CSF SHUNTS GONE BAD!

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Hydrocephalus

Hydrocephalus

Imbalance of absorption and production of CSF

Estimated incidence of 1/500-1000 children

125,000+ shunts

Either due to obstruction of CSF outflow, impaired reabsorption or excess production

Obstructive hydrocephalus

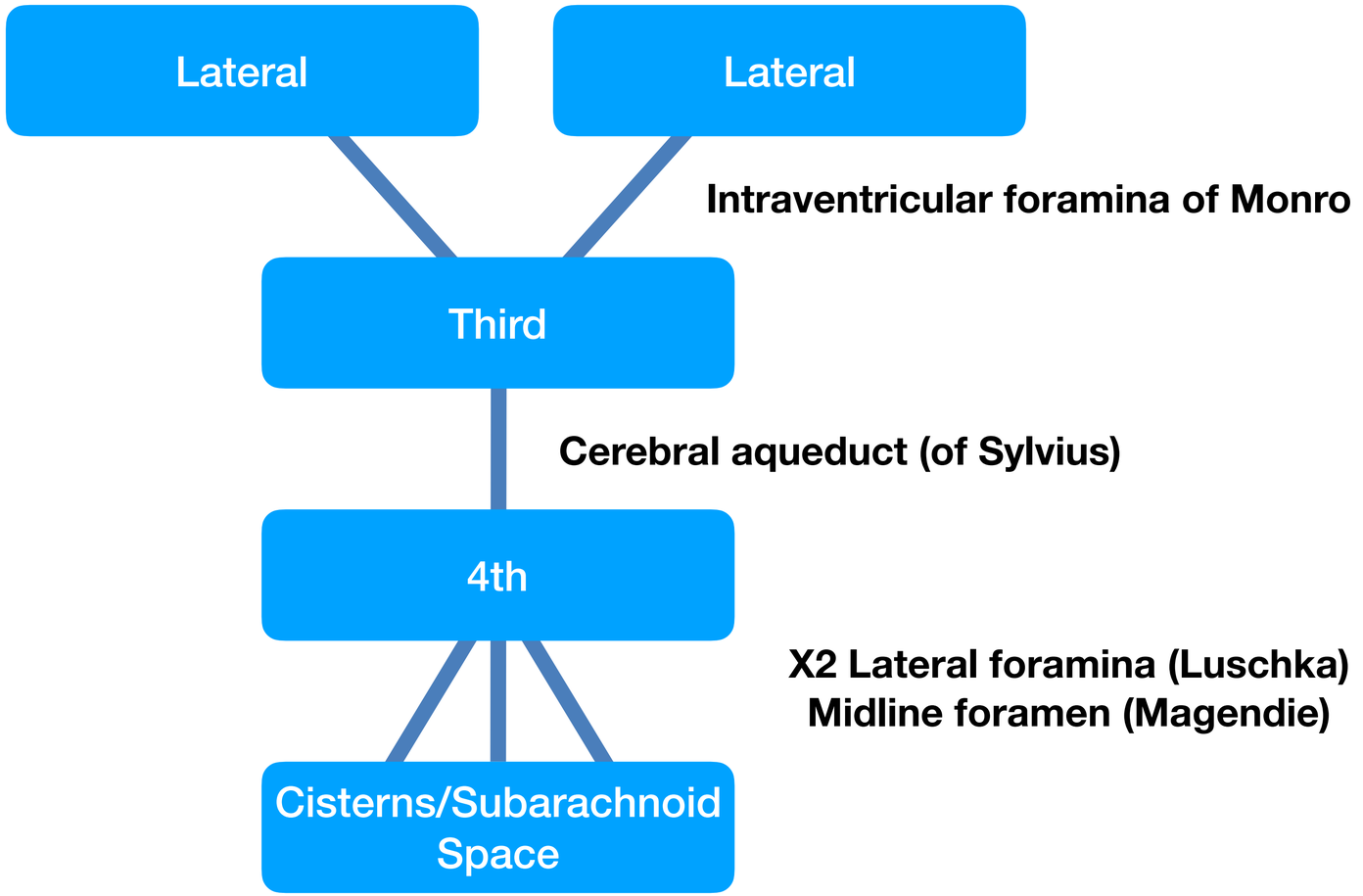
The ventricular system is blocked and CSF accumulates proximal to the blockage

Communicating hydrocephalus

The subarachnoid system is blocked and CSF can’t be absorbed

The entire system fills with CSF

This is less common and due to IVH, Meningitis, Post-inflammatory scarring


Lateral
Lateral
Intraventricular foramina of Monro
Third
Cerebral aqueduct (of Sylvius)
4th
X2 Lateral foramina (Luschka)
Midline foramen (Magendie)
Cisterns/Subarachnoid Space

