Congenital intrahepatic arterio-portosystemic venous shunt mimicking infracardiac total anomalous pulmonary venous connection.

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Learning objectives

- To illustrate the different angiographic aspects in patients with intrahepatic vascular connections between the hepatic artery, the portal vein and the hepatic veins are rare.
- Colour Doppler is a first-line investigation for intrahepatic shunt in early infants. The precise pathway of the shunt has to be confirmed by CT angiography. CT angiography has an important role to guide endovascular treatment.
- Transcatheter coil embolization is the standard management of congenital arterio-venous shunts, TAPVR needs surgery.

Background

Congenital intrahepatic arterio-portosystemic fistula is a high flow vascular malformation between the hepatic artery, portal vein and hepatic or systemic vein or combined forms (1). These abnormal rare communications can be observed in neoplasms (hemangioendothelioma), vascular malformations and infradiaphragmatic total anomalous pulmonary venous return (TAPVR) (2).

Hepatic arteriovenous shunts commonly are acquired (liver biopsy, transhepatic biliary drainage, transhepatic cholangiogram, surgery or trauma). Congenital hepatic arteriovenous shunts are rare and associated with hereditary haemorrhagic telangiectasia, Ehlers-Danlos syndrome and biliary atresia. Four types of intrahepatic shunts are possible: porto-systemic venous (portal vein to hepatic vein or vena cava), arterioporal (hepatic artery to portal vein) and arteriosystemic (hepatic artery to portal vein) (2,5). We describe a case of an abnormal intrahepatic communication between an aberrant feeding artery from the abdominal aorta, the portal vein and the vena cava inferior by a patent ductus venosus (Figure 1).

Ultrasound findings of congenital hepatic AV fistula:

- Cystic structure communicating with vessels (Color Doppler: flow)
- Hepatic artery enlargement with a low resistive index (RI)
- Increased flow velocities in the hepatic or portal veins (arterialized waveform)
- Aortic tapering at the level of the celiac trunk
- Signs of portal hypertension: splenomegaly, ascites, collateral vessels
Arterial-phase contrast-enhanced CT or MRI of congenital hepatic AV fistula (2):

- An abnormal vascular connection
- Marked enhancement of the feeding artery
- Early enhancement of the portal vein (or segmental branches) with the same attenuation or signal intensity of the aorta
- Aortic tapering at the level of the celiac trunk (under feeding artery)
- Signs of portal hypertension: splenomegaly, ascites, collateral vessels

Differential diagnosis congenital interhepatic arterioportal shunt

- Infracardiac total anomalous pulmonary venous return (TAPVR)
- Arterio-venous malformation
- Hypervascular hepatic tumour

Untreated shunts can lead to serious complications including portal hypertension, failure to thrive, right heart failure, portosystemic encephalopathy or gastro-intestinal bleeding. The primary treatment of arteriovenous fistula is to reduce the shunt. In the past the therapeutic option of choice for a symptomatic congenital hepatic shunt was surgical arterial ligation (4). With the development of endovascular techniques embolization of the feeding artery by transcatheter approach (less invasive) is currently possible and is therefore the preferred treatment (3,6). CT angiography has an important role to guide endovascular treatment. Franchi-Abella et al. (7) made guidelines for the treatment of congenital portosystemic shunt in children. Many devices are available including coils, Aga vascular plugs or glue for embolization of vascular malformations and fistulas depending upon the location and the size of the malformations. In our case we had to use multiple devices because of the thickness and the high-flow of the feeding artery.

Images for this section:
Fig. 1: Scheme shows an abnormal intrahepatic communication between an aberrant feeding artery from the abdominal aorta, the portal vein and the vena cava inferior by a patent ductus venosus present in our case.
Findings and procedure details

A 22-day-old boy with progressive respiratory distress and reduced growth was admitted to the paediatrics department of our hospital. There was no fever, rhinorrhea, cough, rash, vomiting or diarrhoea. Physical examination revealed an alert infant with mild tachypnea without retraction. He had a respiratory rate of 45-60 breaths/min, an oxygen saturation of 90-93% on room air and a pulse rate of 152-179 bpm. He had bilaterally normal breath sounds. A cardiovascular examination revealed a systolic ejection murmur III/IV at the left upper sternal border (ICR V) and capillary refill time shorter than 3 seconds. The liver was palpable 2 cm below the right costal margin, the spleen was not palpable. The patient had progressive respiratory failure requiring intubation and was admitted to the intensive care unit (ICU).

