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### When portal blood bypasses the liver: presentation and management of congenital Abernethy extra-hepatic shunts

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#### Introduction

Congenital porto-systemic shunts are rare and hard to diagnose. Clinical manifestation can vary from asymptomatic to extreme forms of cyanosis, pulmonary hypertension, liver insufficiency and mental retardation. Treatment options, depending on the type of abnormality and timing of detection, vary from (stepwise) occlusion to liver transplant. We report our experience.

#### Methods

Test balloon occlusion was performed to confirm portal pressure < 18 mmHg or portal-caval gradient < 8 mmHg. The shunt was then either closed or partially obstructed.

#### Results

5 patients presented between 2006 and 2017. Presenting symptoms were very heterogeneous: antenatal screening (2), cyanosis 75% (1), PMR & pulmonary hypertension by torrential arterio-arterial collaterals (1), and fortuitous in scimitar syndrome (1). The shunts were reached percutaneously either via the jugular or femoral vein (4) or by direct hepatic puncture (1). Four shunts were closed with a device in a one-step-procedure with Amplatzer Vascular or mVSD plugs (3) or a coil (1), depending on the size of the shunt. One patient failed the test occlusion; a very short landing site was determined (12 mm long, 10 mm stretched diameter); a flow-reductor implanted through an 8F sheath using a 6mm Occlutech Muscular VSD occluder (disks 13 mm) preperforated 5/16mm Bentley covered stent (image).



In 2 patients the shunt was completely closed with no residual flow. In one patient the largest shunt was closed but three collaterals became evident at closure requiring later closure; another patient had mini collaterals which remain monitored. In the patient with the flow-reductor, echo Doppler one month later, showed small portal veins appearing in the liver allowing later complete closure.

#### Conclusions

Porto-caval shunt can be managed percutaneously. Early diagnosis allows early and complete closure; this stresses importance of fetal screening. Treatment at late presentation with hypoplastic portal veins may require stepwise occlusion with a flow-reductor, hoping to avoid liver transplantation.