

## Use of steerable delivery catheter to successfully deliver a Ceraflex septal occluder to close an atrial septal defect in a child with interrupted inferior vena cava with azygos continuation

### Yönlendirilebilir taşıyıcı kateter kullanılarak Ceraflex septal occluder ile başarılı bir şekilde kapatılmış olan kesintili vena kava inferiyor-azygos devamlılığı olan atriyal septal defektli bir çocuk

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**Summary**– The closure of a secundum atrial septal defect through the jugular vein in a child with interrupted inferior vena cava with azygos continuation by steerable delivery catheter is described in the present report. The steerable catheter can be used to correct the perpendicular position of the device over the margins of the defect, and is particularly useful in cases of large defects.

**Özet**– Azygos ven devamlılığı olan kesintili vena kava inferiyorda yönlendirilebilir taşıyıcı kateter yardımıyla internal jugular ven yoluyla kapattığımız sekundum tip atriyal septal defektli bir çocuğu sunduk. Bu yönlendirilebilir kateter büyük defektlerde cihazın defektin sınırları üzerinde dikey pozisyonunu düzelterek kapatmaya yardımcı olur.

With advancements in interventional cardiology, transcatheter secundum atrial septal defect (ASD) closure has become the first line of treatment in patients with sufficient rims. The standard procedure is performed through the femoral vein. However, this approach is not always feasible, particularly with large delivery sheaths, in children with interrupted inferior vena cava (IVC), or iliac or femoral vein thrombosis. In such cases, successful transhepatic or transjugular access has been described.<sup>[1–4]</sup> Jugular venous approach using steerable delivery catheter to facilitate orientation of the device to the atrial septum is described in the present report.

#### CASE REPORT

A 12-year-old girl was referred for percutaneous closure of a secundum ASD. She was asymptomatic on admission. Physical examination revealed fixed splitting of second sound and a mid-systolic murmur in the second right intercostal space. Electrocardiogram

showed sinus rhythm and right bundle branch block. Echocardiography revealed left atrial isomerism enlarged right heart cavities, secundum ASD with septal aneurysm, and interrupted IVC with azygos continuation. The initial plan was to close the defect percutaneously through azygos continuation by transesophageal echocardiography. A 6-F sheath was inserted into the femoral vein. Transesophageal echocardiographic colored Doppler examination showed a 23.9-mm secundum ASD with sufficient rims, except for the aortic rim, which was 3.5 mm in diameter. It was possible to place the exchange guidewire into the left upper pulmonary vein through a 6-F Judkins right catheter via the femoral vein. However, it was not possible to advance the sizing balloon catheter (AGA Medical Corp., Golden Valley, MN, USA) through the azygos vein continuation using this route. Therefore, a 6-F sheath was inserted into the jugular vein. A Judkins left catheter was advanced to the left-sided atrium through the jug-

#### Abbreviations:

ASD Atrial septal defect  
IVC Inferior vena cava

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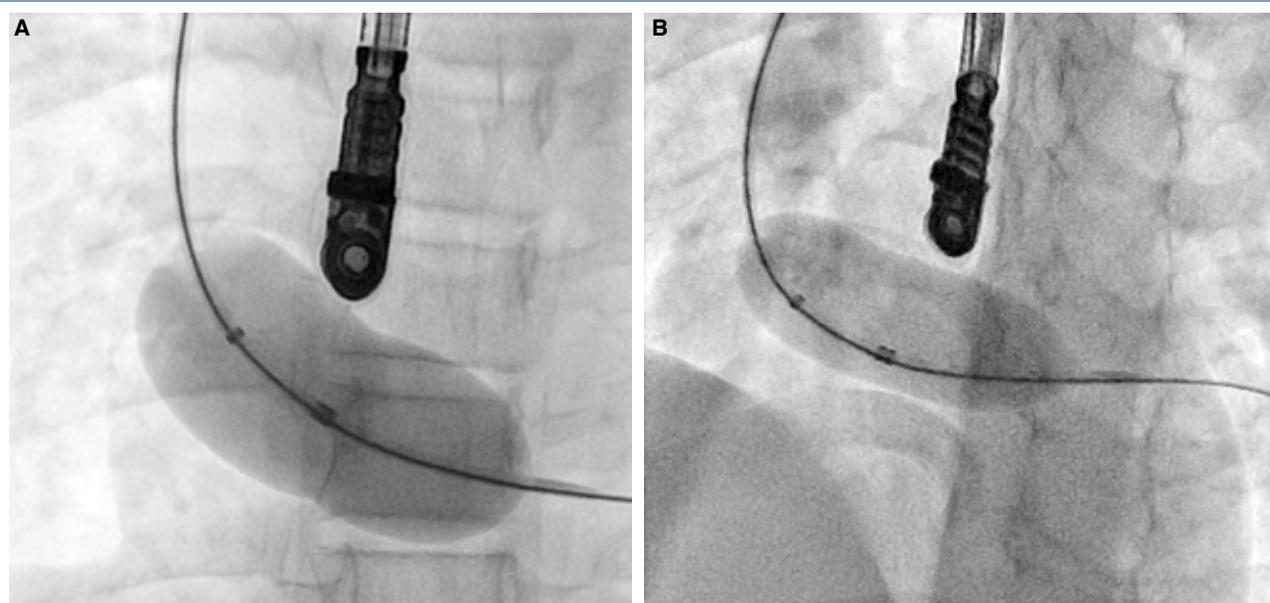
**Figure 1.** Azygos continuation of the inferior vena cava.

