A 32 years old male with dyspnoea and generalised bluish discoloration

Presented By
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Particulars of The Patient

- Name: Md.Rimon
- Age: 32 years
- Sex: Male
- Ward: CS HDU
- Bed: 05
- Address: Sreenagar, Munshiganj
- Date Of Admission: 4.4.2024
- Reg. Number: 55780/130

Chief Complaints

i. Breathlessness since childhood.
ii. Bluish discoloration of the lips and fingers during exertion since childhood.

History of present illness

According to the patient's statement, he has been suffering from breathlessness since his childhood. His breathlessness was less marked in earlier stage, only felt during moderate to severe activity. But for the last 6 months, it is progressively increasing, even during mild exertion. There is no seasonal variation of breathlessness and it is not associated with exposure to dust, pollen or fume. Breathlessness was not associated with fever, cough or sputum production.

There is no history suggestive of paroxysmal nocturnal dyspnea or orthopnea. The patient also noticed bluish discoloration of skin, finger nail, toes and lips which is more marked during physical exertion and becomes less by taking squatting position. This occured several times in childhood but he could not mention the exact number. For the last 3 months this episode has become more frequent. For the last 3 months he has been admitted multiple times in DMCH medicine department for blood letting.

His bowel and bladder habits were normal. His mother mentioned that the patient used to become bluish and breathless while feeding or crying during the first few years of life.

- History of past illness : As mentioned earlier.
- Family history :He is married. Has 2 daughters ,one son. No history of consanguineous marriage. No history of similar illness among family members.
- Personal history : Nothing Contributory
- Socioeconomic history : Middle Class
- Drug history: He is taking metoprolol 25 mg twice daily regularly for last 2 years.
- Immunization History: He is immunized as per EPI schedule & has completed vaccination against COVID-19.

General Examination

- The patient was short in stature, co-operative, suffused conjunctiva
- Dyspnoeic. NYHA class 3.
- Central & peripheral cyanosis (involving tongue, lips, fingers and toes) was present.
- There was generalized clubbing (involving all fingers and toes).

- Pulse: 112/min, low volume, regular in rhythm and normal character
- BP: 100/80 mm Hg
- Temperature: 98ºF
- Respiratory rate: 28/min
- No anemia, leukonychia, koilonychia, edema, jaundice, lymphadenopathy or thyroid enlargement.
- Saturation was 86 by pulse oximetry in room air with out oxygen.

Systemic Examination

Cardiovascular system

- Pulse: 112/min, low volume, regular in rhythm and normal character, no radio-radial or radio-femoral delay.
- BP: 100/80 mm Hg
- JVP: Not raised.

Precordium

- **Inspection:**

Visible cardiac impulse was present in apical region.

- **Palpation:**

Apex beat was palpable in the left 5th intercostal space, 9 cm from midline, normal in character.

- Left parasternal heave —present
- Systolic thrill—was present in pulmonary area.

Auscultation:

- First heart sound— was normal in all the areas
- Second heart sound—P2 component was soft in pulmonary area, A2 component was normal in all areas.
- There was a harsh ejection systolic murmur in the pulmonary area, no radiation, more on inspiration.
- No bilateral basal crepitation.

- Examination of other systems revealed no significant abnormal findings.

Salient features

The patient Mr. Rimon 32-year-old, normotensive, nondiabetic, nonsmoker, presented to us with breathlessness since his childhood. His breathlessness was less marked in earlier stage, only felt during moderate to severe activity. But for the last few months, it is progressively increasing, even during mild exertion. There is no seasonal variation to this breathlessness and it is not associated with dust exposure. It is not associated with cough or sputum production.

There is no history suggestive of paroxysmal nocturnal dyspnea or orthopnea. The patient also had bluish discoloration of skin, finger nail, toes and lips since childhood. These features were more marked during exercise and less by taking squatting position. For the last 6 months this episodes also has become more frequent. His bowel and bladder habits are normal. His mother mentioned that the patient used to become bluish and breathless while feeding or crying during the first few years of life. There is no history of such illness in his family.

On examination patient was dyspnoeic. He had both central and peripheral cyanosis & generalized clubbing. Saturation was 86 in normal room air, without oxygen. On cardiovascular system examination there was no arrhythmia. RV heave & systolic thrill in pulmonary area were present. On auscultation 1st heart sound was audible and normal in all 4 areas.P2 was soft in pulmonary area. There was a harsh systolic murmur was present in pulmonary area with no radiation, more on inspiration. There was no bilateral basal crepitation. All other system examination revealed no significant findings.

Problem list

1. Breathlessness since childhood
2. Cyanosis
3. Clubbing
4. History of blood letting

Provisional diagnosis

My provisional diagnosis is Tetralogy of Fallot.

