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Isolated subpulmonary membrane causing critical neonatal pulmonary stenosis with concordant atrioventricular and ventriculoarterial connections

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Abstract

We report a rare case of isolated subpulmonary membrane leading to critical pulmonary stenosis in an infant. This anomaly needs to be differentiated from valvar pulmonary stenosis as both anomalies require different modality of treatment (surgical/catheter).
MeSH: Neonates, Pulmonary Subvalvular Stenosis, Pulmonary Valve Stenosis

Case

A 22 day old male newborn, product of normal vaginal delivery with birth weight of 3.5 kg after uneventful antenatal and natal period, presented with respiratory distress and feeding difficulty from 2nd week of life. There was no history of pneumonia, sepsis, recurrent vomiting or history of aspiration.
Anthropometric examination was within normal limit. He had tachycardia, tachypnea and uniform central cyanosis in room air (oxygen saturation 83%). Precordial examination revealed no cardiomegaly, normal 1st heart sound, soft and delayed P2, and a grade 3/6 harsh ejection systolic murmur at the left upper sternal border. Perabdominal examination revealed hepatomegaly (liver 3 cm below costal margin).

Electrocardiogram showed heart rate 140/minute, sinus rhythm, right axis deviation (QRS axis +110°) and right ventricular hypertrophy. Chest X ray revealed no cardiomegaly, oligemic lung fields and no parenchymal lesion.

Echocardiography done with Hewlett – Packard Sonos 7500 machine with broad band (8 Hz and 12 Hz) transducers showed situs solitus, levocardia with atrioventricular and ventriculoarterial concordances, left aortic arch with normal branching,and no patent ductus arteriosus. An isolated, discrete, obstructive subpulmonary membrane was seen intimately related to the pulmonary valve in apical five chambered view with anterior angulation, parasternal short axis view at the level of great arteries (figures 1 and 2) and modified parasternal long axis view with anterior tilt.

Figure 1 Two dimensional echocardiography in parasternal short axis view at level of great vessels showing a subpulmonary membrane with restrictive central aperture situated in close proximity to pulmonary valve. (Arrow 1 – Subpulmonary membrane, Arrow 2 - Pulmonary valve)

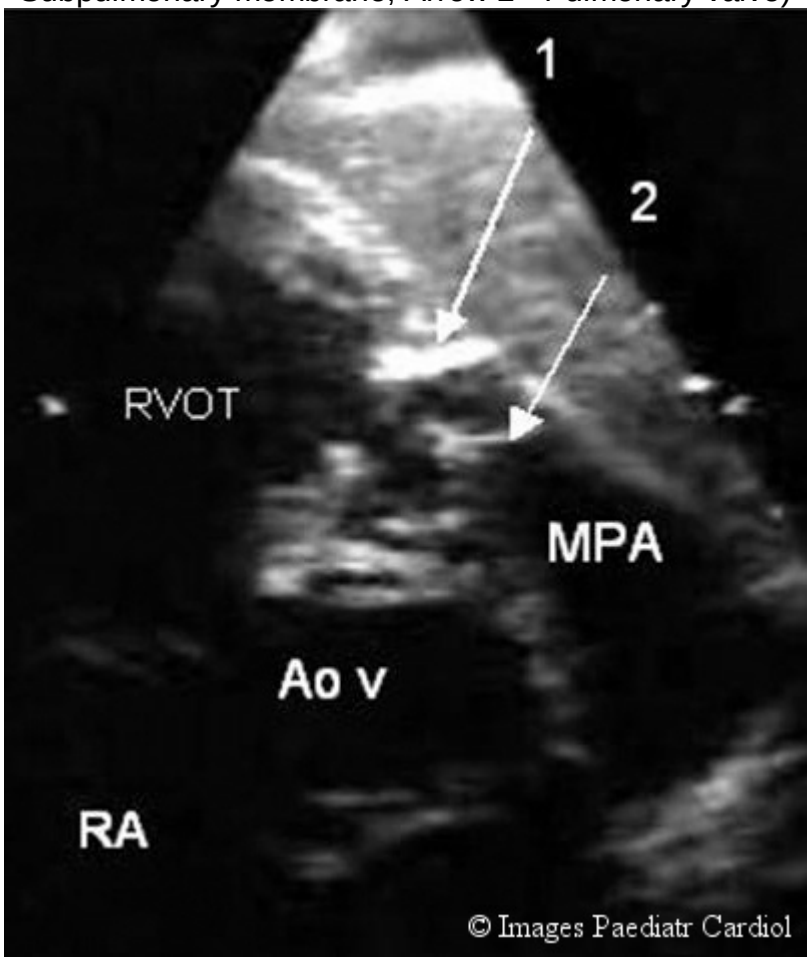
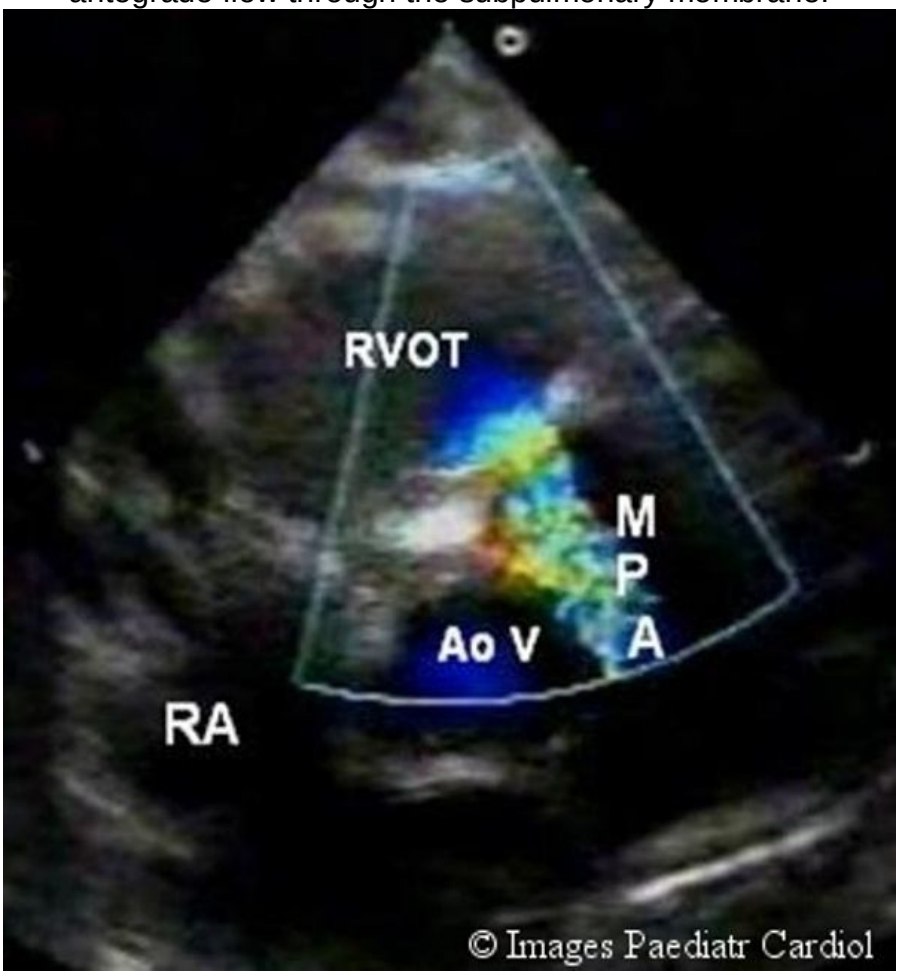
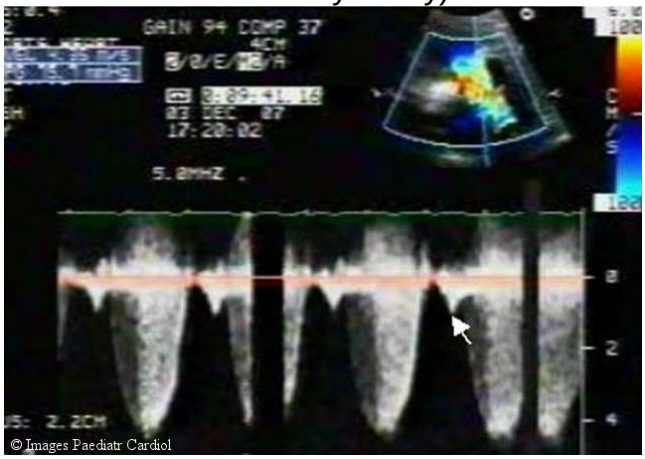


Figure 2 Color flow mapping in same view showing restricted and turbulent antegrade flow through the subpulmonary membrane.



This membrane had a restrictive central aperture causing severe right ventricle outflow tract (RVOT) obstruction (peak gradient = 76 mm Hg) (figure 3).

Figure 3 Continuous wave Doppler echocardiography with cursor placed in RVOT showing severe pulmonary stenosis (peak RVOT PG = 75.7 mm Hg). Arrow marks pulmonary artery 'a' wave indicating RV diastolic dysfunction. (Ao V – Aortic valve, RVOT – Right ventricular outflow tract , MPA – main Pulmonary artery)



The pulmonary valve annulus was hypoplastic (6 mm, Z score minus 2). The pulmonary valve leaflets were secondarily thickened by this stenotic stream but there was no doming of the leaflets. There was no poststenotic dilatation of the main pulmonary artery (MPA) which is a feature of valvar pulmonary stenosis (PS). Biventricular systolic function was preserved. There was a patent foramen ovale (PFO) shunting right to left.

The child underwent surgery under cardiopulmonary bypass and the subpulmonary membrane was resected. However as the membrane was extremely close to the pulmonary valve, the anterior leaflet of pulmonary valve was damaged. It was also resected and short transannular glutaraldehyde treated pericardial patch was placed.

He was hemodynamically stable in the immediate postoperative period. Echocardiography showed laminar flow in the right ventricular outflow tract, normal ventricular function and the PFO shunt left to right. However he developed acute respiratory distress syndrome and succumbed on 14th postoperative day in spite of all resuscitative measures.

Discussion

Although fibrous subaortic stenosis is a well known entity associated with 8 - 20% of left ventricular outflow tract obstructions, to best of our knowledge, the association of subpulmonary membrane with atrioventricular and ventriculoarterial concordances with intact ventricular septum has not yet been described.

On echocardiography, it is important to differentiate this anomaly from valvar PS with intact ventricular septum for the management plan. For valvar PS, balloon pulmonary valvotomy is the treatment of choice with excellent results while for subvalvar PS, surgery is the only option.

Pulmonary stenosis in the newborn is predominantly valvar and sometimes infundibular due to localized muscular hypertrophy. A subpulmonary membrane or ridge on the other hand has been reported in association with transposition of great arteries (complete transposition of great arteries, congenitally corrected transposition) and isolated case reports of doubly committed ventricular septal defect with association of both subaortic and subpulmonary fibrous ridges.[1,2]

Email correspondences with Prof. Richard Van Praagh and Prof. Robert Anderson provided an aid to understand this anomaly .According to Van Praagh, it is an developmental anomaly. Early in embryonic development, the conus is often said to have proximal, middle, and distal endocardial cushions that are forming the conal septum and the semilunar valve (pulmonary or aortic). At the conal cushions stage, there is no obvious difference between what will become the muscular conal septum versus what will become the semilunar valve. But as development proceeds, the conal septal cushions fuse and become muscularised, whereas the semilunar valvar cushions fuse, separating the aortic from the pulmonary valve, but these semilunar valves remain fibrous. They do not become muscularised.

Fibrous subaortic or subpulmonary stenosis is in direct fibrous continuity with the overlying semilunar valve, indicating that the subvalvar ring-like stenosis

really is valvar, not infundibular or conal. If the obstruction becomes muscular, then most probably the origin is infundibular and not valvar.

Anderson thinks that this is not a “membrane” and appears to look more like a fibrous shelf. This is almost certainly an acquired lesion, and is a fibrous accretion exacerbating the narrowing at the mouth of the subpulmonary infundibulum. Anderson and colleagues have illustrated such fibrous accretions in their studies on tetralogy of Fallot, albeit not specifically commenting on the structure.[3]

To conclude, we report this rare case of isolated subpulmonary membrane with intact ventricular septum as a rare cause of RVOT obstruction. Transthoracic echocardiography especially in young infants was helpful to delineate the lesion and differentiate it from the usual valvar pulmonic stenosis and guided us to formulate the management plan. We believe that a careful echocardiography of all similar presentations would enable us to learn more of this rare lesion.

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