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Case Report

Fetal Transposition of the Great Arteries with a Normal Three-Vessel View: Report of Two Cases

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Keywords

- Fetus
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Abstract

Transposition of the great arteries (TGA) affects approximately 2 in 10,000 live births. Babies with this lesion can have significant cyanosis, increased risk of hypoxic cerebrovascular events and death. Fetal diagnosis is important to ensure appropriate postnatal management. Most cases of TGA will have an abnormal three-vessel view on antenatal screening, which would prompt evaluation by echocardiography and prompt appropriate perinatal management. We report 2 unusual cases of TGA with normal three- vessel views that highlight the importance of evaluating outflow views.

ABBREVIATIONS

TGA: Transposition of the Great Arteries; VA: Ventriculo-Arterial; RV: Right Ventricle; LV: Left Ventricle

INTRODUCTION

TGA affects approximately 2 in 10,000 live births and is a common cardiac etiology of neonatal cyanosis [1,2]. Babies with this lesion can have significant cyanosis, increased risk of hypoxic cerebrovascular events and death. Fetal diagnosis is important to ensure appropriate postnatal management. We report 2 unusual cases of TGA that highlight the importance of evaluating outflow views.

CASE PRESENTATION

The mother of case 1 was referred at 30 weeks of gestation for fetal echocardiogram and management after abnormal cardiac screening and an abnormal fetal echocardiogram at a peripheral center. Pregnancy history was unremarkable and no other anomalies were detected. The mother of case 2 was referred at 20 gestational weeks in her second pregnancy for di-chorionic di-amniotic twins following an abnormal cardiac screen with suspicion for TGA in one of the twins. No additional anomalies were detected.

Fetal echocardiography findings of cases 1 and 2 were largely identical. Four-chamber view showed levocardia, normal concordant atrio-ventricular connections, and normal sized structures. Both fetuses had large ventricular septal defects (VSDs) (Figure 1; Video clip 1). Three-vessel and three-vessel-tracheal views were unremarkable showing normally aligned and sized vascular structures (Figure 2; Video clip 2). When performing an upward transverse echocardiography sweep from the apical four-chamber towards the ventricular outflow tracts, the aorta was noted to arise as a first vessel from the right ventricle (RV), followed by the main pulmonary artery from the left ventricle (LV) as the anterior vessel (Figure 3, Video clip 3), compatible with TGA, alias discordant ventricular-arterial

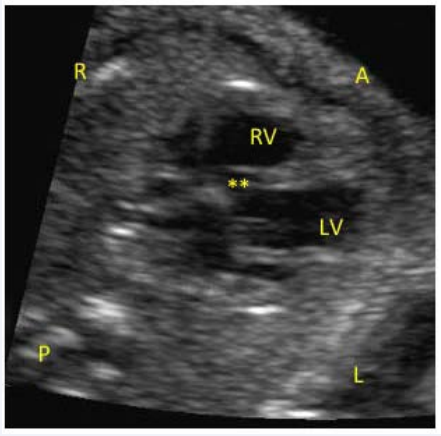
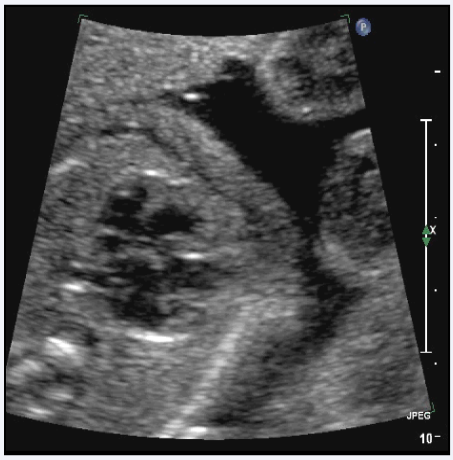


Figure 1 Two-dimensional echocardiographic image of the four-chamber view in a fetus showing a ventricular septal defect (VSD) in the inlet septum shown by the **. A= anterior, P = posterior, R = right, L = left, RV = right ventricle, LV = left ventricle, VSD = ventricular septal defect.



Video 1 Four-chamber view showing a large ventricular septal defect in the inlet septum. The ventricles are balanced and there is normal biventricular systolic function.

