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*Brief Report*

# Successful surgical palliation of a triple-outlet right ventricle: a rare congenital cardiac malformation

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**Abstract** Most commonly, hearts have either a single arterial trunk or two arterial trunks; however, rare reports exist of a "tritruncal" heart, a heart with three outflow tracts. Here, we present one of the first reports of successful surgical palliation of a triple-outlet right ventricle.

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MOST COMMONLY, HEARTS HAVE EITHER A SINGLE arterial trunk or two arterial trunks; however, rare reports exist of a "tritruncal" heart, a heart with three outflow tracts. Diaz-Gongora et al[1] and Pelletier et al[2] have described similar cases of hearts with a right ventricle from which a pulmonary trunk arose (giving rise to a left pulmonary artery and an arterial duct) and a left ventricle from which both an aorta and an "intermediate" trunk (giving rise to a right pulmonary artery) arose. In both cases, the "intermediate trunk" had its own lumen and rudimentary valve and was found anterior to the aorta and posterior to the pulmonary trunk. Tingo et al[3] reported the first case of a triple-outlet right ventricle in which a pulmonary trunk, an "intermediate trunk", and aorta all arose from a morphologic right ventricle. Their patient, who also had an interrupted aortic arch, succumbed to infection 6 months after a complicated surgical repair and postoperative course. Here we present one of the first reports of successful surgical palliation of a triple-outlet right ventricle.

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## Clinical summary

Our patient was diagnosed prenatally with a double-outlet right ventricle with D-transposition of the great vessels with moderate pulmonary valve regurgitation, severe mitral stenosis, an intact atrial septum, and a large ventricular septal defect. Because of the concern for severe mitral stenosis and intact atrial septum the baby was born via caesarean section (38 weeks) and taken emergently to the cardiac catheterisation lab for placement of an atrial stent with successful decompression of the left atrium. Postnatal echocardiogram (Fig 1 and Supplemental Movie Clip 1) showed a triple-outlet right ventricle with a main pulmonary arterial trunk (giving rise to the left pulmonary artery and arterial duct) and a right pulmonary arterial trunk arising side by side, with the aorta just anterior to the right pulmonary arterial trunk. The right pulmonary arterial trunk had rudimentary valve tissue that was severely regurgitant, resulting in right pulmonary arterial dilation. The mitral valve was severely stenotic with trivial inflow into a hypoplastic left ventricle. Angiography performed during the catheterisation confirmed the presence of three arterial trunks arising from the right ventricle (Fig 2). The baby was extubated and had been tolerating oral feeds before surgery.

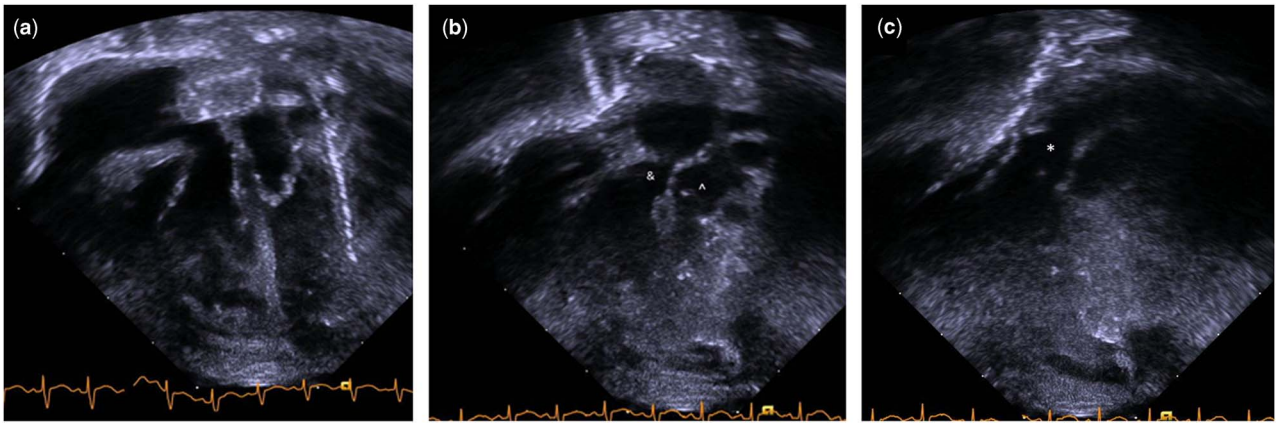


**Figure 1.**
*Transthoracic echocardiogram. (**a**) Apical view demonstrating severe mitral stenosis status post placement of an atrial stent. Three outlets are seen arising from the right ventricle. (**b**) Right pulmonary artery (&) and the main pulmonary artery (^), which gives rise to the left pulmonary artery and patent ductus arteriousus, arise side by side. (**c**) Just anterior to the right pulmonary artery the aorta (*) arises.*

