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 check out Pediatric Emergency Medicine Fellowship Radiology Rounds, which are currently 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: DON'T BE FooLED BY THese casEs!**

**Case 1:** A 14 month old child with an ever increasing abdominal size; the parents think “she’s just getting fat”, but now they’re a little worried after looking up “possibilities” on the internet.

**Case 2:** A 4 month old child also appears to be having some tummy trouble. Her abdomen has been increasing in size. Two in a row?

According to early Roman the calendar, April was the second month but became fourth when they started to use January as the first month. The name originally came from Aprilis, meaning ‘to open’.

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**PEM Fellow Conference Schedule April 2015**

1st - 9AM-10AM Administrative Meeting
10AM-11AM Biostatistics 4..........................Dr Wilkinson
11AM-12PM Toxicology Rounds..................Drs Earp and Mitchell

8th - 9AM-10AM Psychiatric Emergencies.......Guest and Dr Wallin
10AM-11AM Radiology Rounds (Back Pain).....Drs Vezzetti and Gardiner
11AM-12PM M&M..................................Drs Remick, Yee, and Gregg

14th Journal Club.................................................................TBD

15th - 9AM-12PM Poster Presentation, SOC Workday

22nd - 9AM-10AM Biostatistics 5..........................Dr Wilkinson
10AM-11AM Hemoglobinopathies...............Drs Whitaker and Rencher
11AM-12PM....Post-op Heart Patient...Guest Lecturer

29th - 9AM-10AM Adult Emergencies in the PED..........................Dr Higginbotham
10AM-12PM Board Review..............................Dr Earp
12PM-1PM ED Staff Meeting

All Lectures are at Dell Children’s Medical Center in Command Rooms 3&4, unless otherwise specified.

Simulations are held at the CEC at University Medical Center - Brackenridge.

Schedule subject to change.
Case 1: It's a fairly quiet shift in the ED one afternoon (never say the Q word in the Emergency Department) when you pick up the next chart waiting on the slowly expanding rack. It's a 14 month old female with abdominal distention. You greet the family and notice a very cute, playful child who does not appear to be in any acute distress. The family tells you that they have noticed this child's abdomen slowly appear to get bigger over the past 3–4 months, but in the last 4 weeks, the change has been really noticeable. They deny any fever, vomiting, difficulty breathing, weight loss, travel, change in bowel habits, or, for that matter, really anything else. Just the bigger belly. She is a healthy child, born at term, although they do say she had a "brain cyst" on a prenatal ultrasound that seems to have vanished. Her vital signs are unremarkable; she is not febrile, and has age appropriate heart rate, respiratory rate, and blood pressure. Her physical examination is also unremarkable, except for one concerning finding. While she does not appear to have any hepatosplenomegaly, tenderness, guarding, or rebound on her abdominal exam, she does have abdominal distention. This is pretty noticeable and is seen on the right side, between the upper and lower quadrants. What's more, she has a palpable, firm, and nontender mass to the right side and it appears that this mass extends or at least involves the right cava area. Hmmm.....

As you are contemplating what to do next, a bad trauma rolls in and you have to get down to that trauma bay and start resuscitation. The nurse asks you if you want to get any labs or imaging started while you are the trauma team are dealing with the new patient. Thoughts?

Case 2: A 4 month old female comes in for evaluation of non bilious emesis and abdominal distention. Her history is remarkable for being an ex–34 week premie, who spent a couple of weeks in the NICU for feeding issues, which have since resolved. The parents report that they have noticed primarily right sided abdominal distention for the past month or so. There has been no prior history of vomiting, but this has started over the past 1–2 days. There is no history of fever, cough, diarrhea, recent trauma or travel. They report that the child is formula fed and has been vigorously feeding her usual amount. They also report good urine output.

On exam, the child has normal for age vital signs and is afebrile. She appears vigorous and smiles at you. Her exam is unimpressive, with one very obvious and notable exception. She has rather large and disturbing abdominal distention, primarily on the right side of the abdomen. As you palpate the area, you note that there is what appears to be a solid mass, mostly felt in the lower portion of abdomen. The child does not appear to be bothered by palpation of the mass: there is no fluctuance or erythema. There is also no crepitus. You find that the spleen feels normal but it is very difficult to palpate the liver edge, as the distention extends from the right lower quadrant of the abdomen to the right upper quadrant...at least it appears to extend that far.

