Assistance Publique
Hôpitaux de Marseille

"Left subclavian artery to left superior vena cava fistula in a neonate: from pre-natal diagnosis to percutaneous closure"

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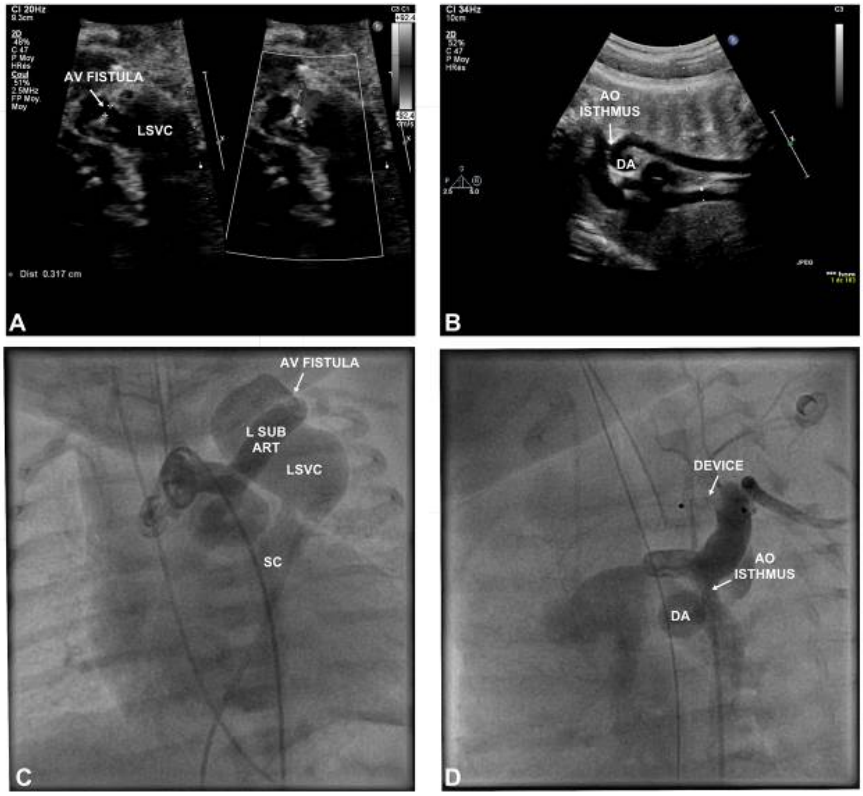
Introduction: Congenital arterio-venous fistulae involving the systemic thoracic vessels and in particular the subclavian artery and vein, innominate veins or superior vena cava are rare. Neonatal congestive heart failure is frequent. To our knowledge, prenatal diagnosis has not been reported so far. **Surgical ligation is the usual treatment but the broad spectrum of devices that are currently available for percutaneous closure offer a concrete alternative.**

Case Report: A **40-y-old woman** was referred **at 19 weeks** of pregnancy for management of a **foetal intra-thoracic mass**. Foetal echocardiography revealed a fistula between the left subclavian artery and an **extremely dilated left superior vena cava** (LSVC) (1A), a **large and tortuous ductus arteriosus** (DA) and **marked aortic isthmal hypoplasia** (1B). Prenatal MRI confirmed the diagnosis. The baby was born at term.

After postnatal echocardiographic confirmation, intravenous prostaglandins were started to maintain the DA patent. Cardiac catheterization was performed on day 6 of life from femoral arterial and venous accesses. Aortographies showed a very dilated proximal left subclavian artery, a short and restrictive fistula (3 mm) and a very dilated LSVC (1C); isthmal hypoplasia was confirmed but without significant peak-to-peak gradient (5 mmHg). The fistula was easily crossed retrogradely and closed with an Amplatzer® ADO II 4 x 4 mm device with **no residual shunt on angiography** (1D) **and echocardiography.**



Legend: Fig. 1 ***At the top: foetal echocardiography showing the fistula between left subclavian artery and LSVC (A), hypoplasia of aortic isthmus and the large ductus arteriosus (B); at the bottom: aortic angiography of the fistula before (C) and after (D) deployment of the device.***

Prostaglandin infusion could be weaned. Clinical evolution was uneventful without significant blood pressure gradient. The baby was **discharged 4 days after the procedure**. Clinical follow-up at **2 months is favorable** with only mild isthmal hypoplasia and obstruction, but **no arterial hypertension**.

Conclusion: Precise prenatal assessment of congenital arterio-venous fistulae involving systemic thoracic vessels **is possible.** It allows **rapid postnatal treatment before development of congestive heart failure.** Transcatheter occlusion is **a safe and effective alternative to surgery**. Aortic coarctation resulting from foetal diminished flow through the isthmus may be associated.

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