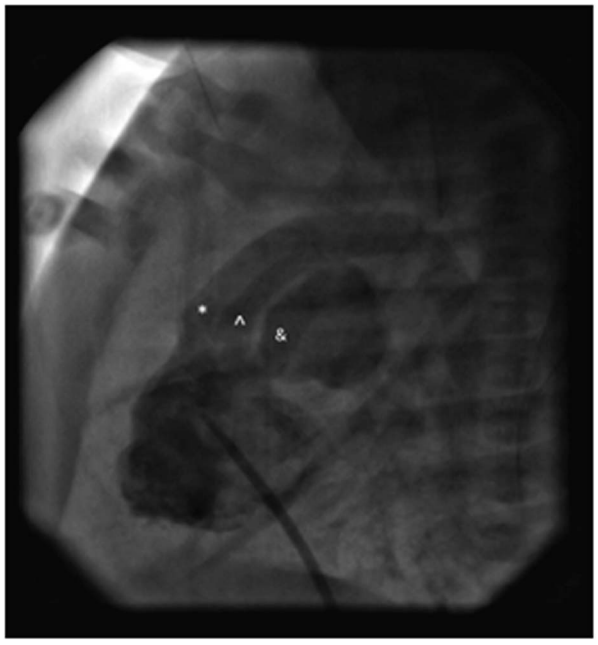


**Figure 2.**
*Cardiac catheterisation. This is a lateral projection of a right ventricular injection demonstrating three distinct outflows, the aorta (*), the main pulmonary artery giving rise to the left pulmonary artery and patent ductus arteriousus (^), and the right pulmonary artery (&), which is severely dilated.*

The patient was taken to the operating room on his 10th day of life where the diagnosis was confirmed. The surgeon appreciated two pulmonary trunks arising from the right ventricle, which shared a medial wall but had two separate lumens. The aorta was confirmed to also arise from the right ventricle just anterior to the pulmonary trunks. The surgical repair consisted of unifocalisation of the right pulmonary artery to the left pulmonary artery, over-sewing of the right pulmonary arterial outlet, ligation of the ductus arteriosus, placement of a 3.5 mm right modified Blalock–Taussig shunt, and removal of the interatrial stent with atrial septectomy. The patient had a relatively uncomplicated postoperative course; he was extubated on postoperative day 5, weaned to room air by postoperative day 8, and discharged home on postoperative day 14, tolerating oral feeds. A predischarge echocardiogram revealed an unobstructed atrial communication, trivial tricuspid valve regurgitation, and a patent Blalock–Taussing shunt with unobstructed flow into the branch pulmonary arteries.

## Discussion

This report describes one of the first successful surgical palliations of a triple-outlet right ventricle. Our case differs from the only other case of presumed triple-outlet right ventricle reported in the literature,[3] in that our patient also had severe mitral valve stenosis precluding a two-ventricle repair.

The embryologic origin of the triple-outlet right ventricle remains speculative. This anomaly may have resulted from a combination of malrotation during aorticopulmonary septation and anomalous fusion of the proximal portions of the right and left branches of the 6th aortic arches to form the pulmonary trunk, as posited by Skidmore.[4] By this mechanism, the left and right 6th arch branches fused with cardiac tissue instead of with each other, yielding two pulmonary outflow tracts. The finding of rudimentary valve tissue at the right ventricle–right pulmonary artery junction is more challenging to explain. On the basis of several theories regarding normal outflow tract and semilunar valve formation, a complex interplay among the myocardial sleeve, the cardiac mesenchyme, and the endocardium of the developing outflow tract

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determines the genesis of semilunar valves from the conotruncal ridges during truncal septation. It is therefore possible that a right 6th arch vessel that did not coalesce with its left counterpart ended up over the right ventricle, so close to its intended location that it was still subject to the same process of semilunar valve and outflow tract formation.[5]

In conclusion, this is one of the first published reports of successful surgical palliation of a triple-outlet right ventricle. The identification and management of this complex cardiac anatomy is intriguing as this combination of defects challenges our understanding of outflow tract development and raises interesting questions for future researchers.

## Acknowledgements

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## Supplementary material

To view supplementary material for this article, please visit http://dx.doi.org/10.1017/S1047951115001237

## References

1. Diaz-Gongora G, Quero-Jimenez M, Espino-Vela J, Arteaga M, Bargeron L. A heart with three arterial trunks (tritruncal heart). Report of a case. Pediatr Cardiol 1982; 3: 293–299.
2. Pelletier GJ, Sokoloski M, Kardon R. Diagnosis and management of tritruncal heart in an infant. J Thorac Cardiovas Surg 2009; 137: 753–755.
3. Tingo JE, Carroll SJ, Crystal MA. Triple outlet right ventricle: a previously unknown cardiac malformation. Cardiol Young 2015; 25: 576–579.
4. Skidmore FD. Development of the right outflow tract and pulmonary arterial supply. Ann R Coll Surg Eng 1975; 57: 186–197.
5. van Den Hoff MJ, Moorman AF, Ruijter JM, et al. Myocardialization of the cardiac outflow tract. Dev Biol 1999; 212: 477–490.