Hypoplastic Right Heart Syndrome - A Case Report

Rekha Matta[1,*], Manjunath Girish[2] and Bhaktavatsala Reddy[3]

[1]*Department of Cardiothoracic and Vascular Surgery;* [2]*Cardiothoracic and Vascular Anaesthesiology;* [3]*Cardiology, Krishna Institute of Medical Sciences, Karad, Maharashtra, India*

Abstract: Hypoplastic right heart syndrome is not a commonly used generic term like Hypoplastic left heart syndrome. It could be in the form of hypoplasia of the tricuspid valve, right ventricle and pulmonary artery or the more common variety i.e. tricuspid atresia.

Keywords: Hypoplastic right heart, Bidirectional Glenn shunt, atrial septal defect.

INTRODUCTION

Hypoplasia of the right heart could occur as an isolated congenital defect or in association with other cardiac anomalies such as atrial septal defect, ventricular septal defect, transposed great arteries, truncus arteriosus etc. The type and timing of clinical presentation depends on the degree of hypoplasia and the presence or absence of associated cardiac anomalies. We present a case of Hypoplastic right heart syndrome.

CASE REPORT

A 14 year old boy, weighing 34kg, presented with history of exertional dyspnoea, functional class 2 since one year. Clinical examination revealed central and peripheral cyanosis, clubbing and a 3/6 ejection systolic murmur audible over the left upper sternal border. His hemoglobin level was 16.4gm% and hematocrit was 50%. Echocardiogram report was Common atrium, sub aortic ventricular defect (V.S.D) with left to right shunt, right ventricular systolic pressure (RVSP) of 52 mm Hg. Cardiac catheterization: systemic saturation off oxygen was 85%, mean pulmonary artery pressure was 15mm Hg. The catheter course was right femoral vein (RFV) to inferior vena cava (IVC) to right atrium (RA) to left atrium (LA) to left superior pulmonary vein (LSPV). Left ventriculogram showed a sub aortic VSD and a closed mid-muscular VSD. The surgical findings were (i) 4cm by 2cm ostium secundum atrial septal defect (ASD) (ii) intact septum primum (iii) single, moderate sized sub aortic VSD (iv) no additional VSD (v) hypo plastic tricuspid valve (annulus diameter-14mm; expected – 24 mm), right ventricle, pulmonary annulus (diameter – 15mm; expected – 19mm), main pulmonary artery (diameter – 15mm; expected – 19 mm). (vi) Right and left pulmonary arteries were adequate size (13mm). (vii) Mitral valve was normal. (viii) no systemic or pulmonary venous anomaly. (ix) absent patent ductus arteriosus. The surgical procedure done was Dacron patch closure of the VSD, pericardial patch closure of the ASD and bidirectional cavopulmonary shunt (Figure **1**, **2**). The child came off cardiopulmonary bypass in sinus rhythm with elective Dobutamine support (five micrograms per kilogram per minute). Right atrial and pulmonary arterial blood gas analysis did not show any step-up in the oximetry. The child was extubated 4 hours after shifting to the intensive care unit. The shunt pressure was 14mm Hg. Post operative echocardiogram (Figure **3**) showed (i) No residual ASD, VSD. (ii) Well functioning Cavopulmonary shunt (peak gradient – 3.9mmHg). (iii) RVSP—20mm Hg (iv) Hypoplastic tricuspid value (annulus = 13mm), pulmonary annulus (14mm) and main pulmonary antery (14mm). (v) No tricuspid regurgitation (vi) Normal mitral value (annulus 26mm). The child was discharged on the eight post operative day.

DISCUSSION

Hypoplastic right heart syndrome comprises 1-3 % of all congenital heart defects [1]. The strategy of operative correction is to establish adequate pulmonary blood flow by diverting the systemic venous return directly to the pulmonary arteries. Cavopulmonary shunts were first described in 1961 by Glenn *et al*. [2]. Their advantage over systemic – pulmonary shunts are they are more physiological as (i) they direct fully desaturated blood into the pulmonary circulation (ii) low pressure venous blood is being fed into the pulmonary circulation and hence carry less risk of pulmonary vascular disease (iii) there is less risk of distortion of the pulmonary artery (iv) there is no risk of volume overload of the left ventricle.

