Cardiology in the Young

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Percutaneous permeabilisation with electrosurgery of intrahepatic inferior vena cava agenesis in a 12-year-old boy

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Brief Report

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Abstract

We describe a 12-year-old boy with agenesis of the intrahepatic segment of the inferior vena cava (a rare congenital anomaly) in whom an electrified coronary guidewire with electrocautery was used percutaneously to perforate and open the atretic segment, followed by stent implantation. This technique may be a safe and feasible option for paediatric patients, offering a therapeutic alternative for complex cardiac anatomies.

Case report

We report an asymptomatic child from Colombia whose immigrant screening bloodwork showed a slight elevation of liver enzymes (gamma-glutamyl-transferase, aspartate aminotransferase, and alanine aminotransferase values of 111, 53, and 51 U/L, respectively). An abdominal ultrasound revealed hepatomegaly with dilated suprahepatic veins. An echocardiogram suggested an atretic intrahepatic inferior vena cava segment, which was later confirmed by CT angiography. The case was discussed and a diagnostic and ± therapeutic cardiac catheterisation was scheduled.

The initial angiography through the extrahepatic inferior vena cava showed an interruption of the inferior vena cava with no visible intrahepatic segment (Fig. 1a, Video-1). Blood from the inferior vena cava reached the right atrium via the superior vena cava through collaterals, including the azygos/hemiazygos systems and some small direct collaterals from the suprahepatic veins. Collateral vessels were insufficient thus causing an increased pressure in the inferior vena cava (12 mm Hg) as compared to the right atrium (4 mm Hg). From the jugular vein, a right atrium angiography was performed and showed a continuous floor with no venous drainage (Fig. 1b) at a 15 mm distance to the inferior vena cava. Thereafter, a Gaia-II coronary guidewire (Asahi Intecc, Nagoya, Japan) connected to an electrocautery was introduced at the inferior vena cava stump and used to progressively vaporise tissue until the right atrium. The coronary guidewire was then snared in the right atrium from the jugular access (Fig. 1c). Sequential dilation with coronary balloons (1.25, 3.5, and 5 mm) was then performed (Fig. 1d). Using a guide catheter, the right atrium was reached from the femoral venous access. The coronary guidewire was replaced by a 0.035′′ Terumo guidewire (Terumo, Tokyo, Japan), which was externalised through the jugular access creating a veno-venous loop. Over the Terumo guidewire, a 12F sheath was advanced to the right atrium. Finally, a 22 mm CP-stent (NuMED, Hopkinton, New York, USA) was implanted and dilated to 10 mm with a balloon without complications (Fig. 2a). An angiography from the extrahepatic inferior vena cava showed a good result (Fig. 2b, Video 2) and pressure normalisation. The patient was discharged on antiplatelet therapy for 6 months, after which he remained asymptomatic and the stent echocardiographically patent. At four months, there was a reduction in the hepatomegaly size and the follow-up blood test showed a slight improvement in the liver enzymes (gamma-glutamyl-transferase, aspartate aminotransferase and alanine aminotransferase values of 67, 30, and 21 U/L, respectively). It is expected that in the future, the stent may require further dilation to accommodate the patient's somatic growth.

Discussion

Agenesis of the hepatic segment of the inferior vena cava is a rare congenital vascular anomaly with an incidence of <0.01%.[1,2] Most patients remain asymptomatic due to the establishment of compensatory collateral circulation (azygos and hemiazygos systems, and superior vena cava).[2] In cases of insufficient collateral circulation, complications related to increased venous pressure may develop, such as lower limb oedema, deep vein thrombosis, or chronic congestion-related