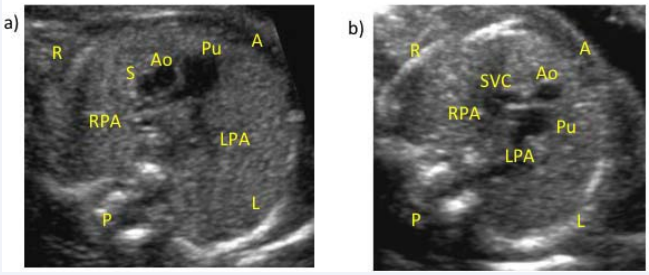
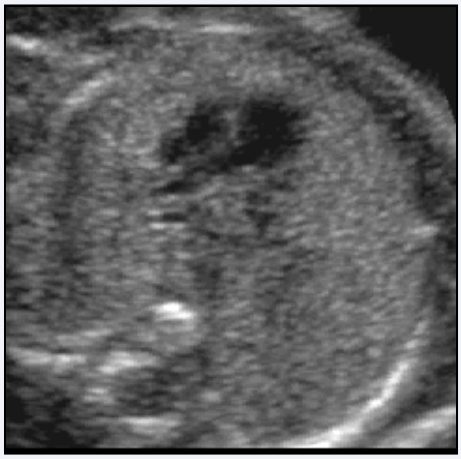


Figure 2 a) Two-dimensional echocardiographic image of the three-vessel view in the fetus of case one showing normal arrangement of the great vessels in posterior TGA. The aorta (Ao) is posterior to the pulmonary artery (Pu) as is seen usually in fetuses with normally related great vessels. In comparison b) shows the abnormal arrangement of the vessels in the three-vessel view in a usual case of TGA, with the aorta anterior and rightward of the pulmonary artery. A = anterior, P = posterior, R = right, L = left, S = superior vena cava, Ao = aorta, Pu = pulmonary artery, RPA = right pulmonary artery, LPA = left pulmonary artery.



Video 2 Three-vessel view showing a normal arrangement of the great vessels. The pulmonary artery is seen leftward and anterior to the aorta, which is leftward and slightly anterior to the superior vena cava.

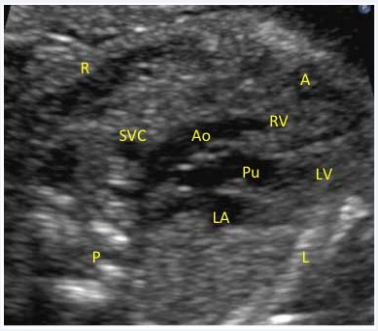
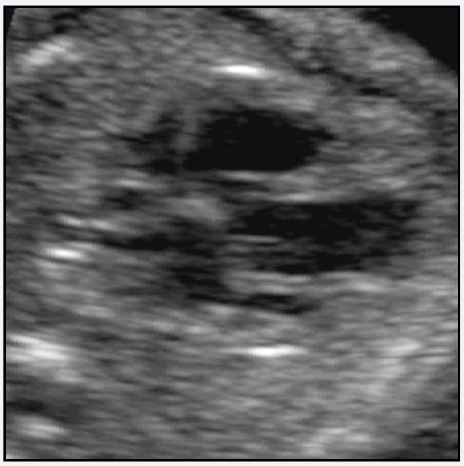


Figure 3 Two dimensional echocardiographic image showing the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle. R = right, L = left, A = anterior, P = posterior, Ao = aorta, Pu = pulmonary artery, RV = right ventricle, L = left ventricle, LA = left atrium, SVC = superior vena cava.



Video 3 Outflow view showing parallel great vessels. The aorta is seen arising from the right ventricle and the pulmonary artery is seen arising from the left ventricle.

(VA) connections. Due to the risk of neonatal compromise in the setting of TGA physiology, delivery at term was arranged with the high-risk pregnancy programs closest to our tertiary care centers. Case 1, a female neonate, presented with an oxygen saturation of 64-70% after birth despite prostaglandin infusion. Oxygen saturation was significantly improved by balloon atrial septostomy. In case 2, a female neonate, oxygen saturation was initially greater than 90%. At one week of life, balloon atrial septostomy was performed due to progressive desaturation. Both babies were discharged home and underwent elective surgery at 8 weeks and 9 weeks of age respectively, including closure of the VSD and ASD and arterial switch operation. In addition to the arterial switch and VSD closure, case 2 required Tran's section and re implantation of the left pulmonary artery with an interposition graft due to compression of the circumflex artery. Surgery in case one was complicated by ventricular fibrillation and asystolic cardiac arrest on postoperative day one and rescue with ECMO, which was discontinued on POD #2. Remainder of her postoperative course was unremarkable. Case 2 had an unremarkable postoperative course but required prolonged admission for poor weight gain. Currently both children remain well and asymptomatic at 3 and 6 years of age.

