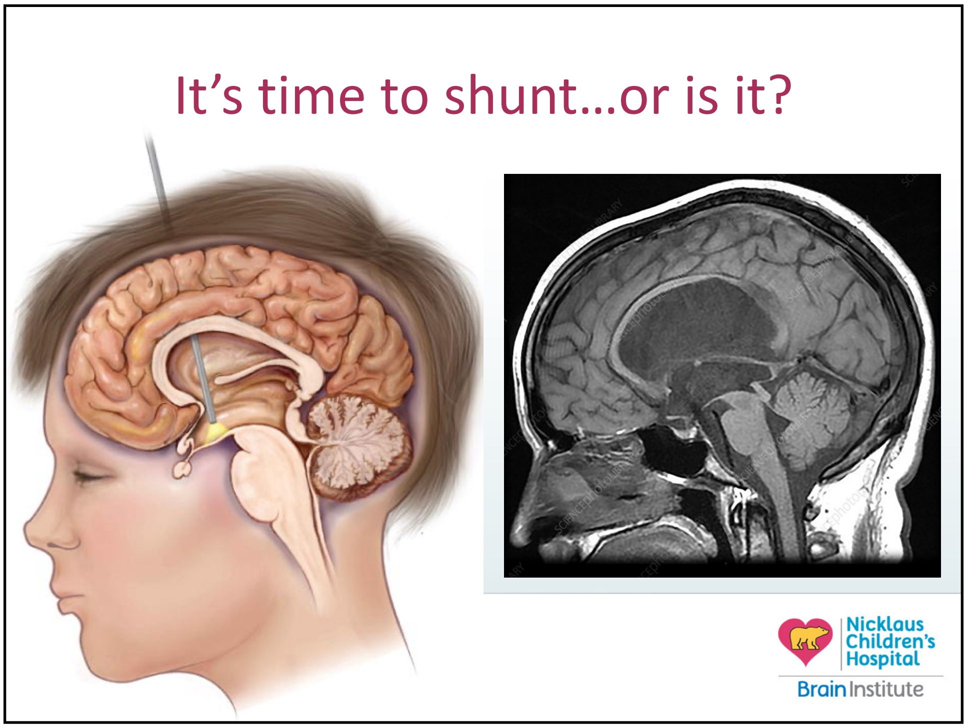
McClugage et al, 2020

Neurosurgical Intervention





It's time to shunt…or is it?

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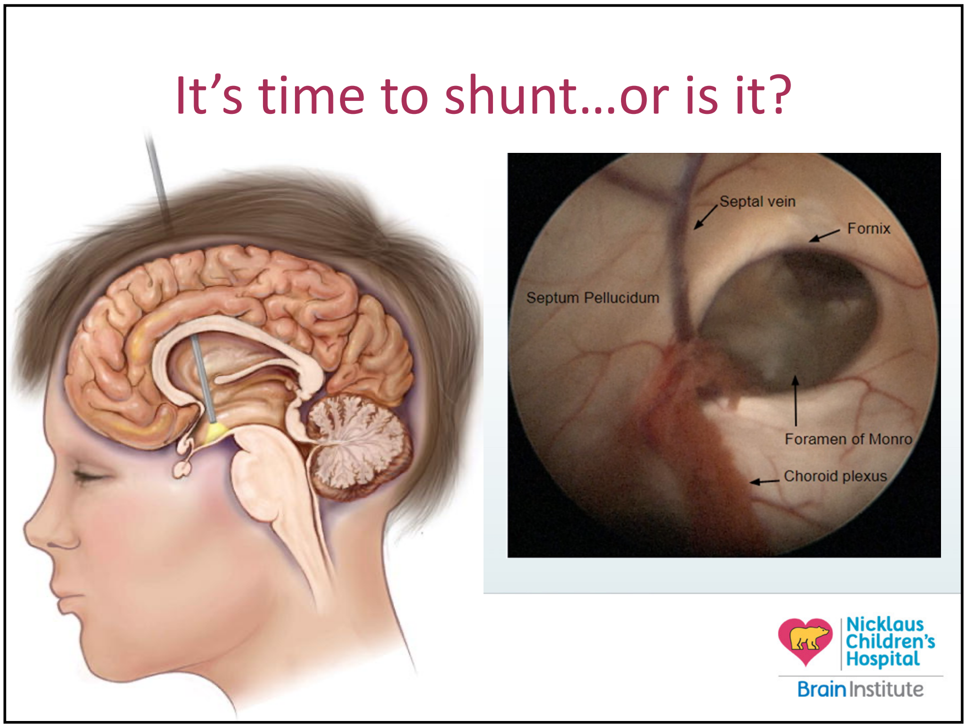

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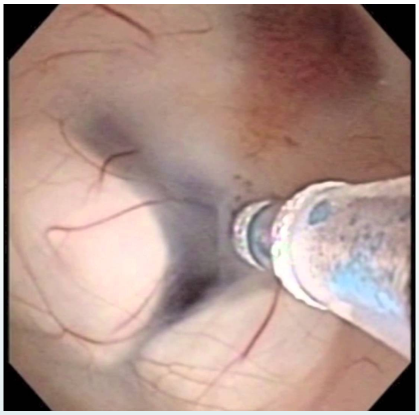
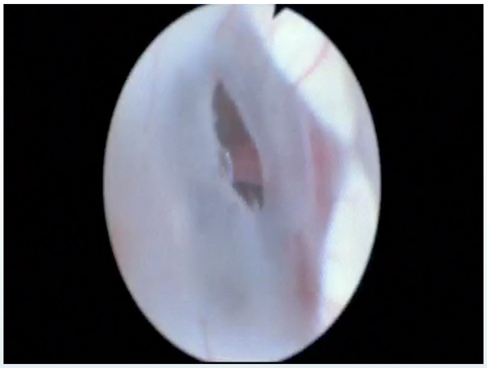




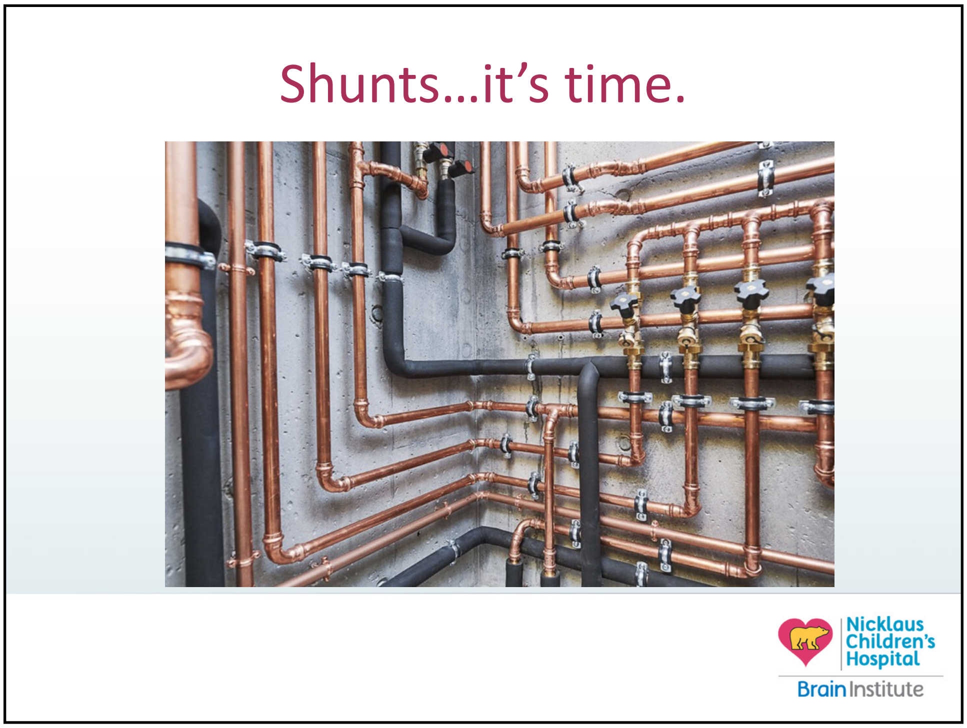
It’s time to shunt…or is it?

