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Case Reports

Percutaneous Atrial Septal Defect Closure in a Child with Interrupted Inferior Vena Cava: Successful Femoral Venous Approach

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Interrupted inferior vena cava (IVC) with azygous continuation to the superior vena cava (SVC) is a relatively common systemic venous anomaly. This anomaly can occasionally complicate transcatheter intervention by rendering more difficult the usual direct route to the systemic venous atrium afforded by femoral venous access. We report our experience with successful transcatheter closure of a large residual atrial septal defect (ASD) using the femoral venous route in a 3-year-old patient with heterotaxy syndrome of left isomerism type, dextrocardia, partial atrioventricular canal defect, and interrupted IVC with azygous continuation to the SVC.

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Key words: atrial septal defect; interrupted inferior vena cava; device closure

INTRODUCTION

Congenital absence of the intrahepatic segment of the inferior vena cava (IVC), also known as interrupted IVC with azygous continuation to the superior vena cava (SVC), is a congenital anomaly found in 0.1–0.6% of the general population, and in approximately 1–3% of patients with congenital heart disease [1,2]. In cases of interrupted IVC with azygous continuation, the IVC is typically interrupted below the hepatic veins (*i.e.*, there is congenital absence of the intrahepatic IVC), and the systemic venous drainage below the interruption then continues via an enlarged azygous vein into the systemic venous atrium through the SVC [3]. The hepatic veins typically drain normally into the inferior portion of the systemic venous atrium, because there is a normally formed hepatic venous confluence and suprahepatic IVC [3].

CASE REPORT

A 3-year-old, 16.5 kg girl with {A, D, S} heterotaxy syndrome (left isomerism type, with polysplenia), dextrocardia, a partial atrioventricular canal defect, an interrupted IVC with azygous continuation to a right-sided SVC with hepatic vein drainage to the left-sided atrium, and multiple small apical muscular ventricular septal defects had previously undergone surgical patch closure of a large primum atrial septal defect (ASD) and diversion of her hepatic veins to the right-sided atrium at 3 months of age. Subsequently, she developed fatigue and exercise intolerance, a prominent right ventricular impulse, and echocardiographic evidence concerning for pulmonary hypertension. The echocardiogram did not reveal interatrial shunting, although the study was limited by lack of patient cooperation and poor transthoracic

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Additional Supporting Information may be found in the online version of this article.

Conflict of interest: Dr. Justino is a physician proctor for AGA Medical.

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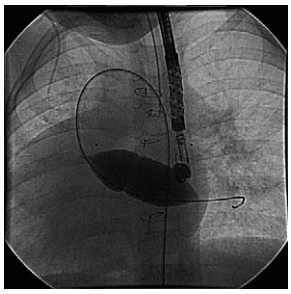

Fig. 1. Balloon sizing of atrial septal defect (waist on balloon measured 19.8 mm).

echocardiographic windows. She therefore underwent cardiac catheterization to obtain hemodynamic data.

A prograde right heart catheterization was performed using right femoral vein access, working through the azygous continuation of the IVC into the right SVC, and a retrograde left heart catheterization was performed using a 4 French pigtail catheter via femoral artery access. Pulmonary artery pressure was mildly elevated at 41/8 with a mean of 22 mm Hg. A large left-to-right shunt at the atrial level (Qp:Qs = 2.7:1) was discovered, and a transesophageal echocardiogram (TEE) was obtained to delineate the anatomic characteristics of the lesion. The TEE showed a large ASD with a deficient superior rim at the level of the SVC, as well as a deficient retroaortic rim, measuring approximately 17 × 17 mm. The edge of the defect appeared irregularly echogenic and thickened, suggestive of a dehisced surgical patch. The area of the atrial septum adjacent to the atrioventricular valves was intact. Abundant left to right shunting was confirmed via color Doppler interrogation and was accompanied by significant right ventricular enlargement. Given the favorable anatomic characteristics, transcatheter device closure was attempted in effort to avoid a second open-heart operation.

Working entirely through the right femoral venous access through the azygous continuation to the right SVC, a 5 French JR 3 catheter was successfully crossed into the left-sided atrium from the right-sided atrium and was advanced deep into the left lower pulmonary vein over a wire. Then, a 0.035 inch Rosen wire (Cook, Bloomington, IN) was selected, and was shaped into a broad 270° loop to ensure stability of the wire deep in the left lower pulmonary vein. The wire was advanced to the left lower pulmonary vein without difficulty. Balloon sizing of the ASD was then performed using a 25 mm sizing balloon (NuMED, Hopkinton, NY). The balloon-sized ASD diameter was 19.8 mm by fluoroscopy (Fig. 1). Once the balloon was removed, a 9 French long delivery sheath and dilator (AGA Medical Corp., Plymouth, MN) were also

