

One Truncal Valve or Two Semilunar Valves: An Unusual Case of Truncus Arteriosus

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INTRODUCTION

In truncus arteriosus, a common arterial trunk arises from the heart and gives rise to the ascending aorta, pulmonary arteries, and coronary arteries. Proximal to the arterial trunk is typically a single truncal valve, the morphology of which varies. Highlighting the embryologic aberration resulting in this disease, we present the unusual finding of a partially septated truncal valve, giving the appearance of two distinct semilunar valves arrested in development, in a neonate with truncus arteriosus and aortic arch hypoplasia.

CASE PRESENTATION

The patient's mother was initially referred to our Fetal Heart Program because of suspicion of congenital heart disease on screening obstetric fetal ultrasound. The mother was in good health, but her history was notable for oral isotretinoin use during the early weeks of pregnancy. Pre- and postnatal genetic testing was normal. Fetal echocardiography at 20 weeks' gestation revealed truncus arteriosus with aortic arch hypoplasia. The truncal valve was enlarged, and some images suggested an unusual central division therein (Figure 1, Video 1). The valve had no stenosis and only trivial insufficiency.

Labor was induced, and the patient delivered at 39 weeks' gestation. The infant was started on prostaglandin E₁ to maintain ductal patency and systemic blood flow. Thereafter the infant was admitted to the cardiac intensive care unit. Transthoracic echocardiography confirmed the anatomy of truncus arteriosus, comprising a large conoventricular septal defect and a single arterial vessel giving rise to the ascending aorta, pulmonary arteries, and coronary arteries (Figure 2, Video 2). The particular anatomic variant was most consistent with Van Praagh type A4, in that there was a large main pulmonary artery segment with a small ascending aorta, aortic arch hypoplasia, and near interruption at the aortic isthmus^{1,2} (Videos 3–6). A large patent ductus arteriosus was in continuity with the descending aorta. Cardiac magnetic resonance imaging performed for preoperative planning confirmed the great vessel anatomy as described (Figure 3).

The truncal valve was extraordinary. In most cases of truncus arteriosus, the truncal valve is tricuspid, bicuspid, or quadricuspid,³ and all valve leaflets are in the same plane. In this case, the valve consisted of two distinct valve orifices separated by a central fibrous raphe. Five

leaflets were identified in total: three grouped posteriorly, with the remaining two grouped anteriorly. From the parasternal short-axis plane, this arrangement gave the appearance of a smaller posterior tricuspid aortic valve partially fused with a larger anterior bicuspid pulmonary valve (Figure 4, Video 7). Although all the leaflets were in continuity, the two groups of leaflets were clearly oriented in separate planes (Videos 8 and 9), with the central raphe dividing the two planes (Figure 2). The systolic motion of the valve leaflets and color Doppler flow patterns also suggested two separate distinct valve orifices (Figure 2, Video 10). The coronary arteries also appeared to originate from the posterior tricuspid aortic valve (Video 11). The truncal valve was surrounded by infundibular muscle, such that there was mitral-truncal discontinuity.

At 4 days of life, the patient underwent surgical repair. This consisted of closure of the ventricular septal defect to the truncal valve, division of the pulmonary arteries, direct anastomosis of the common trunk with the ascending aorta, placement of a femoral vein homograft from the right ventricle to the pulmonary artery branches, division of the nearly atretic aortic isthmus, and arch reconstruction by direct anastomosis. Intraoperative transesophageal echocardiography confirmed the findings of a truncal valve with two effective orifices at the annular level, but no true aortopulmonary septum was identified (Video 12). Intraoperative inspection of the truncal valve confirmed the unusual central thickening that appeared to separate the truncal valve into the anterior and posterior groups (Figure 5, Video 13).

DISCUSSION

From an embryologic standpoint, truncus arteriosus is conventionally thought to be caused by a failure in the development of the fetal conus cordis and truncus arteriosus. This results in failed septation of (1) the right and left ventricular outflow tracts, (2) the semilunar valves, and (3) the aorta and pulmonary arteries. More recently promulgated theories support accompanying abnormal infundibular development as being responsible for the abnormalities seen in conotruncal abnormalities such as truncus arteriosus.⁴ The finding of a partially septated truncal valve—or, depending on one's point of view, two separate semilunar valves—posed a diagnostic conundrum: was the disease a very large aortopulmonary window with only a small remnant of aortopulmonary septum and an accompanying conoventricular septal defect, or was it true truncus arteriosus with an arrested attempt at semilunar valve development?⁵ The presence of truncal-mitral discontinuity is also very unusual for truncus arteriosus,² although it has been described in one case in an autopsy series.⁶ Interestingly, the presence of two semilunar valves has been reported in truncus arteriosus, albeit in the presence of an intact ventricular septum (Van Praagh type B).² The central thickening identified in the truncal valve in this case suggested a potential line of developmental division between an aortic and a