What's going on here? You start to consider the differential diagnosis of abdominal masses in infants, which is easy because you just did it for the 14 month old from case 1! Time to obtain some imaging...what modality are you going to employ?
**Case 1:** Get things started, huh? This child has a concerning history and physical examination findings for some sort of abdominal mass. What next? Well, let’s see: Labs? You bet! A CBC, CMP, LDH, Uric Acid, and UA were all obtained.

Imaging? Sounds like a good idea. Lots of options here:

**Plain films** are a very quick and easy way to get a view of the bowel gas pattern, look for free air, and see if there is any displacement of the solid organs of the abdomen. You might also catch a glimpse of the suspected mass. But, they will be nonspecific and, in the end, more detailed imaging is likely to be needed.

**Ultrasound** is a great and very frequently used imaging modality. Of course, we know that it is painless and radiation free. It could be helpful in getting a quick view of the nature of the suspected mass (i.e., solid or cystic, fluid filled) and a rough location of the mass. You could also get a view of the solid organs of the abdomen and see if the mass either originated from them or involves these structures in some way. But, it is very likely more detail will be needed regarding the anatomic nature of this mass and ultrasound will not provide that level of detail.

**CT** is a great imaging test for abdominal pathology in general and suspected masses in particular. Of course, this involves IV contrast and radiation. In some children, sedation may be needed but not likely, as an abdominal CT is extremely quick. Aside from those drawbacks, CT will provide an excellent level of anatomic detail that will be essential in determining the nature of the mass (and the diagnosis) and possibly guide future studies and/or therapy.

**MRI** is also a viable option and a very effective imaging modality. Excellent anatomic detail can be achieved. Trouble is, and MRI exam of the abdomen is time-consuming and sedation is very likely going to be required. Also, there is an availability issue. An emergent MRI can certainly be obtained, but such a test is not necessarily indicated at this time, especially since other modalities are readily available.

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**So what could be causing an abdominal mass in a child this age? Here’s a handy guide to help sort things out:**

**Most Common Malignant Abdominal Masses In Children:**
- Neuroblastoma
- Wilms Tumor
- Hepatoblastoma
- Lymphoma
- Germ Cell Tumors
- Rhabdomyosarcoma

**Adrenal Neuroblastoma**

**Most Common Benign Abdominal Masses In Children:**
- Renal Masses
- Hydronephrosis
- Multicystic Kidney
- Mesoblastic Nephroma
- Ectopic/horseshoe kidney

**Genitourinary**
- Ovarian cyst
- Bladder distention
- Hematocolpos/Hematometrocolpos

**Liver Masses**
- Hemangioma

**Gastrointestinal**
- Mesenteric cysts, Gut duplication, Choledochoal cyst, Intussusception

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Memorable April Fool’s Day Pranks

1985 - Sports Illustrated runs a 14-page story by George Plimpton about a Mets pitching phenom named Sidd Finch. The reclusive, skinny Finch has a 168-mph fastball (which he credits to meditations in Tibet) and a host of quirks including carrying a French horn at all times and wearing only one hiking boot while pitching.
Memorable April Fool’s Day Pranks That weren’t Pranks!
1950 - When fire station No. 2 in High Point, North Carolina received an emergency call from fire station No. 1, they decided not to respond, thinking it must be an April Fool gag. They were incorrect. It turned out that fire station No. 1 really was on fire. An explosion had resulted in flames which damaged their truck.

Case 1: After careful consideration of all the risks/benefits of the imaging modalities that are available, you elect to perform a CT scan. This child is obviously stable at this time and does not have any physical or historical signs that would make an immediate plain radiograph necessary (to rule out free air or obstruction, for example. Here is the result:

Well, there certainly is something here that should not be here!

The CT was obtained with IV contrast (for a detailed discussion on which CT studies need contrast and which do not, see the February 2015 edition of the newsletter).

There is an obvious, large mass noted on this study (red arrow). It is seen in the lower portion of the abdomen, appearing to involve the right kidney (yellow arrow). There does not appear to be any other masses on these images. The other solid organs seen on the scan also appear to be normal.

At its greatest diameter, the mass measures 10 cm...pretty big!

Renal masses in children have a fairly limited differential. The kidneys do not appear to have hydronephrosis or cysts. The mass is large enough to displace the right kidney. The concern here is if the mass is malignant.

