Heart failure

Definition: HF is a chronic ,progressive condition in which the heart muscle is unable to pump enough blood to meet the body's needs for blood and oxygen .

Causes of HF :

1-**ventricular dysfunction** (structurally normal heart vs congenital heart disease)

2-**preserved ventricular contractility** (volume overload vs pressure overload)

Structurally normal heart e.g.: cardiomyopathy ,myocarditis

CHD e.g.: complex CHD with concurrent ventricular dysfunction

Volum overload e.g: VSD,PDA

Pressure overload e.g.; aortic stenosis , aortic coarctation

Note

in the 1st week of life ,HF usually results from left heart obstruction,(duct dependent)
e.g. sever COA ,hypoplastic left heart syndrome

Clinical presentation:

- In infants includes:

Tachypnea/ diaphoresis during feeds/ easy fatigability/ Irritability

decreased volume of feeds/ poor weight gain

- Older children may present with :

gastrointestinal symptoms (abdominal pain, nausea, vomiting, and poor appetite)

exercise intolerance/ Cough and wheezing/ dyspnea/ edema

Palpitations/ syncope

- **Physical signs:**

Tachycardia, a gallop rhythm, and signs of poor perfusion (dec. capillary refill Cool extremities)

♥ If **left-sided** failure is predominant, tachypnea,dyspnea, orthopnea,wheezing,Pulmonary edema .

♥ If **right-sided** failure is present, hepatomegaly, edema, distended neck veins

- **Investigations**

1- Chest X-ray: - Cardiomegaly

2- Echo:

- Confirm left ventricle dysfunction (decreased ejection fraction & increased ejection time).
- Confirm chamber enlargement.
- May detect cause of failure.

3- ECG: -Detect arrythmias.

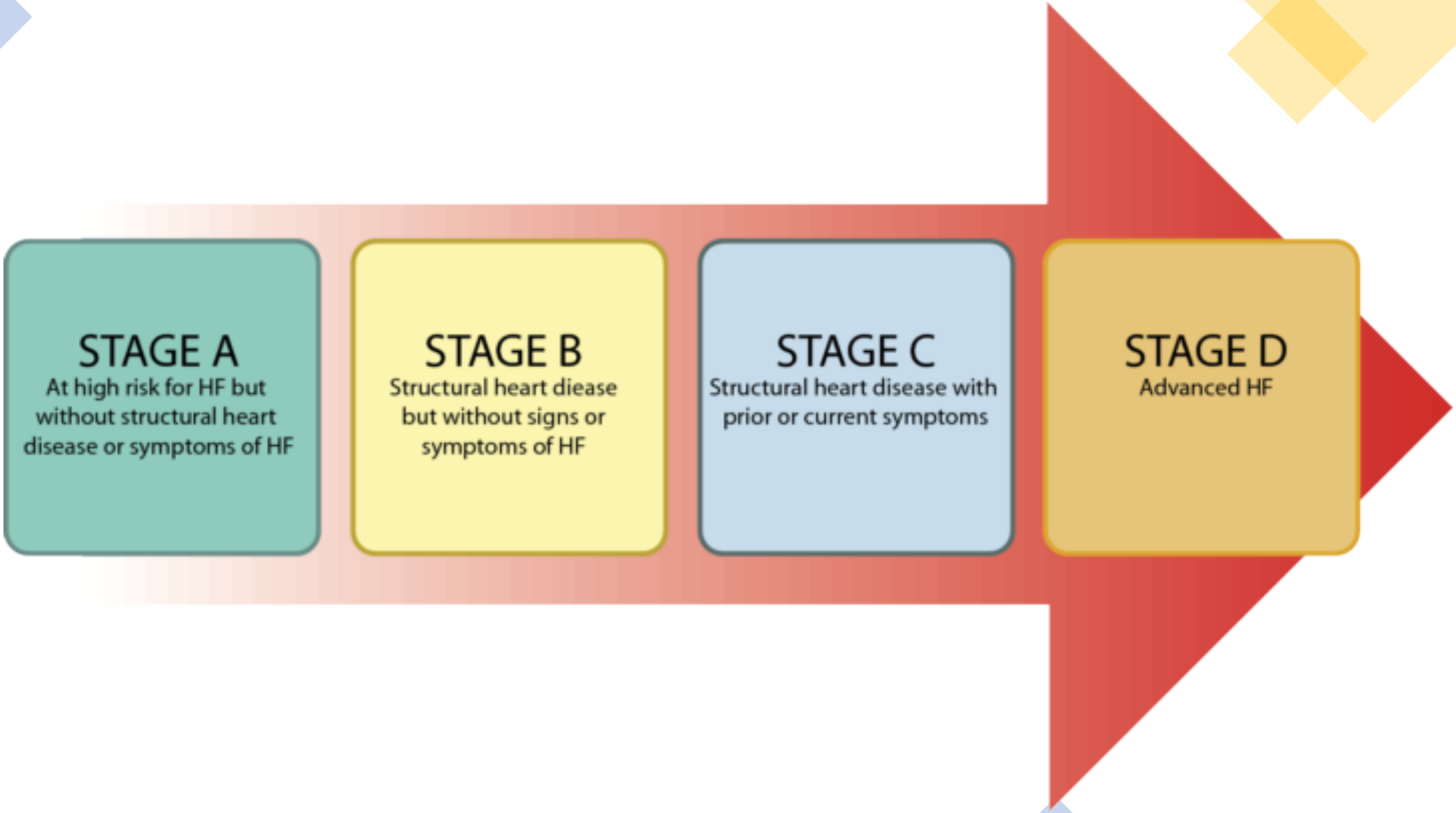
Categorization of the stage and severity of the patient's HF is important for monitoring the disease progression and guiding management decisions:

- **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.
- **Severity** – The two main classification systems used for describing severity of pediatric HF are the New York Heart Association (NYHA) and Ross classifications:

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 on 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 exertion
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.


STAGE A
At high risk for HF but without structural heart disease or symptoms of HF
STAGE B
Structural heart diease but without signs or symptoms of HF
STAGE C
Structural heart disease with prior or current symptoms
STAGE D
Advanced HF

STAGE A	STAGE B	STAGE C	STAGE D
High risk for HF but without structural heart disease or symptoms of HF	Structural heart disease without s&s of HF	Structural heart disease with symptoms	Advanced HF
Exposure to cardiotoxic agents FHX of cardiomyopathy	Aortic insufficiency with LV enlargement	Symptomatic cardiomyopathy	Symptoms at rest despite maximal medical therapy
None	ACE Inhibitors (SE: cough ,angioedema) (2nd line ARBs)	ACEi ,aldosterone antagonist ,duritics After few weeks of stability , a BB is added in pt. With persistent LV dysfunction	iv diuretics ,+- inotropic agents Mechanical circulatory support heart transplantation

Management

- General

Rest/ oxygen/fluid-salt restriction

- Diuretics — Diuretics decrease preload and are used to treat children with stage C or D HF:

1.Loop diuretics – Furosemide is the most commonly used loop diuretic.

Side effects of loop diuretics include electrolyte abnormalities (hyponatremia, hypochloremia, and hypokalemia), metabolic alkalosis, and renal insufficiency. Long-term therapy can lead to nephrocalcinosis and ototoxicity (usually with high intravenous doses). Increased risk of bone fractures has also been reported.

2.Thiazide diuretics –They generally are used as second-line agents and often in combination with a loop diuretic.

- Renin-angiotensin-aldosterone system inhibition

❖ ACE inhibitors: (ACE) inhibitors are an accepted first-line component of therapy for children with stage B and C HF. Blood pressure and renal function should be closely monitored, especially in neonates

❖ Angiotensin receptor blockers:

ARBs are usually reserved for patients unable to tolerate ACE inhibitors due to cough or angioedema.

- Mineralocorticoid receptor antagonists(eg, spironolactone, eplerenone)

Side effects include hyperkalemia (with both drugs) and gynecomastia (with spironolactone).

- Beta-blockers (eg, carvedilol or metoprolol)

is usually added to an established regimen of diuretics and an ACE inhibitor.

Side effects: dizziness, fatigue, hypotension, bradycardia, bronchospasm, and hypoglycemia.

- Digoxin

Digoxin is not recommended for children with asymptomatic ventricular dysfunction; however, it is commonly used in the treatment of infants and children with stage C HF, particularly those with persistent symptoms despite treatment with other agents (eg, diuretics and ACE inhibitors.

Potential adverse effects (arrhythmias) are rare with this low level.

Case presentation 1

Khalid a 2 months-old male in Amman.

He was brought to the ER yesterday because he has feeding problems for two weeks in which he feeds, stops and then feed again.

The mother has noticed that he gets tired, sweaty and tachypneic when he's feeding.

The patient failed to gain weight since birth - his birth weight is 2.700 kg and now his weight is 3 kg.

There's no cyanosis or pallor. There' no cough, wheeze or stridor.

Birth History

Pre-natal: booked case, antenatal visit done, ultrasound was done, multivitamins was taken. There is no H/O diabetes mellitus, Hypertension, swelling of feet, fits during pregnancy. no other drugs taken, no X-ray done.

Natal: term pregnancy, svd at home by dai.

Post-natal: cried immediately after birth. no cyanosis, jaundice or fits.

Normal birth weight according to mother.