Etiology

Congenital

infection: Rubella, CMV, Toxo, Syphilis

X-Linked hydrocephalus stenosis of aqueduct of Sylvius

Acquired

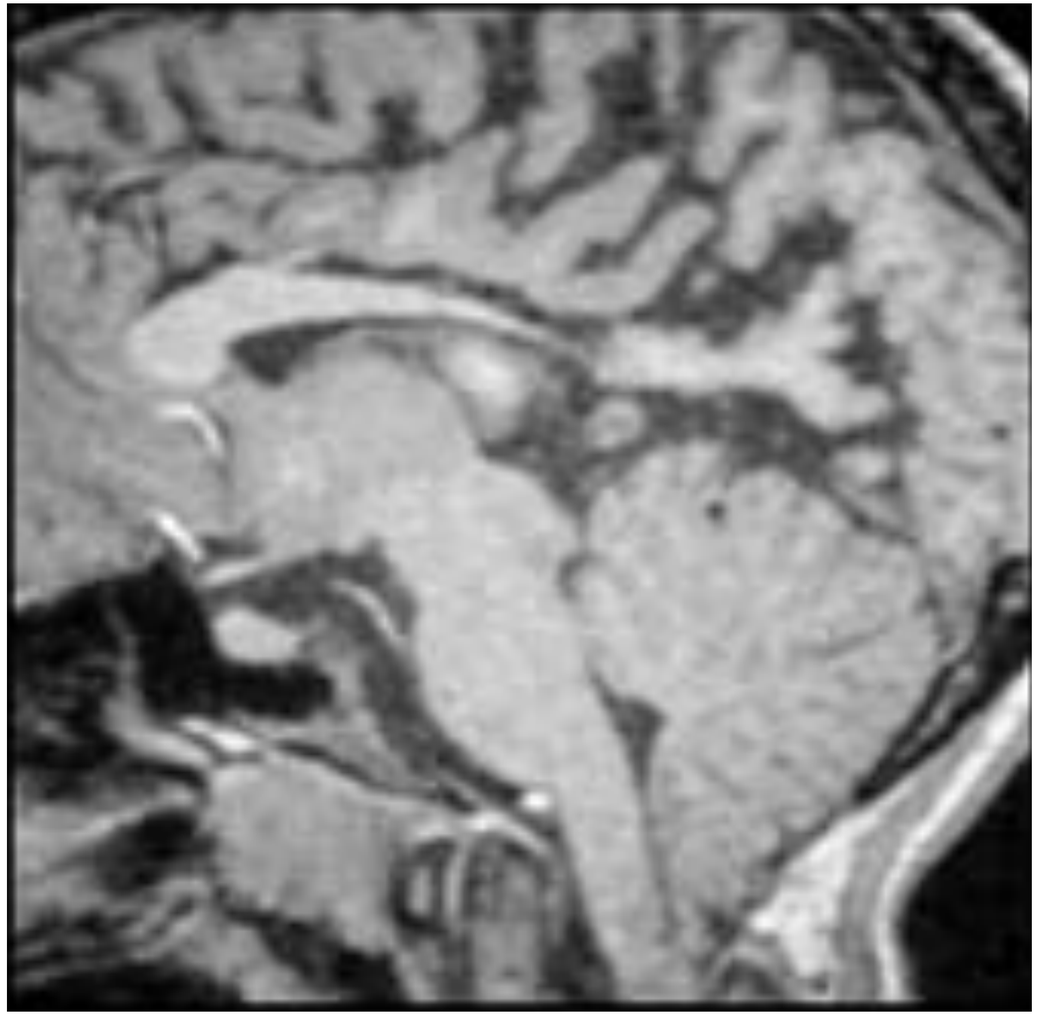
Infection, trauma, tumors, head bleeds

Neural tube defects:

Associated with Chiari or aqueductal stenosis. Linked to teratogens and deficiency of folate.

Isolated

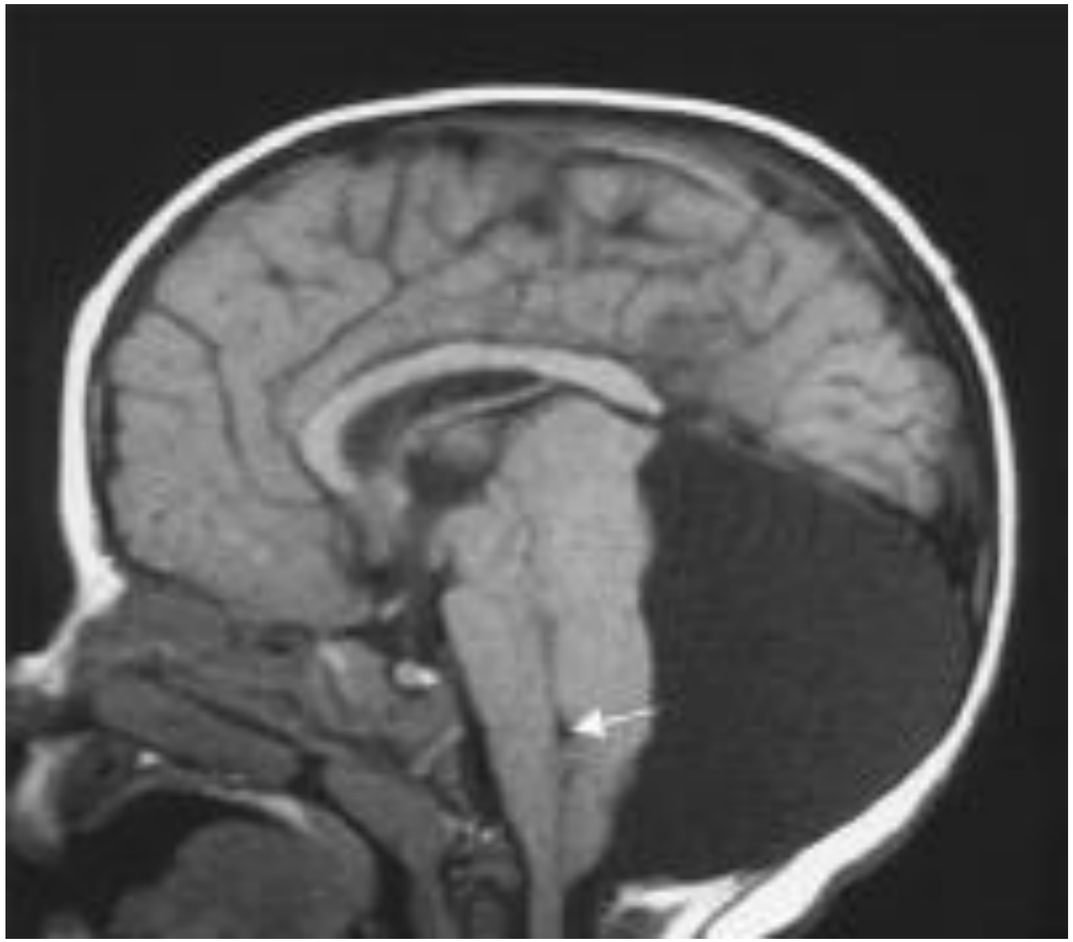
aqueductal stenosis (inflammation d/t intrauterine infection)



Chiari II

Often accompanies NTD

Brainstem and Cerebellum are displaced caudally



Dandy Walker

Large posterior fossa cyst continuous with 4th ventricle

Abnormal cerebellar development

Hydrocephalus in 70-90%

Presenting symptoms of hydrocephalus

Headache

Vomiting: increased ICP in the posterior fossa

Behavioral changes

Drowsiness: midbrain/brainstem dysfunction

Visual changes: Optic Nerve compression

Incoordination

Loss of developmental milestones

Head circumference increases rapidly

“Sunsetting“ eyes: fixed downward gaze

Shunts

Shunt devices

Proximal portion is placed in a ventricle (usually the right)

