Welcome to the DCMC Emergency Department Radiology Case of the Month!

In conjunction with our pediatric radiology specialists from ARA, we hope you enjoy these monthly radiological highlights from the case files of the Emergency Department at DCMC. These cases are meant to highlight important chief complaints, cases, and radiology findings that we all encounter every day.

If you enjoy these reviews, we invite you to check out Pediatric Emergency Medicine Fellowship Radiology rounds, which are offered quarterly and are held with the outstanding support of the pediatric radiology specialists at Austin Radiologic Association.

If you have any questions or feedback regarding the Case of the Month format, feel free to email Robert Vezzetti, MD at rmvezzetti@seton.org.

This Month: BOO!! This month, a young girl with constipation... or is it? This frightening case was brought to us courtesy of Mike Gardiner, MD, one of DCMC’s outstanding PEM Fellows. Enjoy..if you dare!

Conference Schedule: October 2016

5th 9:15 - PEM Radiology: Limping..Drs Vaidya, Huseini, Vezzetti
10:15 - Biostats 2: Inferential statistics..................Dr Wilkinson
11:15 - 12:15
12th 8:00 - Pediatric Multisystem Trauma..................Dr Vezzetti
9:00 - Trauma Sim............Drs Pittman, Vezzetti, Sim Faculty
14th - PEM Fellow Applicant Interview Day!
19th - 9:15 Research: Stats..........................Dr Gardiner
10:15 FAST and Lung US......................Drs Levine, Gorn
12:00 DCMC ED Staff Meeting
21st - PEM Fellow Applicant Interview Day!
26th - 9:15 - M&M..................Drs Schwartz, Vezzetti
10:15 - Board Review: GU Emergencies..............Dr Whitaker
12:15 Research Update..........................Dr Wilkinson

Simulations are held at the CEC at UMC Brackenridge.

Lectures are held at DCMC Command Rooms 3&4.

Locations subject to change.

All are welcome!
Case History

Well, the ED volume is picking up as school is in full swing. You are seeing the usual types of cases typical for this time of year, including falls off monkey bars, head injuries/concussions, and the usual stomachs. No end in sight it seems. That is, until you pick up the next chart.

The chief complaint is listed as abdominal pain and constipation. Not too unusual in the Emergency Department. You walk in to find a 12 year old female who is no acute distress but complains of generalized abdominal pain. The history you obtain is interesting. She has had generalized pain for the past 10 days. Accompanying this pain is difficulty passing stool. She tells you that she has had hard stools consistently during this time; there is pain associated with bowel movements but she denies rectal bleeding or bloody stool. Her mother is with her and she confirms this. There is no history of fever, vomiting, diarrhea, travel or trauma. She denies dysuria or hematuria. This seems pretty straightforward.

You begin your exam and note the vital signs: Temp 98 HR 89 RR 16 BP 110/55. You see a well-appearing child. Her exam is not impressive until you begin the abdominal exam. You note that, while there is no hepatosplenomegaly or obvious masses, she does complain of tenderness, primarily to the bilateral lower quadrants and the suprapubic area. The child also has prominent abdominal distention with possibly palpable stool. The exam, admittedly is complicated by not only the distention, but the fact that she appears very uncomfortable whenever you attempt to palpate the abdomen.

You decide to delve a little deeper into the history. The child has always been healthy and has never had issues with constipation in the past. She denies any recent weight loss and has no other symptoms such as rashes, joint pain/swelling, or decrease in appetite. She has not begun menses yet and denies vaginal discharge or bleeding. She is not sexually active. There is no history of incontinence, weakness, or numbness of the extremities, particularly the lower extremities.

Since she has provided a urine sample, you check a urinalysis which is normal and a urine pregnancy test, which is also normal. The distention is bothersome, so while you think this is probably constipation, perhaps an imaging test may be helpful. While you try to think of what would be indicated, you think about the differential of a child with abdominal distention and reported constipation.

Abdominal distention can have many causes, and while extensive constipation can certainly cause distention, there are other diagnosis to consider. Certainly bowel obstruction can present with pain and distention, but one would expect emesis and 10 days is quite a length of time to have a bowel obstruction, which is usually an acute condition in pediatrics. So, one should consider masses, neurologic causes, and obstetrical/gynological issues as well. Granted, these are more rare, but keep them in the back of your head when evaluating a child with abdominal distention.

It may not be pleasant to think about, but always obtain a beta HCG (urine HCG) in any female who has begun menstruation or females above the age of 10 years. Some of these are positive, and now a whole new conversation has to take place!
Abdominal Plain Radiography

Advantages:
- Inexpensive.
- Available.
- Portable.
- Low radiation.

Disadvantages:
- Nonspecific.
- Limited tissue differentiation
- Sometimes difficult to interpret

Most commonly, this imaging modality is employed when there is clinical concern for obstruction, a swallowed foreign body, or there is vomiting or abdominal distention. With trauma or concerns for intestinal perforation, free air can be seen. In the cases of neonates or very young infants, if necrotizing enterocolitis is in the differential, then intestinal pneumatosis can be seen.