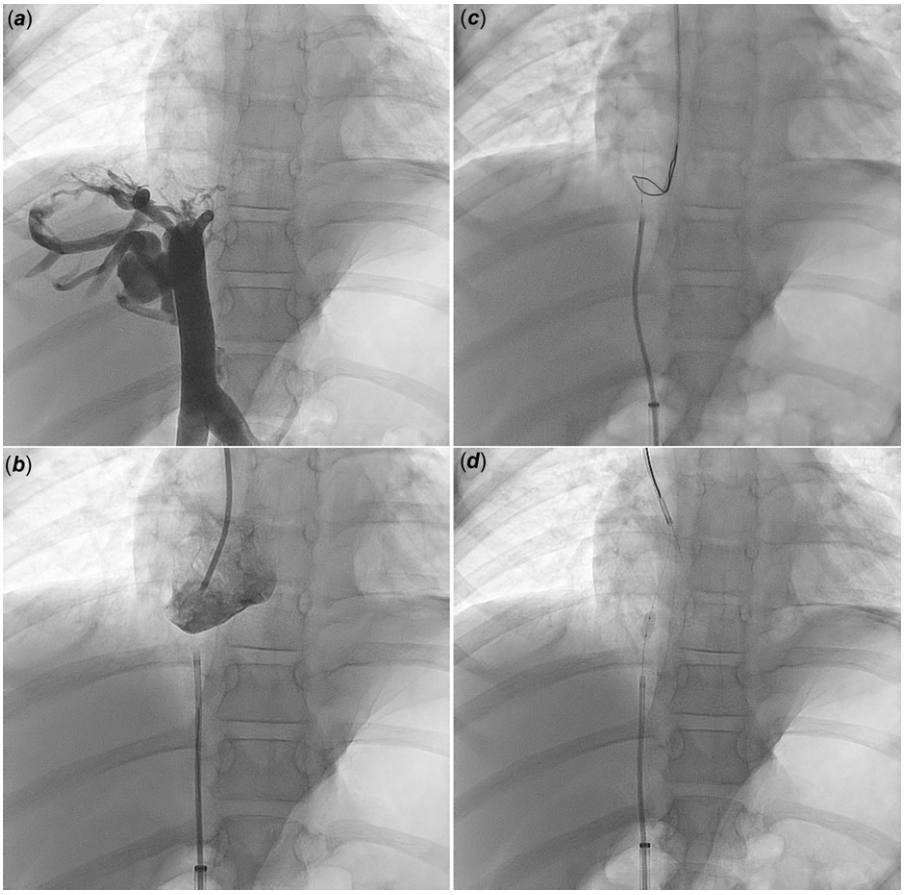

Figure 1. (***a***) Inferior cavography and collateral circulation. (***b***) Angiography in the right atrium. (***c***) Perforation with coronary guidewire into the right atrium. (***d***) Angioplasty of the newly perforated pathway.

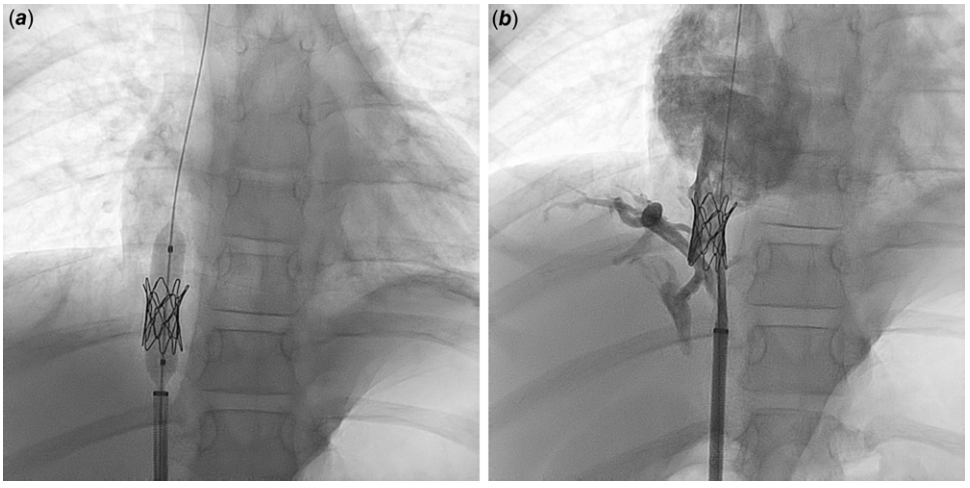

Figure 2. (***a***) Stent implantation. (***b***) Final angiography from the inferior vena cava.

liver disease.[3,4] In paediatric patients, most of these anomalies are diagnosed incidentally through MRI or CT angiography.[1–3]

Perforation using electrocautery combined with coronary guidewires represents an innovative and versatile tool within the therapeutic arsenal of interventional catheterisation, allowing the application of high-frequency energy to the guidewire to cross blood vessels (e.g., transcaval access, vascular recanalization), tissues (atrial septum), lacerate valve leaflets, or cardiac muscle.[5,6] With this technique, tissues in contact with the distal tip of the guidewire are vaporised and cut/dissected without causing nerve or muscle stimulation thereby avoiding pain, muscle contraction, or myocardial fibrillation.[5,7] The percutaneous connection between the superior vena cava and the right pulmonary artery (transcatheter Glenn), or the percutaneous creation of a communication between the descending aorta and the left pulmonary artery (Potts shunt), in a patient with suprasystemic pulmonary hypertension, has been successfully reported using transcatheter electrocautery.[5,6]

The main complication of recanalization, usually without significant consequences, is extraluminal perforation.[5,6] A limitation of the technique can be occlusions of considerable length or highly tortuous anatomies.[7]

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In the described case, an uncovered stent was chosen to avoid interferences with the drainage of the suprahepatic veins close to the area to be treated (Video 1). The reduced collateral circulation in the angiography and the elevated pressure relative to the right atrium suggested an insufficient degree of collaterality. After stent implantation, the pressure was normalised.

Due to the rarity of this condition, there is no established consensus regarding the treatment of choice. The options are endovascular and surgical approaches, which should be considered according to the experience of each centre.[2]

To the authors' knowledge, this is the first paediatric case in which an atretic segment of the inferior vena cava is percutaneously permeabilized de novo using electrosurgery and subsequent stenting.

Thus, percutaneous repermeabilisation of atretic vessels or those affected by chronic occlusion using an electrified guide with electrosurgery followed by stent implantation may be a feasible and safe option in the paediatric population.

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Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the institutional ethics committee of clinical investigation (Comité Ètic d'Investigació Clínica, Hospital Universitari Vall d'Hebron).

Informed consent: a written informed consent was obtained from the child's parents.

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