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Case Report
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Left Sinus of Valsalva to Left Atrial Fistula

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Abstract

A fistula between the left sinus of Valsalva and the left atrium is an extremely rare congenital or acquired cardiac anomaly. This condition involves an abnormal communication between the left sinus of Valsalva, a part of the aortic root, and the left atrium. Such fistulas can result from various etiologies including infectious endocarditis, trauma, or as a complication of surgical or catheter-based cardiac interventions. We present here a neonate presenting with the left atrial fistula.

Keywords: Left Sinus of Valsalva; Left Atrial Fistula; Congenital Cardiac Anomaly; Preterm; Echocardiography; Neonatal Jaundice; Very Low Birth Weight; Patent Ductus Arteriosus

Abbreviations: VLBW: Very Low Birth Weight; LSCS: Lower Segment Cesarean Section; MCDA: Monochorionic Diamniotic; TIFFA: Targeted Imaging for Fetal Anomalies; TR: Tricuspid Regurgitation; PEEP: Positive End-Expiratory Pressure; APGAR: Appearance, Pulse, Grimace, Activity, Respiration; ABG: Arterial Blood Gas; CRT: Capillary Refill Time; CPAP: Continuous Positive Airway Pressure; PFO: Patent Foramen Ovale; PDA: Patent Ductus Arteriosus; LVEF: Left Ventricular Ejection Fraction; NPO: Nothing by Mouth; TEE: Transoesophageal Echocardiography; TTE: Transthoracic Echocardiography

Case

We present a case of a preterm male baby, born at 34+1 weeks, delivered by LSCS due to abnormal Dopplers in both twins. The infant was twin 1 in MCDA twins, with a very low birth weight (VLBW) of 1.44 kg. The mother, a primigravida, had a history of polyhydramnios. A TIFFA scan showed cardiomegaly, right ventricular hypertrophy, and a TR jet across the septal leaflet, along with cardiac contractility dysfunction. There was a 22% discordance between the twins. The mother's blood group was B positive, and she received antenatal steroid coverage. The baby cried immediately after birth but required positive pressure ventilation for secondary apnea and bradycardia, developing respiratory distress soon after birth. The Silverman Anderson score was 3, and delivery room CPAP was administered with a maximum FiO2 requirement of 30% at 5 PEEPS. The APGAR scores were 5, 7, and 8 at 1, 5, and 10 minutes, respectively. Cord ABG showed pH 7.358, PCO2 36.9 mmHg, lactate 2.64 mmol/L, BE -4.6 mmol/L, and HCO3 20.3 mmol/L [1].

The baby was admitted to the NICU for the management of respiratory distress and prematurity. Vital parameters monitored were heart rate 160/min, respiratory rate 74/min, capillary refill time (CRT) < 3 seconds, and SpO2 95% with 21% FiO2. On respiratory system examination, equal air entry was noted, along with subcostal retraction and tachypnea. No murmur was auscultated on the first day of life. Heart sounds S1 and S2 were normal, and no hepatosplenomegaly was observed. The baby exhibited good activity levels, with an open anterior fontanel at level. In the NICU, oxygen support was initiated with bubble CPAP. A chest X-ray revealed an increase in pulmonary vascular markings and cardiomegaly (Figure 1). A diagnosis of transient tachypnea of the newborn was made and managed with bubble CPAP

support. A 2D echocardiogram scan was performed on the first day of life, revealing situs solitus, levocardia, normal pulmonary venous drainage, a dilated right atrium (RA) and left atrium (LA), mild tricuspid regurgitation (gradient 15 mmHg), a dilated right ventricle (RV) and left ventricle (LV) with mild RV dysfunction, a normal great arterial relationship, a patent foramen ovale (PFO) with a left-to-right shunt, and an intact interventricular septum which was hypertrophied (4x5 mm). The aortic valve had three leaflets, the ascending aorta measured 5.9 mm, with a left arch and no coarctation. The patent ductus arteriosus (PDA) measured 2.2 mm with a left-to-right shunt and a gradient of 20 mmHg. The left ventricular ejection fraction was 65%. In light of persistent respiratory distress, a repeat chest X-ray was performed on the third day of life, revealing persistent cardiomegaly (Figure 2)."

