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Case Report

Hypoplastic right heart syndrome

Rajaram M. Weling*, Nilima A. Narkhede

Department of Pediatrics, Bharatratna Dr. Babasaheb Ambedkar Municipal General Hospital, Mumbai, Maharashtra, India

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***Correspondence:**
Dr. Rajaram M. Weling,
E-mail: welingrajaram@gmail.com

ABSTRACT

Congenital heart diseases (CHDs) are of two types-acyanotic and cyanotic. Acyanotic heart diseases include atrial septal defect ASD, ventricular septal defect VSD, patent ductus arteriosus PDA, coarctation of aorta and atrioventricular cushion defects. Cyanotic heart disease includes Fallot's tetralogy, transposition of the great vessels, Total anomalous pulmonary venous return, Tricuspid atresia and single ventricle or hypoplastic left heart or hypoplastic right heart syndrome (HRHS). Hypoplastic right heart is a rarer condition than the hypoplastic left heart. Our patient had a cyanotic heart disease; presented to us with hypoplastic right heart, a very rare association consisting of pulmonary atresia combined with a hypoplastic right ventricle and well-developed pulmonary arteries. Early clinical diagnosis and surgical intervention saved this child.

Keywords: CHDs, HRHS

INTRODUCTION

Congenital heart diseases (CHDs) are of varied types, severity, and complexity.[1-5] Simple scientific approach along with sophistication is required for successful management at all levels. Foetal echocardiography and nondirective genetic counselling are important. Early detection and expert diligence are desirable. The suspicion is clinical. A number of clinical and investigative diagnostic modalities are available and should be utilized as guided by the clinical cues. Medical management includes general health maintenance, immunizations, monitoring, and complications treatment. Paediatric interventional cardiology is making rapid strides and treating many types of lesions. Surgery is of curative, reparative, or palliative types. After surgical correction in early childhood long-term follow up is needed. Here is our case report of a one-month-old child who was diagnosed as HRHS and a timely cardiac intervention was done. HRHS is a condition that is even more rare than HLHS.[6,7] HRHS refers to the underdevelopment of the right-side structures of the heart, which means that the chambers, valves and related blood vessels on the right side of the heart are malformed. This malformation involves the pulmonary valve atresia which has not formed, a very small right ventricle, a small tricuspid valve and a small hypoplastic pulmonary artery. As the ventricle has failed to grow and develop the ventricles muscle structure is poor, so additional problems are encountered as the heart attempts to pump blood to the pulmonary valve for transfer to the lungs. The proper amount of blood pumped from the right atrium is not sufficient and this causes the blood to be not pumped efficiently to the lungs.

CASE REPORT

One month old male child born of second-degree consanguineous marriage, fourth by birth order, no complaints in the previous siblings and no significant birth history was brought to us at Bharat Ratna Dr Babasaheb Ambedkar municipal general hospital with cough and breathing difficulty. In view of the above we did an x-ray chest which showed a pneumonia patch and

raised WBC counts, antibiotics were started for the same and continuous positive pressure oxygen was given for the same. In spite of all this the four-limb pulse oximeter reading showed less than 85% in all four limbs. Also, there was a suck rest suck cycle with cyanosis of the fingers on excessive crying. A clinical suspicion of a congenital cardiac anomaly was thought off. A 2D echocardiogram was done which showed pulmonary atresia, intact interventricular septum, patent foramen with left to right shunt, left ventricular hypertrophy, septal hypertrophy, restrictive duct, small bipartite right ventricle with hypoplastic right heart, moderate tricuspid regurgitation (Figure 1 and 2). The child was referred to cardiac center where patent ductus arteriosus stenting was done followed by PBPV (percutaneous balloon dilatation of the pulmonary valves). A good clinical examination which picked up signs of congenital cyanotic heart disease supported by a timely cardiac procedure saved this child.

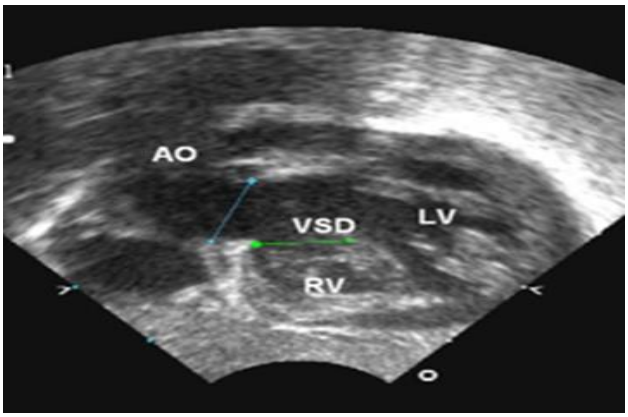

Figure 1: 2D echo-subcostal view showing HRHS.

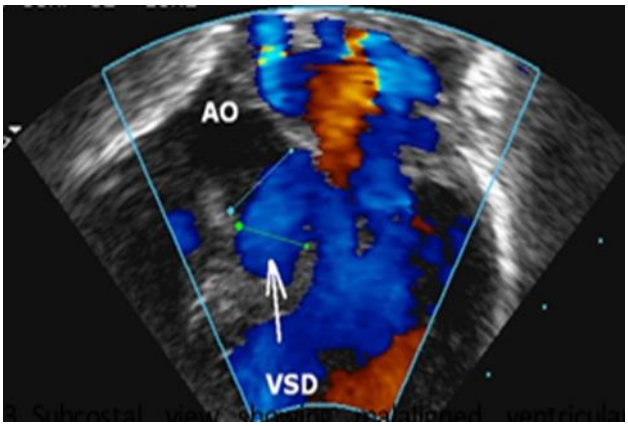

Figure 2: 2D echo with colour contrast showing HRHS.

DISCUSSION

Congenital heart diseases are of two types- acyanotic and cyanotic. Acyanotic heart diseases include atrial septal defects ASD, ventricular septal defects VSD, patent ductus arteriosus PDA, coarctation of aorta and atrioventricular cushion defects. Cyanotic heart disease includes Fallot's tetralogy, Transposition of the great vessels, Total anomalous pulmonary venous return, Tricuspid atresia and single ventricle or hypoplastic left heart or hypoplastic right heart. Hypoplastic right heart is a rarer condition than the hypoplastic left heart. Our patient had a cyanotic heart disease with hypoplastic right heart. Clinically we suspected this rare disease because of central and peripheral cyanosis, suck-rest-suck cycle, all four limb pulse oximeter readings less than 85% in all four limbs and cyanotic spells while crying. Also, in a child up to 6 months of age which presents with recurrent respiratory infections, a high index of suspicion is required for a congenital cardiac disease. 2D echocardiogram is the diagnostic tool to rule out congenital heart. Hypoplastic right-heart syndrome (HRHS)is a rare, cyanotic congenital heart malformation caused by underdevelopment of the right-sided heart structures (tricuspid valve, RV, pulmonary valve, and pulmonary artery) commonly associated with an atrial septal defect, ostium secundum type.[7-9] Pulmonary blood flow is diminished and right-to-left shunting occurs at the atrial level, leading to dyspnoea, fatigue, atrial arrhythmias, right-sided heart failure, hypoxemia, repeated miscarriages that were mostly due to hypoxemia and cyanosis. Two subtypes of HRHS have been characterized: pulmonary atresia-intact ventricular septum and right ventricular hypoplasia. The PDA stenting with percutaneous balloon dilatation of the pulmonary valves (PBPV) was a lifesaving cardiac procedure in this child. The further management will consist of univentricular tract with Glenn in few months and later continuation of the Fontan procedure.[10-13]

CONCLUSION

Our patient had presented to us with a congenital heart disease. A four-limb pulse oximeter reading is helpful in picking up a congenital cyanotic heart disease. 2D echo is the diagnostic tool to establish the type of cyanotic heart disease. Early diagnosis prompted us to refer the patient to cardiac center, which helped the child survive the complicated hypoplastic right heart disease.

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Ethical approval: Not required

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