Could also be in an intracranial cyst or lumbar subarachnoid space

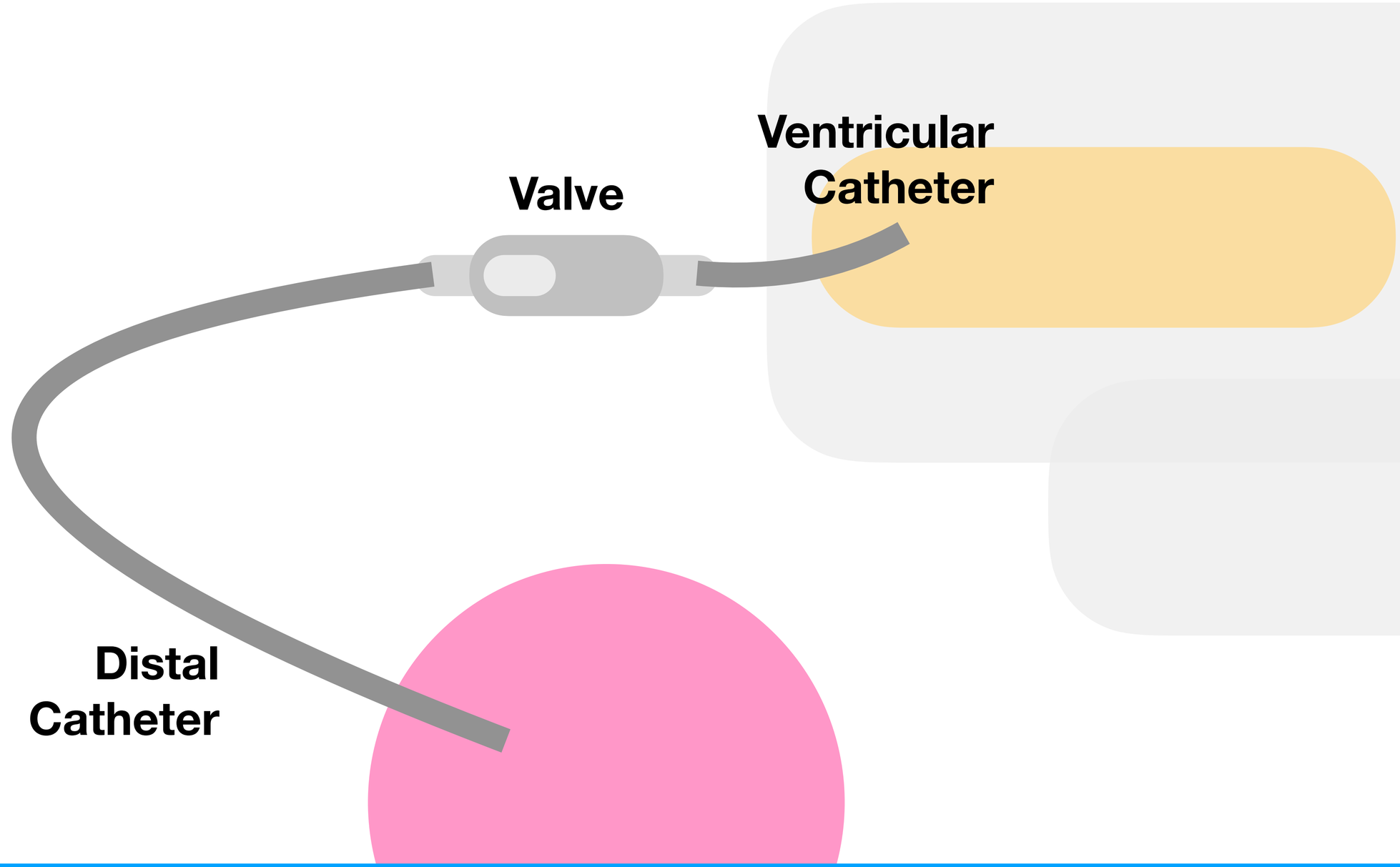
Distal portion

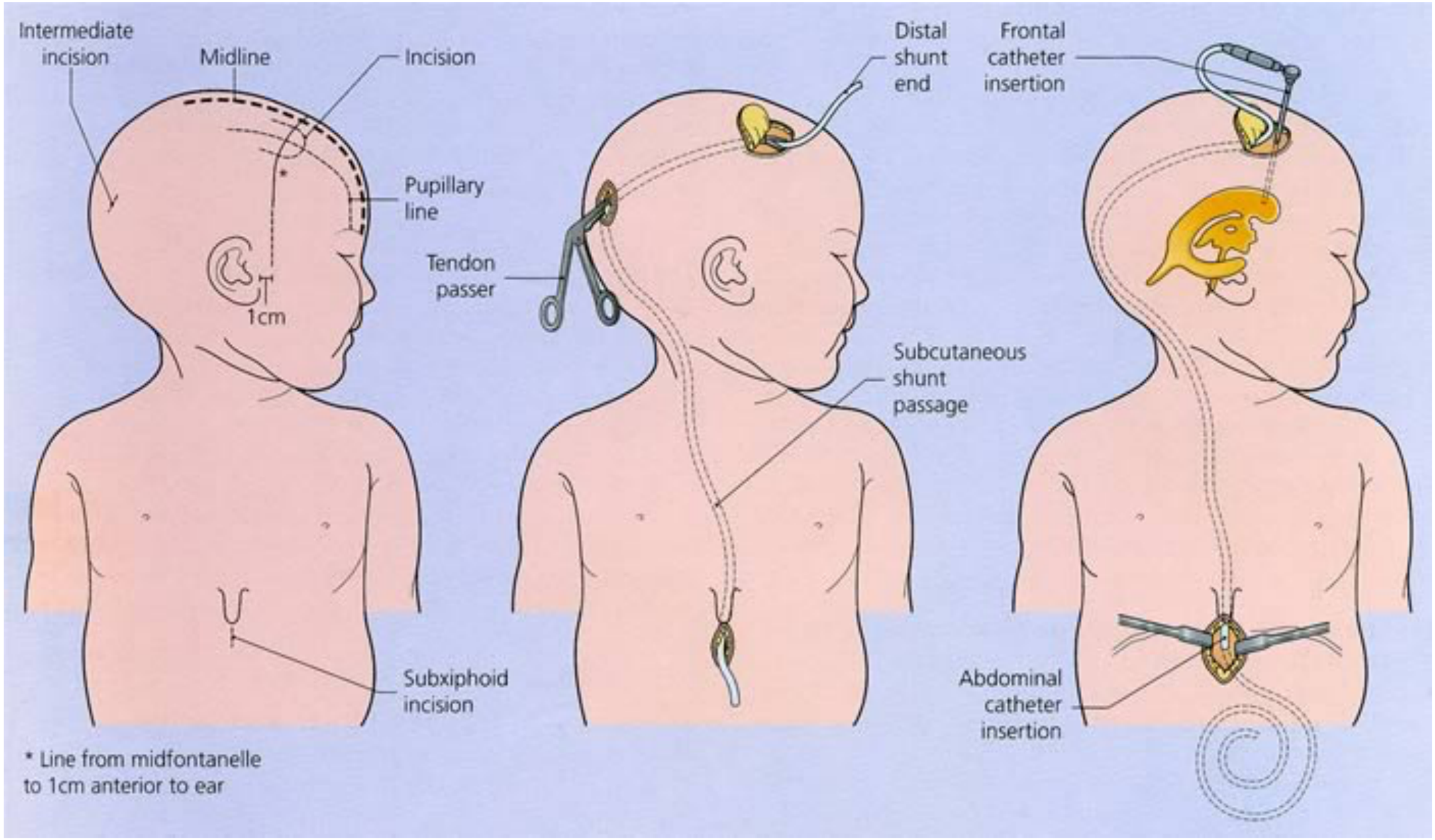
Internalized: peritoneum, pleura, atrium

