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## Brief Report

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### Abstract

Interruption of the inferior caval vein complicates device closure of atrial septal defects. We present a case where a simplified technique was used: from right jugular access the delivery system was directly engaged into the left atrium, where the entire septal occluder was deployed. Both discs were aligned with the interatrial septum, after which the right disc was recaptured and re-deployed in the right atrium under tension. This technique will allow device closure of atrial septal defects from the upper caval vein in selected cases.

## Introduction

Interruption of the inferior caval vein (sometimes referred to as azygos continuation) is a rare congenital variation found in 0.1–0.6% of the population. It occurs either isolated or in combination with CHDs. In itself, lower caval vein interruption does not cause clinical problems. The combination of inferior caval vein interruption with a secundum atrial septal defect-II has been described.<sup>1</sup>

Device closure is an established treatment of atrial septal defect-II and is usually performed from the lower caval vein through femoral vein access. Due to the shape of the delivery systems, closure from the upper caval vein through jugular access is deemed more difficult. Different techniques have been described, ranging from modification of the sheath to deployment of the device along a stiff guidewire placed in the left upper pulmonary vein.<sup>1–7</sup>

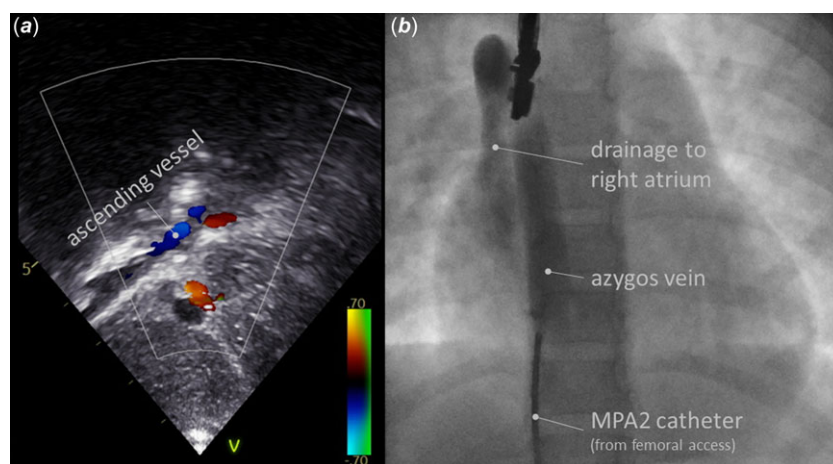
We report a simplified approach, whereby the entire device was deployed from jugular access in the left atrium and aligned to the intra-atrial septum, after which the right disc was recaptured and re-deployed in the right atrium.

## Case report

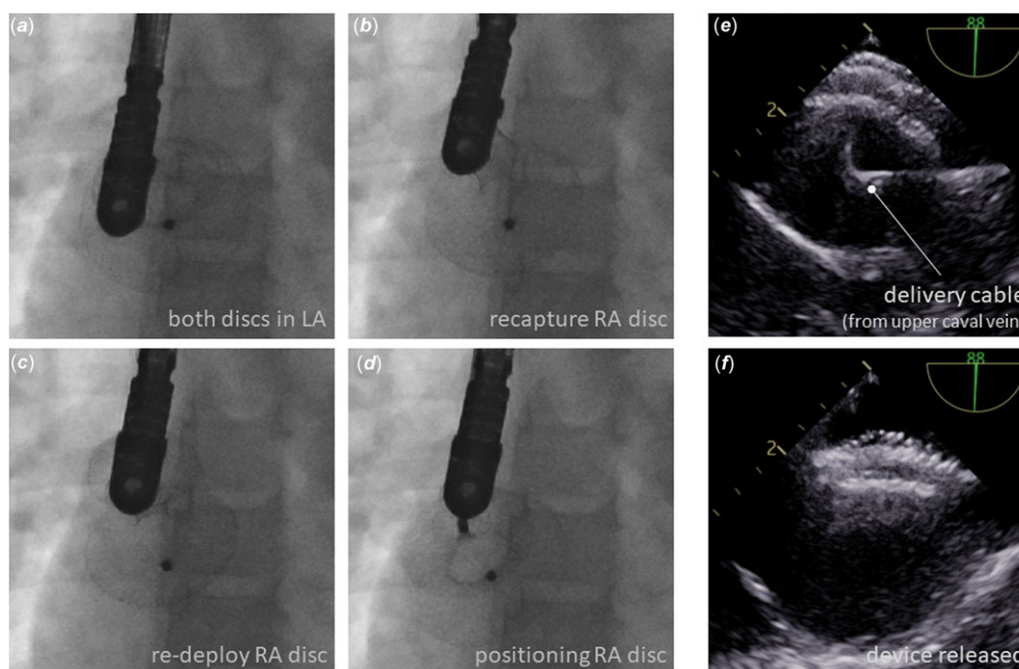
A 4-year-old girl (15 kg) was referred for closure of a secundum atrial septal defect, which was centrally located and on echocardiography measured 15 mm. There were adequate rims. Interrupted lower caval vein was suspected (Figure 1a). The procedure was carried out under general anaesthesia and heparin 100 IU/kg to achieve an activated partial-thromboplastin time (APTT) of > 200 s, which was monitored throughout the procedure. Through femoral venous access, interruption of the lower caval vein was confirmed by angiography (Figure 1b).