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		

Kulkarni AV, *et al*, J Pediatr. 2009



Shunts…it's time.

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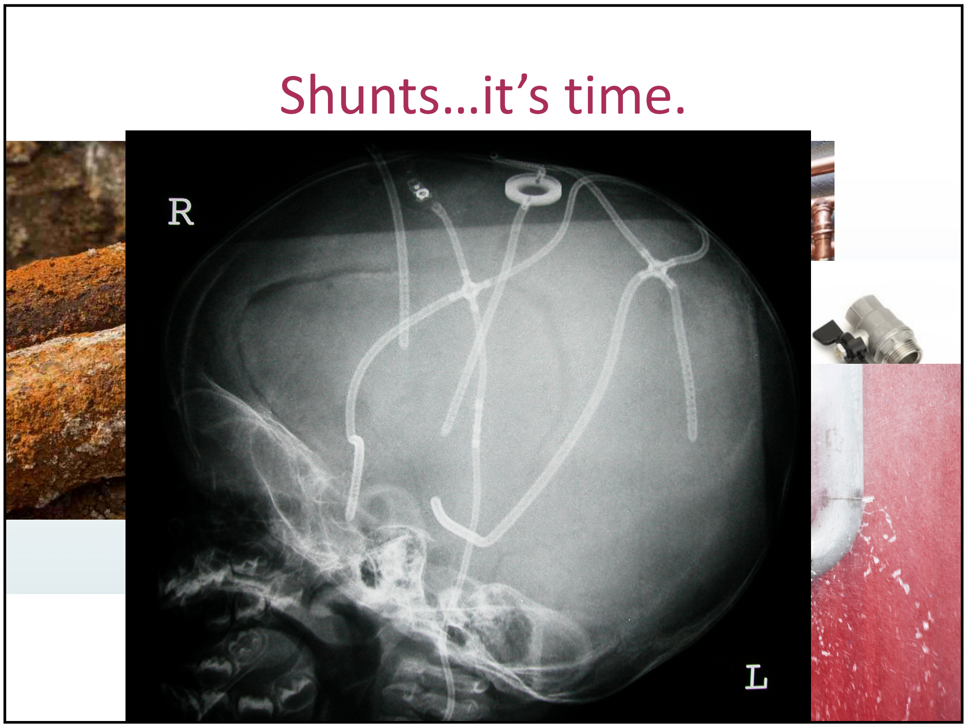
Shunts…it's time.

Shunts…it’s time.

Shunts…it’s time.

Shunts…it's time.

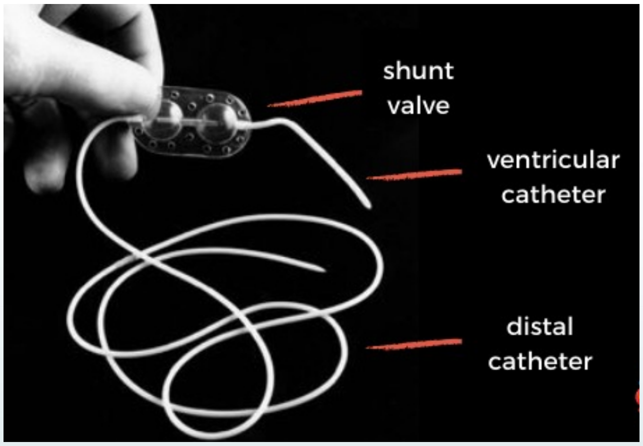
25



Shunts…it's time.

26

It's simple.



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History of Shunts

- Nulsen & Spitz (1951)→ first ball-valve regulated system to the jugular vein
- Refined by Holter (1951)→unidirectional flow, helped to minimize clotting
- Pudenz (1959)→developed one-way silicone slit valve

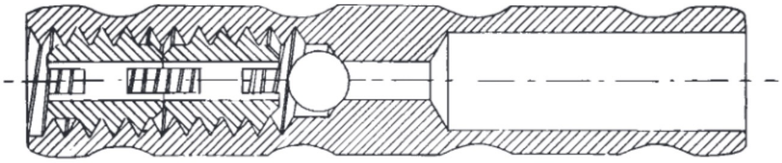
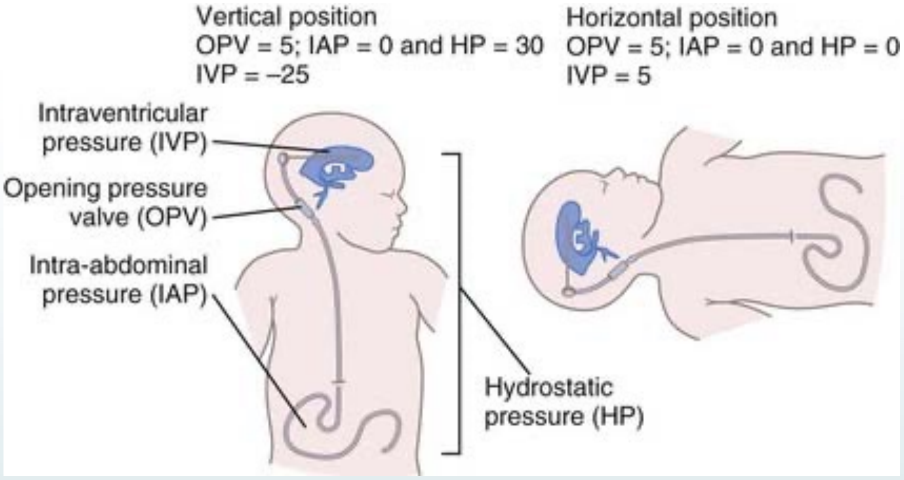


FIG. 1. Schematic drawing of the Johnson Foundation valve with two ball-valve units, platinum springs, and an interposed pumping chamber. Reprinted with permission from Nulsen FE, Spitz EB: Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. **Surg Forum 2**:399–402, 1952.

Shunt Function



It's simple…..?

