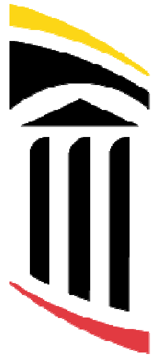

Abnormal Cases of Cardiac Disease Presenting in the Primary Care Office and Emergency Room

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Pediatric Cardiothoracic Surgery

Objectives

- Understand presentations of cardiac disease outside of the neonatal setting
- Understand the initial management, triage, & stabilization of cardiac disease
- Understand important history taking and red flags for possible cardiac disease
- Understand appropriate referrals to cardiology for common cardiac complaints
- Understand the long term issues, medications, and necessity of cardiology follow up
- Understand appropriate imaging studies outside of echocardiography for specific cardiac diseases
- Understand the surgical repair for several specific cardiac malformations
- Understand the common post-operative course for "elective" surgical cases and possible complications

Case #1 – G.R.

- G.R. is a 2 year old who presents to cardiology clinic referred by her endocrinologist for a murmur
- PMH – excessive breast tissue which seen was seen by endocrinology, otherwise healthy
- Birth History – SVD, uncomplicated, born at 39 weeks GA
- Previous Hospitalizations – None
- Diet – Regular
- Past Surgical History – None
- Family history – Maternal Grandmother with hypothyrodism, no family history of cardiac disease or early sudden death
- Medications – Multivitamin and Fluoride daily
- Social History – Lives at home with both parents, a 4 year old brother and 4 cats, not in daycare
- Immunizations – UTD, received flu for 2013
- Allergies – NKA
- Developmental – Appropriate 2 year old

Case #1 – G.R.

- Review of Systems:
  - *Denies fever, decreased activity level*
  - *No recent rhinorrhea or congestion*
  - *No diaphoresis, palpitations, or chest pain*
  - *No shortness of breath*
  - *Denies N/V/D*
  - *No rashes*
  - *No syncope*

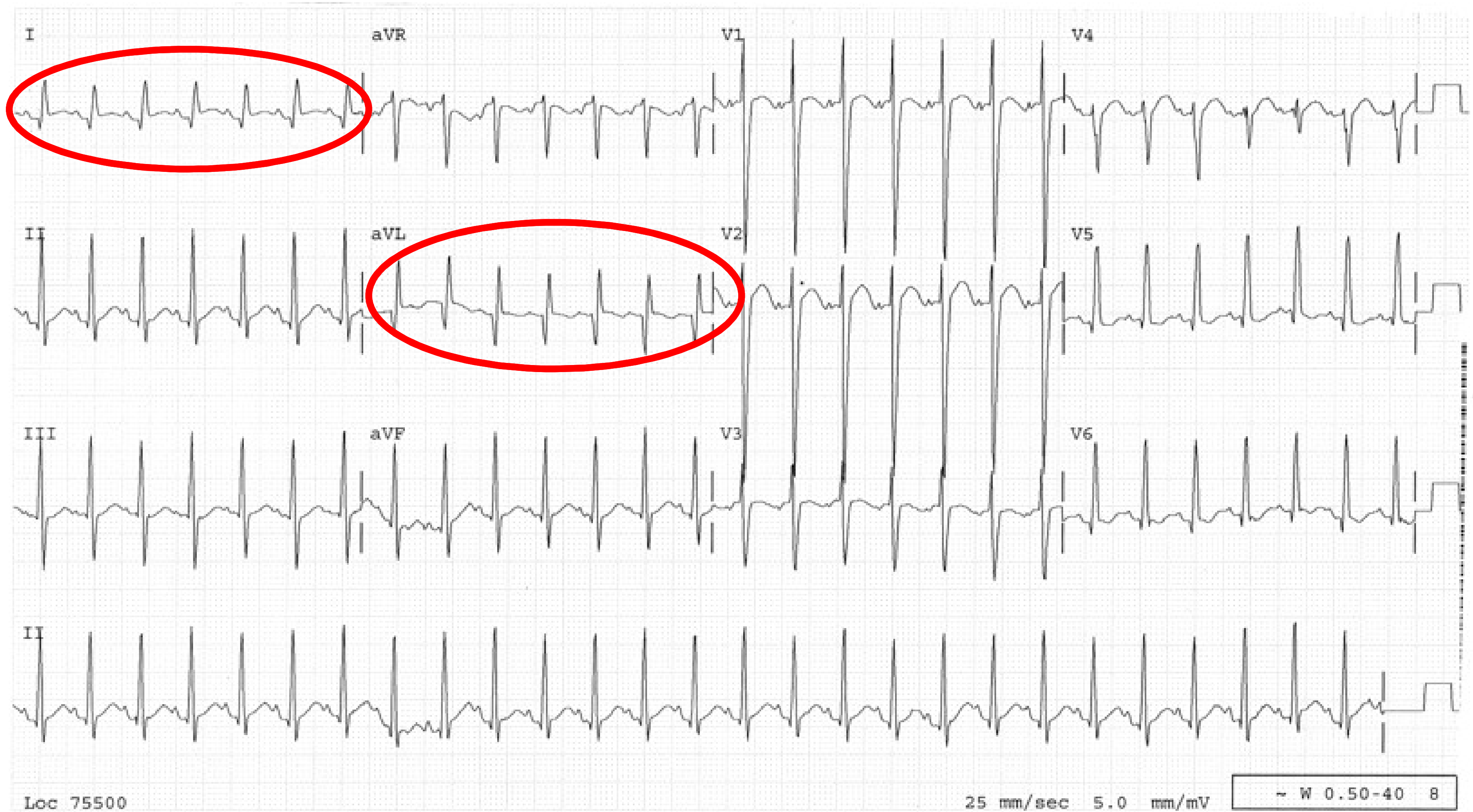
Case #1 – G.R.

- Physical Exam
  - *GEN – alert, nontoxic toddler in NAD, awake, alert & interactive*
  - *HEENT – NCAT, normal nares, hearing grossly intact, no rhinorrhea, MMM, OP patent, no erythema or exudates, pupils nonicteric, noninjected, PEERL, EOMI*
  - *RESP – CTA bilaterally with good aeration, no accessory muscle use, no retractions, no nasal flaring*
  - *CV – Grade 2/6 SEM at RSB, pulses 2+ throughout, no edema GI – abdomen soft, NT/ND, BS active, no HSM, no masses*
  - *LYMPH – no LAD*
  - *MS – moves all extremities, no gross deformities of joints or long bones, no clubbing or cyanosis, strength 5/5*
  - *NEURO – Alert, normal bulk/tone*
  - *DERM – intact, no rashes*

Case #1 – G.R.

- Per Grace's parents, an chest x-ray and EKG were obtained by her pediatrician and were "abnormal"
- Another EKG was obtained in the cardiology clinic and demonstrated:
  - *Normal Sinus Rhythm with ST and T wave abnormalities and Q waves present in leads 1 and AVL…*

Case #1 – G.R.



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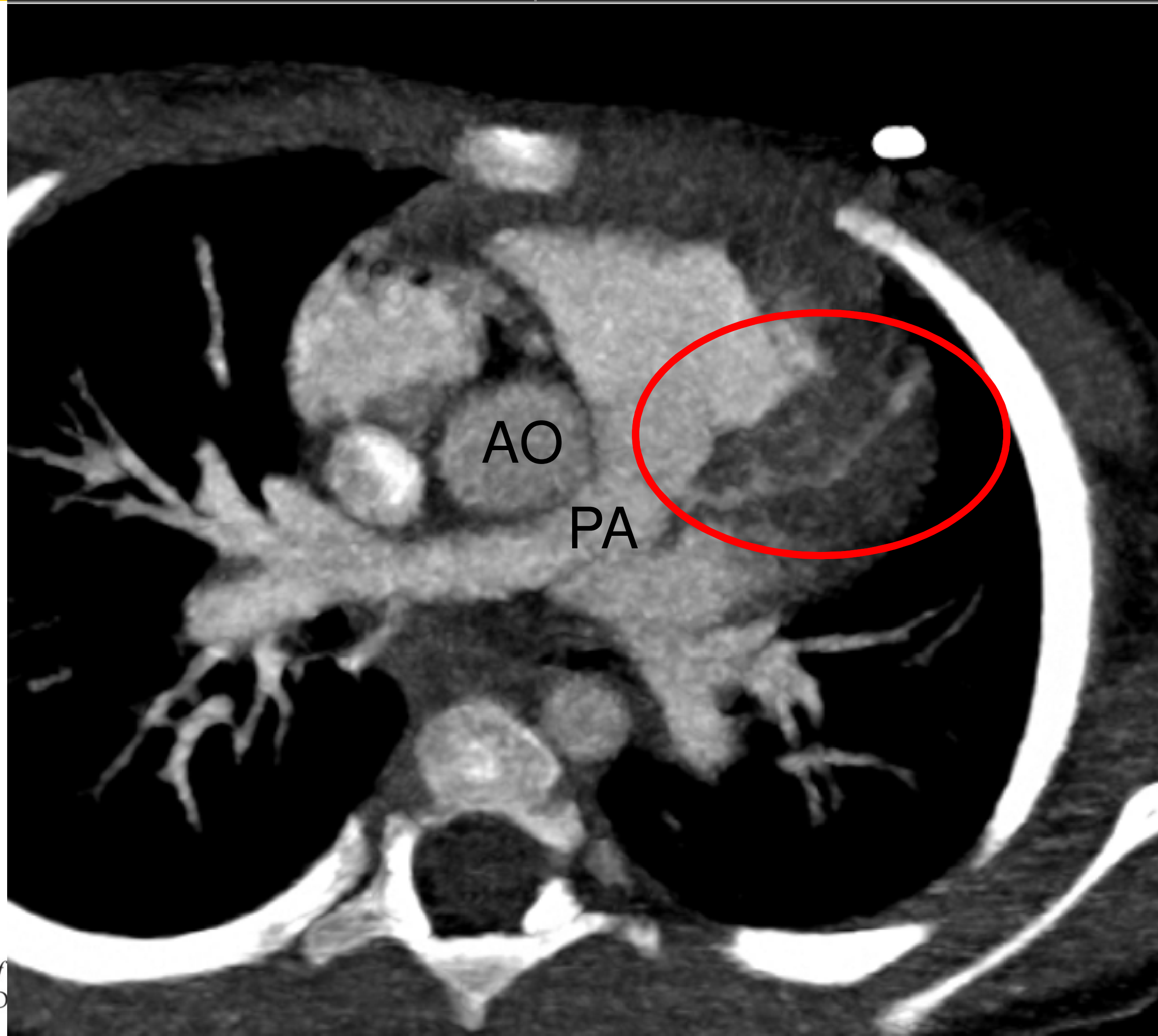
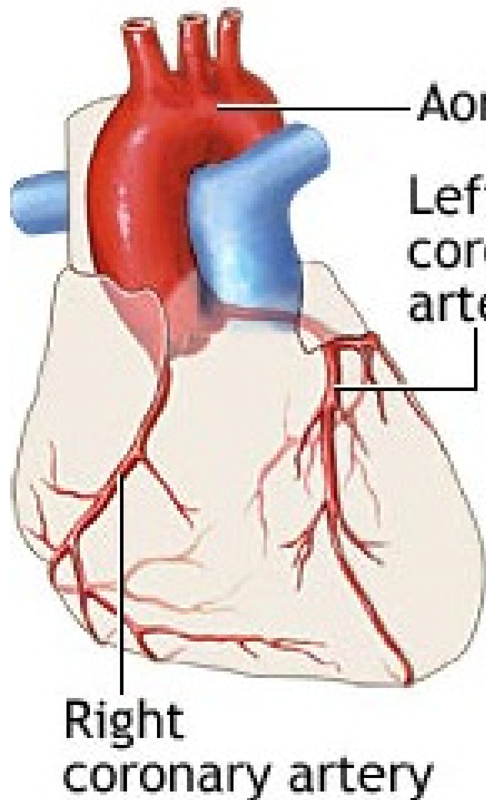
Case #1 – G.R.