Image Selection With Abdominal Radiography

Supine View - obtained by a vertical beam directed at the patient. This includes (or should) the entire abdomen, from the diaphragm to the symphysis pubis.

Upright View - obtained by a horizontal beam directed at the mid abdomen while the patient is standing.

Decubitus View - this is an an AP view where the patient is lying on the left side, ideally for a few minutes. A horizontal beam is directed at the patient's mid abdomen. This is typically done with the left side down, because the liver edge provides a good congrats for detecting free air. This view is very helpful for detecting bowel obstruction, especially if it is not apparent on AP views of the abdomen (intussusception, for example).

Chest Xray - this is added sometimes to look for free air under the diaphragm and is part of an acute abdominal series.

Reading a Plain Abdominal Radiograph

Just like any imaging study, it is helpful to have a system in mind when reading abdominal films. Here's one example:

1. Adequacy - is the film adequately penetrated? do you have the correct views?
2. Free Air - look under the diaphragm in an upright film or AP chest view.
4. Other Gas - retroperitoneal gas, intramural gas.
5. Soft Tissues - check out the solid organs, posses margins, masses, diaphragm.
6. Bones/calcifications - looks at the ribs, spine, and looks for calcifications (gallstones or genitourinary stones can be seen sometimes, as can appendicoliths).
7. Other - foreign bodies and other radiolucent material (PeptoBismol can be seen on radiographs).
Well, you decide to obtain plain radiography of the abdomen, given the distention. This certainly can be helpful in assessing the bowel gas pattern, the presence of constipation, and sometimes one can pick up abdominal masses, although this is not the best imaging modality if that is your objective.

Thriller is probably the most famous music video of all time, at least according to the Library of Congress. It is by far the best selling album in the world. In the United States, it was overtaken by The Eagles Their Greatest Hits 1971-1975, but reclaimed the title after Jackson’s death.

Here is a nice example of an abnormal abdominal film in a young pediatric patient. This child presented with non-bilious emesis and intermittent fussiness. A 2 view abdominal series was obtained, which at DCMC is a supine view with a left lateral decubitus view. The most striking finding on this view is the bowel gas pattern, which is abnormal: the loops of bowel are dilated and stacked, indicating obstruction (red arrow). This child was diagnosed with intussusception. Interestingly, you can see the intussusceptum (blue arrow).

Well, there is a moderate amount of stool noted (red arrow) with a non-obstructive bowel gas pattern. The lung bases are clear. There is no free air or pneumatosis seen. The bones visualized look normal. Was this the cause of the child’s pain?

The film to the left demonstrates dilated bowel loops (red arrows) and pneumatosis (yellow arrows). This child was diagnosed with necrotizing enterocolitis.
You decide to try an enema to see if this will help with the child's pain; this appears to be successful, as the child has two large bowel movements and reports that she feels better. You then go back to re-examine the patient. She appears much more comfortable and would like to go home. On re-examination, however, you note that the child has a large abdominal mass, located at the suprapubic area. This was not easy to detect during your first exam, but you can feel it now. What is this? Does this have anything to do with the child's presenting symptoms? What is the cause of the abdominal mass? Could this be further retained stool? Perhaps something more serious, such as a tumor. Maybe urinary retention (a well-known complication of constipation). Pregnancy, but she had a negative Beta HCG.

You decide to pursue further imaging to delineate the nature of this mass. You could re-image with plain radiography, but at this point one should consider if perhaps a different modality is needed. You could obtain a CT scan, of course, this is radiation. She may need a CT at some point, but the other modality to consider is ultrasound…available, pain free, radiation free, and this can give us a starting point to figure out what is happening with this child.

To the right are selected ultrasound images of this patient. Of note, there is a large, dilated structure in the midline (red arrow) with diffuse internal echoes (blue arrow). This is suggestive of blood filling the uterus. The ovaries (not seen on these images) are normal and there are no associated masses. Arterial and venous Doppler waveforms were normal.

Wow. This fluid collection is consistent with hematometra. The differential for this is limited; typically the most common physical exam finding associated with this is imperforate hymen. Transverse vaginal septum and vaginal atresia may also be associated with this finding. Time to go back and re-examine the child.

Vincent Price narrated the end of Thriller, including the menacing laugh. The line “Must stand and face the hounds of hell” was inspired by Sir Arthur Conan Doyle’s novel, The Hound of the Baskervilles, in which Price had starred in a movie version of the novel.

Vincent Price, while a guest on the Johnny Carson's Tonight Show, laughingly stated that when he did the narration for “Thriller” he had a choice between taking a percentage of the album sales or $20,000; he took the $20,000. Carson told him he could have made millions off of the royalties due to the vast number of copies sold even at that time. Price laughed heartily and said: “How well I know!”
Repeat physical examination is necessary in this child. Specifically, a genital exam needs to be done. You do this, after explaining to the mother and patient what needs to be done. You find a Tanner Stage IV child. The labia major are normal. You are unable to visualize an introitus. There appears to be a very small opening at 3 o’clock but you cannot see into the vaginal tract. You do not appreciate a bulging hymen. Interestingly, you also notice, when you are done, that the child has a hypo plastic right thumb. Now what?

