Atrial septal defect presenting as intractable congestive heart failure in an infant with dismal outcome: A Case report

Ankit Pachauri¹, Shalini Tripathi², Mala Kumar³

From ¹Resident, ²Additional Professor, ³Professor, Department of Pediatrics, King George Medical University, Lucknow, Uttar Pradesh, India

ABSTRACT

Congenital heart disease is a major cause of morbidity and mortality in children. We are presenting a 2-month-old infant with large ostium secundum atrial septal defect (ASD) with congestive heart failure. Isolated ASD usually does not present at this age. We are presenting this case due to unusual early presentation with isolated ASD.

Key words: ASD in infancy, ASD with CHF, Unusual presentation of ASD

The incidence of congenital heart disease (CHD) in live born infants varies between 6 and 8/1000 live births, resulting in 25,000–35,000 infants with CHD in United States [1]. In India, this number is estimated to be 2 lakh/year, with 1/5 of them requiring intervention in the 1st year [1]. This incidence has remained constant over the past several decades. Often late diagnosed and accompanied with high complications which require early surgical intervention [2]. Data from various studies suggest that most common CHDs presenting in children are acyanotic CHD (ACHD) [3] among which ventricular septal defect and atrial septal defect (ASD) are the most common.

CASE REPORT

A 2-month-old female infant presented with respiratory distress and fever, had history of recurrent pneumonia in the past and in one instance got intubated. Patient was managed conservatively, extubated thereafter and discharged. Then this patient came to our side with respiratory distress, cough, and refusal to feed for 2 days. She was first born child, a product of non-consanguineous marriage, delivered at term gestation. Her growth and development were age appropriate. On examination she was febrile, was in shock, tachypneic (RR-64/min) with moderate chest retractions, and nasal flaring with oxygen saturation of 85–88% at room air which later improved to 92–94% on non-rebreathing oxygen mask. There was mild pallor. On auscultation of chest, there was bilateral equal air entry with wheeze, conducted sounds. There was tachycardia (HR-168/min), ejection systolic murmur Grade 2

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with wide and fixed split S_2 . Abdomen was soft, nontender; liver was 6 cm below costal margin. Arterial blood gas analysis showed hypoxemia and metabolic acidosis. A clinical diagnosis of ACHD with congestive heart failure (CHF) was made.

Initial investigations revealed Hb-11 g/dl, TLC-16,000 with neutrophilic predominance. Chest X-ray (Fig. 1) showed cardiomegaly with the right atrial dilatation. ECG was suggestive of tachycardia with the right axis deviation (RAD) with the right ventricular hypertrophy. 2D echocardiography was suggestive of 13 mm ASD with the left to right shunt with RAD with the right ventricular enlargement (RVE) with mild PAH. As ASD does not usually present in this age group, a repeat 2D echocardiography was done which again showed 10 mm ASD with the left to right shunt with RAD with RAD with RVE.

Patient was managed on restricted intravenous fluid, dobutamine, oxygen by mask, furosemide, enalapril, nebulization with salbutamol, and intravenous antibiotics. As the condition of the patient didnot improve with this treatment intravenous digoxin was added. Blood culture showed growth of *Staphylococcus haemolyticus*, so accordingly antibiotics were changed.

The child was planned for early surgical correction and referred to higher center but she developed severe pneumonia, sepsis, and multiple organ dysfunction syndromes and expired despite all conservative measures.

DISCUSSION

Data from large population studies all over the world show that approximately 1/110 live births have CHD and approximately 25% of CHD are considered critical, requiring intervention in the

Correspondence to: Dr. Ankit Pachauri, Department of Pediatrics, King George Medical University, Lucknow, Uttar Pradesh, India. E-mail: ankit_pachauri@yahoo.com

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Figure 1: Chest X-ray s/o cardiomegaly with the right atrial dilatation

1st year of life [1]. Majority of CHD in children are acyanotic with ASD being the second most common CHDs [1,4,5].

The age of presentation of ASD is usually more than 1 year [4-6] and there is female preponderance [7]. Infants with ASDs are usually asymptomatic. In our case a female child presented at 2 months of age with CHF. The most common murmur heard in ASD is ejection systolic murmur due to increased flow across the pulmonary valve [7]. In our case, there was Grade 2 ejection systolic murmur with wide and fixed S2. We managed CHF conservatively but failed, so referred for surgical correction but she expired due to superimposed bacterial infection.

There are reports showing ASD as very severe when associated with genetic syndromes. Wu *et al.* presented a case of 10 mm ASD in an 8 month-old male infant with congenital disorder of glycosylation [8]. In our patient, there was no dysmorphism or features suggestive of any inborn error of metabolism. Lammers *et al.* stated that closure of an ASD should not be postponed to a later age as surgical correction has very good outcome [9]. In our case, child had pneumonia with pulmonary arterial hypertension with ASD. She might have been saved if she could be referred early for surgery but could not be done due to infection.

CONCLUSION

There have been few cases of ASD presenting in 1st year of life and even fewer below 6 months of age. This case we are reporting is a large OS ASD with RAD with RVE with CHF which did not improve on conservative management and required early surgical intervention. Early detection was possible as our hospital is a tertiary care referral hospital in Northern India. However, surgical correction could not be done even in large tertiary institutes in our country.

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