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Case Report
Congenital Heart Defects

A Rare Occurrence of Total Anomalous Pulmonary Venous Return in Dichorionic Diamniotic Twins

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Congenital heart defects (CHDs) represent the most common human birth defect, having a birth prevalence of 7-9 per 1000 singleton births. CHDs are more common in twin pregnancies with a prevalence of approximately 20 in 1000 live births. Monochorionic (MC) twins are at even higher risk compared to Dichorionic (DC) twins. Herein, we report a scenario where Dichorionic Diamniotic (DCDA) twins presented with the same type of CHD (TAPVR: Total anomalous pulmonary venous return) in our Neonatal intensive care unit(NICU). Several familial cases of TAPVR have been reported, but no notable pedigree has been reported. Multiple case reports have shown a genetic background of TAPVR. Screening of all siblings with TAPVR is a burden to the patients due to psychological stress or financial problems for the parents. However, these consequences are outweighed by the benefit of diagnosing this critical disease early so that the patient can be taken for surgery before severe congestive heart failure develops. Therefore, if a genetic background is detected in a case of TAPVR, routine fetal echocardiography or cardiography immediately after birth is reasonable to screen TAPVR.

Keywords: Congenital Heart Defects, Total Anomalous Pulmonary Venous Return, Twins

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Introduction

Congenital heart defects (CHDs) represent the most common human birth defect, having a birth prevalence of 7-9 per 1000 singleton births [1,2]. CHDs are more common in twin pregnancies with a prevalence of approximately 20 in 1000 live births. Monochorionic (MC) twins are at even higher risk compared to Dichorionic (DC) twins [2]. Herein, we report a scenario where Dichorionic Diamniotic (DCDA) twins presented with the same type of CHD (TAPVR: Total anomalous pulmonary venous return) in our Neonatal intensive care unit(NICU). TAPVR is a rare, critical congenital vascular anomaly in which the pulmonary veins drain into the right atrium or one of its venous tributaries [3,4]. Initial symptoms may be non-specific and without a cardiac murmur, symptoms resemble those of respiratory distress syndrome in newborns[3]. Several familial cases of TAPVR have been reported, but no notable pedigree has been reported [2]. A larger genetic study was needed to determine the genetic background of TAPVR.

Case Report

A late preterm, female baby born at 36 weeks gestation, by LSCS, twin 1 in case of DCDA twins with a birth weight of 2.175 kgs, with secondary apnea at birth requiring positive pressure ventilation was admitted because of delayed perinatal transition with respiratory distress. She had cyanosis, subcostal retractions and grunting with a Downe's score at admission being 4/10. The Cardiovasular auscultation did not reveal any murmur. She had central cyanosis which continued despite supplemental oxygen. The abdomen was soft.

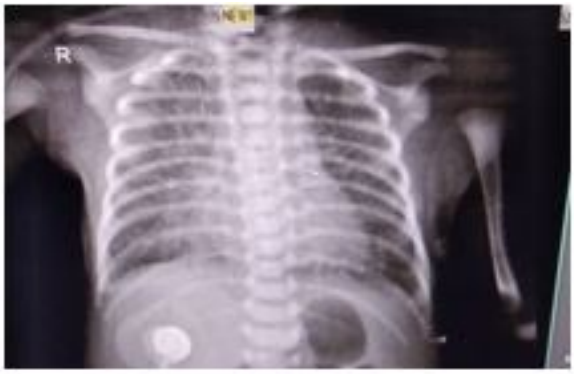

Figure 1: Chest x-ray of twin 1 depicting ground glass opacities and reticulogranular Pattern indistinguishable from Hyaline membrane disease

On CNS examination, she was dull, hypotonic and had depressed reflexes

Because of increasing respiratory distress with low oxygen saturations, she was intubated and connected to conventional mechanical ventilation. As she had high pressures and increasing oxygen requirement on the ventilator, along with the arterial blood gas showing acidosis with hypoxia, with a background chest x-ray suggestive of HMD, one dose of surfactant was given. Despite giving surfactant, she did not show any clinical improvement, with a further drop in saturations even on 100% oxygen, so suspecting severe PPHN she was given a loading dose of Inj. Magnesium sulphate. Because of hypotension with poor perfusion, she was started on inotropic support. Repeat blood gas continued to reveal metabolic acidosis, thus she received sodium bicarbonate infusion.2D Echo done showed Obstructed TAPVC, supracardiac variety. The baby was shifted to an advanced cardiac centre for further management. She successfully underwent TAPVC repair following which she was discharged from the cardiac centre after a post-operative stay of 5 days.

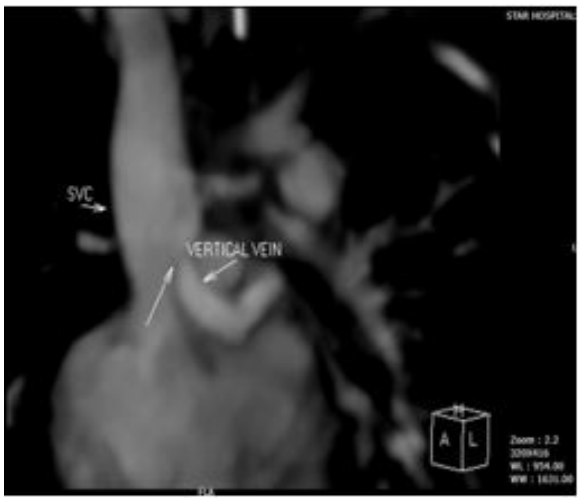

Figure 2: Note the CT angiography image of twin 1 showing the pulmonary veins draining into the superior vena cava(SVC) via the vertical vein revealing the supracardiac variety of TAPVR.