A chest x-ray demonstrated an enlarged cardiac silhouette and increased pulmonary vascular markings indicating increased lung circulation. A 12-lead electrocardiogram (ECG) was performed and revealed right axis deviation and right atrial enlargement.

Echocardiography showed a failure of all four pulmonary veins to make their normal connection to the left atrium. The four pulmonary veins drained into a tortuous tubular structure into the vena cava inferior resulting in drainage of pulmonary venous blood into the systemic venous circulation. The anomalous vein enters the vena cava inferior at an acute angle resulting in pulmonary venous obstruction. There was a severe pulmonary hypertension (115mmHg), right ventricular hypertrophy and a right-to-left shunt through a patent foramen ovale. Given the high pulmonary hypertension and the urgent setting, the patient was accepted for immediate correction of TAPVC of the inferior type. No further imaging assessment was necessary.

During the operation, the surgeon found a normal anatomy of four pulmonary vessels draining into a small left atrium. A process in the liver with one or more arteriovenous fistulas was palpated. During perfusion without ventilation the blood in the vena cava inferior kept a saturation of nearly 100% indicating a severe shunt. The surgeon needed more imaging by the suspicion of an arteriovenous malformation or fistula. Because of the severe pulmonary hypertension the surgeon decided to provide a good circulation (right-to-left shunt) and made a wide atrial septal defect by resection of the flap of the foramen ovale. Postoperative the patient was stable and monitored at the intensive care unit for further diagnostic work-up.

Bedside ultrasonography showed a cystic structure in the left liver lobe communicating with 3 enlarged tubular structures. Doppler US showed pulsatile flow indicating that an arteriovenous shunt must be present (Figure 2). Computed tomography (CT)
angiography revealed an intrahepatic arteriovenous fistula with a large feeder from the abdominal aorta above the level of the celiac trunk. The fistula had a connection with the portal venous system (left portal vein) and with the vena cava inferior through a patent ductus venosus (Arantius ductus). So, there was an early shunting of arterial blood into the portal vein and the vena cava inferior. No soft tissue masses were detected. A reduced aortic diameter below the celiac trunk, ASD and mild cardiomegaly were also noted. The portal vein diameter was normal. There were no signs of portal hypertension.

After multidisciplinary consultation it was decided to coil the arterial feeder by transcather approach through the femoral artery. Angiography shows a large feeding artery from the aorta abdominalis communicating with a large vascular space in the left liver lobe (figure 4A). Retrogradly through a 4-French sheet plugs were positioned in the proximal arterial feeder from the abdominal aorta. Control angiography showed decrease of the flow but persistent arterial shunt. The remaining shunt was again coiled. Control angiography showed persistent flow, thereafter glue was injected. Repeat post-occlusion angiography confirmed complete occlusion (figure 4B). No complications were encountered.

Post occlusion the pulmonary arterial pressure was significantly lower. Follow up US showed no further arterio-portal-venous shunting. The liver parenchyma had a normal sonographic appearance. At a later stage, the ASD have been closed.

Images for this section:

Fig. 1: Scheme shows an abnormal intrahepatic communication between an aberrant feeding artery from the abdominal aorta, the portal vein and the vena cava inferior by a patent ductus venosus present in our case.
Fig. 2: A. Ultrasonography showed a cystic structure in the left liver lobe communicating with 3 enlarged tubular structures. B. Doppler US showed pulsatile flow indicating that an arteriovenous shunt must be present. C. Transversal Doppler sonogram shows arterial waveform in the vena porta (arterialisation of the flow) consisting with arterioportal shunting.
**Fig. 3:** Sagittal CT angiography of the aorta show an aberrant feeding artery above the celiac trunk. The diameter of the aorta is larger above the feeding artery.
Fig. 4: A. Selective angiogram shows an aberrant artery from the aorta feeding a large vascular space drained by a left portal vein. The angiogram demonstrate early filling of the portal vein consistent with arterioportal shunt. B. Post-occlusion angiogram shows a complete occlusion of the feeding artery.
Conclusion

The radiologist has an important role regarding the differential diagnosis of vascular malformations. Color Doppler is the primary imaging modality to find vascular malformations in the liver. Accurate knowledge of imaging features could avoid unnecessary surgery. Transcatheter coil embolization is the standard management of congenital arterio-venous shunts, TAPVR needs surgery.

Personal information

References