CASE REPORTS

Balloon Atrial Septostomy by a Right Internal Jugular Venous Approach in a Newborn with Hypoplastic Left Heart Syndrome with a Restrictive Atrial Septum

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ABSTRACT

Hypoplastic left heart syndrome with an intact or highly restrictive atrial septum requires urgent decompression of the left atrium. Catheter-based interventions from the femoral or umbilical veins represent the standard method of atrial decompression. Restrictive atrial septal defects located at the superior portion of the fossa ovalis can be difficult to cross from these access sites. Here, we describe a successful Rashkind balloon atrial septostomy performed from an internal jugular approach.

Key Words. Atrioseptostomy; Internal Jugular Access; Hypoplastic Left Heart Syndrome

Introduction

Hypoplastic left heart syndrome (HLHS) with an intact or highly restrictive atrial septum is a potentially lethal combination with a historically high morbidity and mortality secondary to marked hypoxemia and pulmonary venous congestion.[1,2] Transcatheter techniques for left atrial decompression are typically performed from a femoral or umbilical venous approach but can be difficult in cases with unusual locations of the atrial septal defect (ASD) or a thickened atrial septum. The internal jugular (IJ) vein has traditionally not been used secondary to concerns about avulsing the superior vena cava, damaging the sinoatrial node, and difficulty in gaining left atrial access through this route.[3,4] However, as presented here, an IJ venous approach for balloon atrial septostomy (BAS) in patients with HLHS with a restrictive atrial septum offers an alternate route to enlarge certain ASDs and has not previously been reported in the literature.

Institution where work performed: University of Michigan C.S. Mott Children's Hospital Congenital Heart Center.

Case Presentation

A newborn without a prenatal diagnosis of congenital heart disease was hypoxic at birth (oxygen saturations 75–80%) with no improvement on supplemental oxygen. Transthoracic echocardiogram (Philips iE33, Philips Medical Systems, Andover, MA, USA) revealed HLHS (mitral stenosis and aortic atresia) with a restrictive pinpoint ASD (mean gradient of at least 13 mm Hg) (Figure 1). Additionally, there was partial anomalous pulmonary venous drainage of the left upper pulmonary vein into the left innominate vein that was mildly obstructive, with a mean gradient of 8 mm Hg. Arterial blood gas at 14 hours of life showed a metabolic acidosis (pH 7.30, pCO_2 41 mm Hg, PaO_2 31 mm Hg, and lactate 2.6 mmol/L).

In the catheterization laboratory, the right IJ vein was accessed under ultrasound guidance and a 4-French sheath placed. Using transthoracic echocardiographic guidance, a 4-French Merit JR 2.5 catheter (Merit Medical Systems, Inc., South Jordan, UT, USA) and a 0.035″ angled glidewire (Terumo Medical Corp., Tokyo, Japan) were used to cross the ASD. The mean left atrial pressure was markedly elevated at 25 mm Hg. Through the JR catheter, the glidewire was exchanged for a 0.018″ V-18 wire (Boston Scientific Corp., Heredia, Costa

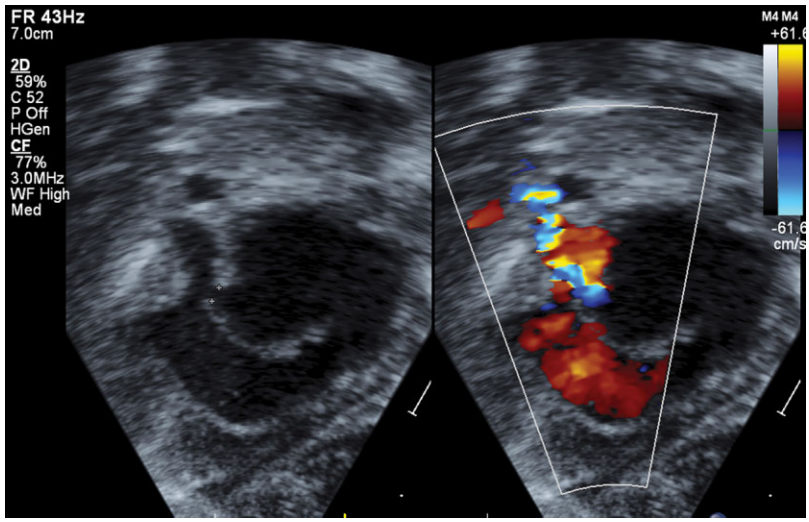

Figure 1. Transthoracic 2D echo with and without color flow mapping documenting a thin intraatrial septum (1.2 mm) and a restrictive superiorly located atrial septal defect (mean gradient at least 13 mm Hg) with left to right flow.