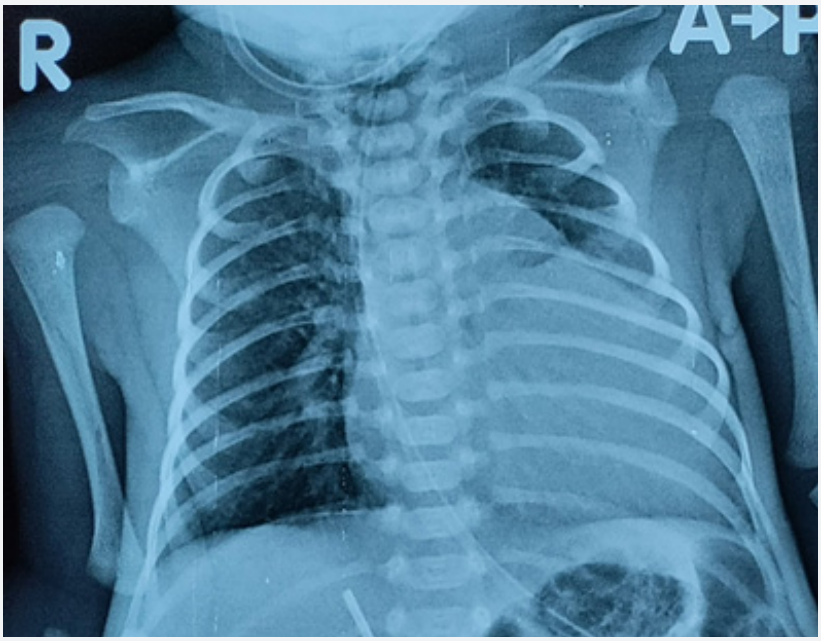

Figure 1: Xray shows increase pulmonary vascular markings.

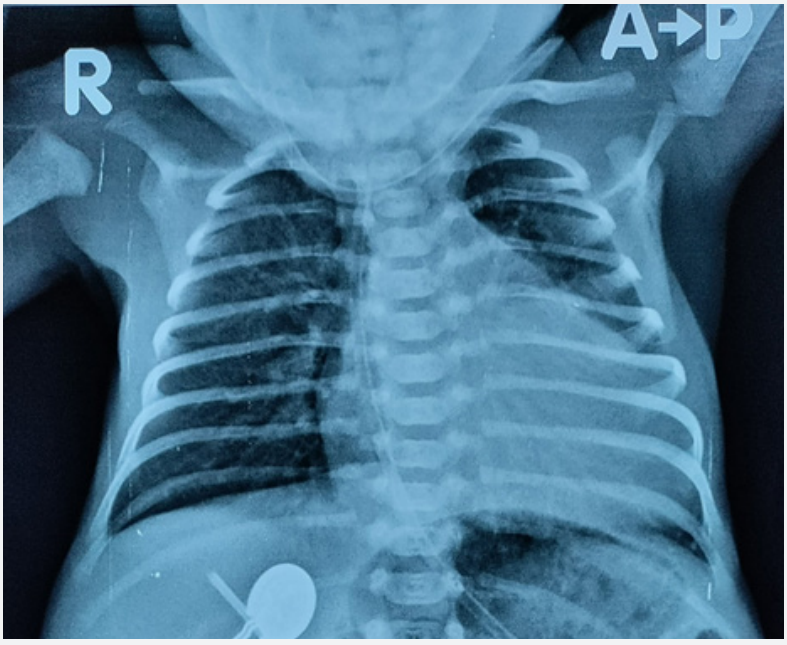

Figure 2: Cardiomegaly with persistent alveolar marking.

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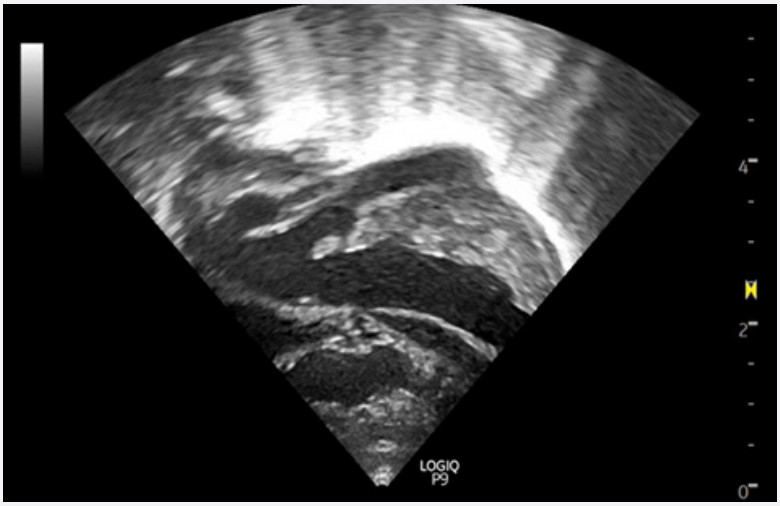

