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Septum Primum Atrial Septal Defect in an Infant with Hypoplastic Left Heart Syndrome

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ABSTRACT

Hypoplastic left heart syndrome (HLHS) is a form of congenital heart disease characterized by severe underdevelopment of the left heart, leading to inadequate systemic blood flow. Several different atrial septal morphologies are observed in HLHS, most commonly a secundum atrial septal defect, patent foramen ovale, intact septum, and leftward displacement of the superior attachment of the septum primum. It has been postulated that atrial septal development is associated with the development of the left heart. We present a case of a newborn infant with HLHS and the unusual finding of a primum ASD.

INTRODUCTION

Hypoplastic left heart syndrome (HLHS) is a form of congenital heart disease in which the left structures are underdeveloped, leading to insufficient systemic blood flow. In HLHS, an interatrial communication allows for oxygenated blood to circulate to the body via a patent ductus arteriosus. In cases of intact atrial septum, anomalous pulmonary venous connections form to provide blood flow. There are different atrial septal morphologies observed in HLHS, such as the secundum atrial septal defect (ASD), patent foramen ovale (PFO), intact atrial septum, and leftward displacement of the superior attachment of the septum primum (LDSP) in relation to the septum secundum. We present the case of a newborn infant diagnosed after birth with HLHS, and found to have the unusual finding of a primum ASD.

CASE PRESENTATION

A term newborn female presented with difficulty feeding, cyanosis, and a loud systolic heart murmur several hours after an uneventful delivery. Prostaglandins were initiated to ensure adequate flow through a patent ductus arteriosus. An echocardiogram revealed severe mitral valve stenosis and hypoplasia of the left ventricle (Figure 1) and an atretic ascending aorta (3-4 mm) (Figure 2), consistent with hypoplastic left heart syndrome (HLHS). In addition to a PFO (†), a large septum primum ASD (*) was present (Figure 3, Clip 1; Figure 4, Clip 2) with left-to-right shunting.

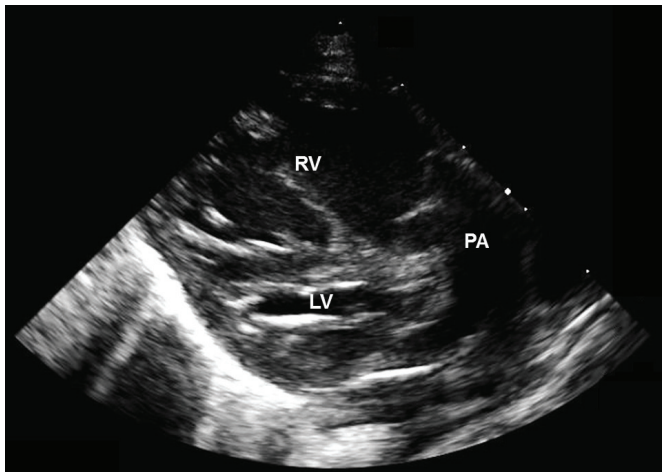

Figure 1. Transthoracic echocardiogram demonstrating hypoplastic left ventricle.

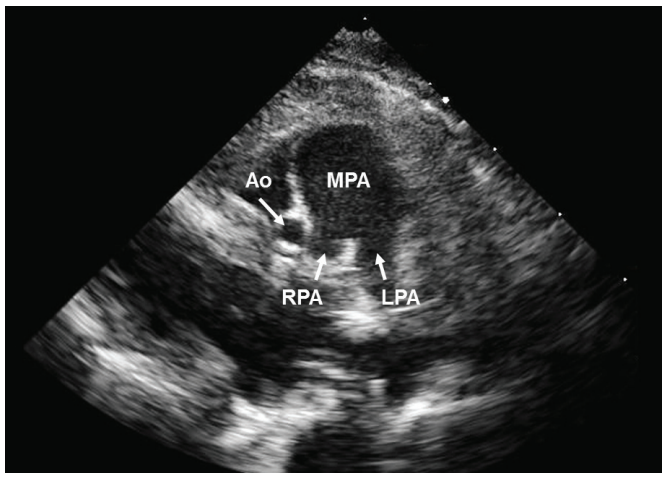

Figure 2. Transthoracic echocardiogram displaying the diminutive ascending aorta.