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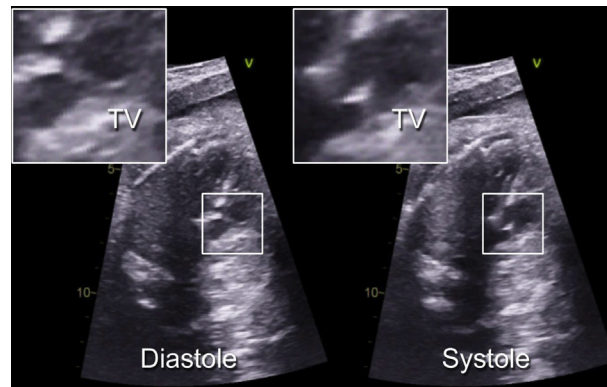


Figure 1 Fetal echocardiogram showing an enlarged truncal valve (TV), with an unusual central division most apparent in systole.

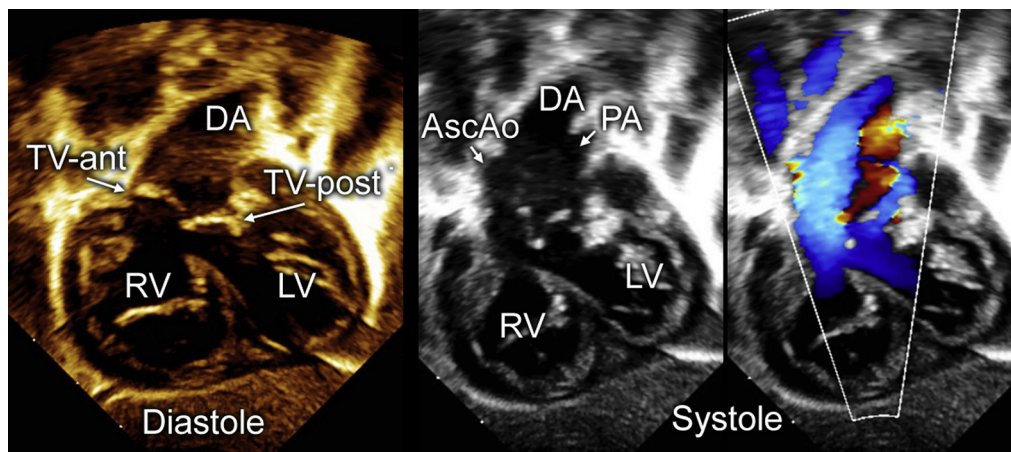


Figure 2 From the subcostal sagittal views, the systolic motion of the valve leaflets and color Doppler flow patterns created the appearance of two separate valve orifices. AscAo, Ascending aorta; DA, ductal arch; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; TV-ant, anterior aspect of truncal valve; TV-post, posterior aspect of truncal valve.

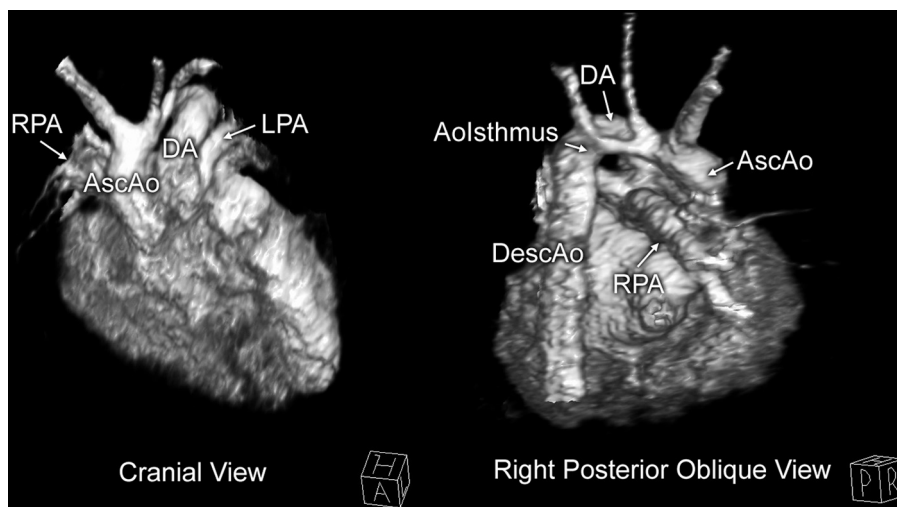


Figure 3 Anatomy of the common trunk via cardiac magnetic resonance imaging. Volume rendering of the magnetic resonance angiogram shows that the common arterial trunk branches off into the ascending aorta (AscAo) and atretic aortic arch, the ductal arch (DA), which supplies the descending aorta (DescAo), and the pulmonary arteries. Aolsthmus, Atretic aortic isthmus; LPA, left pulmonary artery; RPA, right pulmonary artery.

pulmonary valve. This particular case, therefore, appears to represent an intermediate embryologic step between what is typically considered to be truncus arteriosus and aortopulmonary window.

Ultimately, the two abnormalities fall on an embryologic spectrum driven by failure of neural crest cell migration and associated abnormal infundibular development.^{3,4}

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