The Common and the Uncommon: Abdominal Pain in a Child with a Ventriculoperitoneal Shunt

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Outline

• Our patient: Neonatal period
• Ventriculoperitoneal (VP) shunts
• Our patient: One month prior to presentation
• Our patient: Presentation with abdominal pain
• Diagnostic imaging for appendicitis in children
• Our patient: Imaging results and outcome
Our Patient: Neonatal History

• Intraventricular hemorrhage at birth of unknown etiology, which led to:
  – > Obstruction of cerebrospinal fluid (CSF) flow
  – > Hydrocephalus

• Ventriculoperitoneal (VP) shunt placed during neonatal period to relieve the hydrocephalus
Anatomy of Hydrocephalus: Ventricles
Anatomy of Hydrocephalus: CSF Flow

Direction of CSF Flow:
--> Lateral ventricle
--> Foramen of Monro
--> 3\textsuperscript{rd} ventricle
--> Cerebral aqueduct
--> 4\textsuperscript{th} ventricle
--> Foramina of Luschka and foramen of Magendie
--> Subarachnoid space
--> Arachnoid granulations
--> Dural venous sinuses
--> Systemic veins (e.g., internal jugular vein)

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VP Shunts: Function

• Devices that drain CSF away from the ventricles, usually into the peritoneal cavity
  – > Helps relieve excess CSF build-up in the ventricles and prevent abnormally high intracranial pressure
VP Shunts: Mechanics

• Proximal catheter drains CSF from the ventricles (Campellone 2013)
• Valve that controls how much CSF is removed from the ventricles per unit time (Lollis et al. 2010)
• Distal catheter tunneled underneath the skin usually to the abdomen, where it inserts into the peritoneal cavity (Campellone 2013)
  – Extra catheter distance in peritoneal cavity allows for child’s growth (Khan 2015)
VP Shunts: Clinical Issues

In technology-assisted children (e.g., with VP shunts or other devices), consider:

• Common childhood illnesses unrelated to the device
• Device infection
• Device malfunction
  – For VP shunts: Obstruction, catheter disconnection, abnormal positioning, CSF over- or under-drainage

Text: Fein et al. 2010.
VP Shunts: Symptoms of Shunt Pathology

• Neurologic
  – Altered mental status, headache

• Gastrointestinal
  – Abdominal pain, vomiting

Text: Fein et al. 2010.
VP Shunts: Signs of Shunt Pathology

• Fever

• **Cushing’s triad**
  – Increased intracranial pressure --> Hypertension, bradycardia, abnormal respirations

• Sunsetting eyes
  – Compression of midbrain structures that mediate vertical gaze --> Upward gaze paresis

• Signs of cellulitis overlying catheter tract

• Bulging fontanelle or increased head circumference

• Abdominal tenderness

• Abnormal neurologic exam or papilledema

Text: Fein et al. 2010.
VP Shunts: Radiological Work-Up

• Suspected VP shunt malfunction almost always requires diagnostic imaging
  – But there is not consensus as to imaging tests of choice in suspected VP shunt malfunction
VP Shunt Imaging: Cranial CT Scan

• Has been the most commonly used imaging modality for suspected ventricular shunt malfunction (Boyle et al. 2014)
• Cheaper than MRI
  – e.g., Patient charge of $1364 for a head CT versus $1428 for a rapid cranial MRI at one tertiary pediatric hospital (Boyle et al. 2014)
• Easier to access than MRI, especially at night (Boyle et al. 2014)
• **No risk of de-programming VP shunts**
  – MRI can alter the rate of CSF drainage of certain brands of VP shunts that are magnetically programmed. These shunts need re-programming after MRI to prevent CSF over- or under-drainage (Lollis et al. 2010).

VP Shunt Imaging: Cranial MRI

- **No radiation exposure**
  - Compared with adults, children are more sensitive to radiation, with a higher proportion of dividing cells and more life-years remaining in which cancer can develop (Brenner et al. 2007)

- **Rapid cranial MRI technology now exists**
  - Involves fewer MRI sequences with faster image acquisition time (as low as 8 seconds) (Woodfield et al. 2015)
  - Sedation generally unnecessary given minimal time in scanner (Woodfield et al. 2015)
  - Accuracy for diagnosing ventricular shunt malfunction is not inferior to that of cranial CT (Boyle et al. 2014)

