

Double-Orifice Tricuspid Valve in an Infant with Multiple Noncardiac Anomalies



Rachel E. Henning, MD, Victor H. Kieu, MD, and Benjamin H. Goot, MD, *Milwaukee, Wisconsin*

INTRODUCTION

Originally reported by Greenfield in 1876,¹ double orifice of an atrioventricular valve continues to be a rare congenital cardiac anomaly encountered by cardiologists. In comparison with double-orifice of the mitral valve, which has become more recognizable by echocardiogram, double-orifice tricuspid valve (DOTV) is extraordinarily rare, with only 40 reported cases to date.^{1,2} When present, DOTV is often associated with other congenital cardiac anomalies such as divided right atrium, septal defects, mitral or pulmonary valve anomalies, Ebstein's anomaly, tetralogy of Fallot, and transposition of the great arteries.³⁻¹⁵ However, it is not typically associated with abnormalities in other organ systems. This undoubtedly creates a wide spectrum of clinical patient presentation, which contributes to this rare anomaly's frequency and overall morbidity. Here we present a case of a neonate with multiple noncardiac anomalies found to have a DOTV in addition to a bicuspid aortic valve with an otherwise functionally normal heart.

CASE PRESENTATION

A 1-day-old male infant, known to the fetal concerns program due to prenatal concerns, was born at 35 weeks and 0 days via uneventful spontaneous vaginal delivery due to preterm labor. He was clinically well appearing and stable on room air but was transferred immediately to the neonatal intensive care unit (NICU) due to prenatal concerns for duodenal atresia and mega cisterna magna and underwent a screening echocardiogram at 22 hours of life due to these concerns. The transthoracic echocardiogram was notable for DOTV, combined annulus size of 1.33 cm (Z score = 1.42), trivial tricuspid valve insufficiency, mild dilation of his right atrial and ventricular cavities, mild right ventricular hypertrophy, and a well-functioning tricommissural aortic valve with thickening and partial fusion of the right and non-coronary cusps (Videos 1-3; Figure 1). Additionally, a small patent foramen ovale with left-to-right shunting was noted. Multiorgan system screening was completed. His cell-free DNA panel was negative, but further genetic workup yielded a variant of unknown significance in his PEX6 gene. A baseline renal ultrasound was normal, and baseline

head ultrasound was confirmatory for mega cisterna magna of his posterior fossa. After serial exams and testing, he was eventually taken to the operating room due to a concerning imaging that suggested proximal intestinal and rectosigmoid web for exploratory laparotomy. There he was found to have multiple areas of jejunal atresia and a rectosigmoid web requiring extensive bowel resection and duodenojejunal anastomosis on day of life 2. His NICU course was otherwise uneventful from a cardiopulmonary standpoint, with a continually benign cardiac exam and normal hemodynamics. He was ultimately discharged home after a 29-day NICU stay, mostly for surgical recovery and feeding, and has been thriving at latest follow-up.

On recent repeat echocardiogram at 15 months of age his tricuspid valve remains relatively unchanged, with a remeasured combined annular size of 1.43 cm (Z score = -1.62) and continued normal valve function including only trivial regurgitation (Video 4). This study also confirmed a well-functioning bicuspid aortic valve with partial fusion of the right and noncoronary cusps. He continues to grow well, tracking at the 91st percentile for weight, and remains asymptomatic, free from cyanosis, activity intolerance, respiratory distress, and feeding intolerance.

DISCUSSION

A double-orifice atrioventricular valve is an uncommon congenital cardiac anomaly. When discovered, either by autopsy or incidentally by echocardiogram, it more often affects the mitral rather than the tricuspid valve. When DOTV is diagnosed, in most cases it is in association with other congenital heart defects, notably septal defects.³⁻¹⁵ There is no known prior published case report identified of DOTV associated with multifocal jejunal atresias, rectosigmoid web, and/or mega cisterna magna. Like so many previously diagnosed DOTVs, our patient's cardiac anomaly could have easily gone undiagnosed if it had not been for his prenatal concerns for duodenal atresia and mega cisterna magna and need for additional multiorgan screening.

Two double-orifice atrioventricular valve classification models have previously been proposed to better delineate valve anatomy, but no known system exists to help clarify their relative implications toward valve function and eventual patient morbidity. In 1937, Hartmann² focused on double-orifice mitral valve classification and distinguished three types (L, B, S) based on orifice sizing and chordal and papillary attachment. In 1967, Sánchez Cascos *et al*⁹ revised Hartmann's classification to pertain to tricuspid valves, again using similar methodology, but focusing more on accessory orifice size type and location. Sánchez Cascos *et al* described three types as well: a central type, commissural type, and hole type. Based on these classifications, our patient has a type most similar to a Hartmann's type S (orifices are nearly identical in size, but each orifice has an independent set of chordae and papillary muscles that are positioned in the usual locations) and Sánchez Cascos's central type (fibrous band or bridge of tissue divides the orifice into two).

While most double-orifice valves are neither incompetent nor stenotic at diagnosis, it is unclear at this time what implications our

From the Department of Pediatrics (R.E.H.) and Department of Pediatric Cardiology (V.H.K., B.H.G.), Medical College of Wisconsin, Milwaukee, Wisconsin. Keywords: Double-orifice tricuspid valve, Mega cisterna magna, Neonate, PEX6 gene, Duodenal and jejunal atresia

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Correspondence: Benjamin H. Goot, MD, Children's Wisconsin, 9000 West Wisconsin, Milwaukee, Wisconsin 53226. (E-mail: bgoot@chw.org).

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VIDEO HIGHLIGHTS

Video 1: Subcostal sagittal view from initial transthoracic echocardiogram showing a well-functioning symmetric DOTV from en face assessment.

Video 2: Apical four-chamber view from initial transthoracic echocardiogram demonstrating both tricuspid orifices.

Video 3: Apical four-chamber view from initial transthoracic echocardiogram with focus on tricuspid valve with color Doppler demonstrating laminar inflow and just trivial tricuspid regurgitation.

Video 4: Apical four-chamber view from follow-up transthoracic echocardiogram with focus on tricuspid valve with color Doppler demonstrating unchanged tricuspid valve function.

View the video content online at www.cvcasejournal.com.



Figure 1 Subcostal sagittal view from initial transthoracic echocardiogram showing a symmetric DOTV. Arrows point to each separate orifice.

patient's DOTV will have long term based on his Hartmann's and Sánchez Cascos's classifications. At baseline, his DOTV functioned well and continues to function well on follow-up with no color and/or spectral Doppler imaging suggestive of progressive stenosis or regurgitation.

CONCLUSION

We present the very rare finding of DOTV in a patient with multifocal jejunal atresias, rectosigmoid web, and mega cisterna magna. In recognizing that DOTV can exist in relative isolation and not just with other

congenital heart defects, we believe that the emphasis of routine echocardiographic assessment of both atrioventricular valves for an accessory orifice when screening for disease can unmask an otherwise easily missed cardiac anomaly with unknown implications. As there are no current clear guidelines to date for ongoing continued follow-up of isolated DOTV, greater recognition of this anomaly and its natural history can help provide future understanding of the role of counseling and surveillance timeline for potential complications.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2022.03.002>.

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