Proximal Catheter + Valve + Distal Catheter

Ventricle
Cyst
Subdural Space
Syrinx

Abdomen
Heart
Lung
Gallbladder

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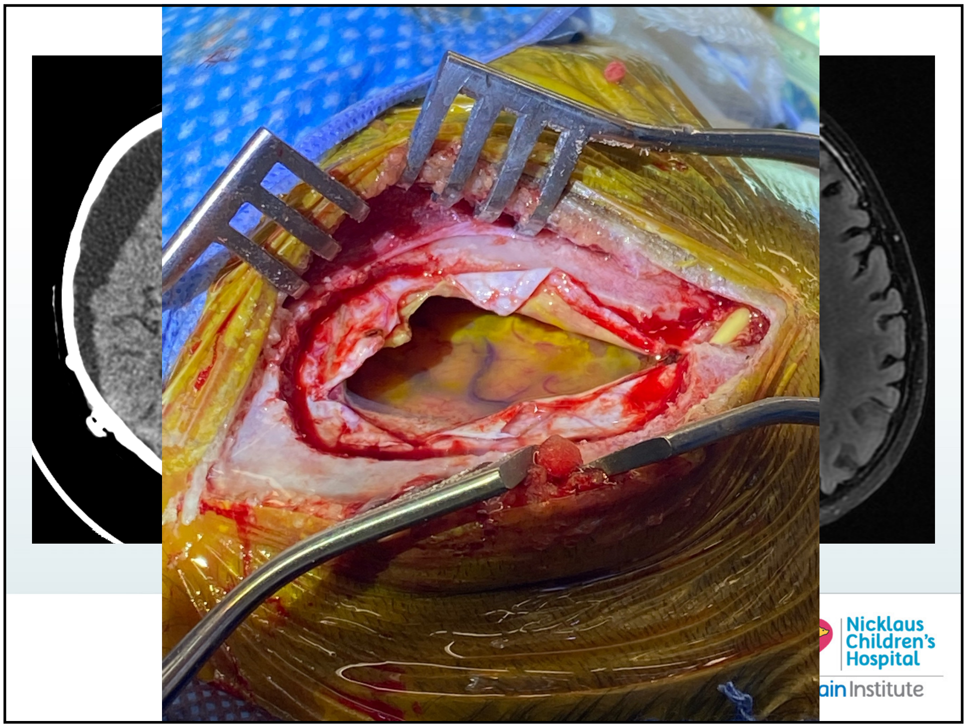
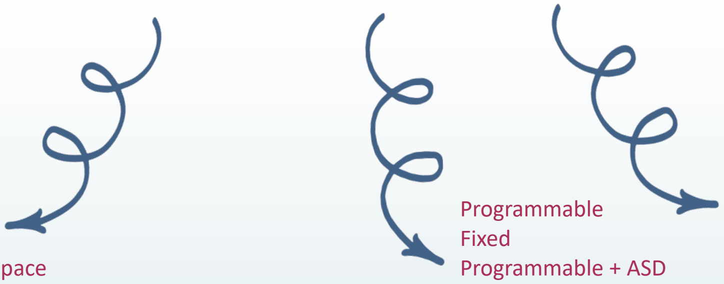


Nicklaus Children's Hospital

ain Institute

It’s simple…..?

Proximal Catheter + Valve + Distal Catheter



Ventricle
Cyst
Subdural Space
Syrinx

Programmable
Fixed
Programmable + ASD
Fixed + ASD
Programmable + Programmable ASD
Distal Slit

Abdomen
Heart
Lung
Gallbladder

Nicklaus Children’s Hospital
Brain Institute

Adjustment and malfunction of a programmable valve after exposure to toy magnets. Case report

Richard C E Anderson [1], Marion L Walker, John M Viner, John R W Kestle

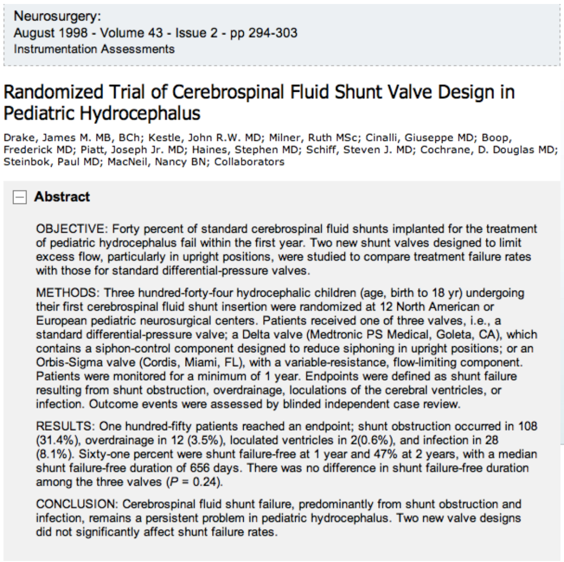
Programmable shunt valve affected by exposure to a tablet computer

Jennifer Strahle [1], Béla J Selzer, Karin M Muraszko, Hugh J L Garton, Cormac O Maher

Programmable shunt valves: in vitro assessment of safety of the magnetic field generated by a portable game machine

Koji Nakashima [1], Takato Nakajo, Michiari Kawamo, Akihito Kato, Seiichiro Ishigaki, Hidetomo Murakami, Yohichi Imaizumi, Hitoshi Izumiyama

Which valve is best?



Neurosurgery:
August 1998 - Volume 43 - Issue 2 - pp 294-303
Instrumentation Assessments

Randomized Trial of Cerebrospinal Fluid Shunt Valve Design in Pediatric Hydrocephalus

Drake, James M. MB, BCh; Kestle, John R.W. MD; Milner, Ruth MSc; Cinalli, Giuseppe MD; Boop, Frederick MD; Piatt, Joseph Jr. MD; Haines, Stephen MD; Schiff, Steven J. MD; Cochrane, D. Douglas MD; Steinbok, Paul MD; MacNeil, Nancy BN; Collaborators

Abstract

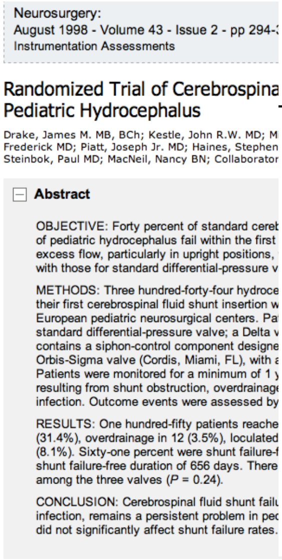
OBJECTIVE: Forty percent of standard cerebrospinal fluid shunts implanted for the treatment of pediatric hydrocephalus fail within the first year. Two new shunt valves designed to limit excess flow, particularly in upright positions, were studied to compare treatment failure rates with those for standard differential-pressure valves.

METHODS: Three hundred-forty-four hydrocephalic children (age, birth to 18 yr) undergoing their first cerebrospinal fluid shunt insertion were randomized at 12 North American or European pediatric neurosurgical centers. Patients received one of three valves, i.e., a standard differential-pressure valve; a Delta valve (Medtronic PS Medical, Goleta, CA), which contains a siphon-control component designed to reduce siphoning in upright positions; or an Orbis-Sigma valve (Cordis, Miami, FL), with a variable-resistance, flow-limiting component. Patients were monitored for a minimum of 1 year. Endpoints were defined as shunt failure resulting from shunt obstruction, overdrainage, loculations of the cerebral ventricles, or infection. Outcome events were assessed by blinded independent case review.

RESULTS: One hundred-fifty patients reached an endpoint; shunt obstruction occurred in 108 (31.4%), overdrainage in 12 (3.5%), loculated ventricles in 2(0.6%), and infection in 28 (8.1%). Sixty-one percent were shunt failure-free at 1 year and 47% at 2 years, with a median shunt failure-free duration of 656 days. There was no difference in shunt failure-free duration among the three valves (*P* = 0.24).