Differential Diagnosis

1. VSD with PS
2. Eisenmenger’s syndrome.
3. Double outlet right ventricle (DORV) with VSD.
4. Ebstein anomaly
5. TGA

Investigations Profile

SN	Name Of Investigation	Reports
01	CBC	
	Hemoglobin	22.2g/dl
	WBC	12.84 (10^3/mm^3)
	Differential Count	
	Neutrophil	81(10^3/mm^3)
	Lymphocyte	14(10^3/mm^3)
	Monocyte	4(10^3/mm^3)
	Eosinophils	0(10^3/mm^3)
	Basophils	1(10^3/mm^3)
	RBC Profile	
	HCT	67.1%
	RDW	17.5%
	TC of RBC	8(10^{12}/L)
	MCV	88fL
	MCH	29pg
	MCHC	33g/dl
	Platelet Count	34(10^3/mm^3)

SN	Name Of Investigation	Reports
02	RBS	5 mmol/L
03	S. Creatinine	0.23 mg/dl
04	S. Electrolytes	Na^{+}128 mmol/L K^{+} 5.6 mmol/L Cl^{-} 101 mmol/L
05	Dengue For IgG & IgM	Negative

SN	Name Of Investigation	Reports
06	SGPT	786 U/L
07	S. Bilirubin(total)	3.1 mg/dl
08	Urine R/E	Normal
09	Anti HIV I&II (ICT)	Negative
10	S. FT3 S. FT4 S. TSH	4.52 pg/ml 0.58 ng/ml 2.13 micro IU/mL

SN	Name Of Investigation	Reports
11	S. Lipase	41.35 U/L
12	Amylase	55.86 U/L
13	ALP	362 U/L
14	Anti HCV	Negative
15	HBsAg	Negative
16	NT proBNP	7066.8 pg/mL
17	Urine C/S	No Growth
18	Blood C/S	No Growth

SN	Name Of Investigation	Reports
19	PROTHROMBIN TIME Control Patient Ratio Index INR	 12 Seconds 15.4 Seconds 1.28 77.9% 1.28

USG of whole Abdomen

Comment:

- Bilateral acute renal Parenchymal Disease.

Chest x-ray- shows

- Boot-shaped heart
- Pulmonary conus is concave (small pulmonary artery)
- Right ventricle enlarged (prominent elevated apex)
- Oligaemic lung.

Color Doppler Echo Report

Comment:

- TOF with 35-40% Overriding of Aorta.
- PFO flow.
- PPG 79 mmhg
- VSD (13mm*20mm)
- Good LV & RV Systolic Function.
- Significant RVOT obstruction with bands'.
- MPA is smaller but branch Pas appears to be of adequate size.

Final diagnosis

- Pentalogy of fallot.

Planned procedure

- Total correction of pentalogy of fallot under cardiopulmonary bypass.

Steps of operation

- Operation was done on 23/4/2024
- General anaesthesia was given with cv line, arterial line & nasal temperature probe.
- Standard median sternotomy was done.
- Establishment of cardiopulmonary bypass was done.
- After Cross clamping of ascending aorta application of cardio-plegic solution in aortic root.
- Mild hypothermia was used.

- Opening of right atrium and incision of abnormal muscle band .

- PTFE patch closure of VSD

- Pulmonary artery transannular patch augmentation

- Closure of PFO and 2 layer closure of RA was done.
- Release of cross clamp after de- airing of heart and regain of normal cardiac rhythm.
- Placement of pacing wire and drain tube
- Chest closure was done with steel wires.
- Total cross clamp time was 110 minute and extracorporeal circulation time was 145 min.

Post –operative events

- Mechanical ventilation with invasive arterial BP monitoring was done in our cardiac surgery ICU.
- Total drain collection in 1st hour was 300 ml and in 2nd hour was 550 ml
- ACT was checked and was 110 which was within normal limit.
- FFP and fresh whole blood were transfused but in 3rd hour again more 400 ml drain came out.
- Patient became haemodynamically unstable.

- So decision to reopen was made and patient was transferred to OT.
- After reopening bleeding was noted from RA suture line . It was secured ,But haemodynamics remained unstable.
- So again cardiopulmonary bypass was established and patient was on cardiopulmonary bypass without arresting the heart for more 2 hours.
- After 2 hours haemodynamics became stable, ABG was within normal limit and saturation became stable.

- Then patient was weaned from cardiopulmonary bypass ,chest was closed & again shifted to cardiac surgery icu.
- Patient was extubated on next morning.

Post-operative follow up

- At 1 month after operation his resting saturation was 96.
- Echocardiography showed: No residual shunt, No pulmonary stenosis, mild pulmonary regurgitation, good biventricular function, PPG was 21 mm hg.

Take home message

- TOF is a common congenital heart disease
- It has 4 components: VSD, RVH, Overriding of aorta,pulmonary stenosis.
- Surgery is the main modality of management.
- Timely & optimum surgery can reduce mortality and morbidity.

Thank you