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**LEFT VENTRICULAR INFLOW OBSTRUCTION BY GIANT ATRIAL SEPTAL ANEURYSM IN A NEONATE WITH HYPOPLASTIC RIGHT HEART SYNDROME: CASE REPORT**

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**SUMMARY**

**Atrial septal aneurysm remains a rare congenital cardiac malformation. In the neonatal age group it can occur as an isolated cardiac malformation or in association with complex hypoplastic cardiac malformations of the right and left heart. In the adult population most aneurysms have been described in association with stroke.**

**Baby H.N delivered on 10/05/2008 by C/S, was cyanosed at birth with systemic desaturation. Chest X-ray showed oligoemic lung fields while two dimensional echocardiograms showed tricuspid atresia with hypoplastic right ventricle, large secundum atrial septal defect, and highly mobile gigantic aneurysms of the atrial septum obstructing the inflow of the mitral valve and entering the left ventricle in diastole. Surgical intervention was not possible and child died on second day.**

**CASE REPORT**

Baby H.N male neonate was delivered in a Mission Hospital by caesarean section due to foetal distress to a Para 1+0 mother. He had low apgar scores of 6/1, 8/5, and was noted to be in respiratory distress after delivery. He became progressively cyanosed and was then suspected to congenital heart disease; a cardiovascular evaluation was then requested.

Physical examinations showed a male infant in obvious respiratory distress with central and peripheral cyanosis, saturating at 60% off oxygen and 88% on oxygen. The birth weight was 3 kgs. The pulses were regular and of low volume. The second heart sound was split, with a continuous murmur along the left upper sternal border. The liver was palpable but the spleen was not palpable. There were no obvious external malformations or dysmorphic features.

Chest X-ray showed normal cardiothoracic ratio but decreased pulmonary vascular marking. Two dimensional echocardiogram showed normal situs solitus with levocardia. The systemic and pulmonary venous returns were normal. There was a large secundum atrial septal defect with redundant membrane in the right and left atrium,

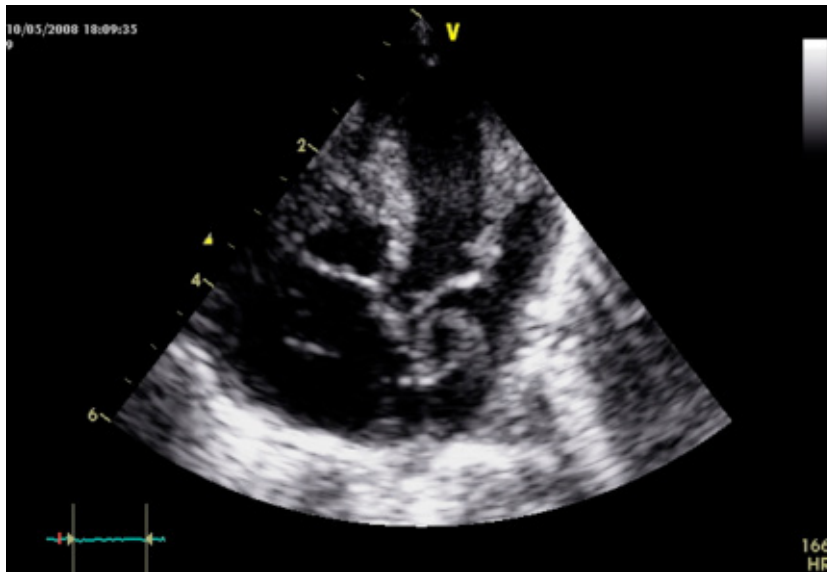
but predominantly filling the left atrium (Figure 1). During the cardiac cycle it occupied the whole of the left atrium before crowding at the orifice of the mitral valve, obstructing the inflow of the left ventricle in early diastole (Figure 2). In mid diastole the gigantic membranes caused turbulent flow as it stretched the mitral leaflets and the subvalvular apparatus. In the latter phase of diastole these membranes could be seen past the mitral leaflets into the mid left ventricular cavity (Figure 3 and 4)

The left atrial size was increased with significant turbulent flow into the left ventricle; there was a gradient of 13mmhg across the mitral valve on continuous wave doppler in keeping with mitral stenosis despite the low output state.