Text: Boyle et al. 2014; Brenner et al. 2007; Woodfield et al. 2015.
VP Shunt Imaging: “Shunt Series” Radiograph

• Captures entire course of **VP shunt** from brain to peritoneal cavity
• May not be needed if other cranial imaging is also obtained
  – When both a cranial CT and a shunt series are obtained, the shunt series alters decisions about the need for neurosurgery in only ~1.5% of cases

Companion Patient #1: Example of a normal shunt series

Text: Vassilyadi et al. 2010.
Image: wikiRadiography website.
VP Shunt Imaging: Ultrasound

Images ventricles through an open anterior fontanelle

Companion Patient #2: Example of an infant undergoing an ultrasound examination

Companion Patient #3: Example of a shunt and enlarged ventricles (*) on ultrasound

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Our Patient: One Month Prior to Presentation

• 11-year-old female presented to emergency department 4 days after falling from a scooter and hitting her head. She had:
  – Headache
  – Pain and swelling behind ear over VP shunt site
  – No other symptoms or signs

• Given these symptoms and signs, there was concern for VP shunt malfunction and imaging was obtained
Our Patient: Enlarged Ventricles on Rapid Cranial MRI

C- axial T2 head MRI from 2 months prior to presentation with **ventricles at baseline size**

C- axial T2 head MRI from 1 month prior to presentation with **larger ventricles**

Images: Boston Children's Hospital.
Our Patient: VP Shunt Discontinuity on Shunt Series Radiograph

New 3 cm **discontinuity** in the VP shunt catheter in the left scalp region

Images: Boston Children's Hospital.
Our Patient: VP Shunt Surgery

• Given her VP shunt discontinuity, she underwent surgical revision of the shunt with placement of a new distal catheter
  – The surgery was uncomplicated
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Our Patient: Presentation with Abdominal Pain

• 11 year-old female presented to emergency department with abdominal pain during the 4 weeks since her VP shunt surgery
  – Worst in **right lower quadrant**, exacerbated by movement, worse in the 3-4 days prior to presentation
  – No fever, anorexia, nausea, vomiting, or diarrhea; last bowel movement the day prior
  – Exam notable for an abdomen that:
    • Was soft, non-distended, diffusely tender (especially in the right lower quadrant)
    • Had rebound and positive psoas sign, but no guarding
Our Patient: Differential Diagnosis

Differential was broad, and included:

- **Appendicitis**
- CSF pseudocyst
  - An abnormal, loculated CSF fluid collection at the tip of the VP shunt catheter in the peritoneal cavity
  - Fluid may become infected
- Renal/urinary and ovarian pathology
Companion Patient #4: Example of a CSF Pseudocyst on Ultrasound

Image: DeFlorio 2014.

Ultrasound showing a **CSF pseudocyst** surrounding multiple **shunt catheter loops (circled)**
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### Appropriateness Criteria for Appendicitis

#### Diagnostic Imaging in Children

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>US abdomen</td>
<td>8</td>
<td>Perform this procedure with graded compression.</td>
<td>O</td>
</tr>
<tr>
<td>CT abdomen and pelvis with contrast</td>
<td>7</td>
<td>This procedure may be useful following negative or equivocal US. Oral or rectal contrast may not be needed depending on institutional preference.</td>
<td></td>
</tr>
<tr>
<td>X-ray abdomen</td>
<td>6</td>
<td>This procedure may be useful in excluding free air or obstruction.</td>
<td></td>
</tr>
<tr>
<td>US pelvis</td>
<td>5</td>
<td>This procedure is appropriate in women with pelvic pain.</td>
<td>O</td>
</tr>
<tr>
<td>CT abdomen and pelvis without contrast</td>
<td>5</td>
<td>Use of oral contrast depends on institutional preference.</td>
<td></td>
</tr>
<tr>
<td>MRI abdomen and pelvis without and with contrast</td>
<td>5</td>
<td>See statement regarding contrast in text under “Anticipated Exceptions.”</td>
<td>O</td>
</tr>
<tr>
<td>MRI abdomen and pelvis without contrast</td>
<td>4</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>CT abdomen and pelvis without and with contrast</td>
<td>3</td>
<td></td>
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</tr>
<tr>
<td>X-ray contrast enema</td>
<td>2</td>
<td></td>
<td></td>
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<tr>
<td>Tc-99m WBC scan abdomen and pelvis</td>
<td>2</td>
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</tr>
</tbody>
</table>