Externalized

EVD: Acute hydrocephalus for pressure monitoring, infected shunt

Ommaya reservoir: Generally for administration of drugs (antibiotics or chemo)


Ventricular
Catheter
Valve
Distal
Catheter


Intermediate incision
Midline
Incision
Pupillary line
1cm
Subxiphoid incision
* Line from midfontanelle to 1cm anterior to ear
Tendon passer
Distal shunt end
Subcutaneous shunt passage
Frontal catheter insertion
Abdominal catheter insertion

Fixed-pressure valves

- **Hakim Microprecision**
  - 0-5 cm H_2O outflow resistance
  - Siphonguard (ball and cone) can prevent overdrainage
  - Codman (Johnson and Johnson) Company

- **PS Medical**
  - Injectable reservoir
  - Nonmetallic
  - Low-low, low, medium, and high pressure outflow available
  - Medtronic

- **Chhabra**
  - Low cost–widely used in developing world
  - Made in India
  - Mechanism: Z flow system of three balls

Flow-regulated valves

- **Delta**
  - Delta Chamber opens for free flow if ICP high
  - Elastomer diaphragm mechanism
  - 0.5, 1.0, 1.5, 2.0, 2.5 levels available
  - Medtronic

- **Orbit-Sigma OSV II**
  - First flow-regulated valve
  - Three stage variable resistance mechanism
  - Magnet neutral/MRI safe
  - Integra

Programmable pressure valves

- **Strata**
  - Ball/spring mechanism with magnet
  - Programmable/noninvasive
  - Incorporated Delta chamber
  - Medtronic

- **Codman Hakim**
  - Noninvasive programs among 18 preset levels
  - Ball/spring mechanism
  - Codman (Johnson and Johnson) Company

- **Sophy**
  - First adjustable valve
  - Siliconecoated polycarbonate chamber
  - Ball-cone mechanism with variable pressure spring
  - Sophysa

- **Polaris**
  - MRI compatible variable valve
  - Self-locking magnetic system
  - Sophysa

Complications

Infection

Malfunction

Over drainage

Under drainage

Subdural hematoma

Multiloculated hydrocephalus

Seizures

Shunt Infections

Infection

5-15% overall risk

Fever is variably present, and meningeal signs are not correlative

Ventriculoperitoneal shunt infections can also present with GI Sx/ peritonitis

VA shunts with endocarditis

Shunt infections are more likely in the first month after placement

Infection

Risk factors

Younger age

Previous shunt infection

Certain causes of hydrocephalus (more likely after purulent meningitis, hemorrhage, or myelomeningocele)

Shunt revision - especially ≥3 revisions

Infection

Risk factors

Less experienced neurosurgeon

More people in the OR

Use of a neuroendoscope

Longer duration of the shunt procedure

For VA shunts insertion of the catheter below T7 vertebral body

Skin preparation/shaving of skin

Infection

EVD

External Ventricular Drain risk is up to 1 in 5

10.6 infections per 1000 catheter days

Risk greatest if in place >5 days

Infection

Usually due to skin flora or more rarely hematogenous spread

50% Coag negative Staph - ⅓ of Staph is Staph aureus

Cutibacterium [FKA *Propionibacterium*] *acnes* and *Corynebacterium jeikeium*

Infection

Most come from the proximal end

Distal site infections are a result of contamination from peritonitis

Gram negatives, Pseudomonas, Streptococci, anaerobes are rare in kids

Infection

Diagnosis requires an organism cultured from the CSF

OR

>1 year of age ≥2 of…

Fever, headache, meningeal signs, or cranial nerve signs

≤1 year of age: ≥2 of…

Fever >38°C or hypothermia <36°C, apnea, bradycardia, or irritability **and** at ≥1 of…

- Increased CSF white blood cell count, elevated CSF protein, and decreased CSF glucose
- Organisms seen on a CSF Gram stain
- Organisms cultured from the blood
- Positive nonculture diagnostic test from the CSF, blood, or urine

Infection

CSF is better obtained via shunt tap

CT or MRI should be performed

Abdominal U/S if the child has GI Sx (looking for pseudocyst)

Infection

Treatment

Device removal and external drainage with replacement once CSF is sterile for ≥48 hours

Parenteral antibiotics for 10-14 days

Empiric Vanc + cefotaxime/ceftriaxone

Shunt Malfunctions

Shunt malfunctions are usually due to mechanical failure

Malfunction

Majority of 1st failures are due to **obstruction at the ventricular catheter**

- Shunt over drains
- Ventricles shrink
- Tip gets clogged against choroid plexus

Other causes include shunt migration and excessive CSF drainage

15% due to fractured tubing

Shunt malfunctions need to be recognized quickly and managed in the operating room

Malfunction

Median survival of a shunt (before need for revision)

- ≤2 years old = 2 years
- ≥2 years old = 8 to 10 years

Malfunction

A decision rule was developed - *Peds Emerg Care,* 2008

Sign/Symptom	+LR	-LR
Bulging fontanel	44.6	1.84
Irritability	13.7	1.75
Nausea/Vomiting	11.1	1.58
Accelerated head	6.02	1.86
Headache	4.28	1.22

