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Review Article

Congenital Extrahepatic and Intrahepatic Portosystemic Shunts: Classifications

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Abstract: Portosystemic shunts occur in children and adults due to varied reasons. Congenital shunts and acquired shunts have different clinical implications and prognosis in either group of patients. Here we describe these shunts, their anatomic and pathogenic mechanisms of development and furthermore shed light on simple and clarified insights on this syndrome and its classifications.
Keywords: Cirrhosis, portal hypertension, portosystemic shunts, hepatic encephalopathy, congenital shunts.

INTRODUCTION

Porto-systemic shunts are congenital or acquired. They are either intrahepatic or extrahepatic in location and origin. Intra-hepatic porto-systemic shunts are rare occurrences and are mostly seen with underlying cirrhosis and portal hypertension and not uncommonly, congenital. Extrahepatic porto-systemic shunts are mostly congenital, and are rare in patients with underlying liver disease. The latter are classically known as Abernethy malformations [1]. In this short review, we discuss briefly, classifications of extra-hepatic and intra-hepatic porto-systemic shunts.

Embryology and Anomalies of Portal Venous System

Portal venous system develops between the 4th and 10th week of embryonic life. It is formed from the vitelline veins, a pair of vessels located over the anterior surface of the yolk sac. In the primitive liver (or septum transversum) these vessels are broken down into sinusoids. At the end of 4th week, the cross communications between the sinusoidal architecture form around the duodenum leading to connections between the two vitelline veins that later selectively involute to form the fully developed portal vein. The superior mesenteric vein is the successor of right vitelline vein and drains the primitive intestinal loop [2, 3].

Umbilical veins course through the septum transversum and lie lateral to the vitelline veins and also become fragmented due to the developing liver with connections to the hepatic sinusoids. Proximal part of both umbilical veins disappears and only left umbilical vein continues to drain blood coming from placenta to the liver leading to ductus venosus bypassing the sinusoidal plexus of liver. After birth, the left umbilical vein and ductus venosus are obliterated, forming the ligamentum teres and ligamentum venosum respectively. Excessive involution of the peri-duodenal vitelline venous loop or complete failure of the vitelline veins in establishing critical anastomosis with hepatic sinusoids or umbilical veins result in extrahepatic portosystemic shunt formation. Shunts are due to persistence of right vitelline vein (draining into retro hepatic inferior vena cava) or left vitelline vein (draining into inferior vena cava or right atrium above level of hepatic vein confluence). Inferior vena cava develops from the sub cardinal veins close to the portal vein. The persistence of subcardino hepatic communications has also been thought to produce congenital extrahepatic shunts. The basis for development of intrahepatic portosystemic shunts is the persistence of communication between the vitelline and omphalo mesenteric venous system and sinus venosus due to focal absence of sinusoid formation [4, 5, 6].

Extrahepatic Porto-systemic Shunts

Congenital extrahepatic porto-systemic shunts are when the porto-mesenteric blood drains into a systemic vein bypassing the liver through a complete or partial shunt. It is most commonly seen in animals, especially described in dogs. It is extremely rare in humans and was first reported in 1793 by John Abernethy in a 10 month old girl on post mortem study in whom, he found the termination of the portal vein in the into the inferior vena cava at the level of the renal veins. It is closely associated with focal nodular

hyperplasia, malignancies of the liver and congenital heart defects and clinically present with hepatic encephalopathy and in the long term, porto-systemic shunt syndrome, leading to hepatic dysfunction. Morgan and Superina classified congenital extrahepatic porto-systemic shunts into 2 types [7, 8]. Figure 1 shows an example of Abernethy malformation type 1b.

Intrahepatic Porto-systemic Shunts

Congenital intrahepatic porto-sytemic shunts are abnormal connections between branches of portal vein and hepatic veins, characterized as per Park classification into 5 types. The most common type of intrahepatic portosystemic shunt is a communication between the portal vein to an extrahepatic systemic vein such as perihepatic vein or inferior vena cava in a patient with portal hypertension secondary to cirrhosis. These are usually very small shunts and tend to be < 2mm in size and larger shunts are considered to occur congenitally. Cutaneous and hepatic hemangiomas are considered common associations with congenital intrahepatic posrtosystemic shunts [9, 10]. Figure 2 shows an example of intra hepatic porto-systemic shunting, Park Type 2.

