

# Atrial Septal Defect (Secundum) : A Case Report of a Family

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### Abstract :

*Atrial Septal Defect (ASD) Secundum type was detected in a female patient Miss S.A at 20 years of age. She was operated from Birla Heart Research Center, Kolkata India in 1995. She got married after two years. Her son Master Z.S was also diagnosed as ASD secundum and was operated from Narayana Hrudayalaya Institute of Cardiac Science, Bangalore, India in the year 2005 when he was 8 years old. Her daughter was born in the same year and was diagnosed as a case of large ASD secundum (almost common atrium). This girl is now waiting for device closure from Combined Military Hospital (CMH) Dhaka, but her weight is still low for such procedure. This family is a rare family where mother and both children have ASD secundum and possibility of genetic inheritance was strongly suspected which lead to the writing of this report.*

*(Cardiovasc. j. 2009; 1(2) : 226-228)*

### Introduction:

Secundum Atrial Septal Defect (ASD) is one of the most common defects occurring as an isolated lesion. It represents about 6 to 10% of all cardiac anomalies encountered and is more common in females than in males. <sup>1, 2, 3, 4</sup> Most cases of ASD occur sporadically, however a few families have the defect as a part of genetic abnormality. <sup>5, 6</sup> Holt and Oram noted association between ASD and certain skeletal abnormalities of the forearms. <sup>7</sup> The lack of early symptoms and the subtlety of the physical findings tend to delay the diagnosis sometimes until well into adult life or even until middle or old age. Spontaneous closure of ASD is rare after one year of age, whereas a small ASD may remain completely asymptomatic and haemodynamically insignificant throughout life. The natural course of ASD except for large one is relatively benign. <sup>8</sup> Late symptoms are atrial fibrillation, congestive heart failure and less often those of pulmonary vascular disease. For major ASD's, elective surgical or catheter closure is the treatment of choice. In this family report, all three members had large ASD's, mother and son had surgical closure of ASD and daughter is waiting for device closure.

### Case Report:

Miss S.A was diagnosed as a case of Atrial Septal Defect (ASD) in the year 1995 when she was 20 years old. She had history of palpitation, shortness of breath for 5 months for which she was investigated. On examination splitting of 2<sup>nd</sup> heart sound and ejection systolic murmur was noticed. Her chest X-ray was normal, ECG showed right bundle branch block. Echocardiography with color flow showed presence of a large ASD secundum of 25 mm size with pulmonary artery pressure of 40 mm Hg. She had pericardial patch closure of ASD on 9<sup>th</sup> May 1995 from Birla Heart Research Centre



**Fig-1:** Photograph of mother and two children with large Atrial Septal Defect.

of Kolkata, India. She is doing very well since then. She got married after two years of ASD closure. Her son was quite well upto eight years of his age. Then he also developed shortness of breath, easy fatigability and chest pain. He was investigated for that like his mother. All investigations except color doppler echocardiogram was normal. His echocardiography showed presence of a large ASD which was 18 mm in size with left to right shunt and pulmonary artery pressure was normal. He had pericardial patch closure of ASD on 21<sup>st</sup> July 2005, 10 years after her mother's surgery. He was operated from Narayana Hrudalaya Institute of Cardiac Science of Bangalore, India. He is doing well and leading a normal life now. Daughter of S.A was born just 3 days before the surgery of her son on 18<sup>th</sup> July 2005 in Combined Military Hospital (CMH) Dhaka. As S.A was very worried about her son and was eager to know about her daughter's heart condition, baby was taken into the Echo laboratory of CMH Dhaka on 2<sup>nd</sup> day of delivery. The baby was diagnosed as a case of Large ASD secundum (almost common atrium). She also had moderate degree of pulmonary hypertension. Calculated pulmonary artery pressure was 50 mm Hg. She was treated with digoxin and captopril. Her ASD size was getting smaller in subsequent echocardiographic examination. She is now waiting for device closure. Her weight is 9 kg now. Her device closure will be performed once she achieves 12-kilogram body weight.

### Discussion:

Defects at the level of fossa ovalis results from deficiency, perforation or absence of the septum primum. There is no known intrauterine events that predispose to a deficiency of atrial septation, and most cases occur sporadically with no family history of congenital heart disease. There is a significant familial incidence in some cases of skeletal abnormalities. A few families has the defect as a genetic abnormality. Autosomal dominant pattern of inheritance is also described.<sup>9-11</sup> Spontaneous closure of ventricular septal defect during infancy is well documented, on the other hand reported instances of spontaneous closure of an Atrial Septal Defect (ASD) are rare especially after one year of age. But Spontaneous closure of ASD after infancy is also reported.<sup>12</sup> The diagnosis of isolated ASD in infancy is uncommon. Twenty six patients were studied in United States; spontaneous closure was documented in 39% of the patients at ages ranging from 2 to 8 years.<sup>13</sup>

Some of the infants become symptomatic in infancy.<sup>14</sup> Congestive cardiac failure, recurrent respiratory tract infection and failure to thrive are important symptoms. Only the daughter of S.A had symptoms like failure to thrive and recurrent chest infection in early infancy. S.A and her son had no symptoms in early infancy. Predictive factors for spontaneous closure were not present in S.A and her children.<sup>15, 16, 17</sup> An ASD with a diameter of 8 mm or more has little chance of closing spontaneously. An ASD is considered haemodynamically insignificant if after the age of one, it demonstrates the following conditions: (1) on echocardiography septal defect is < 6 mm. (2) Systolic pulmonary artery pressure < 30 mm Hg by cardiac catheterization. (3) QP/QS < 1.5:1. In this family report all three patients has haemodynamically significant ASD'S with QP: QS > 1.5:1. For major ASD's elective surgical repair or device closure is the treatment of choice. A major defect is defined as one having a QP/QS ratio >1.5:1. These lesions are usually well-tolerated and elective repair or device closure is usually performed at 4 to 5 years of age. Delaying repair beyond this age is harmful. Long standing volume overload of the right heart causes certain irreversible changes in right atrium and ventricles, arrhythmias and premature death. Patient S.A and her son is lucky that they had no complications before or after surgery. Her daughter was diagnosed in neonatal period and now she is waiting for device closure. Diagnosis of ASD in the mother S.A in 2nd decade of her life, son in 1st decade and daughter in the newborn period also indicates the gradual improvement of services provided to the patient with congenital heart diseases in our country.

### Conclusion:

Elective repair for ASD is safe and simple operation in experienced hand. Trans catheter closure of ASD,s have been available for more than two decades. In Bangladesh it is available since 2001 in Combined Military Hospital, Dhaka. It is a safe procedure in experienced hand. So early detection of the cases and appropriate treatment in time can avoid complications like congestive cardiac failure, arrhythmias and pulmonary vascular disease. An individual born with ASD and treated finely have a normal life expectancy and remain free from related cardiovascular morbidity.

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