Malfunction

Children with a shunt malfunction were less likely to present with…

- Fever
- Seizure

History of multiple prior revisions was also associated with risk for shunt malfunction

Malfunction

Validation of the previous decision rule

146/755 ED visits for 294 kids had a shunt malfunction (19%; 95% CI, 17%–22%)

Children with a ventricular shunt malfunction were more likely to present with…

- Headache
- Nausea and/or vomiting
- Bradycardia
- Mental status change

Boyle et al. *Pediatric Emerg Care*, 2017

TABLE 1. Comparison of Patient Visits Where Ventricular Shunt Malfunction Was Present or Absent

	Ventricular Shunt Malfunction n/N (%) N = 146	No Ventricular Shunt Malfunction n/N (%) N = 609	*P*
Demographics			
Age (y)*	12.1 (8.2–15.8)	9.4 (5.4–15.1)	<0.001
Male sex	93/146 (64%)	334/609 (55%)	0.053
Historical features			
Age at initial ventricular shunt insertion (mo)*	2 (0.5–13.5)	3 (0.75–12)	0.373
No. previous revisions			
None/1	42/146 (29%)	284/608 (47%)	<0.001
2 or more	104/146 (71%)	324/608 (53%)	
Time from insertion or last revision (mo)*	7.9 (1.1–39.6)	16.9 (3.8–39.8)	0.008
Headache	107/125 (86%)	358/470 (76%)	0.023
Nausea and/or vomiting	96/141 (68%)	336/589 (57%)	0.017
Seizure	13/146 (9%)	98/609 (16%)	0.028
Physical examination			
Temperature ≥38.0°C	10/146 (7%)	128/609 (21%)	<0.001
Heart rate below age-based normal range	19/146 (13%)	17/607 (3%)	<0.001
Systolic blood pressure above age-based normal range	9/146 (6%)	48/606 (8%)	0.472
Abnormal pupils	4/142 (3%)	8/584 (1%)	0.264†
Mental status change	59/146 (40%)	184/609 (30%)	0.018
Pain or swelling over shunt	16/80 (20%)	65/360 (18%)	0.685
Diagnostic imaging			
None	3/146 (2%)	29/609 (5%)	0.145
Imaging			
Cranial CT	70/146 (48%)	267/609 (44%)	
Rapid cranial MRI	71/146 (49%)	293/609 (48%)	
Other	2/146 (1.4%)	20/609 (3.3%)	
ED disposition			
Discharge	3/146 (2%)	300/609 (49%)	<0.001
Admission	143/146 (98%)	309/609 (51%)	
Operating room (first ED encounter)	106/143 (74%)	15/309 (5%)	

*Median (interquartile range).

†Fisher exact test.

TABLE 2. Percentage Agreement and Inter-Rater Reliability of the High-Risk Clinical Predictors of Ventricular Shunt Malfunction

Predictor	% Agreement (95% CI)	κ Standard Estimates (95% CI)
Historical features		
Headache	96.9% (90.0–99.2)	0.87 (0.70–1.00)
Nausea and/or vomiting	98.6% (95.9–100)	0.97 (0.92–1.00)
Seizure	100% (95.2–100)	1.00 (n/a)

TABLE 3. Test Characteristics of the Ventricular Shunt Malfunction Clinical Prediction Rule for the Diagnosis of Ventricular Shunt Malfunction

Test Characteristic	n/N (%)	95% CI
Sensitivity	139/141 (98.6%)	94.4%–99.7%
Specificity	38/579 (6.6%)	4.7%–9.0%
NPV	38/40 (95.0%)	81.7%–99.1%
PPV	139/680 (20.4%)	17.5%–23.7%

TABLE 4. Risk of Ventricular Shunt Malfunction for Patients With No, 1, 2, or 3 Ventricular Shunt Malfunction Predictors

Ventricular Shunt Malfunction High-Risk Predictors Present	No. (%) Children Without Ventricular Shunt Malfunction (N = 579)	No. (%) Children With Ventricular Shunt Malfunction (N = 141)
No predictors	38 (6.6%)	2 (1.4%)
1 Predictor	246 (43%)	45 (32%)
Headache	123 (21%)	26 (18%)
Vomiting	64 (11%)	9 (6%)
Mental status change	59 (10%)	10 (7%)
2 Predictors	253 (44%)	65 (46%)
Headache, vomiting	170 (29%)	45 (32%)
Vomiting, mental status change	60 (10%)	13 (9%)
Headache, mental status change	23 (4%)	7 (5%)
3 Predictors	42 (7%)	29 (21%)
Total no. patients with ≥1 predictor	541 (93%)	139 (99%)

Malfunction

Riva-Cambrin et al, 2017 also looked at risk factors for malfunction in a multi-center prospective cohort
344/1036 experienced shunt failure, including 265 malfunctions and 79 infections

Three factors were were independently associated with reduced shunt survival

- Age younger than 6 months at shunt placement (HR 1.6 [95% CI 1.1–2.1])
- Cardiac comorbidity (HR 1.4 [95% CI 1.0–2.1])
- Endoscopic placement (HR 1.9 [95% CI 1.2–2.9])

No independent associations with shunt survival

- Etiology
- Where the surgery was done
- Valve design
- Use of ultrasound or stereotactic guidance
- Surgeon experience and volume

