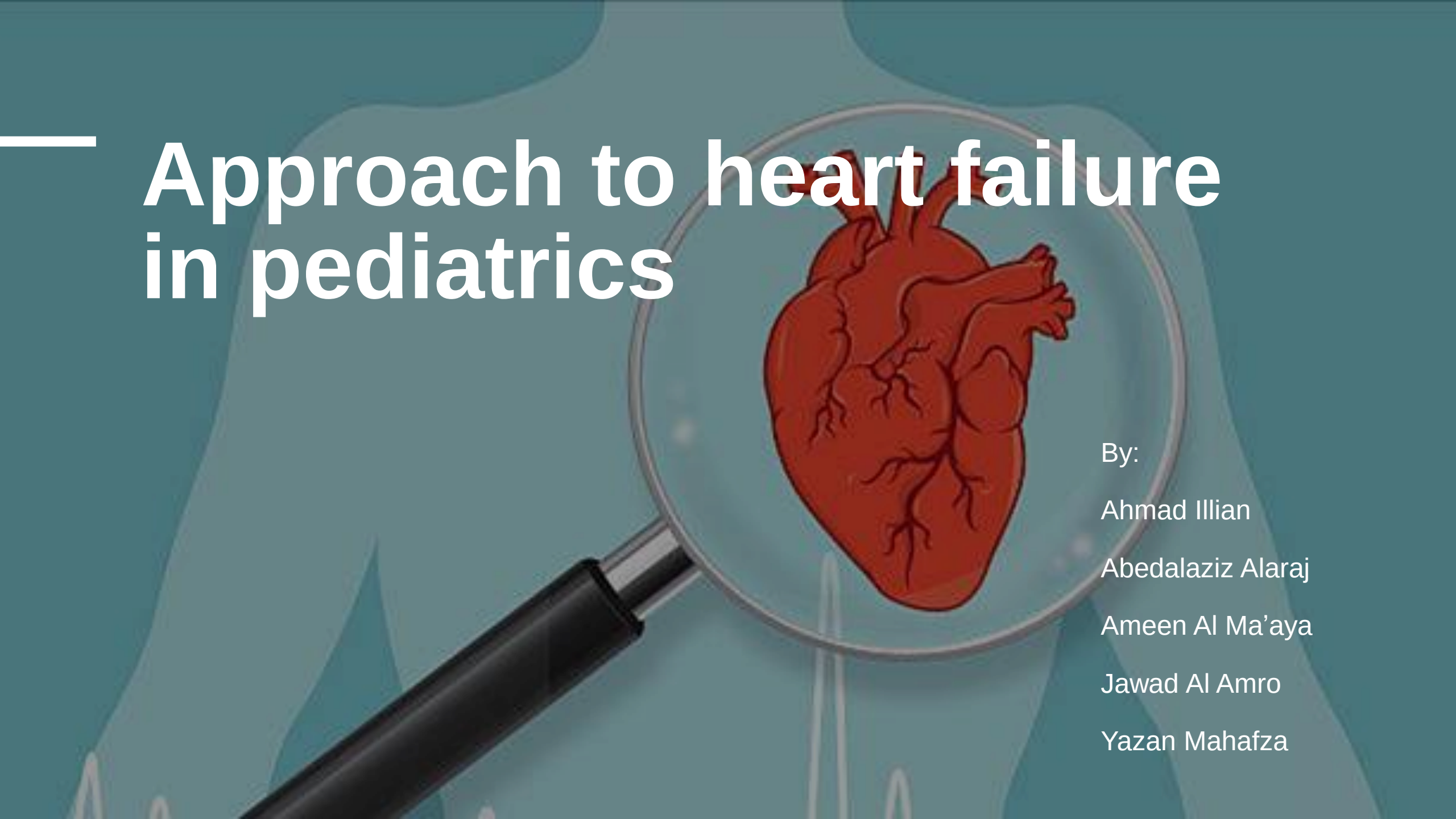
Approach to heart failure in pediatrics



By:

Ahmad Illian

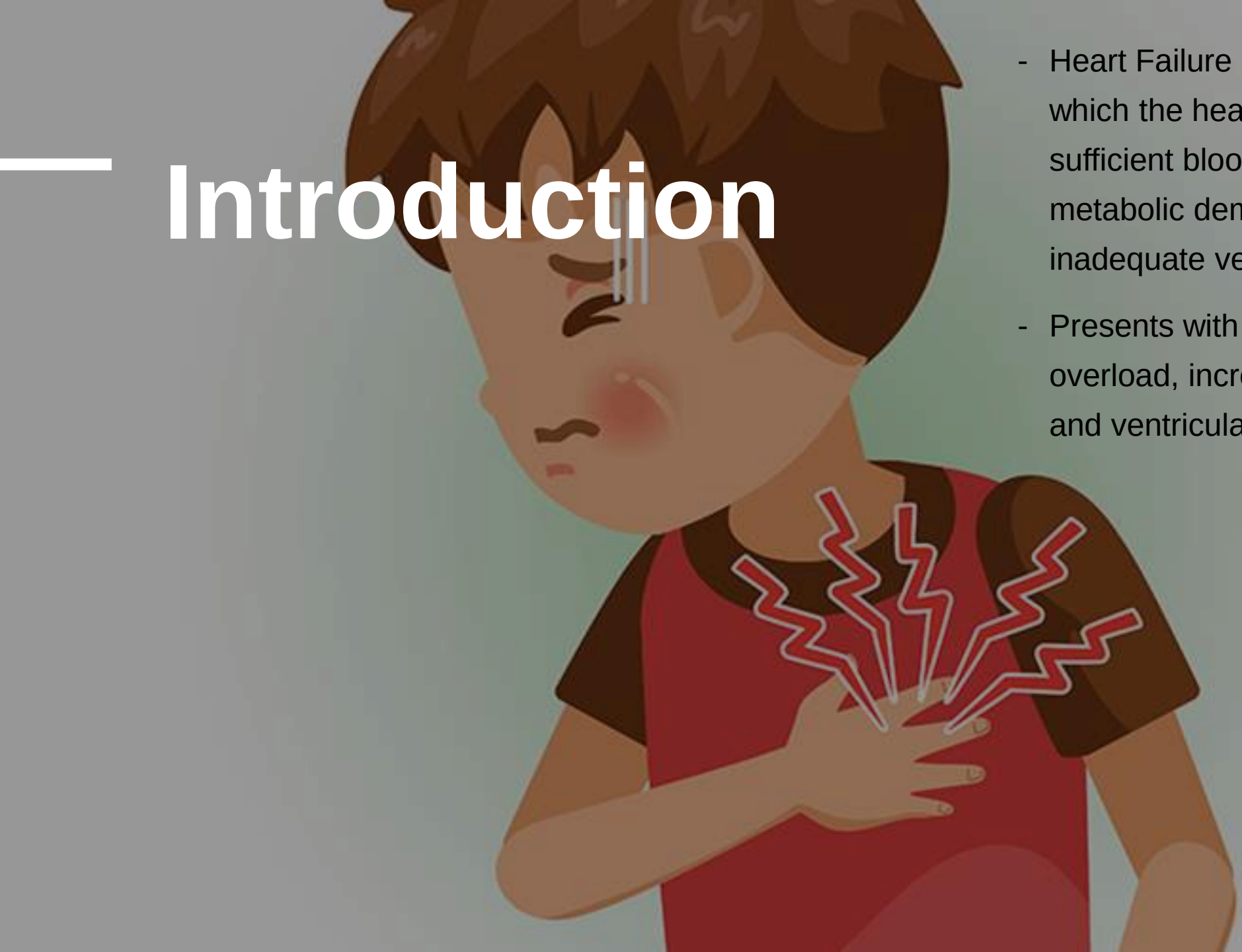
Abedalaziz Alaraj

Ameen Al Ma’aya

Jawad Al Amro

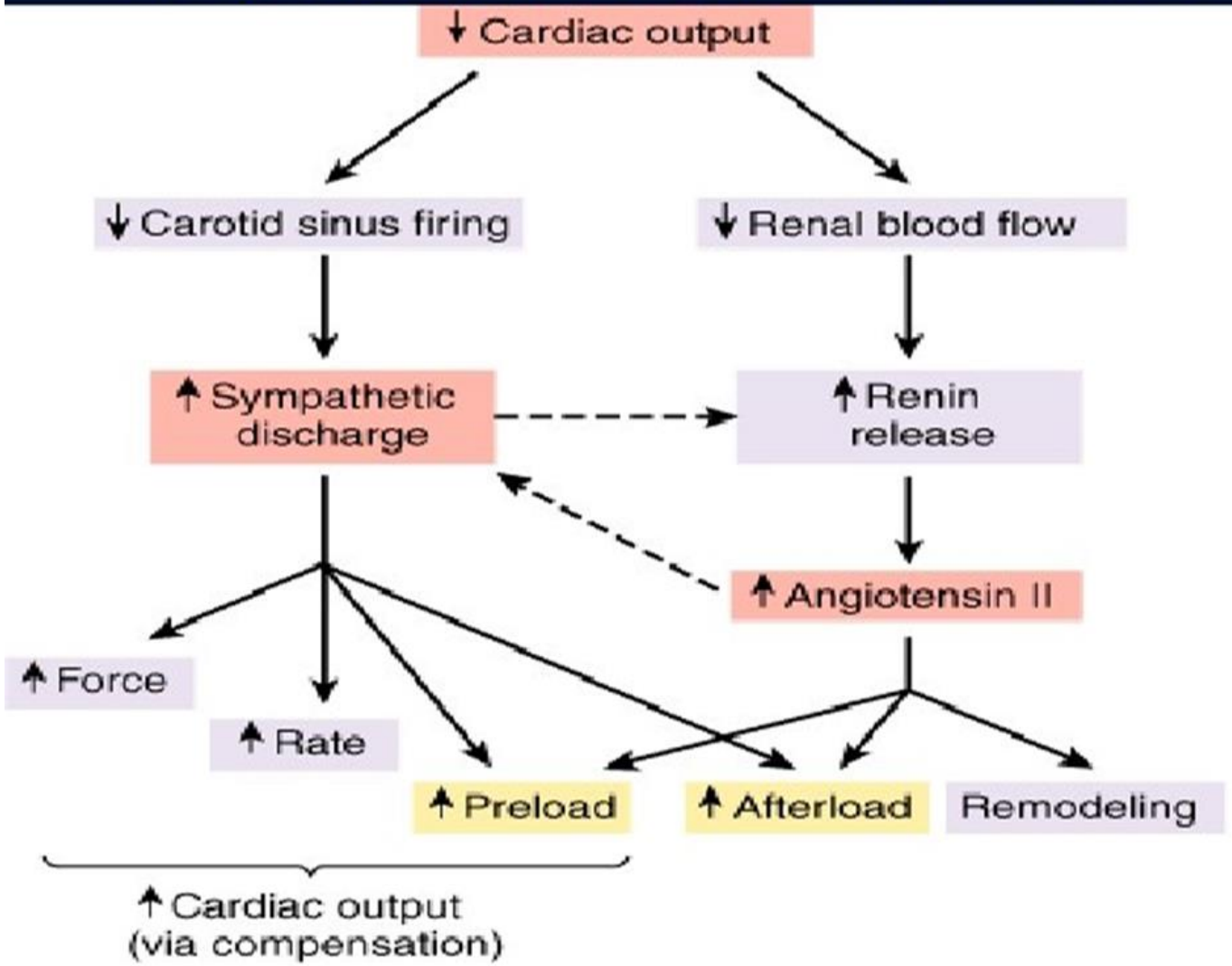
Yazan Mahafza

Introduction



- Heart Failure : a clinical syndrome in which the heart is unable to provide sufficient blood required to meet the metabolic demands of the body due to inadequate ventricular filling or pumping.
- Presents with symptoms of volume overload, increased capillary resistance and ventricular dysfunction.

Compensatory Mechanisms in Heart Failure



Source: Katzung BG, Masters SB, Trevor AJ: *Basic & Clinical Pharmacology, 11th Edition:* http://www.accessmedicine.com

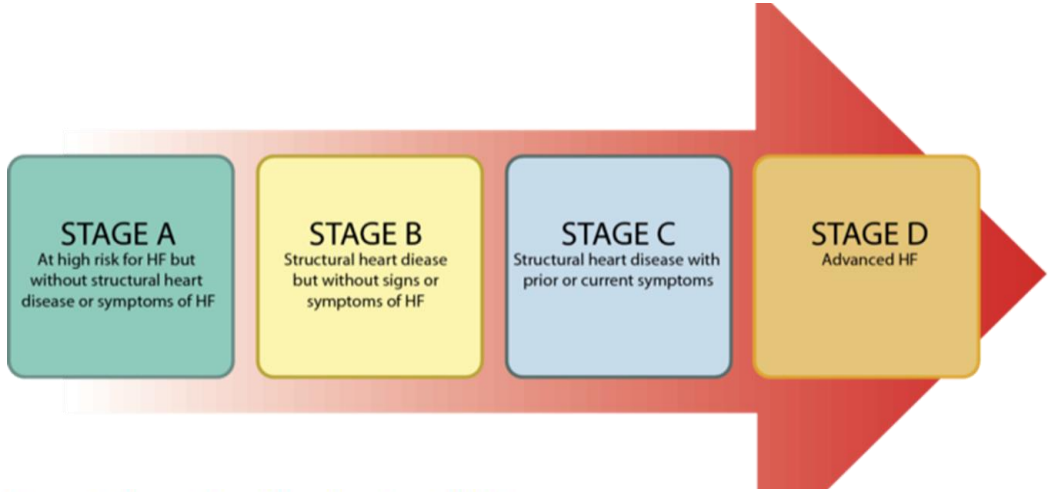
Etiology

1) Congenital heart diseases (VSD , COA ,...)
2) Acquired heart disease (cardiomyopathy , myocarditis (viral , Kawasaki disease), acute rheumatic carditis , anthracycline toxicity ,...)

timing of HF presentation is helpful in determining the cause :

- 1st week : mostly due to left heart obstruction (critical coarctation)
- 4-8 weeks : left to right shunt (VSD , AVSD)
- 6-8 years : acquired heart disease (cardiomyopathy , myocarditis)

CLASSIFIED ACCORDING SEVERITY : THE NEW YORK HEART ASSOCIATION (NYHA) AND ROSS CLASSIFICATIONS



STAGING – THE STAGING SYSTEM OF PEDIATRIC HF (STAGES A TO D) IS USED TO DESCRIBE THE DEVELOPMENT AND PROGRESSION OF DISEASE FOLLOWING EXPOSURE TO A RISK FACTOR FOR HF.

NYHA and Modified Ross Heart Failure Classification for Children

	NYHA	Ross
Class I	No limitations of physical activity	No limitations or symptoms
Class II	May experience fatigue, palpitations, dyspnea, or angina during moderate exercise but not during rest	Infants: Mild tachypnea or diaphoresis with feeding Older children: Mild to moderate dyspnea exertion
Class III	Symptoms with minimal exertion that interfere with normal daily activity	Infants: Growth failure and marked tachypnea or diaphoresis with feeding Older children: Marked dyspnea on exertio
Class IV	Unable to carry out any physical activity because they typically have symptoms of HF at rest that worsen with any exertion	Symptoms at rest such as tachypnea, retractions, grunting, or diaphoresis

HF: Heart failure; NYHA: New York Heart Association.

Management Options

Dependent on its etiology and severity

1- Initial Management :

- Rest / oxygen / fluid-salt restriction
- correction of exacerbating or contributing conditions : Anemia /fever / Hypertension / Renal failure / Acidosis / obesity / Thyroid disorders /…
- Nutritional support

2- Structural heart disease with preserved ventricular function : the mainstay of management involves surgical or catheter-based interventions to correct the underlying defects.

3- Ventricular pump dysfunction : therapy is provided based on the stage of HF

- Stage A : no tttx is needed (treat Predisposing conditions if possible)
- Stage B : ACEI (ARBs if ACEI intolerant)
- Stage C : -ACEI and aldosterone antagonist; oral diuretic (as needed for fluid overload) ; low-dose digoxin
- (if needed for additional symptom relief)
- -After a few weeks of stability, a BB is added in patients with persistent LV dilation & dysfunction
- Stage D :IV diuretics and/or inotropes

Other interventions may include positive pressure ventilation, cardiac resynchronization therapy, mechanical circulatory support, and heart transplantation

Medical Treatment

1) Preload reduction (Diuretics) :

- Loop diuretics (Furosemide)
- Thiazide diuretics
- potassium-sparing diuretic (spironolactone)

- 2) Afterload reduction (vasodilators) :

-ACEI (enalapril)

-Beta blockers (carvedilol /metoprolol)

3) Contractility enhancement (Inotropes) :

- Dopamine

-Epinephrin

-Digoxin

DDX of HF

- Respiratory distress: TTN, RDS , meconium aspiration, CDH, pneumothorax, pneumonia, pulmonary hypoplasia, bronchiolitis, and asthma.
- Poor weight gain : protein-milk allergy ,CF, celiac disease, chronic infections, hyperthyroidism, and metabolic disorders.
- Peripheral Edema : renal failure, venous thrombosis, or adverse drug effects.
- Shock : overwhelming sepsis or hypovolemia.

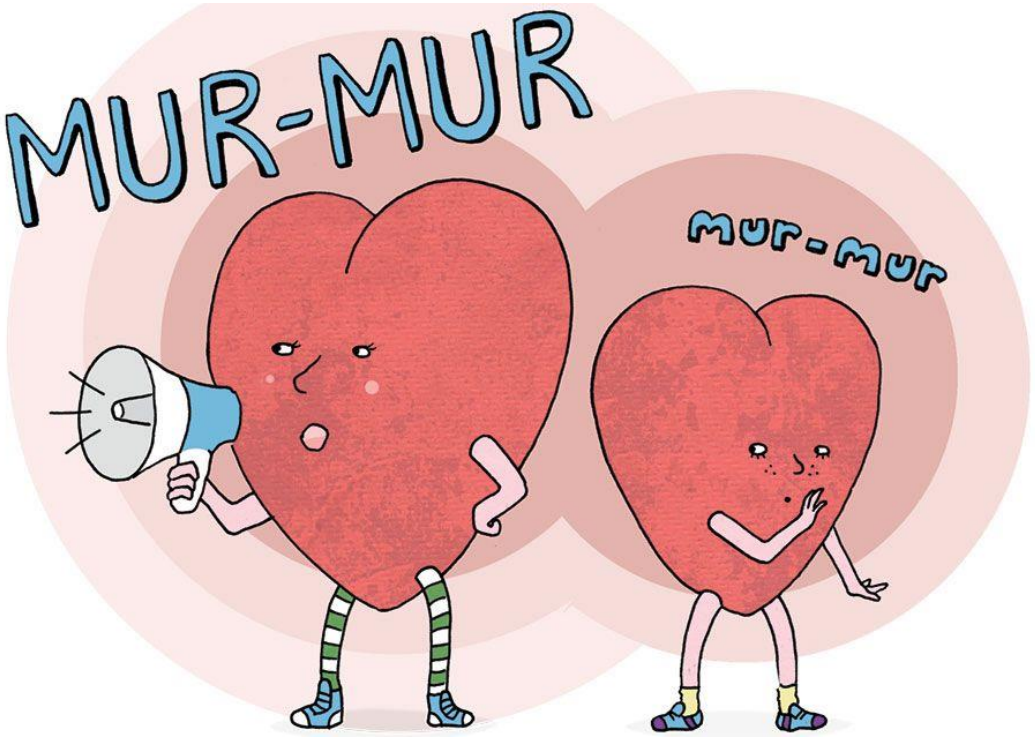
Case 1

Patient History

- 4-year-old boy
- Fatigue on minimal exertion
- shortness of breath on minimal exertion
- Rapid breathing during minor physical activities.
- Recurrent respiratory infections over the past few months
- Perinatal history reveals the patient had a longstanding history of breastfeeding problems (which included sweating, shortness of breath, and perfuse sweating) which were dismissed by the parents
- no significant family history of heart conditions or congenital anomalies

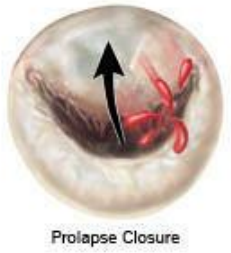
Physical Examination Findings:

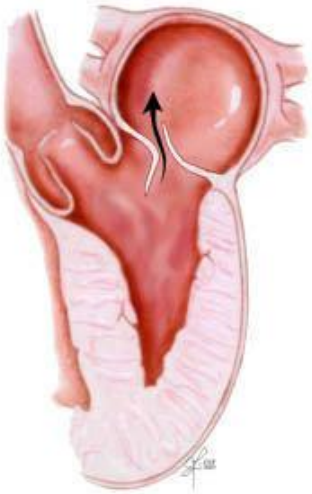
- the child appears mildly underweight but not malnourished.
- The child's heart rate is elevated
- a palpable thrill is felt on the left sternal border and hepatomegaly is noted.
- Auscultation of the heart reveals a loud, harsh, Grade 4 holosystolic murmur heard best at the lower left sternal border.
- There are no other abnormal findings in the examination of other organ systems.

