DCMC Emergency Department Radiology Case of the Month

These cases have been removed of identifying information. These cases are intended for peer review and educational purposes only.

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@ascension.org.

This Month: Blue children?! It happens! This month, we'll look at a case of a child who presents with cyanosis but otherwise seems completely fine. That's strange, isn't it? The differential diagnosis is broad with the cyanotic child. Is there a role for imaging?

PEM Fellowship Conference Schedule: September 2018

5th - 9:00 The IRB…………………………………………Dr Allen
10:00 Rheum Emergencies……Drs Rencher & Sivisankar
11:00 Finding a Job/Contracts……..Drs Allen & Fusco
12:00 Research Update

12th - 9:00 Dermatology in the ED……………………….Dr Diaz
10:00 GU Trauma…………………Drs Friesen & Schwarz
11:00 Grand Rounds……………………………..Dr Cruz
12:00 EKG Series…Dr Yee/DCMC Electrophysiologists

19th - 9:00 Tox: Heavy Metals………………Drs Earp & Gillon
10:00 Coagulopathies…………Drs Friesen & Sivisankar
11:00 Hemoglobinopathies…………………Dr Rencher
12:00 ED Department Meeting
7:00 PM Combined PEM/ED Journal Club

26th - 9:00 M&M………………………………..Drs Ryan & Schunk
10:00 Board Review: Misc………………………..Dr Iyer

Grand Rounds Guest Speaker
Dr Andrea Cruz, Texas Children’s Hospital PEM

Simulations are held at the Seton CEC. Lectures are held at DCMC Command Rooms 3&4. Locations subject to change. All are welcome!
Case History

Kind of a quiet day in the Pediatric Emergency Department. It's Summer, after all, and the volume does tend to slow down. You enjoy it while you can, because you know that soon, school will start, kids start getting coughs and colds, and before you know it you are overrun with URI's, bronchiolitis, and patients needing high flow oxygen.

You are asked to take a call from the transfer line. It is a community pediatrician who tell you that she is sending in a 4 year old child with an unusual clinical presentation. She is cyanotic and has an oxygen saturation of 75% on room air, yet, she is well-appearing. What? You readily agree to see the child when she arrives in the Pediatric Emergency Department. In the meantime, in addition to seeing other patients and a trauma or two, you contemplate what might this child need when she arrives.

And arrive she does. In fact, the patient almost skips down the hallway. She is clearly not toxic and is in no distress, but when you enter the room, you note that the patient, despite her overall well appearance, is visibly cyanotic. Her vitals are more interesting: Afebrile. HR - 113  RR - 30  BP - 106/67  O2 Sat - 73% (RA). She is interactive and even smiling. You quickly examine the child. Her exam is significant for some clinical findings. You note that, aside from the obvious cyanosis (primarily perioral and labial), she has mild tachycardia, an apparently normal S1/S2, non-displaced cardiac PMI, no clicks, no rubs, and no obvious murmur (she is, though difficult to examine because she is constantly moving and speaking loudly to her younger sibling, who has one of her toys). Her pulses are equal throughout. She has brisk cap refill. There is no detectable bruit. There is clubbing to her extremities bilaterally. Her lungs are clear, and while she is mildly tachypneic, there are no retractions. There is no hepatosplenomegaly, but there is something strange about her liver and spleen exam. The liver edge is not detectable and not enlarged, but the spleen edge seems detectable on the left. She is nontender. She has no rash and does not look pale.

What is going on her? You are suspecting a heart issue, but, of course, there are other things to consider, such as pulmonary disease or even a metabolic process. You get a history from the parents. Apparently they are new to the United States. The child was born in Honduras and, the mother states, was diagnose with a “heart problem that might get better” but, to the mother’s knowledge, an exact diagnosis was not given. The child has, despite this, done well, and in fact has been relatively thriving. They arrived in the US 2 months ago. In that time, they have not seen a health care provider, but about a week ago they noted that the child seemed to be more fatigued and “looked more blue than usual”. This prompted a visit to the clinic.