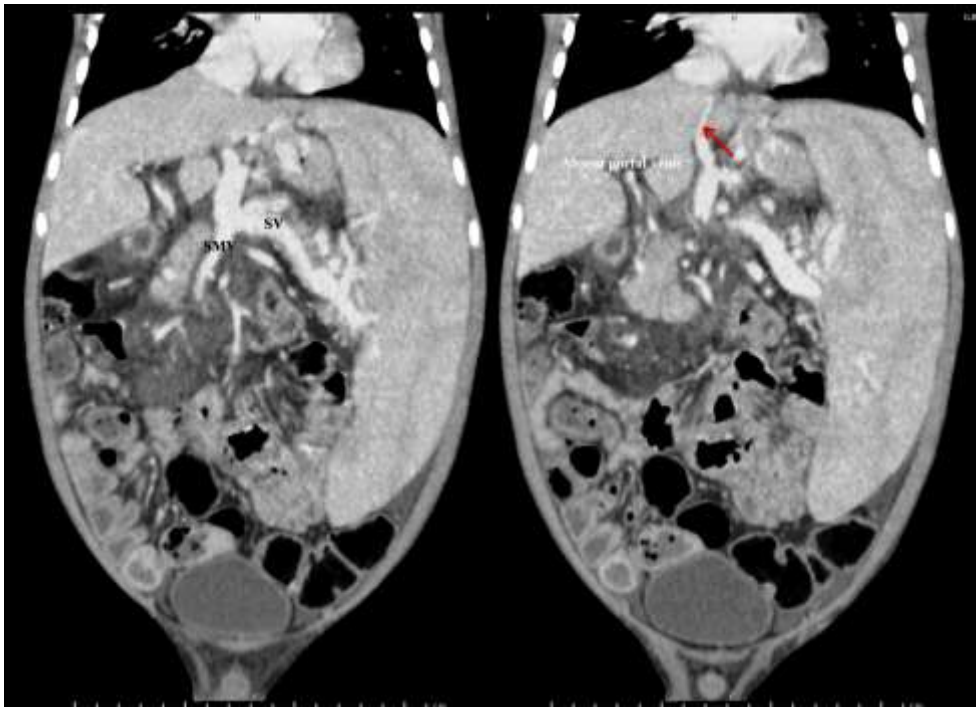

Fig 1: Extrahepatic Portosystemic Shunt: Computed venography imaging of the abdomen in a child showing Abernethy type 1b malformation. The splenic vein (SV) and superior mesenteric vein (SMV) do not form a portal vein (which is completely absent) and instead, forms a small thin shunt (red arrow) that drains directly into the right atrium

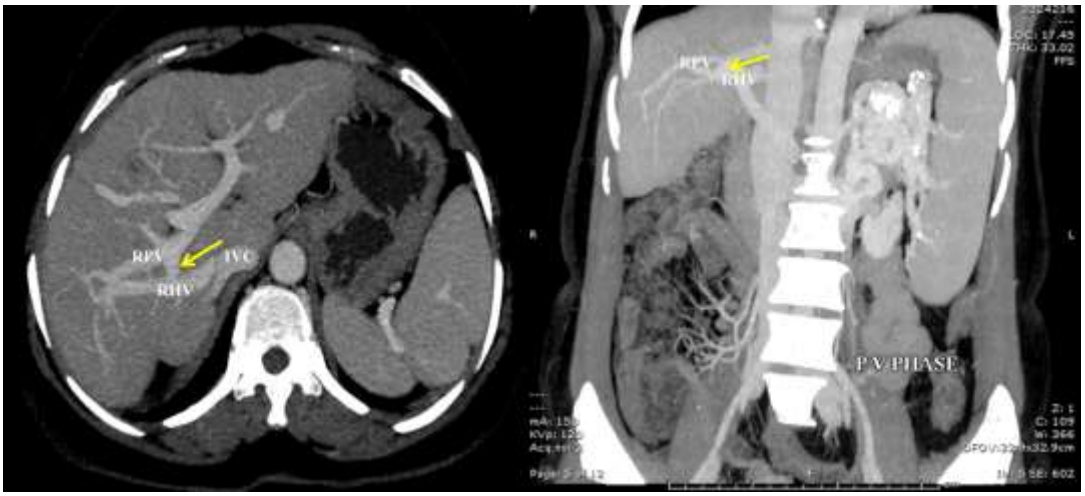

Fig 2: Intrahepatic Portosystemic Shunt: Computed venography with reconstruction in a cirrhotic middle aged woman showing a communication (yellow arrow, shunt) between the right branch of portal vein (RPV) and the right hepatic vein (RHV), suggestive of Park Type 2 shunt

CONCLUSION

Congenital intrahepatic and extrahepatic shunts are seen heterogeneously. The former is associated with cirrhosis, while the latter is most commonly congenital in origin. Clinical symptoms are varied and encephalopathy is one of the most commonly seen presentations. Understanding the anatomical basis of these shunts (Figure 3) is important in planning treatment profiling and clinical follow up of the patient.