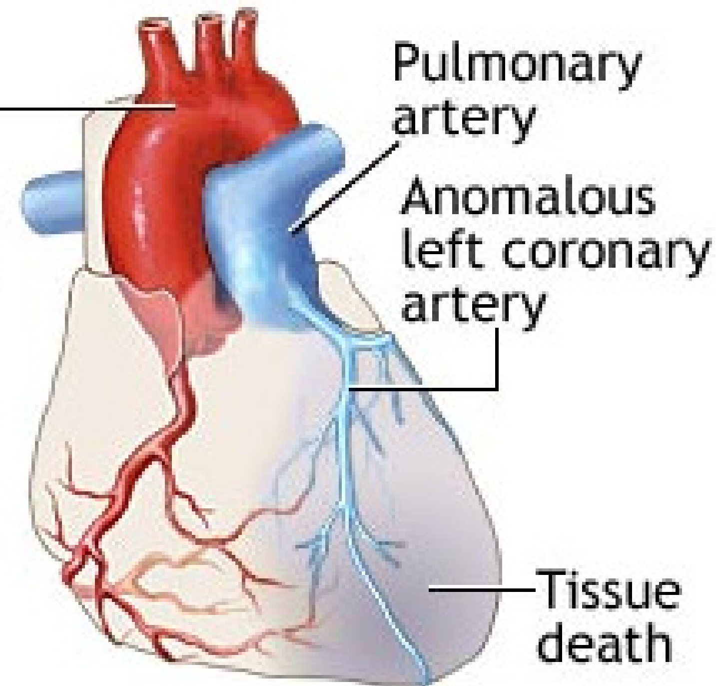
- Due to the Q wave finding, and echocardiogram was obtained which confirmed a diagnosis of ALCAPA – anomalous left coronary off the of the pulmonary artery, with moderately depressed left ventricular function (Ejection Fraction 44%, normal >55%) and right coronary dilation
  - *Deep & wide Q waves in leads I & AVL is pathopneumonic for ALCAPA*
- A chest CTA was also obtained for further imaging prior to surgery

Case #1 – G.R.



Case #1 – G.R.

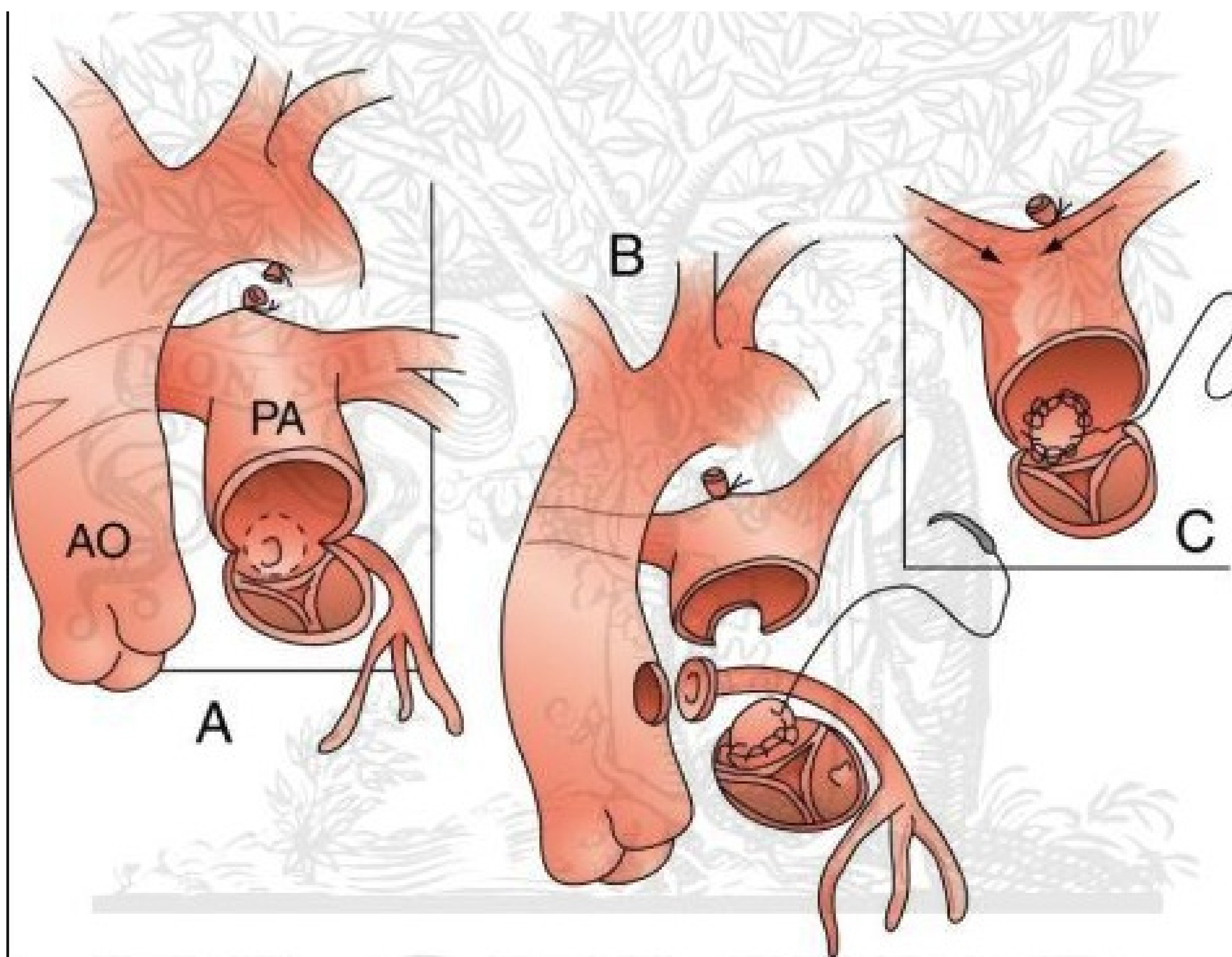




Case #1 – G.R.

- G.R. was therefore taken to the operating room the next day and underwent direct translocation of the left coronary artery button to the aorta
- Her post-operative course was uncomplicated and she was able to be discharged 5 days after admission

Case #1 – G.R.



Case #1 – G.R.

- Long term however, G.R. has continued to have mild to moderately depressed left ventricle function and is on enalapril, aldactone, and aspirin (for coronary patency)
- In a study of 56 patients (31 underwent coronary artery reimplantation), both ventricular function and mitral valve regurgitation normalizes over time; ~95% of patients were alive at 20 year follow-up (Lange et al., 2007)
- However, in a separate study of 11 patients who underwent ALCAPA repair in infancy and were evaluated for exercise performance/capacity (median age of 17 years) and compared to age-matched controls and exercise ability was found to be impaired (Singh et al., 1998)
  - *Potential confounding factors r/t exercise capacity for any child undergoing CT surgery*
    - In a study by Pinto, et al. (2007), >25% of patients with heart disease were obese or overweight with <5% of these patients having single ventricle or transplantation status **meaning 85% of the obese patients had heart disease that should not be limiting their exercise**

Case #1 – G.R.

- *Murmurs – When should you refer??*
- ALL newborn murmurs **MUST** be referred
- *Innocent*
  - Grade I-II/VI
  - Changes with position
  - Can vary in loudness
  - Musical/vibratory
  - Usually systolic except for venous hum
  - Short duration
  - Heard best at LLSB
  - Child looks otherwise healthy
- *Pathologic*
  - >Grade III
  - Heard in pt w/ a syndrome
  - Diastolic
  - Thrill
  - Pansystolic/continuous
  - Clicks or snaps
  - Fixed splitting
  - Loud S2
  - S4 gallop
  - Nonpositional
  - Harsh
- A murmur heard in a school age child with "innocent murmur" qualities can be followed by the primary care provider and does not necessarily need to be referred immediately
  - *If the murmur continues over time, does not go away, or starts to sound different, then referral is necessary*
  - *Up to 80% of all children have a murmur at some point in their life*
  - *3-4 years is most common age*
  - *Innocent murmurs will be louder or present at times of high output states (i.e. fever)*

Case #2 – T. S.

- History of Present Illness:
  - *T. S. is a 9 day old male who was born via SVD to a G2P2 mother at 41 weeks GA. Mother's prenatal history was benign, delivery was complicated by the cord being wrapped around his neck and meconium stained amniotic fluid. Despite this, T. S. did well and was discharge home with his parents at DOL#2. Since then, T. S. has been at home, doing well, breastfeeding with appropriate weight gain until yesterday. Throughout the day, parents felt T. S. was more difficult to arouse and had poor PO intake. Mom states that she was able to breastfeed him ten times throughout the day but he kept falling asleep.*

Case #2 – T.S.

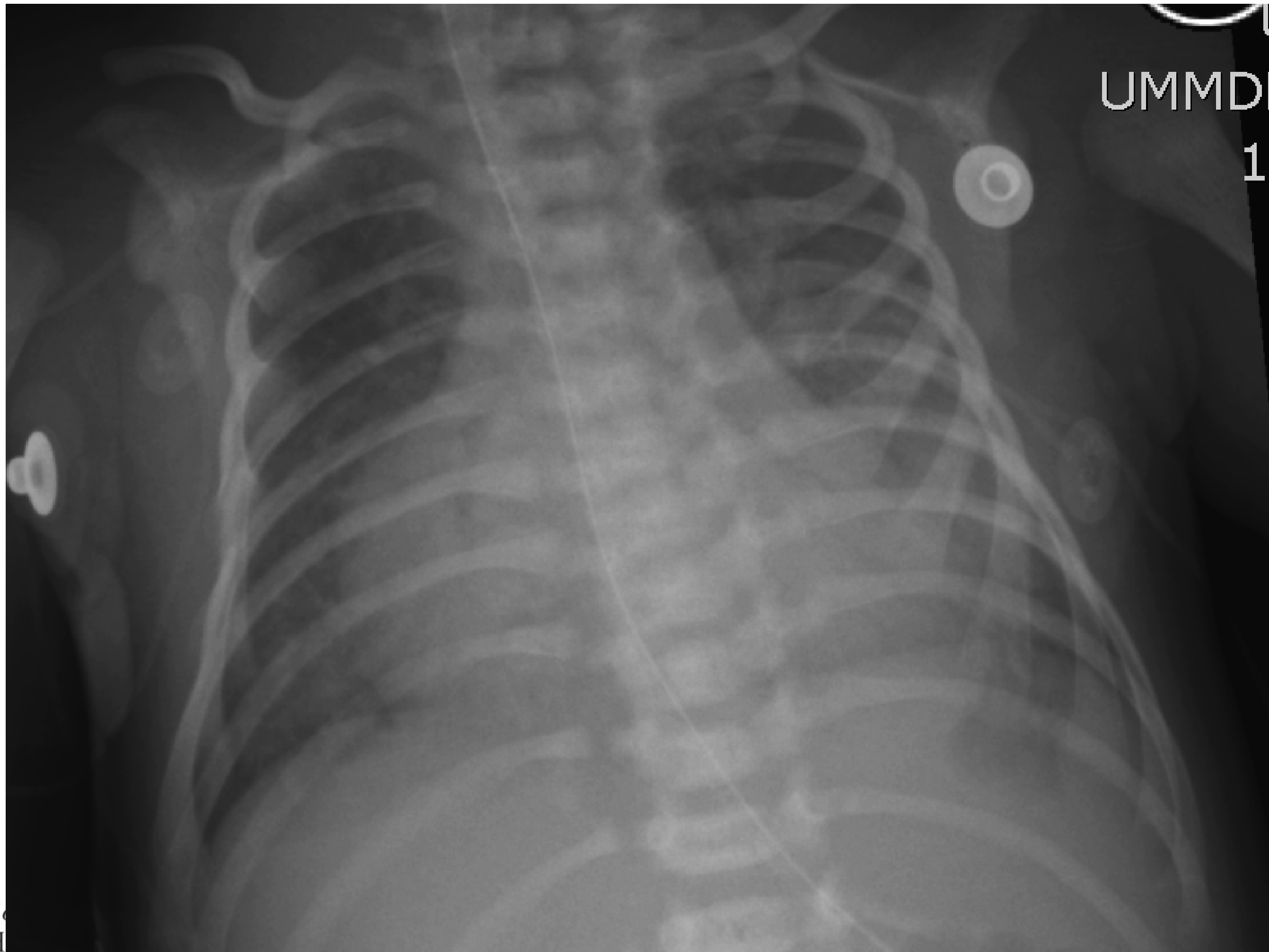
- Past Medical History – full term newborn, no other known medical conditions, prenatal ultrasounds were normal per mother
- Maternal History – born at 41 weeks via SVD to a G2P2 mother who received prenatal care, only medication during pregnancy was prenatal vitamins, +meconium noted in amniotic fluid with no respiratory issues, birth weight 3.97kg
- Past Surgical History – None
- Previous Hospitalizations – None
- Diet – Breastfed ~Q2 hours
- Immunizations – received Hepatitis B at birth
- Social – Lives at home with both parents and 3 year old brother

Case #2 – T.S.

- On limited exam in the PCP office:
- GEN – crying, not easy to calm 9 day old
- HEENT – NCAT, anterior fontanelle O/S/F, PERRL, nonicteric
- RESP – tachypnic to 90s, +retractions and nasal flaring
- CV – S1S2 normal with no murmur noted, cool throughout, pulses barely palpable in lower extremities, cap refill 4-5 seconds in feet
- GI – Abd soft, NT/ND, hypoactive bowel sounds, no HSM
- DERM – no rashes

Case #2 – T. S.

- Chest X-Ray obtained in PCP office

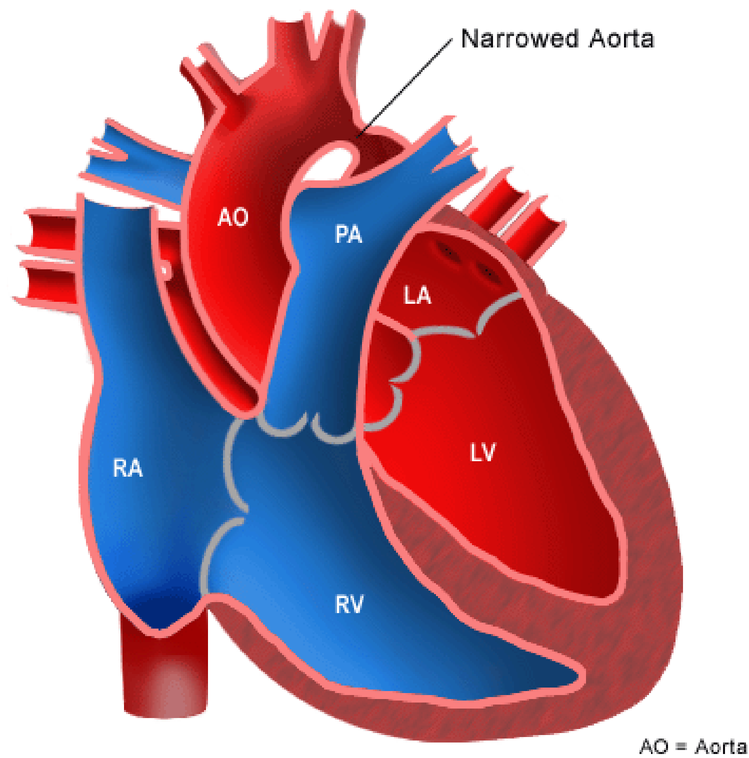


Case #2 – T. S.

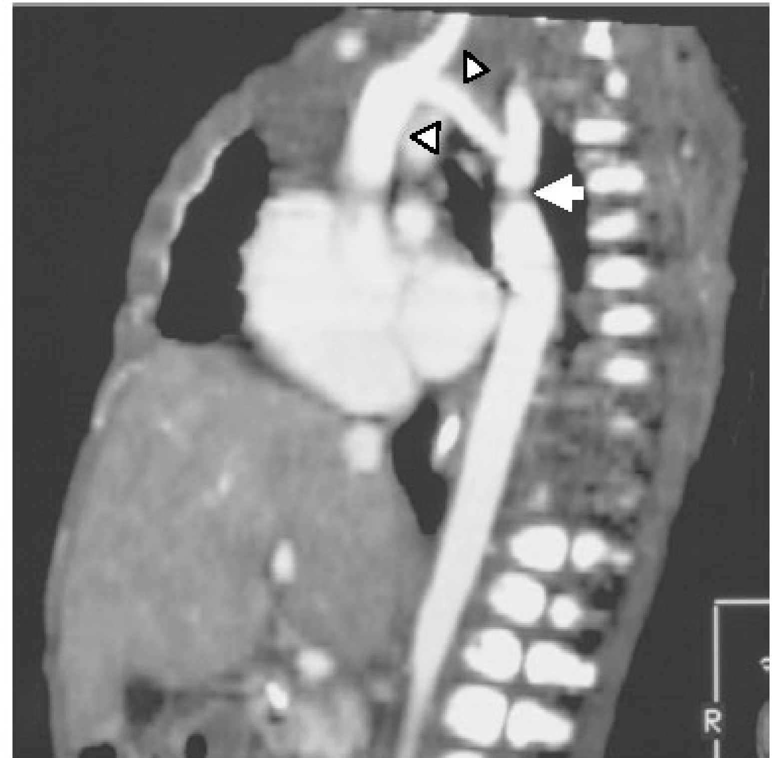
- T. S. was therefore transferred to an outside emergency room where he was intubated for escalating respiratory distress. An echocardiogram was obtained which was concerning for possible coarctation of the aorta. A prostaglandin infusion was initiated and T. S. was transferred via helicopter to the UMMC PICU.
- On arrival to the PICU, T.S. was stabilized and a repeat echocardiogram was obtained which demonstrated hypoplasia of the aortic arch with critical coarctation, multiple muscular ventricular septal defects with low velocity bidirectional shunting, severe right ventricular dilation, right atrial enlargement, right ventricular hypertrophy, and moderately depressed left ventricle and right ventricle function

Case #2 – T. S.

- Critical Coarctation of the Aorta



- Hypoplatic Aortic Arch



Case #2 – T. S.

- Critical coarctation or critical coarctation with aortic arch hypoplasia presenting in cardiogenic shock is defined as "impairment of the left or right ventricular systolic function, respiratory failure requiring tracheal intubation, and metabolic acidosis" (Fesseah et al., 2005).
- Mainstays of management include prompt diagnosis, establishing access and initiating prostaglandins, and early surgical repair
  - *Initiation of PGEs – may be given peripherally, start at 0.1mcg/kg/min, usual ICU dosing once PDA is established in 0.02mcg/kg/min, the higher dose helps to re-open the PDA*
- VBG on admission: 7.38/46/32/27/+1.2; w/ lactate 2.5

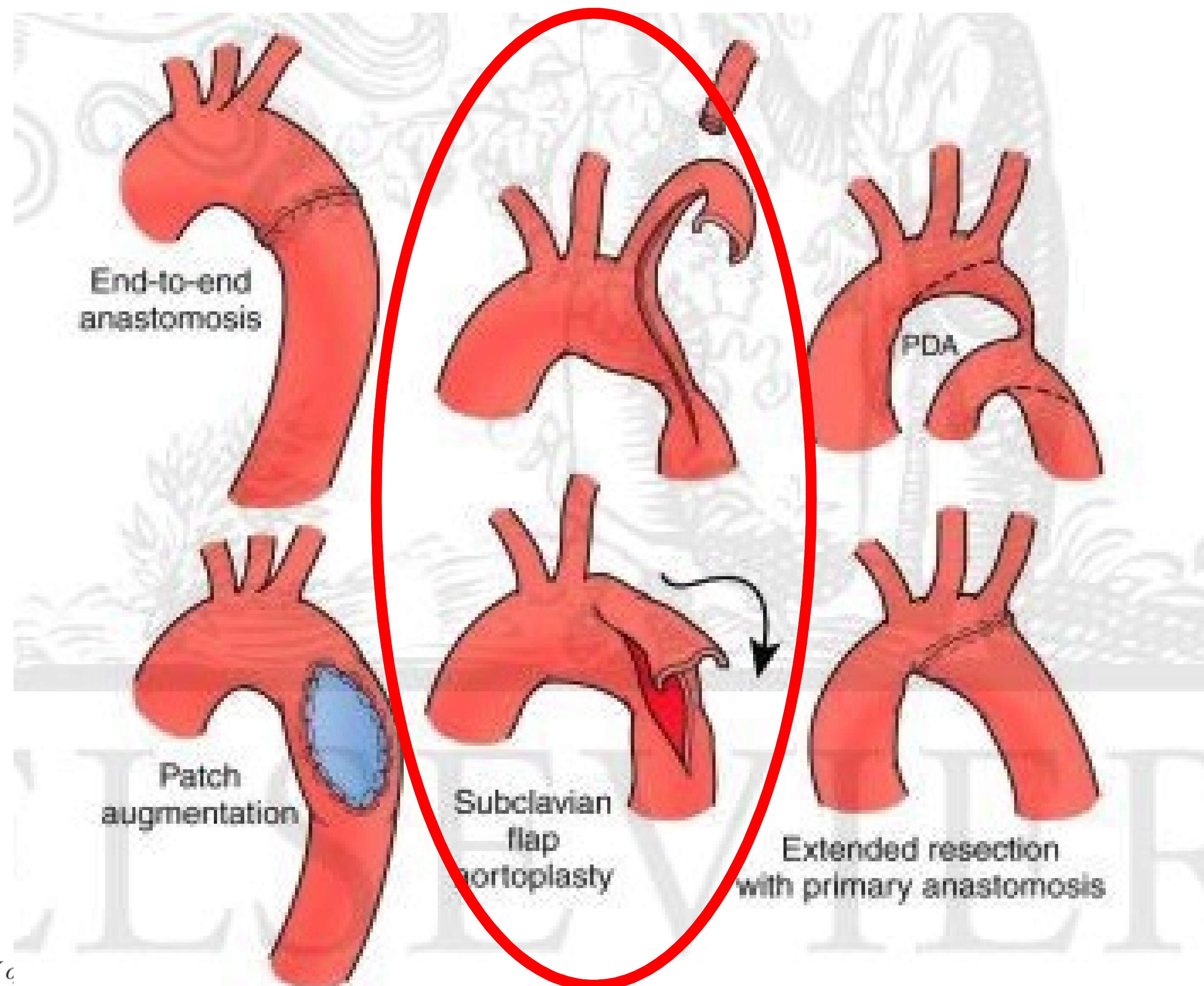
Case #2 – T. S.

- Other important considerations in patients with arch anomalies
- Genetic Syndromes
  - *Turner's Syndrome*
    - 33% of patients have cardiac malformations, 75% of these are coarcation of the aorta or a bicuspid aortic valve
  - *22q11 deletion /DiGeorge Syndrome/Velocardiofacial Syndrome*
    - Seen in 50% of patients with interrupted aortic arch, 34.5% of patients with truncus arteriosus, and 15.9% of pateints with tetralogy of fallot
  - *(Goldmuntz, et al., 1998)*

Case #2 – T. S.

- T. S. was taken to the operating room and underwent hypoplastic aortic arch and critical coarcation repair via subclavian flap
- Postoperative issues included:
- Stridor and increased work of breathing, ENT consult revealed left vocal cord paresis
  - *Due to recurrent laryngeal nerve injury, 35% rate of spontaneous recovery, ~50% chance of aspiration risk, high incidence of need for gastrostomy tube feeding (Truong et al., 2007)*
- Difficulty feeding but eventually resumed full PO breastfeeding
- Superficial wound infection and dehiscense requiring Keflex and collagenase therapy
- Hypertension necessitating discharge home on captopril
- Follow up echo at post-op visit demonstrated normal biventricular systolic function

Case #2 – T.S.



Case #2 – T. S.

- Balloon/catheter based intervention versus surgery for PRIMARY repair
  - *Study by Rao et al. (1996) demonstrated that balloon angioplasty is safe for neonates and infants although a significant incidence of recoarctation was seen which was amendable to repeat balloon angioplasty*
  - *Study by Cowley et al. (2005) demonstrated a higher incidence (50%) of aneurysm formation artery injury compared to primary surgical intervention in infants and neonates*
  - *Meta-analysis by Hu et al. (2014) demonstrated that balloon angioplasty provides comparable immediate results to surgery but does not provide better medium or long term results; increased incidence of aneursym formation*

Case #2 – T. S.

- Coarctation of the aorta in the long term
- Two main issues: reintervention and hypertension
  - *Excellent long term results for patients who undergo subclavian flap repair during infancy compared to other techniques (~6% need for reintervention) (Adams et al., 2013).*
  - *~10% of all patients with isolated coarctation of the aorta require reintervention by 30 years; younger age at primary repair and end-to-end anastamosis technique have a lower rate of reintervention (Brown, et al., 2013).*
  - *~50% of patient with coractation of the aorta have hypertension as assess by 24 hour blood pressure monitoring at~12 years after initial surgical repair (O'Sullivan, et al., 2002).*
    - Despite successful surgical repair, due to abnormalities in the shape of the aortic arch, changes in baroreceptor reflexes, and neurohuromal factors (Hauser, et al., 2000)

Case #2 – T. S.

- Ventricular septal defects and spontaneous closure
  - *Location, location, location!*
    - Small muscular VSDs have the greatest likelihood of spontaneous closure with rates of ~80-90% by 2 years of age
  - *Patient sex*
    - Girls win! Boys have a slightly decreased chance of VSDs closing spontaneously
  - *Patient age*
    - Higher chance of spontaneous closure if seen in patient <2 years of age
  - *That said, do all VSDs that don't close spontaneously need to be surgically closed??*
- (Taylor, 2013)

Case #3 – S.C.

- S. C. presents to the cardiology clinic with a complaint of chest pain
  - *Per his mother, S.C.'s chest pain was first noted approximately 4 weeks ago. He is very physically active, and plays soccer. Approximately 4 weeks ago, he developed an acute onset of chest pain. This was not associated with activity. He described the pain as sharp and like a pressure. He grabbed his chest, but did not appear pale. He did not faint. Also, his mother notes that he has difficulty swallowing bulk foods and routinely takes 30+ minutes to eat his meals due to prolonged chewing.*
  - *He has previously been evaluated by his PCP and in the emergency room with no abnormal diagnostic testing so he was referred to cardiology for further work-up.*

Case #3 – S.C.

- Past Medical History – occasional constipation, no chronic medical conditions, no previous surgeries or hospitalizations
- Medications – Miralax 17gm PRN for constipation
- Allergies – NKA
- Family History – Mother with SVT, maternal grandmother died from MI at age 42, father's family history is unknown
- Social – S.C. lives at home with his mother, only child, in 2nd grade—likes school and is doing well
- ROS – all systems negative except for aforementioned chest pain and difficulty swallowing solid foods

Case #3 – S.C.

- Physical Exam:
- Vitals: BP 109/60 | Pulse 81 | Resp 26 | Ht 4' 4.84" (1.342 m) | Wt 61 lb 9.6 oz (27.942 kg) | BMI 15.52 kg/m2 | SpO2 99%
- General: NAD, no dysmorphic features
- Eyes: Conjunctiva are non-injected, PERRL.
- ENT: The external ears and nose are normal. The mucous membranes are moist
- Respiratory: Normal respiratory effort, clear lung fields bilaterally. No stridor.
- CV: The precordium is normal to palpation. There is a regular rate and rhythm with a normal S1 and S2. There are no murmurs, rubs or gallops.
- Abd: The abdomen is soft and nontender. There is no hepatosplenomegaly.
- Ext: The extremities are warm and well perfused. There are 2+ pulses with no brachiofemoral delay. There is no edema. There is no cyanosis or clubbing.
- Neuro: The patient is alert and oriented with a normal affect.
- Muscl: No focal tenderness to palpation of his chest wall.

Case #3 – S.C.

- Chest Pain
- In a study of family or patient's understanding of chest pain
  - *Cardiac – 52-56%; 44% of children presenting with chest pain thought that they were having a heart attack*
  - *Musculoskeletal – 13%*
  - *Respiratory tract – 10%*
  - *Skin infection – 3%*
  - *Breast – 3%*
  - *Cancer – 0 to 12%*
  - *Unsure – 10 to 19%*
- (Geggel & Endom, 2014)

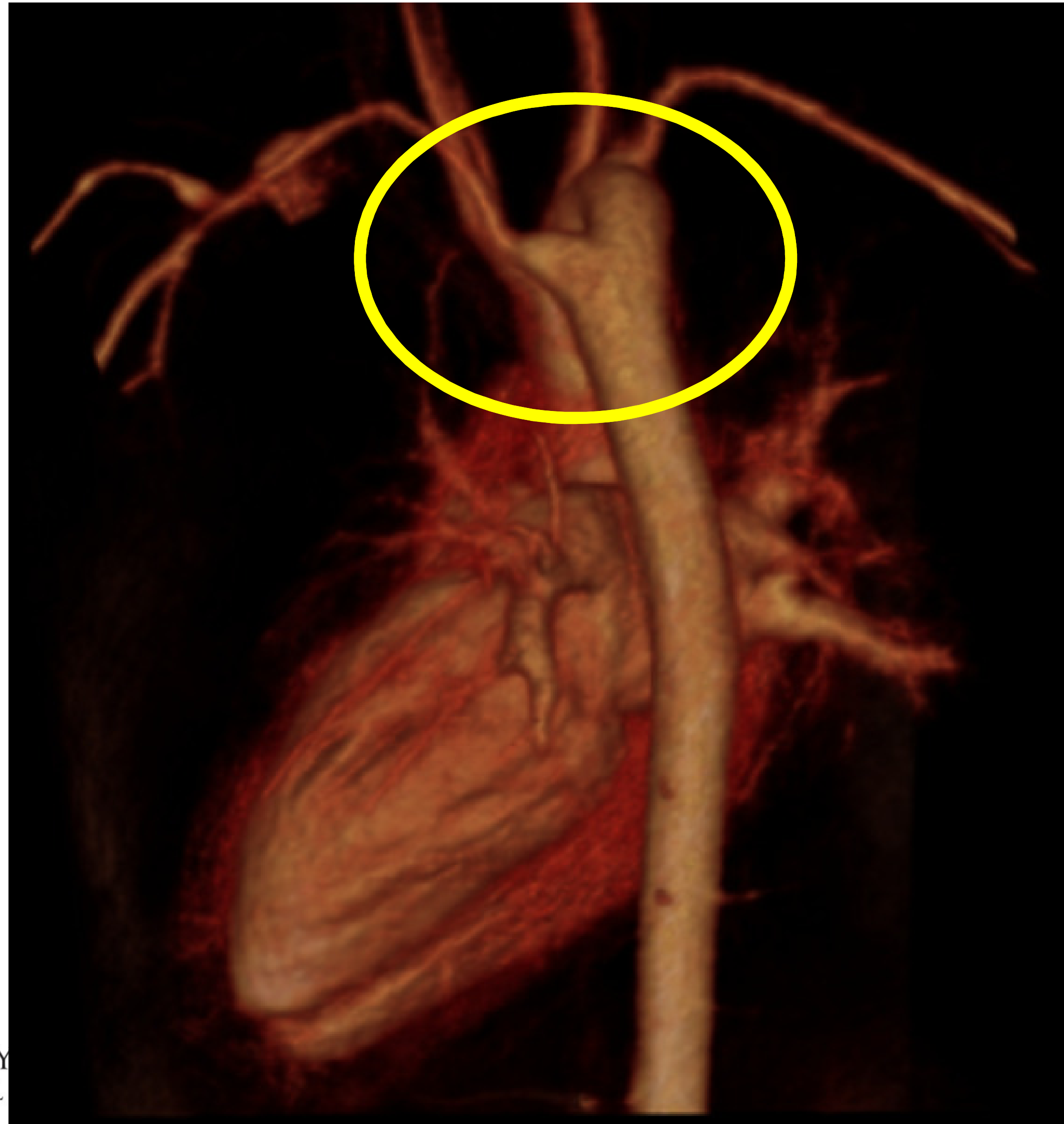
Case #2 – C. S.

- Chest Pain
- Whereas the true causes of chest pain are:
  - *Idiopathic – 21 to 45%*
  - *Musculoskeletal – 15 to 31%*
  - *Hyperventilation/psychiatric – 0 to 30%*
  - *Breast related – 1 to 5%*
  - *Respiratory – 2 to 11%*
  - *Gastrointestinal – 2 to 8%*
  - *Cardiac – 1 to 6%*
  - *Miscellaneous – 9%*
- (Geggel & Endom, 2014)

Case #3 – S.C.

- EKG obtained in cardiology clinic demonstrated normal sinus rhythm with a QTc of 405ms and no evidence of chamber hypertrophy
- Further probing by cardiologist revealed that chest pain occurred after meals
- Due to history of difficulty swallowing food, an echocardiogram was obtained which demonstrated a right aortic arch with an aortic arch anomaly. Therefore, an MRI/MRA of the chest and a barium swallow was obtained.

Case #3 – S.C.



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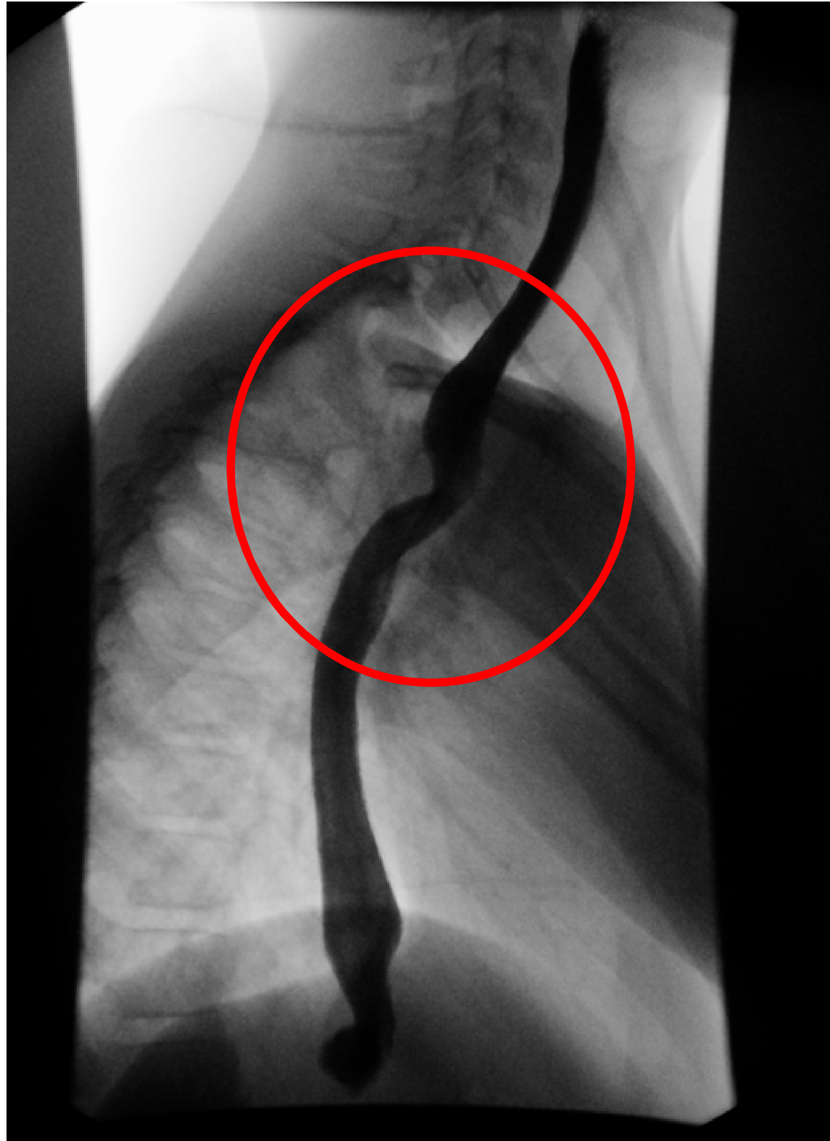
Case #3 – S.C.



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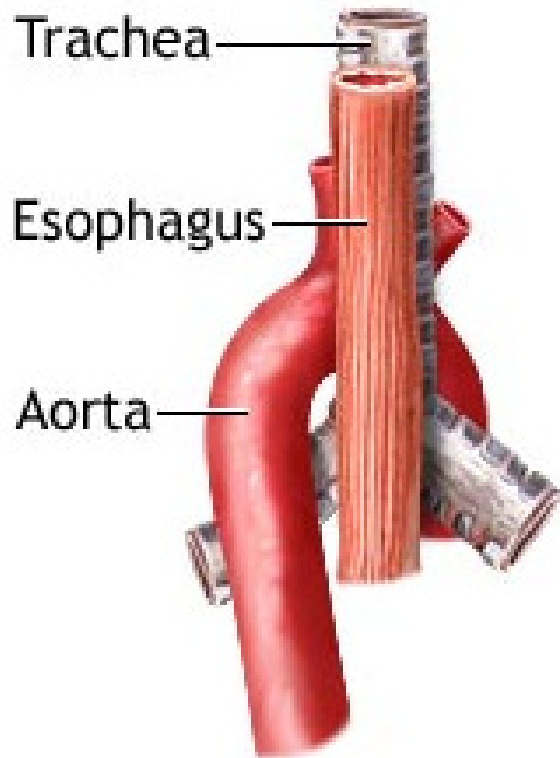
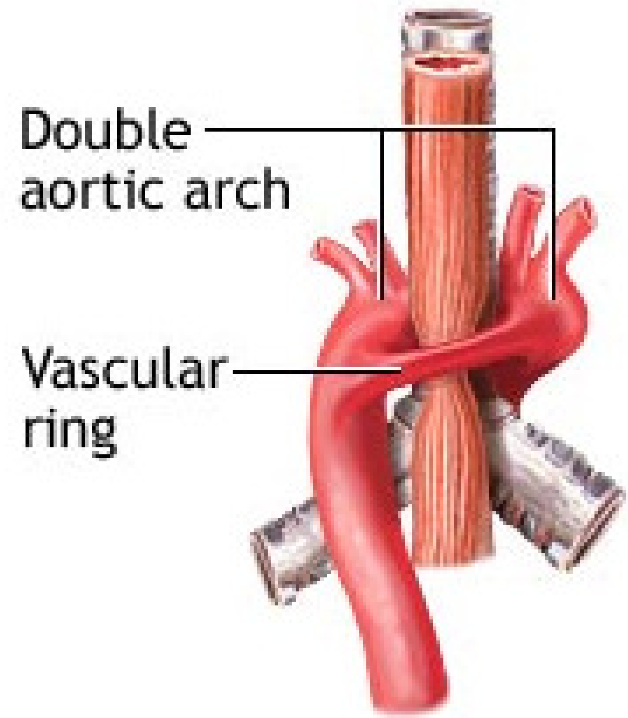
Case #3 – S.C.

Normal

Vascular ring





View from the back

A.D.A.M.

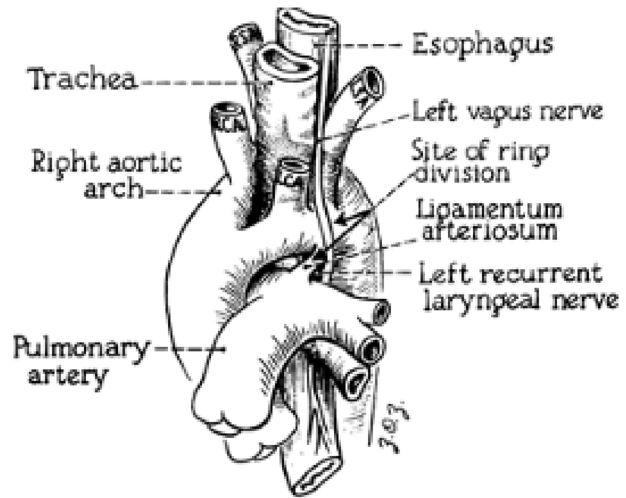
Case #3 – S. C.

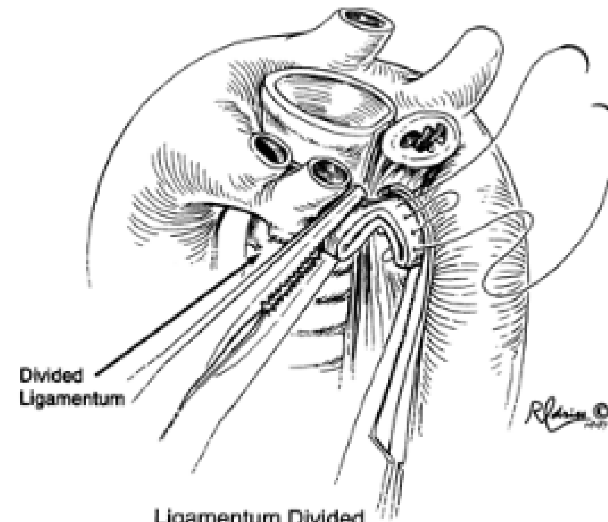
- S. C. was seen again in cardiology for follow-up from his imaging tests, results were shared with the mother and explained to her that he would need surgery to fix the vascular ring
- Additionally, since his last visit, S.C.'s mother reports that she has had time to reflect on his eating habits. She reports that as a baby, S.C. was not able to tolerate stage 2 or stage 3 baby foods. Currently, she states that he eats meat infrequently and prefers vegetables and softer starchy foods. He frequently takes a long time to chew his food. For instance, that morning, he had waffles and 2 sausage links. His mother states that it took him 1 1/2 hours to eat this meal. She states that he chews his food "beyond the (softened) consistency that others would consider appropriate for swallowing." All of his foods are either of a creamy consistency or it takes his a long time to eat. When asked, S.C. states that he has to chew his food to a very fine consistency to avoid choking.

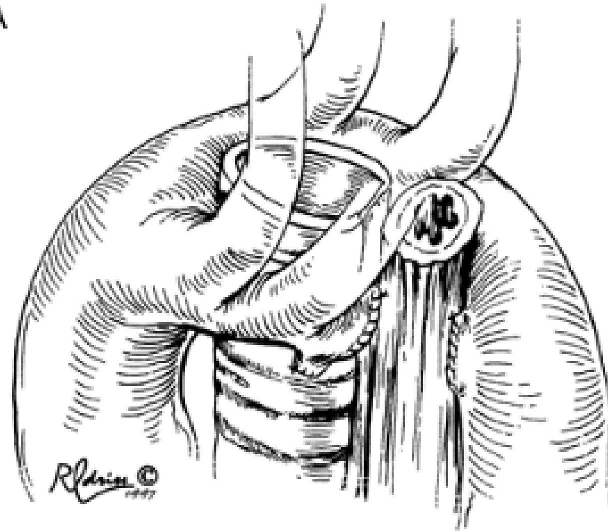
Case #3 – C.S.

- S.C. was seen by Pediatric GI prior to CT Surgery to confirm diagnosis and for further work-up.
- Upper GI – "Findings compatible with the patient's known right aortic arch and aberrant left subclavian artery with characteristic impressions on the right and posterior aspect of the esophagus. Questionable minimal indentation of the left aspect of the esophagus on the AP projection, may reflect the abnormal tenting of the origin of the left subclavian seen on prior MRI exam, raising the possibility of an atretic vascular ring."
- A PPI (Prevacid) was trialed for 30 days with no improvement in symptoms
- Therefore, S.C. was referred for cardiac surgery

Case #3 – S.C.







Case #3 – C.S.

- Post-operatively, C.S. did well and was discharged from the hospital the next day following his surgical repair
- At his first post-operative appointment, his mother reported, “He is eating much better. He eats multiple meals and at times he is eating too fast. He often asks for second and third helpings within a short period of time.”
- His chest pain has also resolved…

Case #4 – J. B.

- J. B. is a 10 year old female who was previously healthy who presents with a 4 day history of increasing right ankle pain and fever. Her symptoms started 4-5 days ago when she woke up with ankle pain. No history of trauma. She went to Patient First and an XR done was negative, so she was diagnosed with an ankle sprain and sent home with an aircast. The next day she developed another fever, noticed her ankle was swollen with continued pain and now trouble bearing weight. She went to an outside ED with low grade fever and nausea. An ankle X-ray was done negative again for fracture. J. B. did receive treatment at that time with an albuterol neb x1 for some heavy breathing. She was sent home. The following day she developed vomiting and fever up to 104. She continued to have increasing pain in her ankle, decreased PO intake, persistent fevers >102. The family therefore took her to another outside ED where she was admitted to a pediatric floor for increasing pain and refusal to ambulate, fever to 104, and nausea.

Case #4 – J. B.

- At the OSH, her admission labs included positive blood cultures, acute thrombocytopenia, abnormal coags, and clinical respiratory distress. A CXR demonstrated bilateral lower lobe infiltrates. Blood cultures grew gram + cocci in clusters, and her platelets dropped from 101 to 39. She further developed hypoxia and tachypnea requiring oxygen via NC.
- She was therefore transferred to the UMMC PICU for concern for SIRS. She received vancomycin, ceftriaxone, and zosyn prior to transfer.

Case #4 – J. B.

- Past Medical History – asthma, anxiety, depression, obsessive-compulsive disorder, ADHD
- Prenatal History – born at 36 weeks via c-sxn, mother with gestational diabetes, newborn jaundice
- Past Surgical History – Tonsil & adenoidectomy in 2010
- Family History – No history of congenital heart disease, mom with T2 diabetes, grandmother with HTN, uncle with HTN, DM, and CAD
- Previous Hospitalizations – None
- Social History – Lives at home with both parents and sister, in 4th grade
- Medications – Wellbutrin 300mg Qam/150mg Qpm; Adderall 25mg Qam/10mg Qpm; Hydroxyzine 20mg BID, Albuterol inhaler PRN for SOB/wheezing
- Allergies – NKA
- Immunizations - UTD

Case #4 – J. B.

- ROS – +fever at home x4 days, tachycardia, tachypnea upon transfer, + complaints of periumbilical pain, nausea, vomiting, complaints of bilateral knee pain, difficulty with ambulation, multiple scabs and open skin lesions on extremities, and anxiety.
- Of note, she was healthy prior to her acute illness. Mom and Dad do report that she was camping about 2 weeks ago in Philadelphia but they have not noticed any rashes since then. She is an overall nervous person and will scratch at her arms when she is anxious, and she has multiple healing scabs from bug bites, with some excoriations from picking.

Case Study #4 – J. B.

- Physical Exam (Admission to UMMC PICU):
- Wt 58kg, Temp 36.9, BP 105/55, RR 43, P 135, 96% on 1L NC
- General: anxious appearing but resting, cooperative, no acute distress
- HEENT: NCAT, ext ears nml, no rhinorrhea, PERRLA, EOMI, no conjunctival injection, MMM, OP clear, no oral lesions, normal dentition, no erythema or exudate
- Neck: supple, no masses
- Resp: tachypneic but equal air movement bilaterally, decreased at the bases, rales appreciated L>R
- CV: tachycardic, no appreciable murmur, normal S1, S2, no rubs, no gallops, 2+ peripheral pulses, CRT<2s
- Abd: soft, mild tenderness at periumbilical area, no guarding
- Lymph: no lymphadenopathy appreciated
- MSK: decreased ROM of right leg at the ankle compared to the left, mild swelling of ankle R>L, temperature equal both sides, no surrounding erythema, left knee also tender to palpation with no increased swelling compared to the right, also with decreased range of motion, normal ROM of upper extremities
- Neuro: CN's II-XII grossly intact, no focal deficits, anxious but alert and oriented
- Skin: multiple patches of healing bug bite wounds on extremities, scabbed over excoriations on wrists bilaterally, no surrounding erythema or swelling, no active bleeding