You consider imaging options. Clearly, an echocardiogram is in order but will plain imaging be useful for this child? If so, what findings on imaging can lead you to suspect congenital heart disease in this child? Will a plain film (ie chest xray) suffice, or do you need a chest CT or chest angiogram? What about a cardiac MRI? And why did the child look more cyanotic and her oxygen saturations drop to the 50’s when an IV was placed to obtain the bloodwork that you decided to order (CBC, CMP, BNP, Type and Screen). She also did not like the ECG that you asked for. Why does this always happen on a Friday afternoon?

A Word on Pulse Oximetry

In 1935 Karl Matthes (1905-1962) developed the first 2 wavelength ear O2 saturation meter, using red and green filters. In 1940, the first oximeter was developed by Glen Allan Millikan. In 1972, Takuo Ayoagi and Nihon kohden developed what we now know as the contemporary pulse oximetry device. By the 1987, pulse oximeter use can become the standard of care when administering anesthesia. By 1995, the biotech company Masimo introduced Signal Extraction Technology that could more accurately provide measurements during patient motion and low perfusion states. Interestingly, no manufacturers of pulse oximetry devices guarantee readings during such states.

We use this clinical tool on practically every patient we see but what exactly is a pulse oximetry reading telling us? Well, the oximetry passes two wavelengths of light through the body (usually a finger or toe) to a photodetector. This allows the oximeter to measure what percentage of hemoglobin is loaded with oxygen. Remember that this is a measurement of oxygenation not ventilation. This does not provide a complete measure of respiratory sufficiency. Falsely low reading can be caused by hypo perfusion or vasoconstriction, movement, or certain disease states, such as carbon monoxide poisoning, cyanide poisoning, or methemoglobinemia.

"So, does anyone wanna play Parcheesi?"
-Rick Moranis, Ghostbusters.
Rick improvised this, and all the lines, during this scene.

"Lozemgael!"
-Mel Brooks, Blazing Saddles. This entire scene was improvised, in Yiddish, by Mel Brooks, who plays an Indian Chief.
“It’s a Cinderella story”.
Bill Murray, Caddyshack. Director Harold Ramis let Murray improvise this entire scene.

I’m in a glass case of emotion”.
-Wil Ferrell, The Anchorman. This movie was famous for its cast of improvisers.

“We’re gonna need a bigger boat”.
-Roy Scheider, Jaws. This was actually an inside joke because the actors felt that the production crew was using a too small boat to film the scene. Roy tried it in different scenes.

Here are a few images of our patient. Note that there is clubbing of the fingers and the toes. Additionally there is cyanosis of the lips and perioral area. Her ECG is also noted. Perhaps RVH is present, but otherwise it looks normal (sinus rate of 110, normal QTc, normal PR). You decide to obtain a plain 2 view chest X-ray to see if you can get a hint as to what is going on with this patient. Now that she is calm, her oxygen saturations are back to the low 70’s.

Here are the chest X-ray images that were obtained on this child. There are a few striking findings. The lungs look. But, note that the gastric air bubble is on the right (yellow arrow), the liver is on the left (silver arrow), the left lung has an appearance that suggests it is tri-lobed (orange arrow), the right lung is bi-lobed (purple arrow), and there appears to be a right sided aortic arch (red arrow). There is levocardia. The cardiac size is normal.

Well, these are interesting findings! The child has levocardia with situs inversus.

Situs Inversus: This is a shortening of the phrase “situs inversus viscerum”, meaning an inverted position of the chest and abdominal organs. When there is total transposition of the chest and abdominal organs, this is called situs inversus totalis. Situs inverses is usually associated with dextrocardia (although not in our patient) and an incidence of congenital heart disease of up to 5% (transposition of the great vessels is the most common heart defect). In patients with situs inverses with levocardia (much more rare, less than 1%) , congenital heart disease is very common, up to 95%.