ular vein, superior vena cava, and right-sided atrium route, then placed into the inferior pulmonary vein with guidance of a hydrophilic guidewire, which was then exchanged for an extra-stiff guidewire. Over the guidewire, a 24-mm sizing balloon catheter was glided across the defect in order to measure the stop flow and stretched sizes of the defect. The stop flow and stretched sizes were 22.7 mm and 25.6 mm, respectively. A 24-mm Ceraflex septal occluder (Lifetech

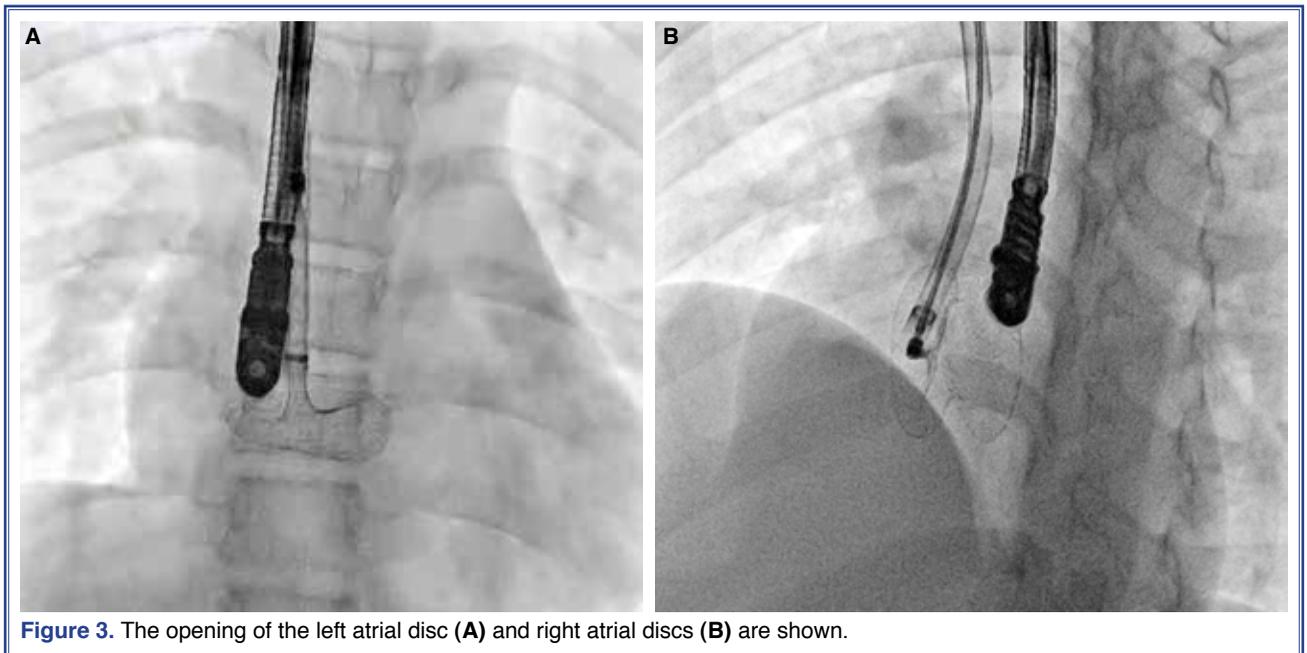
Scientific Corp., Shenzhen, China) and 12-F delivery sheath were selected to attempt closure. Attempts at deployment of the device failed due to prolapse of the retention disks. Left atrial disk tended to be nearly perpendicular to the septum when it was pulled against the defect to capture the rims. Therefore, a Fustar steerable guiding catheter (Lifetech Scientific Corp., Shenzhen, China), which can be bent by a handle located on the sheath to create a better angle, was used in order to easily anchor and deploy the device. The 24-mm Ceraflex septal occluder was loaded and delivered to the left-sided atrium through the 12-F steerable curved long sheath and was easily implanted due to perfect alignment of the device with the interatrial septum, which was established by use of a steerable guide catheter. The patient was discharged the following day with oral aspirin and experienced no problems during 10-month follow-up.