Nutrition and immunization:

- Exclusive breast feeding
- Adequately immunized according to the Jordanian program

Past medical, surgical and drug history:

-no previous admissions.
-No surgeries
- He doesn't take any drugs

Family history:

Family history of congenital heart diseases.

Physical examination:

General examination General Impression An ill looking, irritable male child with no dysmorphic features of normal build and height lying on bed uncomfortably with mild intercostal recessions, and having IV cannula on his left hand.

Vitals:

HR: 170 BPM (Normal HR 80-140)

RR: 45 (Normal RR: 20-30)

BP: 60/30 in all 4 limbs (Normal systolic blood pressure is 70-100)

Growth parameters:

Weight: 3 kg (below 5th percentile)

Hight: 56 cm (47th percentile)

Head circumference: 39 cm (50th percentile)

There was no signs of dehydration

RS:

Signs of respiratory distress of subcostal retractions and tachypnea -

There was bilateral crepitations but no wheeze or stridor. -

No cyanosis -

CVS- inspection and palpation

Trachea is in the midline -

No scars/ dilated veins -

Apical impulse is felt in the left 5th ICS -

Capillary refill: 1S -

Femoral pulses were palpable -

No edema -

CVS- auscultations:

Grade 3 pansystolic murmur with fixed split s2 in the lower left sternal border

GI:

- There was hepatomegaly and the liver span was felt 3 fingers below the costal margin.
- There’s no ascites

Nothing significant in the rest of the physical examination

Deferential diagnosis?

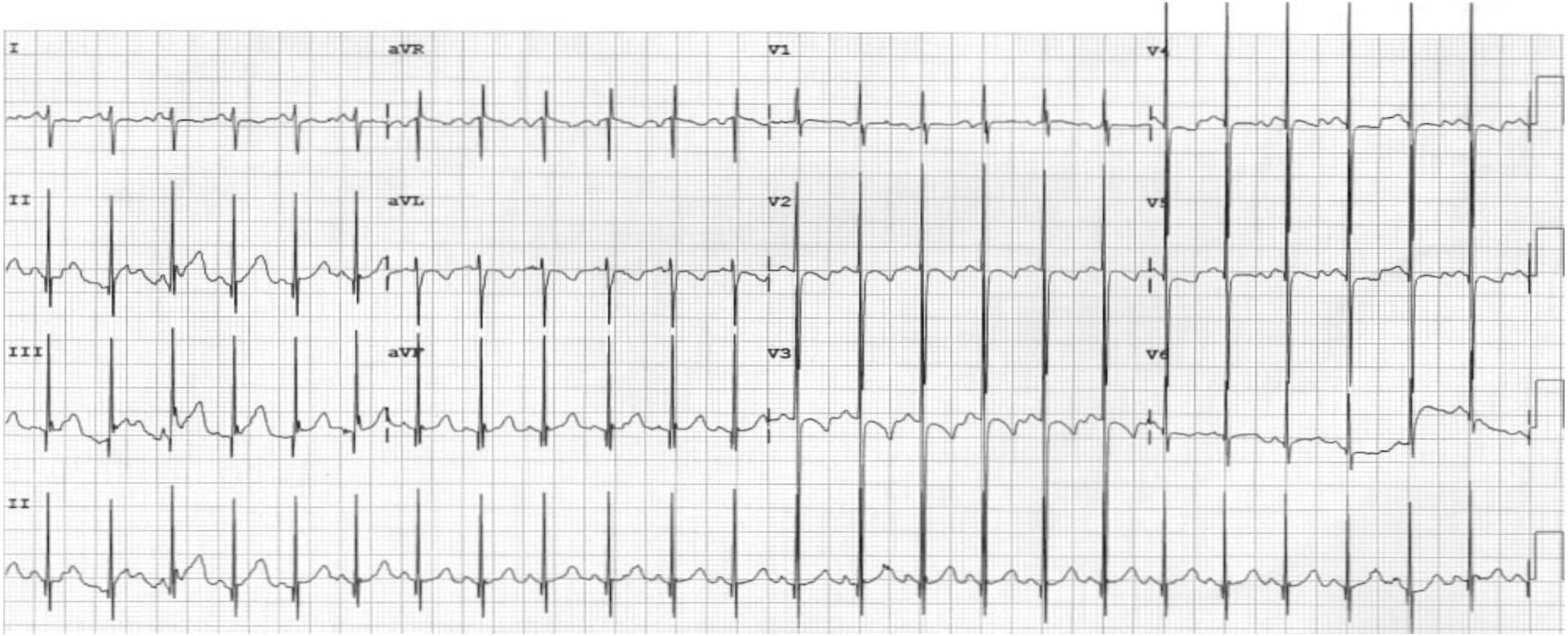
Deferential diagnosis

Left to right shunt

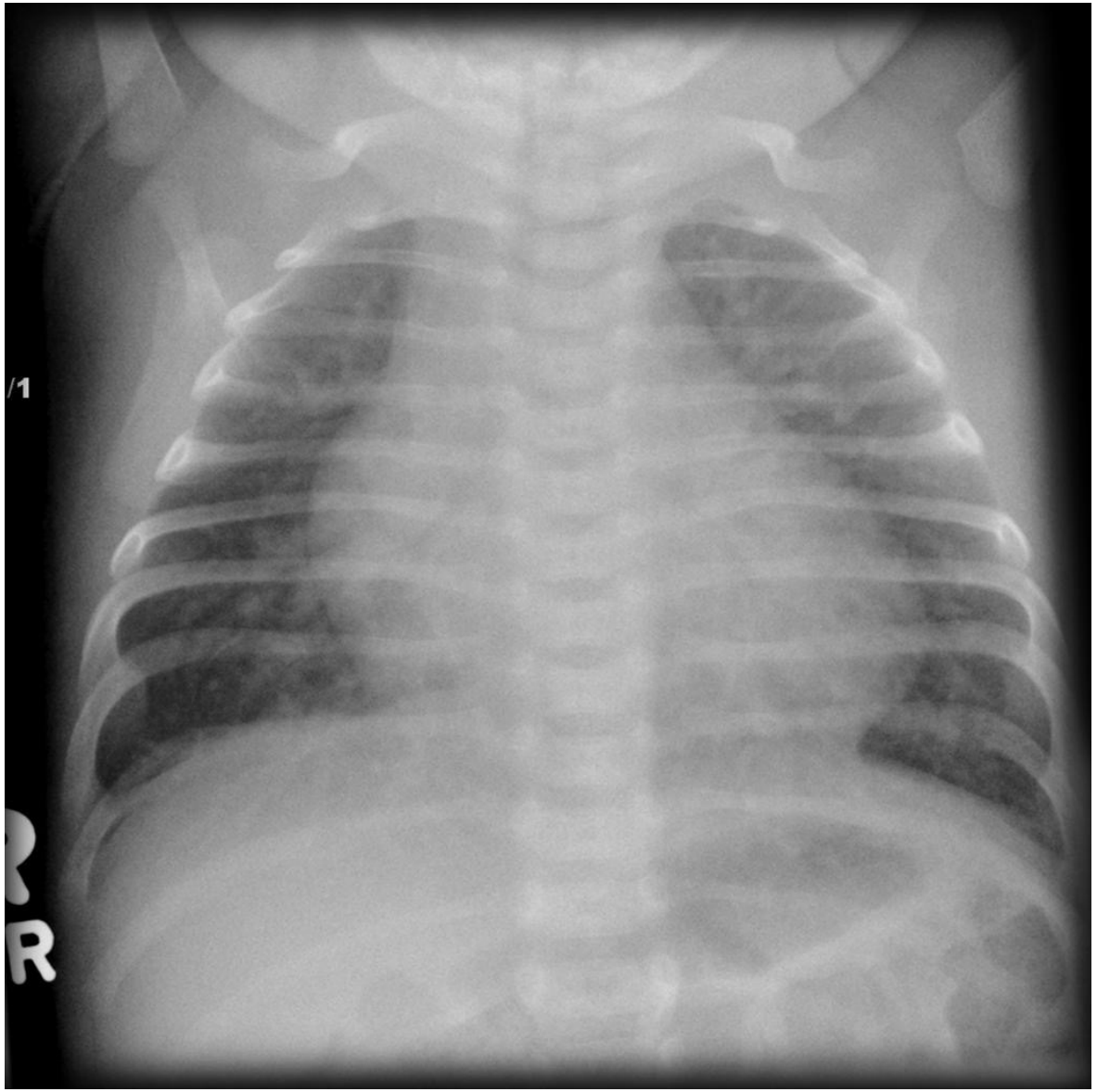
1. **VSD**
2. Large PDA
3. Aortopulmonary window
4. AV canal

Investigations:
ECG .1

Biventricular hypertrophy



2. ECHO
3. X-ray:



Ventricular septal defect

Most common congenial heart disease.

is a developmental defect in the interventricular septum allowing shunt between the left & right ventricles.

Types:

Peri-membranous (MC) .1

Muscular .2

Supracrustal .3

Inlet .4

Complications:

Heart failure .1

Pulmonary HTN .2

Endocarditis .3

Clinical Features:

In small defect:

Usually asymptomatic

Normal growth and development.

Incidental detection of a pansystolic murmur at left 3rd and 4th intercostal spaces.

Treatment:

1. Diuretics to decrease the preload
2. ACE inhibitors to decrease the peripheral resistance
3. Digoxin to improve the cardiac contractability

Case presentation 2

A 12-day-old girl was referred to our hospital because of tachypnea and retractive breathing.