Situs Classification: this best basically a spectrum but there are three primary categories:
1. Situs Solitus: normal configuration of organs.
2. Situs Inversus: mirror image of normal.
3. Situs ambiguous: intermediate configuration with duplication. Also known as heterotaxy.
“Sir what? Where you about to call me an a@#$**e??!”
- R Lee Ermey, Full Metal Jacket.
Initially hired as a consultant, the retired US Marine sergeant did so well he was hired to play Sgt Hartman. He improvised his entire dialog in the film.

“You can’t handle the truth”.
-Jack Nicholson, A Few Good Men. The original line was “You already have the truth” but he thought this would work better instead.

“Leave the gun. Take the cannoli”.
-Richard Castellano, The Godfather. I guess that’s one way to lighten up a scene.

Imaging the Pediatric Patient With Suspected Congenital Heart Disease

Chest Xray - Always a good option! As has been indicated in the past, this modality is inexpensive, available, has a low exposure to ionizing radiation, and requires no sedation. Plain chest radiography can be sensitive for the detection of a cardiac anomaly. Note though, that a normal radiograph does not eliminate the possibility of congenital heart disease (depending on the age of the patient and the heart lesion) but it is a great first step!

Echocardiogram - The first step gold standard in congenital heart lesion imaging. Depending on the institution this may be readily available (at DCMC it is) and requires no ionizing radiation. It is useful for looking at bloodflow, cardiac anatomy, and the presence of any effusions.

CT - Computed tomography with angiography is helpful to delineate cardiac anatomy and is rapid (sedation is not needed). But this requires ionizing radiation.

MRI - Great for examining cardiac anatomy but requires sedation and is not rapid. May also not be readily available (at DCMC, luckily, it is).

Tetralogy of Fallot

This is a classic plain chest radiograph of an infant with Tetralogy of Fallot (VSD, Overriding aorta, RVH, Pulmonary stenosis). In this child, the cardiac apex is elevated, suggesting RVH (red arrow); the patient also has a right sided aorta (green arrow). The heart also has a “boot” shape to it. There may be normal to diminished pulmonary blood flow which may be seen as decreased pulmonary vascular markings on a plain film. This condition is the most common cyanotic congenital heart illness. It was first described by French physician Etienne-Louis Fallot in 1888.

Transposition of the Great Arteries

Here, the aorta rises from the right ventricle and the pulmonary artery from the left ventricle (there are variants). In order to survive this cardiac lesion, the child must have an ASD, VSD, or PFO (sometimes they have all of them). Initially, neonates with this lesion can seem normal but as pulmonary pressures decrease after birth, cardiac enlargement occurs and pulmonary over circulation develops. On plain chest radiographs, this can be seen by the “egg on a string” appearance of the heart (red arrow). This is due to thymic atrophy related to the stress of cyanosis, the great arteries causing a narrow vascular pedicle (green arrow), and cardiomegaly.

Cardiomegaly

Chest radiography can be helpful in detecting cardiomegaly. Using the cardiothoracic ratio helps to determine if cardiomegaly is present. This is the ratio of maximal horizontal cardiac diameter to maximal horizontal thoracic diameter (inner edge of ribs/edge of pleura). A normal measurement should be less than 0.5.

Total Anomalous Pulmonary Venous Return

There are 4 types of this congenital anomaly, with Type I being the most frequent (this is the supra cardiac type). Patients with severe obstruction usually present in the neonatal period but those with less severe obstruction can present later on in life. The underlying pathology is the pulmonary veins do not drain into the left atrium, instead they drain into the right atrium either directly or by a systemic venous connection. Radiographically, one can see the “Snowman Sign”. The body of the snowman (red arrow) is the enlarged right heart (atrium) and the head (green arrow) is formed by the widening of vertical vein on the left, the brachiocephalic vein on top, and the SVC on the right.

