Development anomalies of the liver: What went wrong?

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Learning objectives

This exhibit classifies, describes and illustrates various development anomalies of the liver.

We present a comprehensive review of the imaging of development anomalies of the liver.

This exhibit will be a core learning tool for the residents and radiology physicians.

Background

Development anomalies may occur within the liver in regard to the arrangement of the liver cells, the lobules, the intrahepatic bile ducts, and also in the mesenchymal architecture of the portal areas.

The liver is frequently involved in the spectrum of situs anomalies.

The tongue-like accessory lobe of the liver (Riedel's lobe) that is related to the inferior portion of the right hepatic lobe is a common finding on nuclear medicine imaging of the liver (up to 20% in females).

Anomalous vascular branching variants are common, being present in up to 59% patients.

Biliary variants are also common, occurring in 42% of the population.

Imaging findings OR Procedure details

1) Location (situs anomalies)

Situs inversus is a rare anomaly that is characterized by mirror-image location of the abdominal organs and, in most cases, the cardiac apex relative to situs solitus. The liver is located in the left upper quadrant, whereas the spleen and stomach are located in the right upper quadrant. An extremely rare variant of situs inversus is situs inversus with levocardia. In this anomaly, there is mirror-image location of the abdominal organs with a left-sided cardiac apex.

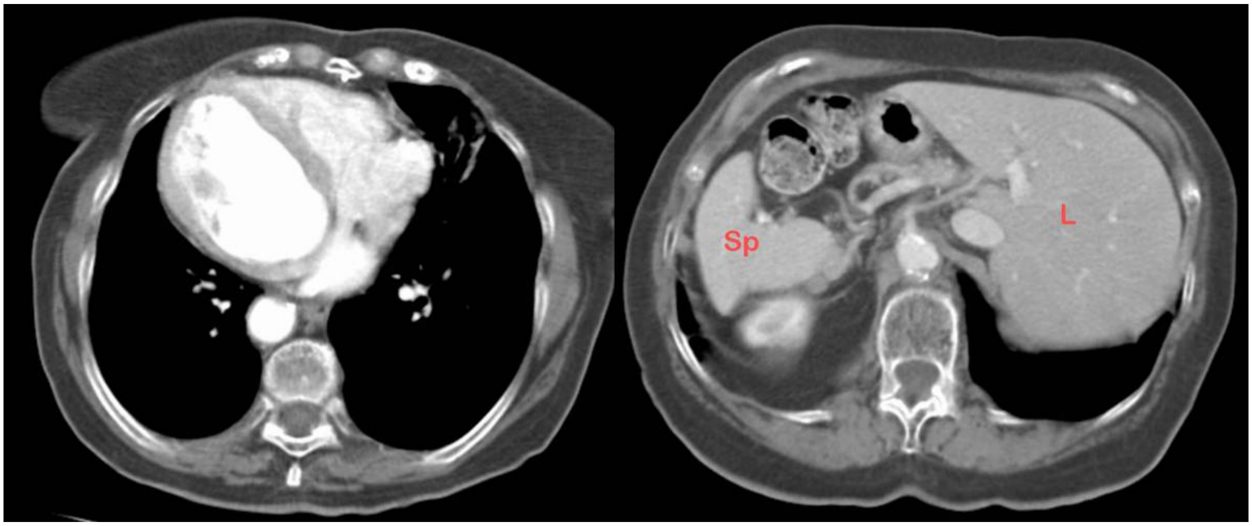

Fig.: Contrast-enhanced CT scan of the lower chest reveals dextrocardia. Transverse contrast material-enhanced CT scan of the abdomen demonstrates mirror-image location of the abdominal structures relative to situs solitus (normal). The liver (L) is located in the left upper quadrant, whereas the spleen (Sp) is located in the right upper quadrant.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Situs ambiguous with polysplenia, often referred to as left isomerism or bilateral left-sidedness, is a subcategory of situs ambiguous that is usually characterized by an abnormal arrangement of the solid organs and bowel and the presence of multiple spleens. However, some studies report patients with situs ambiguous who have a single, lobulated spleen or even a normal spleen.

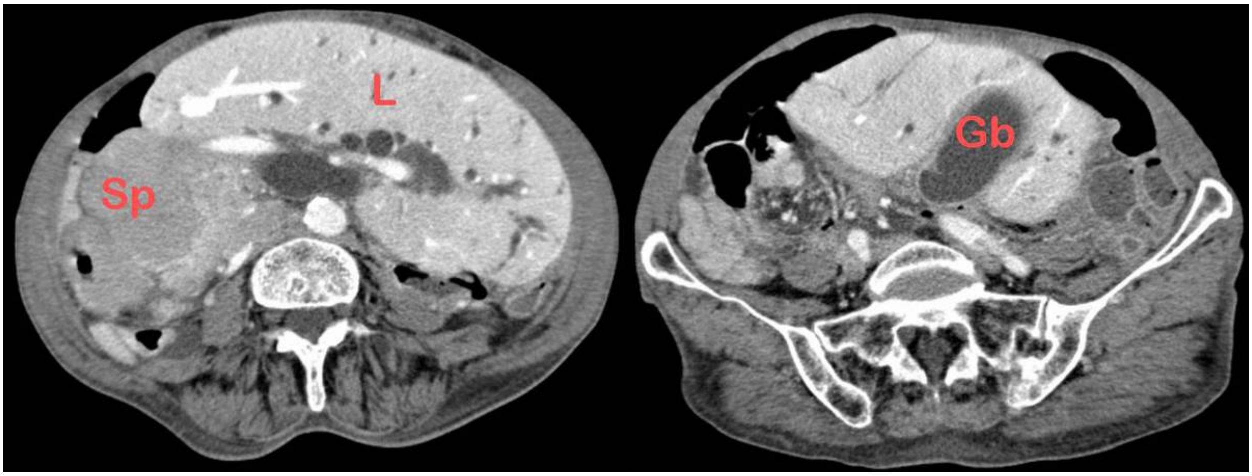

Fig.: Transverse contrast-enhanced CT scan of the abdomen shows a midline liver and multiple spleens (Sp) in the right upper quadrant. The low attenuation of the spleens is related to infarctions. Note the absence of splenic tissue in the left upper quadrant. Transverse CT scan through the midabdomen reveals a midline gallbladder (Gb).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Situs ambiguous with asplenia is a second subcategory of situs ambiguous that is generally characterized by an abnormal arrangement of the abdominal organs and absence of the spleen.

2) Segmentation

Liver anatomy can be described using two different aspects: morphological anatomy and functional anatomy. Naming of the parts of this complex inner organ is still highly varied: parts, halves, lobes, divisions, sectors, segments, and subsegments.

The most widely used classification system was proposed in 1957 by Couinaud. Hepatic segmentation is based on the distribution of the portal pedicles and the location of the hepatic veins. Couinaud's system distinguishes two lobes and four segments. Each segment has a branch (or a group of branches) of the portal vein at its center and a hepatic vein at its periphery. Each of these afferent territories is separated from the others by planes corresponding to portal scissures which allow surgical removal of parenchyma without impairing the blood supply of the remaining liver tissue.

Fig.: Claude Couinaud
References: Saulius Rutkauskas et al (2006) Clinical and anatomical basis for the classification. Medicina (Kaunas) 2006; 42(2)

The liver is divided into eight segments, both longitudinally along the hepatic veins and transversely through the right and left portal pedicles. Seen from the front, they are numbered clockwise from 1 to 8, with 1-4 in the left lobe and 5-8 in the right.

From a functional point of view, the caudate lobe (or segment I) must be considered an autonomous segment, for its vascularization independent of the portal division and three main hepatic veins. It receives vessels both from the left and right branches of the portal vein and hepatic artery; its hepatic veins are independent and drain directly into the inferior vena cava.

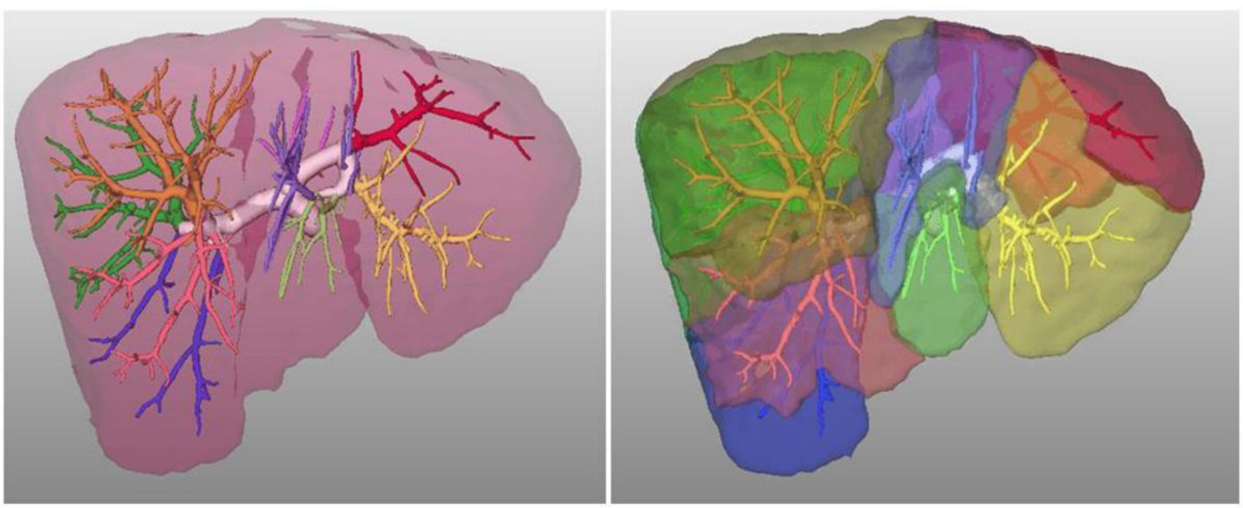

Fig.: Hepatic segmental anatomy. Frontal views of color-coded three-dimensional CT images from a liver donor show the various portal ramifications and corresponding liver segments.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

The tongue-like accessory lobe of the liver (**Riedel's lobe**) is a common finding on nuclear medicine imaging of the liver. This anatomic variant has been reported in some detail by Reitmeier et al. (1958) and was first described by Riedel (1888), a French surgeon.

The Riedel's lobe is usually closely related to the inferior portion of the right lobe and the area or plane of cleavage between the Riedel's lobe and right lobe can be sharply demonstrated. It may extend in a number of directions. Although anterior extension is the most frequent presentation, there may be variable degrees of extension posteriorly. The Riedel's lobe appears to vary considerably in size, too. In some images it is seen as a small, thin, tongue-like structure while in others it may have the appearance of a large rounded or globular mass. Interestingly, it has been observed almost exclusively in females.

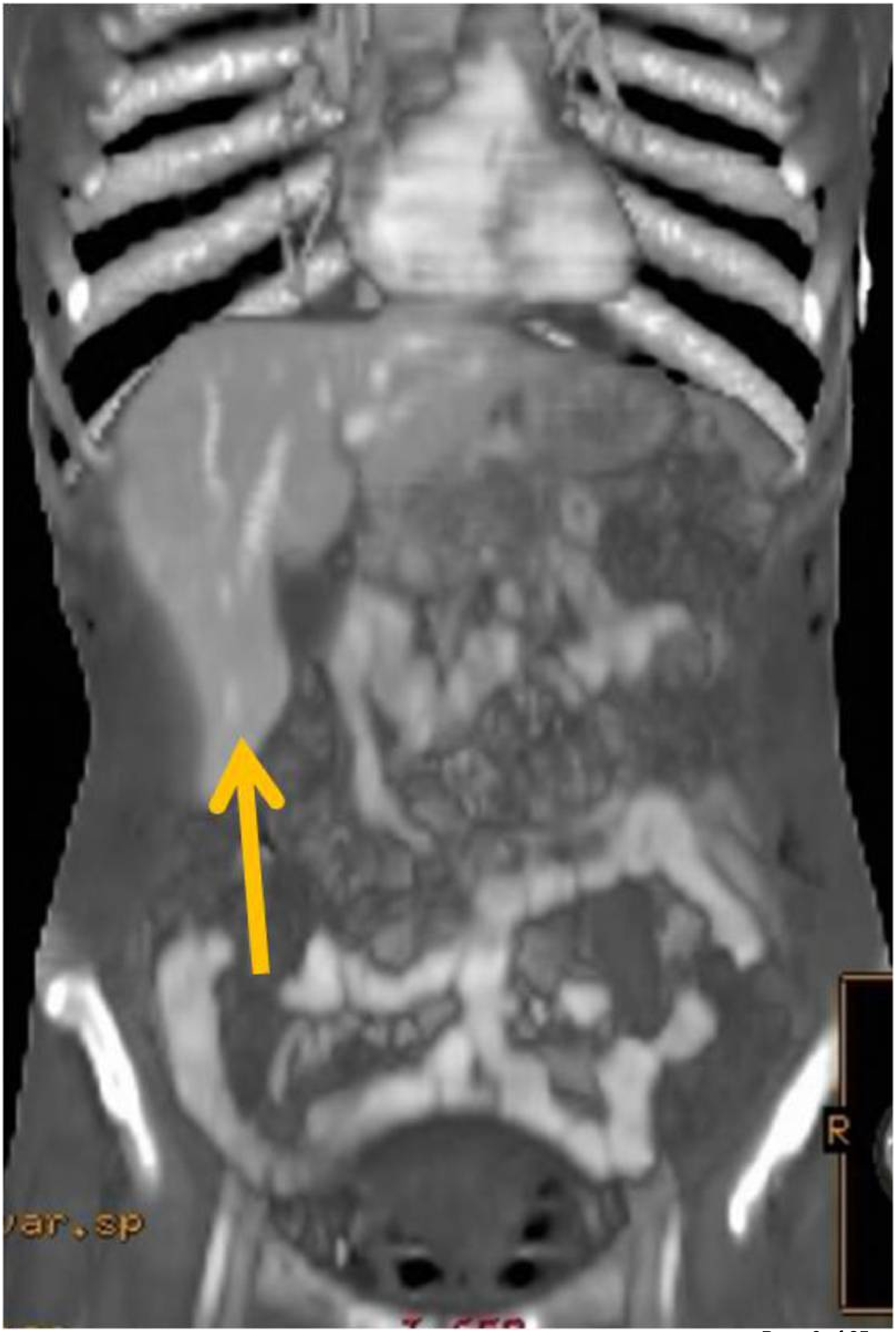
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Fig.: The tongue-like Riedel's lobe (arrow) is clearly demonstrated in the anterior and right lateral positions.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

3) Vascular system

Hepatic artery

Variations in the origin or the course of the hepatic artery are known to occur in as frequently as over 40% of cases. Variations in the origin are thought to result from developmental changes in the primitive ventral splanchnic arteries.

All the classical variations of the hepatic artery can consequently be explained by either abnormal disappearance of an arterial segment that normally persists, persistence of an arterial segment that normally disappears, or both phenomena.

An aberrant hepatic artery refers to a branch that does not arise from its usual source. Under variant patterns, the lobes may receive blood supply from the superior mesenteric artery (SMA), left gastric artery, aorta, or other visceral branches. These vessels may be accessory, occurring in addition to the normal arterial supply, or replaced, representing the primary arterial supply to the lobe.

In Michels' classic autopsy series of 200 dissections, published in 1966, the basic anatomical variations in hepatic arterial supply were defined and this classification has served as the benchmark for all subsequent contributions in this area.

Table 1 Michels' classification of hepatic arterial anomalies

Type	Description
I	Normal anatomy
II	Repl. LHA from LGA
III	Repl. RHA from SMA
IV	Repl. LHA from LGA and repl. RHA from SMA
V	Acc. LHA from LGA
VI	Acc. RHA from SMA
VII	Acc. LHA from LGA and acc. RHA from SMA
VIII	Acc. LHA from LGA and RHA from SMA
IX	CHA from SMA
X	CHA from LGA

LHA left hepatic artery, *LGA* left gastric artery, *RHA* right hepatic artery, *SMA* superior mesenteric artery, *CHA* common hepatic artery, *Repl.* replaced, *Acc.* accessory

Fig.: Michels' classification of hepatic arterial anomalies.
References: Carlo Nicola De Cecco et al (2009) Anatomic variations of the hepatic arteries. Eur Radiol (2009) 19: 2765-2770

The most common variant seen is a **replaced right hepatic artery** originating from the SMA. Whereas the right hepatic artery usually courses anterior to the right portal vein, the replaced right hepatic artery originates from the SMA, courses posterior to the main portal vein in the portacaval space, and classically ascends posterolateral to the common bile duct.

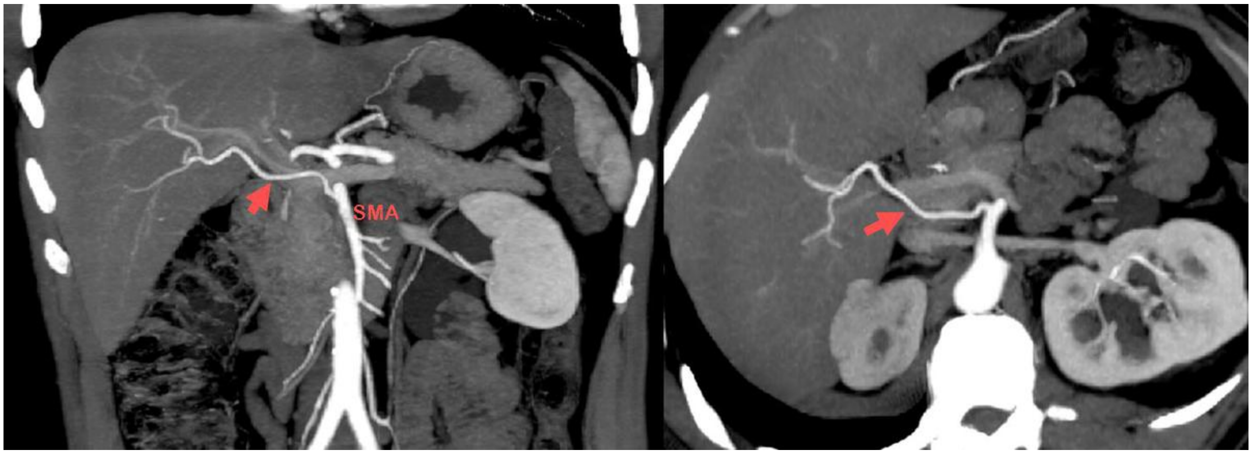

Fig.: Coronal and axial MIP reformatted images show right hepatic artery arising directly from superior mesenteric artery and coursing posterior to the main portal vein in the portacaval space.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

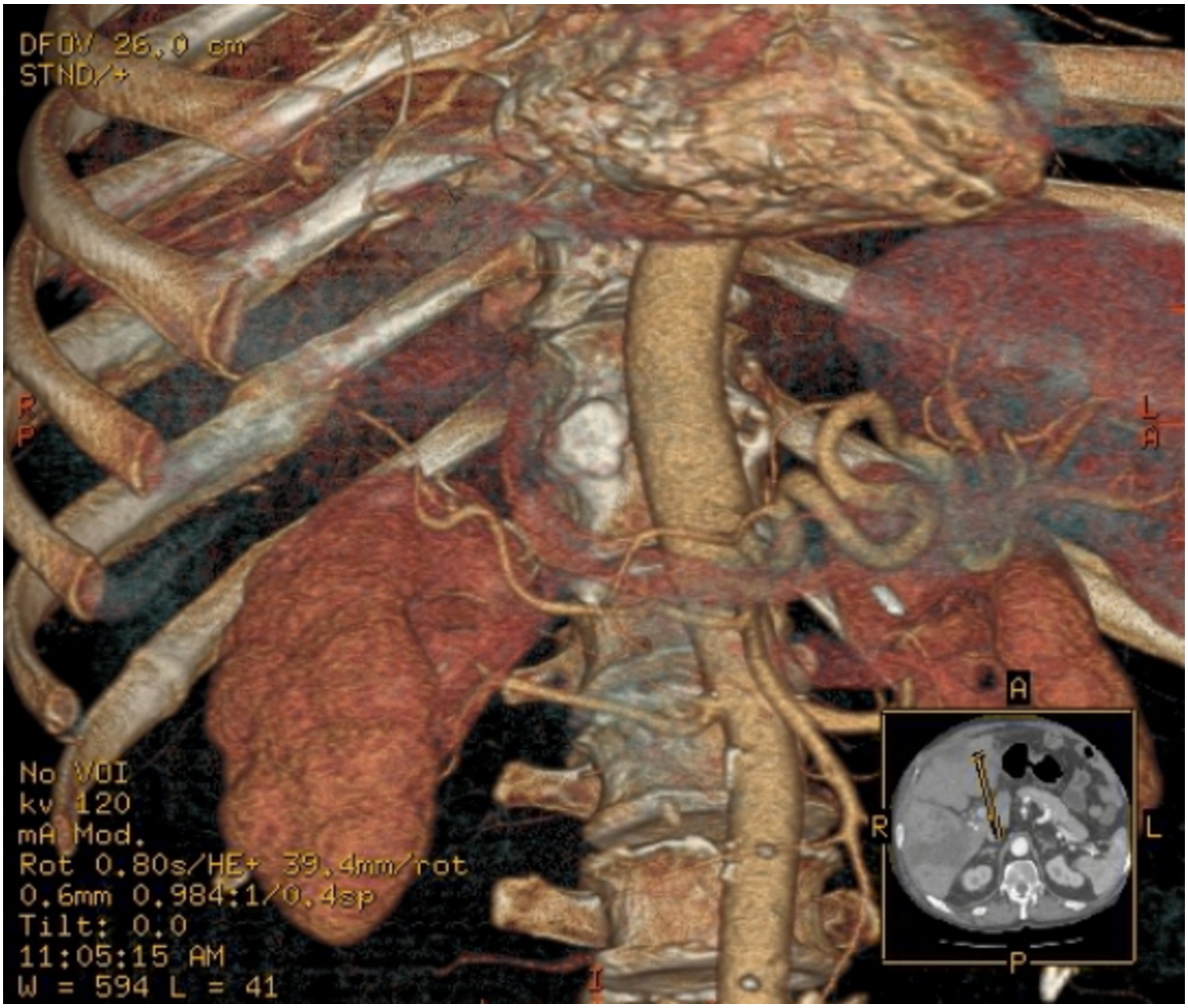

Fig.: Three-dimensional volume-rendered CT angiography image showing a replaced right hepatic artery.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

The second most common arterial variant identified is a **replaced left hepatic artery** originating from the left gastric artery. The replaced left hepatic artery originates off the left gastric artery and courses to the right through the lesser sac, through the fissure for the ligamentum venosum, and into the umbilical fissure to perfuse the left hepatic lobe.

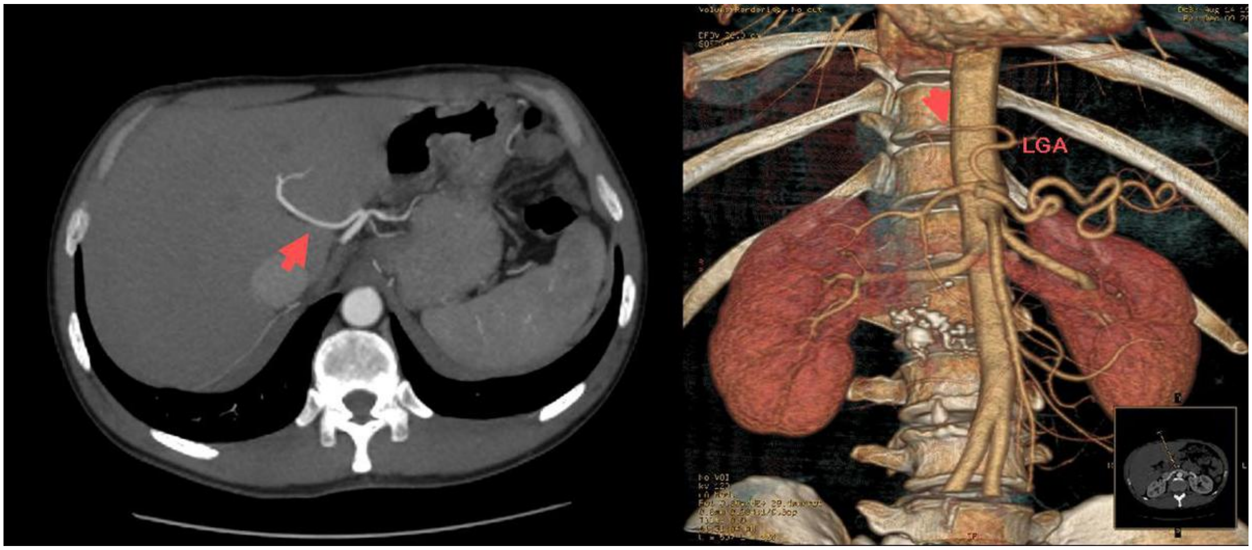

Fig.: MIP and three-dimensional volume-rendered and maximum intensity projection CT angiography images showing a replaced left hepatic artery (arrows) arising from LGA and a normal right hepatic artery.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

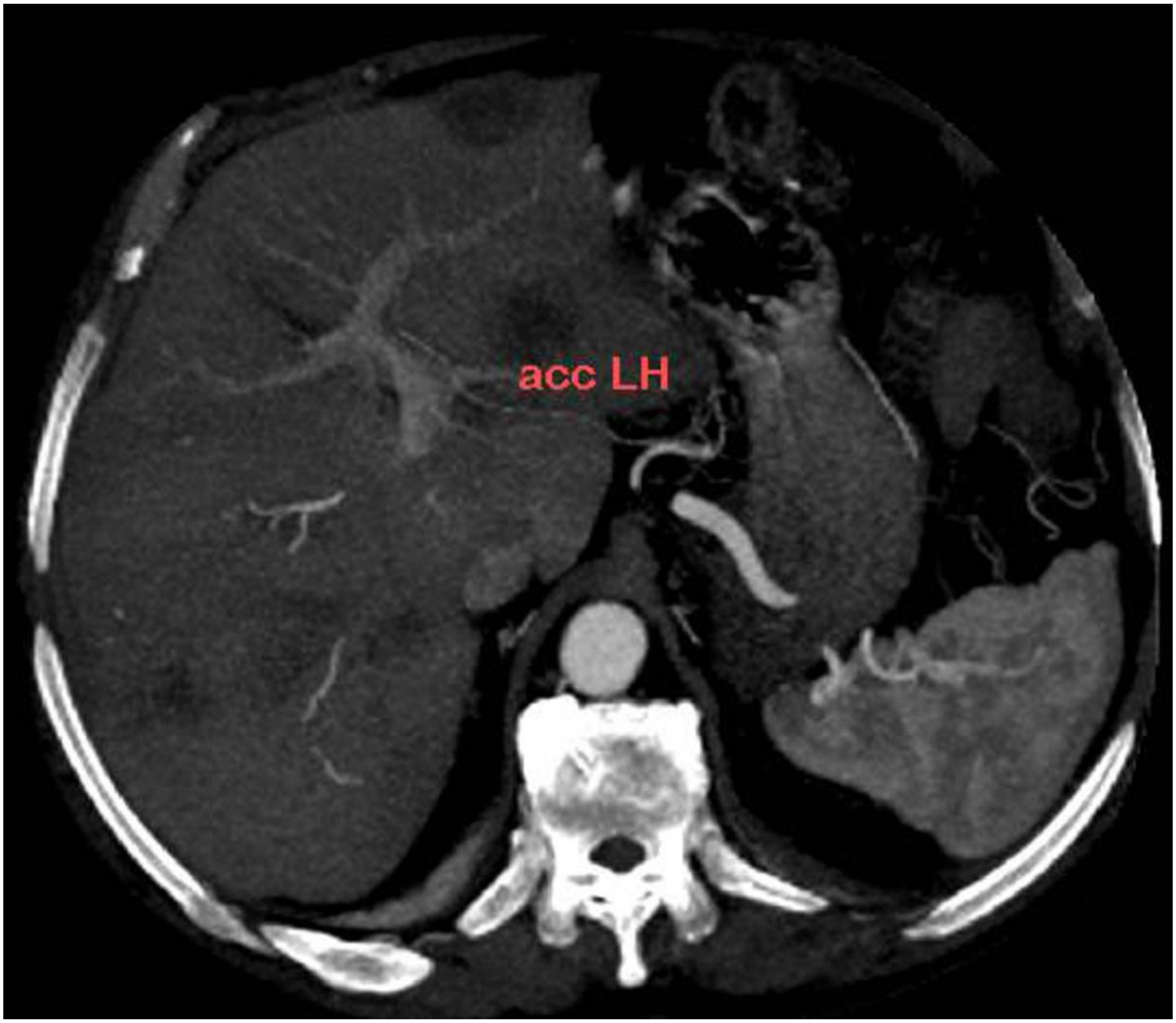

Fig.: Axial MIP reformatted image shows a very thin accessory left hepatic artery arising from left gastric artery.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

It is also possible to find other anatomical variants, not included in Michels's classification, including a replaced RHA that originating directly from the aorta, or the main hepatic artery also originating from the aorta.

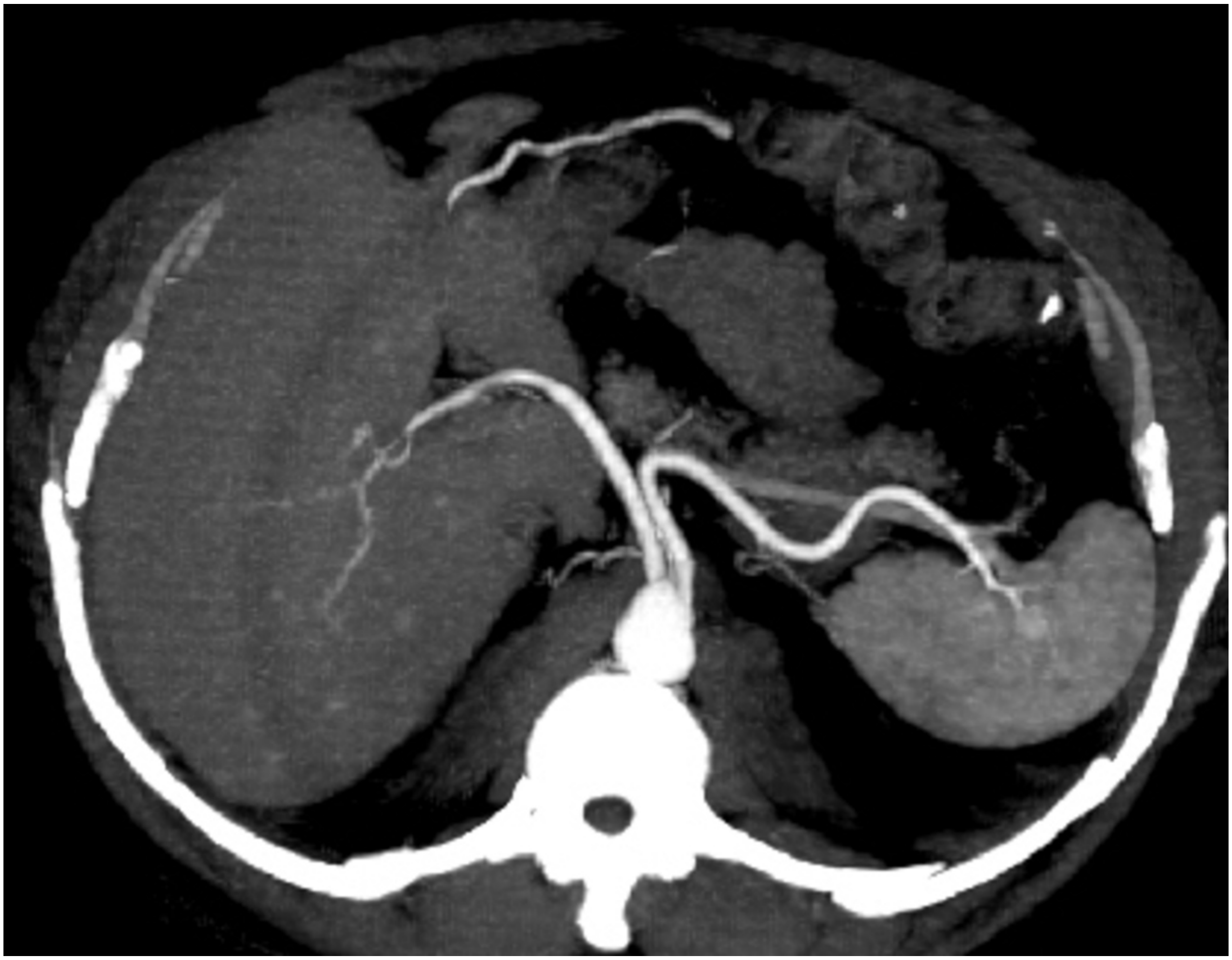

Fig.: MIP image depicts a main hepatic artery that arises directly from the aorta.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

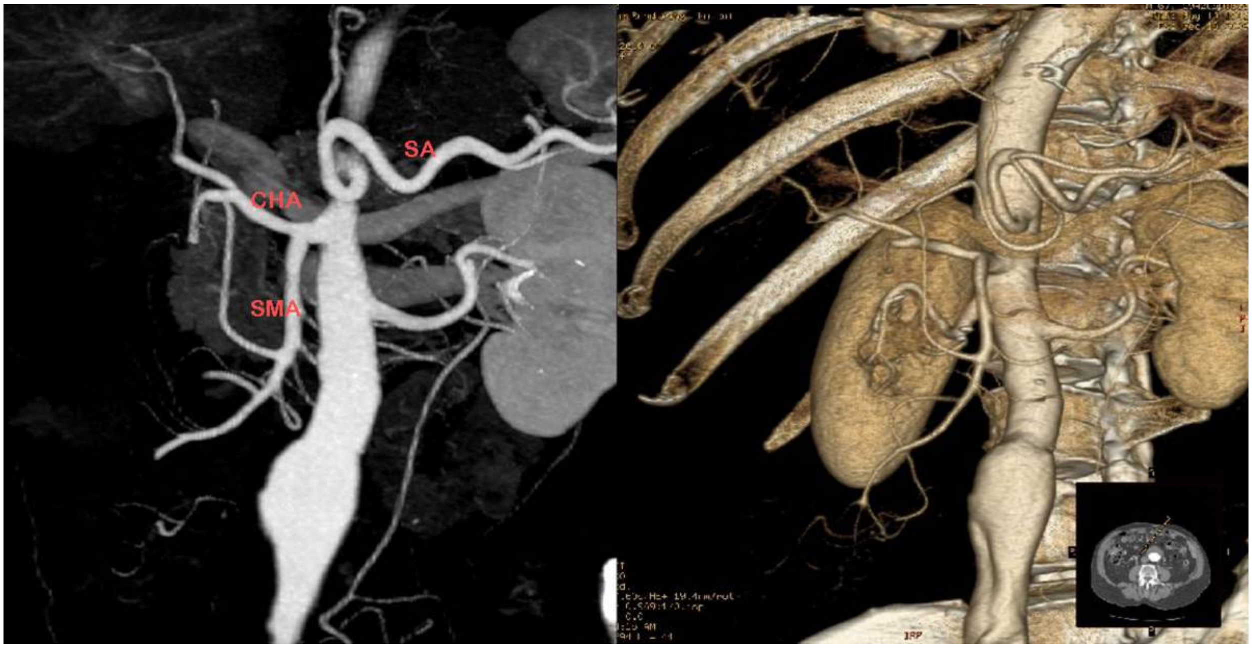

Fig.: MIP and volume rendered image depict clearly Michels variants type IX. The hepatic artery originates from the superior mesenteric artery. Splenic artery (SA) arises directly from the aorta.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

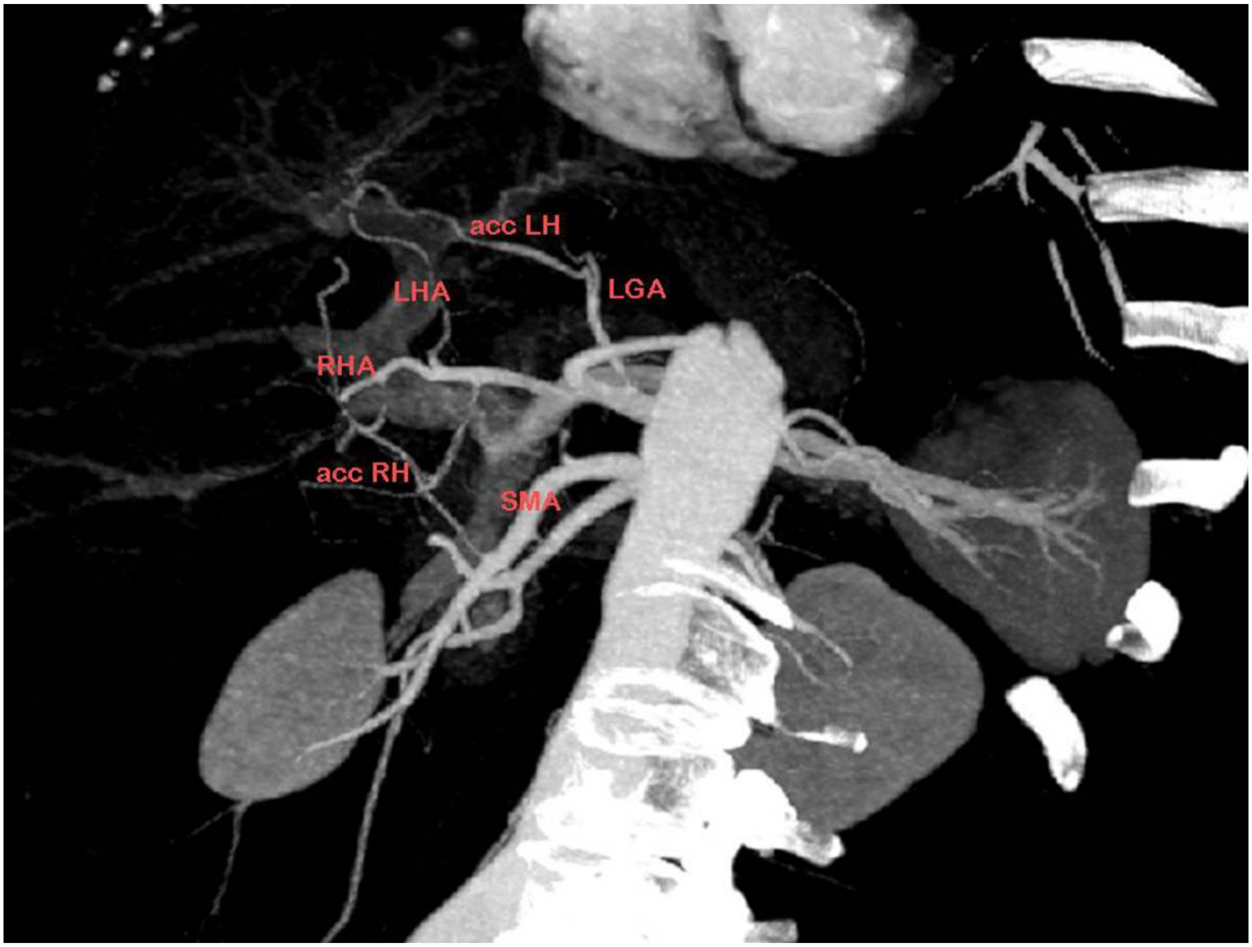

Fig.: Maximum-intensity-projection CT scan obtained in oblique plane shows accessory right hepatic artery (acc RH) arising from superior mesenteric artery (SMA) and accessory left hepatic artery (acc LH) from left gastric artery (LGA). Common hepatic artery gives off gastroduodenal artery and then bifurcates into left hepatic artery (LHA) and right hepatic artery (RHA).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Even the anatomy of the artery (or arteries) that feeds segment IV may be of interest, because of its crucial position in surgical procedures. The arterial supply for segment IV may be quite variable. It is possible to observe a single, double, and triple supply for segment IV, originating from RHA, LHA, and proper hepatic artery.

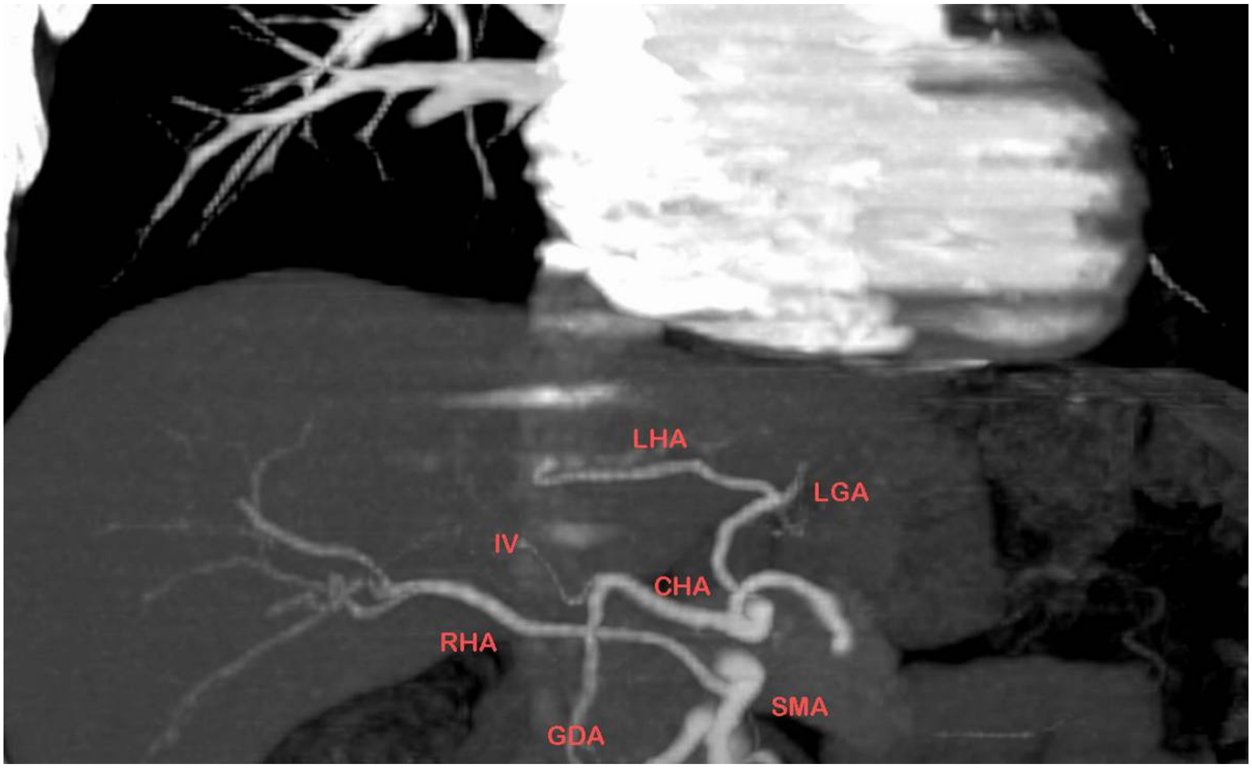

Fig.: MIP image showing a replaced right hepatic artery (RHA) arising from the SMA and a replaced left hepatic artery (LHA) arising from the LGA. CHA, common hepatic artery; GDA, gastroduodenal artery; IV, artery for segment IV.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Portal vein

Branching anomalies of the main portal vein (PV) at the hepatic hilum are known to be less frequent than those of the hepatic arteries, hepatic veins, and biliary ducts. Embryologically, the PV is formed during the second month of gestation by selective involution of vitelline veins, which have multiple bridging anastomoses, anterior and posterior of the duodenum. Alterations in the pattern of these anastomoses result in PV variations.

Conventional portal anatomy is accepted as the main PV bifurcating into the right portal vein (RPV) and left portal vein (LPV), the RPV then dividing into the right anterior portal vein (RAPV) and right posterior portal vein (RPPV). Any deviation from this pattern was regarded as variant anatomy.

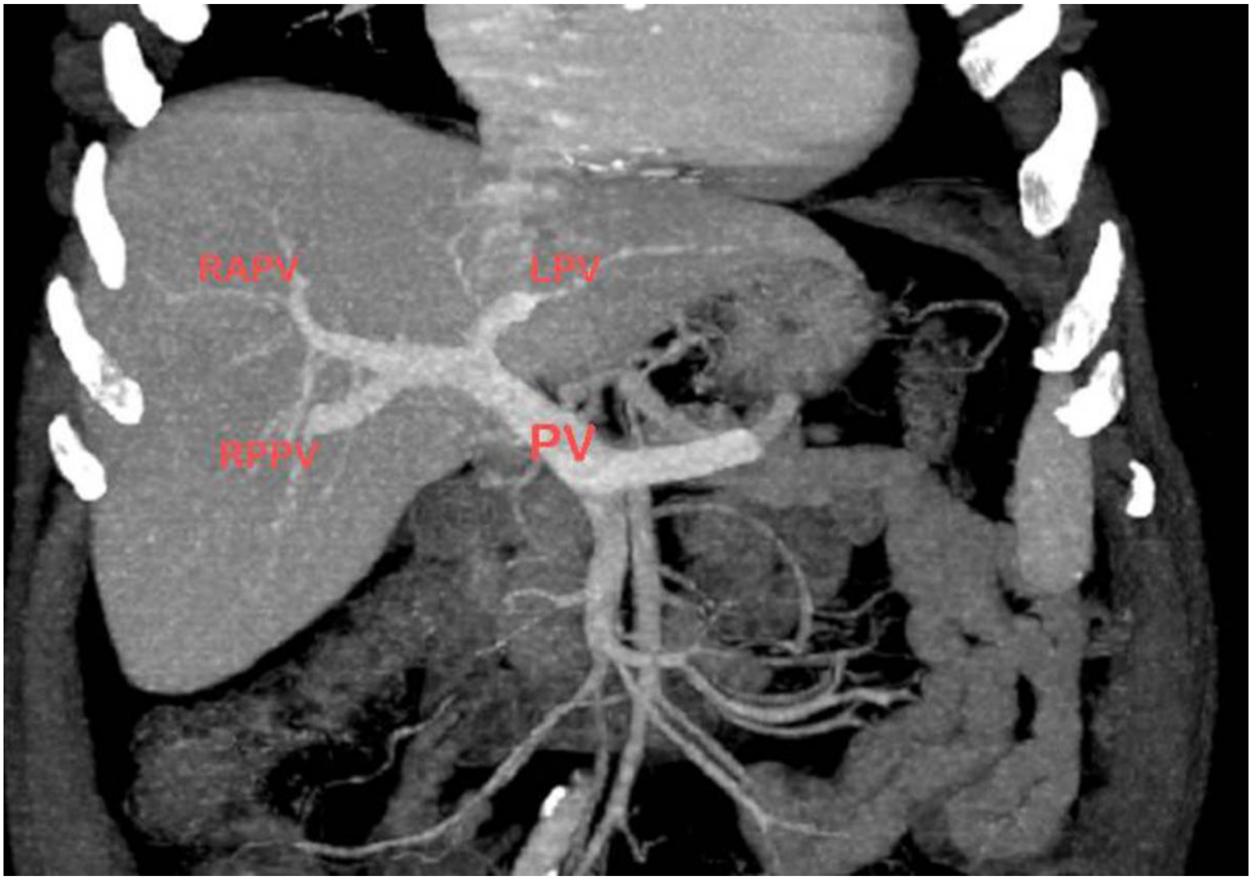

Fig.: Normal (classic) main portal vein branching pattern. Image shows the portal vein (PV) branching into the left portal vein (LPV) and right portal vein. The latter divides into the right anterior portal vein (RAPV) and right posterior portal vein (RPPV).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Variants in the normal branching pattern of the intrahepatic portal vein occur in approximately 20% of the population. The most common patterns include trifurcation of the main portal vein (7.8%-10.8%), right posterior segmental branch arising from the main portal vein (4.7%-5.8%), and right anterior segmental branch arising from the left portal vein (2.9%-4.3%).

- **Trifurcation of the main portal vein**: In these cases, the main portal vein divides into three branches after entering the porta hepatis; right anterior segment, right posterior segment, and left portal vein.

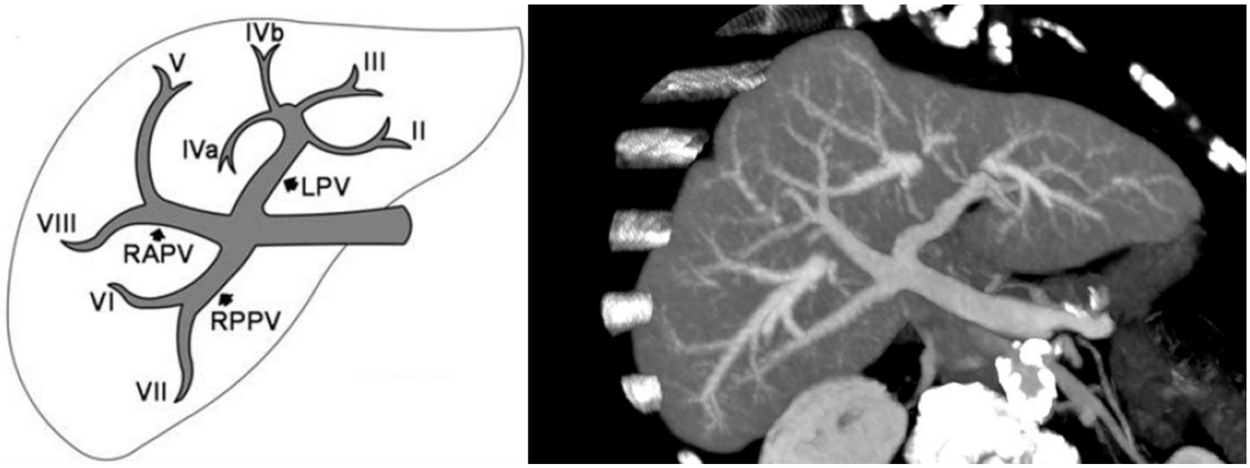

Fig.: Maximum-intensity-projection CT scan obtained in oblique plane shows trifurcation of the portal vein into the right anterior portal vein (RAPV), right posterior portal vein (RPPV), and left portal vein (LPV).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

- **Origin of the right posterior segmental branch from the main portal vein**: The main portal vein gives rise to the right posterior segment, continues to the right for a short distance, and then divides into the right anterior segmental branch and the left portal vein.

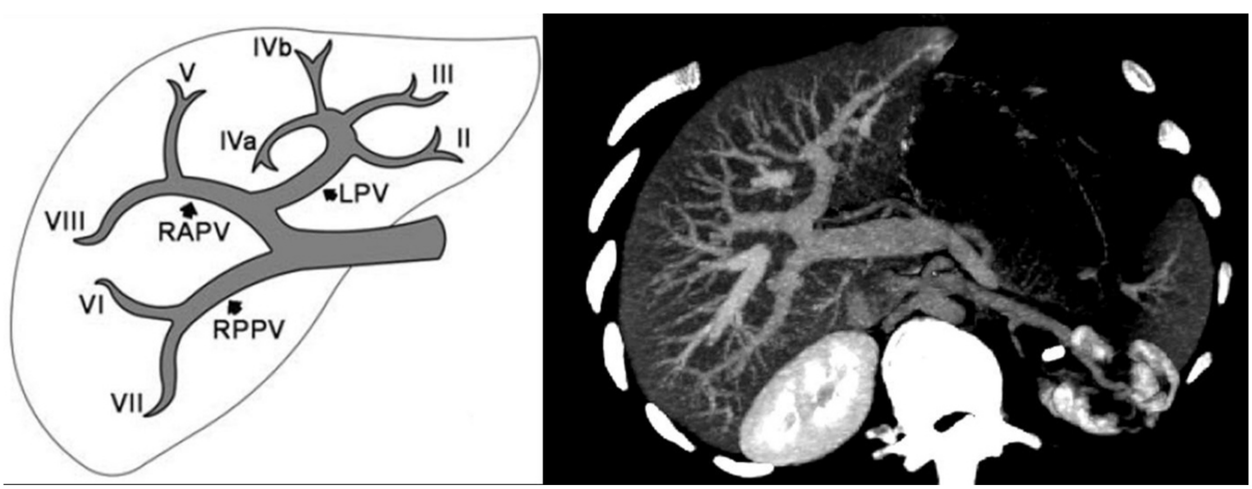

Fig.: Maximum-intensity-projection CT scan obtained in oblique plane shows accessory right inferior portal vein (RPPV) arising from main portal vein. Main portal vein then bifurcates into left portal vein (LPV) and right anterior portal vein (RAPV).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

- **Origin of the right anterior segmental branch from the left portal vein**: In these cases, the main portal vein divides into the right posterior segment and the left portal vein. The right anterior segmental vein originates from the left portal vein.

Slight variations of the "normal" distribution of portal vein are commonly seen. These include short main right portal vein, a short horizontal portion of the left portal vein, disproportionate size of different segmental branches, and a small accessory branch (arising from the main portal vein) to the right posterior segment. Some of the latter variations correlate with differences in the size of some segments of the liver, where a hypoplastic segment receives small branches.

An **aberrant right gastric vein** directly draining into the liver is a well-known variation of the portal vein system. It was described as early as the 19th century by Sappey as an "accessory portal vein" and is also known as one of the "parabiliary veins" described by Couinaud. Aberrant right gastric vein, as a significant abnormality in portal blood flow, has been related to either focal fatty infiltration of the liver and focal sparing in fatty liver.

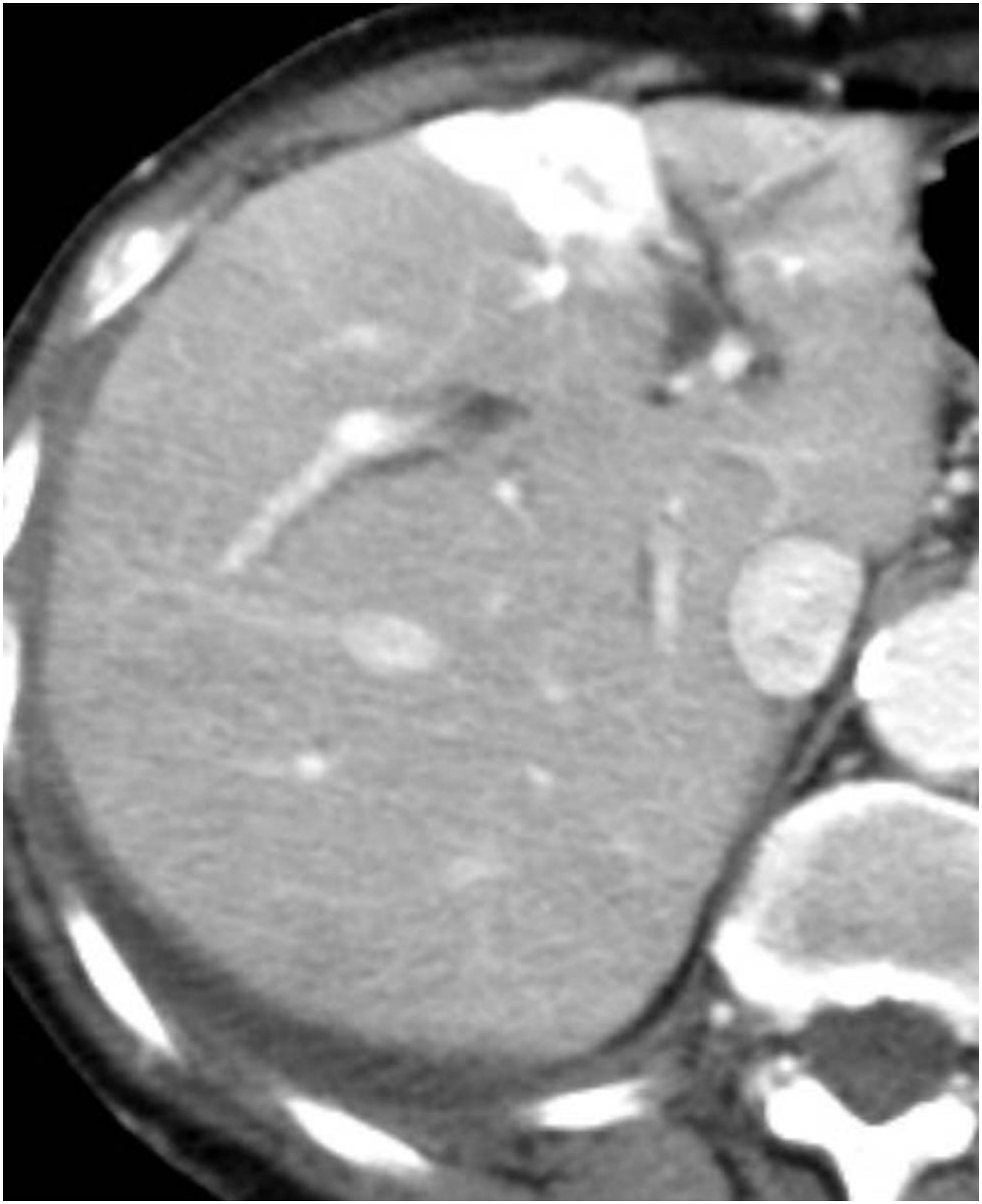

Fig.: Hot liver sign. Patient with superior vena cava syndrome, with intense and early enhancement of segment IV of the left liver lobe, mimicking a hypervascular focal lesion, corresponding to an early arrival of minimally diluted contrast agent to those areas(through an aberrant right gastric vein).

References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Hepatic veins

The 3 main hepatic veins (right, middle, and left) drain into the inferior vena cava (IVC) approximately 1 cm below the diaphragm and 2 cm inferior to the lower border of the right atrium. The right hepatic vein (RHV) is the largest of these veins as it drains the largest volume of hepatic parenchyma. The middle hepatic vein (MHV) courses along the major lobar fissure. It forms a common trunk with the left hepatic vein (LHV) in 85% of the population, which then drains into the anterior left lateral aspect of the IVC. The LHV is the smallest of these veins.

The anatomy of the major hepatic veins is widely available. However, the **minor hepatic veins or accessory hepatic veins** (AHVs), which are additional veins draining into the retrohepatic portion of the inferior vena cava between the right supra-adrenal vein and the openings of the main hepatic veins, have only been rarely mentioned and without any real systematization. AHVs are small and abundant, but they are very important in the liver surgery. The pathophysiologic importance of the minor hepatic veins is of great significance in situations where the drainage of the liver through the main hepatic veins becomes insufficient, such as in the Budd-Chiari syndrome or in large tumors of the liver.

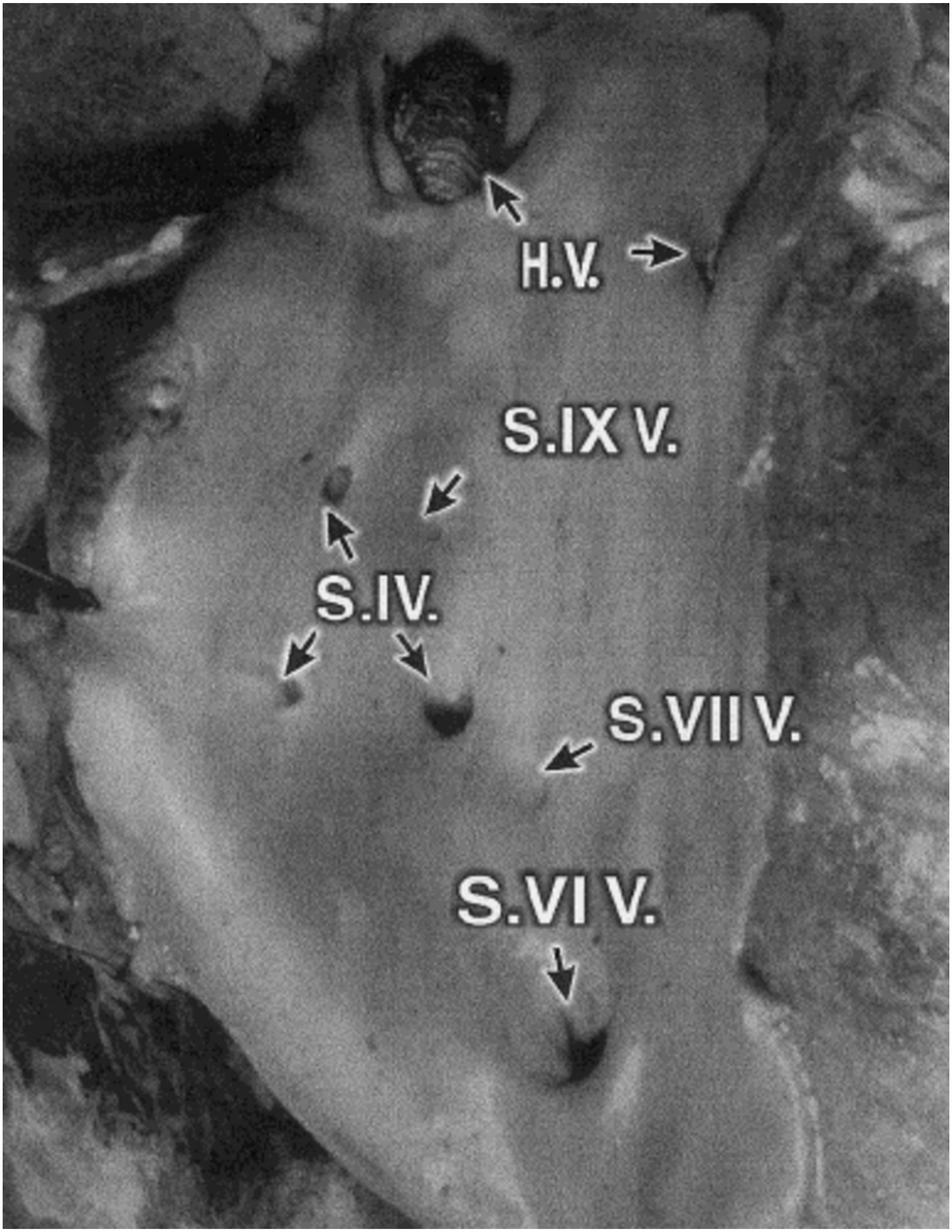

Fig.: The retrohepatic inferior vena cava with the opening of the hepatic veins. H.V., main hepatic veins; S.IV; Segment I veins; S.VI V., Segment VI vein; S.VII V., Segment VII veins; S.IX V, Segment IX vein.

References: Reza Mehran et al (2000) The Minor Hepatic Veins: Anatomy and Classification. Clinical Anatomy 13 2000; 416-421

The largest of these AHVs is the **inferior right hepatic vein**, which can sometimes be the dominant draining vein of the right lobe. A large inferior RHV is the dominant drainage of the posterior segment of the right lobe in approximately 3%-5% of patients. It enters the IVC well below the right atrium; the superior RHV may be atretic or absent in such a case.

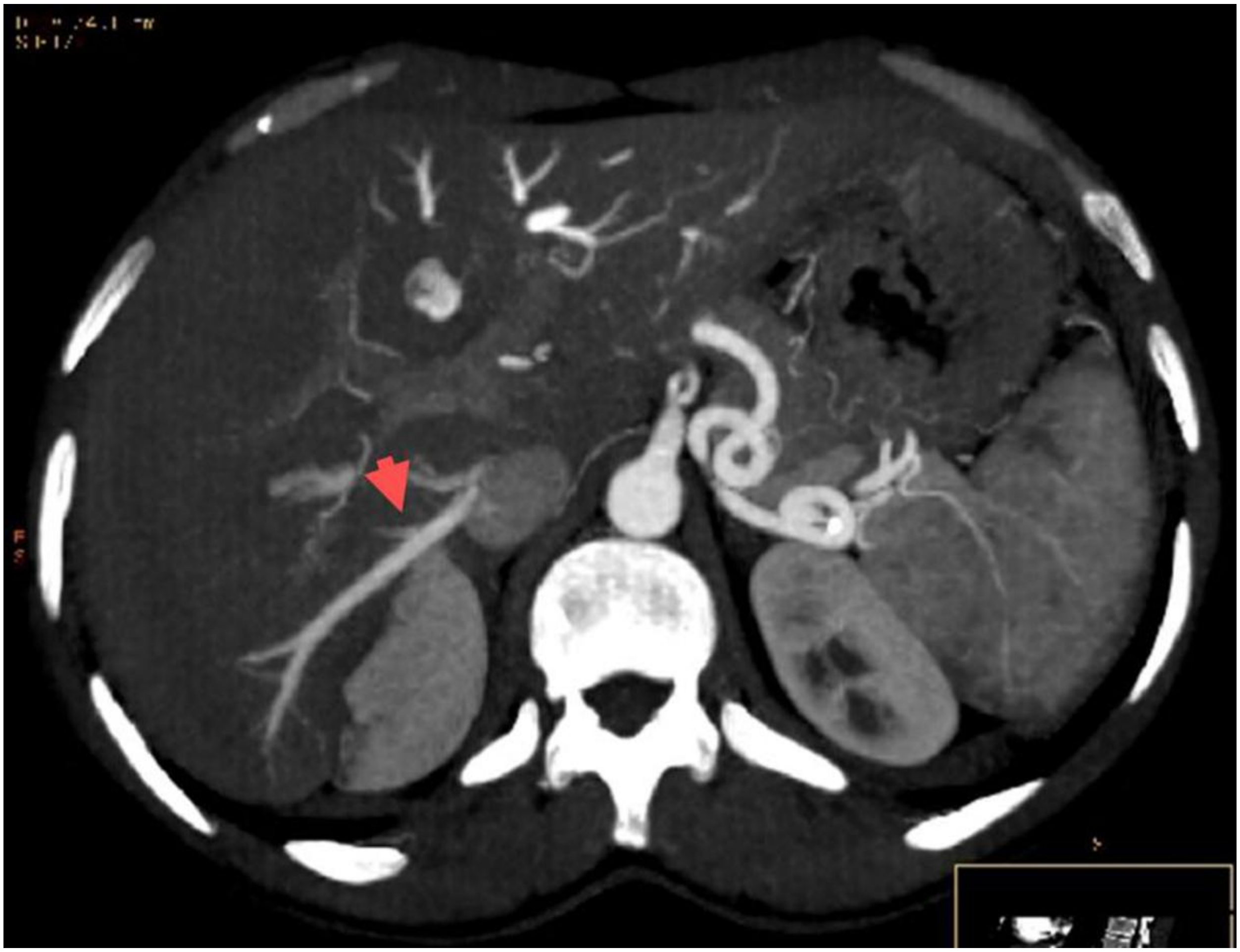

Fig.: MDCT axial image shows an accessory venous branch (VI segment) draining directly to inferior cava vein (arrow).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Accessory hepatic veins also drain the caudate lobe directly into the IVC.

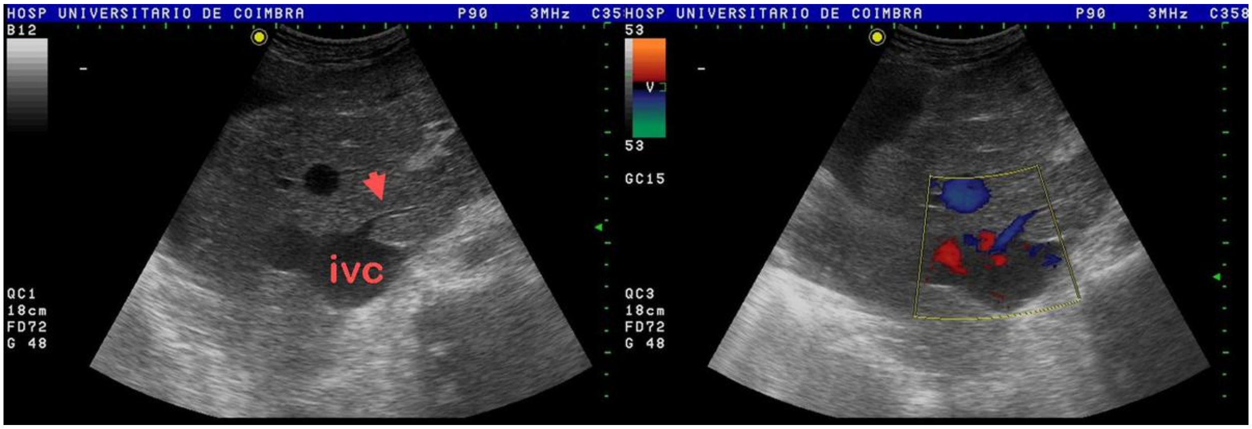

Fig.: Ultrasound and Color Doppler images show hepatic vein draining the caudate lobe directly into the inferior vena cava.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

4) Biliary system

Anatomic variants of the biliary anatomy are common, with the classic anatomy being found in only 58% of the population.

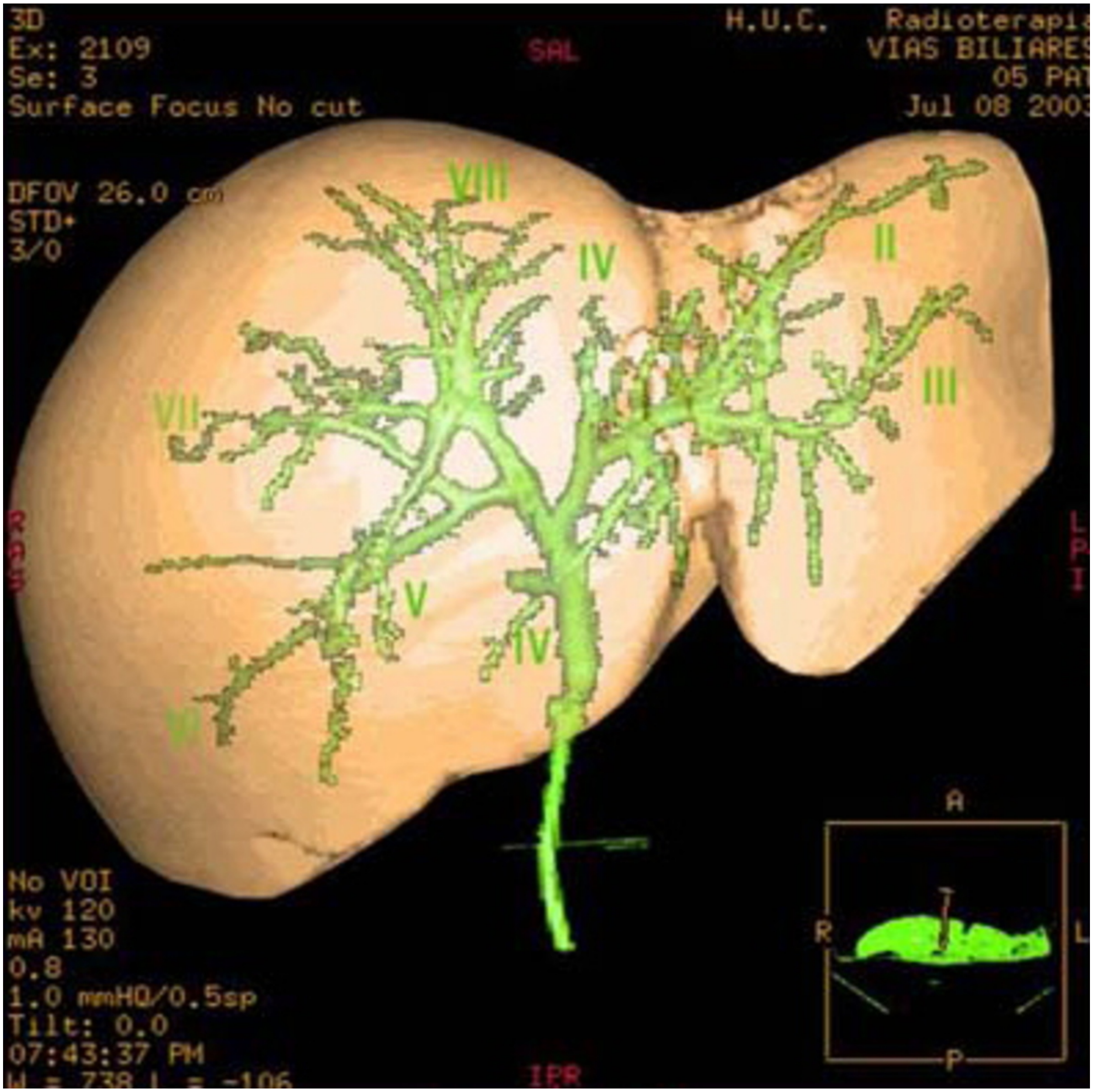

Fig.: Three-dimensional rendered model of a liver with the common bile duct and its branches injected. Classic biliary anatomy is shown: the left hepatic duct has a transverse orientation and it is formed by the lateral branch (from segment II) and the medial branch (from segments III and IV). The right hepatic duct is short and almost vertical and it is formed by a posterior branch (from segments VI and VII) and an anterior branch (from segments V and VIII).

References: P. Donato et al. (2007) Normal vascular and biliary hepatic anatomy: 3D demonstration by multidetector CT. Surg Radiol Anat 29:575 - 582

The most common anatomic variants in the branching of the biliary tree described involve the **right posterior duct** and its fusion with the right anterior or left hepatic duct. Drainage of the right posterior duct into the left hepatic duct before its confluence with the right anterior duct is the most common anatomic variant of the biliary system and reported to occur in 13-19% of the population.

Fig.: MR cholangiogram shows drainage of right posterior duct (arrow) into left hepatic duct before joining right anterior duct.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

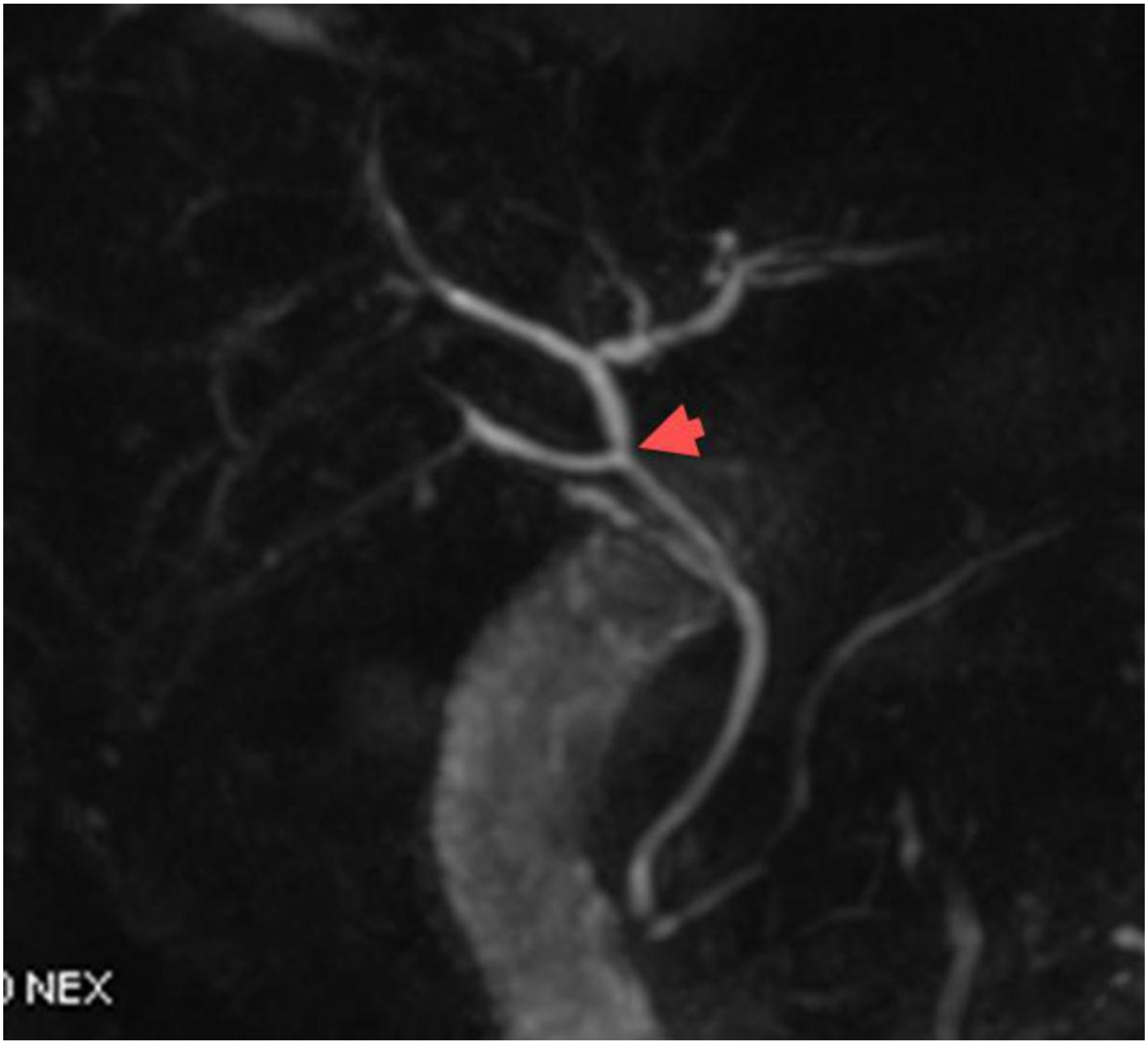

Fig.: MR cholangiogram shows drainage of the posterior branch from segments VI and VII directly into the common hepatic duct (arrow).
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Other common clinically relevant anatomic variants of the biliary tract include a **biliary trifurcation**. This is an anomaly characterized by simultaneous emptying of the right posterior duct, right anterior duct, and left hepatic duct into the common hepatic duct. In patients with this variant, the right hepatic duct is virtually nonexistent.

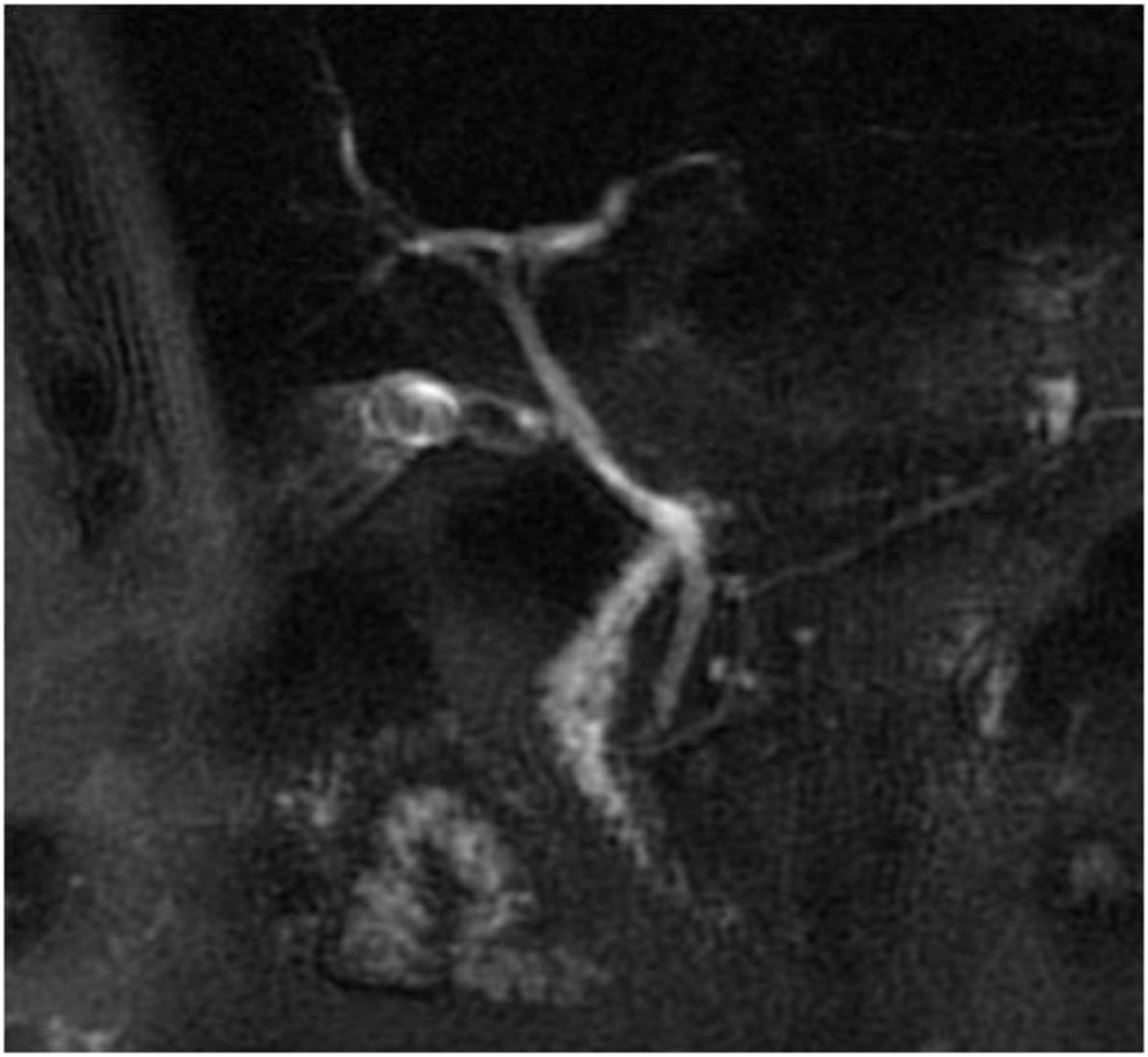

Fig.: MR cholangiogram shows triple confluence of right anterior duct, right posterior duct, and left hepatic duct.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Several less common and usually more complicated anatomic variations of the bile ducts have been described and consist of both **aberrant and accessory bile ducts**: an aberrant bile duct is the only bile duct draining a particular hepatic segment, whereas an accessory one is an additional bile duct draining the same area of the liver.

Fig.: Complex biliary variant. MR cholangiogram shows drainage of right posterior duct into left hepatic duct before joining right anterior duct. There is also an accessory left duct (arrow) draining into left hepatic duct.
References: J. F. Costa; Radiology, Hospitais da Universidade de Coimbra, Coimbra, PORTUGAL

Conclusion

Recognizing these different anomalies is essential since many of them can mimic pathological conditions and others are important in surgical planning, such as living donor liver transplantation.

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