The patient was born via elective cesarean section at 36 weeks and 0 days of gestation, weighing 2,500 g, with Apgar scores of 6/9.

On day 6 after birth, Tachypnea, retractive breathing, and tachycardia were observed.

*History :

-Gradual increase of SOB in the last few days

-Sweating with crying or eating

-cynosis

-No fever

Birth History

Pre-natal: booked case, antenatal visit done, ultrasound was done, multivitamins was taken. There is no H/O diabetes mellitus, Hypertension, swelling of feet, fits during pregnancy. no other drugs taken, no X-ray done.

Natal: preterm pregnancy.

Post-natal: cried immediately after birth. cyanosis, no jaundice or fits.

*Physical examination :

-Vital signs :

BP 68/30 mmHg (wide pulse pressure)

HR 170 beat\min (tachycardia)

RR 68breath\min (tachypnea)

Temperature 37.3c

O2 saturation 92%

*Cardiac examination:

-Tachycardia

-Continuous murmur machine like murmur was heard at left infraclavicular border

-Palpable femoral pulses

*Respiratory examination:

-Tachypnea

-Mild subcostal and intercostal retraction

-Mild crepitation bilaterally

*Abdominal examination:

- Liver palpable 3 cm below costal margin

*Extremities examination:

- No edema

DDX:

- Large significant PDA
- Non-restrictive large VSD
- Av canal with large inlet VSD
- Large aorto-pulmonary window

*Investigation:

-**Blood tests** revealed(mild metabolic acidosis, mild hyponatremia, increased urea)

ECG :

Twelve-lead electrocardiography showed that the heart rate 160 beats\min and regular , with right-axis deviation and left atrial overload with the downward deflection of the P wave in lead V1.

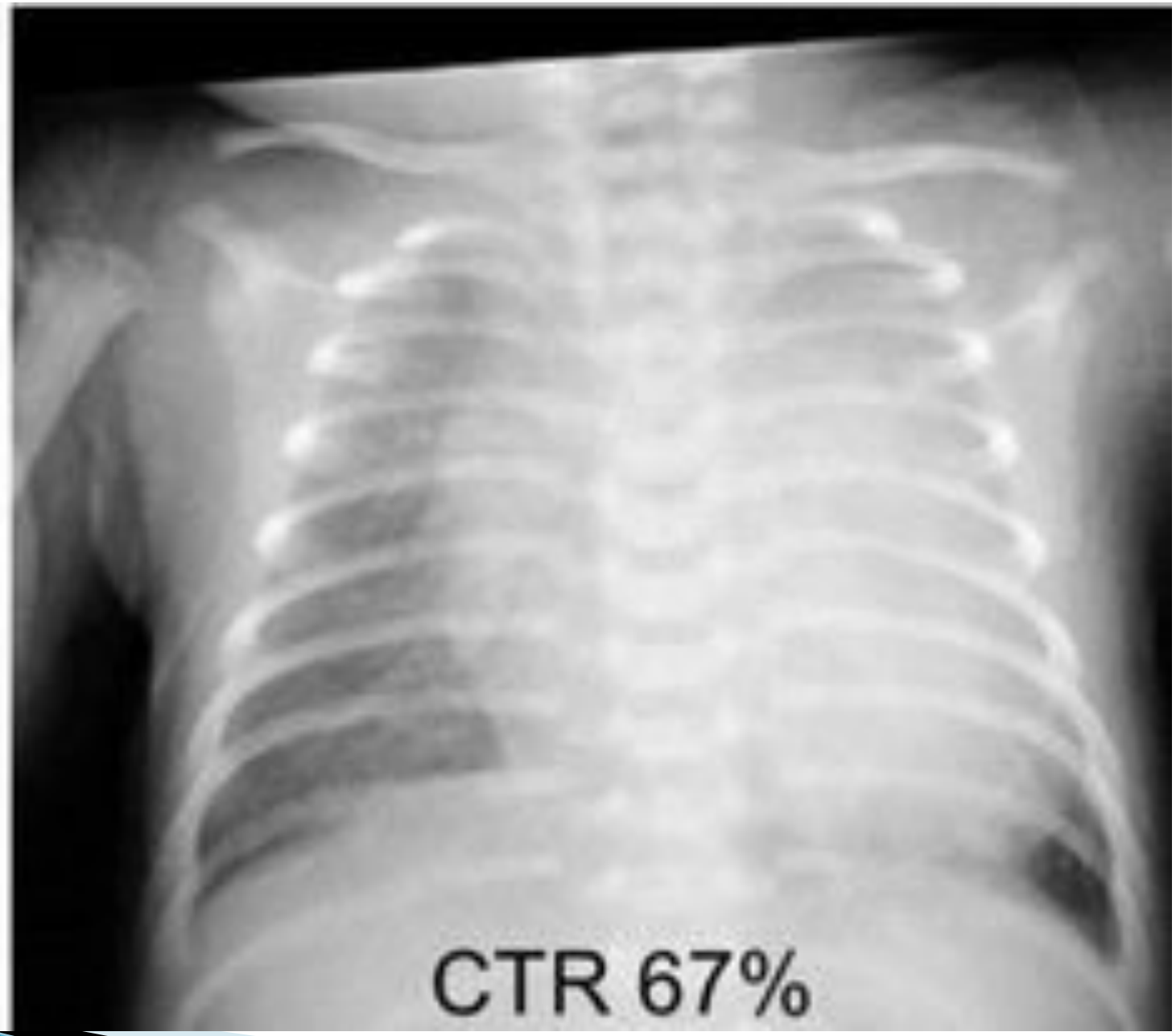
-**Imaging study** :

Chest x-ray showed that the cardiothoracic ratio 67% indicating cardiomegaly and lung permeability was reduced bilaterally.

Echo:

Large PDA

Left to right shunt of blood flow


CTR 67%

Patent ductus arteriosus (**PDA**) is a medical condition in which the ductus arteriosus fails to close after birth: this allows a portion of oxygenated blood from the left heart to flow back to the lungs by flowing from the aorta, which has a higher pressure, to the pulmonary artery. Symptoms are uncommon at birth and shortly thereafter, but later in the first year of life there is often the onset of an increased work of breathing and failure to gain weight at a normal rate. With time, an uncorrected PDA usually leads to pulmonary hypertension followed by right-sided heart failure.

Cause:

Large significant PDA with large left to right shunt

Management :

1. Ant failure medication (Lasix , digoxin)
2. Medical trial for PDA closure by:
   - Indomethacin (now not given)
   - Or Ibuprofen
   - Or Paracetamol
3. If no effects were obtained , we would perform surgery(PDA clipping).

Case #3

History

- A 5-month-old female presents to the ER with shortness of breath, sweating, interrupted feeding, grunting on feeding, failure to thrive and decreased urine output for the last month
- The infant lost 0.5 kg over the last month.

Vital signs

- HR: 160 bpm
- RR: 55 bpm
- BP: 90/50 mmHg
- Temp: 36.7 c
- O2 sat: 96% on room air

Physical Examination

- Hypoactive & Sleepy
- Diaphoresis
- Tachypnea w/ retractions and grunting
- Tachycardia
- Displaced apex
- Audible S3
- Mild crepitus
- Apical systolic murmur (Mitral regurgitation)
- Enlarged liver