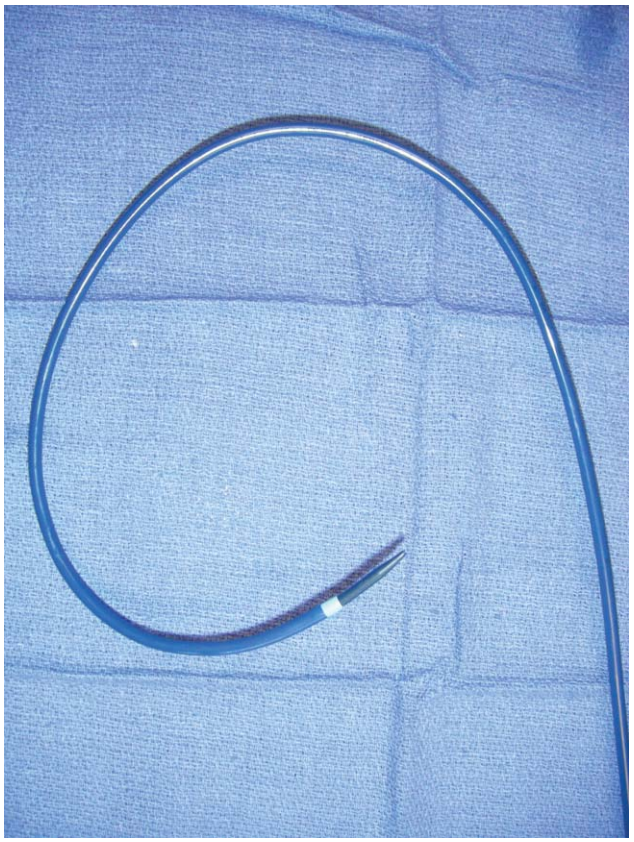

Fig. 2. Photograph of 9 French AGA delivery sheath and dilator pre-shaped into a very broad 270° loop so as to remain stable in the mid left atrium after guidewire removal. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

pre-shaped into a very broad 270° loop so as to remain stable in the left-sided atrium once the guidewire would be removed (Fig. 2). The sheath and dilator were advanced over the Rosen wire without difficulty, and the dilator and wire were then removed, leaving the delivery sheath in stable position within the left-sided atrium (Fig. 3). A 20 mm Amplatzer Septal Occluder device (AGA Medical Corp.) was advanced with its delivery cable through the sheath. The device was successfully advanced within the delivery sheath into the mid left-sided atrium, and the left atrial disc was gradually exposed. The entire assembly was then withdrawn to appose the atrial septum, and the right atrial disc was formed by carefully pushing on the delivery cable while slowly withdrawing the sheath (Fig. 4). Transesophageal echocardiography confirmed excellent position of the device within the atrial septum, with no significant residual shunt and no impingement of the device on any of the surrounding cardiac structures, including the right and left atrioventricular valves, the pulmonary veins, the SVC, and the baffled hepatic veins. Stability testing was performed by gently

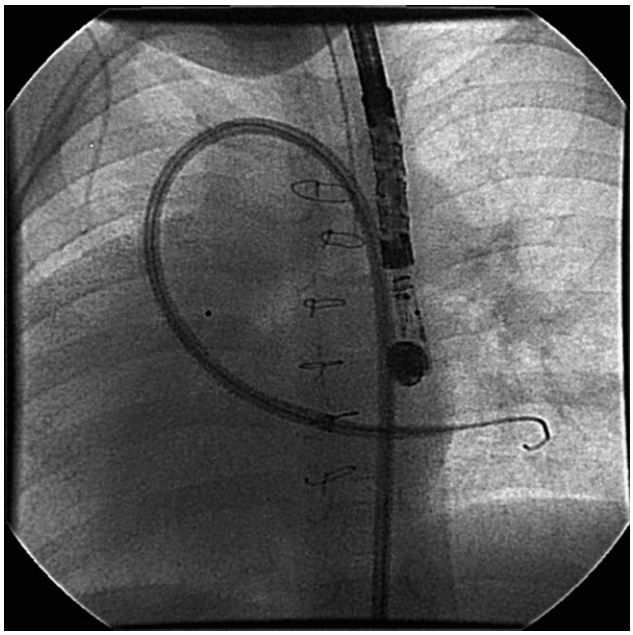

Fig. 3. 9 French long delivery sheath, dilator, and wire in stable position within the left-sided atrium.