CONCLUSION: Cerebrospinal fluid shunt failure, predominantly from shunt obstruction and infection, remains a persistent problem in pediatric hydrocephalus. Two new valve designs did not significantly affect shunt failure rates.

Which valve is best?



Neurosurgery:
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Pediatric Hydrocephalus

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Patients were monitored for a minimum of 1 y
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among the three valves (P = 0.24).

CONCLUSION: Cerebrospinal fluid shunt failu
infection, remains a persistent problem in pec
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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

[a]Division of Pediatric Neurosurgery, Primary Children's Medical Center, University of Utah, Salt Lake City, Utah, USA; [b]Division of Neurosurgery, Hospital for Sick Children, Toronto, Canada; [c]Service de Neurochirurgie, Hôpital Necker Enfants Malades, Paris, France; [d]Division of Neurosurgery, Arkansas Children's Hospital, Little Rock, Ark.; [e]Division of Neurosurgery, Oregon Health Sciences University, Portland, Oreg.; [f]Division of Neurosurgery, University of Minnesota, Minneapolis, Minn.; [g]Division of Neurosurgery, Children's National Medical Center, Washington, D.C., USA; [h]Division of Neurosurgery, B.C.'s Children's Hospital, Vancouver, Canada

Key Words
Randomized clinical trial · Hydrocephalus · Ventriculoperitoneal shunt

Abstract
Background: A previously reported multicenter randomized trial assessed whether 2 new shunt valve designs would reduce shunt failure rates compared to differential pressure valves. The study did not show a significant difference in the time to first shunt failure. Patients entered the trial between October 1, 1993, and October 31, 1995. The primary results were based on the patients' status as of October 31, 1996 (a minimum follow-up of 1 year). This report describes the late complications based on the patients' most recent follow-up. ***Methods:*** Three hundred and forty-four hydrocephalic children at 12 North American and European centers were randomized to 1 of 3 valves: a standard differential pressure valve; a Delta valve (PS Medical-Medtronic) or a Sigma valve (NMT Cordis). Patients were followed until their first shunt failure. Shunt failure was defined as shunt surgery for obstruction, overdrainage, loculation or infection. If the shunt did not fail, follow-up was continued until August 31, 1999. ***Results:*** One hundred and seventy-seven patients had shunt failure. Shunt obstruction occurred in 131, overdrainage in 13, loculated ventricles in 2 and infection in 29. The overall shunt survival was 62% at 1 year, 52% at 2 years, 46% at 3 years, 41% at 4 years. The survival curves for the 3 valves were similar to those from the original trial and did not show a survival advantage for any particular valve. ***Conclusions:*** Prolonged follow-up to date does not alter the primary conclusions of the trial: there does not appear to be one valve that is clearly the best for the initial treatment of pediatric hydrocephalus.

Introduction

A multicenter randomized trial (The Shunt Design Trial [1]) assessed whether either of 2 new shunt valves (compared to differential pressure valves) could decrease the 1-year shunt failure rate from 40 to 20%. Twelve North American and European centers randomized 344 patients. The study did not show a significant difference in the time to first shunt failure among the 3 valve

The collaborators are listed in the appendix.

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1016–2291/00/0335–0230$17.50/0

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Which valve is best?

Neurosurgery:
August 1998 - Volume 43 - Issue 2 - pp 294-3
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ORIGINAL PAPER

Management of neonatal hydrocephalus: feasibility of use and safety of two programmable (Sophy and Polaris) valves

Juan F. Martínez-Lage · María-José Almagro · Isabel Sanchez del Rincón · Miguel A. Pérez-Espejo · Claudio Piqueras · Raúl Alfaro · Javier Ros de San Pedro

Received: 19 August 2007 / Published online: 9 October 2007

Abstract
Background Neonates represent a unique group of pediatric patients with special peculiarities. Hydrocephalus valves have not always been designed to meet the requirements of these small children. Few series have addressed the problem of cerebrospinal fluid shunting in newborn babies.
Objectives We aimed (1) to evaluate the feasibility of the use of two programmable valves (Sophy and Polaris) in hydrocephalic neonates and (2) to ascertain complications and safety issues arising from their use.
Materials and methods We performed a prospective study of 100 consecutive preterm and term babies (<2 months of age) given a programmable valve. Valves' settings were readjusted at different pressure levels as required. Outcomes were obtained from the records of our Outpatient Clinic.
Results The study group was formed by 60 term and 40 preterm infants (average weight 2,440 g, mean age of 25 days). Mean follow-up was 55 months. Only one fifth deaths was shunt-related. In 70 babies, no complications occurred, and hydrocephalus was successfully controlled. Proximal catheter obstruction presented in 20% and infection in 5% of cases. Several external adjustments of the valves apparently avoided several surgical shunt revisions.
Conclusions (1) Both programmable valves (Sophy and Polaris) can be safely used for treatment of neonatal hydrocephalus, introducing some technical modifications. (2) Both valves are comparable to other shunts with regard to indications, performance, and safety. (3) The possibility of modifying their working pressure seems to constitute their main advantage. Prevention of late overdrainage syndromes with these valves needs a longer follow-up.

Keywords Neonatal hydrocephalus · Programmable hydrocephalus valves · Pediatric hydrocephalus · Sophy valve · Polaris valve · CSF shunts · Programmable valve safety · Programmable valve efficiency

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Introduction

Recent advances in the treatment of hydrocephalus comprise the development of a variety of valve designs that include antisiphon devices, flow-regulated shunts, and hydrostatic and programmable valves [1, 5, 12–14, 20–22, 26, 28, 35–37, 39–41, 44, 48–50]. Adjustable pressure shunts incorporate a valve that can be regulated externally by a magnet, thus allowing modifying noninvasively their flow characteristics. Programmable valves have proven to be beneficial in patients with normal pressure hydrocephalus, arachnoid cysts, and in the management of shunt complications such as the slit ventricle syndrome and

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ORIGINAL PAPER

Shunt survival rates by using the adjustable differential pressure valve combined with a gravitational unit (proGAV) in pediatric neurosurgery

Ulrich-W. Thomale · Anna F. Gebert · Hannes Haberl · Matthias Schulz

Received: 5 June 2012 / Accepted: 24 October 2012

Abstract
Object Overdrainage is a chronic complication in shunted pediatric patients with hydrocephalus. The use of adjustability of differential pressure (DP) valves in combination with antisiphoning devices may help to overcome this sequela and may diminish the rate of possible shunt failures. The purpose of this retrospective study is to report our experience on shunt survival and infection rate with an adjustable DP valve with integrated gravitational unit in pediatric hydrocephalus.
Methods The proGAV consists of an adjustable differential pressure (DP) valve and a gravitational unit. During the time period of July 2004 and December 2009, a total of 237 adjustable gravitational valves were used in 203 children (age, 6.5±6.54; 0–27 years). In the follow-up period, valve and shunt failures as well as rate of infection were recorded.
Results Within the average follow-up time of 21.9± 10.3 months (range, 6–72 months), the valve survival rate was 83.8 %. The overall shunt survival rate including all necessary revisions was 64.3 %. Looking at the group of infants (<1 year of age) within the cohort, the valve survival rate was 77.3 % and the shunt survival rate was 60.9 %. The overall infection rate was 4.6 %.
Conclusion In a concept of avoiding chronic overdrainage by using the proGAV in hydrocephalic children, we observed a good rate of valve and shunt survival. Compared to previous reported series, we experienced the proGAV as a reliable tool for the treatment of pediatric hydrocephalus.