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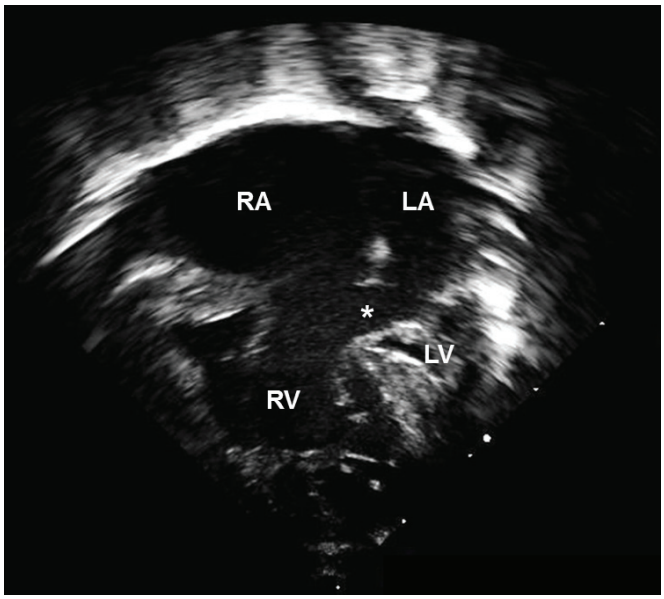

Figure 3. Echocardiogram, 4-chamber view demonstrating a large septum primum atrial septal defect.

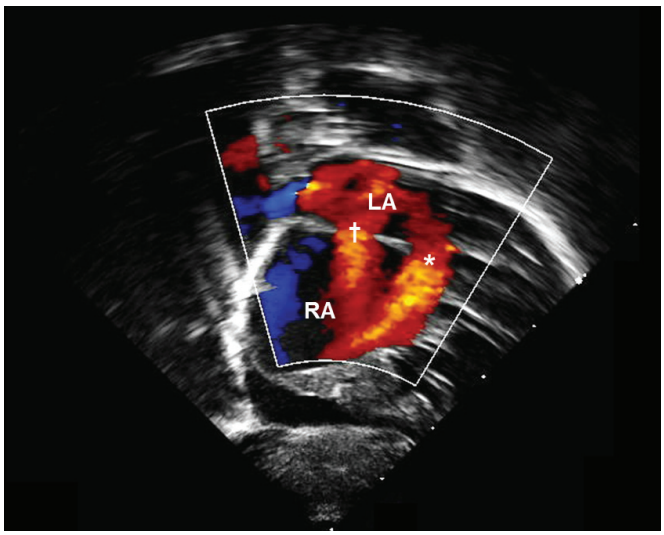

Figure 4. Echocardiogram, 4-chamber view demonstrating left-to-right shunting through the septum primum atrial septal defect.

The infant remained on a prostaglandin infusion until stage 1 Norwood palliation, (aorta-pulmonary artery anastomosis, aortic arch reconstruction, right ventricle to pulmonary artery shunt). Presence of a primum ASD was confirmed at surgery. The muscular tissue between the PFO and primum ASD was excised, followed by excision of the remaining secundum tissue, thereby creating a large unrestricted ASD. The postoperative course was uncomplicated. At 4 months of age, the infant underwent a successful bidirectional cavopulmonary anastomosis and right ventricle to pulmonary artery shunt takedown. She has done well since then with normal systemic ventricular function and unrestricted atrial shunting. She is currently 22 months of age and awaiting a Fontan procedure in the next year.

DISCUSSION

A primum ASD is an uncommon finding associated with HLHS. Weinberg and Weindling examined 110 post-mortem cases of HLHS in which they identified 4 atrial septal morphologies [Weinberg 1988]. The observed morphologies included LDSP, secundum ASD, PFO, and malaligned AV canal. The latter morphology refers to a primum ASD with a common atrioventricular (AV) valve and left ventricular hypoplasia, now generally referred to as unbalanced AV canal with right ventricular dominance.

A prior case series reviewed 31 post-mortem specimens from infants with mitral valve atresia, only two of which had predominant atrial shunting via a septum primum ASD [Williams 1974]. One child had mitral atresia with a primum ASD in addition to a right atrium-to-left ventricle communication and normal development of the left ventricle. The other child had traditional HLHS, similar to our case, with mitral atresia and an underdeveloped aortic arch and left ventricle.

A more recent article from Park et al. studied the atrial septal morphologies of 71 consecutive patients with HLHS [Park 2013]. The most common morphology seen was LDSP (64%). Primum ASD was not observed in any patients among this cohort. The authors concluded that LDSP correlates with more severe maldevelopment of the left heart structures in HLHS. This supports the speculation that septum primum formation may play a role in the initial morphologic maldevelopment in HLHS. The growth of the septum primum predates the embryologic formation of the semilunar valves, which leads to the thought that impeded/altered flow through the normal PFO may contribute to underdevelopment of the left heart [Lev 1952].

The presence of a septum primum ASD is rare in HLHS, as it has been previously reported only once collectively from 3 prior studies. From a surgical perspective, the management of this case necessitated additional excisions of the atrial septum to provide completely unimpeded trans-atrial blood flow. Interestingly, our case had mitral stenosis and aortic atresia, which is associated with the highest mortality rate among HLHS subtypes [Tweddell 2002].

In contrast to other associated atrial septal morphologies, presence of a primum ASD is a rare finding with HLHS. To our knowledge, this is the first described case of primum ASD in a living child with HLHS.

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