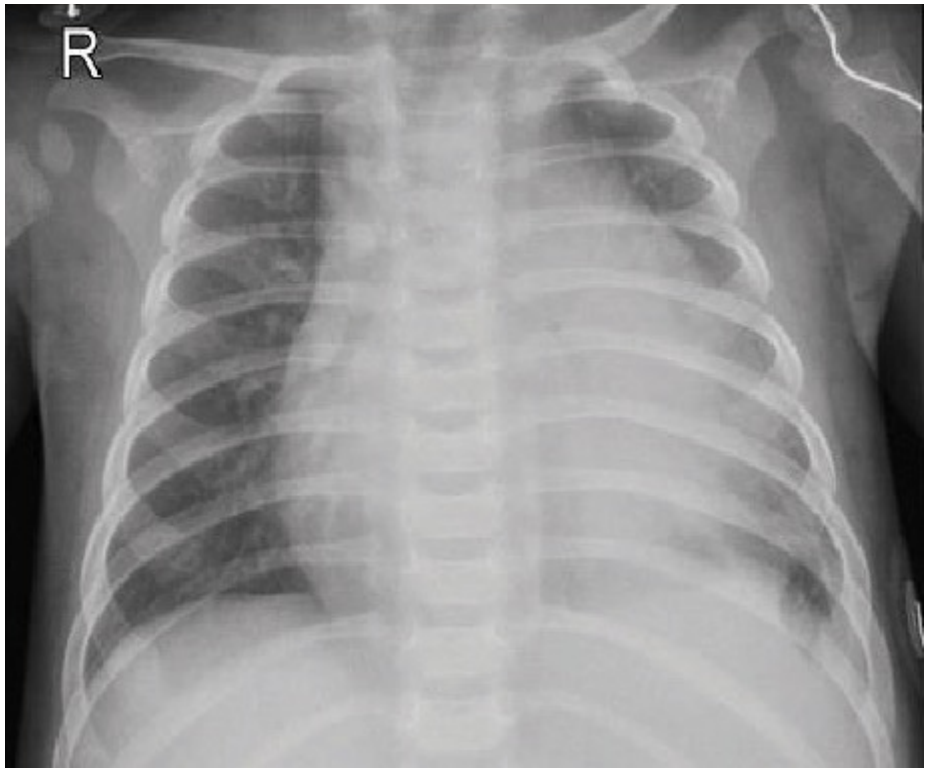
Labs

Troponin: Elevated

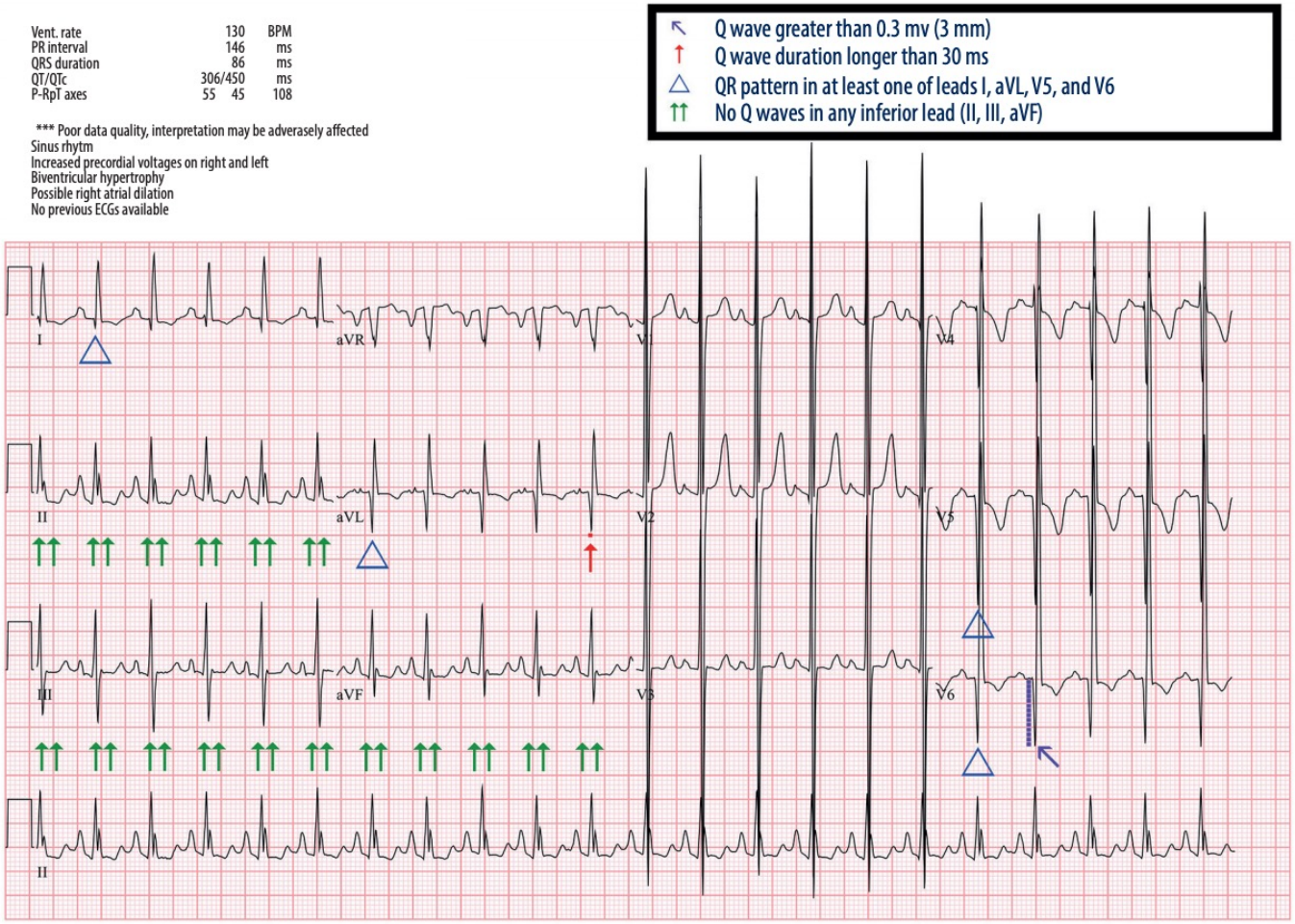
CKMB: Elevated

ABG : Metabolic Acidosis

Chest x-ray

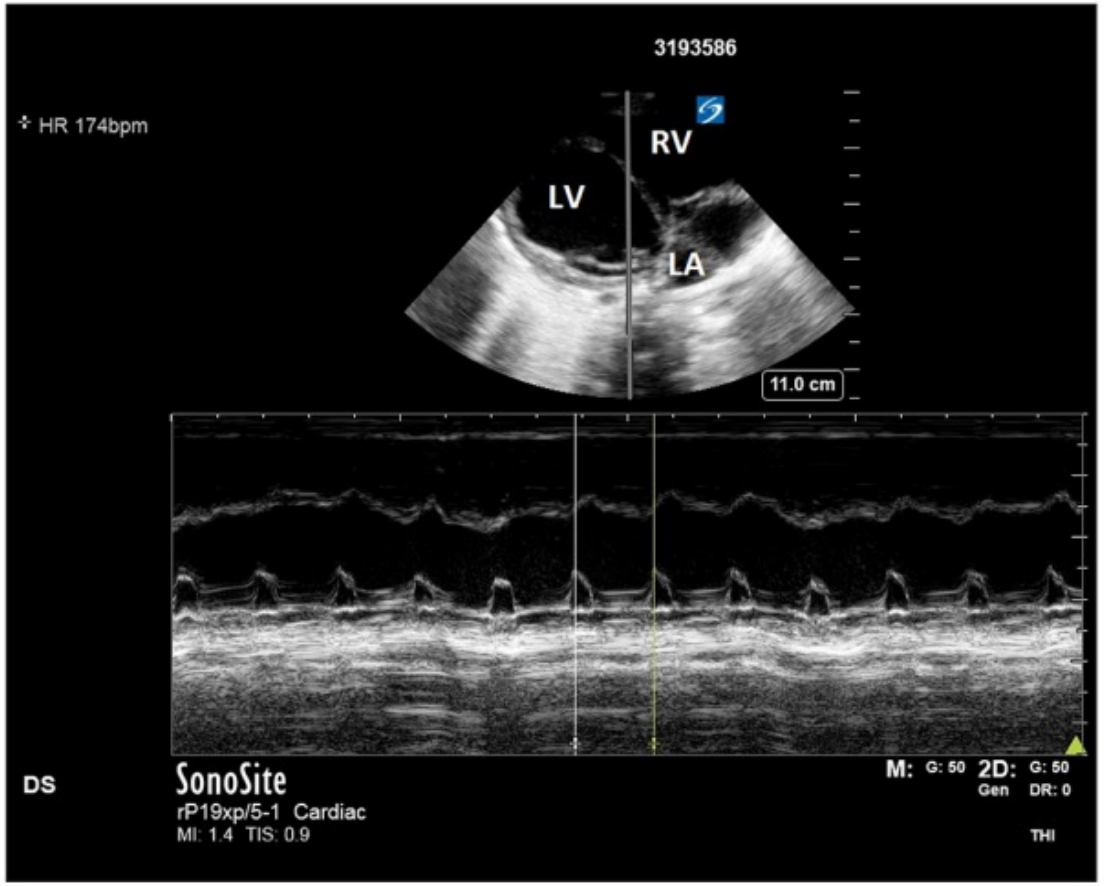


ECG



Biatrial enlargement, Biventricular hypertrophy Deep Q waves in I, AVL, V5–V6

U/S

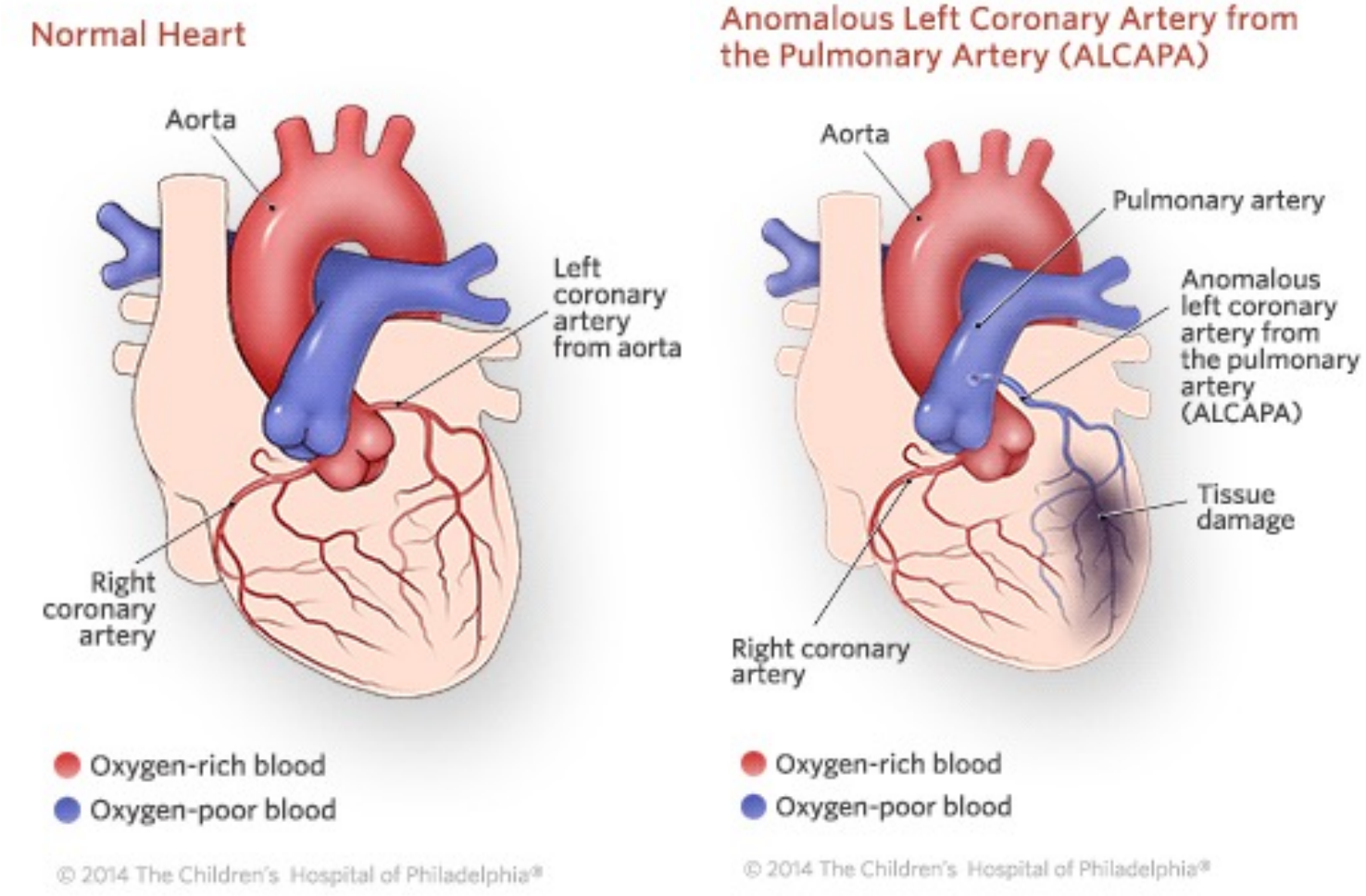


Left ventricle (LV) is dilated.

Echocardiography: short axis aortic root view

- Large origin of RCA evident from usual coronary cusp
- LCA origin from aortic root not evident
- Linear tracking of LAD/CX coursing to a main LCA arising from posterior aspect of MPA
- Extensive network of collateral flow from RCA connections distally

Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA)



Medical Management:

- Correct ABG + Supporative
- Lasix + Captopril but if severe Digoxin

•Surgery is required to fix ALCAPA., including:

•Detaching the left coronary artery from the pulmonary artery and suturing (stitching) it into the correct position on the aorta.
•Creating a tunnel from the aorta to the anomalous left coronary artery, and then closing the connection between the left coronary artery and the pulmonary artery.
•Removing the faulty left coronary artery, then using a vein from the leg to create a new left coronary artery.
•Creating a connection between the left subclavian artery (a large artery that carries blood to the left arm and upper body) and the left coronary artery. This allows some of the very oxygen-rich blood from the subclavian artery to feed the left coronary artery and the heart.

Case #4

History

- A 4 years old girl presented into the ER with SOB, the mother complains that her child is fatigued easily and has been suffering of tachypnea, cough, excess sweating and fatigue for the past 2 days, irritability. However, there were no history of recent infection and no fever.

Physical Examination

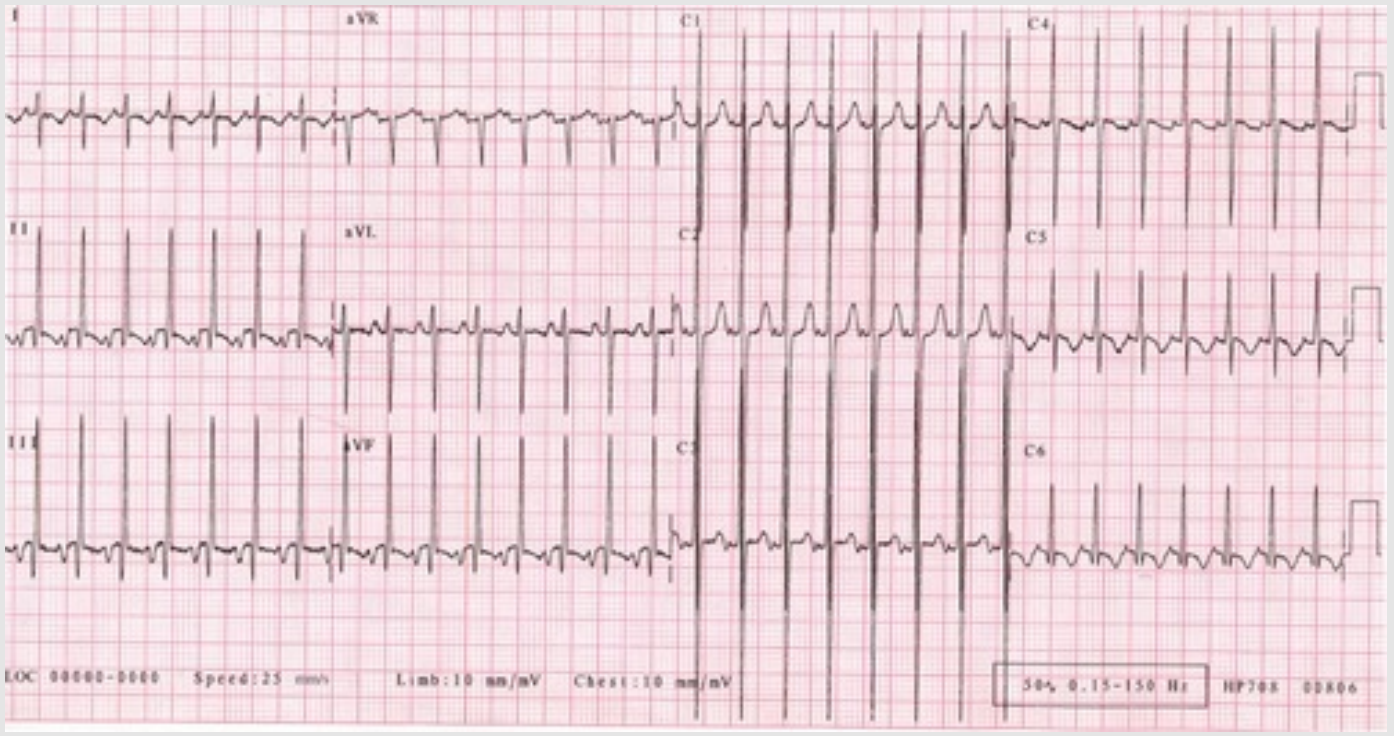
The patient vital signs were taken

- Normal blood pressure 100/60 mmHg
- HR 180 bpm
- RR: 50 bpm
- Temp: 37 c
- CNS was normal
- hepatosplenomegaly

Auscultation:

- Apical grade II pansystolic murmur
- Audible S3
- Basal crackles

ECG



- ECG showed an incessant narrow complex tachycardia with inverted P waves in leads II, III and aVF, as well as the left lateral leads, and an RP interval longer than the PR interval

Biochemical and hematological parameters were within the normal limits.

Viral markers were negative.

Laboratory tests for inborn errors of metabolism were all negative.

Cardiac magnetic resonance imaging was performed to detect possible myocarditis and it was normal.