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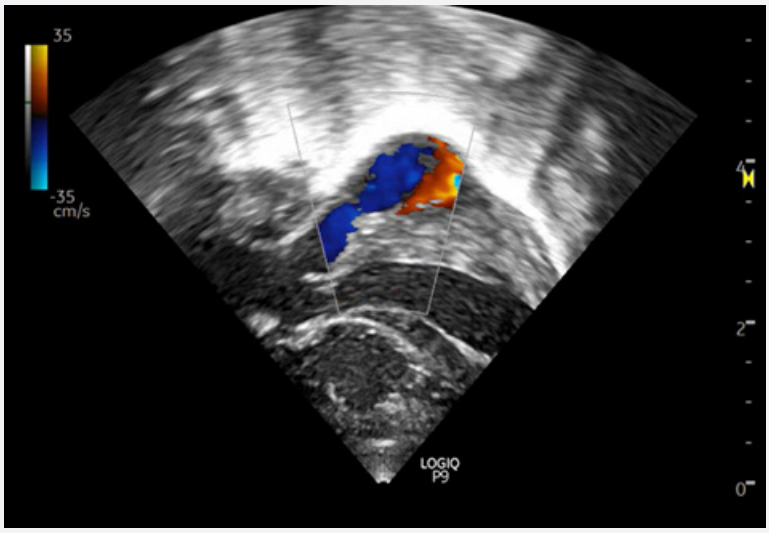

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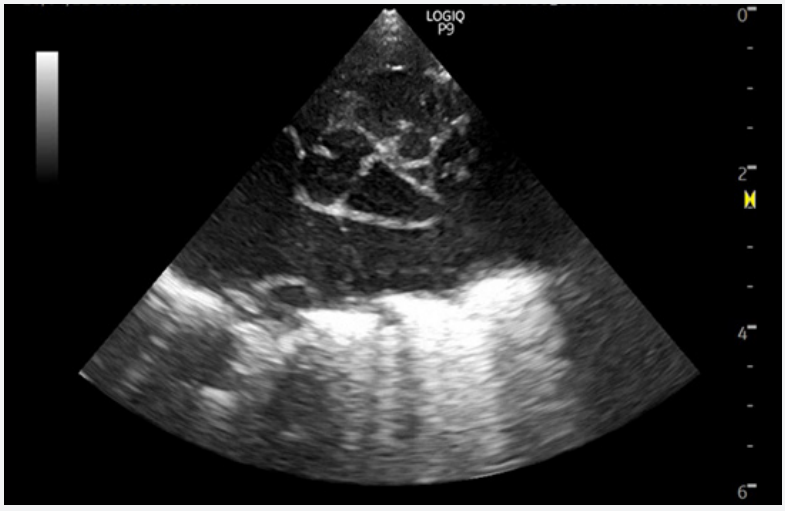

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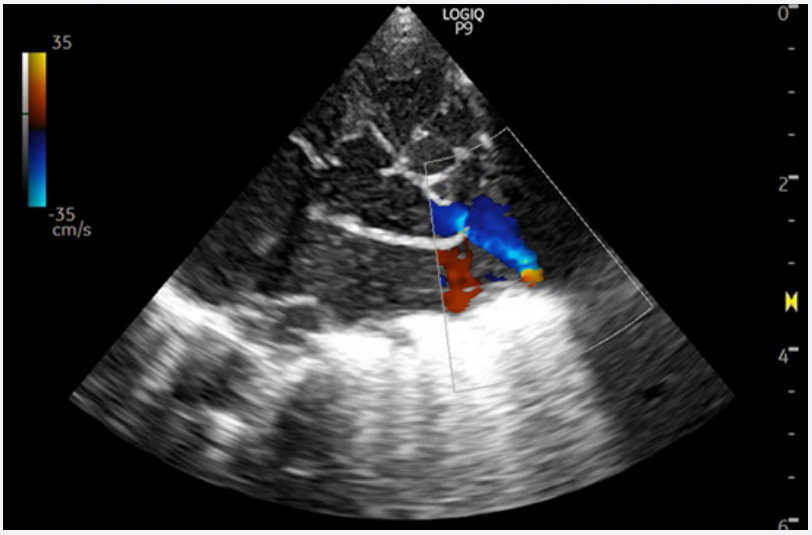

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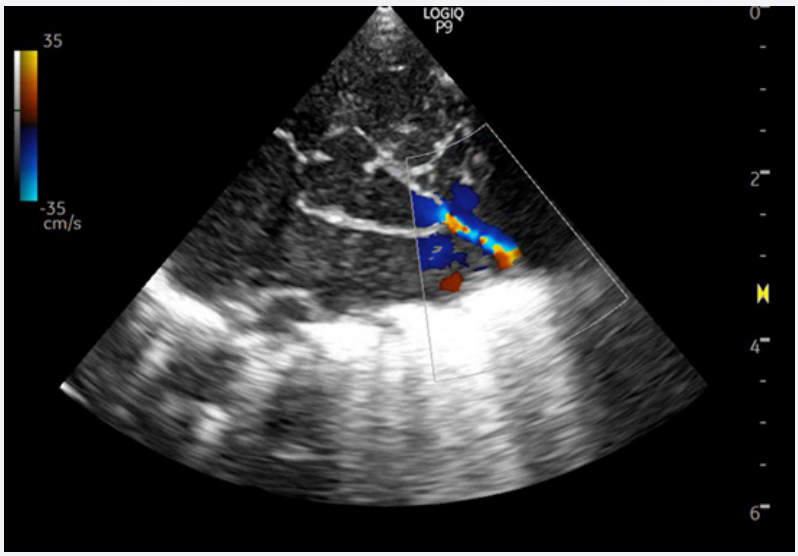

(E)

Figure 3: Echocardiography findings left sinus of Valsalva to left atrial fistula.

