Long-term Survival in Tricuspid Atresia after Palliative Surgery

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ABSTRACT

Here we report a rare case of tricuspid atresia. A 46 years old male treated palliatively by pulmonary artery banding. Patient is still surviving and is almost asymptomatic with limited exertion for so many years in spite of chronic hypoxia.

Keywords: Tricuspid atresia, Pulmonary artery, Hypoxia.

INTRODUCTION

Tricuspid atresia (Figs 1A and B) is a type of congenital heart disease in which the tricuspid heart valve is missing or abnormally developed. It was first described in 1817. Frequency is equal in males and females.

The defect blocks blood flow from the right atrium to the ventricle. Incidence is 0.06 per 1000 live births with prevalence in clinical series of congenital heart disease of 1 to 2.4%.1

This defect is contracted during prenatal development, when the heart does not finish developing. In usual variety, there is agenesis of morphologic tricuspid valve so that no communication exists between morphologic right atrium and ventricle. An atrial septal defect (ASD) must be present to maintain blood flow. Systemic venous blood is received by the anatomic right atrium and passes through an interatrial communication to converge with pulmonary venous blood in the morphologic left ventricle.

Thus, the physiologic consequences of tricuspid atresia are:

- Obligatory right to left shunt at atrial level.
- The left atrium receives the entire systemic venous return via interatrial communication and receives the pulmonary venous blood directly.
- Mixture flows directly into the left ventricle.
- If the great vessels are normally related to pulmonary arterial flow is usually reduced, because a restrictive VSD constitutes a zone of subpulmonic stenosis. Left ventricular volume overload is curtailed but at the cost of cyanosis.
- This anatomic condition accounts for about 90% of cases. Approximately, 6% of infants born with tricuspid atresia are premature. The lifespan is shortest less than 6 months. One patient lived 21 years.

In tricuspid atresia with normally related great arteries presence of VSD is desirable when pulmonary blood flow is appropriately regulated but this balance is exceptional rarely favorable balance is achieved permitting survival from 2nd to 5th decade. Excessive pulmonary arterial flow results in volume overload of left ventricle and CCF. Death in these patients is due to Hypoxia and CCF.

Tricuspid atresia is the third most common cyanotic congenital cardiac lesion, with a mortality rate of 90% before the age of 10 years.2 The Fontan operation is the usual goal of therapy for children with tricuspid atresia. According to one retrospective study, 204 patients (86%) were judged suitable for a future Fontan procedure at presentation. However, 99 (48%) of these are known to have died before a Fontan operation or became unsuitable for such surgery during follow-up, mostly because of death after palliative surgery (23 patients, 11%), sudden death (18 patients, 9%), and new adverse features (32 patients, 16%), such as sub aortic stenosis, pulmonary arterial distortion and ventricular dysfunction.3

CASE REPORT

A 46 years old male patient (Fig. 2) was admitted in MGM Hospital, Kamathe and CBD Belapur, Navi Mumbai, Maharashtra, India for plasmodium vivax Malaria. Patient was diagnosed to have congenital heart disease in early childhood which was investigated at John Hapkin Hospital in United States. While performing cardiac catheterization in 1968, it was confirmed that the patient was suffering from tricuspid atresia, therefore, he was subjected to pulmonary artery banding. Later on, the patient was re-evaluated at Southern Railway Hospital, Perambur, Tamil Nadu, India, at
Figs 1A and B: (A) Normal heart and (B) Tricuspid atresia (Courtesy: Adam)

Fig. 2: Patient’s photograph

Fig. 3: Central cyanosis

Fig. 4: Clubbing

Fig. 5: Polycythemia
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the age of 16, in 1980. Findings after cardiac catheterization were as follows:
• Tricuspid atresia with post pulmonary artery banding.
• Large left ventricle with small hypoplastic right ventricle.
• High flow in pulmonary artery, (Large pulmonary artery with banding).

The patient was advised not to undergo any further procedure, since he was not found suitable for Fontan procedure.

Since, the patient was having high fever, he was admitted at MGM Hospital, CBD Belapur, Navi Mumbai, Maharashtra, India and was diagnosed that he was suffering from plasmodium vivax positive malaria.

His clinical evaluation reveals Central Cyanosis (Fig. 3), Grade 3 Clubbing (Fig. 4) and Hypoxia with SpO$_2$ 84 to 85%.

His hemoglobin was 23 gm% and he was having polycythemia (Fig. 5) with PCV of 75% without any evidence of iron deficiency.

His uric acid level which was 11 mg% has come down to normal with tablet Zyloric. Other blood reports including LFT, RFT, sugar, lipid profile were normal except serum triglycerides 195 mg%.

X-ray (Fig. 6) chest shows CTR of 55% with near normal vascularity.

PFT showed minimal obstructive lung defect with an additional restrictive lung defect.

The cardiovascular examination revealed grade 4/6 systolic murmur with thrill.

**Echocardiography (Figs 7A and B) revealed**
- S/P pulmonary banding.
- Tricuspid atresia.
- Large ASD with right to left shunt.
- Large muscular VSD with left to right shunt.
- Significant subovolular RVOT obstruction (Gradient 95 mm Hg).
- Dilated hypokinetic LV with EF 40%. Dilated LA.
- Good size pulmonary artery and branches.

**Electrocardiogram (ECG) (Graph 1)**

**SUMMARY**

Thus in summary, patient is a middle aged gentleman, who was diagnosed to have tricuspid atresia with high flow in pulmonary arteries, subjected to pulmonary artery banding at the age of 4 years, still surviving where mortality rate is more than 90% without definitive surgery, now developed significant subpulmonary obstruction and is being considered for TCPC or Bidirectional Glenn or BT shunt. Though patient is having chronic hypoxia (SpO$_2$ 84-85%), he can do his routine activities without feeling breathless or fatigued.
Graph 1: Electrocardiogram (ECG)

REFERENCES