Holter ECG confirmed long R-P tachycardia with a rate of 140-160 beats/minute.

Echocardiogram showed:

- an enlarged and spherical left ventricle with diminished systolic functions
- ejection fraction: 27%

Holter ECG confirmed long R-P tachycardia with a rate of 140-160 beats/minute.

Permanent Junctional Reciprocating Tachycardia (PJRT)

Management

Acute termination of PJRT can be achieved by a wide array of antiarrhythmic agents, including adenosine, calcium channel blockers (antegrade block), and class I antiarrhythmic drugs as flecainide, propafenone, and ajmaline (retrograde block)

However, the choice of how to terminate PJRT should be based on the patient's clinical characteristics including underlying cardiac or non-cardiac diseases, haemodynamic status, and co-medication.

Although drugs with a longer half-life, such as calcium channel blockers, flecainide, and propafenone, have the advantage of preventing immediate recurrence of PJRT, they should not be used in patients with depressed left ventricular ejection fraction due to the risk of further haemodynamic deterioration because of their negative inotropic effects.

In infancy and early childhood, medical therapy is considered as a first option, and various successful therapies including flecainide, propranolol, amiodarone, and sotalol

Medical therapy in infancy and early childhood may allow delay of the highly effective radiofrequency ablation treatment until the children have reached adequate growth

In patients with atrioventricular reentrant tachycardia, oral flecainide or propafenone, preferably in combination with a beta blocker, may be considered if structural or ischaemic heart disease is absent, and oral beta blockers, diltiazem, or verapamil may be considered if no pre-excitation sign on a resting ECG are present

Case 1

- A 13 year old athletic female patient presents to the hospital after suddenly collapsing during exercise . The patient was rushed to the hospital via an ambulance in which the patient was evaluated by the paramedics and was found to be pulseless and an automated external defibrillator advised shock, she was defibrillated thrice. Return of spontaneous circulation was achieved after electrical cardioversion. On arrival to the hospital patient was stabilized, history and physical exam were obtained. Patient had previous episodes of syncope especially on exertion she also complained of shortness of breath. Family history was positive for undiagnosed cardiomyopathy, no sudden deaths in the family and no QT prolongation or any other electrical problems.

