



# 门脉系统疾病影像学诊断

北京协和医院

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2017-6-4, 福州

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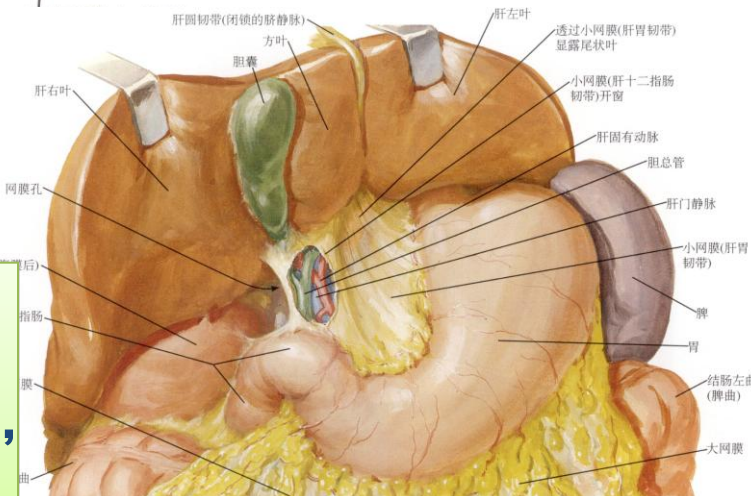
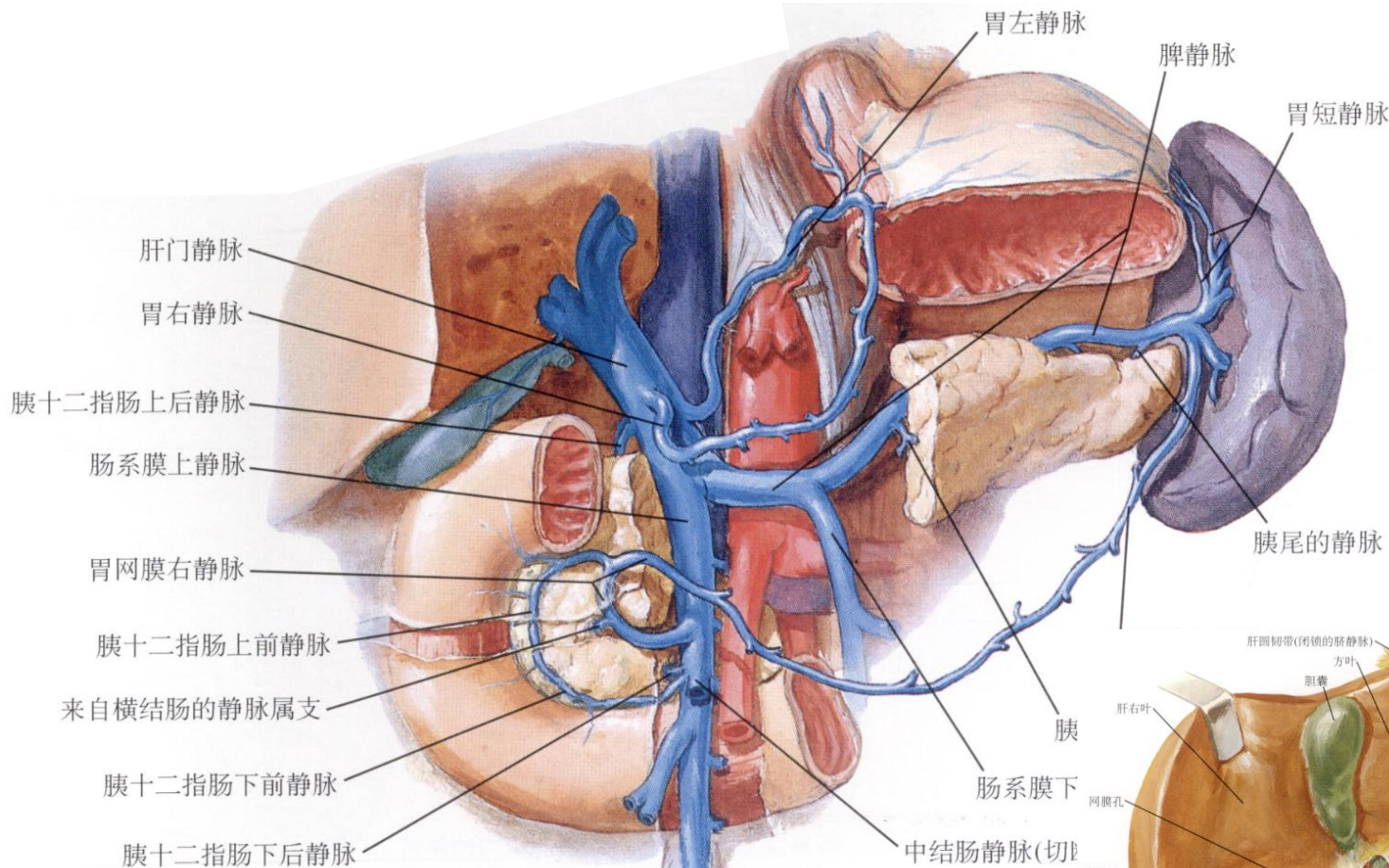
1 • 门脉解剖、变异与胚胎学

2 • 门脉发育及结构异常

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# 门静脉位置毗邻

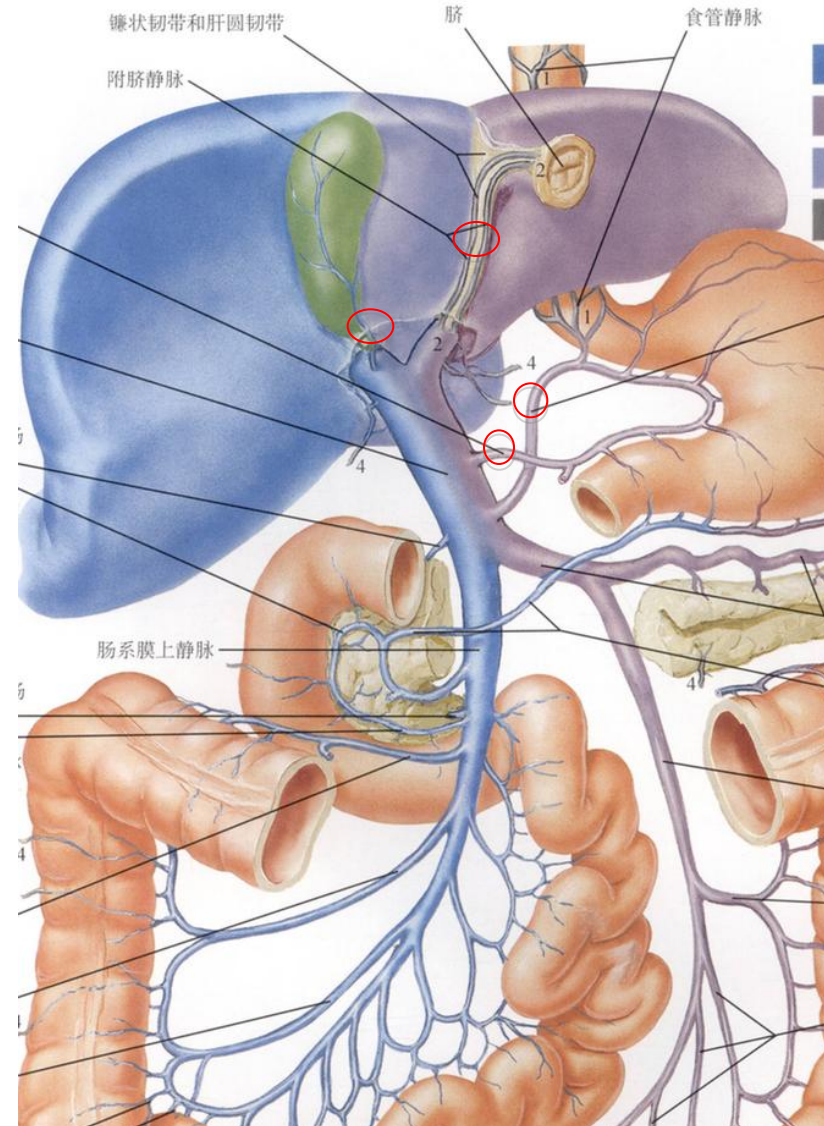


自胰颈的后方上行，通过十二指肠上部的深面后进入肝十二指肠韧带，上行至第一肝门  
 在肝十二指肠韧带内，肝门静脉行的右前面为胆总管，左前面为肝固有动脉

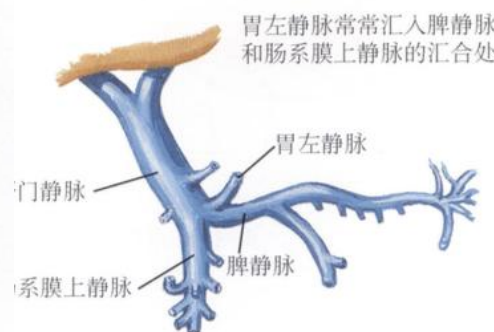
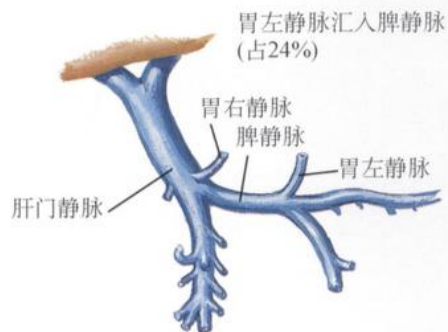
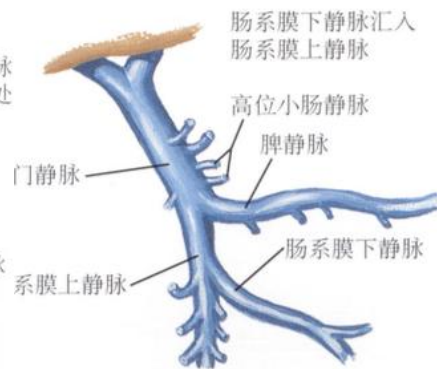
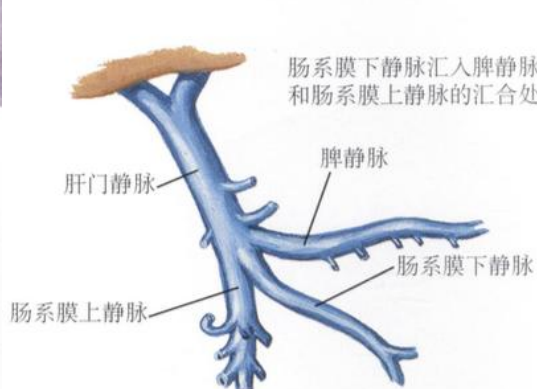
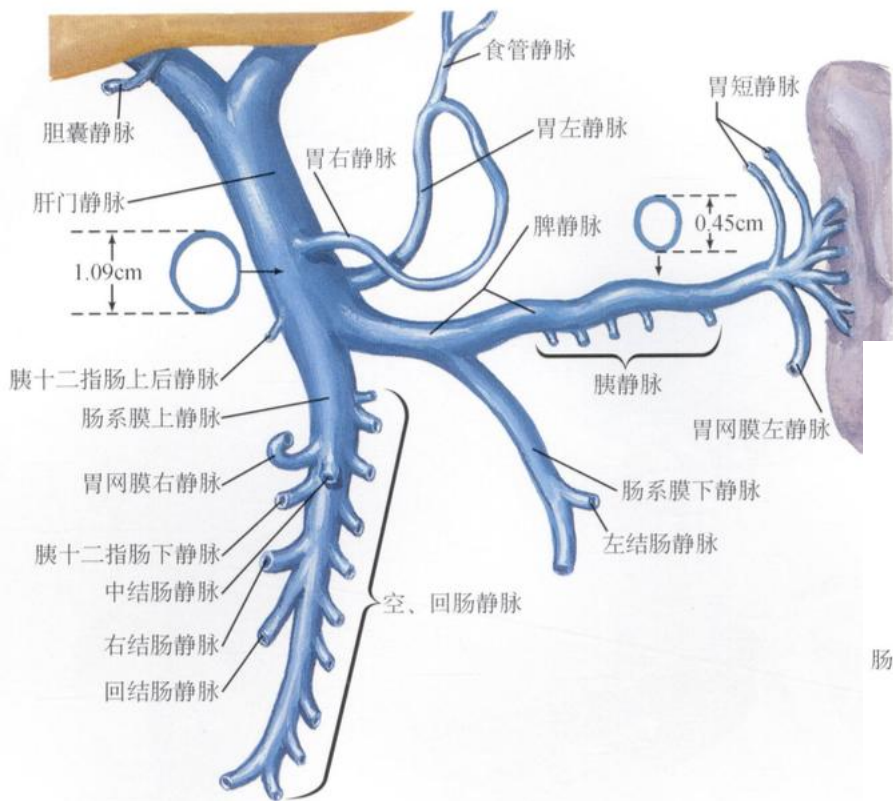
# 门静脉的主要属支

- (一) 肠系膜上静脉
- (二) 脾静脉
- (三) 肠系膜下静脉
- (四) 胃左静脉，也叫胃冠状静脉，沿胃小弯左行再转向右后汇入门静脉干。在贲门处食管静脉丛有小支汇入胃左静脉，其主支食管静脉汇入奇静脉或半奇静脉，从而使门静脉系和上腔静脉系沟通
- (五) 胃右静脉，汇入门静脉干
- (六) 胆囊静脉，收集胆囊壁的血液，汇入门静脉干或其右支
- (七) 附脐静脉，为数条细小的静脉，起于脐周静脉网，沿肝圆韧带走行，汇入门静脉或其左支

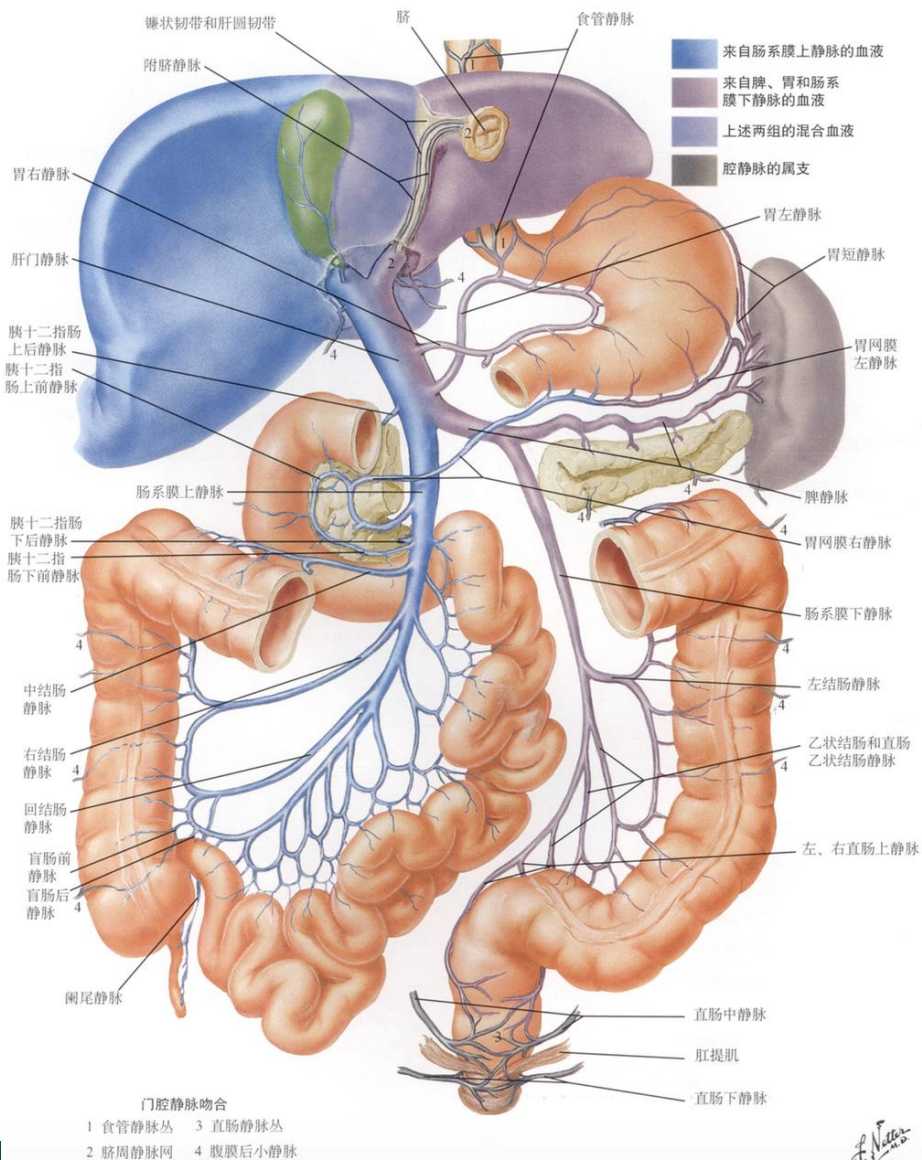
收集腹腔不成对脏器(除肝脏外)的静脉血  
肝门静脉两端均为毛细血管，门静脉系缺乏静脉瓣，血液易逆流



# 门静脉系统解剖变异



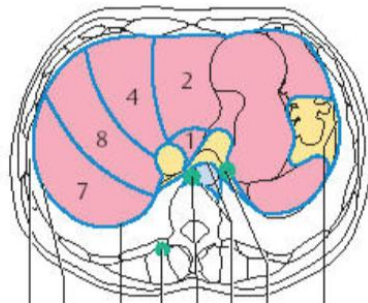
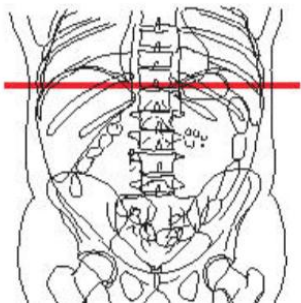
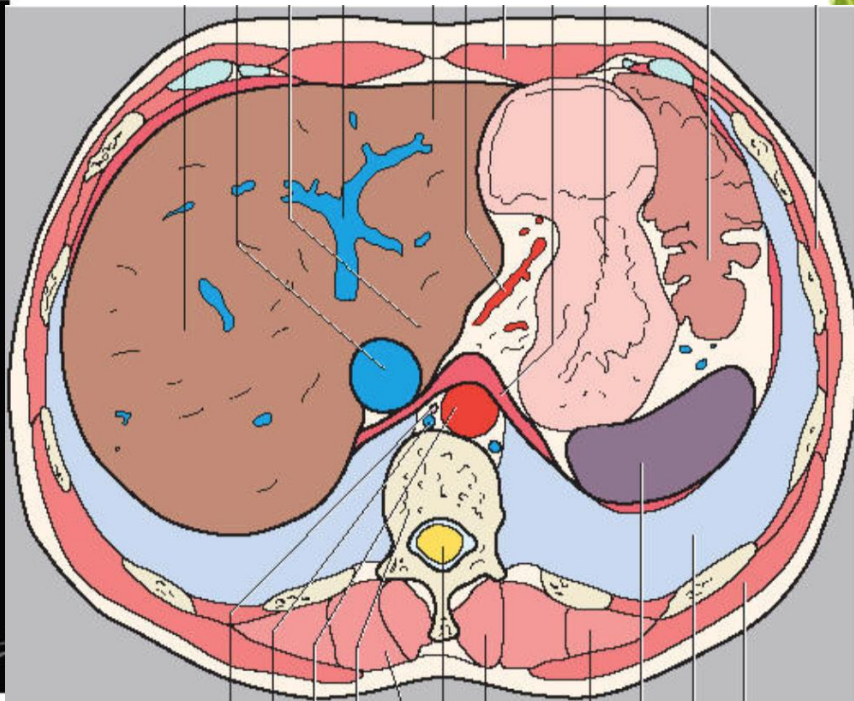
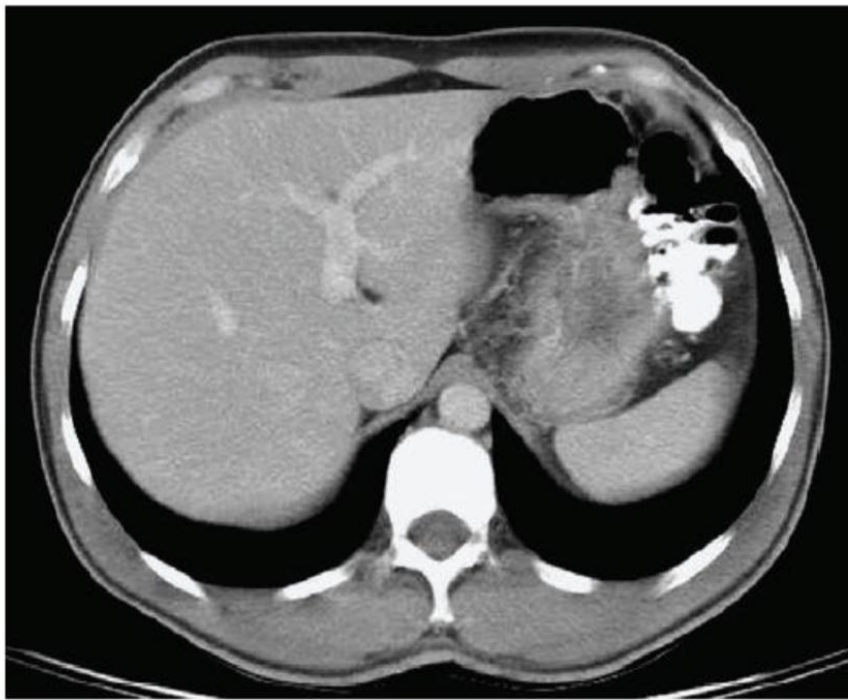
# 门脉的分流现象



临床意义:

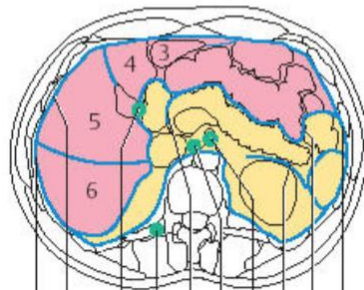
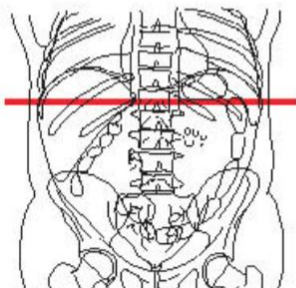
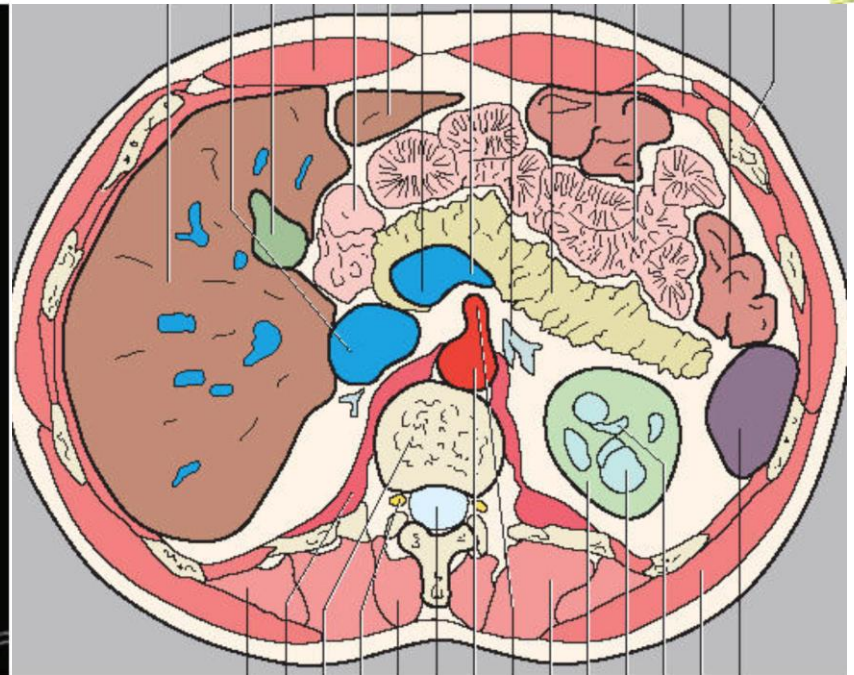
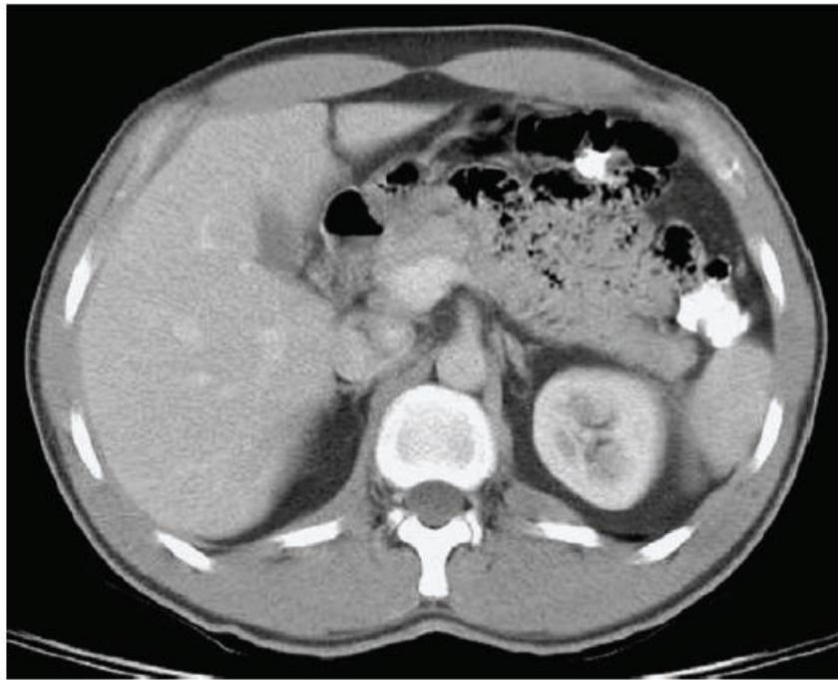
- 肝脓肿、脂肪肝等，右半肝好发
- 动脉期门静脉强化不均(与门静脉血栓鉴别)

# 门静脉相关断层解剖



Pocket Atlas Sectional Anatomy Computed Tomography and Magnetic Resonance Imaging

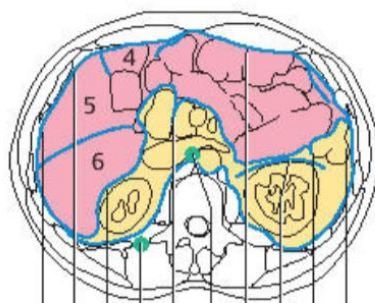
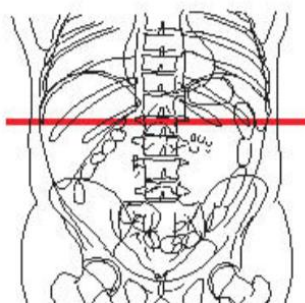
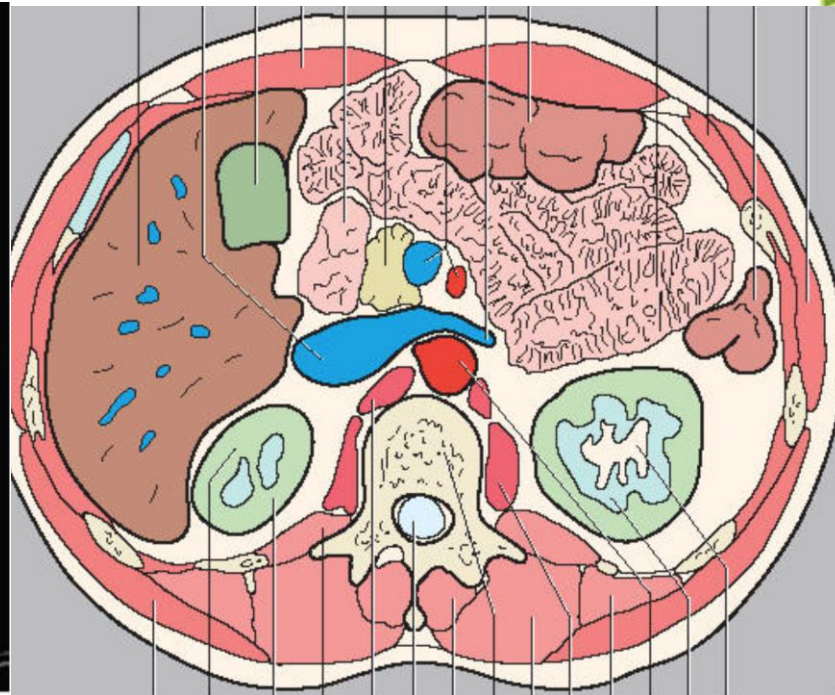
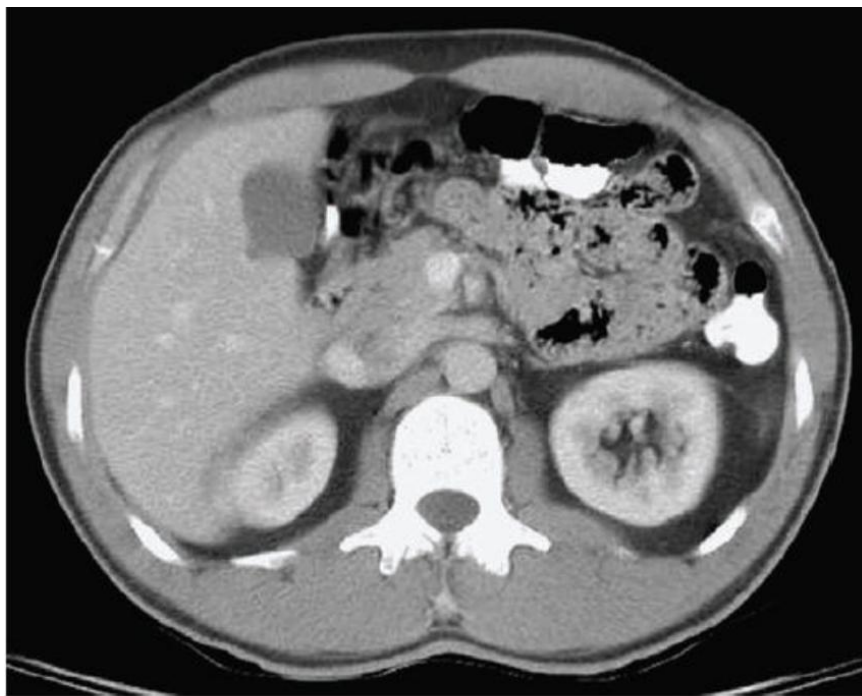
# 门静脉相关断层解剖



Pocket Atlas Sectional Anatomy Computed Tomography and Magnetic Resonance Imaging

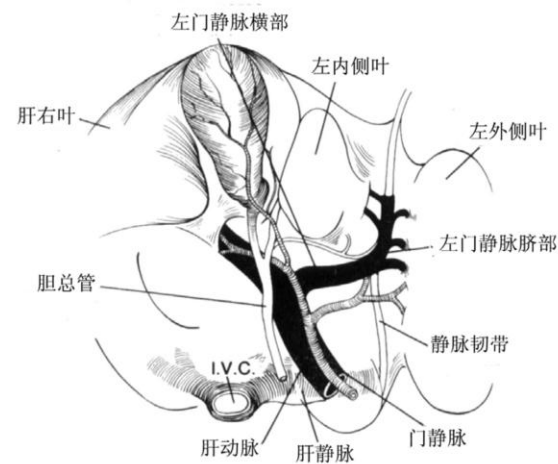
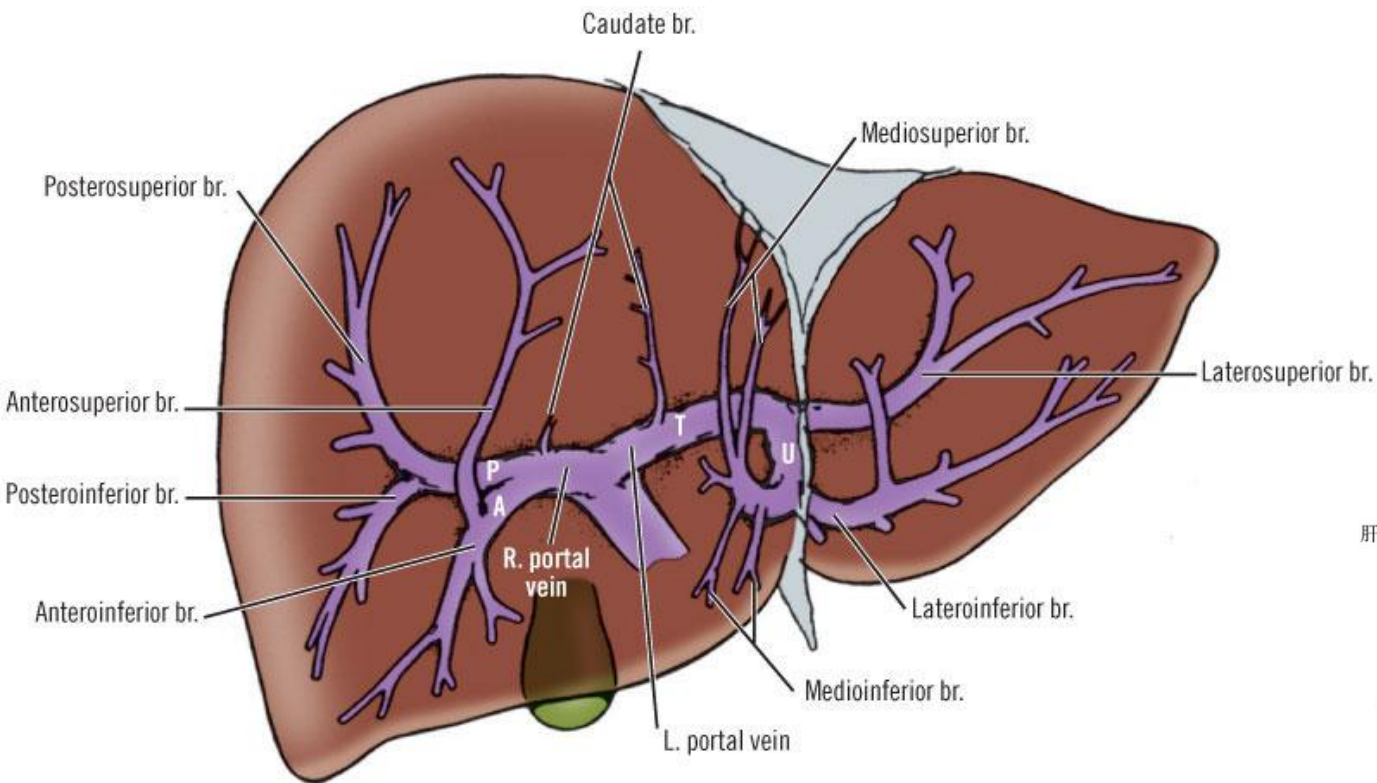


# 门静脉相关断层解剖



Pocket Atlas Sectional Anatomy Computed Tomography and Magnetic Resonance Imaging

# 门脉肝内分支



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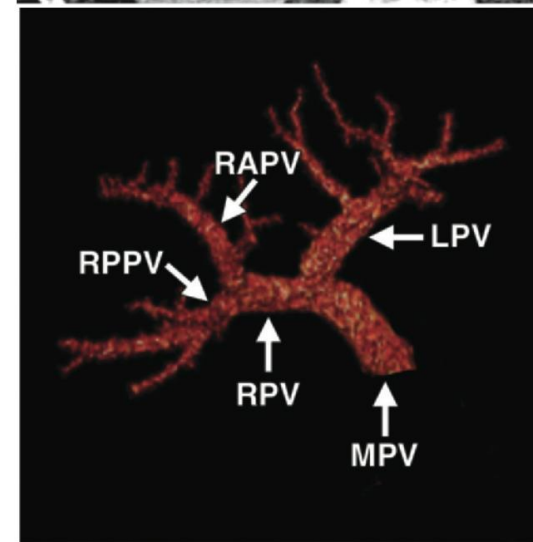
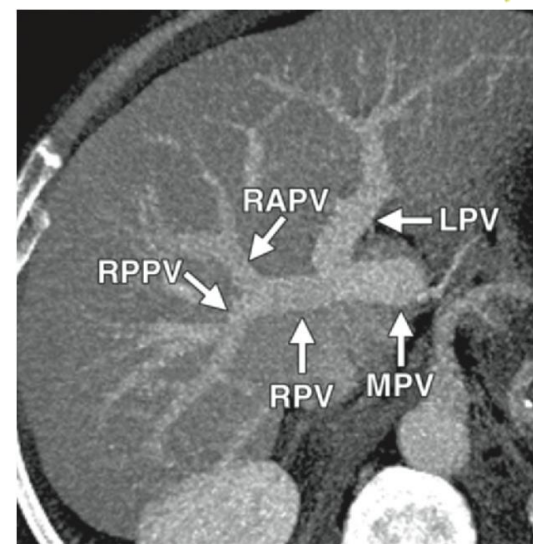
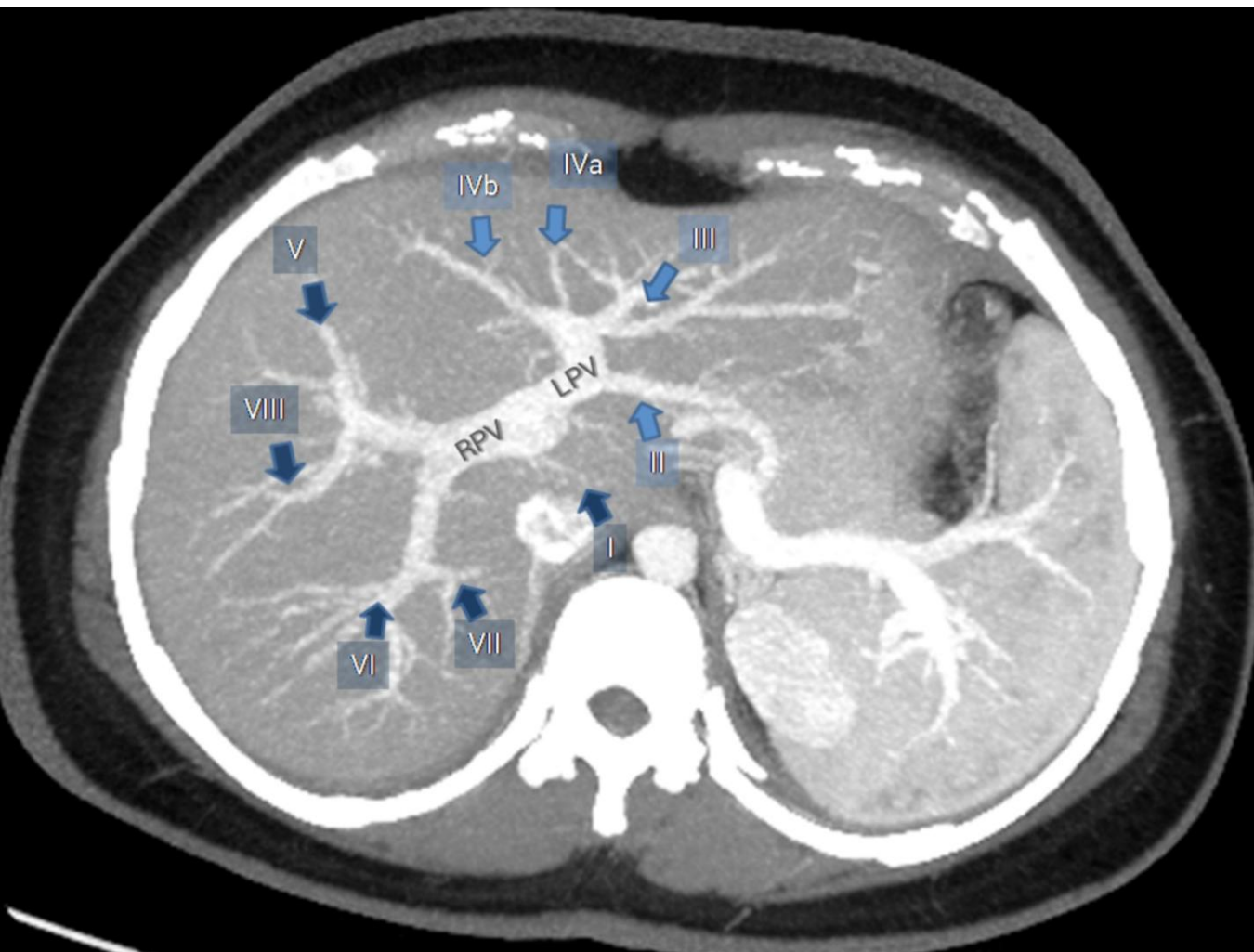
右支短、宽，变异较多

- 前支、后支

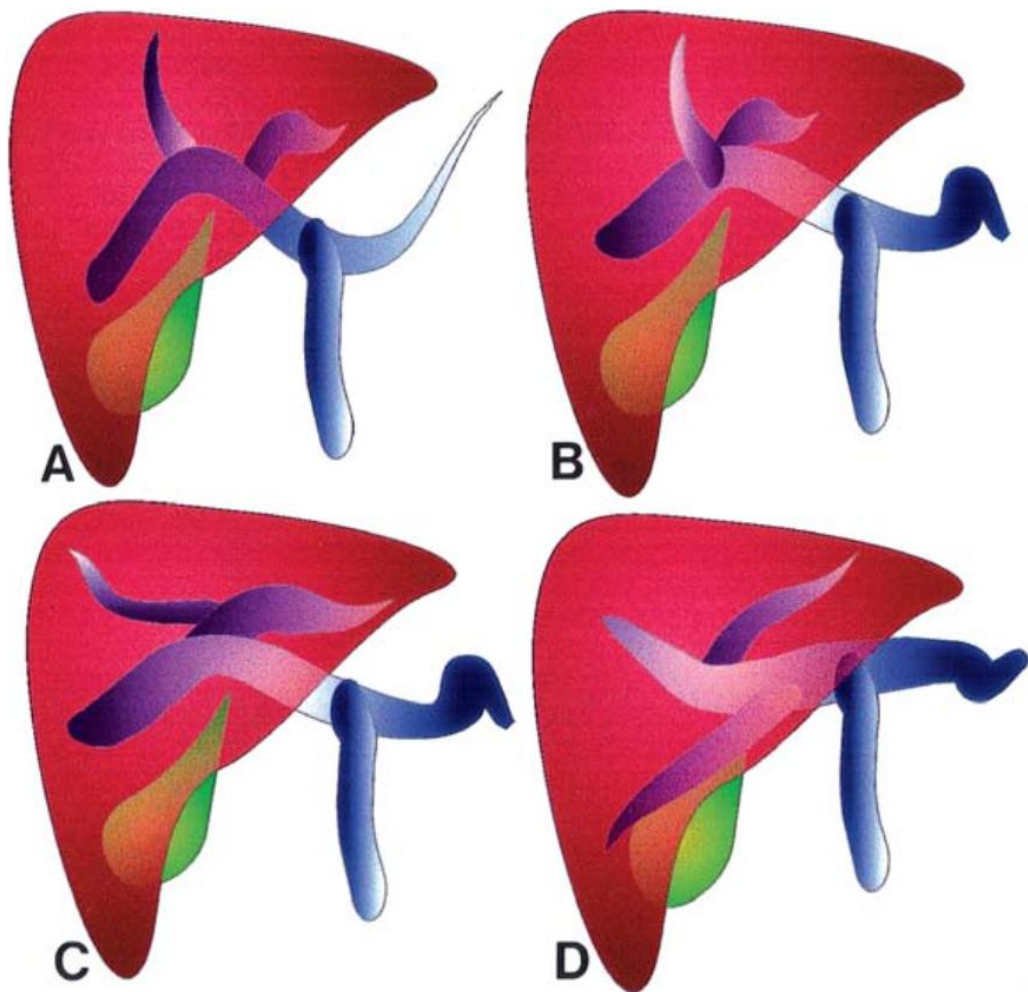
左支长、细，变异较少

- 主干沿横沟左行（横部）至左纵沟，弯向前上方，移行于脐部，发出外侧和内侧支

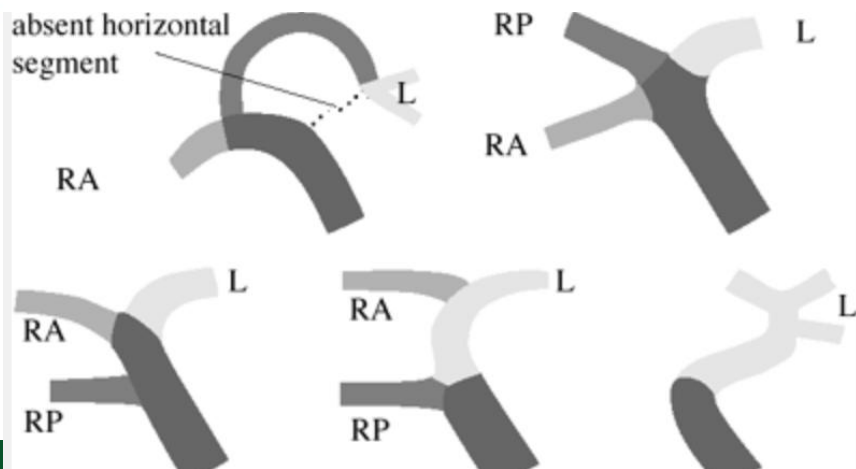
# 门脉肝内分支



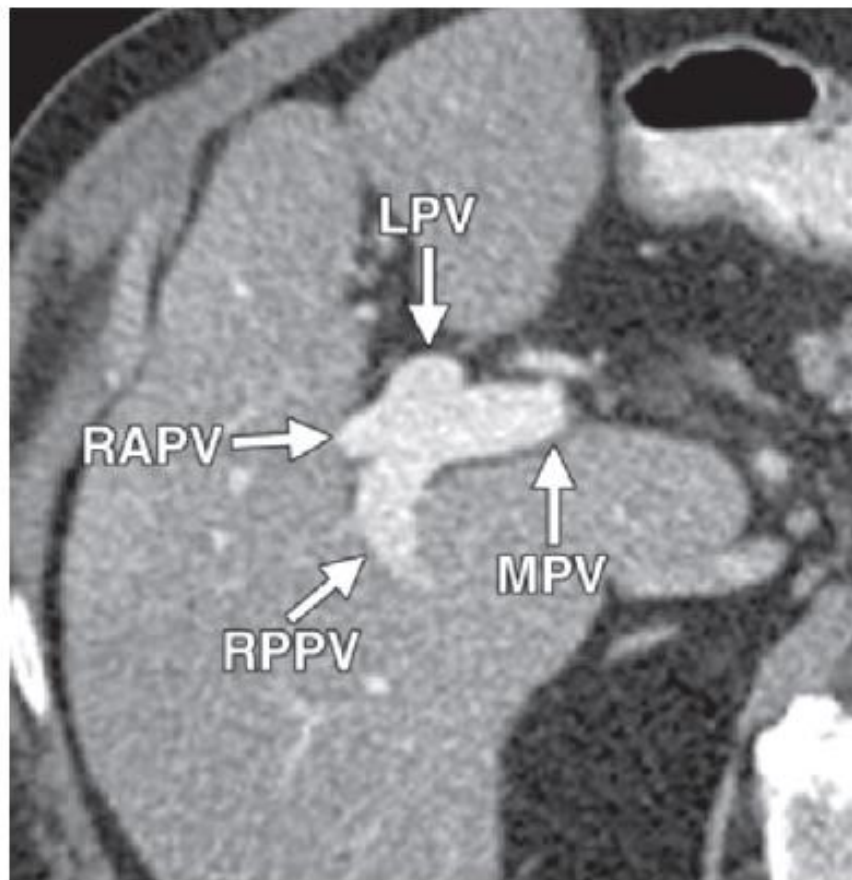
# 门脉肝内分支变异



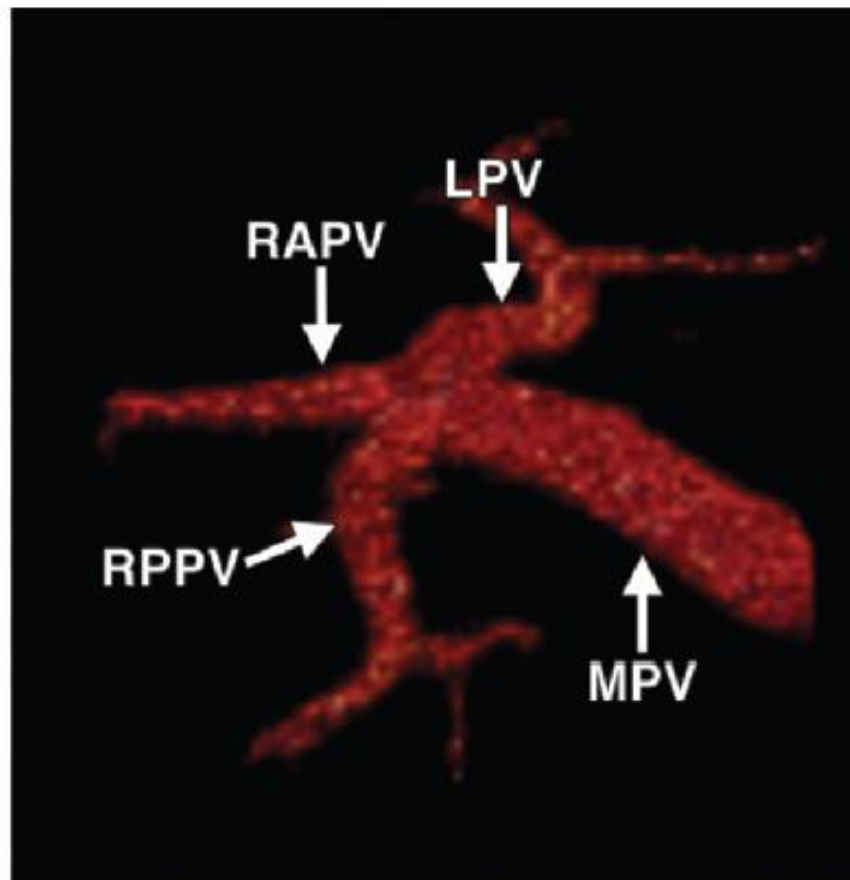
- 右支的变异主要包括
  - 三叉状：门静脉主干在肝门处直接分为左支、右前支和右后支
  - 主干发出右后支，然后向左侧略弯曲斜行，演变为左支，右前支起自左支
  - 主干发出右后支，然后继续向右上行分为左支和右前支
- 左支的变异主要是左支水平段缺如
- 门脉右支主干、右前支及右后支缺如，主干分出肝右叶分支后，转向左侧，延续为左支



# 门脉肝内分支变异



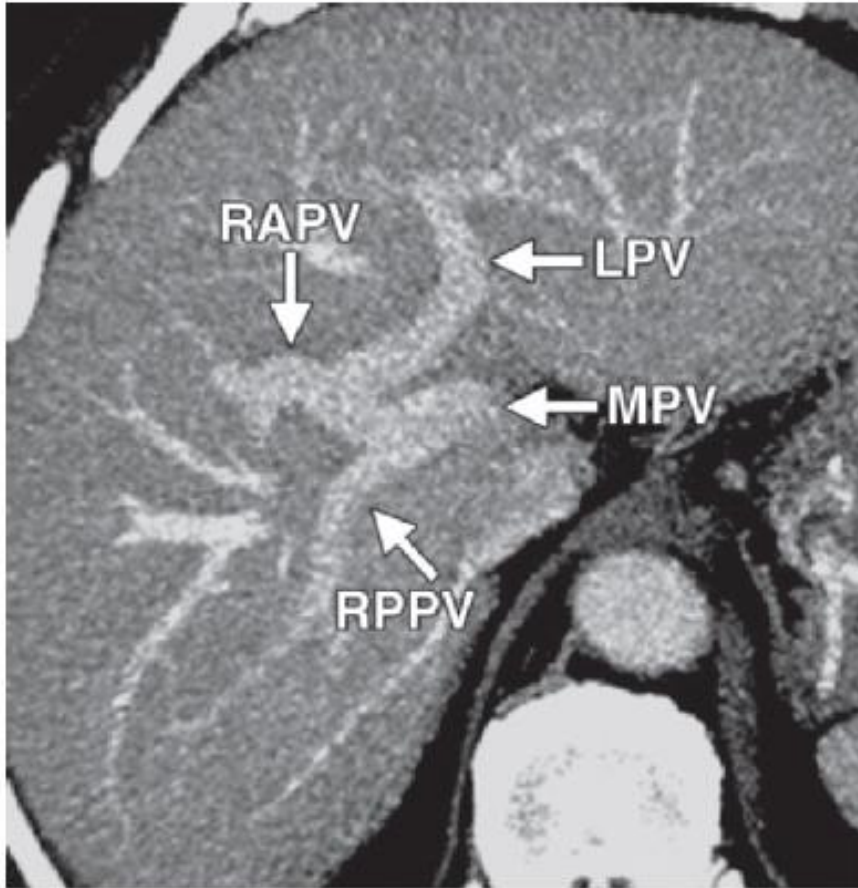
a.



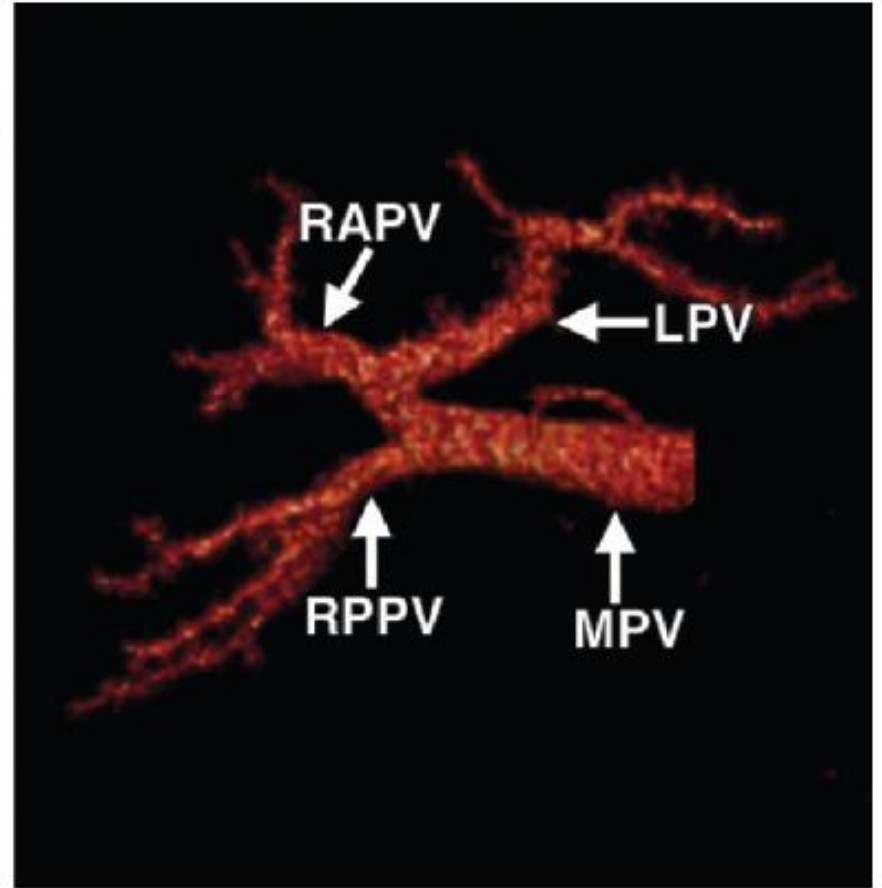
b.

门静脉主干同时分出**3**支血管

# 门脉肝内分支变异



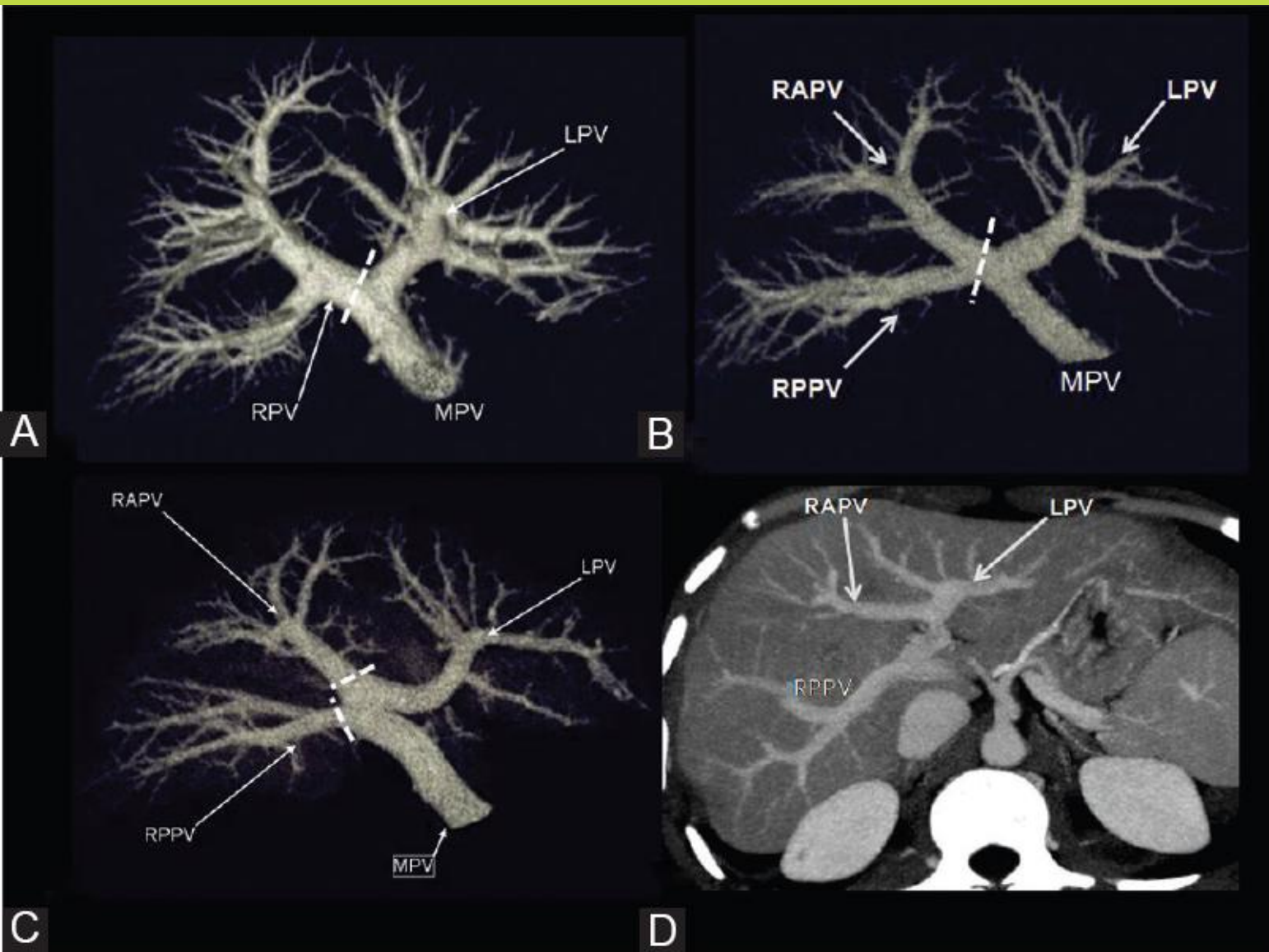
a.



b.

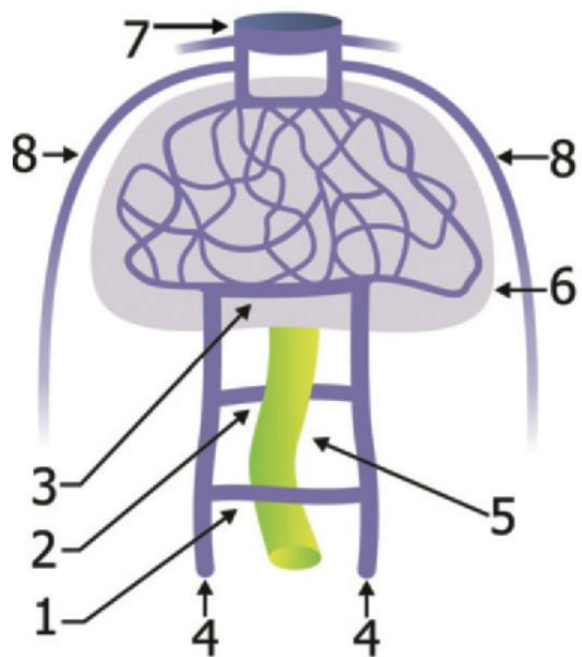
门脉右后支是门脉主干发出的第一个分支，门脉左支和右前支共干

# 门脉肝内分支变异

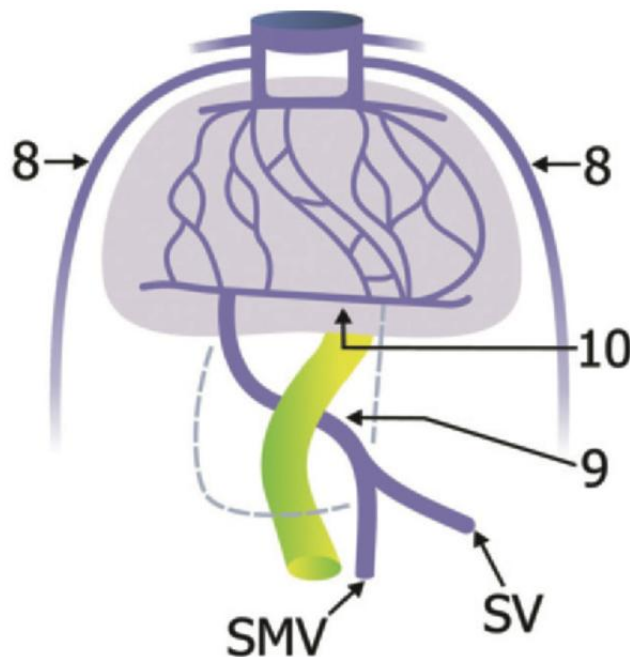


Indian J Radiol  
Imaging. 24(4):  
350-359, 2014

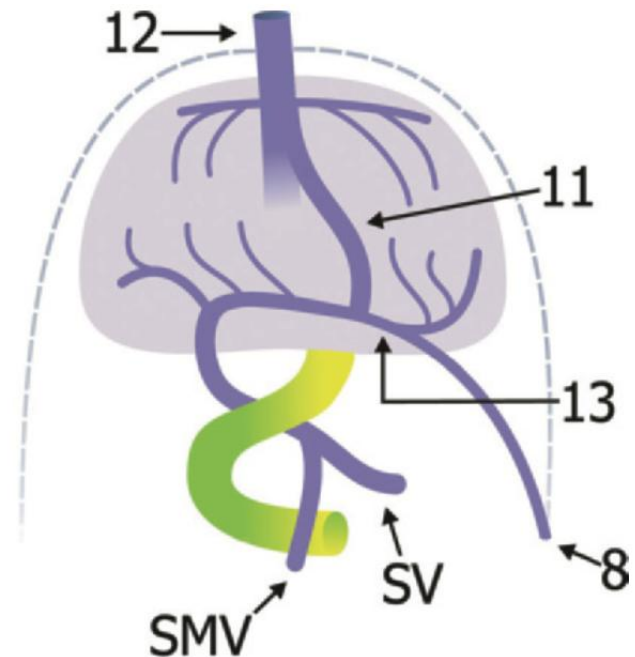
# 门脉的胚胎发育学



- [4]成对卵黄静脉
- 头腹侧[3]、背侧[2]、尾腹侧
- [1]吻合静脉
- [8]成对脐静脉，
- [7]静脉窦（原始心脏），
- [5]十二指肠，
- [6]横隔（原始肝脏）



- [9]门脉主干，
- [10]门脉左支



- [11]静脉导管，
- [12]下腔静脉，
- [13]门脉左支与左脐静脉间交通，
- [8]左脐静脉



# 门脉发育及结构异常



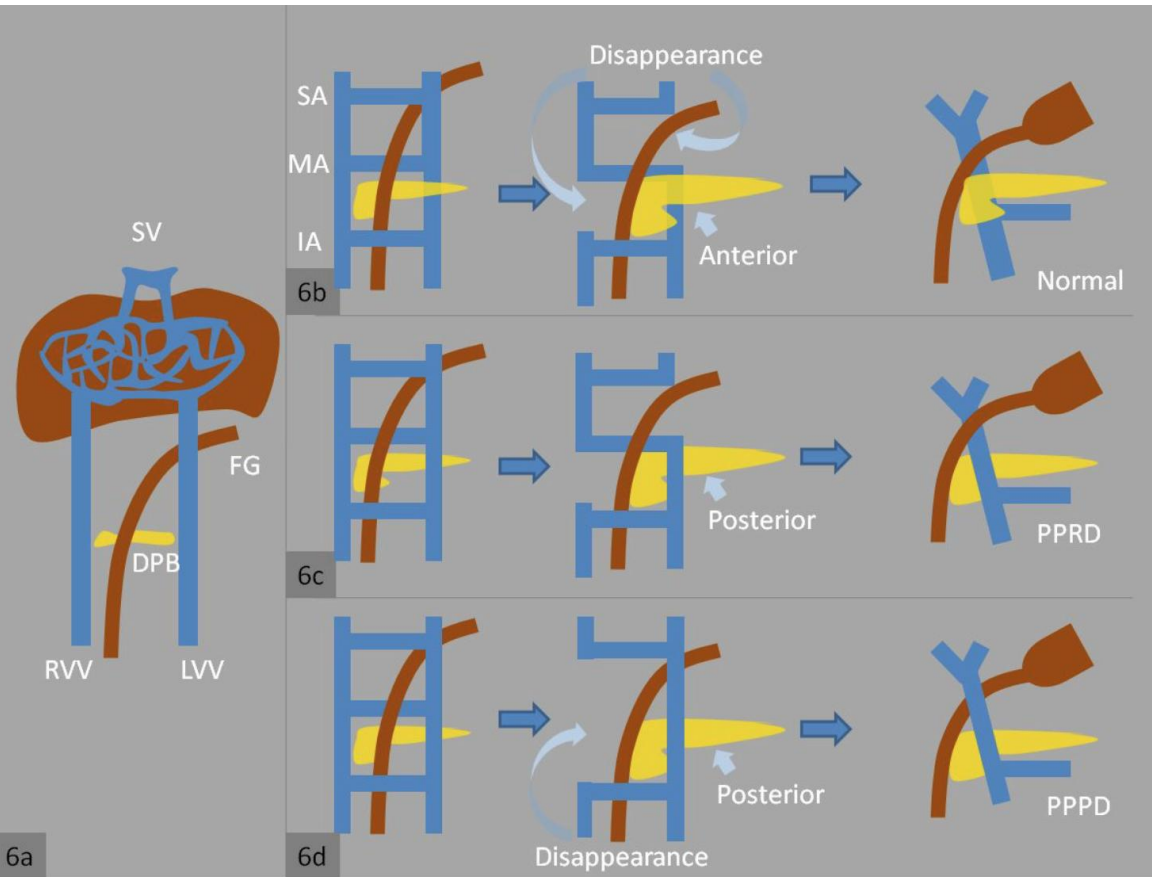
胰腺前方门静脉

双门静脉

先天性肝外门体分流

肝内门体分流

# 门脉发育及结构异常 - 罕见病变



正常：

门静脉位于胰腺及十二指肠后方

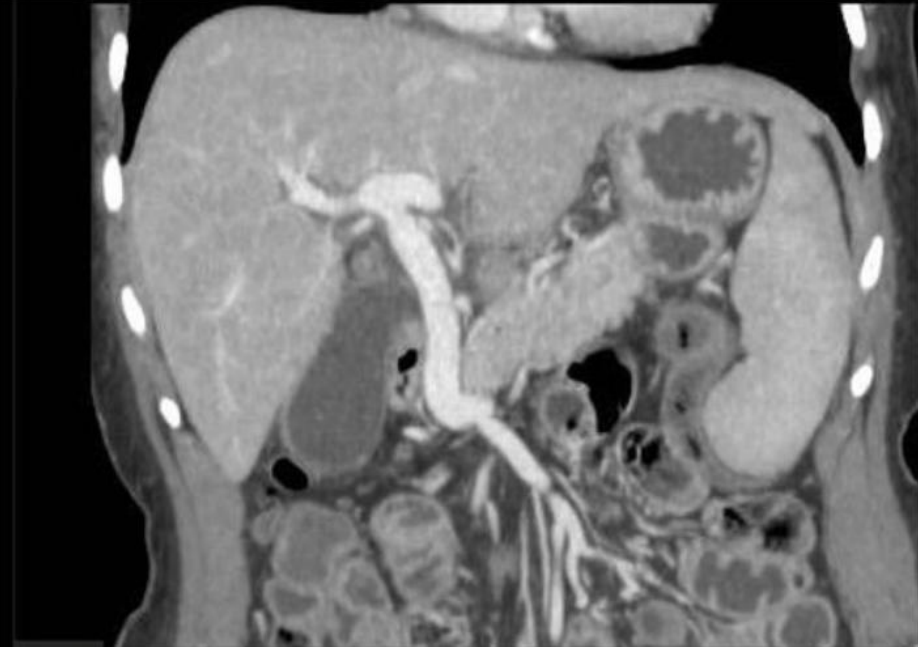
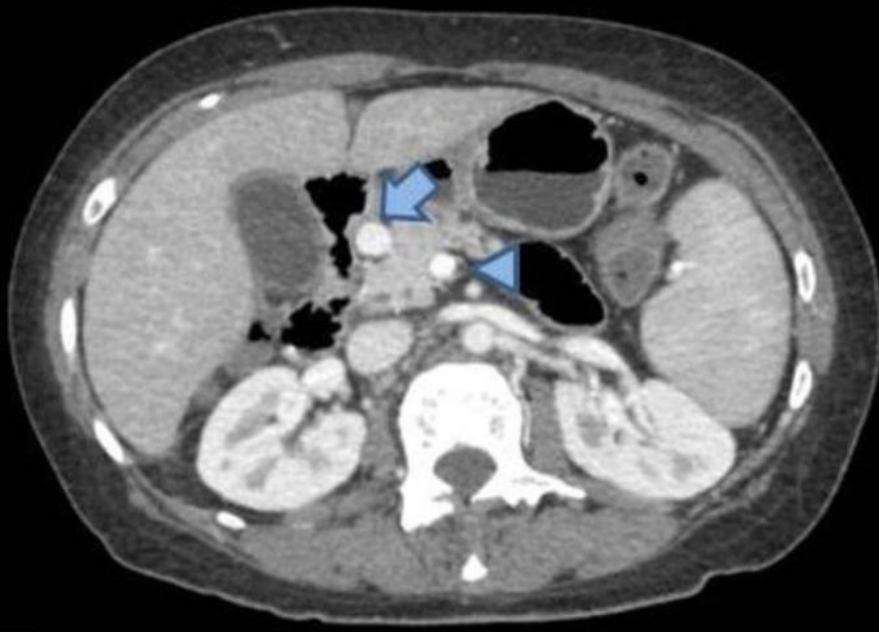
胰腺前方十二指肠后方门静脉  
Prepancreatic retroduodenal  
(PPRD) portal vein

