

A case of cyanotic L-transposition with complete heart block in an adult female who had three in-hospital normal deliveries

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ABSTRACT

A 48-year-old female presented with complete heart block. On evaluation, it was diagnosed as a congenital cyanotic heart disease, namely, L-transposition of great arteries (L-TGA) with Fallot's physiology. She led the normal life of a manual laborer and had three hospital deliveries and yet escaped detection of her cardiac condition.

Key words: Complete heart block, congenitally corrected transposition of great arteries, ventricular septal defect

INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) is a rare condition in which systemic venous blood returns to normally positioned atria. However, the atria are connected to the opposite ventricle [right atrium to left ventricle (LV) and left atrium to right ventricle (RV)], the so-called atrioventricular (AV) discordance. In addition, the ventricles are inverted (right to left change in position) and are connected to the opposite great artery [LV to pulmonary artery (PA) and RV to aorta], thus forming ventricular–arterial (VA) discordance. The aorta is anterior and to the left of the PA, L-transposed. AV discordance plus VA discordance results in normal blood flow (i.e. congenitally corrected). The RV with the tricuspid valve (TV) is the systemic ventricle. Common associated conditions are ventricular septal defects (VSDs)^[1-3] pulmonary stenosis, and congenital

heart block. Major issues related to management revolve around the status of the systemic RV, which can develop dysfunction with increasing age, and tricuspid regurgitation, which can increase in severity with age and contribute to ventricular dysfunction. One emerging treatment is the double-switch operation. In patients with no pulmonary obstruction, it is possible to switch the systemic and pulmonary venous return using an atrial baffle procedure followed by an arterial switch procedure. This results in the anatomical LV now functioning as the systemic ventricle. In those patients with associated pulmonary obstruction and a VSD, another type of double switch can be performed in which the LV is tunneled through the VSD to the aorta, the RV is connected to the PA with a homograft or other conduit, and the atrial baffle procedure is performed. The most difficult challenge is choosing the patient who is a candidate for the double-switch operation and the timing of that operation, or the timing of a more classical operation for associated defects.

CASE REPORT

A 48-year-old female presented to the Medical OPD with complaints of giddiness, chest pain and breathlessness. She had history of such problems from childhood, except

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giddiness. Chest pain was squeezing in type, retrosternal location, aggravated by doing work and radiated to both hands and lower jaw. Dyspnea progressed from grade I to grade III New York Heart Association grading.^[2] Giddiness was of recent onset, on and off with one episode of syncope. There was history of cyanosis with varying intensity noted from childhood. She gave history of working as a headload worker until the age of 30 years without much difficulty, but had to stop thereafter due to increasing chest pain and dyspnea. She had three uncomplicated normal deliveries at a local government hospital (heart disease not diagnosed then).

On examination, the patient was found to have central cyanosis and digital clubbing. Pulse rate was 36/min, regular. Blood pressure and respiratory rates were normal. Jugular venous pulse showed the presence of canon waves. Auscultation revealed RVS₃ and an ejection systolic murmur in pulmonary area, of grade IV intensity.

Investigations

ECG showed 3rd degree heart block with right ventricular hypertrophy (RVH) and left ventricular hypertrophy (LVH).

Repeat ECG showed sinus rhythm with biventricular hypertrophy.

Chest X-ray showed cardiomegaly with biventricular enlargement.

Echocardiography showed:

- Situs solitus, levocardia and L-loop ventricles;
- AV and VA discordance; [Figures 1 and 2]
- Large inlet VSD with bidirectional shunt; [Figure 3]
- PA from right-sided LV;
- PA to left with side by side great vessels;
- Aorta from left-sided RV;
- Pulmonary valve thickened and calcified with severe pulmonary stenosis (PS) and
- Mild right AV valve regurgitation.

Diagnosis

Congenital cyanotic heart disease, L-transposition of great arteries (L-TGA), VSD with severe PS and intermittent complete heart block were detected.

In other terms, there was Fallot's physiology.

Management

Since the patient was not willing for corrective or any other type of interventions (including pacemaker) and she was doing relatively well except for occasional giddiness due to