Keywords Hydrocephalus · Pediatric · CSF shunt · Adjustable valves · Gravitational unit · Over drainage · Under drainage

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Introduction

For the treatment of pediatric hydrocephalus, the technical quality of new valve designs continues to improve by the introduction of adjustability in differential pressure (DP) valves and overcoming the siphoning effect by, e.g., antisiphon devices or gravitational units. It is an evident fact to keep in mind, that the quality of shunts used at the first implantation may significantly influence the neurocognitive development of children by its biomechanical construction as well as by any risks for possible complications or the need of further revisions.

Overdrainage is a well-known chronic complication after long-term shunting in pediatric hydrocephalus [5–8, 11, 29, 33]. The compensatory capacity of the pediatric skull and central nervous system anatomy leads to long-term changes of the cerebrospinal fluid (CSF) tissue ratios which are able to cause microcephaly, calvarial hyperplasia, slit ventricle syndrome, and shunt-induced Chiari malformations [9, 12, 19, 28, 36]. In older children, frequent complications do not only lead to further surgeries and shunt revisions but also to chronic headaches and additional neurological impairments [1, 10, 14, 36, 40]. Since these changes are only recognized after years of treatment, this problem seems to be rather neglected [15].

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Neurosurgery:
August 1998 - Volume 43 - Issue 2 - p
Instrumentation Assessments

Randomized Trial of Cerebro
Pediatric Hydrocephalus

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Frederick MD; Piatt, Joseph Jr. MD; Haines, S
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OBJECTIVE: Forty percent of standa
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METHODS: Three hundred-forty-four
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Orbis-Sigma valve (Cordis, Miami, FL
Patients were monitored for a minimu
resulting from shunt obstruction, over
infection. Outcome events were asse

RESULTS: One hundred-fifty patients
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shunt failure-free duration of 656 days
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It doesn't matter!

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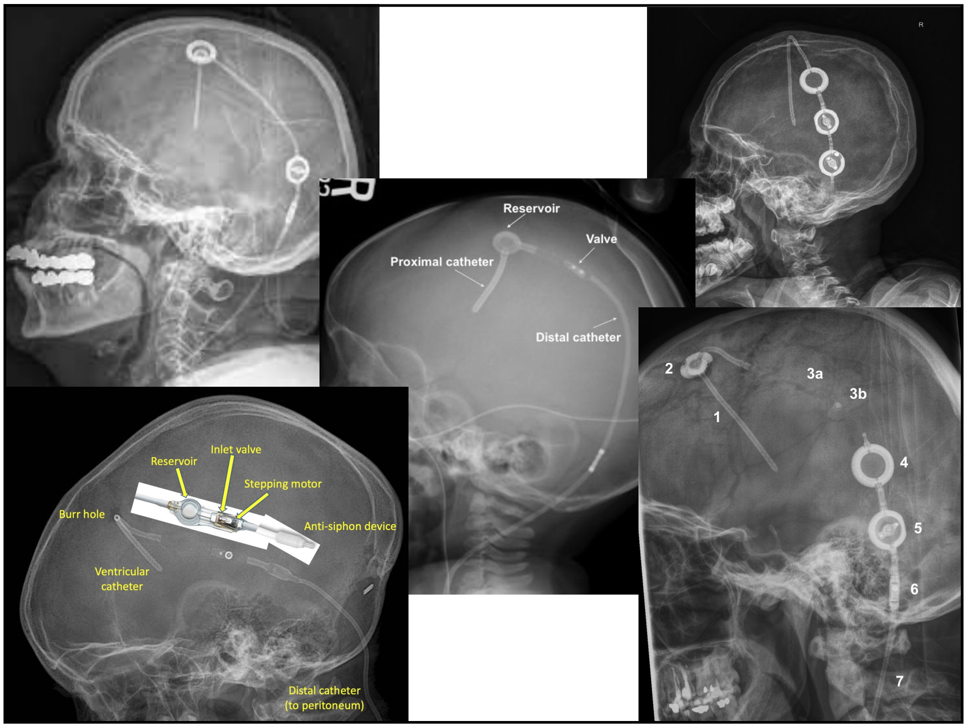

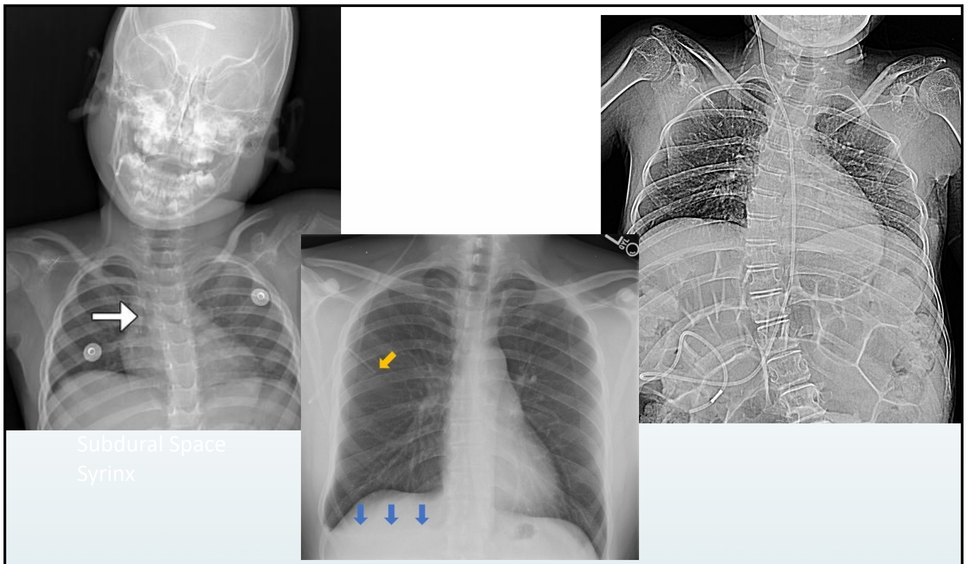

Subdural Space
Syrinx

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Context Matters

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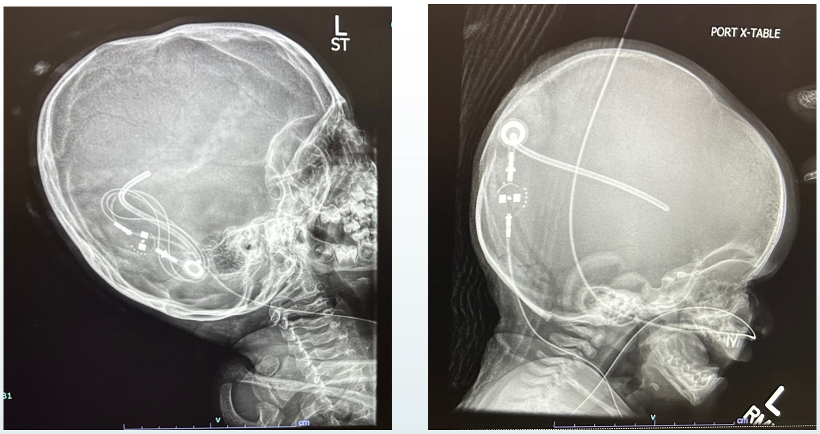
Context Matters



