CASE REPORT

Pentology of Fallot's presenting with complete heart block

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SUMMARY

A 17-year-old male patient presented with cyanosis, repeated squatting since childhood and haemoptysis since the past 1 month. He had central cyanosis with clubbing. Cardiovascular examination revealed ejection systolic murmur in the pulmonary area with single S2. ECG showed right ventricular hypertrophy (RVH) with right atrial enlargement and first-degree heart block. Two-dimensional echo showed ventricular septal defect, overriding aorta, RVH, right ventricular enlargement (RVE) and right atrial enlargement with infundibular and valvular pulmonary stenosis and 1.9 cm ostium secundum atrial septal defect. There was no evidence of atrioventricular canal defect. The patient was diagnosed with pentology of Fallots. Follow-up ECG showed complete heart block (CHB) that again reverted to first-degree heart block. A diagnosis of pentology of Fallot with intermittent CHB was made with an awake heart rate of 50/min. This case report shows association of CHB with tetralogy of Fallot.

Figure 2 Chest X-ray posteroanterior view showing typical boot-shaped heart in a patient with tetralogy of Fallot.

BACKGROUND

This case holds clinical importance as complete heart block has not been reported with tetralogy of Fallot (TOF) or pentology of Fallot so far. The rarity and novelty of this association starts concern about importance of heart blocks in TOF.

CASE PRESENTATION

A 17-year-old male patient presented to our emergency with cyanosis since childhood with repeated episodes of squatting for the past 5 years. The patient also presented with repeated episodes of

Figure 1 ECG showing right ventricular hypertrophy with right atrial enlargement with first-degree heart block.

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Figure 3 Parasternal long axis view with colour Doppler showing right ventricular hypertrophy, overriding aorta and subaortic ventricular septal defect.

Figure 4 Continuous wave doppler showing infundibular and valvular pulmonary stenosis.

Figure 5 Three-dimensional echo showing atrial septal defect and ventricular septal defect.

haemoptysis since the past 1 month. There was no history of syncopal attacks and the patient was not taking any drugs at the time of presentation. On examination, the patient had central cyanosis with clubbing. Cardiovascular examination revealed apex beat in the fourth left intercostal space in the midclavicular line and a grade 4/6 ejection systolic murmur in the pulmonary

area with single second heart sound. ECG at the time of presentation showed right ventricular hypertrophy (RVH) with right atrial enlargement and first-degree heart block ([figure 1A](#)). Chest X-ray showed boot-shaped heart ([figure 2](#)). Two-dimensional echo showed a large subaortic ventricular septal defect with 50% aortic override ([figure 3](#)), RVH, right ventricular enlargement (RVE) and right atrial enlargement. There was evidence of infundibular and valvular pulmonary stenosis ([figure 4](#)). There was also a 1.9 cm ostium secundum atrial septal defect present ([figure 5](#)). There was no evidence of atrioventricular (AV) canal defect ([figure 6](#)). The patient was diagnosed as having pentology of Fallot and given anxiolytics, cough suppressants and was advised complete rest. There was no history of any surgery. Routine follow-up ECG showed complete heart block ([figure 7](#)) that again reverted to first-degree heart block. The patient was not taking any rate-limiting drug. A diagnosis of pentology of Fallot with intermittent complete heart block (CHB) was made with an awake heart rate of 50/min. Congenital complete heart block associated with TOF is very rare. The patient continues to be free of presyncope or syncope and has refused any intervention.

TREATMENT

Since the patient refused any intervention and he is having an awake heart rate of 50/min, he has been given cough suppressants and anxiolytics. He has not been given β -blockers.

OUTCOME AND FOLLOW-UP

The patient is on routine follow-up and is free of syncope and presyncope.

Figure 6 Two-dimensional echo image apical view of the ostium secundum atrial septal defect.

Figure 7 ECG showing complete heart block.

DISCUSSION

This is a very rare association of CHB with unrepaired pentology of Fallot. AV blocks are frequently seen with congenitally corrected transposition of the great arteries^{1 2} and AV canal defects. In this case, both these anomalies were particularly excluded. Although, a large ostium secundum ASD was present, tricuspid valve was apically displaced excluding the presence of

AV canal defect. Moreover, CHB has been reported after corrective surgery for TOF^{3–5} and pacemaker implantation may be needed, but association of unrepaired pentology of Fallot has not been reported so far. Hence, this case highlights this clinically important finding.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- Association of complete heart block (CHB) with pentology of Fallot is a rare but clinically important finding.
- The CHB can be intermittent or can persist.
- If the patient develops syncope/presyncope and heart rate falls pacemaker implantation should be performed.

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