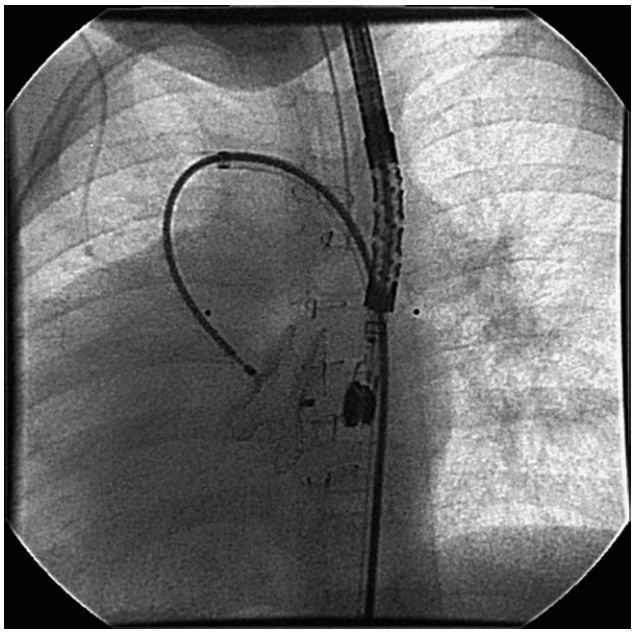

Fig. 4. Placement of 20 mm Amplatzer Septal Occluder device across atrial septal defect.

pushing and pulling the delivery cable, followed by careful reassessment of device position by TEE. When stable positioning of the device was confirmed, the cable was unscrewed, releasing the device into position. TEE assessment revealed that the device remained in stable position.

At 1 year of follow-up, the patient has done very well, with resolution of the right ventricular dilation, and no residual shunting or impingement of the device on surrounding cardiac structures on transthoracic echocardiography. She has had no evidence of atrioventricular block on ECG or Holter.

DISCUSSION

Interrupted IVC with azygous continuation to the SVC may complicate the femoral venous route typically used for diagnostic or interventional cardiac catheterization because of the abrupt 180° turn at the level of the azygous arch. Particularly challenging are interventions that require access to the pulmonary veins and pulmonary venous atrium because of a second abrupt turn required to cross the atrial septum once the catheter has entered the systemic venous atrium from the SVC.

Percutaneous femoral venous access is widely accepted as the standard approach for transcatheter ASD closure. However, this approach tends to be abandoned when interruption of the IVC is encountered, in favor of transjugular access [4,5] or transhepatic puncture [6–11]. There has only been one previous publication describing successful transfemoral device closure of a small ASD in a case of interrupted IVC in an adult [12]. Ours is the first report of successful percutaneous transfemoral closure of an ASD in a child with a large ASD and an interrupted IVC with azygous continuation to the SVC.

Certain technical considerations made transcatheter ASD closure from the femoral venous route possible in our patient despite an interrupted IVC with azygous continuation to the SVC. First, a Rosen wire was selected for balloon sizing of the ASD as well as for subsequent advancement of the delivery sheath; this wire was chosen because it is not excessively stiff. An excessively stiff wire might have dislodged the JR3 catheter from the left lower pulmonary vein when advanced through the complex loop formed by the IVC, SVC, and ASD. Second, the wire was pre-shaped with a broad 270° loop to approximate the anatomy of the ascending IVC, the 180° turn at the azygous arch, and the subsequent 90° turn across the ASD. Then, an adequate inferior rim of the ASD was particularly important, such that despite the complex arc of the sheath, the device could be anchored in the inferior septal rim. This patient had undergone previous repair of a large primum ASD. By definition, primum ASDs have complete absence of the antero-inferior rim (rim bordering the atrioventricular valves), and are therefore not amenable to transcatheter device closure because of the risk of damaging the atrioventricular valves. Although our patient suffered a severe patch dehiscence leading to a large residual ASD, it was somewhat fortunate that the dehiscence occurred superiorly, leaving a sufficient antero-inferior rim to allow the device to anchor well, and to prevent the

device from impinging on the atrioventricular valves. Finally, the fact that the Amplatzer septal occluder can be so easily deployed, recaptured if necessary, and then redeployed, allowed us to attempt device closure with little risk to the patient. Had there been evidence of instability of the device during attempted deployment, or inability to deploy the device because of the tortuous course of the delivery system, the patient could have undergone attempted device closure using an alternate route, such a the internal jugular or transhepatic route. Ultimately, if no device could be safely implanted, the patient would have been referred for surgical closure of the residual ASD.

CONCLUSION

Transcatheter ASD closure using the femoral venous route may be possible despite an interrupted IVC with azygous continuation to the SVC. An attempt using the femoral venous route is a reasonable initial approach, followed by conversion to an internal jugular or transhepatic approach if the femoral venous route proves impossible. Surgical ASD closure should be considered as a final option if transcatheter approaches are unsuccessful.

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