**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level

Table: ACR 2013.
Pediatric Appendicitis Imaging: Ultrasound

• No radiation

• Sensitivity of 88% and specificity of 94% (Doria et al. 2006)
  – Similar to CT sensitivity of 94% and specificity of 95% (Doria et al. 2006)

• **Initial imaging of choice** (ACR 2013)
Pediatric Appendicitis Imaging: CT Scan

- Among females ages 5-14 “a radiation-induced solid cancer is projected to result from every 300 to 390 abdomen/pelvis [CT] scans” (Miglioretti et al. 2013:700)
- Consider obtaining a CT scan if ultrasound findings are equivocal (ACR 2013)
Pediatric Appendicitis Imaging: Abdominal Radiograph

- Shows free air and obstruction (ACR 2013)
- Shows findings that can be associated with appendicitis, e.g.:
  - An appendicolith (Lieberman 2005)
  - A sentinel loop of bowel, which is a focal ileus in an area of inflammation (Lieberman 2005)

Pediatric Appendicitis Imaging: MRI

• Not commonly used, given availability, potential need for sedation, and cost
Companion Patient #5: Example of Appendicitis with Hyperemic Appendiceal Wall on Doppler Ultrasound

Incompressible, blind-ended, fluid-filled structure, with diameter $> 6$ mm and thickened/hyperemic wall

Image: Gaitini 2011.
Companion Patient #6: Example of Appendicitis with Appendicolith on Ultrasound

Enlarged appendix with appendicolith apparent as an echogenic focus with adjacent acoustic shadowing (*)

Image: Rybkin et al. 2007.
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Our Patient: Appendix Not Seen on Abdominal/Pelvic Ultrasound

Right lower quadrant view with **appendix not visualized**

Images: Boston Children's Hospital.
Our Patient: Other Findings on Abdominal/Pelvic Ultrasound

• Large stool burden in rectum
• Unremarkable kidneys, bladder, uterus, and ovaries
• VP shunt tip not definitely identified, but no appreciable loculated fluid collection suggestive of CSF pseudocyst
Our Patient: Clinical Reassessment

• Patient’s status after her ultrasound
  – Afebrile with normal vital signs
  – Abdomen soft without rebound or guarding
  – Labs (CBC, chem-7, LFTs, amylase, lipase, urinalysis) unremarkable

• **Clinical suspicion** for appendicitis or other acute abdominal/pelvic pathology was **low** enough to defer CT scan

• Abdominal radiograph was obtained mainly to evaluate further for obstruction, constipation, and VP shunt pathology
Our Patient: Abdominal Radiograph

There are 3 main findings on our patient’s upright and supine abdominal radiographs. Let’s look at these findings one by one.
Our Patient: New VP Shunt Catheter on Abdominal Radiographs

New VP shunt distal catheter placed 1 month prior, which does not have discontinuities or kinks.

Images: Boston Children's Hospital.
Our Patient: Old VP Shunt Catheter on Abdominal Radiographs

There is a piece of the patient’s old VP shunt distal catheter, which does not appear to extend past the T8 vertebral level. This free piece of catheter was calcified and could not be removed during her surgery 1 month prior.

Images: Boston Children's Hospital.
Our Patient: Stool on Abdominal Radiographs

There is no bowel obstruction, but the right colon contained a moderate-to-large amount of stool.

Images: Boston Children's Hospital.
Our Patient: Summary of Findings on Abdominal Radiographs

Images: Boston Children's Hospital.
Our Patient: Diagnosis and Outcome

• She was discharged from the emergency department with a diagnosis of constipation and instructions for the outpatient treatment of this condition.
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Summary of Learning Points

We reviewed the:

• Anatomy pertinent to hydrocephalus

• Function and malfunction of VP shunts
  – And the warning signs of shunt malfunction

• Imaging options for diagnosing VP shunt malfunction
  – And the radiological appearance of VP shunt pathologies on ultrasound, radiograph, and cranial MRI
Summary of Learning Points Continued

We reviewed the:

• Imaging options for diagnosing appendicitis
  – And the radiological appearance of appendicitis on ultrasound

Finally, we were reminded that common diagnoses (e.g., constipation) are indeed common.
References


• DeFlorio RM, Shah CC. Techniques that decrease or eliminate ionizing radiation for evaluation of ventricular shunts in children with hydrocephalus. Semin Ultrasound CT MR. 2014;35:365-373.


References Continued


Acknowledgements

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