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Brief Report

Occlusion of anomalous systemic arterial supply in Scimitar syndrome using the new Amplatz vascular plug IV

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Abstract Scimitar syndrome is a rare condition often with a separate systemic arterial supply from the abdominal aorta. Occlusion of this systemic arterial supply is frequently performed, though it can be difficult in small patients or in those with tortuous vessels. This case documents use of the new Amplatz vascular plug IV for arterial occlusion. It has major advantages in being able to deliver the device through a 4F catheter without the need to upsize to a dedicated delivery sheath. This is particularly appealing to paediatric practice or to those with difficult anatomy in older patients.

Keywords: Intervention; cardiac catheterization; Scimitar syndrome

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A 7-month-old child presented on day 2 of life with tachypnoea. A diagnosis of Scimitar syndrome was made echocardiographically. Scimitar syndrome is a rare condition of an abnormal lung segment with anomalous venous drainage usually to the right atrium–inferior caval vein junction,

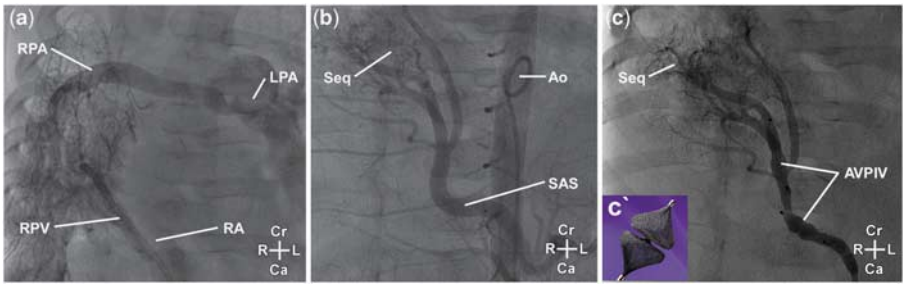

Figure 1.
*(**a–c**) Antero–posterior angiograms taken in a right anterior oblique projection (30 degrees) to show occlusion of systemic supply to the sequestered lung using the new Amplatz Vascular Plug IV (AVPIV) device (AGA Medical Corp., Golden Valley, MN, USA). (**a**) The catheter is traversing the inferior caval vein (IVC) to reach the Scimitar vein (right pulmonary vein, RPV). The Scimitar vein drains anomalously to the inferior caval vein–right atrium junction and is stenosed at this point. A wedge hand injection of layered contrast is used to define the abnormal sequestered lung right back to the pulmonary artery bifurcation. There is no venous return to the left atrium. (**b**) The pig-tail catheter lies in the aorta and a large anomalous vessel (systemic arterial supply (SAS)) arising adjacent to the superior mesenteric artery is seen. The vessel trifurcates before supplying the sequestered lung (Seq). (**c**) Embolisation with two Amplatz Vascular Plug IV devices one deployed at the bifurcation (6 millimetres) of the anomalous vessel and the other more proximally (5 millimetres; **inset figure c′**) image of Amplatz Vascular Plug IV device deployed. Cr, cranial; Ca, caudal; R, right; L, left; LPA, left pulmonary artery; Ao, descending aorta.*

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frequently with a separate systemic arterial supply from the abdominal aorta.[1,2] It is often associated with dextroposition of the heart, hypoplasia of the right pulmonary artery, and atrial septal defect or more complicated structural cardiac disease. Treatment of Scimitar syndrome is controversial though occlusion of the anomalous systemic arterial supply is frequently performed in order to encourage compensatory growth of the remaining normal lung tissue and to reduce both the associated left-to-right shunt and also the risk of chronic and recurrent infection in the abnormal lung in later life[3–5] (Fig 1).

This is the first reported case of using the new Amplatz Vascular Plug IV device (AGA Medical Corp., Golden Valley, MN, USA) for arterial occlusion and it has several advantages, particularly in difficult cases. They can be delivered easily through several 0.038 4F catheters, which, as in this case, took some time to manipulate into position with obvious reluctance to exchange for a delivery sheath. They also allow the entire procedure to be performed through a 4F arterial sheath attractive to paediatric intervention. Successful occlusion of the vessel was confirmed the following day echocardiographically.

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