The spectrum of hypoplastic left heart syndrome spans a continuum from borderline hypoplasia to extreme forms with retrograde blood flow from the ductus arteriosus to the cerebral and coronary arteries. A male neonate, born at 37 weeks' gestation, had an antenatal diagnosis of hypoplastic left heart syndrome. Postnatal echocardiographic assessment confirmed the diagnosis of hypoplastic left heart syndrome with minimal antegrade flow in the aortic sinus, retrograde arch perfusion, and nondemonstrable ascending aorta. As a result of initial instability with recurrent infections, bilateral pulmonary artery banding was performed. Intraoperatively, a thread-like ascending aorta was found (Figure 1A), which warranted further investigation. Focused echocardiographic assessment could not visualize flow in the ascending aorta but demonstrated antegrade flow through a severely stenotic aortic valve with laminar color Doppler flow in the coronaries (Figure 2). Computed tomography angiography confirmed that the ascending aorta ended blindly above the sinotubular junction with antegrade coronary artery perfusion (Figure 3).

Because of the unusual anatomy, he was deemed unsuitable for single-ventricle–type palliation. Worsening episodes of insufficient coronary perfusion with severe distress and irritability, associated with ST-segment changes and elevated troponin levels, further aggravated the course. Subsequently, he underwent heart transplantation at 2 months of age. Pathological examination of the explanted heart confirmed a blind-ending atretic ascending aorta with a relatively well-developed aortic root and usual coronary artery origins.

In this rare anatomic anomaly, we speculate that competing blood flow antegrade through the stenotic aortic valve and retrograde through the aortic arch in fetal life created no flow and atresia of the ascending aorta, as has been suggested previously. In our case, the Norwood-type aortic reconstruction was abandoned mainly as a result of the short height of the aortic root, which makes Damus-Kaye-Stansel anastomosis impossible without coronary artery distortion. The short height of the aortic root and the muscle structure between the great arteries would also have precluded direct implantation of the ascending aorta to the pulmonary trunk. Weidenbach et al reported a similar anatomy in which a neonate had the atresia of the ascending aorta with a large blind pouch of the proximal aortic root. This patient underwent the modified Norwood procedure with the Damus-Kaye-Stansel anastomosis and died of myocardial dysfunction presumably caused by poor myocardial protection. We believe that this anomaly essentially contraindicates conventional Norwood procedure and primary transplantation would be recommended unless the blind pouch is large and long enough to perform Damus-Kaye-Stansel anastomosis without coronary artery distortion.

Disclosures

None.

References

Figure 1. Intraoperative photograph and corresponding ultrasound image. A, Intraoperative findings of the thread-like ascending aorta. B, Two-dimensional image showing the thread-like atretic ascending aorta in a modified parasternal long-axis view. Arrows show the atretic aortic segment. PA indicates pulmonary artery.

Figure 2. Preoperative echocardiographic images. A, Parasternal short-axis image at the aortic sinus level showing the origin of the left coronary artery. B, Color Doppler image of A demonstrating antegrade flow in the aortic root (red flow). Ao indicates aorta; LMCA, left main coronary artery; and PA, pulmonary artery.

Figure 3. Computed tomography (CT) angiography and pathological photograph. A, CT angiography confirmed that the ascending aorta ended blindly above the sinotubular junction with antegrade coronary artery perfusion. B, Gross pathology of the explanted heart showing the aortic root and coronary arteries. Asterisks indicate left and right coronary arteries. Ao indicates aorta; and LV, left ventricle.
Atresia of the Ascending Aorta in Hypoplastic Left Heart Syndrome
Christoph Haller, Osami Honjo, Timothy Bradley, Andreea Dragulescu and Glen S. Van Arsdell

Circulation. 2015;131:925-926
doi: 10.1161/CIRCULATIONAHA.114.013069

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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