Top malignant renal masses in children:
- Wilms tumor (6%-7% of all childhood renal cancers).
- Clear cell carcinoma of the kidney (the second most common cause, accounting for approximately 3%-5% of all childhood renal cancers).
- Renal cell carcinoma is also a possibility, but this is typically found in adolescent patients. Additionally, this type of cancer is usually not asymptomatic, as hematuria, flank pain, and a palpable mass are the usual symptoms.
- Rhabdoid tumor; this is a very aggressive neoplasm and rare, representing less than 2% of renal malignancies in children.
- Ewing Sarcoma (remember last month’s issue?). These tumors are very aggressive and invasive. They typically have areas of hemorrhage and necrosis and the majority of them have translocations involving the ESWR1 gene on chromosome 22.

Memorable April Fool’s Day Pranks
1996 - Taco Bell Corp runs a full-page ad in several major newspapers claiming it has purchased the Liberty Bell and is renaming it the “Taco Liberty Bell.”
Pediatric Surgery was consulted regarding the mass. Given the location and age of the child, the most likely pre-op diagnosis was Wilms tumor. Additionally, Pediatric Hematology-Oncology was consulted. Further imaging of the chest was obtained and there were no signs of other masses. After discussion with the Tumor Board (a multi-disciplinary team at DCMC), this child went to the operating room for resection of the mass. Removal was successful, and the mass was sent to pathology, which, some days later, confirmed that the diagnosis was indeed Wilms Tumor, Stage II. Chromosomal studies were also sent, looking for loss of heterozygosity (LOH) at 1p36 and 16q22 locations (see below). Mutations of the WT1 gene on chromosome 11p13 are observed in approximately 20% of Wilms' tumors.

**Wilms Tumor**
Also known as nephroblastoma, this is the most common pediatric renal malignancy, occurring primarily in the 2-5 year old age range. It can occur bilaterally and may be associated with congenital anomalies, such as WAGR and Beckwith-Wiedemann syndromes. Radiographically, Wilms Tumor looks like a large smooth mass that's intrarenal and well-defined. Neighboring structures are often displaced due to the tumor's size. Pre-operative imaging tests of Wilms Tumor include CT or MRI scans of the chest, abdomen, and pelvis. The actual stage of the mass is determined by not only these studies, but the surgical appearance of the mass and the results of pathology studies.

**Wilms Tumor histology**

**Favorable Histology:** exactly that; no evidence of anaplastic process or nephrogenic rests.

**Anaplastic Histology:** marked nuclear enlargement and hyperchromasia; single most important predictor of response to treatment and survival. Widely seen anaplasia correlates with poor prognosis.

**Nephrogenic Rests:** kidney precursor cells that are abnormally retained and increase the risk for tumor formation in any remaining kidney.

**Wilms Tumor Staging**

I - Limited to kidney, no metastasis.
II - Extends beyond kidney but completely excised; tumor may involve blood vessels or spillage is present.
III - Unresectable, lymph node metastasis.
IV - Metastasis.
V - Bilateral involvement.

There is substageing based on histology as well.

The Genetics of Wilms Tumor: About 20% of patients will have mutations on the WT1 gene on chromosome 11p13. Half of mutations in WT1 will also have mutations in the CTNNB1 gene (regulates cell-cell adhesion and transcription). Tumor-specific loss of heterozygosity (LOH), if present, increases the risk of relapse as well as a poor prognosis. This can be used to plan and target therapy.

**Other conditions associated with Wilms Tumor:**

1. **Beckwith-Wiedemann Syndrome:** Defect (5 different ones) on 11p15 gene leading to overproduction of IGF-2 and neonatal hypoglycemia, macroglossia, macrosomia, and abdominal wall defects (omphalocoele most common). Around 10% of children will have associated Wilms Tumor. Screening with US is recommended up to age 8.

2. **WAGR:** Wilms tumor, Aniridia, Genitourinary anomalies, Retardation: results from a deletion on chromosome 11 (11p13), involving the WT gene and PAX6 gene (ocular development). Screening for Wilms Tumor is recommended up until age 8. Female patients can have streak ovaries, which places them at risk for gonadalblastomas.

3. **Denys-Drash Syndrome:** triad of Wilms Tumor, Nephropathy, and gonadal dysgenesis. Very rare!