- Normal lung auscultation and normal liver.

Physical examination

- Palpatioln : apical heave was noted.
- Auscultation : grade 3 left sternal systolic ejection murmur and an S4 sound

DDX

➢ Hypertrophic cardiomyopathy

➢ Electrical Abnormalities

➢ Restrictive cardiomyopathy

Distinguishing factors:

- Kussmaul sign on physical exam
- ECG with low voltages

➢ Dilated cardiomyopathy

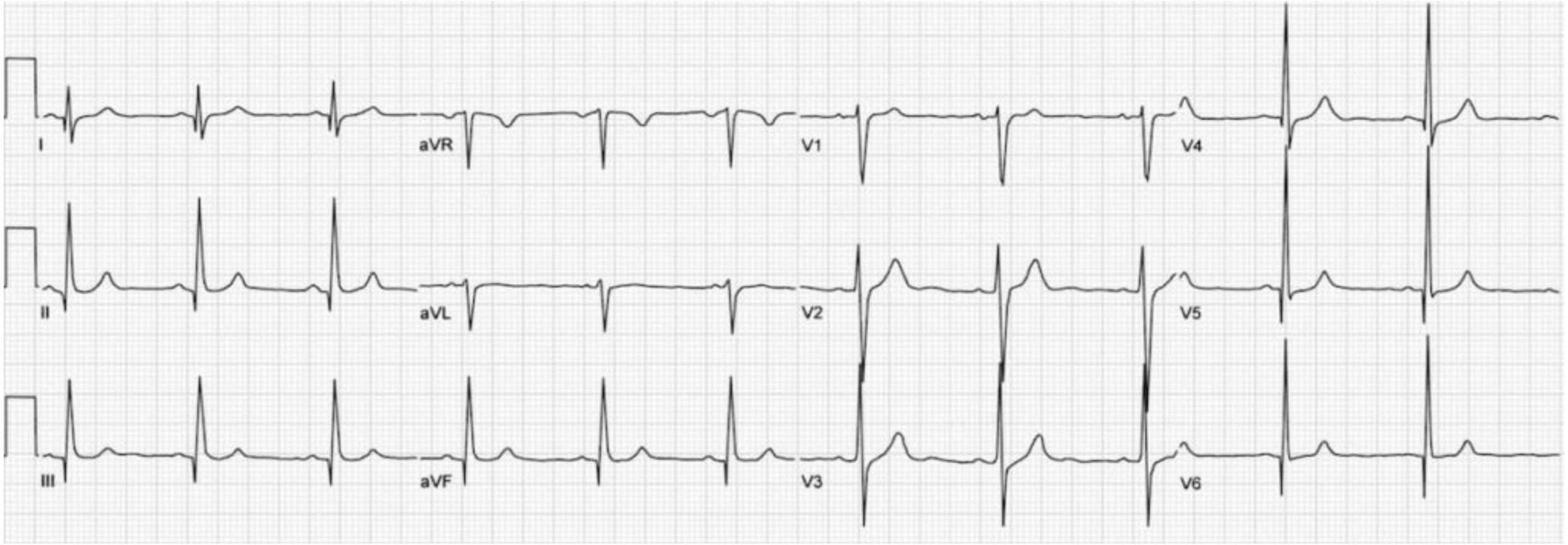
Distinguishing factors:

ECG with reduced ejection fraction

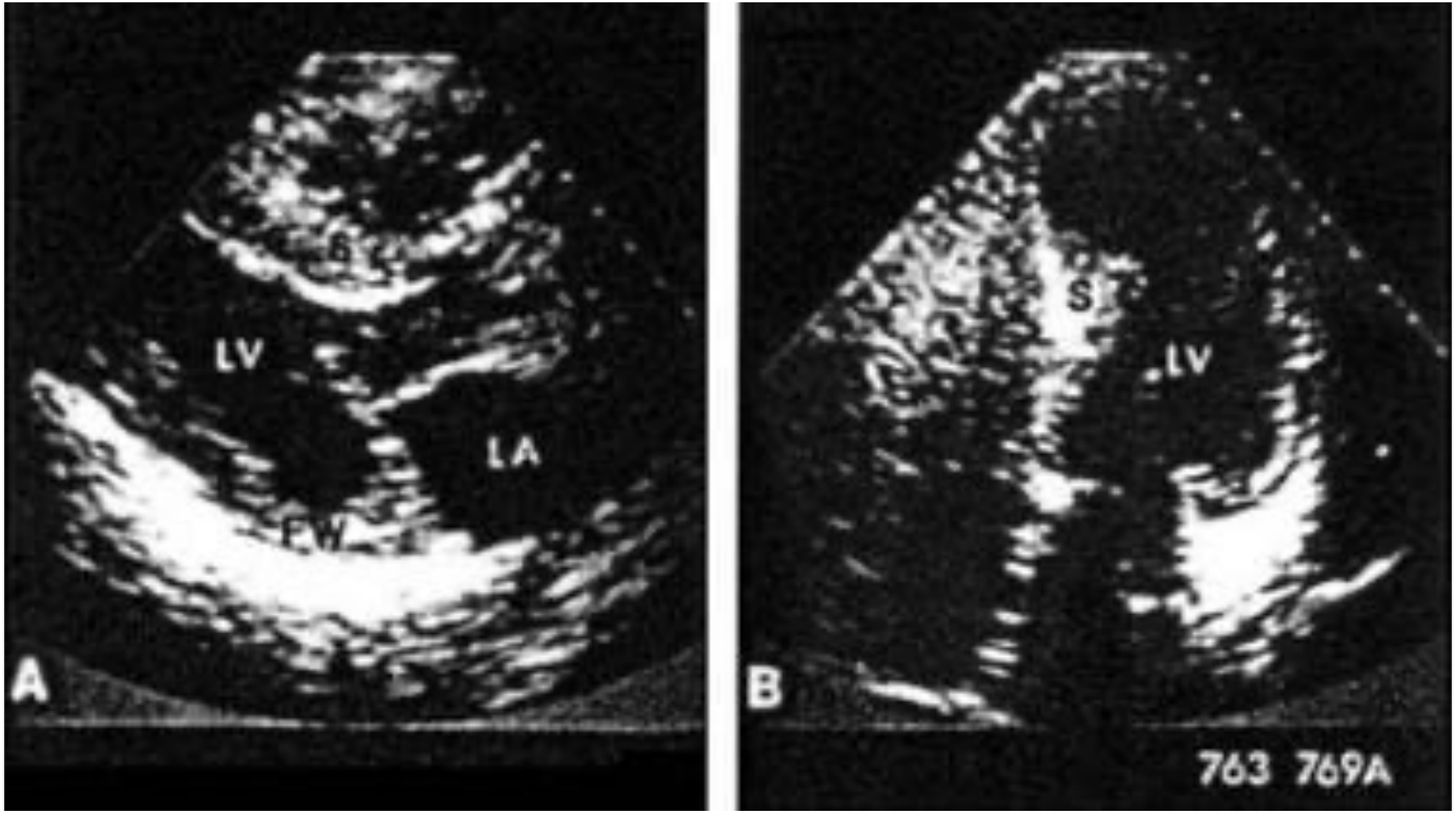
Progressive heart failure

Work up

- Holter monitor for 24 hours or more
- Chest Xray: Cardiomegaly
- ECG : Left ventricular hypertrophy
- Echocardiogram: Normal systolic function
- Ejection Fraction 72%
- Fractional shortening 35%
- LVH with left ventricular outflow obstruction (gradient – max 20 mmhg)
- Diastolic Dysfunction Grade 2



