Retroaortic Innominate Vein in Hypoplastic Left Heart Syndrome: A Case Report

John Martinez, MD, David Lehenbauer, MD, and Elaine Maldonado, MD, San Antonio, Texas

INTRODUCTION

The innominate vein is formed by the union of the left internal jugular and subclavian veins, normally coursing superoanterior to the aortic arch. Embryologically, the left cardinal veins regress, allowing venous drainage of the left head, neck, and arm into the right anterior cardinal vein via two transverse plexi located superior and inferior to the fourth aortic arch. Typically, this arch shortens and eventually fills the area occupied by the inferior plexus, leading to its regression. Venous blood is then shunted through the superior plexus, leading to development of the normal anatomic course of the innominate vein. The superior capillary plexus may regress in cases of right-sided aortic arch or high-riding aortic arch or in abnormal pulmonary arterial development. These lesions allow persistence of the inferior plexus, leading to formation of a retroaortic innominate vein.1,2 Retroaortic innominate vein is a rare finding in congenital heart disease (CHD), with a reported incidence of approximately 0.55%.1 The presence of this anatomic variant has been described in congenital cardiac lesions with right ventricular outflow tract obstruction, most commonly tetralogy of Fallot.1,3 This finding has been described previously in cases of double-outlet right ventricle with mitral atresia and a hypoplastic left ventricle,1 in addition to coarctation of the aorta.3 Herein, we describe a case of retroaortic innominate vein in an infant with hypoplastic left heart syndrome (HLHS).

CASE PRESENTATION

Our patient was a term female infant born at 40 weeks and 2 days to a 25-year-old G3 mother with negative results on serology via repeat cesarean section. The infant was diagnosed in utero with HLHS at 36 weeks’ gestation. Apgar scores were 8 and 9 at 1 and 5 min, respectively. No resuscitation was necessary in the delivery room. Initial physical examination was remarkable for 1/6 systolic murmur. The infant was stabilized in the neonatal intensive care unit and subsequently transferred to the pediatric cardiac care unit, where she was started on a dinoprostone and milrinone drip at 0.01 and 0.3 µg/kg/min, respectively. Initial transthoracic echocardiography performed shortly after birth confirmed HLHS with aortic and mitral atresia. The presence of a retroaortic innominate vein draining to the right superior vena cava at the level of the ayzygous vein was noted (Figures 1-4, Videos 1 and 2). The right ventricle was dilated, with normal systolic function. A left-sided aortic arch with a severely hypoplastic ascending aorta was observed (Figures 1-4, Video 3). On day of life 6 the patient underwent a Norwood procedure with Sano shunt placement. The presence of a retroaortic innominate vein was confirmed intraoperatively, leading to difficulties with aortic arch reconstruction and venous drainage. The patient underwent delayed sternal closure on postoperative day 8. The initial postoperative course was complicated by hypotension with progressive supraventricular tachycardia and increased central venous pressure, requiring epinephrine and subsequent extracorporeal membrane oxygenation support. After 3 days, echoencephalography revealed a right-sided intraparenchymal hemorrhage, and the patient was separated from extracorporeal membrane oxygenation support. The patient remained hospitalized until postoperative day 84.
Figure 2  Retroaortic innominate vein and right pulmonary artery in short-axis view with Doppler.

Figure 3  Anteroposterior and sagittal angiographic view of retroaortic innominate vein.

Figure 4  Anteroposterior and sagittal angiographic view of left-sided aortic arch.
DISCUSSION

Diagnosis of CHD has improved with the use of routine obstetric sonographic surveillance. Fetal echocardiography has further improved the prenatal diagnosis of CHD. Transthoracic echocardiography performed shortly following delivery serves a dual purpose: confirmation of the suspected diagnosis and proper visualization of cardiac anatomy and function. Preoperative knowledge of anatomic variants allows cardiothoracic surgeons to anticipate potential intraoperative difficulties.

The retroaortic innominate vein in CHD is a rare variant, with a reported incidence of <1%.1 Historically, it has been described in CHD with the presence of right ventricular outflow tract obstruction, most commonly tetralogy of Fallot.1,3 Additionally, right-sided aortic arch has been described previously as a common association.1 The finding of a retroaortic innominate vein in obstructions of the left ventricular outflow tract is rare. A thorough search of the literature failed to reveal a case in a patient with HLHS. Herein, we have described a case of retroaortic innominate vein in an infant with prenatally diagnosed HLHS.

CONCLUSION

This case highlights the importance of prenatal anatomic surveillance in CHD. Furthermore, it emphasizes the importance of accurate anatomic and functional characterization of infants born with congenital lesions. Knowledge of anatomic variation may assist surgeons in anticipation and avoidance of intraoperative difficulties.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.12.002.

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