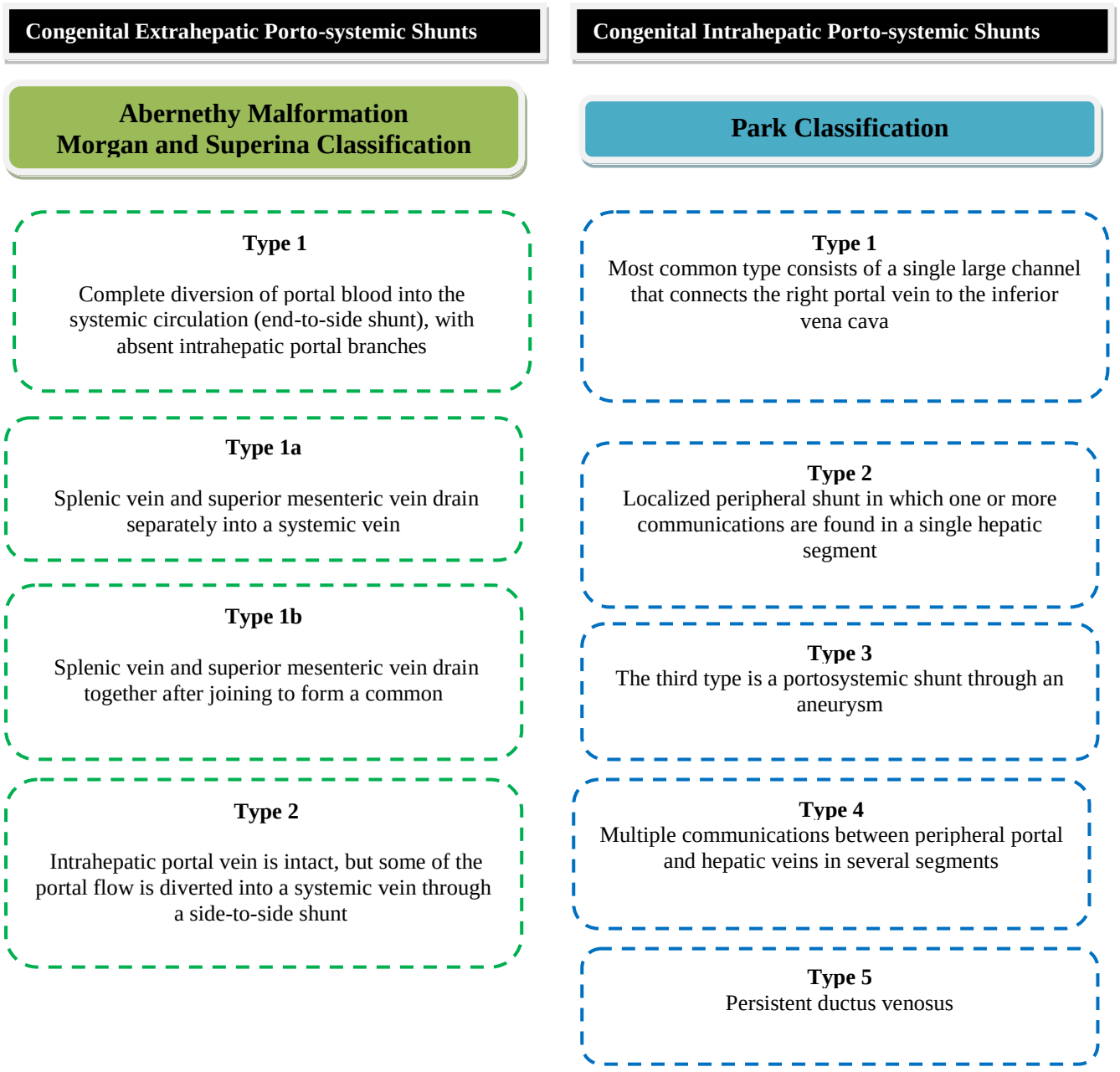

Fig 3: Classification of Congenital Extrahepatic and Intrahepatic Portosystemic Shunts

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