Increased QRS voltage

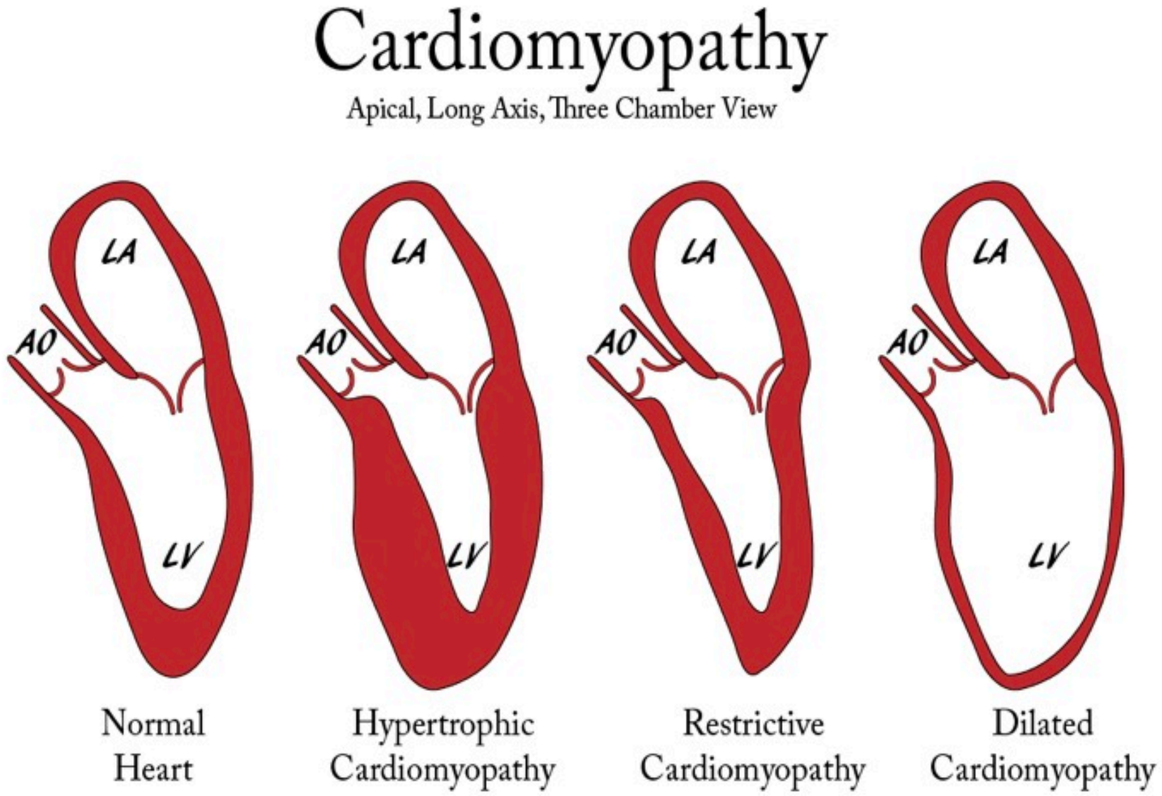

LV
LA
PW
A
S
LV
B
763 769A

Treatment

- Avoid overexertion especially athletic activities.
- Managed with Beta blockers and/or calcium channel blockers (if BB contraindicated)
- Implantable cardioverter defibrillator (ICD) used for prevention of sudden arrhythmic death.

Hypertrophic cardiomyopathy

- Results from genetic defects in sarcomeric proteins of cardiac myocyte.
- Diastolic failure, preserved systolic
- Ventricular hypertrophy
- 2nd most common cardiomyopathy after dilated cardiomyopathy
- LV wall increase thickness and appearance may indicate chronicity
- Increased thickness leads to outflow obstruction



Fpnotebook.com

Functional Pattern	Dilated	Hypertrophic	Restrictive
LV EF	<40%	50-80%	45-90%
Mechanism	Impaired Contractility	Impaired Compliance	Impaired Compliance
Dysfunction	Systolic	Diastolic	Diastolic
Cause	Volume Overload	Pressure Overload	Amyloidosis, Sarcoidosis, post-radiation fibrosis, and eosinophilic syndromes of the heart
Associated Heart Sound	S_3	S_4	S_4

CASE 2

A 2-year-old boy is brought to the emergency department due to fever and respiratory distress.

A week ago the patient had rhinorrhea and nasal congestion that spontaneously resolved. For the past 3 days he has had per

History:

-General fatigue
-Fever 39c
-SOB
-Maculopapular skin rash
-Bilateral non-suppurative conjunctivitis
-Abdominal pain
-Decrease urine output
-Decrease oral intake
-No history of syncope
-No history of palpataion
-History of recent infection

Physical examination:

-General appearance : pt was ill-looking , irritable, hypoactive.

-Vital signs :

1-BP 85\50 mmhg (hypotensive)
2-HR 140 beats\min (tachycardia)
3-RR 30 breaths\min (tachypnea)
4-Delayed capillary refill (>3 second)
5-Scattered, mobile, lymph nodes are palpable in the anterior cervical chain bilaterally.

Cardiac examination:*

-Pansystolic murmur was heard at the apex
-S3 heart sound
-Elevated JVP + positive hepatojuguler reflux

****Respiratory examination:***

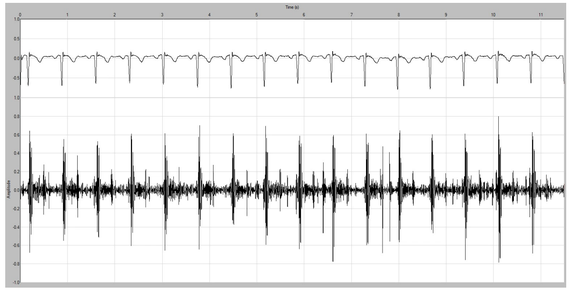
-Tachypnea
-Decrease breath sound
-Bilateral lung crepitation

****Abdominal examination:***

-No ascites
-Liver palpable 4 fingers below costal margin

Extremities examination:*

-Cold extremities
-No lower limb edema



DDX.

- Acute rheumatic fever
- Kawasaki disease
- Viral myocarditis

*Investigation:

CBC(leukocytosis)

C-reactive protein , ferritin, LDH, D-dimer, ESR(elevated)

ABG (metabolic acidosis)

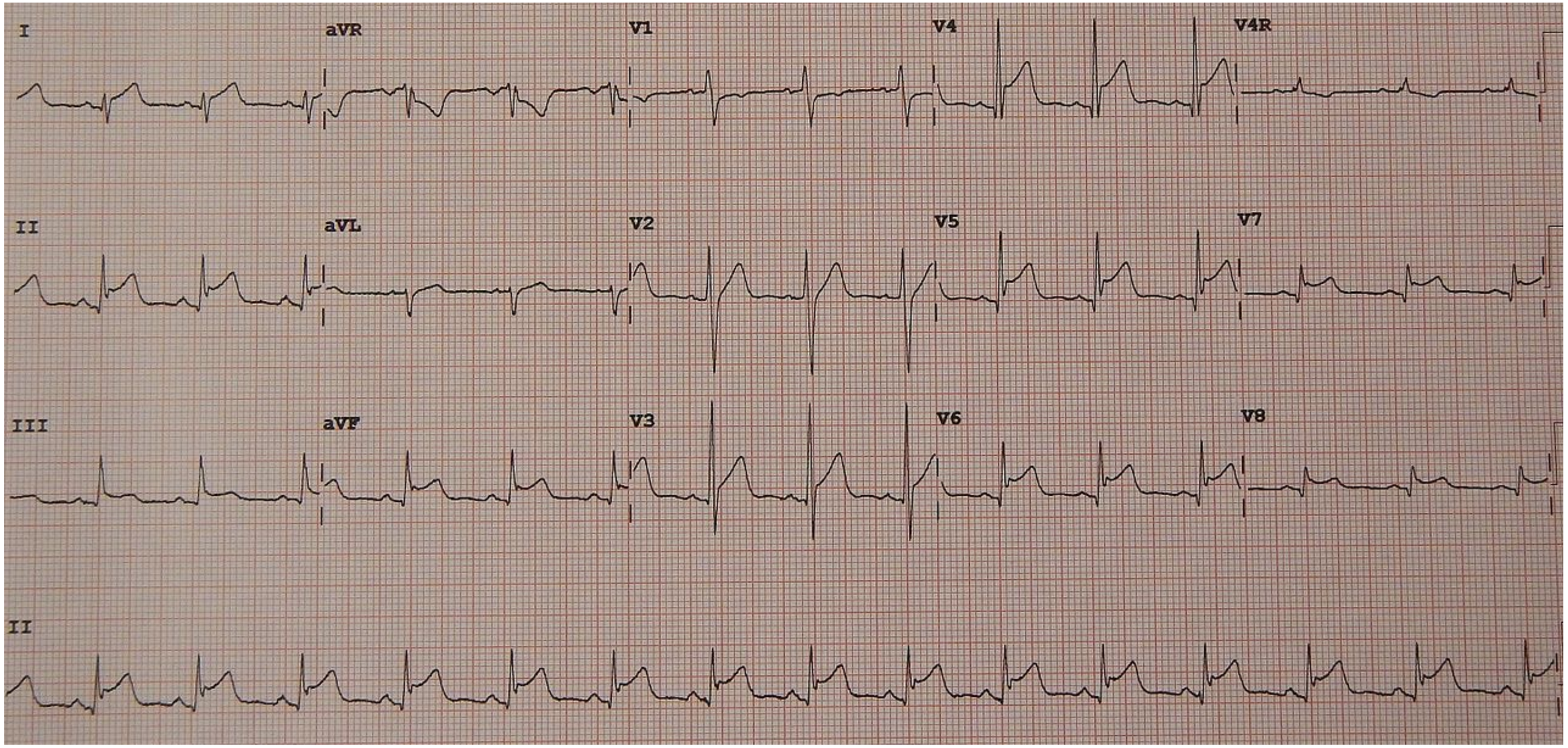
Electrolytes(hyponatremia)

Cardiac enzymes (elevated troponin)

KFT(elevated creatinine)

LFT(elevated AST. ALT)

ECG: decreased QRS voltage and diffuse ST elevation may suggest myocardial edema or pericardial effusion and may be present in children with myocarditis

