

Successful repair of complex atrio-ventricular septal defect in an Egyptian child.



Fig 9 Baby Ebtisam Ibrahim

Introduction:

Atrio-ventricular septal defects are a spectrum of uncommon congenital cardiac anomalies comprising of 1-3% of all congenital heart defects¹. An Egyptian child, diagnosed to have a complete AV septal defect was referred to us for further management. On evaluation, she was found to have complete AV septal defect with straddling of the left AV valve. She underwent successful two-patch repair under total circulatory arrest at Frontier Lifeline in July this year.

Case Report:

An 8-month-old female child, first born of a non-consanguineous marriage, presented with complaints of tachypnoea and sweating. There was no history of cyanosis, poor feeding, or failure to thrive. There was no history of any perinatal insult. On examination, the child was alert, saturating 81% on room air, and weighed 6 kgs. Features of Down's syndrome were noticed. Cardiovascular examination revealed normal S1, widely split S2 and a pan -systolic murmur at the left lower sternal border. Chest radiograph showed cardiomegaly and pulmonary plethora. Electrocardiograph showed normal sinus rhythm, extreme left axis deviation and evidence of right ventricular volume overload. Echocardiography showed Rastelli Type A complete AV septal defect with mild AV valve regurgitation and good bi-ventricular function.

The heart was approached through a midline sternotomy. On standard cardiopulmonary bypass, the child was cooled to 18°C and circulation was arrested. Through a right atriotomy, the internal morphology was visualized. Complete AV septal defect was observed. Additionally there was straddling of the left superior leaflet of the AV valve. A two-patch repair was carried out. Goretex patch was used to close the ventricular septal defect and bovine pericardial patch was used to close the ostium primum defect. Two competent AV valves were reconstructed during the process by committing the bridging

leaflet towards the left side, Circulation was re-started. The right-atriotomy was closed, the child re-warmed and weaned off bypass in normal sinus rhythm with acceptable hemodynamics. The post-operative period was uneventful and the child was discharged on the eighth post-operative day. Echocardiogram at discharge showed two competent AV valves, no inter-atrial or inter-ventricular communications and good bi-ventricular function.

Discussion:

Faulty development of the endocardial cushions leads to a spectrum of anomalies ranging from a simple ostium primum defect to complex complete atrio-ventricular defects. There is a strong association between Down's syndrome and complete AV septal defect, with 45% of Down's babies having AV septal defect², and more than 50% of babies with AV septal defect have Down's syndrome^{3,4}. The clinical symptoms in these children, is due to left to right shunting at the atrial and ventricular levels. Poor feeding, failure to thrive, tachypnoea, sweating are some of the common clinical features. Congestive cardiac failure is very common early in life and is the most common cause of death. Diagnosis is easily accomplished trans-thoracic echocardiography. Attention is given in particular to the number, morphology and competency of the AV valves, and the ventricular function. The presence of pulmonary artery hypertension can be ascertained by echocardiography, although cardiac catheterisation may sometimes be necessary to quantify pulmonary vascular resistance especially in older children.

Surgical correction has to be done early to prevent the development of irreversible pulmonary artery hypertension, which occurs rapidly in Down's children. The surgical technique employed at Frontier Lifeline is the two-patch technique. At surgery, care should be taken to reconstruct competent AV valves. In addition, this child had significant straddling of the left AV valve, which in most instances will preclude a bi-ventricular repair. Two competent AV valves were reconstructed by leaving more valve tissue on the left side (i.e.) committing the common bridging leaflet towards the left. Ensuring competency of the valves is essential as the presence of regurgitant AV valves post-operatively may necessitate re-operation. Long-term results of surgery are excellent with survival rates approaching 80 % at 10 years⁶.

This child was the first patient from Egypt to be operated at Frontier Lifeline. Our centre caters to patients from several countries where complex congenital cardiac anomalies are diagnosed, but infrastructure to correct them does not exist. Prior to coming to India, the parents had consulted at many European centres and finally decided to come to Frontier Lifeline; this is a positive pointer towards the increasing confidence of overseas patients in our medical facilities and promotes the growth of medical tourism in India.

References:

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