Cardiomegaly, Pneumonia, or a Mass?

Nope, none of the above. This is a thymus demonstrating a thymic shadow and it is often mistaken for a number of different pathologies. Remember that the thymus is present at birth, undergoes considerable growth (reaching its maximum weight during adolescence) and then involutes after that. This is the Sail Sign. Note that there is no mass effect on other structures. It is often on the right but can be bilateral and is seen in up to 15% of cases.

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“I’m funny how? I mean, funny like I’m a clown?”
-Joe Pesci, Goodfellas. This line was actually based on a real-life story and the scene was almost entirely improvised with Ray Liotta.
Remember our patient? With chest radiograph findings, Pediatric Cardiology was consulted. An echocardiogram was performed, which revealed very complex cardiac anatomy:

1. An unbalanced AV canal with single ventricle (the aorta arose from this ventricle and there is no obstruction).
2. No identifiable pulmonary arteries (consistent with pulmonary atresia with MAPCAs (major aortopulmonary collaterals)).
3. Large primum ASD
5. Normal ventricular function.
6. Single coronary artery raising from the left cut and giving rise to the LAD/Circumflex and RCA.

In order to further delineate the child's anatomy, a CT angiogram of the chest was performed. Images are noted to the left. A CT was utilized because of the rapidity of the study and this modality obviates the need for sedation, as opposed to cardiac MRI. Note the large single ventricle (red arrow) and the unobstructed aorta (yellow arrow).

Case Resolution:
This child was admitted to the Pediatric Intensive Care Unit for further workup/imaging. The child, as we've seen, underwent echocardiography and CT angiogram imaging of the chest. While she remained stable in the PICU, Pediatric Cardiology and Pediatric Cardiothoracic Surgery were consulted. She was able to be discharged home with followup cardiac catheterization. This was done several days later and confirmed the child's very complex cardiac anatomy (including the absence of central pulmonary arteries and pulmonary atresia but with very small collateral blood supply to the pulmonary vasculature, and single ventricle physiology). Her bloodwork (remember that) revealed polycythemia (hemoglobin was 23 and hematocrit was 69) most likely secondary to chronic hypoxia. This child with heterodoxy syndrome has extremely complex anatomy and surgical repair, at this time, does not seem possible. Pediatric Palliative Care was engaged while the case was continued to be reviewed by Pediatric Cardiothoracic Surgery. As of this writing, though, surgical intervention as a treatment option does not seem possible.

Teaching Points:
1. Cyanosis in children has a very broad differential but can be categorized into central or peripheral etiologies. The basic mechanism is either desaturation of systemic arterial oxygen or increased oxygen extraction by the tissues. Don't forget that cyanotic mimics exist, including dyes on the skin, pigmented lesions (Mongolian spots), drugs (silver application).
2. Cyanosis can be due to pulmonary causes (including airway obstruction), cardiac causes (specifically congenital heart disease), acute chest syndrome in sickle cell patients, environmental exposures (methemoglobinemia), and hypoperfusion states (shock). A good history and physical exam can help sort out the most likely cause.
3. If suspecting congenital heart disease, consider the age of the patient, the history/presentation, and the physical examination. For example, neonatal ductal dependent lesions often present around 2-3 weeks of age (when the ductus arteriosus completed closes) with signs of cyanosis and shock.
4. Plain chest radiography is an excellent first line imaging to modality when a child has cyanosis, especially if a congenital heart lesion is in your differential.
5. Echocardiography is indicated if the diagnosis of congenital heart disease is being considered and if myocarditis or a pericardial effusion is in your differential. This may not be readily available, so know the resources at your institution.
6. Chest angiography or cardiac MRI imaging can help further define suspected complex cardiac anatomy. These modalities may not always be indicated and are situation dependent.
7. Early consultation with Pediatric Cardiology should be done on children with suspected congenital heart disease. Be sure to obtain and ECG as well!

References: