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Review Article

A REVIEW ON ABERNATHY MALFORMATION

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ABSTRACT

The first account of an absent portal vein and a congenital mesenteric-caval shunt was given by John Abernethy in 1793. Five new cases of congenital extra hepatic Porto caval shunts are described in this report .One end-to-side shunt with congenital absence of the portal vein (type 1 shunt). Four are side-to-side (type 2) shunts of which there have been only two previous reports. Type 1 shunts are associated with other congenital abnormalities and have only been recognized in girls. Five of these cases developed liver tumors. Type 2 shunts are rarer, and four of five have been boys. They are not usually associated with other anomalies. Encephalopathy may be associated with these shunts in adults, and surgical closure of the side-to-side shunts is therefore recommended. End-to-side shunts are not correctable but the abnormal anatomy may create a problem for hepatic transplantation, should this be necessary for associated disorders such as biliary atresia. "The Abernethy Malformation" would seem to be a suitable eponym for congenital extra hepatic Porto caval shunts.

Keywords: *Congenital Porto caval shunt, Encephalopathy, hepato pulmonary syndrome,*

INTRODUCTION

A port systemic shunt (PSS) also known as an Abernethy malformation or a liver shunt is vascular connection effect the body's circulatory system. In most people, blood from the stomach, intestines, spleen and pancreas drains into the liver through a large vein called the "portal" vein. A port systemic shunt (PSS) also known as an Abernethy malformation or a liver shunt is vascular connection effect the body's circulatory system. Abernethy malformation is a very rare congenital vascular malformation defined by diversion of portal blood away from liver.

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It is commonly associated with multiple congenital anomalies. Congenital extra hepatic port systemic shunt, also known as Abernethy malformation. Paraffin-embedded tissue, liver tumor resections, and liver biopsies were evaluated using eosin stains, reticule in, elastic, and immune histology chemistry for D2-40.The Abernethy malformation is an extremely rare anomaly of the venous system. It is named after John Abernethy, who first reported the anomaly in 1793.

The Abernethy malformation is divided into two types.

Type I anomalies may be further divided into subtypes A and B, defined as the superior mesenteric and spleen veins draining separately into the inferior canal vein in type IA and draining from a common trunk in type IB.

A type II shunt is defined as a malformation of the portal vein leading to perfusion of the liver via a partial shunt.

Earlier case reports focused on the widely variable forms of clinical presentation and the difficulties encountered in diagnosing the malformation. Malignant lesions such as hepatocellular carcinoma (HCC) also can develop, however, and they are most often described in patients with a type I shunt. Abernethy malformation complicated by the development of pulmonary arterial hypertension (mean pulmonary artery pressure 48mmHg), which had been diagnosed three years before and treated successfully with the endothelia receptor antagonist.

Computed tomography (CT) displayed the presence of a tumor, and magnetic resonance imaging (MRI) was performed for characterization of the lesion. The lesion was hypo intense on T1-weighted images and hyper intense on T2-weighted images in the venous phase, and arterial enhancement and washout were demonstrated, as is typical for a HCC .The presence of a shunt of 2 cm between the left extra hepatic portal vein and the inferior vena cava was confirmed.

Histological examination of the respected specimen revealed features of grade II fibrosis. A tumor with a diameter of 16cm was characterized as HCC with the presence of micro vascular invasion. A tumor-free surgical resection of 5mm was recorded.

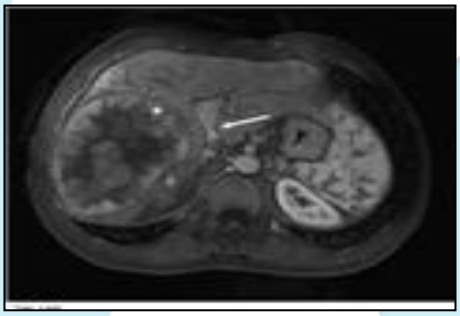

Figure 1:- Magnetic resonance angiography showing hepatocellular carcinoma and a Porto caval shunt

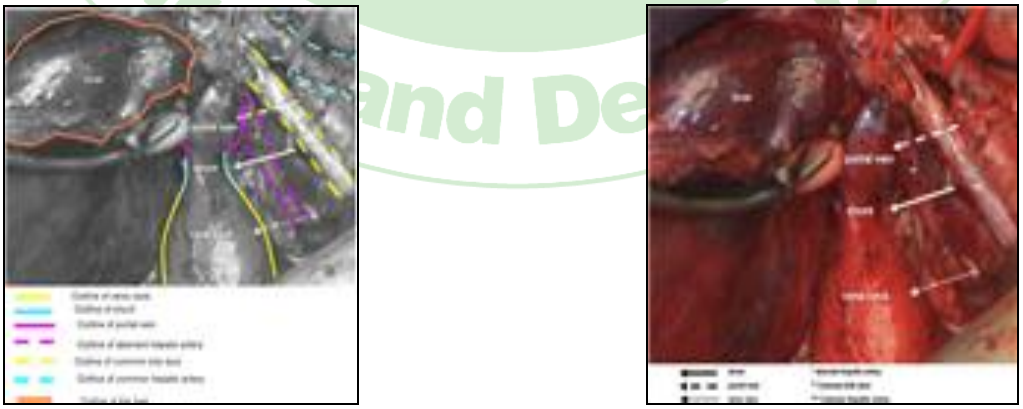

Figure 2:- The Porto caval shunt peri-operatively.

The follow-up of the patient consisted of a routine serum tumor marker AFP determination and contrast-enhanced CT or MRI at six months intervals starting three to six months after surgery. After nearly two years of follow-up, the patient is doing well and is free of HCC recurrence. His cardiac output is normalized, and his pulmonary hypertension is stable.

ANATOMY & PHYSIOLOGY OF LIVER

Anatomy of Liver

The liver is the largest organ of the human body , weighs approximately 1500 g, and is located in the upper right corner of the abdomen. The organ is closely associated with the small intestine, processing the nutrient-enriched venous blood that leaves the digestive tract. The liver performs over 500 metabolic functions, resulting in synthesis of products that are released into the blood stream (e.g. glucose derived from glycol genesis, plasma proteins, clotting factors and urea), or that are excreted to the intestinal tract (bile). A second blood supply to the liver comes from the hepatic artery, branching directly from the celiac trunk and descending aorta. The portal vein supplies venous blood under low pressure conditions to the liver, while the hepatic artery supplies high-pressured arterial blood. Blood from the hepatic artery on the other hand, originates directly from the aorta and is, therefore, saturated with O2. Blood from both vessels joins in the capillary bed of the liver and leaves via central veins to the inferior canal vein.

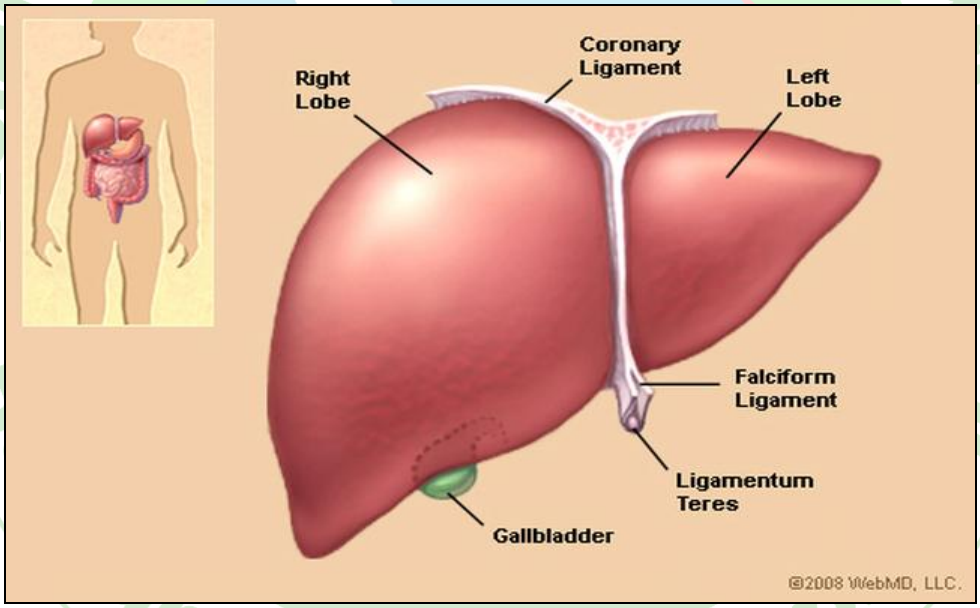

Figure. 3:- Liver

Basic Liver Architecture

The major blood vessels, portal vein and hepatic artery, lymphatic's, nerves and hepatic bile duct communicate with the liver at a common site, the hills. Here, in the sinusoids, blood from the portal vein joins with blood flow from end-arterial branches of the hepatic artery. Once passed through the sinusoids, blood enters the collecting branch of the central vein, and finally leaves the liver via the hepatic vein. The hexagonal structure with, in most cases, three portal canals in its corners draining into one central vein, is defined as a lobule.

The functional acnes can be divided **into three zones**:

- The per portal zone, which is the circular zone directly around the portal canal,
- The central zone, the circular area around the central vein, and
- A mid zonal area, which is the zone between the per portal and per central zone.

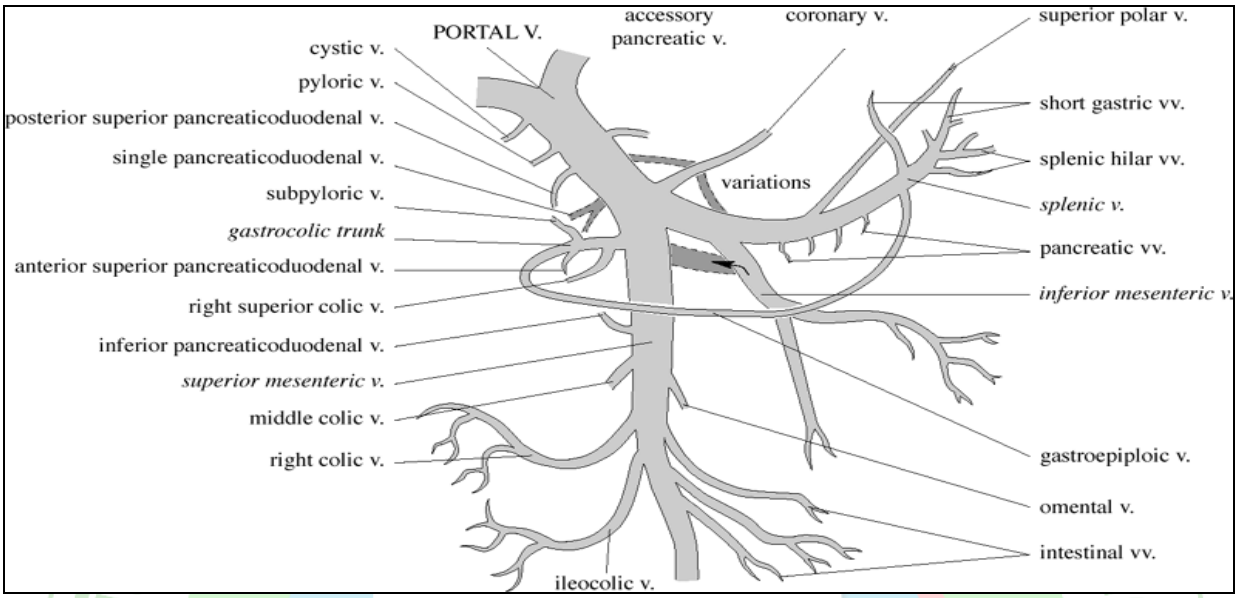

Figure.4:- Veins of Liver

PHYSIOLOGY OF LIVER

Pressure distribution

Blood pressure in afferent vessels and pressure distribution inside the liver, is essentially similar for most species. Pressure in the hepatic artery, originating from the descending aorta and the celiac trunk, is considered to be the same as aortic pressure. This includes a high palatial pressure between 120 and 80 mmHg with a frequency equal to the heart rate.

Flow distribution

Total human liver blood flow represents approximately 25% of the cardiac output, up to 1500 ml/min. Hepatic flow is subdivided in 25-30% for the hepatic artery (500 ml/min) and the major part for the portal vein (1000 ml/min). Assuming a human liver weighs 1500 g, total liver flow is 100 ml/min per 100 g liver. Comparing this normalized flow rate to other species, it can be concluded that total liver blood flow is 100-130 ml/min per 100 g liver, independent of the species. The hepatic artery originates directly from the descending aorta, and is therefore

saturated with oxygen. It accounts for 65% of total oxygen supply to the liver. One of the most important triggers for sphincter function is the need for constant oxygen sup-ply. If the rate of oxygen delivery to the liver varies, the sphincters will react and the ratio of arterial: portal blood flow alters.

Liver Preservation

Liver transplantation requires a period of preservation time, during which the liver after explanation is stored and transported outside its natural environment. Nowadays this time zone is kept between 6-15 hours of preservation, while the liver is cut from its life sustaining mechanisms, i.e. blood flow and oxygen supply, and consequently, ischemic damage will occur.

Three periods of ischemia are distinguished in a donor and transplantation procedure

- The first period is defined as the time in between clamping of the liver's vasculature

and start of the wash-out with ice-cold preservation solution during the procurement phase in the donor. During this period of ischemia the liver is still at body temperature, for which reason this is called warm-ischemia. During this warm-ischemic time, mainly damage to the hepatocytes occurs, and it is therefore very important to keep this period as short as possible. Especially in non-heart-beating donors (NHBDs).

- The second period of ischemia starts at the moment when the ice-cold preservation solution enters the microvasculature of the liver to wash-out the blood. A good wash-out of blood cells is important to obtain optimal and uniform perfusion and cooling of the liver. This second period takes place at 0-4±C, including the wash- out with ice-cold solution and cold storage preservation period, and is defined as the period of cold ischemia.
- The last period of this bridging ischemia is the period needed to complete the vascular anatomizes during implantation. This procedure could take 30-60 minutes, a period in which the liver lies in the abdominal cavity and isthus rearmed by temperature of the body.

ANATOMY & PHYSIOLOGY OF HEART

Anatomy of Heart

The main function of the cardiovascular system is to transport nutrients and oxygen to the entire body. The heart can be thought of as two pumps in series that send a fluid through a series of tubes that eventually return to the pump. One pump sends the blood to the lungs to pick up oxygen in the lungs, and the other sends the blood through the rest of the body. Eventually the blood returns to the heart and the process is repeated. Each "pump" in the heart is made up of two chambers; an atrium and a ventricle, giving the heart a total of four chambers. The atria are the smallest of the four chambers. The four heart chambers can be seen in Fig. The two atria are the top chambers, and the ventricles are on the bottom

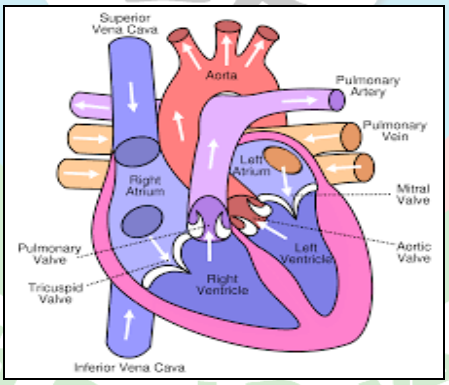

Figure. 5:- Heart

The heart uses a series of valves to ensure that blood flows in one direction into and out of the heart. Heart valves are made of tough, flexible tissue that is oriented in such a way that blood can only go through the valve in one direction. Blood flows in the heart chambers there are four heart valves .Two atria ventricular (AV) valves and two semi lunar valves. An AV valve is located between each atrium and ventricle, with the tricuspid valve on the right and the mitral valve on the left. When ventricular pressure exceeds atria pressure, the valve closes again. A semi lunar valve is located between the right ventricle and pulmonary artery (pulmonary valve) and the left ventricle and aorta (aortic valve).

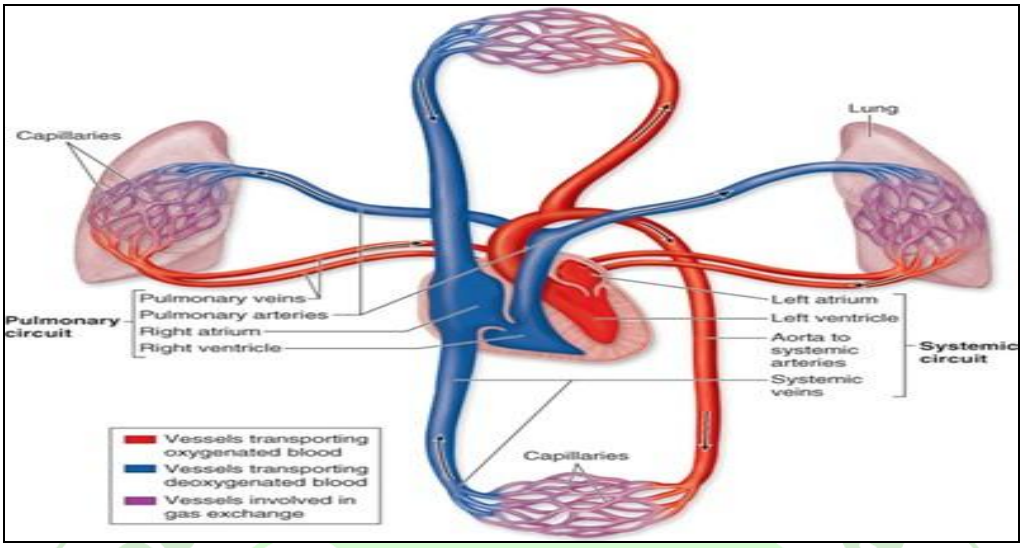

Figure. 6:- Circulation of Blood

Similar to the AV valves, when left or right ventricular pressure exceeds aortic or pulmonary artery pressure, the valve opens. When ventricular pressure decreases, the three cusps of the valves close, preventing blood from flowing back into the ventricle. The heart has two atria and two ventricles because there are two different blood circulation paths. The circulation path controlled by the right side of the heart is a low-pressure system known as the pulmonary circulation. Blood pressure in the systemic circulation is pustule due to contraction and relaxation of the heart. Pulsatility can be seen in the pressure and flow wave forms Aortic pressure also fluctuates when the heart beats.

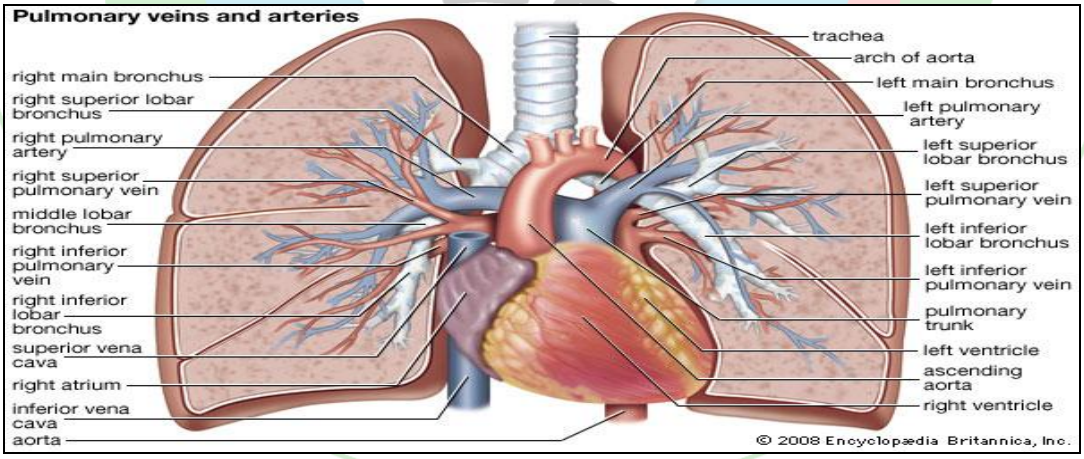

Figure.7:- Lungs

The aorta stretches as the ventricles pump blood into the aorta and the increased volume increases aortic pressure. Eventually the ventricle is emptied and the aorta contracts, causing a decrease in pressure. When ventricular pressure is lower than aortic pressure, the semi lunar valve closes and the walls of the aorta constrict.

Physiology of Heart

Pulsatility in the heart is caused by periodic contraction and relaxation, known as the cardiac cycle. The two main phases of the cardiac cycle are systole, or ventricular contraction (ejection) and diastole, or ventricular relaxation (filling). The entire cardiac cycle lasts approximately

800ms. Waveforms of Left Ventricular Pressure (LVP), Aortic Pressure (AOP), Left Atria Pressure (LAP), and Left Ventricular Volume (LVV)

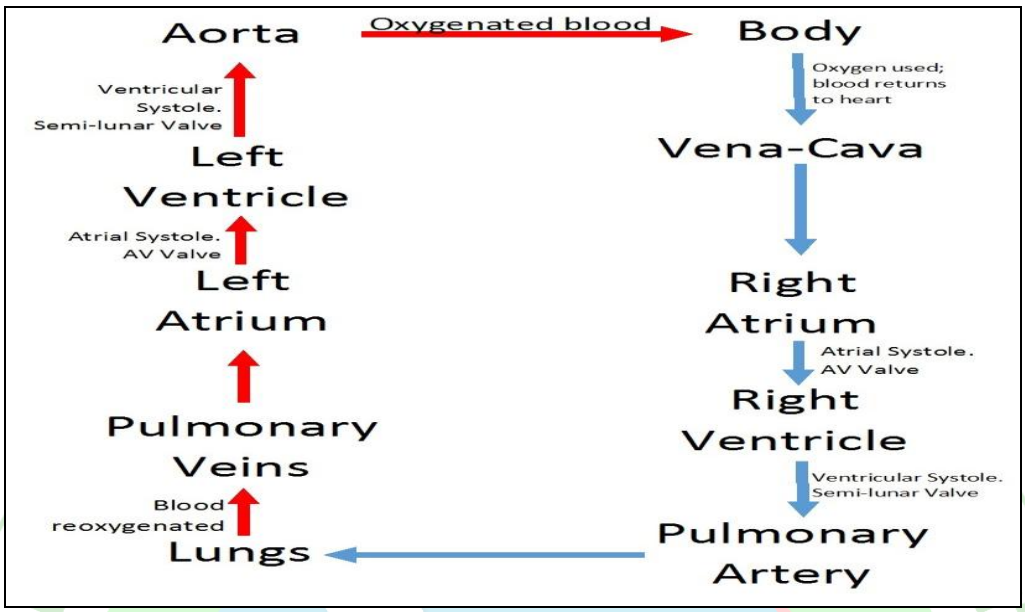

Figure.8: Flow Chart of circulation

LVP is less than AOP, causing the aortic valve to also remain closed. This point marks the beginning of the next phase of the cardiac cycle, rapid ejection, indicated by phase two on Fig. 4. During this phase blood flows from the ventricles into the aorta and pulmonary artery. Also in this phase LVP and AOP reach their maximum values, with LVP always being slightly larger than AOP. There is a decrease in left atria pressure (LAP) due to expanding of the atria. LVP and AOP increase to their maximum values then begin to decrease. The blood flow into peripheral arteries eventually exceeds the blood flow coming from the ventricles, which causes the decrease in AOP that begins to occur towards the end of the phase. The decreasing LVV causes the LVP to decrease .that as AOP and LVP decrease through the reduced ejection phase, AOP becomes larger than LVP. Changes in LVP and LVV are generally summarized using the pressure-volume (PV) loop seen in. LVV is plotted on the x-axis against LVP. Point A corresponds to the onset of diastole. The ventricle fills from A to B. At point B, the mitral valve closes, and from point B to C the heart is in the volume contraction phase. At point C, the aortic valve opens, and the ventricle ejects blood into the aorta. This decrease in LVV is seen from points C to D, indicating the rapid ejection phase. The volume decreases at a lower rate from points D to E for the reduced ejection phase. At point E, the aortic valve closes, and the heart enters the relaxation phase, completing the cardiac cycle. One of the main goals of an LVAD is to help the weakened heart provide Cardiac Output (CO). CO is defined as the blood pumped by each ventricle in one minute. The CO of the body depends on the patient's level of activity, emotion, and various physiological factors. Providing adequate CO is the motivation behind designing improved LVADs. The volume of blood pumped by each ventricle in one heartbeat is the Stroke Volume (SV). CO is related to SV and heart rate (HR) by the

Eq CO = HR x SV

The CO of the heart is related to preload, the venous blood returning to the heart. The volume of blood pumped out of the heart is directly related to the volume of blood returning to the heart, or the preload. This concept is known as the Frank-Starling law.

Definition

A Porto systemic shunt (PSS) also know as an Abernethy malformation or a liver shunt, is a vascular connection affecting the body's circulatory system. In most people, blood from the stomach, intestines, spleen and pancreas drains into the liver through a large vein called the "portal" vein. With a Porto systemic shunt, Blood draining from the portal system bypasses ("shunts" past) the liver and directly enters the general circulation. As a result the liver does not get a chance to clean or filter the blood. A Porto systemic shunt can be present at birth (i.e. congenital) or acquired as the result of an underlying liver disease that was not present at birth. This blog discusses a congenital Porto systemic shunt.

CLASSIFICATION

Table: 1 Classification According to *Dr.Morgan and Dr.Superina*

Type I	Absence of intra hepatic portal veins
Type I a	Superior mesenteric and spleen vein drain separately into inferior vena cava
Type I b	Superior mesenteric vein and spleen vein form a common Trunk before draining into the inferior vena cava
Type II	Important collateral, patent intra hepatic veins
Type II a	Arising from left or right portal vein (includes the patent ducts venous)
Type II b	Arising from main portal vein (including its bifurcation or steno mesenteric confluence)
Type II c	Arising from the mesenteric, gastric, or spleen veins

Symptoms

- Hyperammonemia:-
- Failure to gain weigh
- Hypoglycemia
- Elevated liver enzymes
- Seizure activity Coagulopat
- Regenerative liver nodules
- Regenerative liver nodules

Tests

- Ultra-sonography
- Liver Enzyme Test
- Ammonia Blood Test
- Oxygen Saturation Test
- Ct Scan & Magnetic Resonance Imaging (MRI)
- Angiogram

Treatment

The reaction of the liver to an uneven blood supply usually results in focal nodular hyperplasia. This is monitored yearly by checking alpha fetoprotein levels to check for liver cancer and with a yearly CT scan. Intelligence may be normal, but some may have a learning disability. Limiting protein intake is advised in some circumstances. Lacunose or Xifaxan (Rifaximin) can be used if there are signs of Hyperammonemia.

A test in radiology is performed to temporarily block the shunt to see if a portal vein is present and to monitor pressure as the shunt is closed. If the results indicate a higher pressure then the doctor is comfortable with, then the process may

be done in stages to slowly expand the portal vein. Children are more resistant to hepatic encephalopathy than are adults, and most children with CEPS are asymptomatic. In this younger population, Porto systemic encephalopathy may develop either spontaneously or after a precipitating event such as gastrointestinal bleeding, constipation, or an infectious systemic disease.

Therefore, the shunt ratio seems to be an important factor in determining the age at which the onset of encephalopathy occurs. It has been reported that patients with shunt ratios of 30%–60% are prone to develop encephalopathy after precipitating events, whereas those with shunt ratios of greater than 60% (especially older patients) are at high risk for developing spontaneous encephalopathy

Case presentation

A 34-year-old Caucasian man was referred to our hospital with complaints of abdominal pain in the right upper quadrant and pain in his right shoulder. The pain was not associated with nausea or vomiting, and there was no history of fever or jaundice. His medical history included an open ducts venous and an Abernethy malformation complicated by the development of pulmonary arterial hypertension (mean pulmonary artery pressure 48mmHg), which had been diagnosed three years before and treated successfully with the endothelia receptor antagonist bosental.His laboratory test showed a hemoglobin level of 9.2mmol/L, an elevated liver enzyme enzyme profile with aspartate aminotransferase 46U/L, alanine aminotransferase 50U/L and bilirubin 32μmol/L. α- Fetoprotein (AFP) concentration was normal (3μg/L) as was hepatic synthetic function

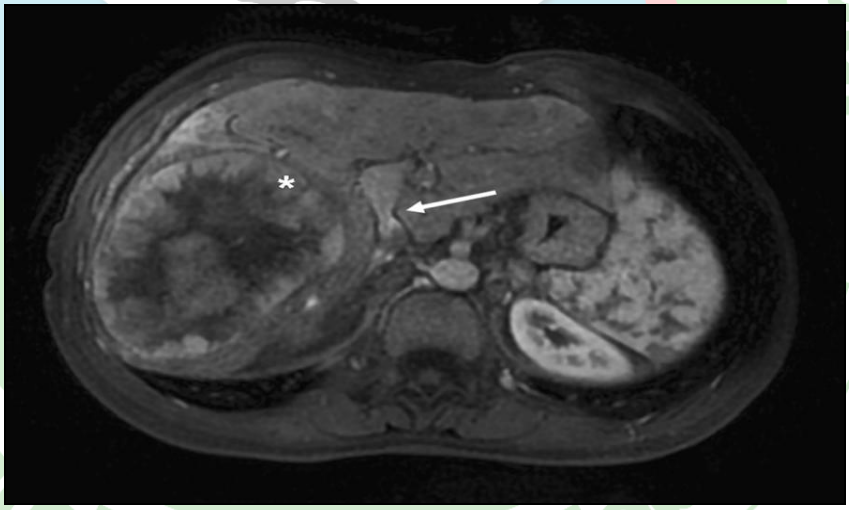

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Figure 9. :- Computed Tomography

Computed tomography (CT) displayed the presence of a tumor, and magnetic resonance imaging (MRI) was performed for characterization of the lesion. The lesion was hypo intense on T1-weighted images and hyper intense on T2-weighted images in the venous phase, and arterial enhancement and washout were demonstrated, as is typical for a HCC (Figure 1). The presence of shunt of 2 cm between the left extra hepatic portal vein and the inferior vena cava was confirmed Preoperatively , workup included several tests, such as lung capacity (lung function test), cardiac function (electrocardiogram) and hepatic synthetic function (laboratory tests). If the results of these tests were normal, the patient would be eligible for surgery. In the preoperative workup, no biopsy of the tumor neither a biopsy of the normal liver parenchyma was found necessary. Because of his pulmonary hypertension, he was

categorized as an American Society of Anesthesiologists (ASA) patient . Histological examination of the resented specimen revealed features of grade II fibrosis.

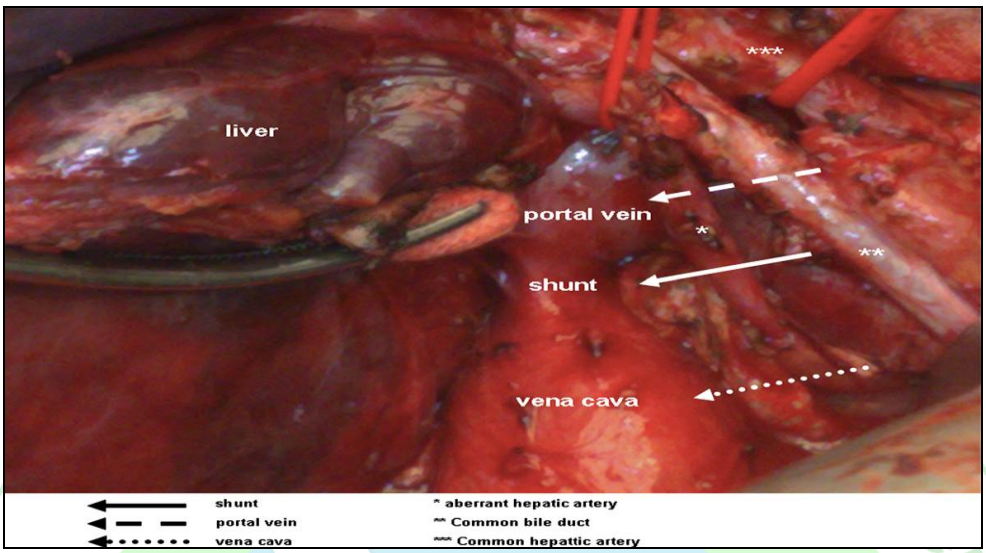

Figure 10:- The Porto caval shunt perioperatively

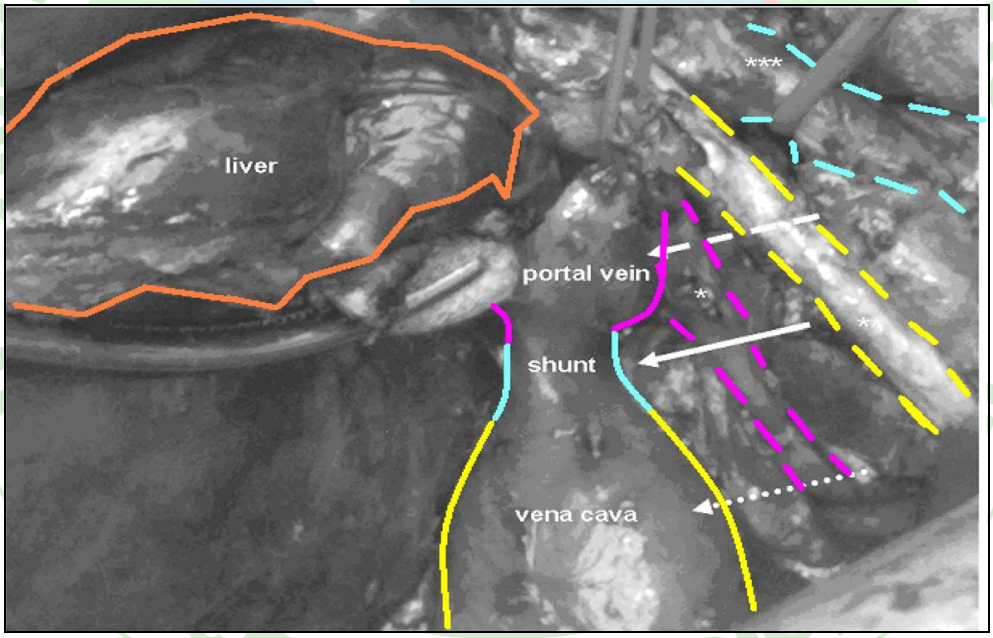

Figure 11:- The Porto caval shunt peri operatively.

Diameter of 16cm was characterized as HCC with the presence of micro vascular invasion. A tumor-free surgical resection of 5mm was recorded. The follow-up of the patient consisted of a routine serum tumor marker AFP determination and contrast enhanced CT or MRI at six months intervals starting three to six months after surgery. After nearly two years of follow-up, the patient is doing well and is free of HCC recurrence.

Discussion

Treatment of congenital malformations of the portal system depends on the type of Porto caval fistulas, the presenting symptoms, complications and co morbidity. Treatment may vary from surgical correction of the shunt to even liver transplantation. Patients with the Abernethy malformation have been described in several case reports, most (80%) of them involving

children ages 18 years and younger. Management strategies for children with a type I shunt have been developed, with close monitoring of clinical, biochemical and radiological parameters in follow-up being advocated. To date, no management strategy has been described for adult patients with a type II shunt. We propose regular follow-up of patients with a type II shunt as well. Absence of a decent portal circulation and systemic diversion of portal vein flow may have consequences for hepatic development, function and regenerative capacity, thus predisposing such patients to the development of fibrosis, nodular dysplasia or HCC. Patients infected with hepatitis B or patients with cirrhosis due to hepatitis C virus infection, alcohol abuse or another cause are patients at risk for the development of HCC. The patients at risk are screened at six to 12 months intervals using liver ultrasonography and serum AFP levels . Liver resections of large malignant tumors have been described before . In cases of a small tumor, patients can be treated with curative intention more often, and five years survival rates over 70% can be achieved The Abernethy malformation comprises a rare vascular malformation with resultant congenital extra hepatic Porto systemic shunt (CEPS). The first known case was described by John Abernethy in 1793, where he described a postmortem evaluation of a 10monthold girl that, among other congenital anomalies, revealed the portal vein to be terminating in the inferior vena cava.1 The eponym referring to congenital extra hepatic Porto systemic shunts was suggested by Howard and Davenport in 1997 in recognition of Abernethy.2 Congenital extra hepatic Porto systemic shunts are characterized by Porto mesenteric blood draining directly to a systemic vein, bypassing hepatic flow either completely or partially.3 In 1994, Morgan and Superina proposed a classification system for CEPS according to the presence or absence of hepatic parenchyma perfusion with mesenteric venous blood.4 In Type 1, there are no intra hepatic portal branches and all portal blood is diverted into the systemic circulation by end to side shunt. This is sub classified into type 1a, where spleen and superior mesenteric veins have separate drainage to the systemic venous system (congenital absence of the portal vein) and 1b, where they drain together after forming a common trunk. Pathogenesis of type 1 CEPS is thought to be a result of excessive involution of the per duodenal vitalize venous loop or failure of vitalize veins to form appropriate an astomosis. Type 2 CEPS may be caused by persistent sub cardio hepatic an astomosis or persistence of the right vitalize vein

CONCLUSION

This case underlines the importance of regular examination of patients with an Abernethy malformation, even In older patients, to prevent complications and to detect liver lesions at an early stage.

Persistence of jaundice should be investigated at all ages. Ultrasound is useful tool for screening of congenital anomalies associated with liver. In resource poor settings the diagnosis of Porto systemic shunts can be missed or significantly delayed due to lack of resources need for the diagnosis, including availability of trained ultrasonologists. The prognosis of the patients with congenital Porto systemic shunts depends on the site of the shunt as determined by Morgan and Superina classification, the associated congenital anomalies and the extent of liver disease. Many patients will benefit from shunt surgery. The extent of associated abnormalities should not deter pediatricians to refer patients for treatment. A long term follow up is indicated for all asymptomatic patients of Abernethy malformation.

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