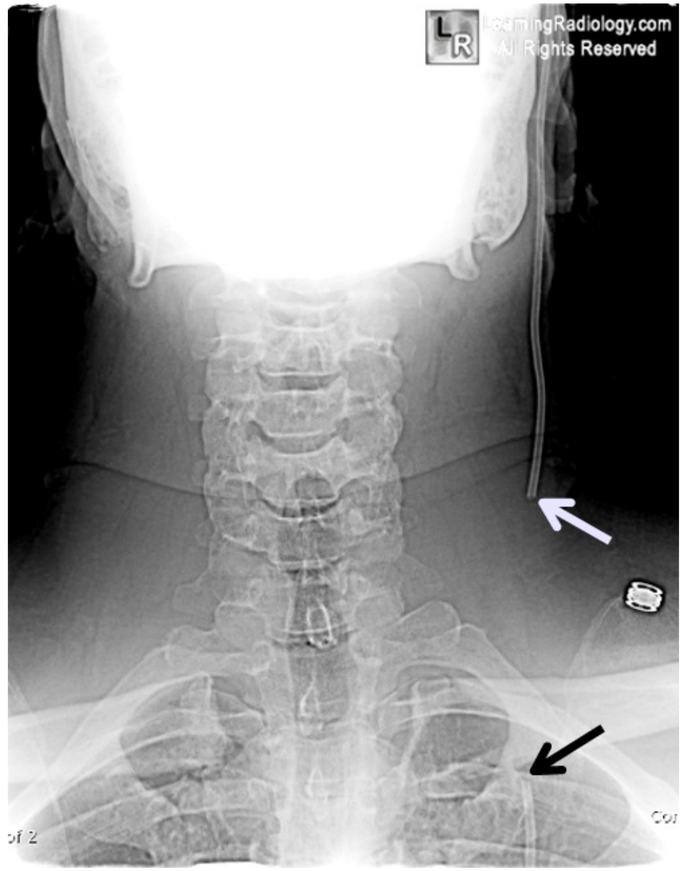
Malfunction

Workup includes **head CT** and **shunt series** + **Neurosurgery consult**

Malfunction

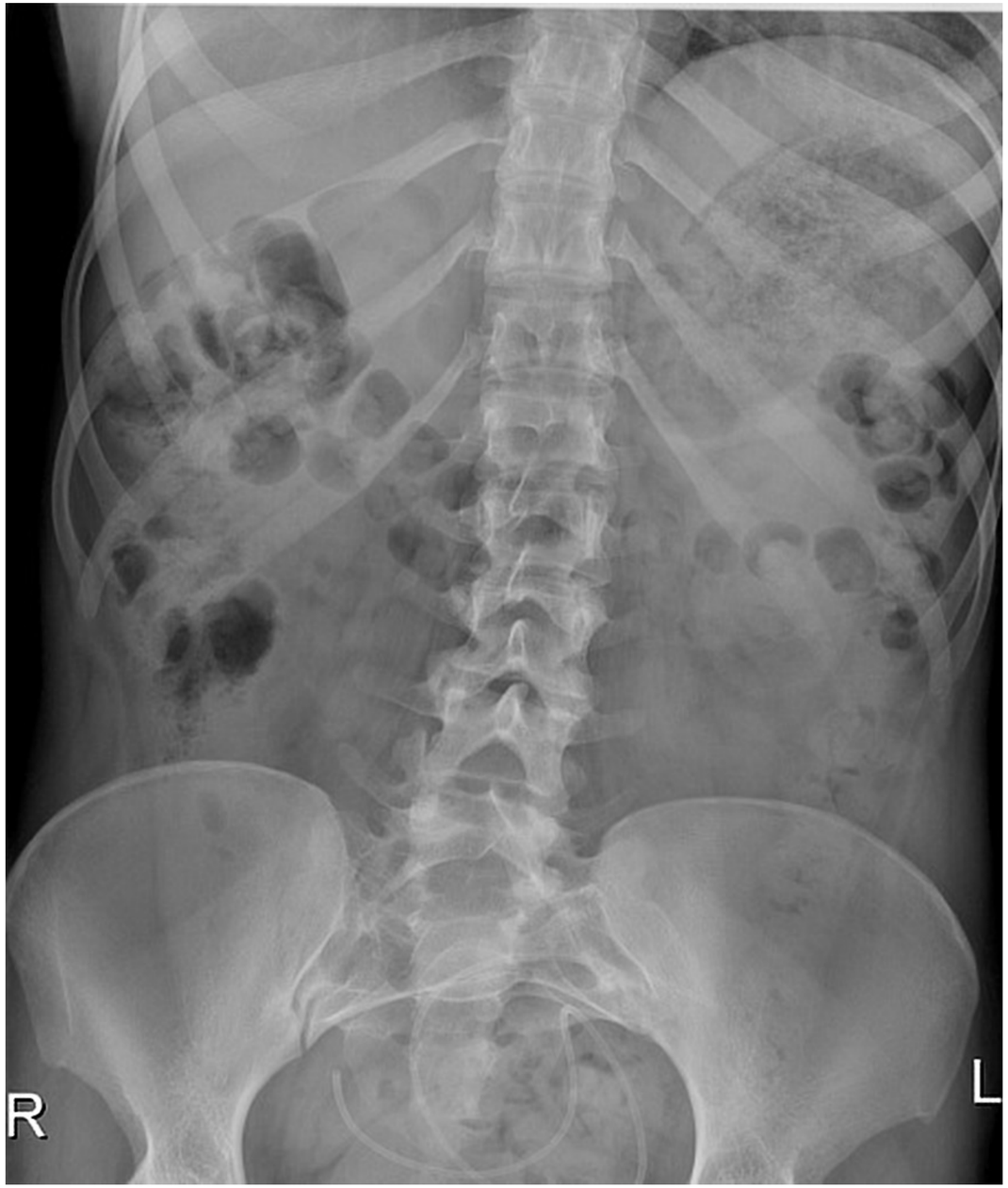
Shunt series

- Radiographs of the skull, neck, chest, and abdomen
- Look for mechanical breaks, kinks, and disconnections in the shunt - most common in the neck



From **Radiopedia**

The VP shunt on the right side of the neck, seen best on the lateral neck/skull and AP neck views appears discontinuous. The shunt is not seen on the chest or abdominal wall with the remainder of the tubing is noted coiled in the abdomen.