It is obvious that this child has a genital issue that is preventing her from having menses; perhaps a GYN consult is in order.

Amenorrhea
The diagnosis of primary amenorrhea is made when a female has reached the age of 15 years and has normal growth and secondary sexual characteristics, but has not yet had menses. It is also considered if, by age 13, there is no menses and a lack of secondary sexual characteristics.

Causes:
Gondola dysgenesis (Turner’s syndrome): Most Common
Mullein dysgenesis
Physiological delay of puberty
Polycystic ovarian syndrome
GnRH Releasing dysfunction
Transverse vaginal septum
Anorexia/weight loss/athletes
Hypopituitarism
Imperforate hymen (< 1% of cases)
Pituitary tumors

One of the first steps in evaluating primary amenorrhea is an ultrasound, looking for a uterus or hematometra/hematometrocolpos, in addition to bloodworm (FSH, LH). Out patient does not meet the criteria for primary amenorrhea (she is 12) but there is clearly something wrong, since her ultrasound reveals hematometra. GYN is consulted and they recommend an MRI. This modality will visualize the pelvic organs and anatomy quite well.

In these selected MRI images there are some interesting findings. The images to the left, above, and the following ones on the next page demonstrate hematometrocolpos (red arrow); there is a longitudinal vaginal septum (yellow arrow) that separates the vaginal into two hemivaginas (blue arrow); the lower portion of the vagina is also filled with blood products, suggesting communication to either hemivagina, or both (white arrow). The right adnexa demonstrates a dilated, tortuous right fallopian tube, also containing blood products (green arrow). The left uterus is normal size; the right is either atretic or nonexistent. The lower vagina area look normal. The right kidney (not seen in these images) is difficult to identify and may be absent. There is also no identifiable distal sacrum and coccyx (purple arrow). Lots of anatomy issues here! What happened to the patient? Read on…

The song “Mr Crowley” by Ozzy Osborn, is about Aleister Crowley, a practitioner of black magic and once called “The Wickedest Man Alive”
Female reproductive tract development is a complex process and, let's face it, lots can go wrong. A detailed review of the whole developmental process is beyond the scope of this newsletter, but here is a very brief review. The process begins at week 3 of gestation and continues into the second trimester of pregnancy. The tract is derived from the muellerian ducts and the urogenital sinus. The uterus and fallopian tubes are derived from the are derived from this structure, as is vagina. Gonadal development occurs independently of the genital tract. An important point to remember is that the development of the genital tract is closely related to the development of the urinary system.

Abnormalities of the Vagina

Transverse Vaginal Septum
Failure of fusion of the urogenital sinus and the muellerian ducts. This is not a common occurrence: approximately 1 in 30,000-80,000 woman. Most septa are found in the upper portion of the vagina. Externally the patient appears normal; the vagina is shortened. Hematocolpos develops and a mass may be palpable.

Longitudinal Vaginal Septum
This condition is usually associated with uterine anomalies, including a septate uterus. Patients with this anomaly may have difficulty with tampon insertion or uterine bleeding. Some patients may be asymptomatic.

Obstructed Hemi-Vagina
This is typically associated with ipsilateral renal agenesis. Treatment for this condition is surgical correction.

Vaginal Agenesis
This is also known as Mayer-Rokitansky-Kuster-Hauser syndrome. This is congenital absence of the vagina. The uterus is often not developed or poorly developed as well. Interestingly, the WNT4 gene seems to play a roll in female genital development, and a mutation in this gene may cause this condition. This condition is also associated with other anomalies, such as renal agenesis (isilateral), horseshoe kidneys, and skeletal anomalies (typically the extremities). Treatment is controversial, because there is no agreement about the best method for surgical correction.

Oh, by the way.....I hate it when parents say that! Remember the MRI read about the right kidney? The child's mother (AFTER the read), mentioned that the child was diagnosed with a “missing kidney” as an infant! Are you kidding me?! Makes sense, though...
Teaching Points

1. The differential diagnosis for female abdominal pain is vast! History is extremely important, with particular attention paid to onset (acute or chronic), degree of pain, and associated symptoms.

2. Always consider ovarian causes of pain in females, particularly females of adolescent age or females who have already had menses. Ovarian torsion (discussed in another edition of the newsletter), is an extremely time sensitive diagnosis and should be considered in cases of acute abdominal pain.

3. Always obtain a Beta HCG and complete sexual history on adolescent females. Beta HCG’s are typically obtained in females over the age of 11-12. It is important to rule out pelvic inflammatory disease and other causes of sexually transmitted diseases in any female who is sexually active. It is best to obtain this history without the parent in the room and ensure the patient that this history is obtained in complete confidence.

4. Pelvic masses have a broad differential as well and this can be narrowed by location, symptoms, and imaging.

5. Ultrasound is a great initial imaging test when evaluating females with amenorrhea, suspected pelvic masses, suspected abdominal masses, and is easily obtained.

REFERENCES

2. Lauder M. Diagnosis and management of congenital anomalies of the vagina. In Up To Date. 2015.