There was a small bulbo-ventricular foramen from which an atretic pulmonary valve arose. The pulmonary trunk was present with confluent good size but hypoplastic branches. There was a small patent ductus arteriosus of 0.2cm in size with high velocity continuous flow (Figure 5). The aortic valve was normal with no evidence of systemic outflow tract obstruction, normal coronary arteries and normal left sided aortic arch. The left ventricle was of normal size with good contractility and there was no pericardial effusion.

**Figure 1**

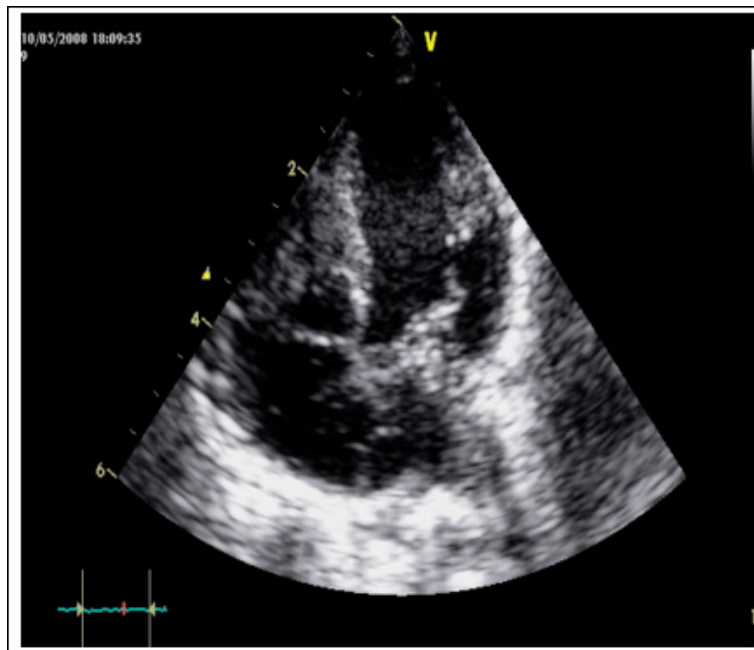
*The arrow shows a giant atrial septal aneurysm rolled in the left atrium.*



The picture above (systolic) showing atrial septal aneurysm in the left atrium associated with a large secundum atrial septal defect. The right ventricle is small while the left ventricle is of normal size.

**Figure 2**

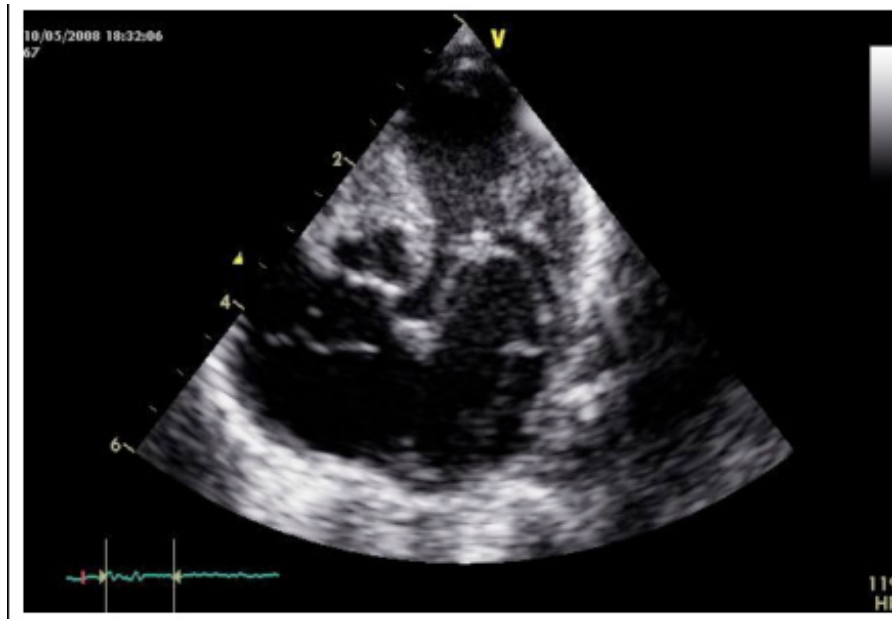
*The arrow shows the aneurysm completely obstructing the left ventricular inflow (mitral valve-MV)*



Above frame in early diastole, the aneurysm at the supramitral region virtually obstructing the orifice of the mitral valve

**Figure 3**

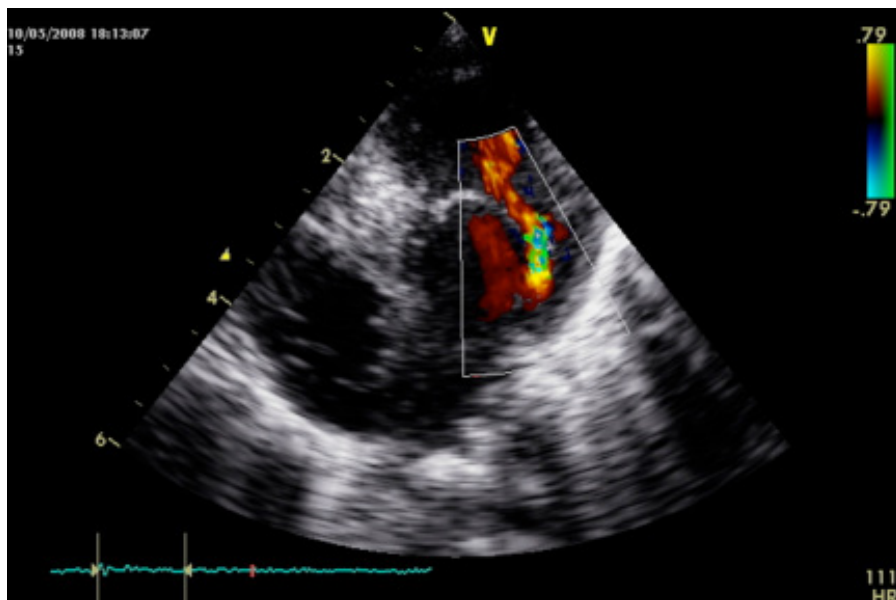
Arrow showing Aneurysms stretching into the left ventricular cavity and a large secundum atrial septal defect



In the above mid diastolic frame (Figure3) the aneurysm has stretched itself into the mitral valve pushing its chordal attachments into the left ventricular cavity. The margins of the atrial septum are distinct and the atrial septal defect is large. Parts of the aneurysmal tissue remained attached into the superior margin of the atrial septal defect while the other parts are in the left atrium

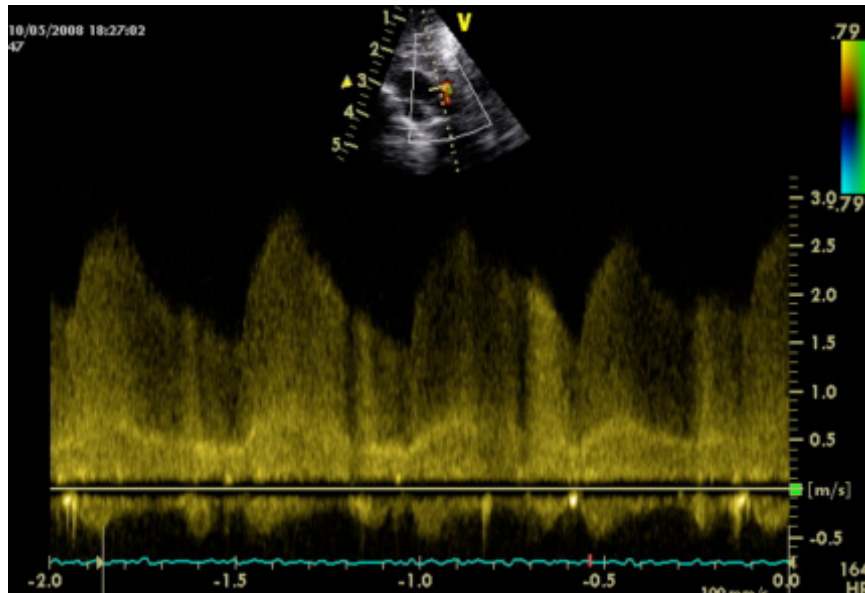
**Figure 4**

Colour flow Doppler showing turbulence as the mitral valve apparatus are stretched.



**Figure 5**

*Parasternal Short axis view across the pulmonary trunk, spectral Doppler shows continuous flow in keeping with ductus.*



The baby remained in sinus tachycardia and low output. The parents were counselled about the baby's condition and transfer to the tertiary referral hospital for further management was recommended but they declined further intervention. The baby passed away on the second day. A post-mortem examination was not performed.

## DISCUSSIONS

Majority of atrial septal aneurysms are described in the adult population where they are defined echocardiographically as transient bulging of the fossa ovalis region of the intra atrial septum greater than 15mm in the absence of chronically elevated atrial pressures. Most of these lesions are small, have limited motions within the plane of the atrial septum and are commonly associated with stroke (1). Similar smaller aneurysms have also been described in the neonatal age group as relatively benign (2).

Gigantic atrial septal aneurysm in the neonate however have been described in patients with complex cardiac malformations the left and right heart like tricuspid atresia, pulmonary atresia intact septum, hypoplastic left heart syndrome and hypoplastic right heart syndrome (3). When giant atrial septal

aneurysm are associated with hypoplastic left or right heart syndrome the intra-atrial communications have been described as restrictive and associated with premature closure of the foramen ovale (4). Our baby had complex malformations of the right heart which included tricuspid atresia with a very small right ventricular chamber, a small bulbo-ventricular foramen, absent pulmonary infundibulum and an atretic pulmonary valve associated with a small ductus. The secundum atrial septal defect was quite large unlike previous reports with significant right to left shunting. The aneurysm had attachments to both edges of the atrial septal defect. During the cardiac cycle as blood flowed from the right to left, the aneurysms also moved into the left atrium and in early diastole formed a tumour like mass at the orifice of the mitral valve, while in mid diastole it stretched and prolapsed into the mitral valve causing obstruction, turbulent flow and a significant gradient. Keeping its attachments the aneurysms were seen stretching the Mitral valve attachments to occupy the mid ventricular cavity. Previously described echocardiograms have documented giant atrial septal aneurysm in the right and left atrium that mimicked a right atrial mass (3, 4,) we hereby describe these aneurysms causing significant left ventricular inflow obstruction. Hypoplastic right heart is a ductal dependant lesion which if left untreated is a uniformly

fatal lesion. We believe that among other factors, this obstruction further worsened the already precarious clinical state contributing to the rapid downhill course with death on the second day of life.

Appreciations- Parents of baby H.N, Dr Peter Messner who worked in the newborn unit Kijabe Hospital then, Management and Staff Newborn Unit Kijabe Mission Hospital.

#### REFERENCES

1. Gallet B, Malergue MC, Adams C, Saudemont J.P, *et al.* Atrial septal aneurysm- a potential cause of systemic embolism. *Br. Heart J.* 1985; **53**: 292-7.
2. Ozcelik N, Atalay S. Tutar E, Ekiei F. Prevalence of Intratrial septal aneurysm in Newborns and their Natural Course. *Pediatr. Cardiol.* 2006; **27**: 343-346.
3. DJ Sahn, HD Allen, R. Anderson and SJ Goldberg. Echocardiographic diagnosis of atrial septal aneurysm in an infant with hypoplastic Right heart Syndrome. *Chest* 1978; **73**: 227-230 .
4. Chrysostomou C. Romaguera R. Rodrigueuz M . Giant Aneurysm of the atrial septum associated with premature closure of foramen ovale. *Cardiovascular Ultrasound* 2005; **3**:20