Malfunction

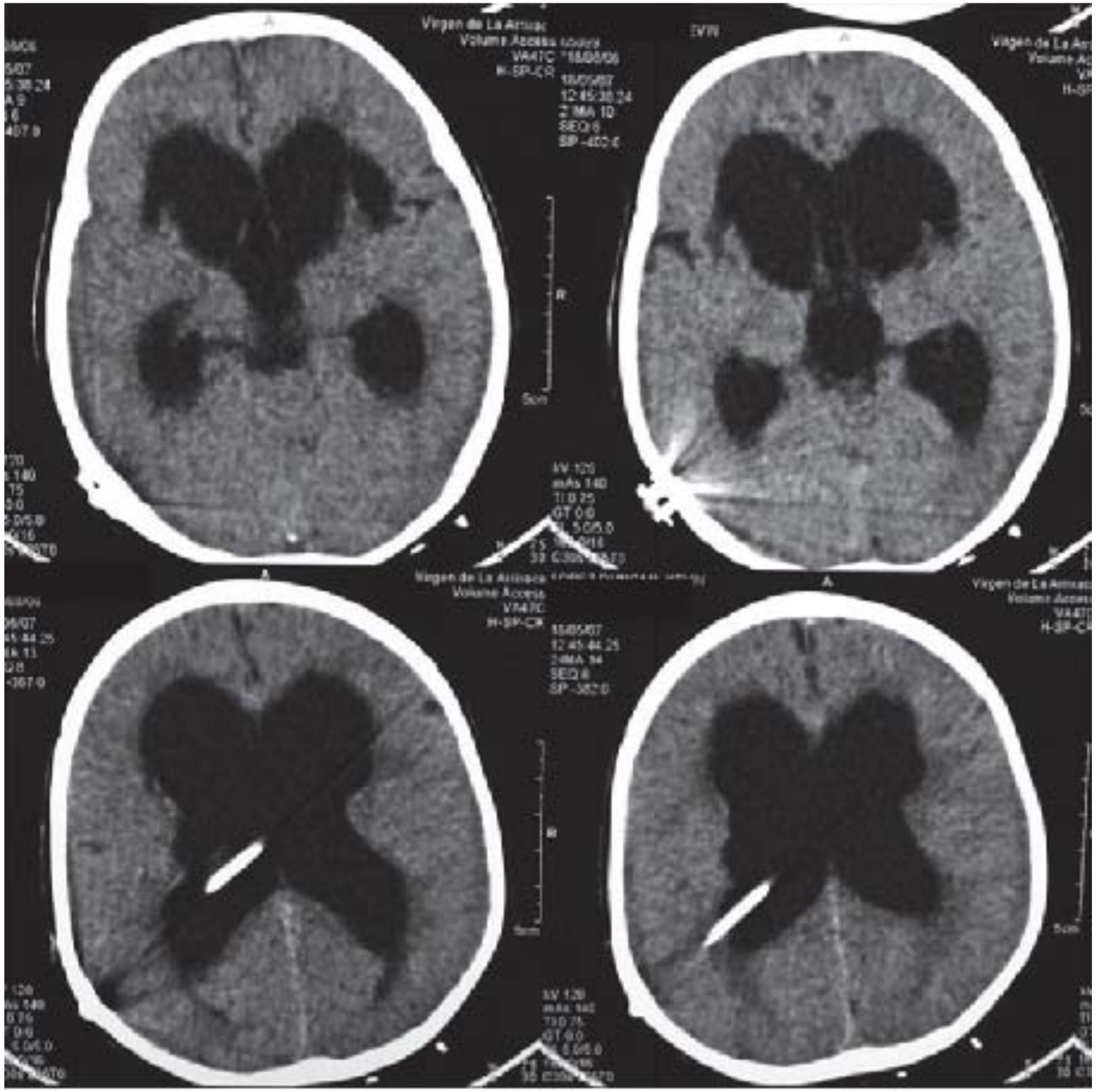
Pitetti, *Pediatr Emerg Care*, 2007 – Retrospective review of 291 kids (461 ED visits)

- 78% had a shunt series
- 15% (71/291) Dx with malfunction
  - 22 of these 71 had a normal head CT
  - 6 of these 22 had an abnormal shunt series

Head CT

Not always diagnostic - sensitivity 54-83%

Size of ventricles can help, but in up to ⅓ of cases of shunt malfunction the CT is nondiagnostic (especially Chiari II/MM)



Malfunction

Zorc, 2002

TABLE 1

Results of shunt series and head computed tomography scan and clinical outcomes

Radiographic results	Clinical outcome			
	Obstruction ($n = 60$)	No obstruction ($n = 173$)	Sensitivity	Likelihood ratio
Shunt series				
Findings associated with obstruction				
Disconnection of distal catheter	6	1	10%	17
Retraction of distal catheter tip	4	1	6%	5.5
Discontinuity near shunt bulb	2	2	3%	2.8
Any abnormality	12	4	20%	8.6
Findings not associated with obstruction				
Kink or coil in shunt tubing	1	7	3%	0.82
No tip movement from prior exam	2	12	3%	0.48
Head CT scan				
Increased ventricles since prior CT scan	29	8	48%	10.1
Possible shunt dysfunction	4	6	6%	1.9
No prior comparison CT scan	17	27	28%	1.8
Any abnormality	50	41	83%	3.5

CT = computed tomography.