A late preterm, male baby born at 36 weeks gestation, by LSCS, twin 2 of DCDA twins, having a birth weight of 2.040 kgs, with

Secondary apnea at birth requiring bag and tube ventilation was admitted to the Neonatal Intensive care unit (NICU). He continued to require bag and tube ventilation during the period of transport from the birthing centre to the NICU. His cardiovascular auscultation did not reveal any murmur. His chest x-ray showed a reticulogranular pattern with a ground glass pattern suggestive of HMD, hence surfactant was administered.

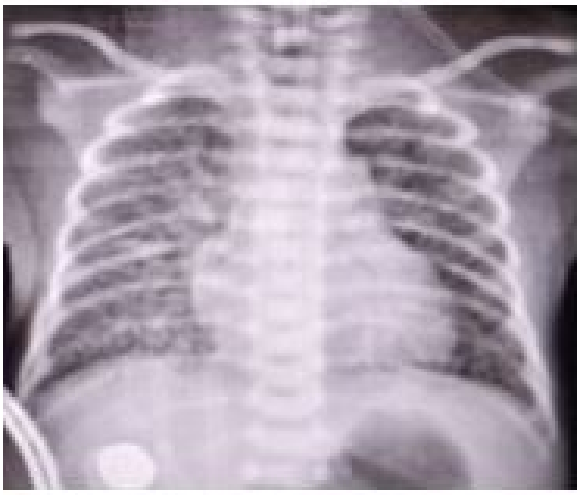

Figure 3: Chest x-ray of the 2nd twin depicting reticulogranular pattern indistinguishable from Hyaline membrane disease

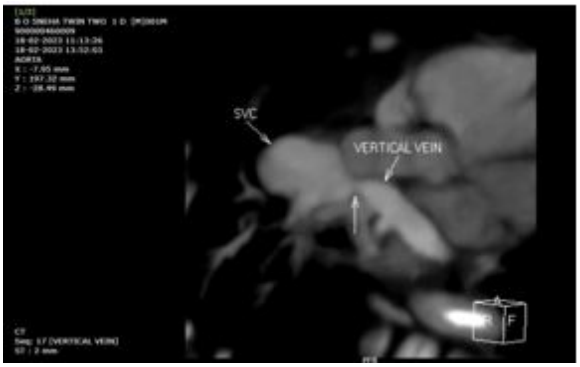

Figure 4: Note the CT angiography image of twin 2 showing the pulmonary veins draining into the SVC via the vertical vein revealing the supracardiac variety of TAPVR

Despite the above measures, his oxygen requirement increased with repeat Blood gas post surfactant having severe hypoxemia; hence he was shifted to high-frequency ventilation. Suspecting severe PPHN he was given a loading dose of Inj. Magnesium sulphate.

He received sodium bicarbonate infusion in view of severe metabolic acidosis. 2D Echo done showed Obstructed TAPVC, supracardiac variety. The baby was shifted to an advanced cardiac centre for further management. However, he had a very stormy post-operative case with secondary bacterial pneumonia in the post-operative period and he could not be weaned off from ventilatory support. He succumbed after a hospital stay of 20 days.

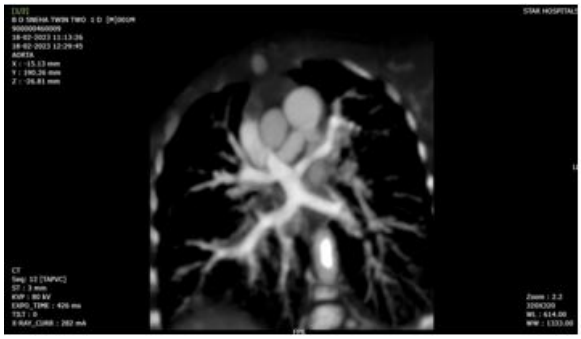

Figure 5: Both twins had the same pattern on the CT angiogram showing the pulmonary veins draining into the SVC via the vertical vein.

Discussion

Patients with TAPVR have occasionally been reported with single gene disorders such as Holt-Oram syndrome, Noonan syndrome, Ivermark syndrome, and associated chromosomal aberrations (22 partial trisomies) [5]. Several familial cases of TAPVR have been reported, but no notable pedigree has been reported, except in the case of the Utah-Idaho family in 1994 [2]. Bleyl et al. described a 4p13-q12 chromosomal abnormality in the aforementioned Utah-Idaho family [6]. Several chromosomal or gene abnormalities associated with TAPVR have been reported. Ramer et al. reported five children with del(2) (q31q33) and one individual with dup(2)(q31q33) from a single family. [7]. Wu et al. suspected a deletion of 11q24.2-qter was related to TAPVR.[8]. Degenhardt et al. reported that the SEMA3D gene is associated with TAPVR or partial anomalous pulmonary venous return. They identified semaphorin 3D as playing a crucial role in pulmonary venous patterning. Although no other abnormality was associated with a congenital disorder in our case, we did propose a genetic study for these siblings, however, the parents did not desire further evaluation.

Conclusion

Based on the above case scenario and the aforementioned studies, it is very clear that a larger genetic study was needed to determine the genetic background of TAPVR. Screening of all siblings with TAPVR is a burden to the patients due to psychological stress or financial problems for the parents. However, these consequences are outweighed by the benefit of diagnosing this critical disease early so that the patient can be taken for surgery before severe congestive heart failure develops. Therefore, if a genetic background is detected in a case of TAPVR, routine fetal echocardiography or cardiography immediately after birth is reasonable to screen TAPVR.

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