Due to the size of the child progression of a delivery sheath through the azygos vein into the superior caval vein was deemed unfavourable. Right internal jugular venous access was established, via which the defect was easily crossed with the MPA catheter. An exchange was made for an 8F Amplatzer Trevisio 45° delivery system (9-ATV08F45/80, Abbott, Plymouth, USA). It was not possible to reach a stable wire position in the left heart, therefore, the delivery system was carefully advanced directly into the left atrium. An 18 mm Amplatzer Septal Occluder (9-ASD-018, Abbott, Plymouth, USA) was then completely deployed within the left atrium paying meticulous attention to avoid interference with the mitral valve. With gentle pull on the delivery cable the entire device was fully aligned to the interatrial septum (Figure 2a). The right disc was then recaptured keeping the left disk in contact with the interatrial septum (Figure 2b). Keeping the tension on the Trevisio wire, the right disc was re-deployed in the right atrium (Figure 2c–d). With release of the tension, the device was configured nicely and adequate position was confirmed echocardiographically (Figure 2e). With release, tension on the interatrial septum was fully relieved and device position was excellent (Figure 2f). Transthoracic echocardiography confirmed good position the next day. The child was discharged in good clinical condition.

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**Figure 1.** *a)* Echocardiography suggestive of a para-aortic ascending vessel. *b)* Angiography: confirmation of azygos continuation by contrast injection.



**Figure 2.** *a-f)* Fluoroscopic images of the deployment of the device. *a)* Alignment of the device to the interatrial septum; both discs within the left atrium. *b)* Right atrial disc recaptured into the sheath in the right atrium. *c)* Right atrial disc re-deployed in the right atrium. *d)* Right atrial disc positioned at the right side of the interatrial septum. *e-f)* Echocardiography of release of the device. *e)* Delivery cable coming from the superior caval vein with an acute angle generating tension on the device. *f)* After release the device configures in normal shape and lines up with the interatrial septum.

## Discussion

Device closure of secundum atrial septal defect's from the superior caval vein can be challenging. Our simplified approach made use of the mechanical properties of the most recent delivery system (Trevisio) of the Amplatzer Septal Occluder.<sup>8</sup> Whilst deployment of the entire device within the left atrium could have been achieved with any delivery system, alignment with the interatrial septum requires a flexible delivery cable, as the angles of the septum differ between a jugular and a femoral access. Once the right disc was recaptured and re-deployed, it was important to be able to release some of the wire tension to allow the septum to configure more naturally, allowing echocardiographical assessment of device configuration. Devices from other manufacturers with flexible delivery systems can potentially also be used in the described manner, but we haven't found any report in the literature yet. A

stiffer delivery cable might have interfered with device configuration prior to release, independent from the type of connection of the cable to the device. A traditional wiggle manoeuvre is not feasible from the upper caval vein.

In previously reported techniques, it was attempted to reach the left upper pulmonary vein with the delivery sheath in order to deploy the device directly in line with the septum.<sup>2,3</sup> This requires substantial modifications of the delivery process and can result in tension on the device prior to release. The latter was also the case during a procedure reported by Flosdorff et al., who closed an atrial septal defect-II via femoral access, through the azygos vein, in a 3-year-old boy.<sup>4</sup> A venoarterial wire loop with over-the-wire technique to deploy the device has also been described.<sup>5</sup> This again complicates the procedure as the alignment between device and septum is difficult to achieve. Partial device deployment within the

left ventricle has been reported, too, in order to align the device against the septum, but this carries a risk of damage to the mitral valve apparatus.<sup>6</sup>

Sheath modifications for both jugular and femoral approaches have been described.<sup>1,7</sup> Although sheaths can be shaped outside the patient, this can complicate the procedure. Furthermore, in our own experience, inserting the sheath over a stiff guidewire usually configures the sheath to the shape of the guidewire, and after removal of the wire, the sheath tends to regain its original shape.

In our patient, no sheath modifications were necessary, as the interatrial septum was directly crossed with the angled delivery sheath. Our approach of deploying the entire device within the left atrium and pulling it against the left side of the interatrial septum overcomes above described difficulties, but of course requires a left atrium big enough to accommodate the whole device. The described technique of deploying the whole device in the left atrium and aligning it with the septum, recapturing the right disk and re-deploying it in the right atrium, could also be performed in cases with difficult rims from femoral access.

### Conclusion

We describe a simplified method for closure of atrial septal defect-II via jugular access with an Amplatzer Trevisio 45° delivery system. The delivery system is directly advanced over the interatrial septum defect, where entire device is deployed in the left atrium and aligned with the septum. Then, the right atrial disk is recaptured and re-deployed in the right atrium. This technique may be applicable for other selected cases of atrial septal defects without adequate inferior vena cava access.

**Competing interests.** None of the authors has any conflicts of interest.

**Consent statement.** Parents gave consent to publish the data.

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