Shunt Evaluation

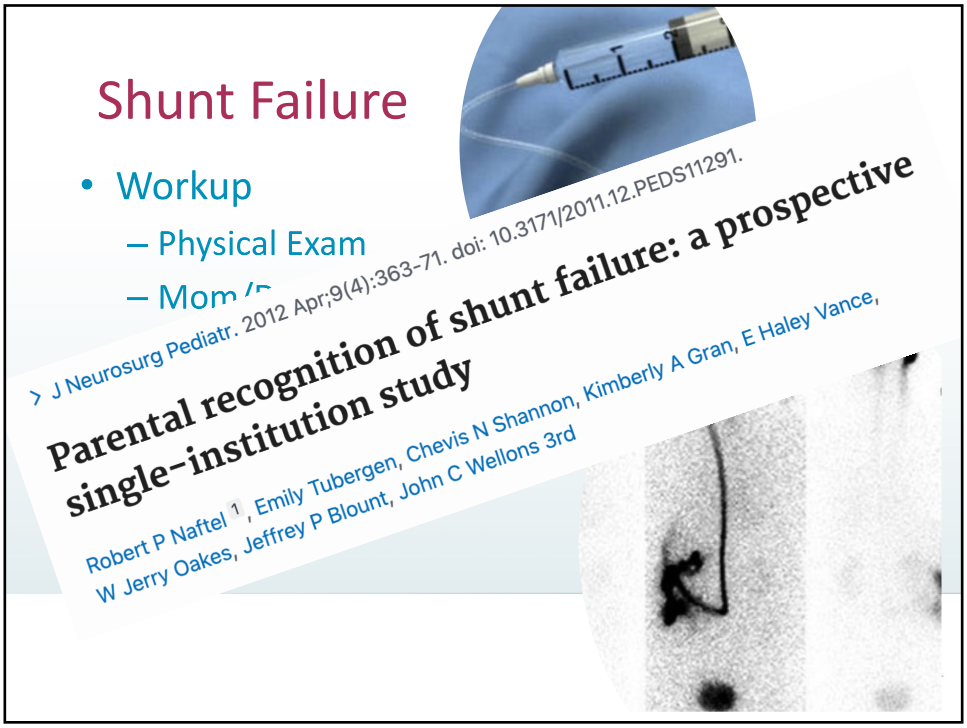
- Shunt History
  - Reason
  - Timing
  - Revisions
  - Type
  - Setting
  - Radiographic change*

Shunt Failure

- Workup
  - Physical Exam
  - Imaging
  - Shunt tap/Nuclear Medicine
  - Surgical Exploration
  - Mom & Dad *

Shunt Failure

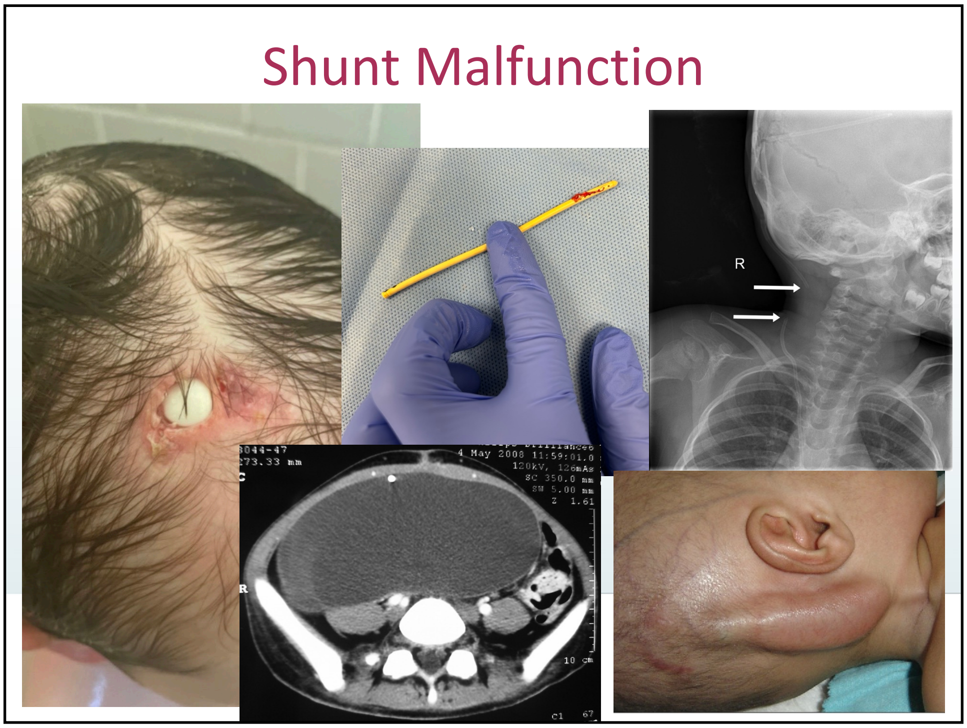
- Workup
  - Physical Exam
  - Mom /



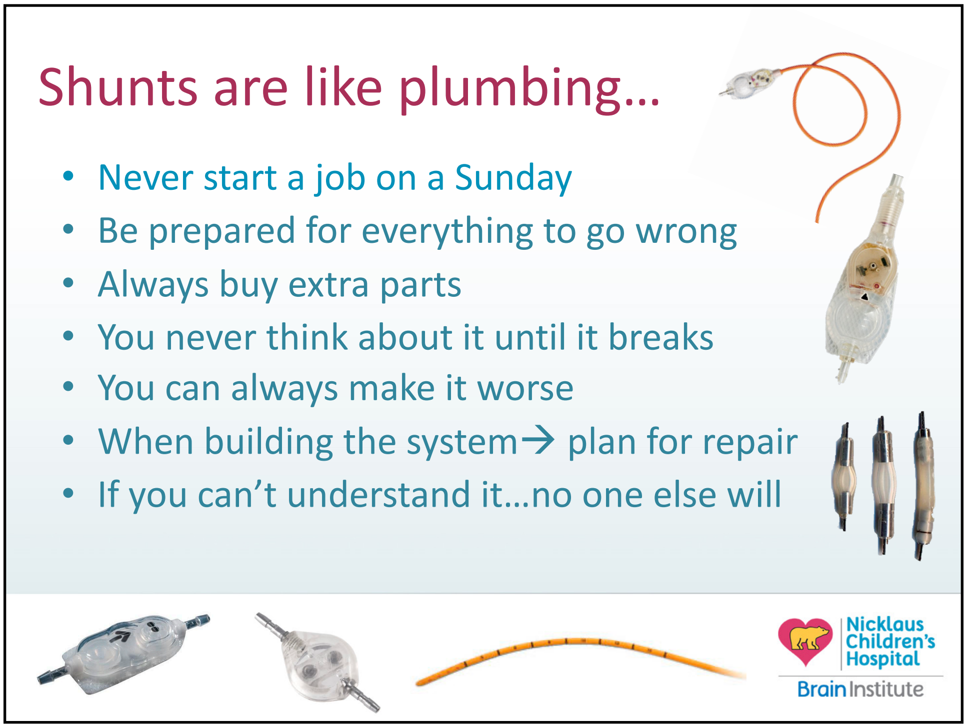
> J Neurosurg Pediatr. 2012 Apr;9(4):363-71. doi: 10.3171/2011.12.PEDS11291.