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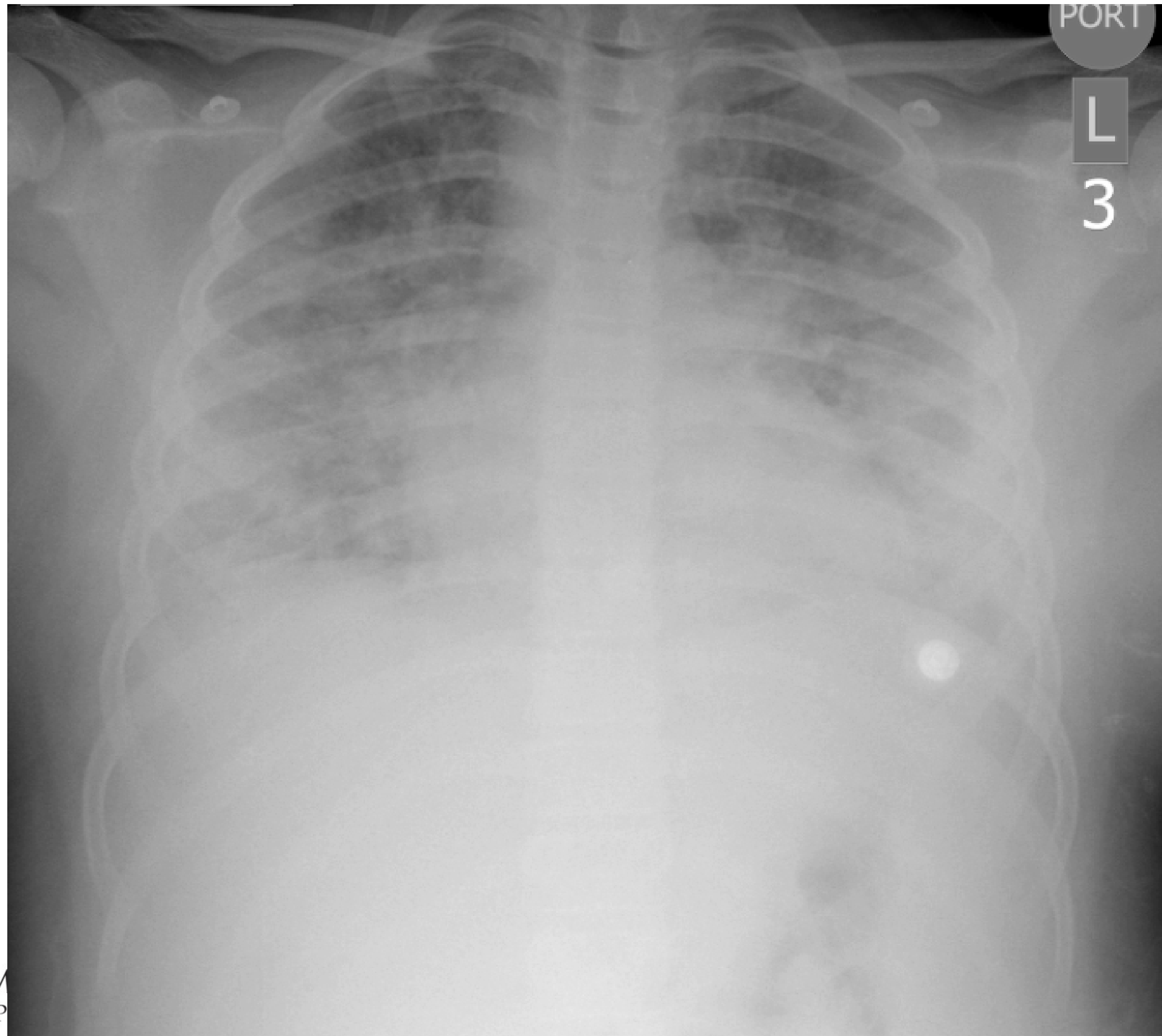
Case #4 – J.B.

- Laboratory Data (UMMC PICU Admission)
- WBC 5.2/ Hgb 10.8/ Hct 31.5/ Plt 37
- S 57/ B 11/ L 26/ M 4
- CRP 22.1
- Na 145/ K 3.5/ Cl 108/ BUN 8/ Cr 0.48/ glc 96
- Ca 8.3/ Mag 1.9/ Phos 3.4
- TPr 5.1/ Alb 2.6/ AST 73/ ALT 58/ Alk Phos 211/ TBili 0.7
- Fibrinogen 349/ PT 15.5/ INR 1.2/ PTT 40
- ESR 12

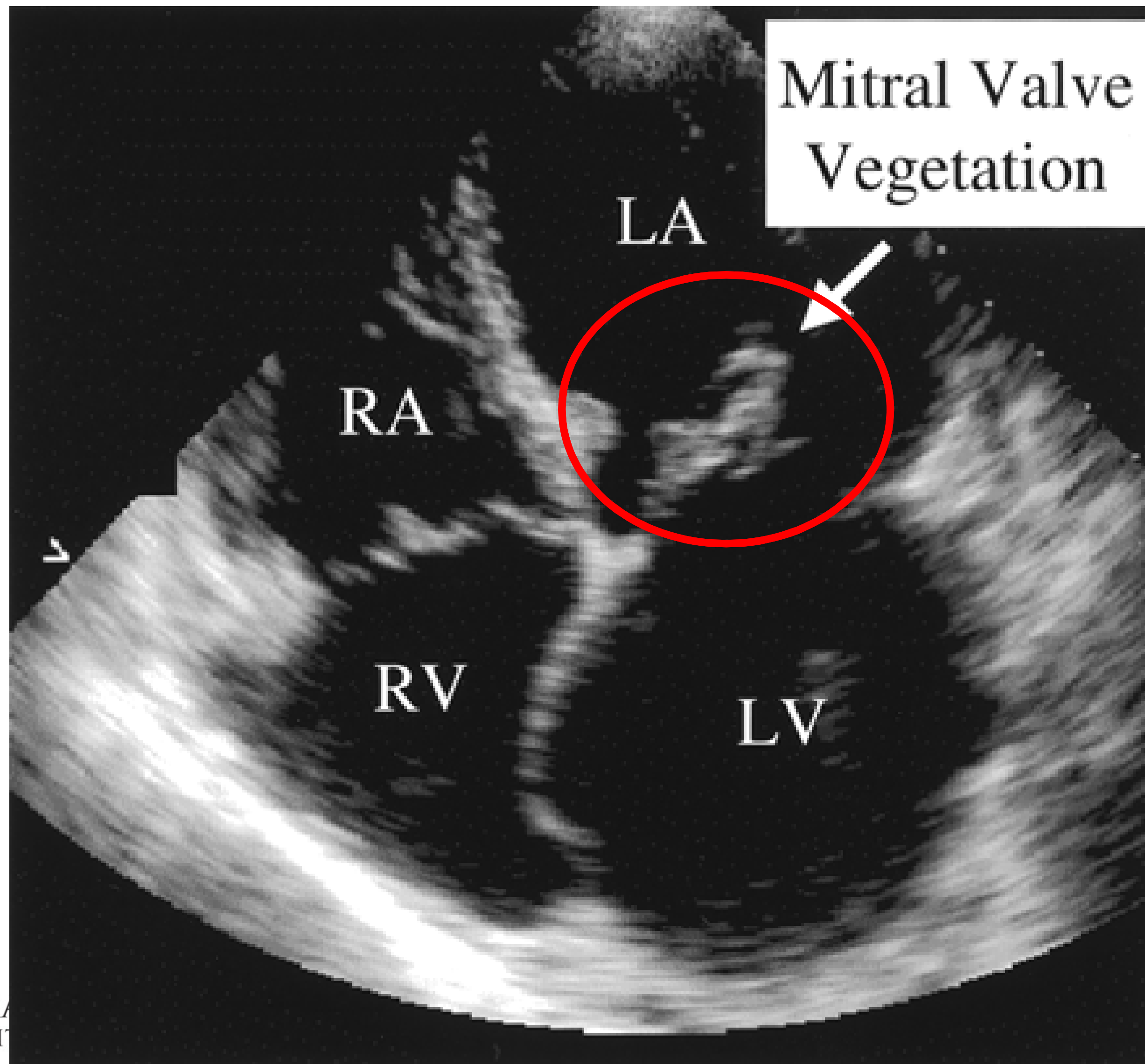
Case #4 – J.B.



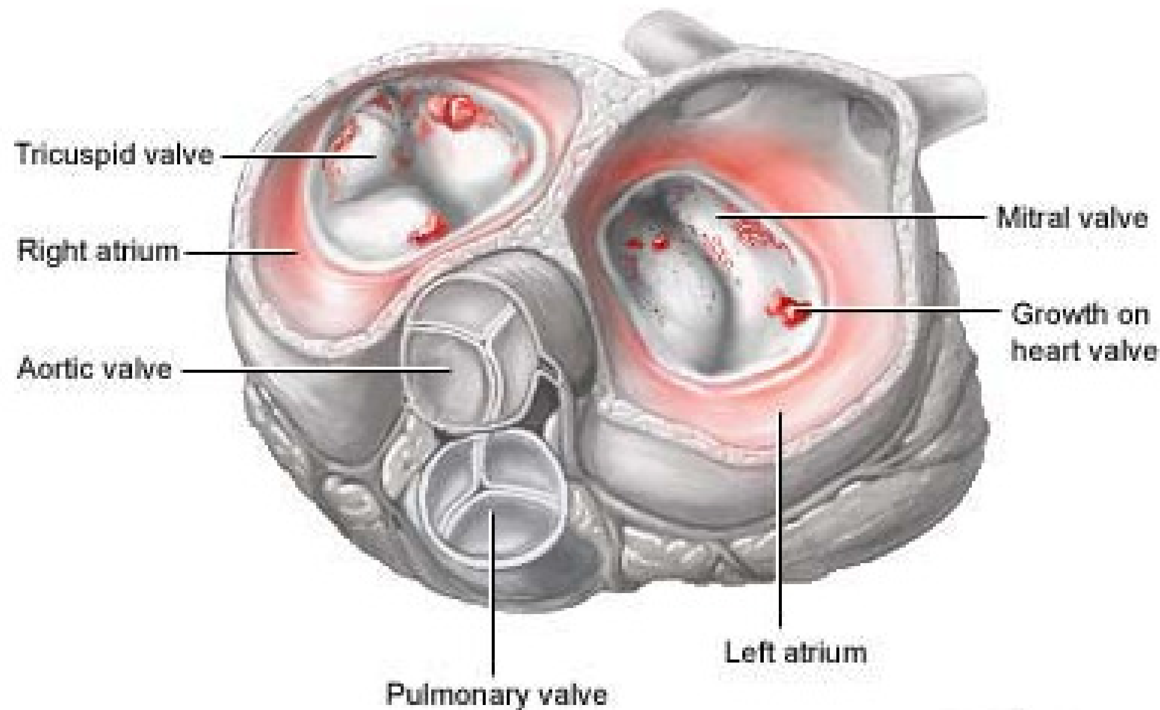
Case #4 – J.B.