*Address correspondence to this author at the Professor & Head of Department of Cardiothoracic and Vascular Surgery, Krishna Institute of Medical Sciences, Karad –41 5539, Maharashtra, India; Tel: +91 8275259763; Fax: 02164 – 241866; E-mail: rekhamatta@gmail.com

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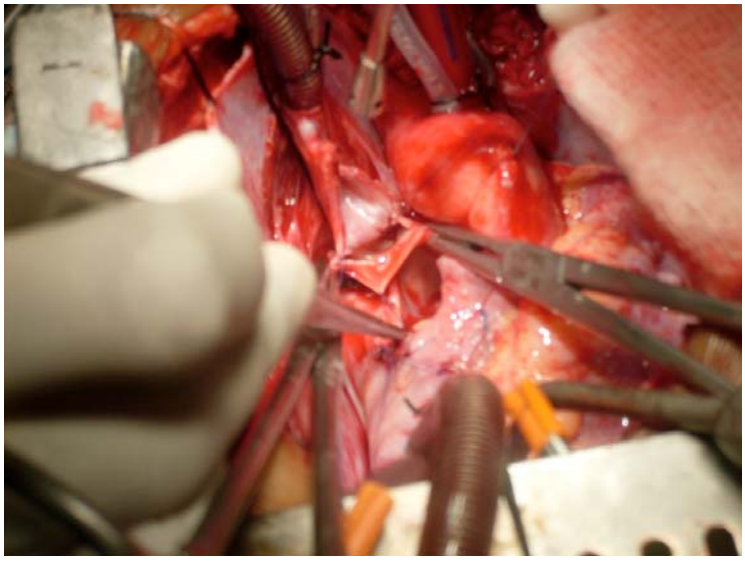

Figure 1: Bidirectional Glenn shunt being constructed.

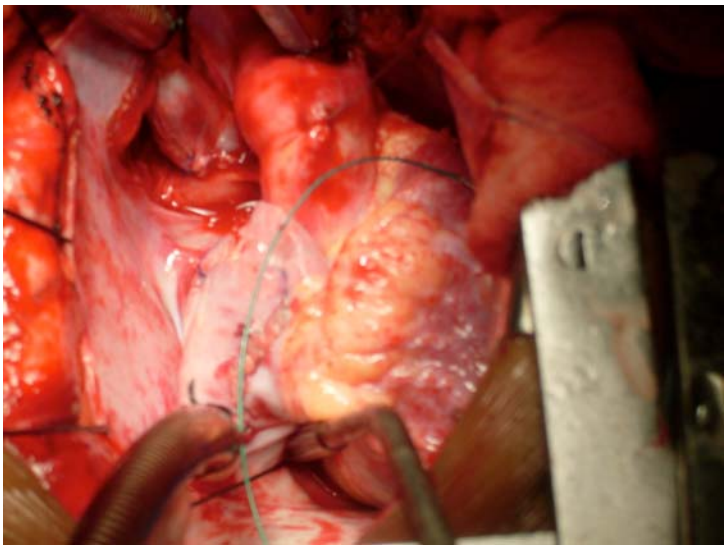

Figure 2: Bidirectional Glenn shunt completed.

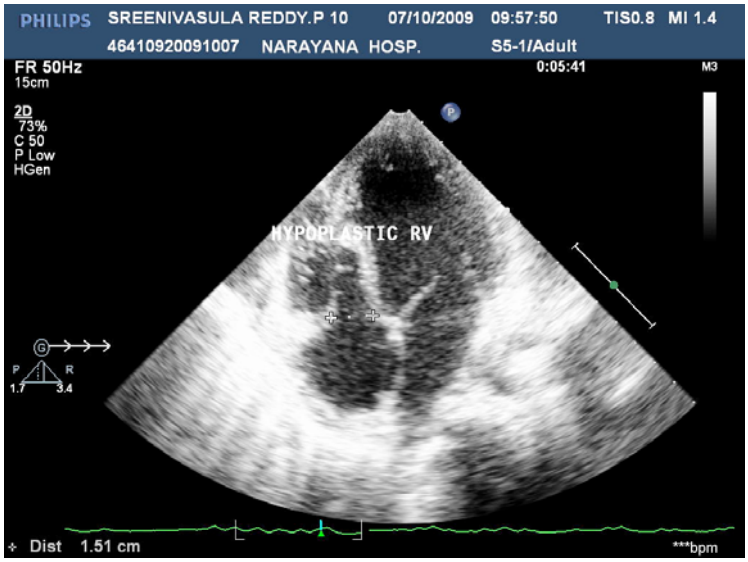

Figure 3: Echocardiography showing hypoplastic tricuspid valve and right ventricle.

The pre-requisites for construction of cavopulmonary shunts are: (i) hypoplastic right heart (ii) adequate size branch pulmonary arteries (iii) normal pulmonary artery pressure (pulmonary vascular resistance index less than 4 Wood units / m^2 or mean pulmonary artery pressure less than 15 mm Hg) (iv) good left ventricular function and (v) absence of tricuspid regurgitation.

Bidirectional cavopulmonary shunt diverts blood from the superior vena cavae to the pulmonary arteries with preservation of the right and left pulmonary arteries. It can be constructed with or without the use of cardiopulmonary bypass (CPB). When CPB is not used, it is prudent to decompress the upper torso venous system by heparinising the patient and placing cannulae in the superior vena cava and in the right atrium and connecting them after deairing [3].

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