On day 6 of life, due to persistent respiratory distress, a repeat 2D echocardiogram was performed. The findings included a patent foramen ovale with a left-to-right shunt, a small patent ductus arteriosus with a left-to-right shunt, a congenital left sinus of Valsalva to left atrial fistula with a pressure gradient of 60 mmHg, a dilated left atrium and left ventricle, good biventricular function, and a left arch of the aorta with no coarctation (Figure 3). Following the initiation of hydrochlorothiazide, respiratory distress improved, and oxygen support was discontinued. On day 10 of life, a repeat 2D echocardiogram revealed a persistent of congenital left sinus of Valsalva to left atrial fistula measuring 2.8 mm. The left coronary artery was visualized, while the right coronary artery and left anterior descending artery were not visualized. The aortic valve showed thickening of the trileaflet, and there was no evidence of coarctation of the aorta. The aortic arch was left-sided with right descent, and both the left atrium (18 mm) and left ventricle (18 x 10 mm) were dilated. The left ventricular ejection fraction (LVEF) was measured at 58%. An ECG performed which was normal. The baby was kept NPO (nothing by mouth) for 24 hours due to abnormal Doppler findings. Minimal enteral feeding was initiated on Day 2 of life, but the baby experienced feed intolerance manifested as bilious vomiting and abdominal distension, leading to a cessation of oral feeding. Feeding was gradually reintroduced on day 4 of life and

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increased incrementally to full feedings as tolerated. Additionally, the baby developed neonatal jaundice on day 2 of life, which was investigated and treated with phototherapy for 4 days. The maximum total serum bilirubin level reached 15.2. The baby was discharged home at 16 days of life with hydrochlorothiazide and scheduled follow-up with a cardiologist for closure of the left sinus of Valsalva to left atrial fistula [2].

Discussion

A fistula between the left sinus of Valsalva and the left atrium is a relatively rare cardiac anomaly1. It involves an abnormal connection between the aorta's sinus of Valsalva and the left atrium, two crucial structures of the heart. Patients with this condition may present with symptoms such as shortness of breath, chest pain, palpitations, fatigue, and signs of heart failure. However, the clinical presentation can vary depending on the severity of the fistula and associated complications. Diagnosing a left sinus of Valsalva to left atrium fistula typically involves a combination of imaging studies and cardiac catheterization. Echocardiography, including transthoracic echocardiography (TTE) and transoesophageal echocardiography (TEE), is often the initial diagnostic modality of choice. These imaging techniques can provide detailed information about the anatomy of the fistula, its hemodynamic significance, and any associated abnormalities such as aortic valve regurgitation. Cardiac catheterization may be performed to confirm the diagnosis and assess the extent of the fistula. Untreated left sinus of Valsalva to left atrium fistula can lead to various complications, including heart failure, aortic valve regurgitation, arrhythmias, endocarditis, and stroke2.

The abnormal flow of blood from the aorta into the left atrium can cause volume overload on the heart, leading to myocardial dysfunction and heart failure over time. Aortic valve regurgitation may develop due to the retrograde flow of blood from the left atrium into the aorta through the fistula. Arrhythmias can occur due to the disruption of normal cardiac electrical conduction pathways. Furthermore, the abnormal communication between the systemic and pulmonary circulations increases the risk of thromboembolic events such as stroke. Surgical repair is typically the treatment of choice for a left sinus of Valsalva to left atrium fistula. The surgical approach aims to close the abnormal connection between the sinus of Valsalva and the left atrium, thereby restoring normal cardiac anatomy and function3. The specific surgical technique may vary depending on the size and location of the fistula, as well as the presence of associated cardiac abnormalities. In some cases, transcatheter closure techniques may be considered as an alternative to surgical repair, especially for patients deemed high-risk for surgery [3].

Conclusion

In conclusion, a left sinus of Valsalva to left atrium fistula is a rare cardiac anomaly that can lead to significant morbidity and mortality if left untreated. Early diagnosis and appropriate management are essential to prevent complications and improve patient outcomes. Collaboration between cardiologists, cardiac surgeons, and other members of the healthcare team is crucial in providing comprehensive care for patients with this condition.

Declarations:

Consent for publication - Written informed consent was obtained from the parent as patient is minor.

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