**Treatment for Wilms Tumor**
Nephrectomy followed by chemotherapy, using Vincristine, Dactinomycin, Doxorubicin, or other agents, depending on which treatment protocol is used. Radiation therapy is used in advanced stage disease in some cases. In cases of bilateral tumors, pre-operative chemotherapy is often used and preservation of renal parenchyma is the goal.

Memorable April Fool's Day Pranks That Weren't Pranks!

2006 - Holiday-makers flying back to Britain from Tenerife were told by their pilot that they could not land because an air traffic controller was on a tea break. The passengers initially thought his announcement was an April Fool's Day joke, but it wasn't. The plane had to circle for almost half an hour before the air traffic came back to work.
Case 2: Well, the same imaging considerations that you applied to the child from Case 1 apply to this patient as well. The one important consideration, though, is the age of this patient. 4 months of age is pretty young, and the long term implications of ionizing radiation (CT, for example), are well-known and well-documented.

The physician treating this child elected to obtain plain radiographs first. Here they are:

This radiograph does indeed have some concerning findings. Notice that there appears to be a paucity of air on the entire right side of the film. More concerning, though, is displacement of bowel to the left (yellow arrow), presumably from something on the right (green arrow). This film is concerning primarily for a mass lesion.

Further imaging is needed. Again, CT is an option, as is MRI and US. In this patient, after discussion with the pediatric radiologist, an ultrasound was obtained. Here is the result:

This ultrasound shows a large septated (purple arrow) cystic structure arising from the pelvis and extends towards the liver. There is also bladder compression, causing bilateral hydronephrosis (red arrow).

Well, there is obviously a lesion causing mass effect on the bladder, leading to bilateral hydronephrosis. The lesion is cystic, rather than completely fluid filled or completely solid.

Bloodwork was obtained and is normal; a urinalysis was also obtained and looks normal as well.

Given the age of the child and the extent of the mass, a pediatric surgical consult is obtained. The surgical team evaluates the patient and decides an exploratory laparotomy is the best option for this patient.
Case Resolutions:

Case 1: This child had a rocky post-operative course; a few days after the initial resection, the child developed fever and abdominal pain. A microperforation of the bowel was discovered and a return to the OR was needed for repair. This went smoothly, and a course of antibiotics was completed. As of this writing, chemotherapy was begun with vincristine and actinomycin, and the child is currently cared for by the pediatric oncology department. So far, so good. Further imaging (chest, brain, etc) was negative. As a plus, the pathology was reported as diffuse anaplasia) to 98% (lower stage, favorable histology). Most children do well.

Case 2: The teratoma was removed uneventfully. A followup coccygectomy was performed to remove the residual teratoma as well as any residual cysts that were present. The child was also followed by pediatric urology and nephrology during her hospitalization. Of note, the alpha-fetoprotein levels were initially elevated and have since returned to normal; pediatric oncology is also following the child.

Currently, she is doing well.

Teaching Points:

1. Abdominal masses in children require immediate attention!
2. Most masses are of renal origin in neonates, and include typically include multicystic kidney disease or congenital conditions causing hydronephrosis. Abdominal masses in infants and children are often retroperitoneal in origin (Wilms or neuroblastoma) or enlargement of the liver or spleen (as can be seen in leukemia or lymphoma).
3. Physical exam hints: hypertension (kidney mass, Wilms tumor, neuroblastoma), proptosis/ecchymosis (neuroblastoma), sporic aniridia or hemihypertrophy (Wilms tumor), imperfurate hymen (hydrocolpos, hematocolpos).
4. Useful laboratory studies: CBC (anemia, leukocytosis, pancytopenia), Uric Acid (can be elevated in tumor lysis syndrome or elevated in non-hodgkin’s lymphoma), electrolytes (hypercalcemia in kidney neoplasms), LDH (elevated in lymphoma, neuroblastoma, and germ cell tumors), urinalysis (hematuria in Wilms tumor or renal cell carcinomas). Don’t forget pregnancy in adolescent patients!
5. Useful imaging studies include plain x-rays, ultrasound, CT and MRI. Consider patient age, prior exposure to ionizing radiation, and suspected diagnosis. Consultation with a Pediatric Radiologist can be useful to determine which test to use.
6. Treating abdominal masses requires a multi-team approach, including pediatric surgery, pediatric hematology-oncology, and pediatric intensivists. Referral to the Emergency Department is always appropriate, especially in ill children.

References: