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AORPA: A Rare Congenital Heart Disease

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1. Introduction

The anomalous origin of the pulmonary artery from the ascending aorta (AOPA) is also referred as ''hemitruncus'' and accounts for 0.1% of all congenital heart diseases [(4).] Anomalous origin of pulmonary artery (AOPA) from the ascending aorta is an unusual and critical cardiovascular anomaly, which frequently involves the right pulmonary artery (RPA). Hemitruncus occurs alone in roughly 60% of cases of right pulmonary artery abnormality, which is 6 times more frequent than left [(5).] The most common related condition is patent ductus arteriosus, however tetralogy of Fallot, an atrial septal defect, a ventricular septal defect, and coarctation of the aorta have also been described. The known underlying aetiology of this anomaly is a partial or total developmental failure of the left sixth arch, despite the fact that the pathophysiology of this anomaly is poorly understood. As a result, there is a significant left - to - right shunt, which causes pressure and/or volume overload in both lungs. One lung receives the whole cardiac output from the right ventricle, while the other receives blood from the aorta at a systemic pressure [(8).]

2. Case Report

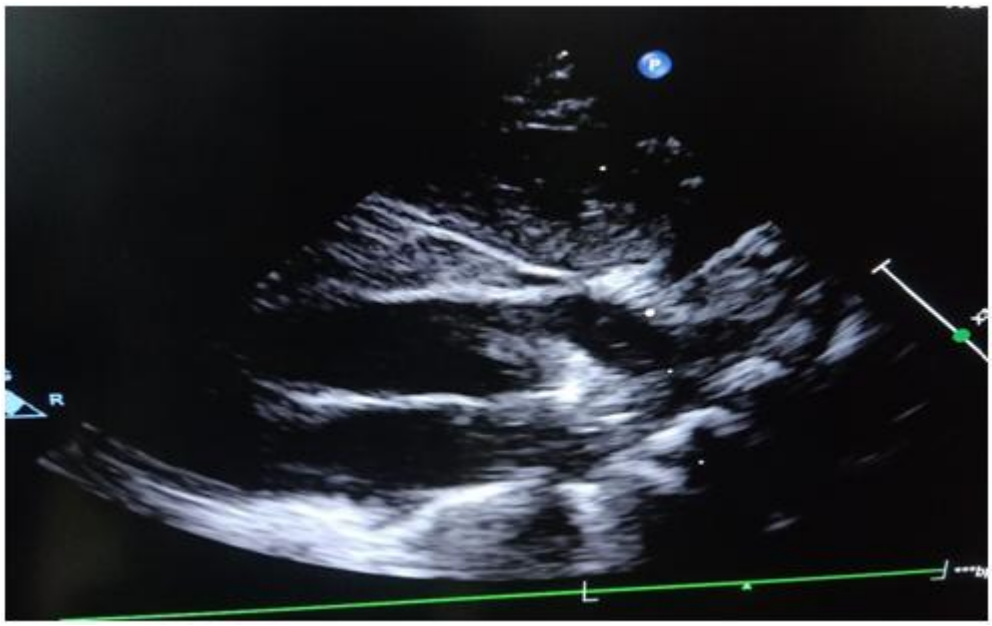
4 days old late preterm female baby of birth weight 2.5kg born to G3P2L2mother through LSCS (previous LSCS). Baby cried at birth and was noted to have respiratory distress since birth. As baby was having persistent desaturation and cyanosis baby was referred to our centre.

On admission, HR - 156, RR64, PVnormal, CRT<2sec, Spo2 in room air - 84%Rt UL, 80%Rt LL, Spo2 with 2L O2 - 95%UL, 94%LL. Baby with B/L air entry (+) with no added sounds; S1, S2 heard and no murmurs; AF (+), nobulge, moderate activity, tone normal in all limbs, B/L pupil NSRL. Baby was started on O2 inhalation with nasal prongs with 2L O2, iv fluids, Sildenafil 1mg/kg/dose tid (in view of suspecting PPHN). Baby sepsis screen - negative, blood culture - negative, 2D ECHO - Right pulmonary artery from ascending aorta (12 mm from annulus), PFO with bidirectional shunt, mild PAH, intact IVS, no PDA. On pediatric cardiology referral, baby was planned for AORPA repair (right pulmonary artery translocation).

3. Discussion

The clinical manifestations of the condition usually occur in the infant or, more rarely, in the newborn including respiratory distress or congestive heart failure due to increased pulmonary resistance. The lung connected to the abnormally arising PA is exposed to both pressure and volume overload due to unrestricted shunting from the aorta. The vasoconstrictive reaction due to the abnormal state of the vascular bed on the abnormally connected lung is responsible for the development of pulmonary hypertension [(8).]

This is the first example of anomalous right pulmonary artery origin from the ascending aorta, noted in our institutuion. This case will serve as a teaching tool so that similar cases in the future will be swiftly diagnosed and aggressively treated. Despite its rarity, infants with congenital heart disease symptoms should be checked for hemitruncus arteriosus after ruling out more frequent disorders such patent ductus arteriosus, tetralogy of Fallot, an atrial septal defect, and a ventricular septal defect.



2D ECHO showing AORPA

4. Conclusion

Anomalous origin of pulmonary artery is dismal but surgically correctable disease. Early diagnosis and timing of operation is important as pulmonary hypertension can start early in life.

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