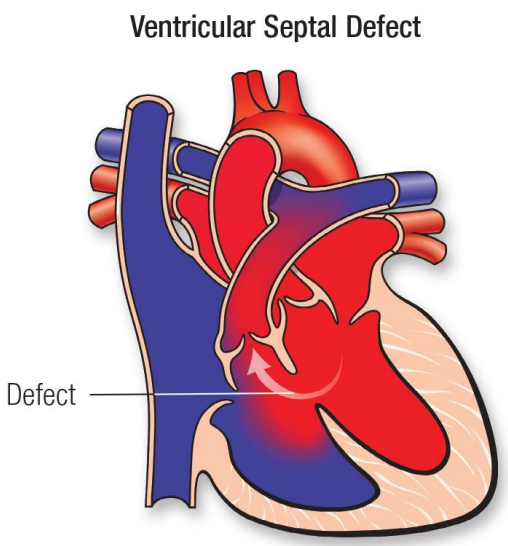


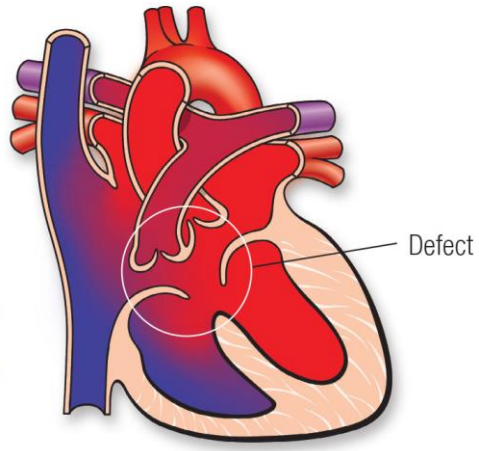
DDX and management:

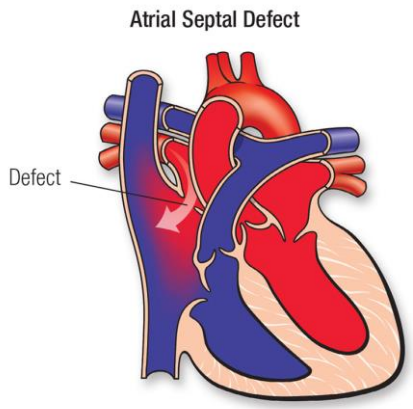
- **Ventricular Septal Defect (VSD)**
- **Atrial Septal Defect (ASD)**
- **Patent Ductus Arteriosus (PDA)**
- **Pulmonary stenosis**
- **Valve prolapse**
- **AV canal**

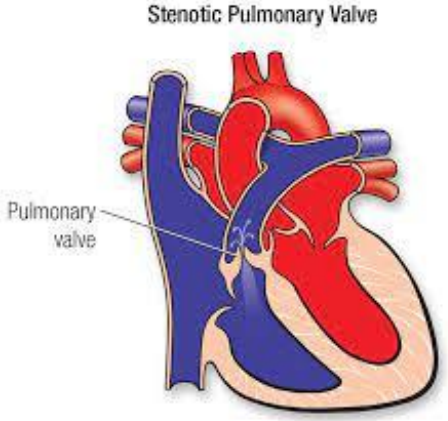


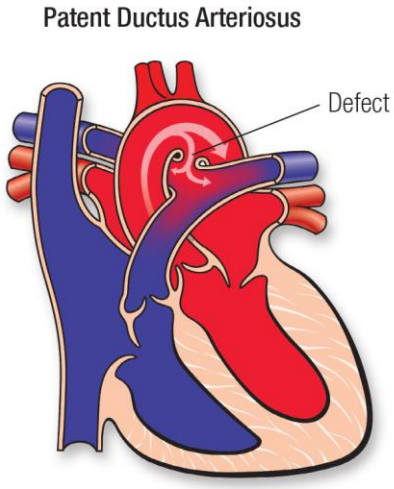










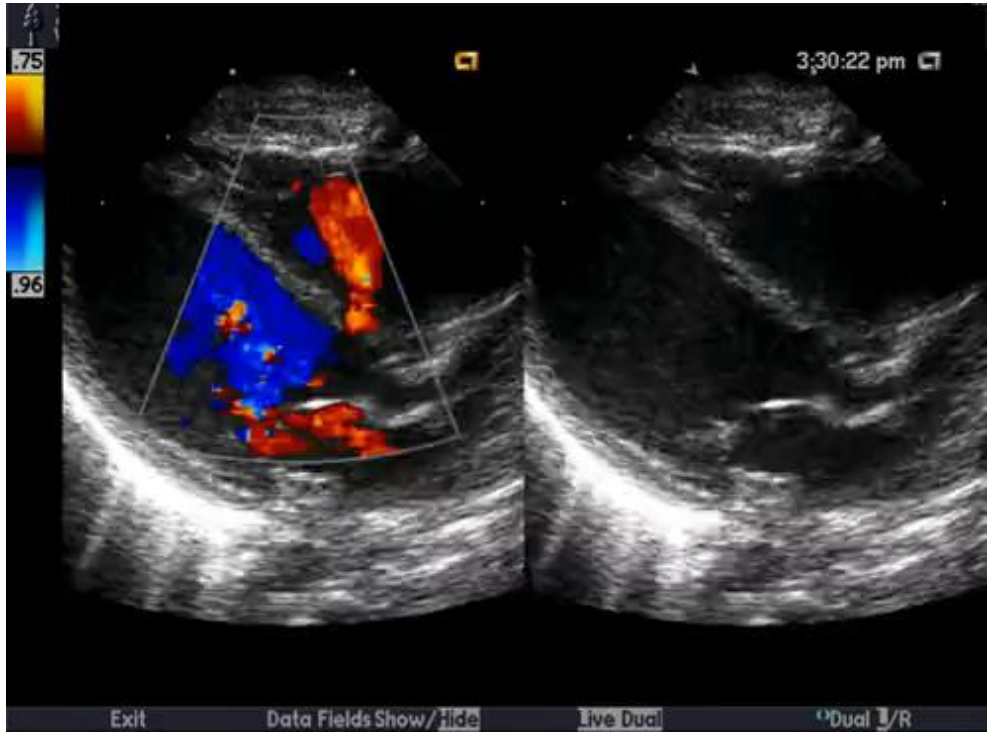


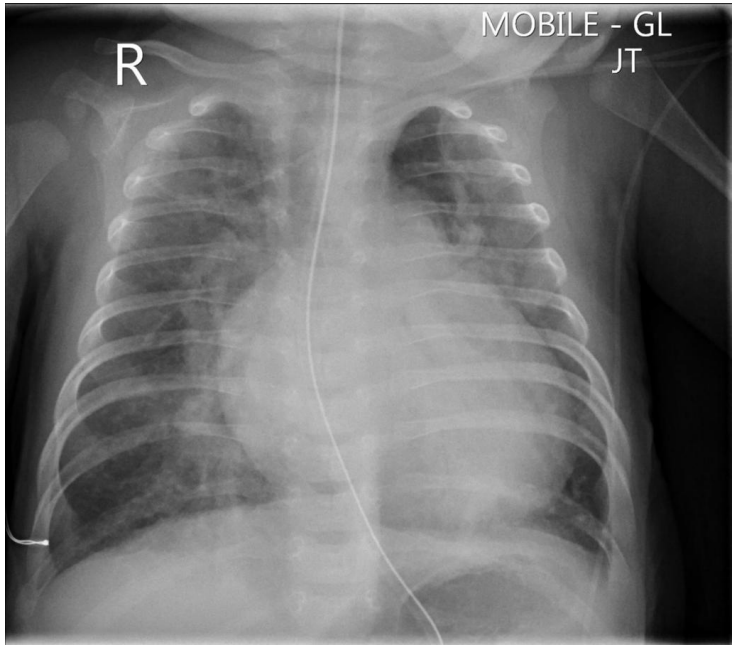
Investigations and Imaging

- Based on the clinical presentation and physical examination findings, The following tests are ordered:

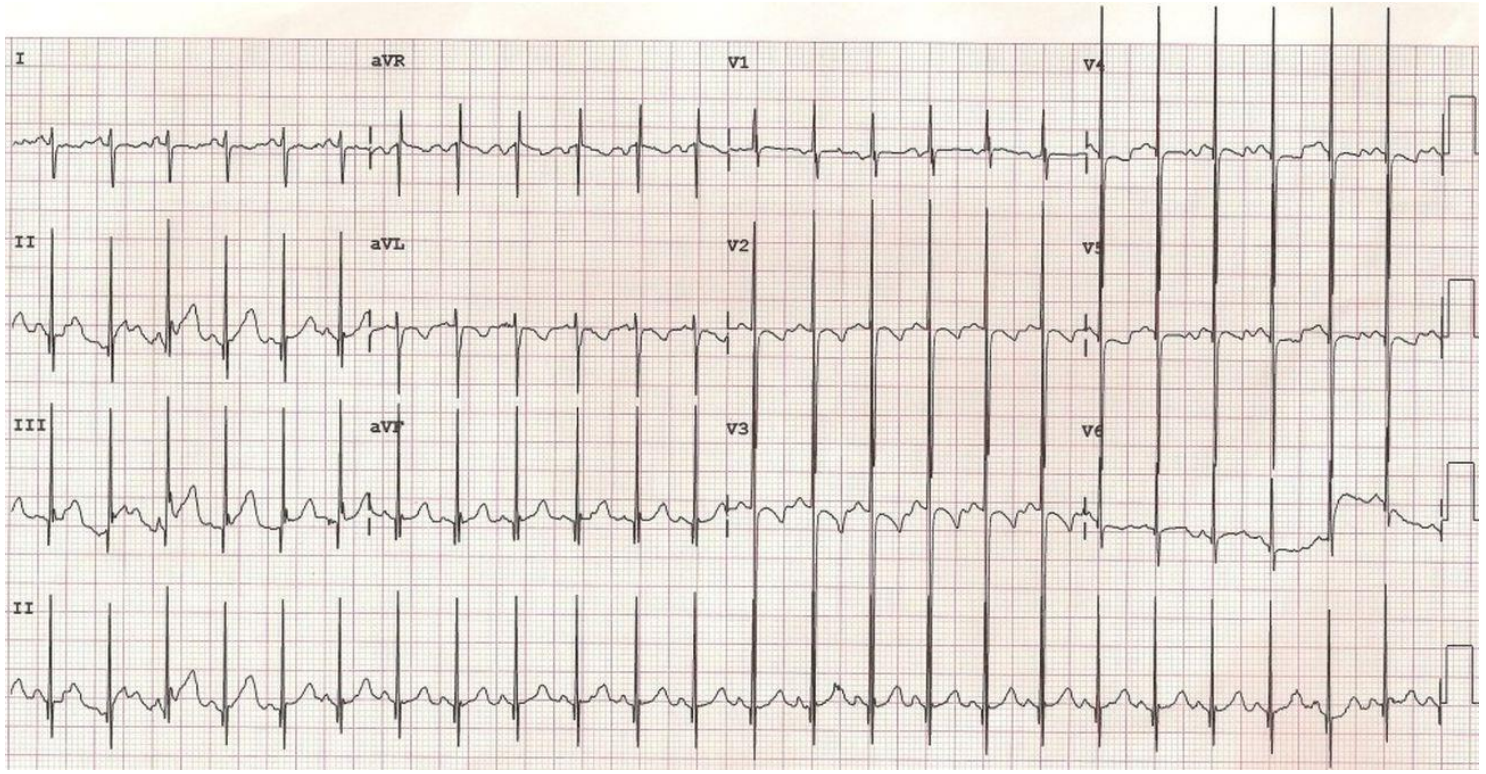
- Transthoracic echocardiography : This will provide a detailed visualization of the heart structures and help in confirming the presence of a structural defect. It will also determine the size/location of the defect and the direction of the possible shunt.

- Chest X-ray: To assess the heart's size and look for any signs of pulmonary congestion and hear failure. Electrocardiogram (ECG): To evaluate the heart's electrical activity and identify any associated arrhythmias or conduction abnormalities.





ECG findings: This large defect is associated with large left to right shunting which will have left ventricular volume overload with small q, tall R and upright T waves in lateral leads. It can progress to biventricular overload when there is hyperdynamic pulmonary hypertension.



Etiology, Classification

- VSDs are the most common congenital heart defects in children. The etiology is likely congenital. MC congenial heart disease.

- Types:
  - Peri-membranous (MC)
  - Muscular
  - Supracrustal
  - Inlet

Complications:
- Heart failure
- Pulmonary HTN
- Endocarditis

Treatment:

For small uncomplicated VSDs > Observation: Small VSDs may close on their own over time. With regular follow-up with the pediatric cardiologist to monitor the defect's size and assess any changes in symptoms.

For large/complicated VDSs without congestive HF > Surgical/Percutaneous repair: performed through open-heart surgery or minimally invasive techniques.

For complicated VSDs with congestive HF > Medical: Diuretics to decrease the preload, ACE inhibitors to decrease the peripheral resistance, Digoxin to improve the cardiac contractability.

Case 2: Viral myocarditis

DONE BY :AHMAD ILLIAN

Congestion at Rest

Low Perfusion at Rest	Congestion at Rest: NO	Congestion at Rest: YES
NO	Warm & Dry (PCW normal, CI normal)	Warm & Wet (PCW elevated, CI normal)
YES	Cold & Dry (PCW low/normal CI decreased)	Cold & Wet (PCW elevated CI decreased)

Signs/Symptoms of Congestion:
- Orthopnea / PND
- JV Distension
- Hepatomegaly
- Edema
- Rales
- Abd-Jugular Reflex

Signs/Symptoms of Low Perfusion:
- Narrow pulse pressure
- Sleepy / obtunded
- Low serum sodium
- Cool extremities
- Hypotension with ACE inhibitor
- Renal/hepatic dysfunction

HISTORY:

Jacob is 6 years old male, he was doing well until he was brought to the pediatric emergency department by his parents due to a sudden onset of symptoms that started about a week ago. He had a fever, fatigue, and a persistent cough that worsened over the last few days. Jacob's parents noticed that he was breathing rapidly , had difficulty catching his breath, even while at rest and orthopnea. He also complained of chest pain and feeling very weak and dizziness. Jacob did not take covid-19 vaccine.

PHYSICAL EXAMINATION:

-Jacob appeared pale, tired, and had labored breathing.
-His heart rate was elevated at 180 beats per minute, and his blood pressure was slightly low.
-capillary refile was 3 sec
-There was crackling sounds in his lungs,.
-pan-systolic murmur over the apex of the heart
-Jacob had lower limb edema and ascitis.
-hepatomegaly

DIFFERENTIAL DIAGNOSIS:

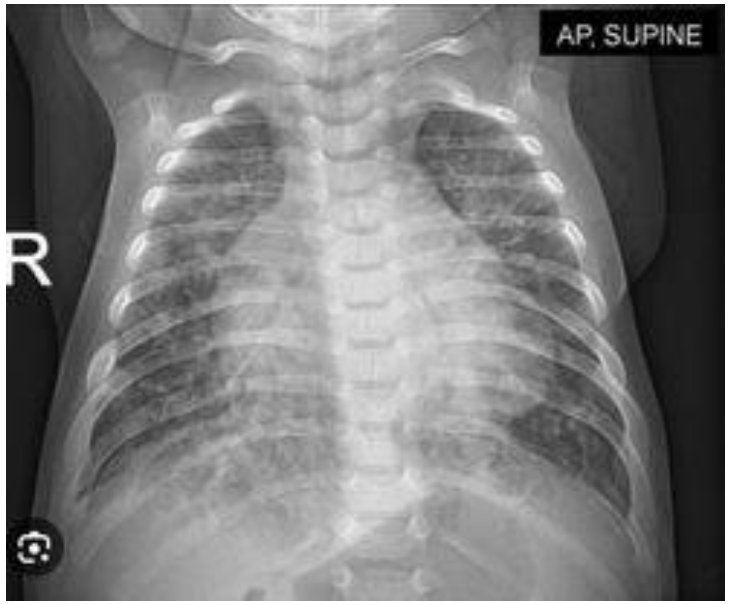
1- pneumonia
2-Multisystem inflammatory syndrome in children(MIS-C)
3-sever anemia
4-sepsis
5-viral myocarditis

LAB TESTS:

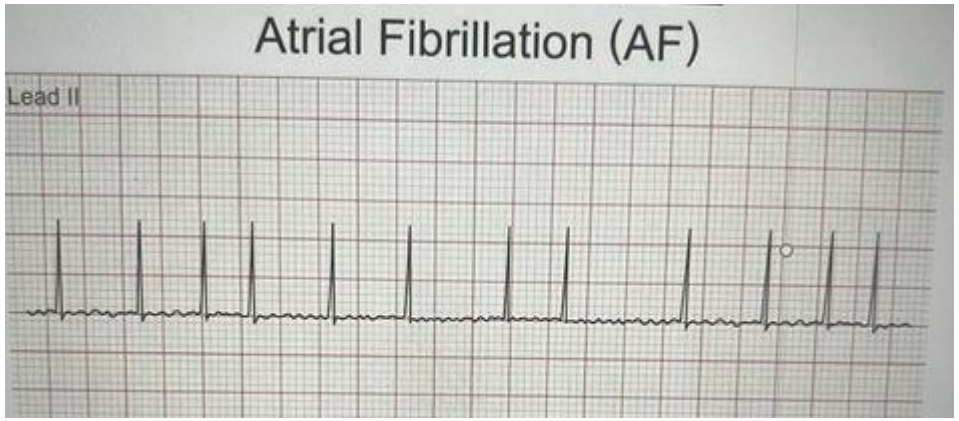
1- CBC →luekocytosis / RBC was Normal
2-blood cultures→are negetive
3-CRP→is elevated
4-brain natriuretic peptide (bnp) → was increased
5-troponin → was increased
6-SARS-CoV-2 Testing→negative IgG antibody

IMAGING:

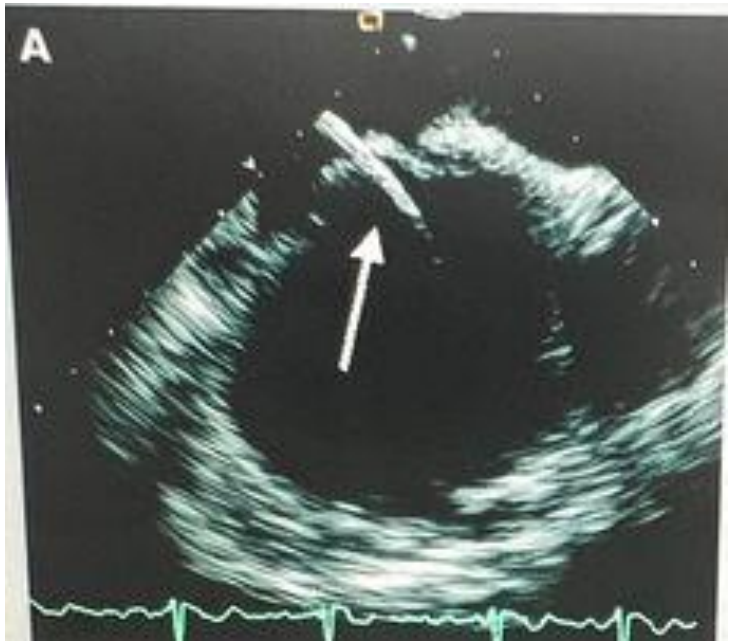
1-CXR

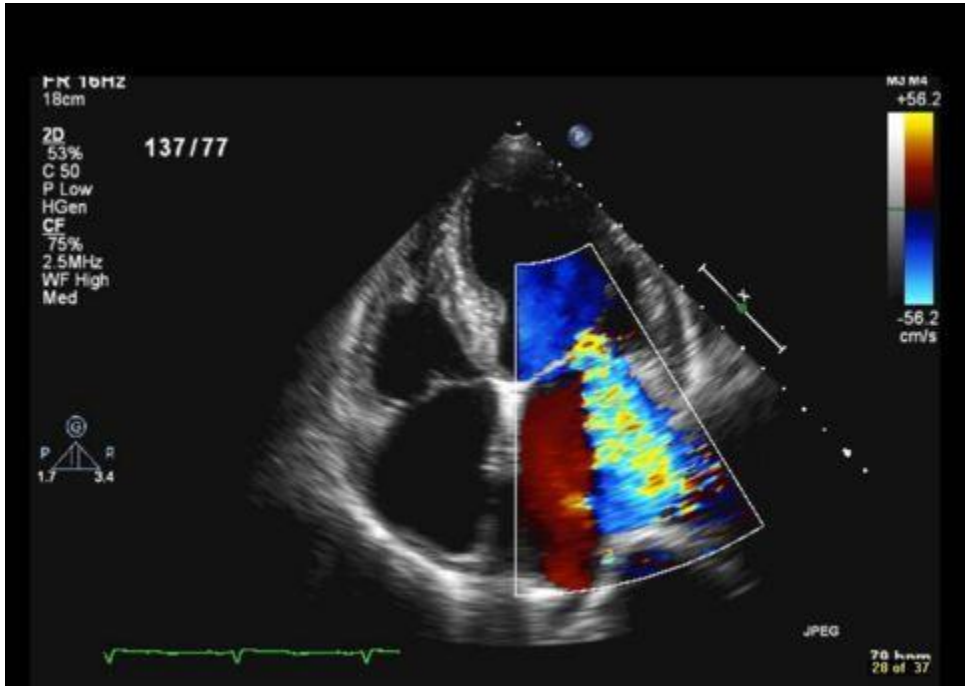


2-ECG(image)→



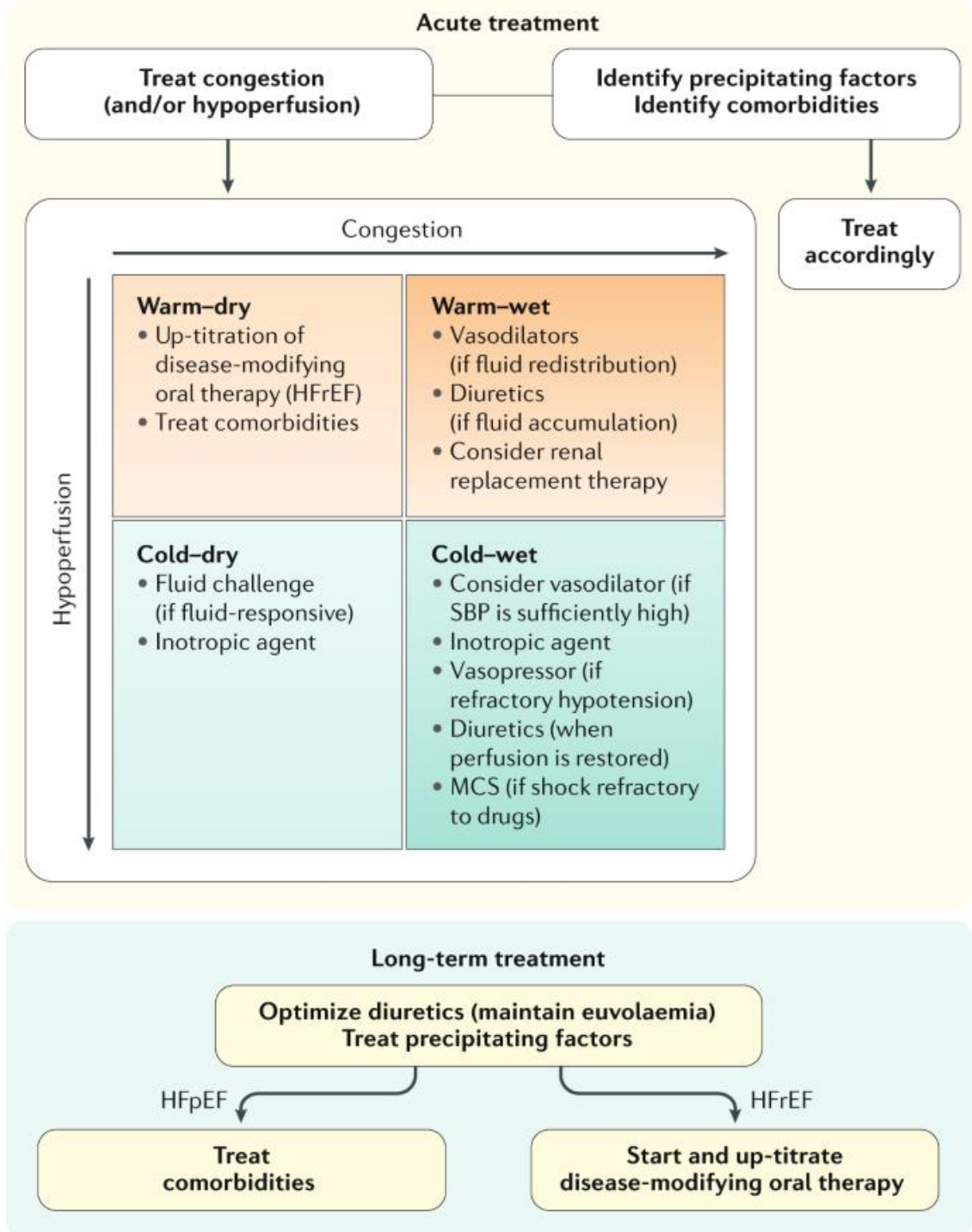
3-Echocardiogram→





AETIOLOGY:

-Coxsackievirus B
-Parvovirus B-19
-Adenovirus
-Influenza virus
-Human herpes virus 6 (HHV-6)


Acute treatment
Treat congestion (and/or hypoperfusion)
Identify precipitating factors
Identify comorbidities
Treat accordingly
Congestion
Hypoperfusion
Warm–dry
• Up-titration of disease-modifying oral therapy (HFrEF)
• Treat comorbidities
Warm–wet
• Vasodilators (if fluid redistribution)
• Diuretics (if fluid accumulation)
• Consider renal replacement therapy
Cold–dry
• Fluid challenge (if fluid-responsive)
• Inotropic agent
Cold–wet
• Consider vasodilator (if SBP is sufficiently high)
• Inotropic agent
• Vasopressor (if refractory hypotension)
• Diuretics (when perfusion is restored)
• MCS (if shock refractory to drugs)
Long-term treatment
Optimize diuretics (maintain euvolaemia)
Treat precipitating factors
HFpEF
HFrEF
Treat comorbidities
Start and up-titrate disease-modifying oral therapy

MANAGEMENT:

1-Oxygen Therapy →supplemental oxygen to help him breathe more comfortably and improve oxygenation.

2-Intravenous Medications→ Medications such as diuretics should be administered to reduce fluid buildup in the lungs and other tissues, relieving pulmonary edema and peripheral edema.

3-Inotropic Support →Inotropic medications should be used to improve Jacob's heart's contractility and pumping function.

4-Continuous Monitoring of Jacob's vital signs, oxygen saturation, and cardiac function should be closely monitored using continuous telemetry and other medical devices.

5-Cardiology Consultation: A pediatric cardiologist was involved in Jacob's care to provide specialized expertise in managing heart failure in children.

FOLLOW UP:

After discharge, The patient would require regular follow-up visits with his cardiologist to monitor his heart function and continue appropriate medications to prevent further complications. Cardiac rehabilitation might be recommended to improve his exercise tolerance and overall cardiac health.

case report 3

- History : A 2.3-month-old female patient, presented to the ER with SOB, constant crying, feeding problems, tachypnea,afebrile and sweating for 3-4 days.

- Her prenatal history was unremarkable. The patient was born(NVD) on term to a 22-year-old mother, her birth weight was 3kg. There was no parental consanguinity. Weight and height percentiles were normal for the patient’s age.

- Review of systems was unremarkable.

physical examination

- The patient’s general condition was fair, she appeared to be in distress.
- Vitals : body temperature of 36.8 °C, heart rate of 170 bpm, blood pressure of 80/50 mmHg, oxygen saturation of 96% in room air and respiratory rate of 62/min.

- CVS : displaced apex beat, systolic murmur at apex and audible S3 heart sound.

- RS : tachypnea with retractions

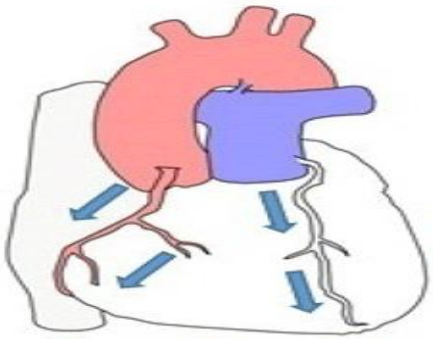
- GI : liver span was normal to her age

DDX

- ALCAPA (Anomalous origin of the left coronary artery arising from pulmonary artery).
- DCM (Dilated cardiomyopathy).
- Delayed presentation of moderate left to right shunt.
- Viral myocarditis.
- Arrhythmia.
- Mitral regurgitation.
- Coronary artery fistula.
- CHF

Investigations

- Laboratory tests results were as follows; hemoglobin: 11.6 g/dL, WBCs: 6200/mm3, thrombocytes: 318000/mm3, KFT and LFT was normal, electrolytes and acute phase reactants, elevated troponin-I at 27.3 ng/mL (normal range: 0.02-0.06 ng/ mL) and elevated creatine kinase-MB at 12.6 ng/mL (normal range: 0-5 ng/mL).
- CXR : cardiomegaly, LV enlargement and pulmonary venous congestion (cardiothoracic ratio of 0.65).
- ECG : 176 bmp sinus tachycardia, deep Q waves (Q wave width>30ms & depth>3mm) and inverted T waves in leads 1, aVL and V4-6 as well as left axis deviation.
- ECHO : LV dilation, impaired systolic function (13% shortening fraction), first degree mitral regurgitation, The right coronary artery/aortic annulus ratio was 0.2 and her RCA outlet was dilated and LCA arose from the PA.

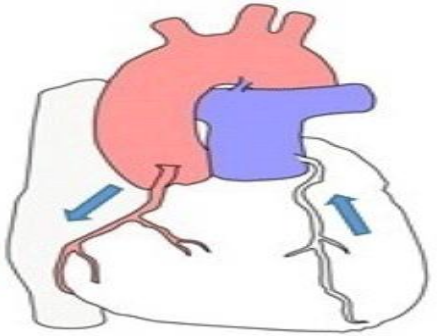


Neonatal period
MAP pressure = Ao
Antegrade flow in LMA

No symptoms
Pressure decrease in MAP
Ability to develop collaterals between MAP and Aorta

2 ALCAPA presentations

Infantile

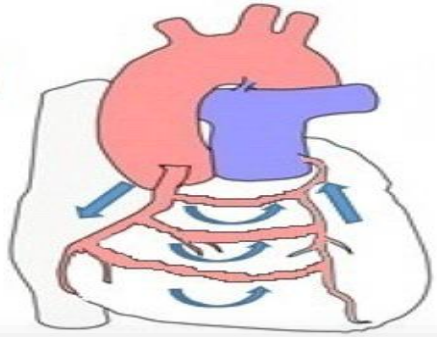


MAP pressure < Ao
Retrograde flow to LMA
No collaterals between RCA and LMA
RCA and LMA normal size

Ischemic cardiomyopathy secondary to infarction

90% death within the first year

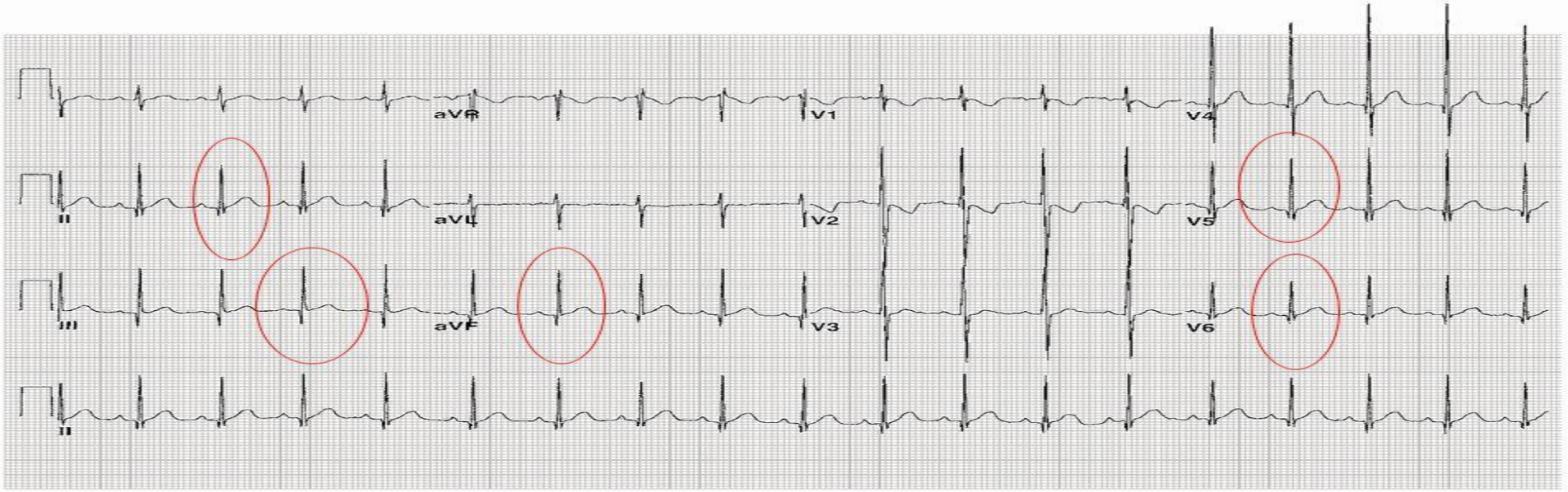
Adult



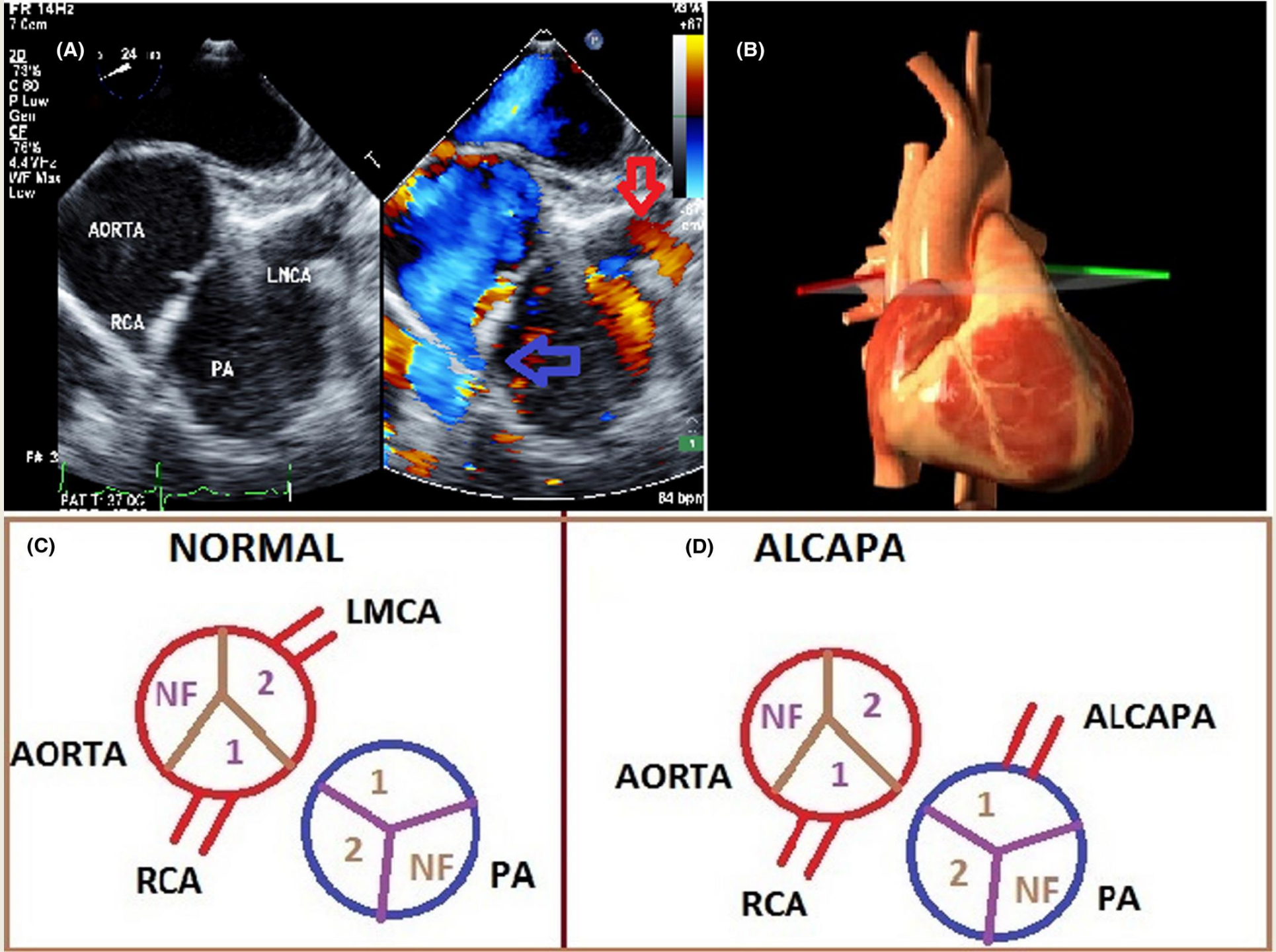
MAP pressure < Ao
Retrograde flow to LMA
Collaterals from the RCA to the LMA
Marked dilation of the LMA and RCA from the increased volume

Chronic myocardial ischemia
Dysrhytmia

Sudden death



Q waves in inferior/lat leads


(A)
AORTA
LMCA
RCA
PA
(B)
(C)
NORMAL
LMCA
NF
2
1
AORTA
RCA
1
2
NF
PA
(D)
ALCAPA
NF
2
1
AORTA
RCA
ALCAPA
1
2
NF
PA

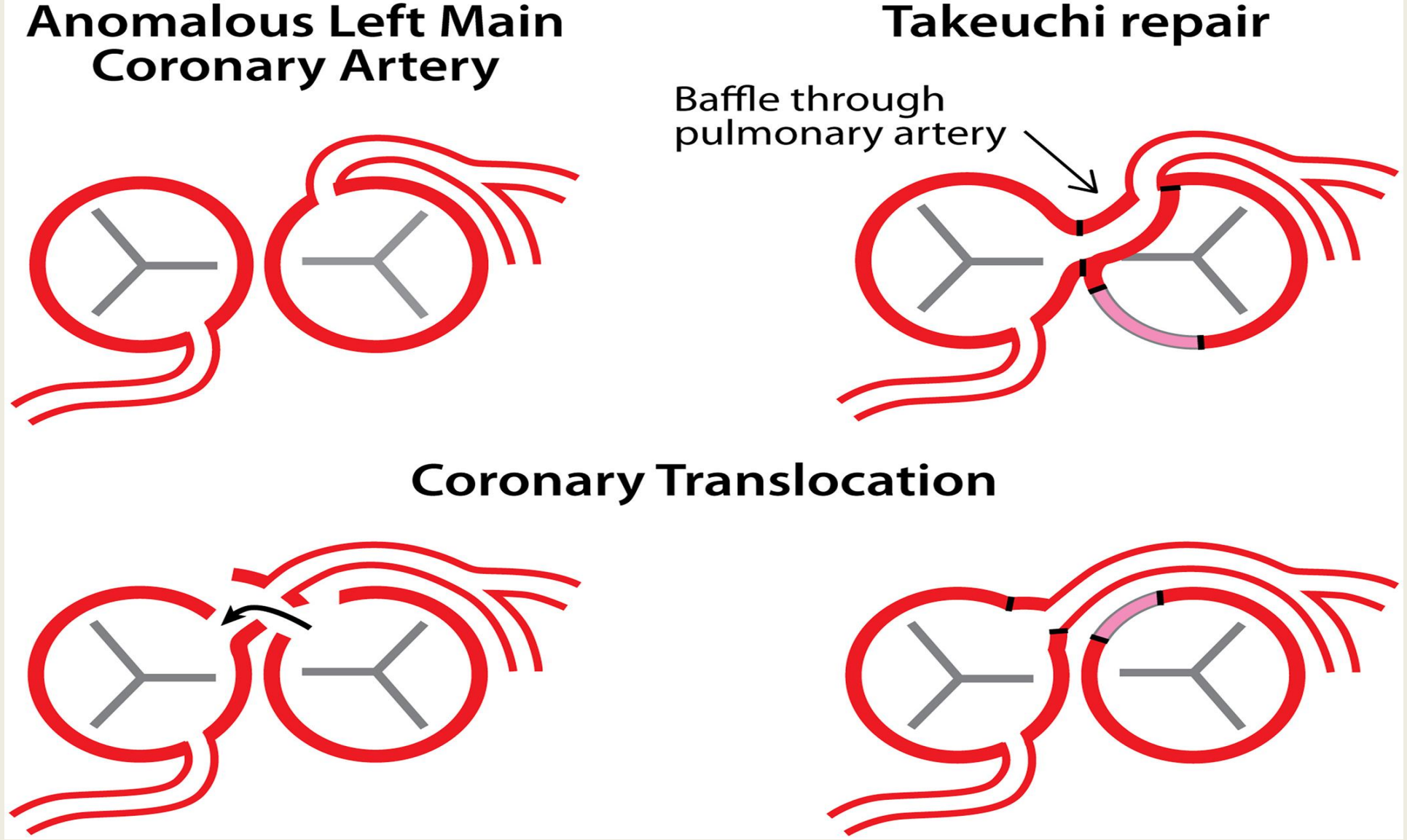
Management

- 100% oxygen
- Medical : usually post-op

  Diuretics(furosemide), and afterload reduction may be necessary until there is significant improvement in left ventricular systolic and diastolic function with resolution of mitral valve insufficiency.(nitroprusside vasodilator, captopril ACEI) These medications improve cardiac output and eliminate the preoperative symptoms of congestive heart failure.

- Surgical :

1. Direct reimplantation.
2. Takeuchi procedure (creation of an aortopulmonary window and an intrapulmonary tunnel extending from the anomalous ostium to the window).
3. A saphenous vein bypass graft.
4. A left subclavian artery-coronary artery anastomosis.



Done by : Jawad Samer Al-amro

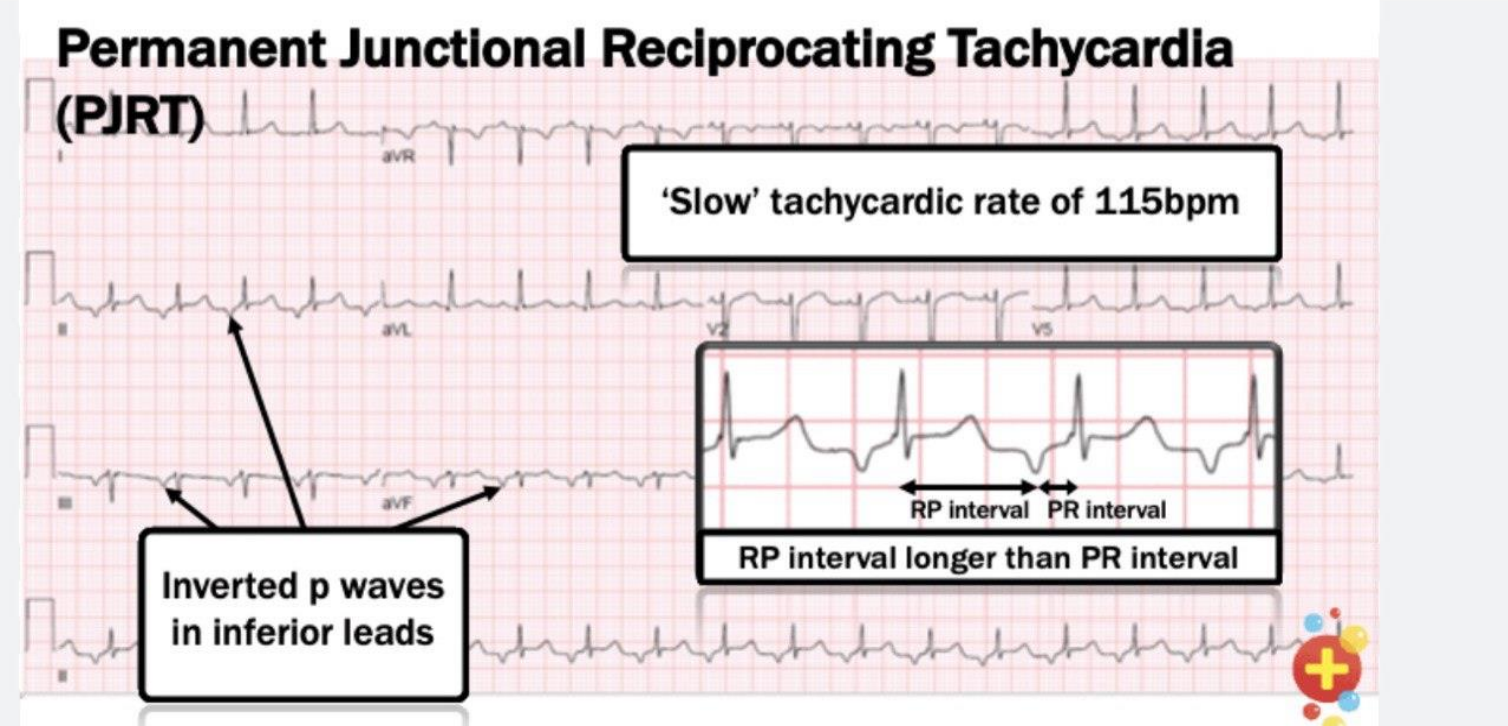
Case 4

Case scenario

- Saleh khaled, a 12-year-old male, presented to the pediatric clinic with complaints of intermittent episodes of rapid heartbeats and occasional dizziness over the past few months. During these episodes, he feels his heart racing and sometimes experiences lightheadedness. The episodes last for a few minutes and resolve spontaneously. There is no associated chest pain, shortness of breath, or fainting.
- His past medical history is unremarkable, with no known heart-related conditions or chronic illnesses. There is no family history of cardiac arrhythmias or congenital heart diseases. Saleh lives in a smoke-free home, is a 7th-grade student, and denies any substance use or exposure to harmful environmental factors.
- Upon physical examination, Saleh's vital signs, including blood pressure, heart rate, and respiratory rate, are within normal limits for his age. Auscultation of his heart reveals a regular heart rhythm, with no murmurs or abnormal sounds detected during the current visit.
- The parents' concern and the symptoms described warrant further evaluation by a pediatrician to determine the underlying cause of Saleh's rapid heartbeats and dizziness. Additional diagnostic tests and investigations may be necessary to reach a definitive diagnosis and develop an appropriate treatment plan.

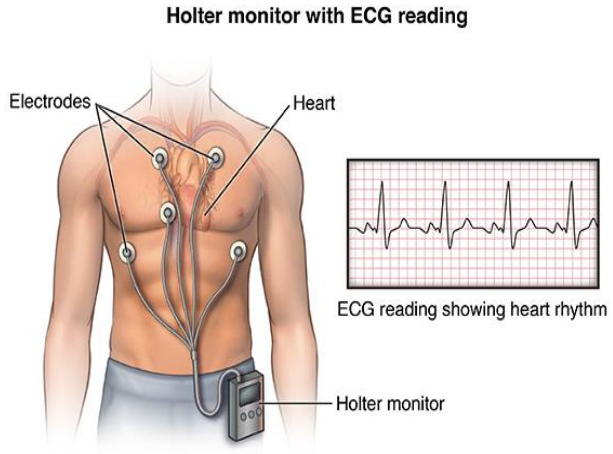
Diagnostic Workup

1.Electrocardiogram (ECG): An ECG is performed, which reveals the characteristic findings of permanent junctional reciprocating tachycardia (PJRT) in a pediatric patient. The ECG shows a narrow-complex tachycardia at a rate of approximately 115 beats per minute, with retrograde P-waves suggesting involvement of the atrioventricular (AV) node in the reentrant circuit.



- 2. Holter Monitor: A 24-hour Holter monitor is recommended to capture and analyze Saleh's heart rhythm over an extended period, given the intermittent nature of his symptoms.

What is a Holter monitor? The Holter monitor is a type of portable electrocardiogram (ECG). It records the electrical activity of the heart continuously over 24 hours or longer while you are away from the doctor's office.



3. Echocardiogram: An echocardiogram is performed to assess the heart's structure and function. It aims to rule out any structural heart abnormalities that could be contributing to the arrhythmia.

Differential Diagnosis

- The diagnosis of permanent junctional reciprocating tachycardia (PJRT) is based on the characteristic findings observed on the ECG, which show a ***narrow-complex tachycardia*** with retrograde P-waves, ***indicating a reentrant circuit involving the AV node and an accessory pathway***. However, to ensure an accurate diagnosis and rule out other potential causes of tachycardia in pediatric patients, the following differential diagnoses are considered:

  1. Atrioventricular Nodal Reentrant Tachycardia (AVNRT):
  AVNRT is another form of supraventricular tachycardia (SVT) caused by a reentrant circuit within the AV node itself, ***without involvement of an accessory pathway***. Differentiating AVNRT from PJRT can be challenging based solely on the ECG findings, as both conditions may show similar narrow-complex tachycardias with retrograde P-waves.

  2. Atrioventricular Reentrant Tachycardia (AVRT):
  AVRT occurs when there is an extra electrical pathway, known as a bypass tract (e.g., Wolff-Parkinson-White syndrome), connecting the atria and ventricles. This accessory pathway allows for rapid conduction and reentry of electrical impulses, leading to tachycardia. In AVRT, the ECG may show a ***wide-complex tachycardia*** due to the presence of the accessory pathway.

- 3. Inappropriate Sinus Tachycardia (IST):
  IST is a condition where the sinus node, the heart's natural pacemaker, generates an abnormally fast heart rate in response to various triggers such as exercise, stress, or fever. IST typically presents with a ***regular, sinus-originating rhythm on the ECG.***

  4. Sinus Tachycardia:
  Sinus tachycardia is a normal response to physiological factors such as exercise, fever, pain, anxiety, or dehydration. It is characterized by a ***regular rhythm with a heart rate higher than the normal range for age.***

Treatment

- The treatment and management plan for Saleh's PJRT will involve the following steps:
- Acute Episode Management: During episodes of PJRT, the immediate focus will be on terminating the tachycardia and stabilizing Saleh 's condition. Vagal maneuvers (e.g., carotid sinus massage, ice to the face) and, if necessary, intravenous administration of adenosine or other antiarrhythmic medications, may be employed to restore normal sinus rhythm.
- Long-term Medication: Saleh will be prescribed long-term antiarrhythmic medication to prevent future episodes of PJRT. Commonly used medications include beta-blockers (e.g., propranolol) or calcium channel blockers (e.g., verapamil). The medication will be carefully titrated to achieve optimal control of heart rate while minimizing side effects.
- Ablation Therapy: If medical therapy proves inadequate or intolerable, catheter ablation may be considered. During this procedure, a specialized catheter is used to destroy the abnormal electrical pathways responsible for the tachycardia, restoring normal heart rhythm.
- Regular Follow-up: Saleh will require regular follow-up appointments with her pediatric cardiologist to monitor her response to treatment, assess for medication side effects, and ensure there are no disease progression or complications.

Conclusion

The diagnosis of Permanent Junctional Reciprocating Tachycardia (PJRT) was made based on the characteristic ECG findings. Further investigations, including the Holter monitor and echocardiogram, were recommended to confirm the diagnosis and rule out other potential causes of pediatric tachycardia. The treatment plan is tailored to Saleh's individual needs to manage his condition effectively and ensure his well-being.

Case Scenario 5

History

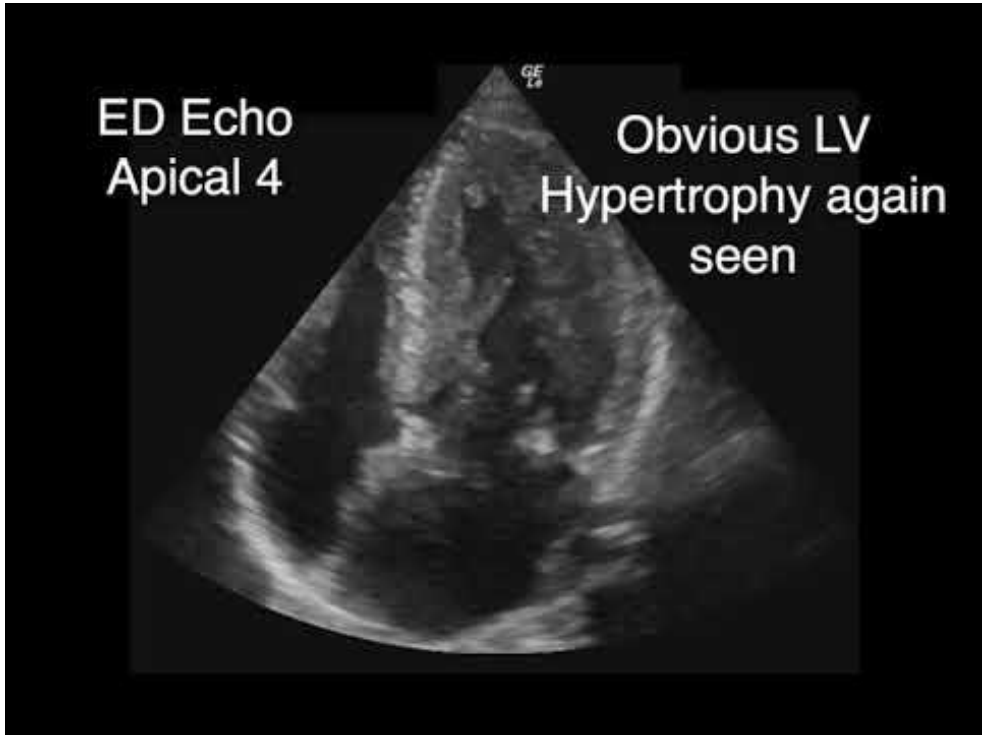
*Name: Sarah. *Age: 8 years old. *Gender: Female. *Chief Complaint: Chest pain during physical activities, fatigue, shortness of breath and syncope. *History of Present Illness: Sarah's parents have noticed that she gets tired easily during playtime and complains of chest pain after running or playing for a short period. Her symptoms seem to worsen over the past few months until she passed out few days ago when she was running after her younger brother. *Past Medical History: Sarah was healthy with no significant medical history. She had the usual childhood illnesses, such as occasional colds and ear infections, but no chronic conditions. *Family History: There is a family history of heart disease, with her maternal grandfather having had a heart attack in his early 50s.

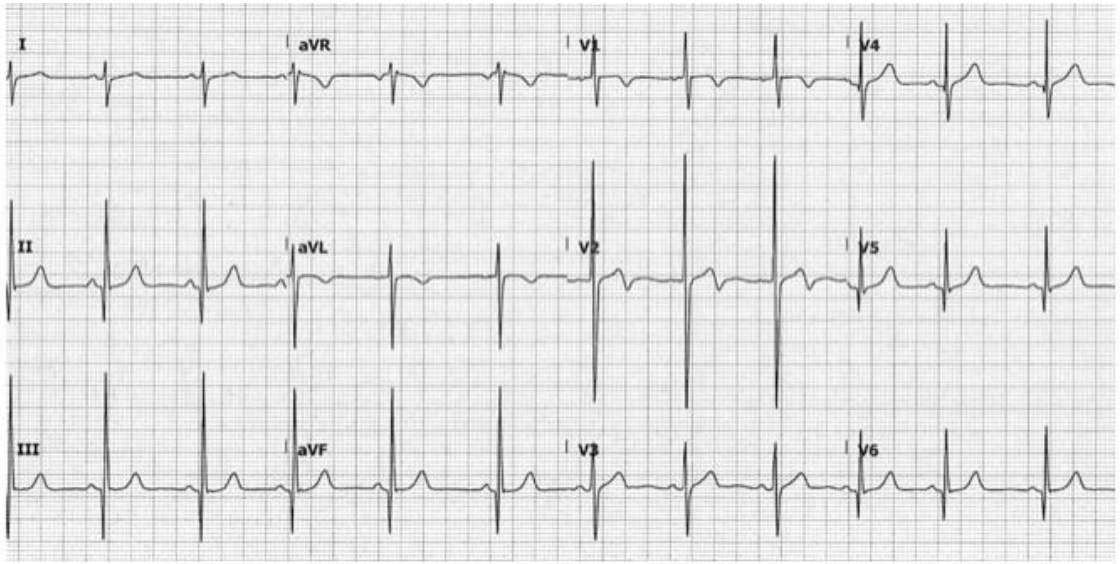
Physical Exam

- Upon examination, it was noticed that :*Blood pressure: Within normal limits for her age.*Heart rate: Normal for her age.*Respiratory rate: Normal at rest, but she demonstrates increased respiratory effort after exertion.*Signs of heart failure: Hepatomegaly, distended neck veins, and bilateral pedal edema are absent at this stage.*Growth: Sarah's growth parameters are within normal limits for her age.
- CVS exam: on palpation: double apical impulse. *on auscultation: normal S1, split S2, clear S3 sound and loud and harsh crescendo-decrescendo holo-systolic murmur is heard during auscultation at the left sternal border.
- RS exam: was insignificant except of basal crackles.
- Differential Diagnosis: *Asthma: Shortness of breath and fatigue could be mistaken for asthma, especially if no heart murmur is appreciated on initial examination. *Anemia: Fatigue and tiredness might be related to anemia, which is a common condition in children.

Investigations and Imaging

- CBC: to rule out anemia and it was all normal.
- Chest X-ray: shows increased cardio-thoracic ratio and pulmonary edema.
- ECG: It shows narrow deep “dagger-like” Q waves in the inferior and lateral leads.
- Echocardiogram: it shows left ventricular hypertrophy, reduced end diastolic volume, reduced stroke volume and normal ejection fraction.





Management

- Management:*Beta-blockers: Sarah may be prescribed beta-blockers to reduce the heart's contractility and alleviate symptoms. *Calcium channel blockers: These drugs can also be used to manage symptoms by relaxing the heart muscle and reducing obstruction. *Lifestyle modifications: Sarah will need to avoid strenuous physical activities until her symptoms are better controlled. Gradual exercise programs may be recommended as tolerated.

Etiology and Type of Heart Failure

- Etiology: HOCM is a genetic condition caused by mutations in genes that encode for proteins in the cardiac sarcomere, the contractile machinery of the heart muscle.
- Type of Heart Failure: HOCM falls under the category of diastolic heart failure, where the heart muscle becomes stiff and has difficulty relaxing during the filling phase.

Follow-up

- Sarah will require regular follow-up appointments with her pediatrician and a pediatric cardiologist. The frequency of visits will depend on the severity of her symptoms and her response to treatment. Follow-up care will include monitoring her growth, symptoms, and response to medication. Echocardiograms may be repeated periodically to assess changes in the heart's structure and function. Additionally, Sarah's parents should be educated about recognizing any worsening symptoms or potential complications of HOCM, such as arrhythmias. They should be encouraged to seek immediate medical attention if Sarah experiences chest pain, fainting, or palpitations.

- **Thank you**