A Novel Technique of Ilio-Portal Venous Bypass to Relieve Symptoms of Pelvic Vein Congestion in a Patient with Inferior Vena Cava Agenesis

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Introduction

Inferior vena cava agenesis (IVCA) is a rare pathology that may remain latent for several years.1) Patients may tolerate this condition well as long as there is adequate collateral flow from the ilio-femoral veins towards the azygos and hemiazygos systems.2) Symptoms occur when deep vein thrombosis (DVT) affects these collateral pathways.3)

Case Report

A 37 year old male patient with IVCA has been treated in our department from lower limb insufficiency, C3 stage of CEAP (comprehensive classification system for chronic venous disorders), controlled by conservative treatment (previous anticoagulation, compressive stockings). He had no antecedent perinatal surgical or medical history. He was suffering from severe symptoms of lumbar and abdominal pain that were exacerbated by physical activity requiring opiate medication. He also experienced recurrent rectal bleeding, recurrent diarrhoea, haematuria and episodes of erection after physical activity.

Duplex ultrasound scan and computed tomography angiography (CTA) were performed. The patient had an agenesis of the infra-renal and supra-renal section of inferior vena cava (IVC) and, of the two common iliac veins with occlusion of the renal veins (Figs. 1A and 1B). Radical surgical approach was considered to reduce venous pressure in the lower limbs and the pelvis. We suggest an alternative approach: a bypass between the right iliac vein and the portal vein.

The right external iliac vein was chosen because the bypass in this location is short and straight. An abdominal approach was used with a midline incision towards the right inguinal region. The right external iliac vein was exposed at the more distal point in the retroperitoneal space and the portal vein in the intra-peritoneal space.

An autogenous graft (superficial right femoral vein) was used to conduct the bypass. Initially, we performed an end-to-side portal anastomosis then we did an end-to-side right external iliac vein anastomosis (Figs. 2 and 3). In order to enhance venous flow through the bypass an infrainguinal arterio-venous fistula was made.

The patient was followed up with clinical visits and duplex scanning at one month and then every six months for up to 36 months. Graft patency was also documented with magnetic resonance angiography (MRA) after 6 months. The majority of the patient’s symptoms resolved during follow up. The arterio-venous fistula closed spontaneously within three months. Three years following surgery, he had unrestrained professional and physical activity. This paper shows that the bypass is technically possible in specific clinical situations.
Discussion

Given the rarity of the IVCA, there are currently no guidelines for the treatment, and the conservative treatment was most often reported. Surgical treatment of IVCA has been reported for severe lower limb vein stasis.\(^4\)

Dougherty et al.\(^5\) describe a prosthetic bypass between the external iliac vein and the intra-thoracic azygos vein. The patient had a complete symptomatic relief at 30 months follow up period. Sagban et al.\(^6\) treated 15 patients with an heterogeneous pattern of the IVCA. A combination of surgical procedures was used and most of them involved a long prosthetic bypass replacing the missing part of the vena cava.

In our opinion, the ilio-portal vein bypass has several advantages; the right iliac vein is dissected in the retroperitoneal cavity and the portal vein in the intraperitoneal cavity. This permits the creation of a short bypass with less risk for twisting and occlusion. At the same time it allows the utilization of native material. We are not sure if a simultaneous arteriovenous fistula creation is always necessary.

The use of portal vein in cases of venous insufficiency has been demonstrated in experimental and clinical settings; Child and Glenn\(^7\) used a bypass to the portal vein in experimental settings for prevention of renal hypertension. Starzl et al.\(^8\) described a cava-portal transposition for the treatment of glycogen storage disease. And we have previously reported a similar reconstruction in the context of iatrogenic injury of the suprarenal IVC with good technical and clinical results.\(^9\) Also a cavo-portal or reno-portal bypass has been used in liver transplantation in the presence of portal vein thrombosis.\(^10-11\)
Conclusion

Patients with symptomatic IVC agenesis/obstruction should be considered for surgical treatment. We suggest ilio-portal venous bypass as an alternative if hepatic veins and the supra-hepatic vena cava are without obstructive lesions.

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Disclosure Statement

None.

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