## DISCUSSION

Interrupted inferior vena cava with azygos continuation can render occluder delivery difficult or impossible. Several routes for closure of atrial septal defects in patients with interrupted IVC have been reported.<sup>[4-8]</sup> This anomaly can occasionally complicate transcatheter intervention by rendering more difficult the usual direct route from the femoral venous access. This has been overcome by use of several reported



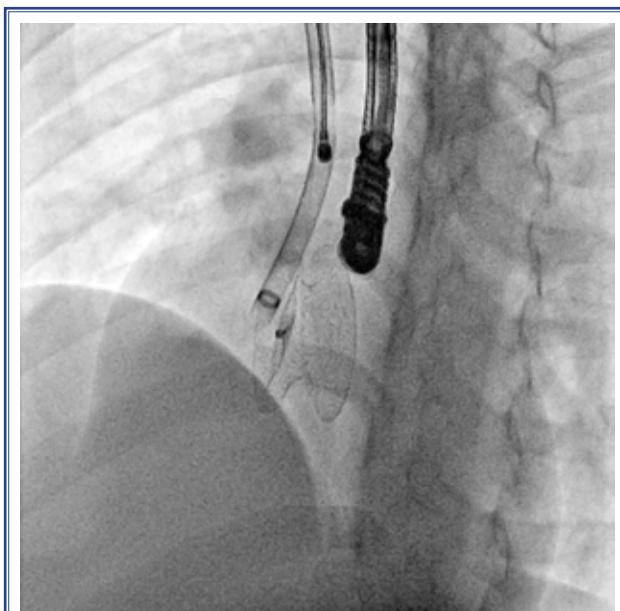
**Figure 2.** Over the guidewire, a 24-mm sizing balloon catheter was glided across the defect in order to measure the stretched size (A) and stop flow size (B) of the defect. The stop flow and stretched sizes were 22.7 mm and 25.6 mm, respectively.



**Figure 3.** The opening of the left atrial disc (A) and right atrial discs (B) are shown.

techniques.<sup>[9,10]</sup> Flosdorff et al. reported a successful transfemoral closure in a 3-year-old boy with an 8x10-mm secundum ASD and interrupted IVC with azygos continuation.<sup>[4]</sup> The authors were the first to demonstrate that percutaneous closure of ASD via azygos continuation is practicable in children. How-

ever, the procedure may not be performed when the defect is larger, as a larger delivery catheter is needed, and it may be impossible to advance a larger catheter through the azygos vein. It would be additionally challenging to perform deployment and alignment maneuvers, which are usually crucial for percutaneous closure of large ASDs, with large delivery sheaths through the azygos vein. Transhepatic approach for closure of secundum ASD has also been described as an option in pediatric patients. However, dangerous and challenging complications, such as hepatic abscess, retroperitoneal bleeding, and hemobilia have been reported.<sup>[8,12,13]</sup> Therefore, the transhepatic route was not selected in the present case. It was agreed that complications associated with transjugular access would be easier to handle.



**Figure 4.** 24-mm Ceraflex septal occluder was loaded and delivered to the left-sided atrium through a 12-F steerable curved long sheath, and was easily implanted due to perfect alignment of the device with the interatrial septum, established by use of a steerable guide catheter.

In cases of azygos continuation, either through the femoral or jugular vein, it can be difficult to pass through the ASD to the left atrium and place an extra-stiff guidewire in the upper pulmonary vein.<sup>[14]</sup> Furthermore, the guidewire already placed in the upper pulmonary vein may fall back into the left atrium when delivery catheter is advanced to the left atrium.<sup>[7]</sup> Use of a Judkins left catheter was convenient for passing the defect through the jugular vein. The primary curve of the left Judkins catheter engaged the base of the right-sided atrium, and the secondary curve directed the guidewire to the left-sided atrium. Passing through the defect was made easy by this

double-curved shape. The guidewire was placed in the left-lower pulmonary vein. We believe this approach provided a more stable and straight pathway for the delivery catheter to be advanced into the pulmonary vein, and reduced the risk of the guidewire falling back into the left- and right-sided atria, respectively.

Even with experience, it is not always possible to establish perpendicular orientation of a delivery system to the atrial septum using conventional approaches and methods. In the present case, device and total septum measured 24 mm and 40 mm, respectively. It was acknowledged that different maneuvers, slide-out techniques including right or left upper pulmonary vein approaches would have been impossible to perform due to large detention discs and limited space in the left atrium. Useful methods of overcoming this problem have been described; use of a Hausdorff sheath is a good means of reaching the desired orientation of devices to the septum. In addition, use of ancillary balloon catheter, modified Cook sheath, and steerable guide catheter in adult patients have been described by Flores et al.,<sup>[1]</sup> Spies et al.,<sup>[2]</sup> and Nounou et al.,<sup>[11]</sup> respectively.

A steerable guide catheter was selected in the present case. The Agilis introducer is primarily used in electrophysiologic procedures, and is available in different curl sizes, allowing access to various sites in the heart that are difficult to reach. Use for percutaneous closure of complex secundum atrial septal defects makes sense, as establishing parallel orientation of a device to the septum is challenging in most such cases.

To our knowledge, the ASD presently described was the largest to have been closed percutaneously through the jugular vein in a child. It was found that transjugular use of a steerable guide catheter for percutaneous closure in a pediatric patient with a large ASD and interrupted IVC was feasible and convenient.

**Conflict-of-interest issues regarding the authorship or article: None declared.**

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**Keywords:** Atrial septal defect; closure with internal jugular route; steerable delivery system.

**Anahtar sözcükler:** Atriyal septal defekt; juguler ven yoluyla kapatma; yönlendirilebilir taşıyıcı sistem.