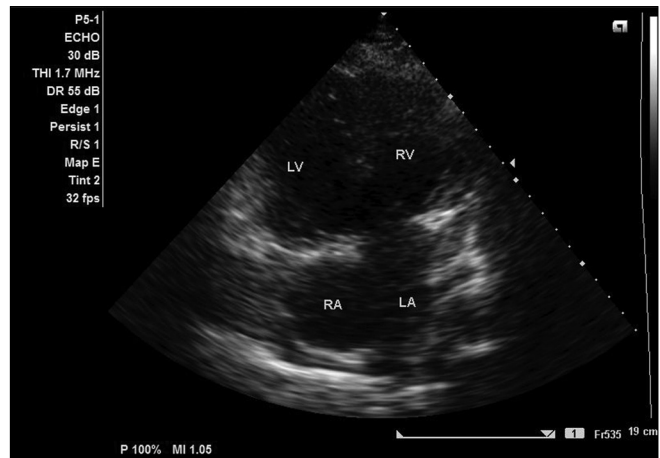


Figure 1: Echo: A-V discordance

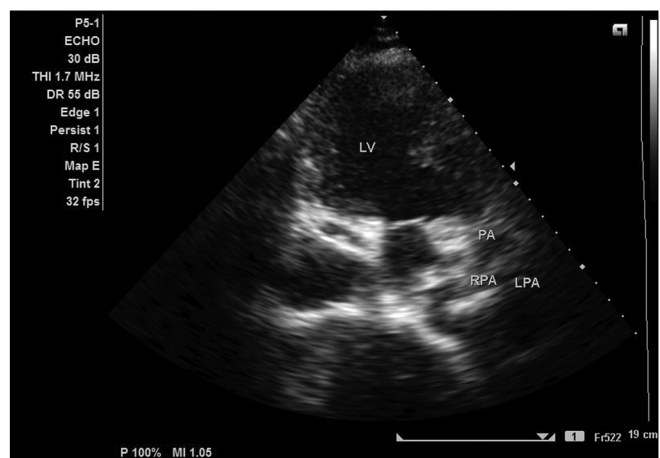


Figure 2: Echo: V-A discordance

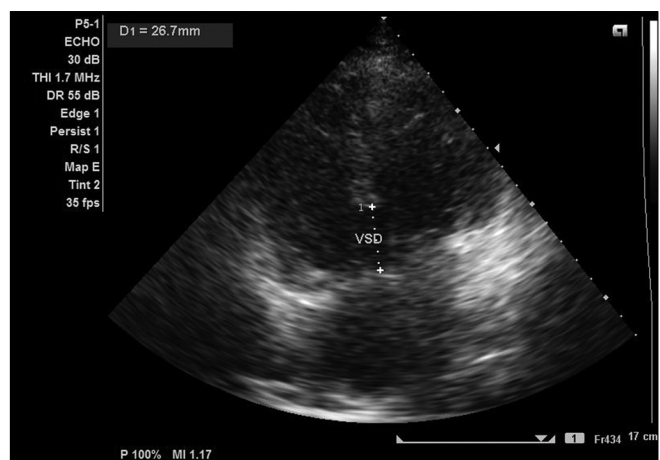


Figure 3: Echo showing VSD

complete heart block, she was put on medical management with diuretics, orceprealin and supportives.

DISCUSSION

This patient remained undetected with congenital cyanotic heart disease well into adulthood and also had three

successful uncomplicated normal deliveries in hospital. A patient with cyanotic congenital heart disease with Fallot's physiology, delivering normally thrice, evading detection of the condition, though the deliveries occurred in hospital is a rare occurrence! She used to work with a road contractor and was doing heavy manual labor though she occasionally had symptoms and cyanosis.

Few reports of pregnancy are available about women with L-TGA.^[4] In the present series, pregnancy was well tolerated in all, but two women. One woman required AV valve replacement in the early postpartum period. The second had congestive heart failure during three pregnancies and toxemia, endocarditis and a myocardial infarction during three other pregnancies. L-TGA does not inhibit fertility in women before or after surgical repair. Successful pregnancy can be achieved in most women, although there is increased risk of maternal cardiovascular morbidity and fetal loss. Data regarding pregnancy in cyanotic L-TGA are lacking.

Natural history

The initial physiology of isolated L-TGA is normal. It is the potential late failure of the RV and TV, both of which face the higher-resistance systemic arterial circuit that most frequently brings these patients to attention as young adults. In this group, progressive dilation of the RV, as the myocardium fails, typically leads to enlargement of the TV annulus and worsening of the tricuspid regurgitation. The volume load of the tricuspid regurgitation, in turn, worsens RV chamber dilation, which further stretches the tricuspid annulus. Longitudinal studies have described outcomes in patients with and without surgical intervention.^[5,6]

Complete heart block is a frequent accompanying or presenting symptom in this population owing to the associated abnormal development of the conduction system.^[7] Other associations include an Ebstein like displacement of the left-sided TV (common), which may contribute to TV dysfunction, and ventricular noncompaction (rare), which may contribute to ventricular dysfunction.^[8]

- VSD: This is the most common associated cardiac malformation, with an incidence of 60–70% in clinical series and nearly 80% in reviews of autopsied cases.
 - The defect is usually large and perimembranous in location but can occur in any position along the ventricular septum.
 - The perimembranous VSD tends to be subpulmonary.

- Subarterial VSDs, roofed by the semilunar valves, have been described in Asian patients but are uncommon in the Western world.
- The resulting left-to-right shunt is usually large.
- Conduction system abnormalities: The sinus node is positioned normally but the anatomical situation precludes normal conduction because the AV conduction tissue is profoundly abnormal. The normal AV node cannot give rise to the penetrating AV bundle. An anomalous second AV node is the functional AV conduction system in many patients, generally located beneath the opening of the right atrial appendage at the lateral margin between the pulmonic valve and the mitral valve; thus, the node has an anterior position and gives rise to the AV bundle immediately underneath the right anterior pulmonic valve leaflet. This accessory node is not always present and may be hypoplastic or nonfunctional.
 - Complete heart block occurs in 30% of patients and may be present at birth or develop at a rate of 2% per year.^[9] Other conduction disturbances described include sick sinus syndrome, atrial flutter, re-entrant AV tachycardia due to an accessory pathway along the TV annulus, and ventricular tachycardia.
- Left ventricular outflow tract obstruction: Left ventricular outflow tract obstruction (pulmonary outflow tract) occurs in 30–50% of patients and is typically associated with a ventricular septal defect.^[10] Freedom reported that of the patients with pulmonary outflow tract obstruction and a VSD, approximately one third have TV deformities.

Mortality/morbidity

- Ten-year survival rate ranges from 64 to 83% from the time of diagnosis and is dependent on associated anomalies.^[6]
- A rare patient without associated cardiac anomalies may have a much more benign course, and literature documents many examples of these patients being diagnosed in the sixth and seventh decades of life.^[6]
- A median age at death of 40 years has been reported in both patients who have undergone operation and those who have not.

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Source of Support: Nil, **Conflict of Interest:** This patient with a complex congenital heart disease had a near normal life of a manual laborer. She had three normal deliveries in hospital (and several other hospital visits), yet evaded diagnosis of her cardiac problem.