胰腺前方十二指肠前方门静脉  
Prepancreatic preduodenal  
(PPPD) portal vein

- 胰腺前方门静脉 prepancreatic portal vein
- 十二指肠前方门静脉 preduodenal portal vein

# 门脉发育及结构异常-罕见病变

胰腺前方十二指肠后方门静脉  
Prepancreatic retroduodenal (PPRD) portal vein



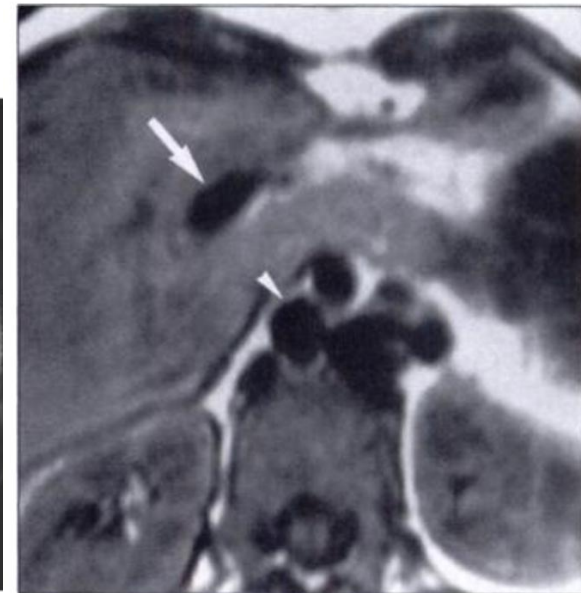
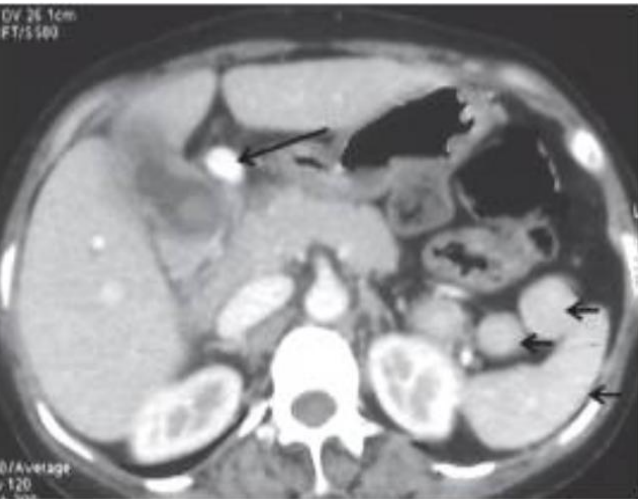
PPRD呈L型或反L型  
PPRD位于胆总管的前方或与其平行  
大多数PPRD不合并肠道、胆系与胰腺的发育异常



# 门脉发育及结构异常-罕见病变

胰腺前方十二指肠前方门静脉

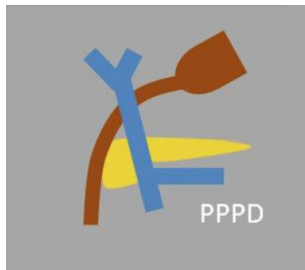
Prepancreatic preduodenal (PPPD) portal vein



PPPD

位于胆总管前方

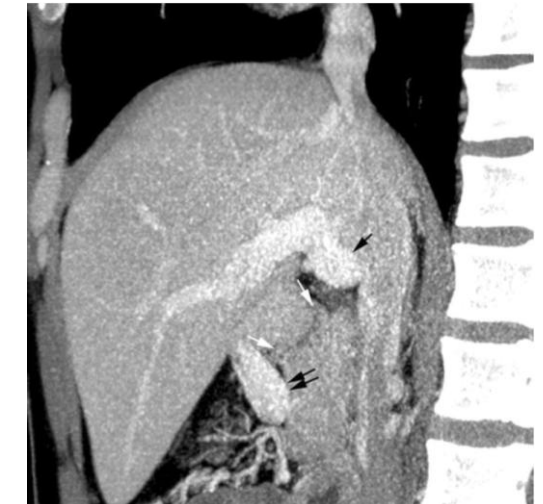
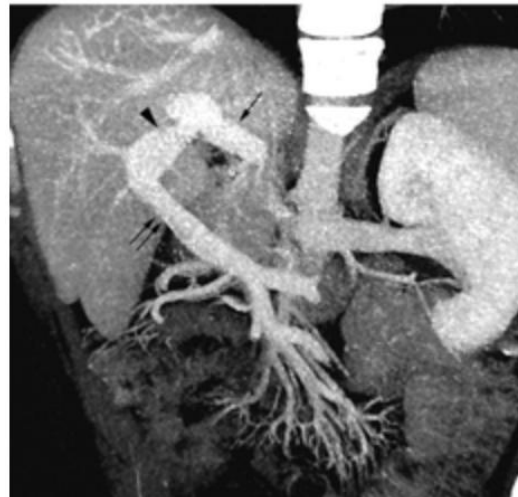
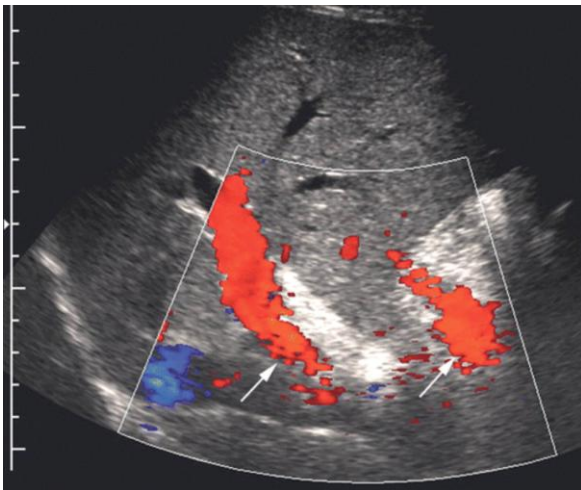
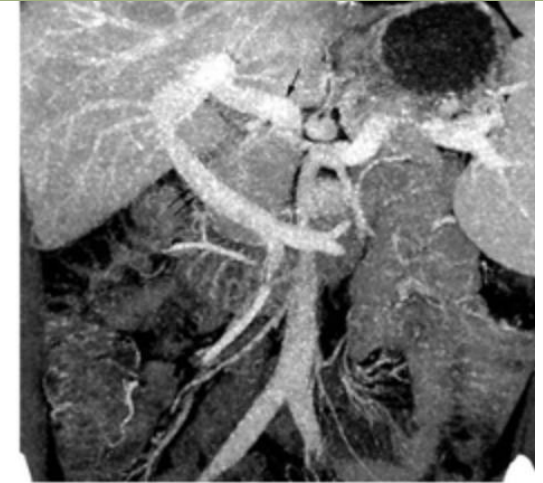
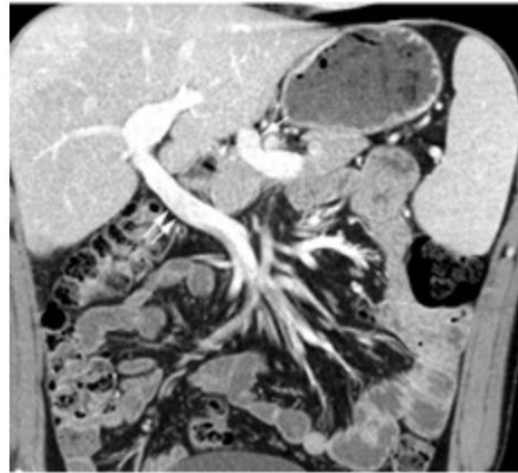
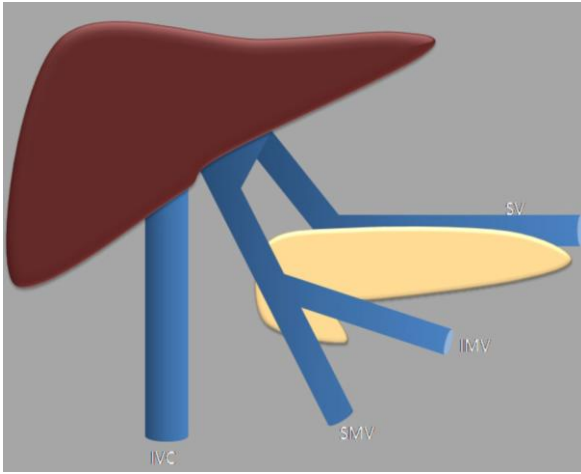
常合并有肠道、胆系、胰腺、脾脏、心血管系统的异常



Polysplenia syndrome with preduodenal portal vein. Annals of Gastroenterology, 2013  
Imaging of Congenital Abnormalities of the Portal Venous System, AJR, 1997

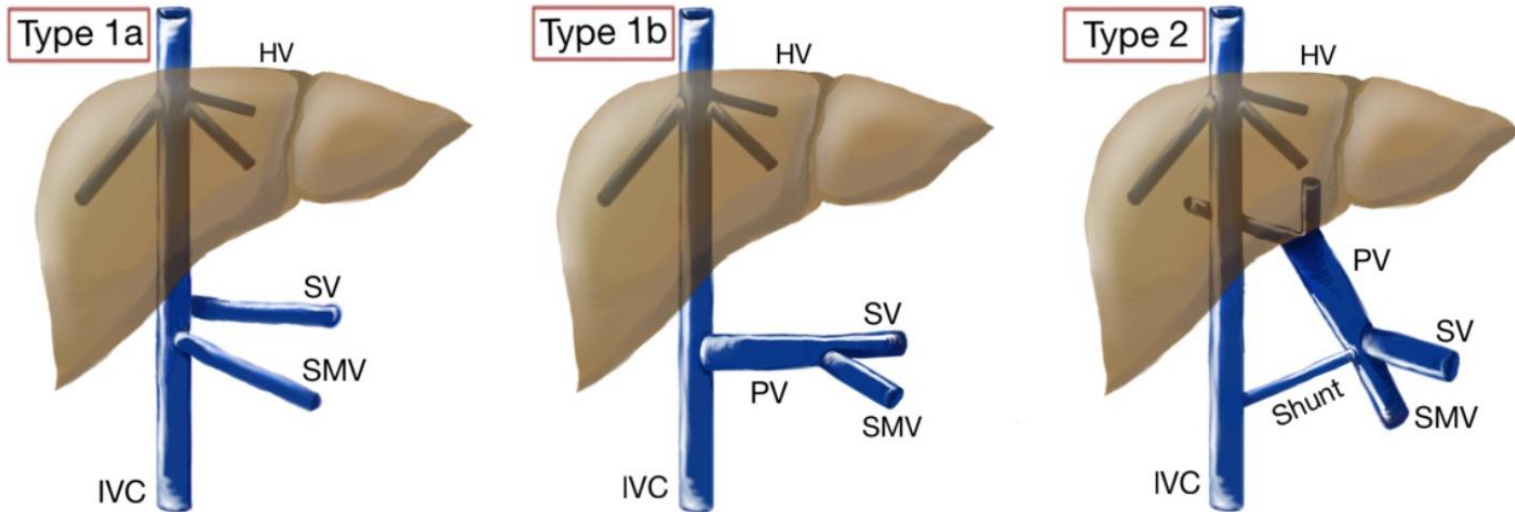
# 门脉发育及结构异常-罕见病变

## 双门静脉 Portal vein duplication



# 门脉发育及结构异常-罕见病变

先天性肝外门腔分流 (congenital extrahepatic portocaval shunt, Abernethy malformation)  
(门静脉缺如, congenital absence of the portal vein)