Parental recognition of shunt failure: a prospective single-institution study

Robert P Naftel [1], Emily Tubergen, Chevis N Shannon, Kimberly A Gran, E Haley Vance, W Jerry Oakes, Jeffrey P Blount, John C Wellons 3rd



Shunt Malfunction



Shunts are like plumbing…

- Never start a job on a Sunday
- Be prepared for everything to go wrong
- Always buy extra parts
- You never think about it until it breaks
- You can always make it worse
- When building the system→ plan for repair
- If you can't understand it…no one else will

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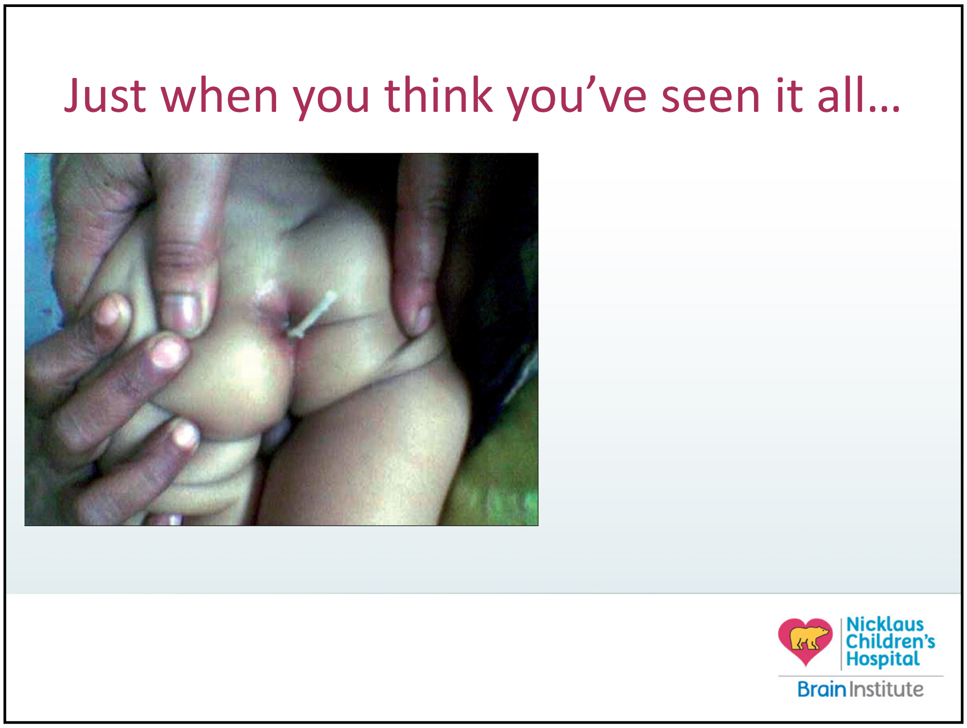

Just when you think you've seen it all…

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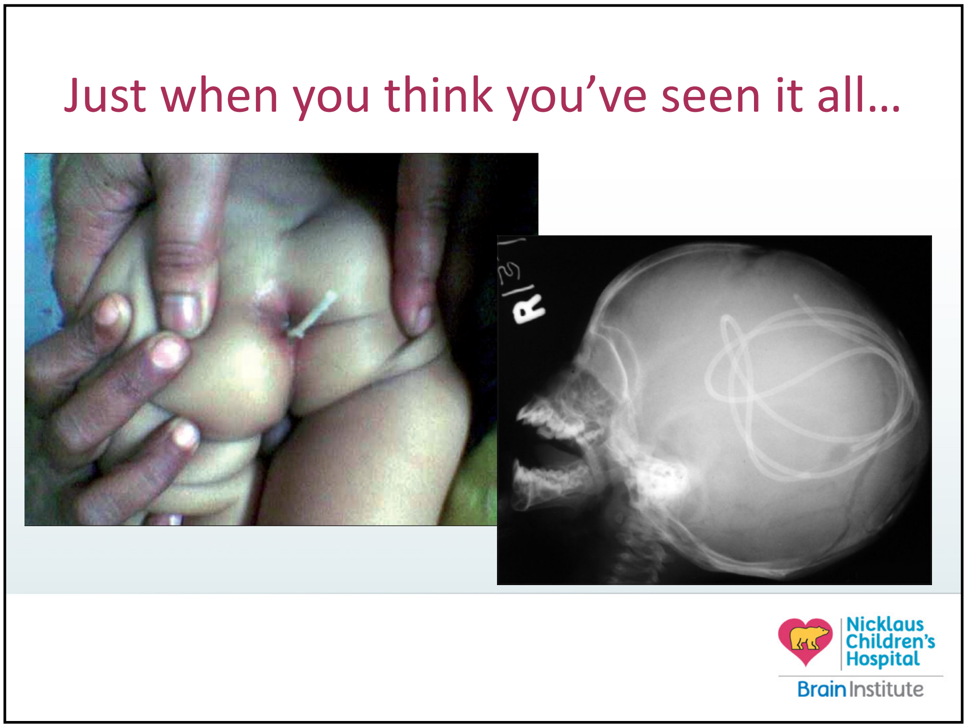

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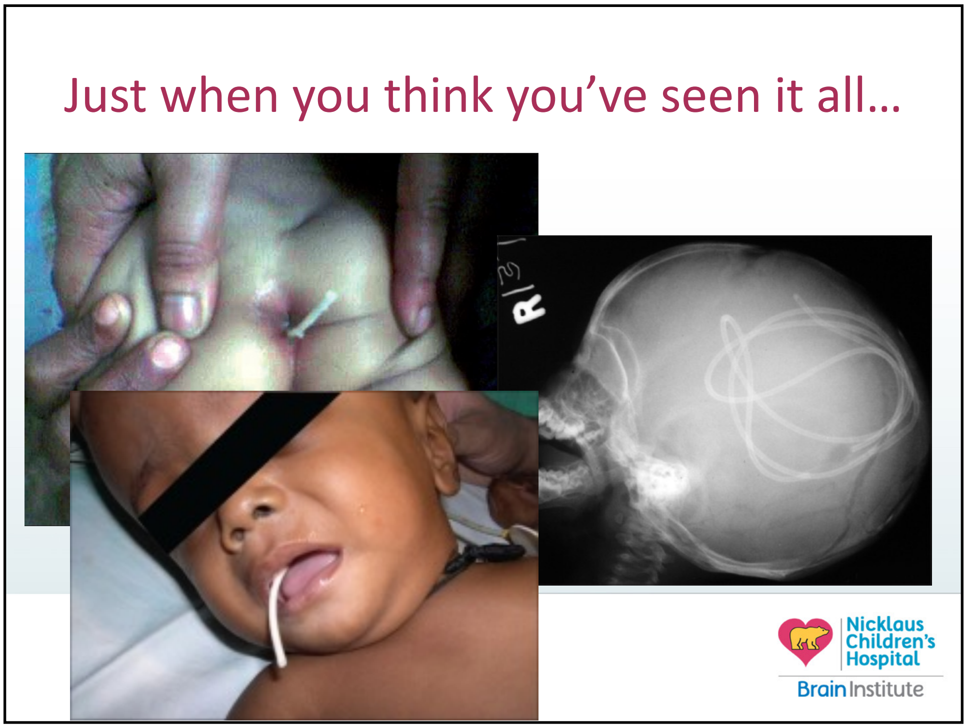