Rica), which was positioned in the left atrium. The IJ sheath was then upsized to a 5.5-French sheath, which was positioned within the right atrium at the superior vena cava–right atrial junction. The JR catheter was advanced back into the left atrium and the 0.018″ wire was exchanged for a 0.014″ Stabilizer XS wire (Cordis Corp., Miami, FL, USA). On fluoroscopy, the 0.014″ wire was outside the heart border, consistent with microperforation of the left atrium. The wire was repositioned, and transthoracic echocardiogram showed a small pericardial effusion, which remained small throughout the case. A 5-French B.Braun Z-5 (9.5 mm) septostomy balloon (NuMed, Inc., Hopkinton, NY, USA) was advanced into the left atrium over the wire (Figure 2), and dynamic atrial septostomy was performed with attention to keeping the sheath tip below the superior vena cava—right atrial junction so that injury would not occur to the sinoatrial node and to prevent avulsion of the superior vena cava. The post-BAS mean left atrial pressure was 10 mm Hg, the mean gradient across the atrial septum was 1 mm Hg, and, by echocardiogram, the defect was significantly larger with improved flow (Figure 3). Postprocedure, the PaO_2 was 40 mm Hg with systemic saturations of ~80%. Serial post-BAS echocardiograms revealed a mildly restrictive ASD (mean gradient 3 mm Hg) and near resolution of the small pericardial effusion. Oxygen saturations and PaO_2 remained stable at ~75–85% and ~35–45 mm Hg, respectively. Three days post-BAS, the patient underwent a Norwood procedure with Sano shunt, repair of the partial anomalous pulmonary venous return, and atrial septectomy.

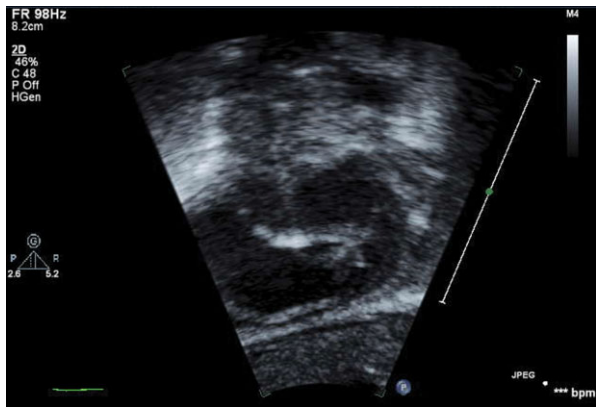

Figure 2. Transthoracic 2D echo with inflated septostomy balloon in the left atrium shows the catheter course from the superior vena cava across the superiorly oriented atrial septal defect into the left atrium.

Postoperative PaO_2s were ~35–42 mm Hg, and there was no evidence of elevated pulmonary vascular resistance throughout the postoperative period.

Discussion

HLHS with a restrictive or intact atrial septum is associated with a significant mortality and morbidity if decompression of the left atrium is not performed. The higher mortality in infants with a restrictive atrial septum may be secondary to changes in the pulmonary vascular histopathology resulting in increased pulmonary vascular resistance, which can be irreversible.[2] Rapid catheter-

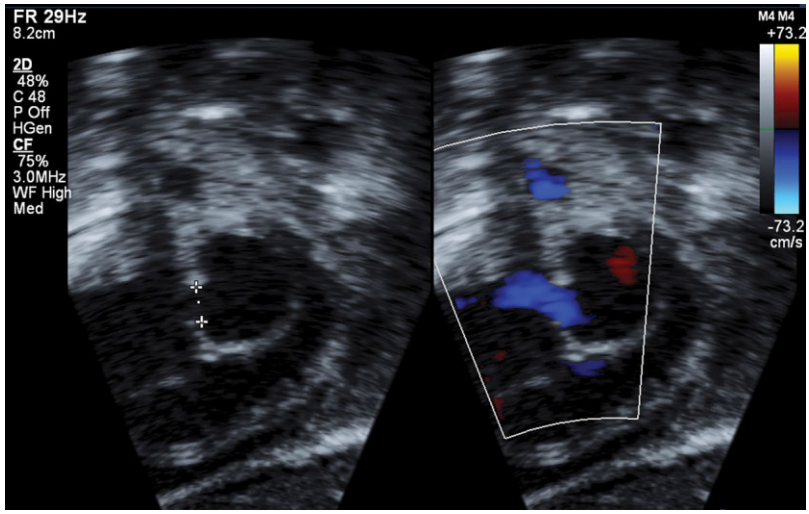

Figure 3. Transthoracic 2D echo with and without color flow mapping after Rashkind balloon atrial septostomy documenting an enlarged atrial septal defect (4.3 mm) with low velocity flow (mean gradient 2 mm Hg).

based interventions are standardly performed in these critically ill infants and include static balloon atrial septal dilation, Rashkind BAS, and intra-atrial stent placement.[5] Factors which may play an important role in the success of atrial septostomy include thickness of the septum, size of the left atrium, and location of the defect. In infants with HLHS with a restrictive but patent and superiorly located ASD, an IJ approach may allow for easier access to the left atrium, potentially avoiding the need for transseptal perforation.

Padhi et al. described successful left atrial decompression from an IJ approach in a 45-day-old with transposition of the great arteries with a restrictive ASD and occlusion of external iliac vein and distal inferior vena cava.[3] However, there are no previous reports in the literature of IJ access for HLHS with a restrictive atrial septum. This case illustrates that an internal jugular venous approach for BAS can be performed as an alternative route in selected patients with HLHS and superiorly located, restrictive ASDs, with particular procedural care to ensure appropriate positioning of the sheath below the superior vena cava—right atrial junction to help avoid superior vena cava avulsion and damage to the sinoatrial node.

Author Contributions

Stewart Mackie collected data, interpreted data, and drafted the article. Ranjit Aiyagari did concept design, helped draft portions of the article, and provided critical revision of the article. Jeffrey Zampi also aided in concept design and drafting portions of the article and provided critical revision of the article.

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