Malfunction

MRI

May replace CT

Protocols exist for fast MRI scans

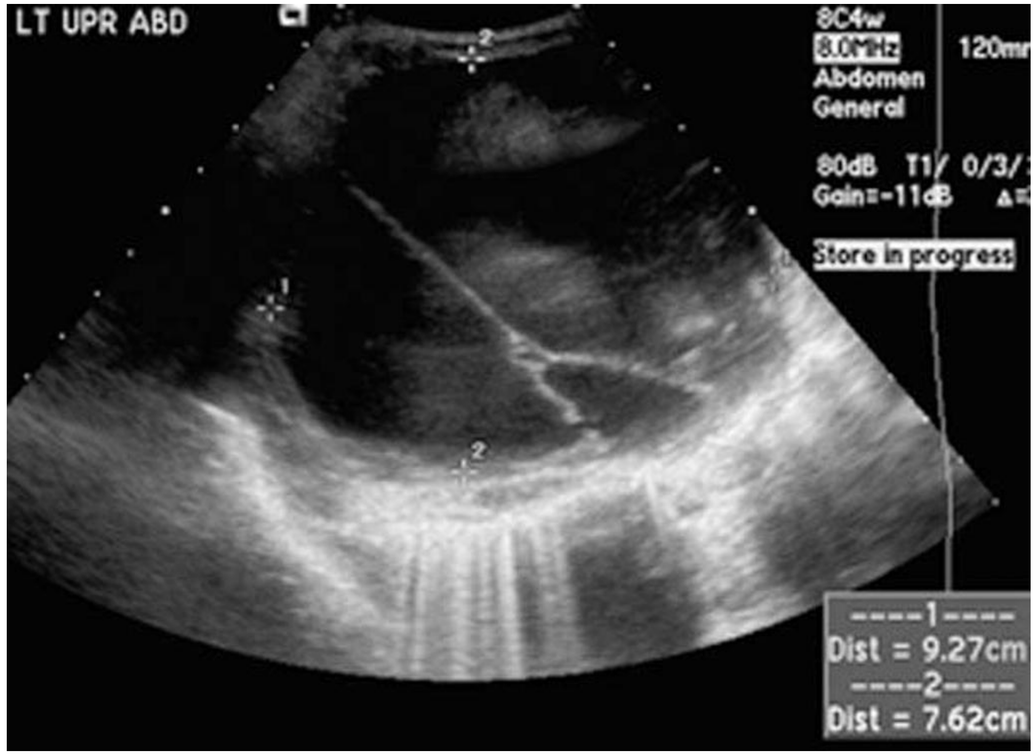
Malfunction

Abdominal Ultrasound

A pseudocyst is a false pocket in the abdomen at the distal end of the shunt

Fluid collects and may cause obstruction

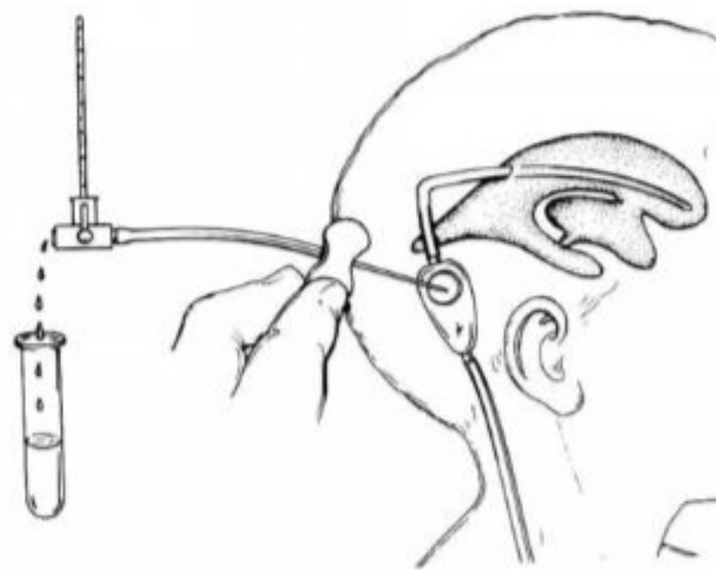
Consider in patients with GI symptoms and concern for shunt malfunction, but usually obtained at discretion of Neurosurgeon



Malfunction

Shunt Tap?

- Opening pressure >25cm H_2O associated with distal obstruction in 90%
- Poor flow associated with proximal shunt in >90%



Shunt Tap?

Contraindications

- Skin infection over shunt site
- Coagulopathy
- Lack of shunt imaging/info

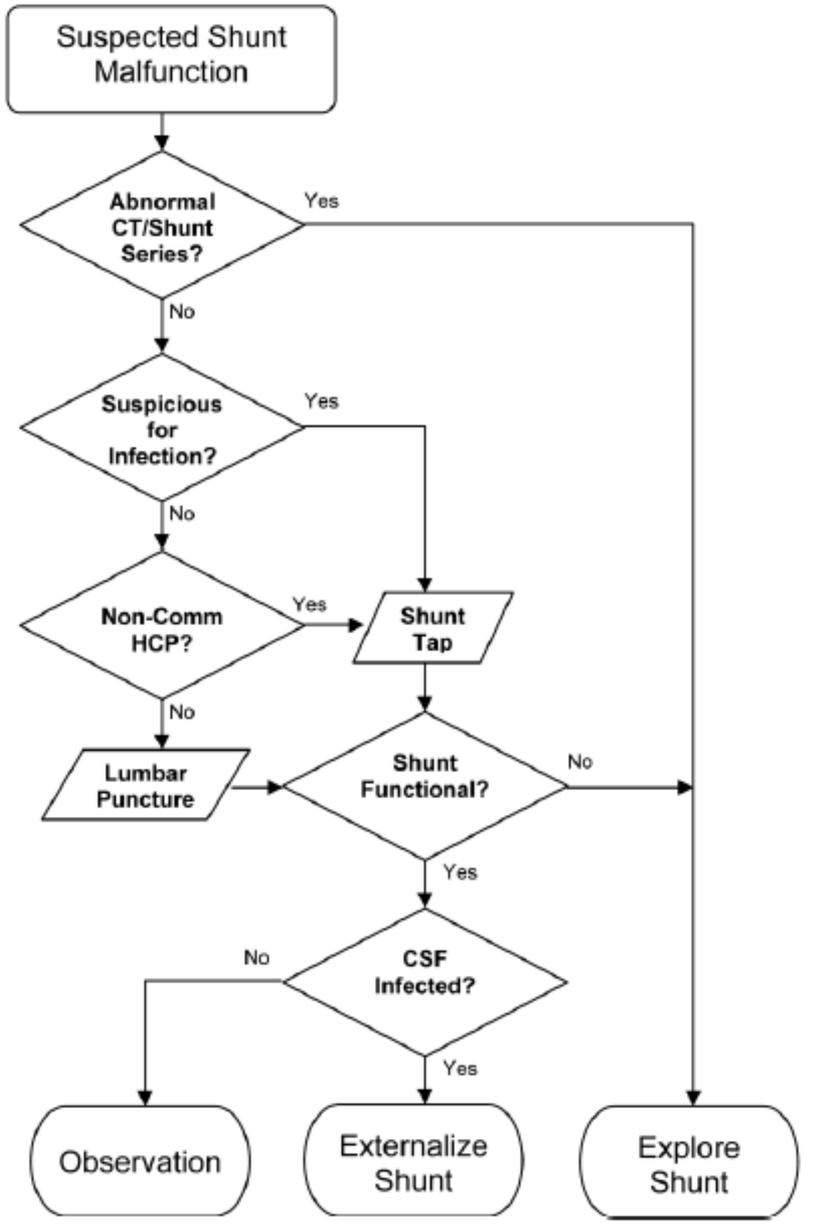


FIG. 1. Flow chart showing algorithm used for management of suspected shunt malfunction. Non-Comm HCP = non-communicating hydrocephalus.

Miller, J. Neurosurg Pediatrics 2008

Malfunction

ABCs

Head midline, elevated 30 degrees

Manage hypoxia (sats >95%), hypercarbia , hypotension, and hypoglycemia

Temperature control

Mild sedation (don’t cause hypotension)

Control severe shivering w/ paralytics

Prophylactic AEDs to patients at risk for seizures

3% Saline/Mannitol

No intervention is more important than a trip to the OR in shunt malfunctions!

Take Home Points

Shunts infections are far more common in the initial month after placement

Shunt malfunctions are usually mechanical, and proximal

Get a head CT and shunt series unless you can find another cause for the child's symptoms