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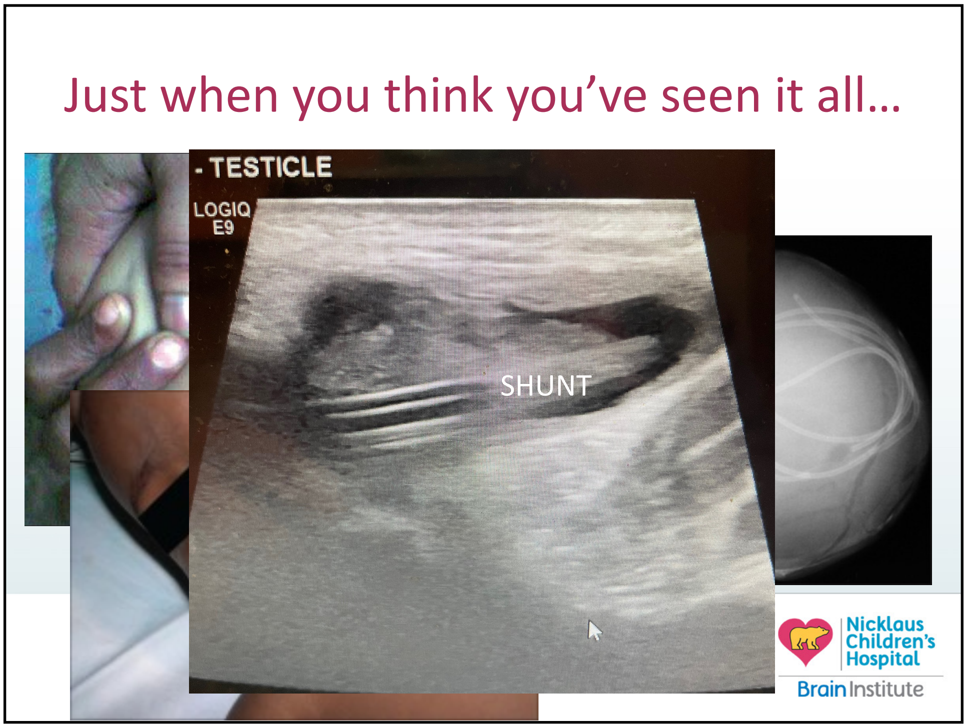

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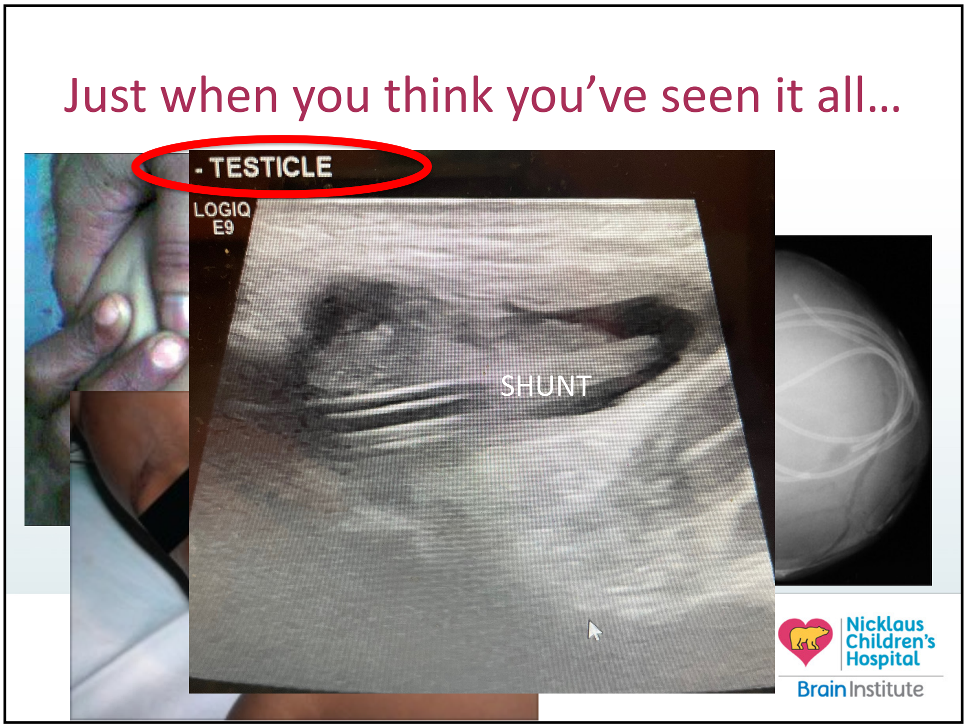

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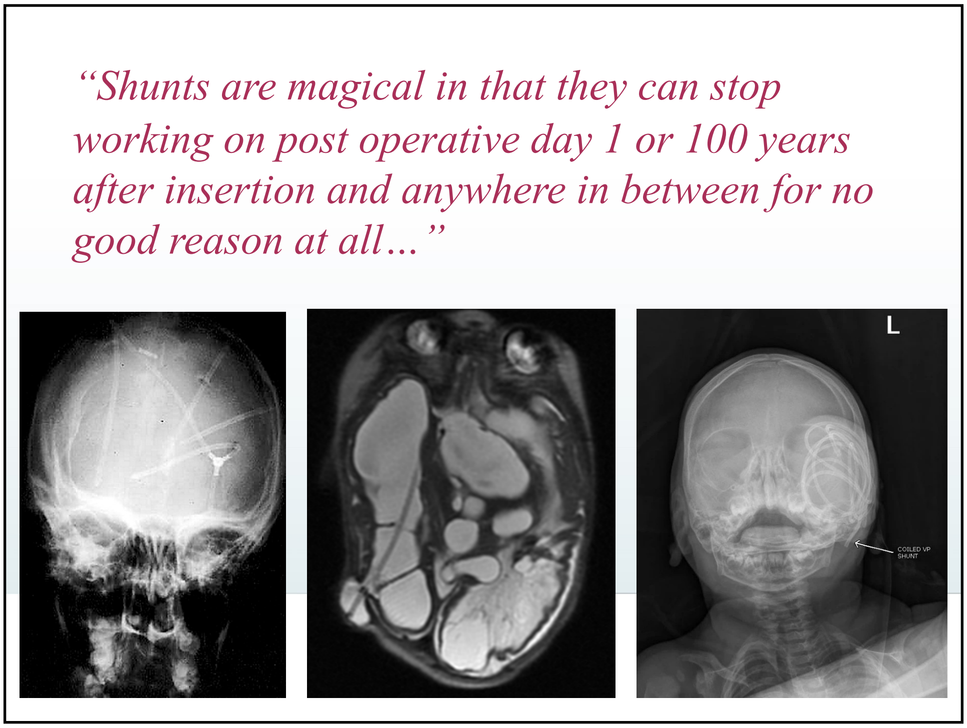

"Shunts are magical in that they can stop working on post operative day 1 or 100 years after insertion and anywhere in between for no good reason at all…"

Thank you!