- Hospital Course by Systems
  - *Respiratory – J.B. was intubated on hospital day #2 for increasing WOB and tachypnea and ultimately respiratory failure*
  - *GI – Abdominal CT demonstrated splenic infarct*
  - *ID – known gram + cocci in 2/2 bottles – further speciation to MSSA; with ID guidance, J.B.'s antibiotic regimen included nafcillin, clindamycin, & gentamycin*
  - *Heme – thrombocytopenia resolved without intervention and appropriate treatment of sepsis*
  - *Neuro – head CT demonstrated possible embolic infarcts*
  - *And CV – an echocardiogram was obtained due to cardiomegaly on CXR…*

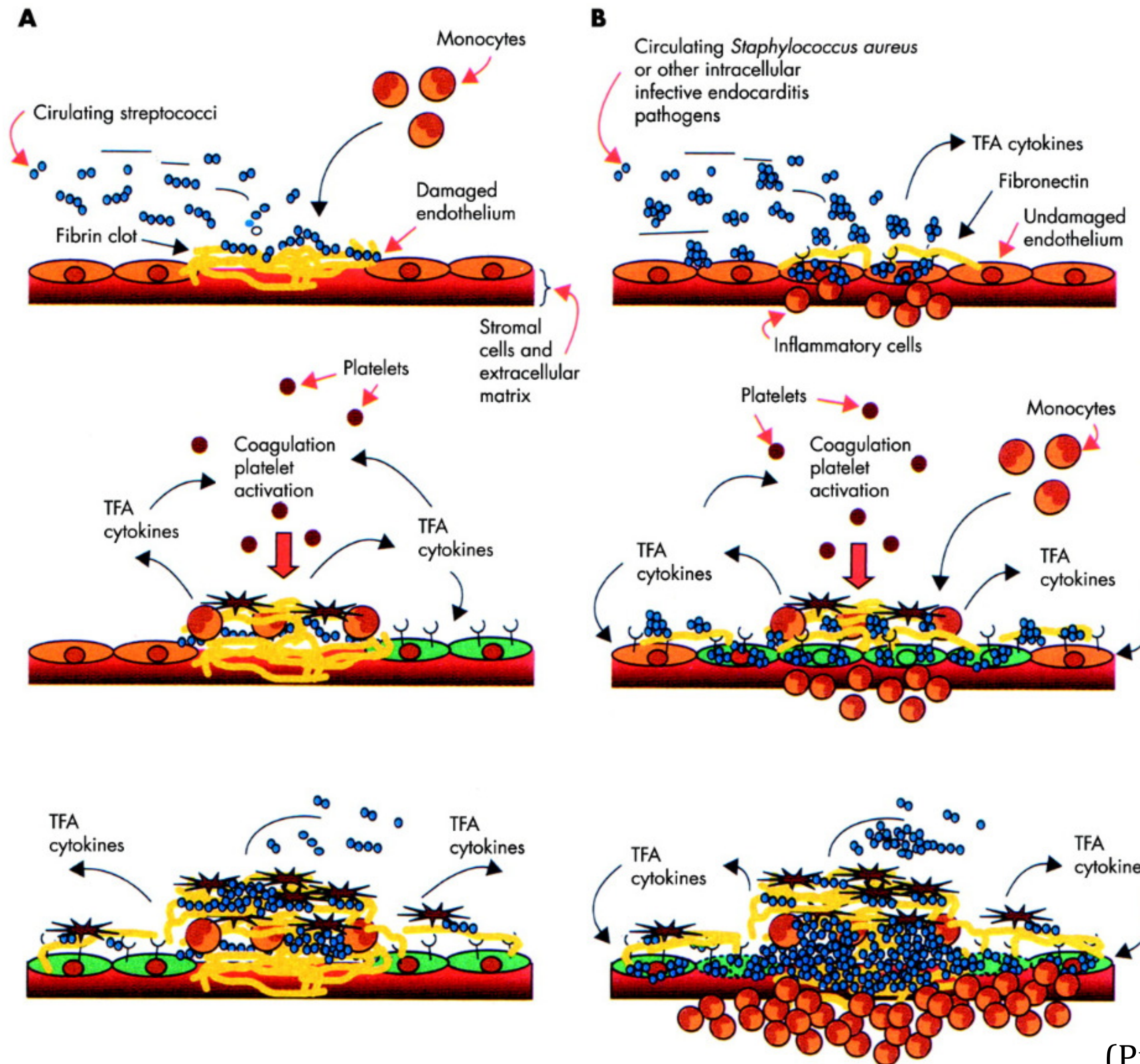
Case #4 – J. B.



Case #4 – J. B.



Case #4 – J. B.



(Prendergast, 2006)

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Case #4 – J. B.

- Modified Duke Criteria for the Diagnosis of Infective Endocarditis (IE)
  - *Definite IE*
    - – Pathologic Criteria
      - Microorganism: demonstrated by culture or histology in a vegetation, or in a vegetation that has embolized, or in an intracardiac abscess **OR**
      - Pathologic lesions: vegetation of intracardiac abscess, confirmed by histology showing active endocarditis
    - – Clinical Criteria
      - 2 major criteria **OR** 1 major and 3 minor criteria **OR** 5minor criteria
  - *Possible IE*
    - – 1 major criterion and 1 minor criterion **OR** 3 minor criterion
- (Li et al., 2000)

Case #4 – J. B.

- Major Criteria
  - *Positive blood cultures for IE*
    - From 2 separate blood cultures
    - Persistently positive blood culture, defined as recovery of a microorganism consistent with IE from
      - Blood cultures drawn more than 12 hours apart **OR**
      - All of 3 or a majority of 4 or more separate blood cultures, with 1st and last drawn at least one hour apart
  - *Evidence of endocardial involvement*
    - Positive echocardiogram for IE
    - New valvular regurgitation

- Minor Criteria
  - *Predisposition – predisposing heart condition or IV drug use*
  - *Fever of >38*
  - *Vascular phenomena – major arterial emboli, septic pulmonary infarcts, mycotic aneursym, intracranial hemorrhage, conjunctival hemorrhages, Janeway lesions*
    - Janeway lesions – small, non-tender, erythematous or hemorrhagic macular or nodular lesions on the plams or soles (only seen in IE)
  - *Immunologic phenomena – glomerulonephritis, Osler's nodes, Roth spots, rheumatoid factor*
    - Osler's nodes – painful, red, raised lesions on hands or feet
    - Roth spots – retinal hemorrhages with white or pale centers
      - Both are not only seen in IE, also with other immune complex mediated vasulitises
  - *Microbiologic evidence – positive blood culture not meeting major criterion*
- (Li et al., 2000)

Case Study #4 – J. B.

- The "demographics" of infective endocarditis (IE):
  - *Most common organisms: Streptococcus viridans & Staphlococcus aureus*
  - *~75% of patients have previously known congenital heart disease, ~66% of which had previous cardiac surgery*
  - *~30% of patients require full valve replacement (bioprostethic or mechanical)*
  - *~35% of patients have complications which include mycotic aneurysms, myocardial abscess formation, or emboli*
  - *Overall mortality for IE is 4% with fungal infections having the highest mortality*
- (Johnson, et al., 2012)

Case #4 – J. B.

- Indications for surgical intervention vs. medical management of infective endocarditis:
- Heart failure
- Unable to control infection; continued fever and elevated inflammatory markers despite appropriate antibiotics treatment x1 week
- Previous embolic episode; vegetation >10mm in size and mobile
- Associated perivalvar infection/aneurysms
- Valvar obstruction by vegetation
- Prosthetic valve endocarditis
- Fungal endocarditis
- Difficult to treat or highly resistant organisms
- Associated neurologic complications, if hemorrhagic stroke—delay surgery 3-4 weeks due to anticoagulation with bypass; if ruptured mycotic aneurysm—it must be resected/clipped/embolized prior to cardiac surgery
- (Delahaye et al., 2004)

Case #4 – J. B.

- J.B. was therefore taken for cardiac surgery for repair of her mitral valve and removal of the vegetation (HD#3)
- Continued hospital course by systems
- CV – did well hemodynamically following surgery, minimal inotrope need, post-operative echo with good result and normal biventricular function
- RESP – extubated on POD #3 but did have long high flow cannula requirement due to persistent pleural effusions and respiratory distress, effusion necessitated pigtail placement with resultant resolution of increased WOB and hypoxia and decreased oxygen requirement
- ID – vegetation removed also grew MSSA, daily blood cultures + for MSSA until HD#10, rifpampin and nafcillin continuous infusion was needed for better penetration with resultant negative blood cultures; immunologic w/u was normal; was discharged home with PICC on continuous nafcillin infusion for 6 weeks
- FEN – full enteral (tube feed) diet by POD#3 and advanced to regular diet PO diet after resolution of respiratory distress; repeat abdominal US with continued splenic infarcts however repeat ultrasound at outpatient follow up demonstrated near resolution of infarcts
- OPTHO – mild optic nerve damage of both eyes; outpatient exam obtained 1 month after discharged demonstrated resolution of optic nerve edema with no residual effect
- NEURO – repeat head/neck MRI with punctate lesions within developing abscesses, neurosurgery/neurology consults determined no acute surgical management was needed; keppra added for seizure prophylaxis
- ORTHO – MRI of ankle was not concerning for osteomyelitis

Case #4 – J. B.

- Sub-Acute Bacterial Endocarditis (SBE) Guidelines
  - *Required for:*
    - Prosthetic valves
    - History of infective endocarditis
    - Unrepaired cyanotic CHD, including palliative shunts and conduits
    - Completely repaired defect with prosthetic material or devices done via surgery or cardiac cath (x6 months)
    - Repaired CHD with residual defects at the site of or adjacent to the repair
    - Cardiac transplant patient with valvular disease
  - *Antibiotic regimen (taken once, 1 hour prior to dental procedure/cleaning):*
    - Amoxicillin 50mg/kg
    - If allergic: Clindamycin 20mg/kg

Case #4 – J. B.

- Red Flags from this case's presentation:
  - *Fever!!*
  - *Nausea & Vomiting*
  - *No history of trauma; negative x-rays*
  - *"Heavy Breathing"*

Pearls/Take Home Information

- Always stress the importance of cardiology follow-up, I like to use the phrase "you're married to cardiology for life!"
  - *We unfortunately use the term "fixed" quite frequently, this gives the family a false sense of everything being perfect forever*
  - *~25% of patients are not followed by CHD specialists with a median duration of loss to follow-up being 22 years (!); ~50% of late deaths related to CHD have occurred in patients not under specialist care (Wray, 20103)*
- Most kids do not need to be restricted from sports/activities, obesity is a real problem in children with heart disease
- An echo always isn't the answer, sometimes it's not needed and other times other imaging is more appropriate
- Thorough history taking is important
- Not all murmurs and chest pain needs to be referred
  - *Kids are not having heart attacks!!*

Questions???

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