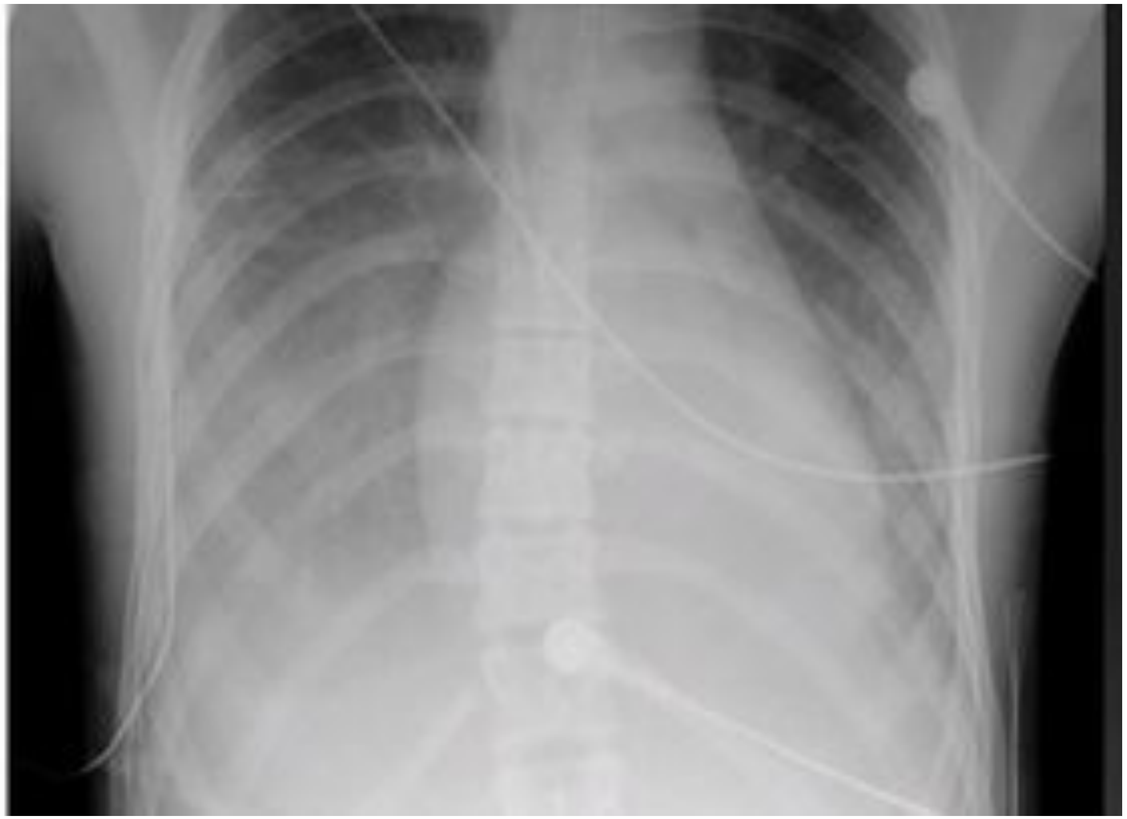


Chest x-ray:

(mild cardiomegaly , bilateral plural effusion , prominence of central pulmonary vessels and interstitial haziness suggestive of pulmonary edema

-Echo

(decreased EF 40%-left ventricular systolic dysfunction, mild mitral regurgitation, mild pericardial effusions)



Viral myocarditis

Incidence	• <1 per 100,000 per year
ETIOLOGY	• Infectious(The most common causes of viral myocarditis are enterovirus (coxsackie group B), adenovirus, parvovirus B19, Epstein-Barr virus, cytomegalovirus, and human herpes 6 (HHV-6)) • Toxic • Autoimmune.
Presentation	• Chest pain (45 percent) • Respiratory distress (28 percent) • Gastrointestinal symptoms (27 percent) • Hepatomegaly (27 percent) • Gallop rhythm (20 percent) • Poor perfusion/diminished extremity pulses (16 percent) • Viral prodrome (41 percent)
History	• Prodrome of fever, myalgia, and malaise several days prior to the onset of symptoms of heart dysfunction. • Recent respiratory or gastrointestinal illness within the previous two weeks.
Complication	• Acute myocarditis may be followed by either a complete recovery of LV function or a secondary dilated cardiomyopathy with chronic HF.

-Myocarditis often results in ventricular dysfunction(dilation)

-presentation of heart failure in 6-8 years of life could be due to HF after myocarditis

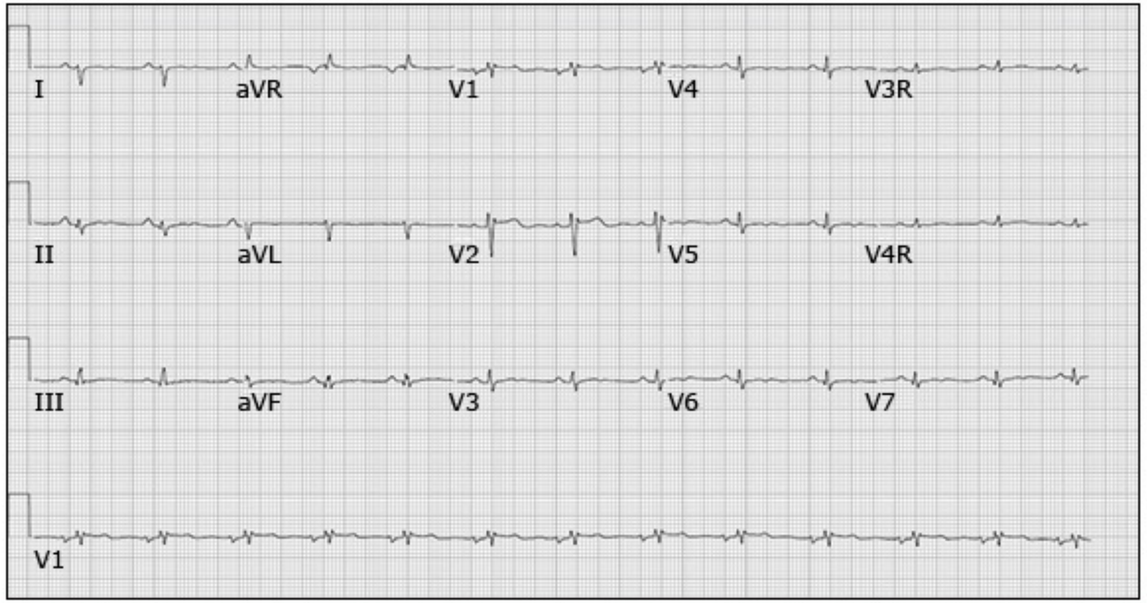
-Cardiac troponin I and troponin T are sensitive biomarkers for myocyte injury,Troponin levels are elevated in myocarditis

Among children presenting with LV dysfunction, an elevated troponin level may suggest acute myocarditis rather than dilated cardior

-BNP levels can help discriminate between cardiac disease and noncardiac causes of HF symptoms

-Left atrial and ventricular size may be helpful in determining chronicity:
LV dilation, a spherical appearance, and eccentric hypertrophy (increased mass with normal wall thickness), often described as components of remodeling, usually suggest longstanding dilated cardiomyopathy rather than an acute process (eg, myocarditis

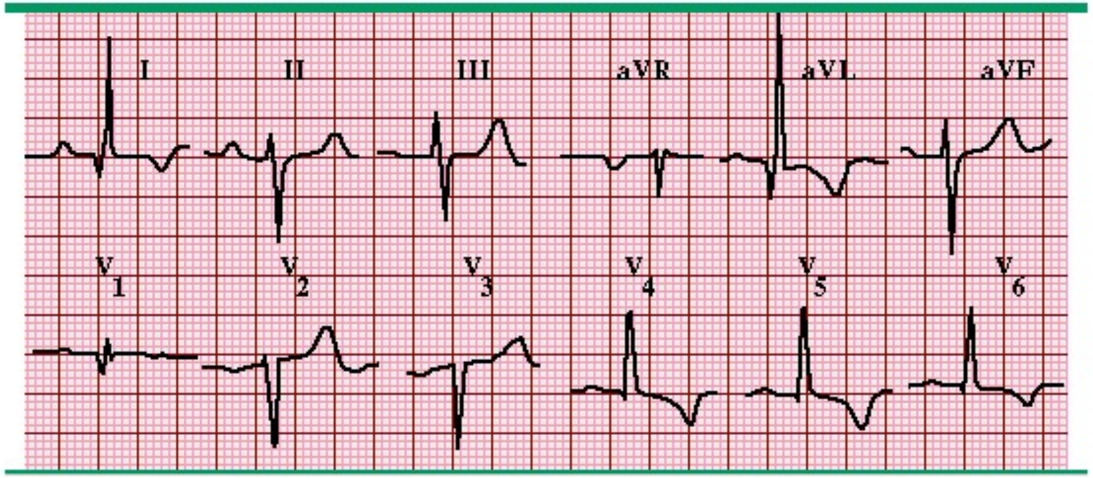
Electrocardiogram in a 17-year-old female with myocarditis



Electrocardiogram demonstrating decreased QRS voltage with ST segment and T wave abnormalities in a 17-year-old female with myocarditis.

Decreased QRS voltage may suggest myocardial edema or pericardial effusion and may be present in children with myocarditis

Left ventricular hypertrophy with strain pattern



The ST-T wave abnormalities secondary to LVH (often termed "strain") are most often seen in the anterolateral leads (eg, I, aVL, V4-V6). Typical abnormalitites include a horizontal or downsloping ST segment and T wave inversions. In some cases, there is concavity to the ST segment, which has a final downward turn that blends into an inverted T wave.

ST segment and T wave abnormalities are common in all forms of cardiomyopathy and myocarditis.