Table 1:

Author	Case	Additional Lesions	Mode of Diagnosis	Outcome
Van Praagh et al 1971[9]	1 2 3 4	VSD, PFO VSD, ASD VSD, PFO VSD, PFO, PS	Autopsy	Death 43 days Death 20 days Death 2 days Death 6 months
Wilkinson et al 1975[8]	5 6 7 8 9 10 11	VSD VSD VSD VSD VSD, bicuspid pulmonary valve VSD, bicuspid pulmonary valve with severe stenosis VSD	Angiography and autopsy	Death 1 month Death 3 weeks Death 7 weeks Death 6 months Death 4 months Death 6 months Death 3 weeks
Buchler et al 1984[4]	12 13 14	VSD, PFO VSD, ASD No VSD, ASD	Autopsy	All demised Tme of death unknown
Virdi et al 1988[5]	15	Multiple VSDs	Echocardiography (day 1)	Complete repair Death day 1 postop
Benatar et al 1990[6]	16	VSD, sub-aortic obstruction	Echocardiography (day 1)	Complete repair Death day 5 postop
Miyake et al 1990[7]	17	VSD	Angiography (day 13)	Complete repair

Table 1: Previous cases of posterior TGA that have been found in the literature. All cases have been diagnosed postnatally; the first 11 cases were diagnosed on autopsy while only 3 cases were diagnosed correctly without post-mortem examination. Our cases are the first ones to be diagnosed in the fetus.
Abbreviations: VSD: Ventricular Septal Defect; PFO: Patent Foramen Ovale; ASD: Atrial Septal Defect. PS: Pulmonary Stenosis

DISCUSSION

The majority of babies with discordant VA connections will have the aorta positioned anteriorly to the pulmonary artery. In a cranial sweep from an apical transverse four-chamber view, the pulmonary artery is seen rising as the first vessel from the LV, followed by the more anterior aorta from the RV. The ascending aorta and main pulmonary artery are not crossing over but have parallel course in the longitudinal outflow tract view. In the three-vessel view, the pulmonary artery, aorta and superior vena cava form a triangle, with an anterior central aorta, a posterior-left pulmonary artery and a posterior right superior vena cava (Figure 2b) [3]. In the three-vessel tracheal view, only the aortic arch as the most superior vessel is seen rather than the ductal and aortic arch forming a V sign in cases with a normal VA connection. Thus in TGA, the outflow tract, three-vessel and three-vessel tracheal views will be abnormal which facilitates the prenatal detection.

TGA with aorta posterior to the pulmonary artery is extremely rare. Despite widespread use of fetal echocardiography there have been no earlier reports of fetal cases with posterior TGA partly because the diagnosis is challenging. Our literature research resulted in less than 20 cases with a postnatal diagnosis of posterior TGA that had been reported in case reports or small case series (Table 1) [4-9]. Key differences from the classical TGA include: a) the posterior origin of the aorta from the RV; b) crossover of the great arteries in an “inverted” pattern; c) a normal alignment (from left anterior to right more posterior) of the main pulmonary artery, aorta and superior vena cava in the three vessel view; and d) a normal V-sign formed by the transverse aortic arch and the ductal arch in the three vessel tracheal view. Thus in posterior TGA, only the outflow tract view will be abnormal while the three-vessel and three-vessel tracheal views will appear normal. A large VSD may help with prenatal identification, as it seems to be a common association with posterior TGA. Despite differences in appearance, newborns with posterior TGA carry the same risk of compromise and cyanosis as classical TGA. Perinatal management including in-utero transfer for delivery should be provided by a tertiary care center with expertise in the care of high-risk cardiac patients. Whereas a fetus with a usual TGA would be identified by three-vessel view and receive appropriate perinatal management, a fetus with posterior TGA may not be identified, thus may not be delivered in a tertiary care center and receive prompt initiation of prostaglandin. Timing of balloon atrial septostomy may also be delayed. Fetuses with posterior TGA identified correctly would receive appropriate perinatal management and significant cyanosis can be potentially avoided. Surgical management would include an arterial switch and outcomes would expect to be the same as a usual TGA.

In summary, we report two cases of posterior TGA. Careful evaluation of outflow tract views allowed prenatal diagnoses of discordant VA connections. Careful attention should be made in examining the outflows; the pulmonary artery should be shown to bifurcate into the branch pulmonary arteries and the head and neck vessels should be seen arising from the aorta. Once the outflows are correctly identified, the VA connection should be evaluated to ensure the great arteries are not transposed. Posterior TGA is a rare example of a conotruncal abnormality that will not be detectable in the three-vessel and three-vessel tracheal views. While fetal echocardiography may stress the importance of evaluating the outflow tracts, three-vessel and three-vessel tracheal views, these routine views may not all be performed on the detailed anatomic obstetric scan.

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