先天性肝外门腔分流分型

Type 1a: 端侧门腔吻合，肠系膜上静脉（SMV）及脾静脉（SV）单独汇入IVC

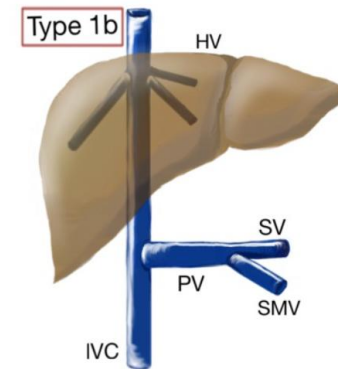
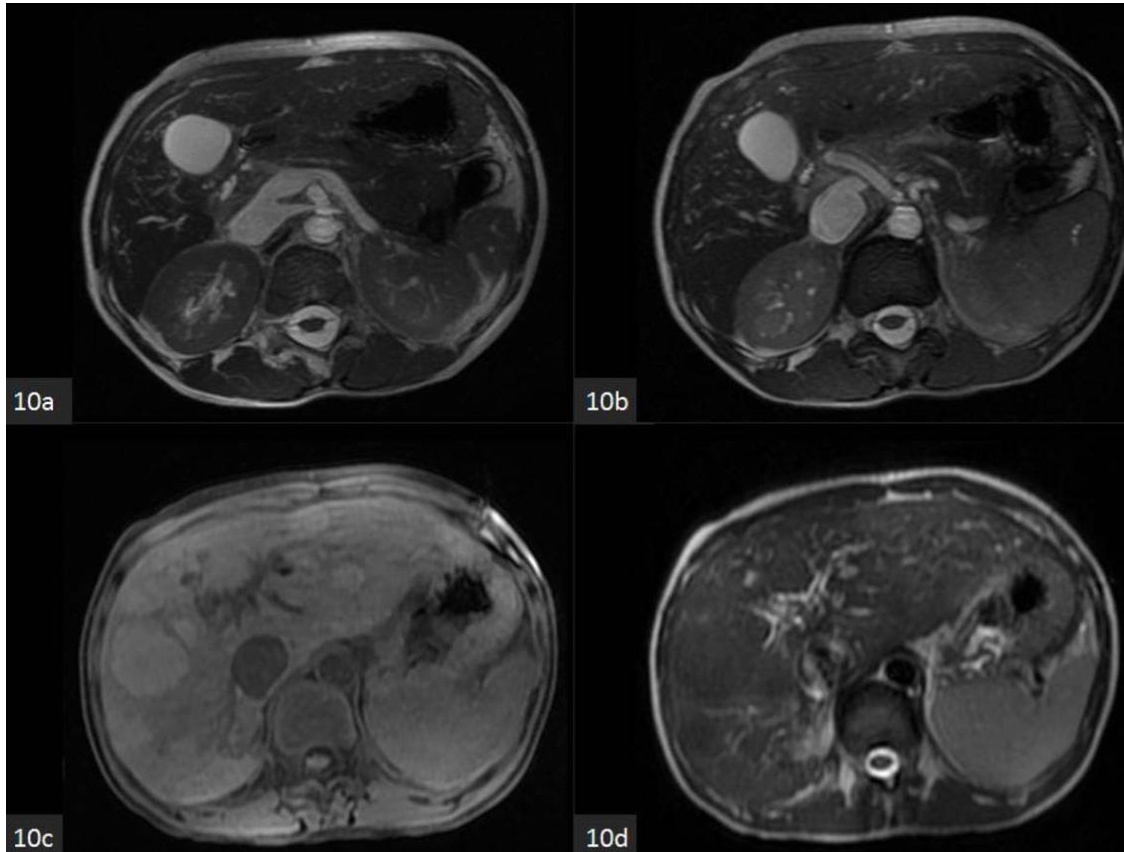
Type 1b: 端侧门腔吻合，肠系膜上静脉与脾静脉共干成门脉（PV）后汇入

Type 2: 侧侧门腔吻合，IVC与门脉间

肺动脉高压，肝硬化及肝性脑病（较晚出现）

# 门脉发育及结构异常-罕见病变

## 先天性肝外门腔分流 (congenital extrahepatic portocaval shunt)



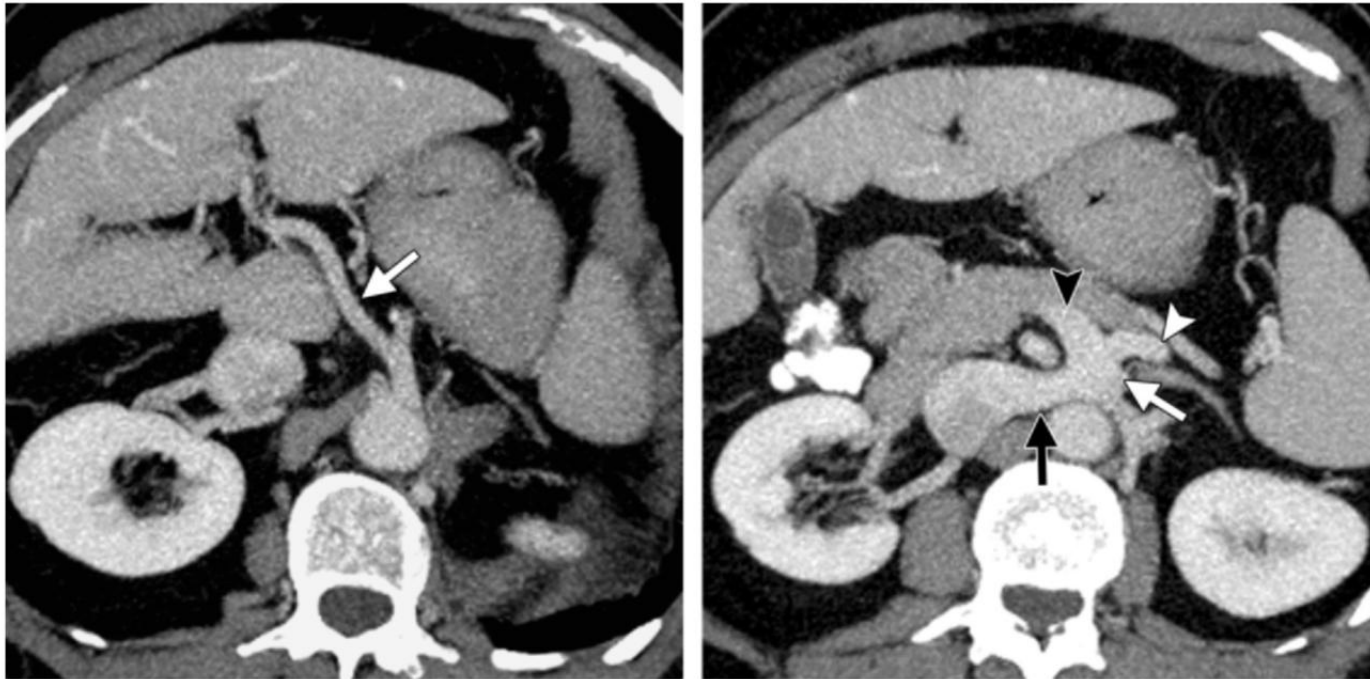
先天性肝外门腔分流分型

Type 1: 端侧门腔吻合

I型畸形多发生于女性，常合并其他脏器的先天畸形（如心脏的畸形、胆道闭锁和多脾）、肝脏结节样再生性增生和肿瘤

# 门脉发育及结构异常-罕见病变

## 先天性肝外门腔分流 (congenital extrahepatic portocaval shunt)



a.

b.

**Figure 5.** Extrahepatic portosystemic shunt in a 55-year-old man with congenital absence of the portal vein and pulmonary arterial hypertension. **(a)** Axial contrast-enhanced CT image at the level of the celiac artery shows a single vessel at the hepatic hilum (arrow) that represents an enlarged hepatic artery (7 mm in diameter). No intrahepatic or extrahepatic portal vein is depicted. **(b)** Axial contrast-enhanced CT image obtained at the level of the left renal vein shows the splenic vein (white arrowhead) and superior mesenteric vein (black arrowhead) uniting to form an abnormal splenomesenteric venous confluence (white arrow) that empties into the left renal vein (black arrow), findings indicative of an Abernethy type 1b portosystemic shunt.



# 门脉发育及结构异常-罕见病变

## 先天性肝内门体分流 (congenital intrahepatic portosystemic shunt)



肝内门体分流**4**种类型：

- a 门脉右支与下腔静脉间粗大通路
- b 某一肝段中门静脉与肝静脉间单发/多发交通
- c 外周门静脉与肝静脉间瘤样连通
- d 左右叶门静脉与肝静脉间有多发连通

分流量小：无明显症状

分流量大：肝功能异常、高血氨症与肝性脑病

治疗：

保守（有可能自发闭合）

介入栓塞、手术结扎或部分肝切除

# 门脉发育及结构异常-肝内门体分流



A



B



先天性肝内门体分流  
(congenital intrahepatic  
portosystemic shunt)

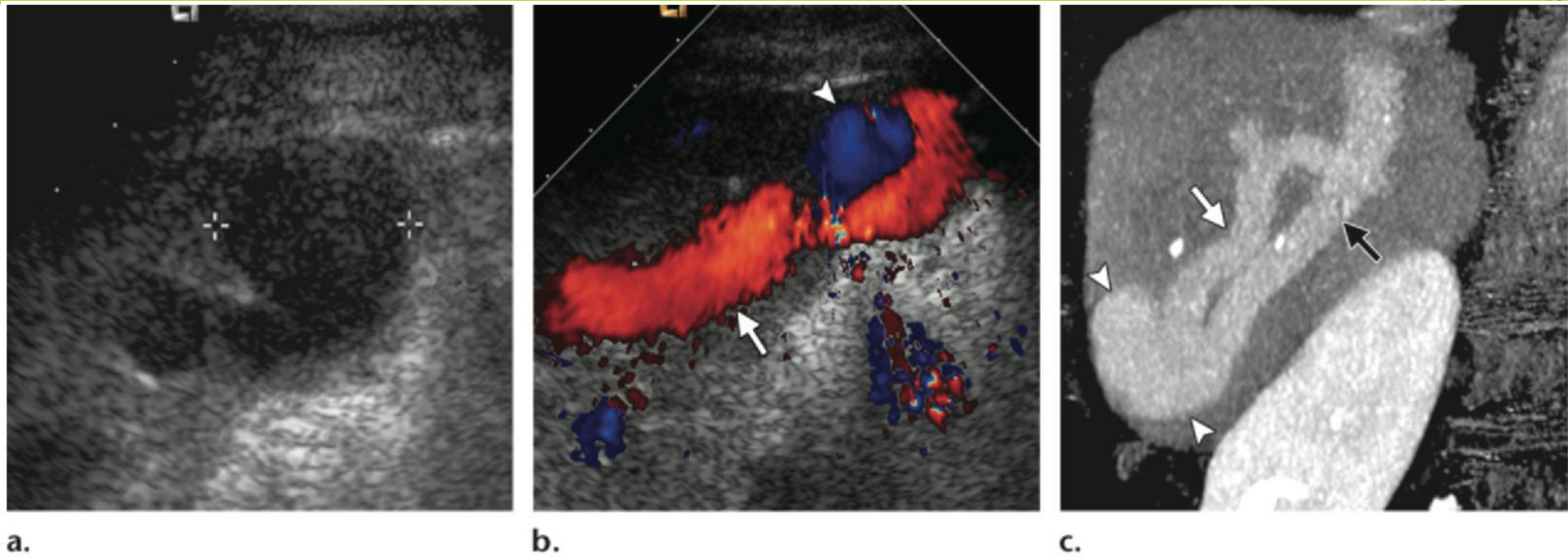
M,8y

门脉左支与肝左静脉间分流

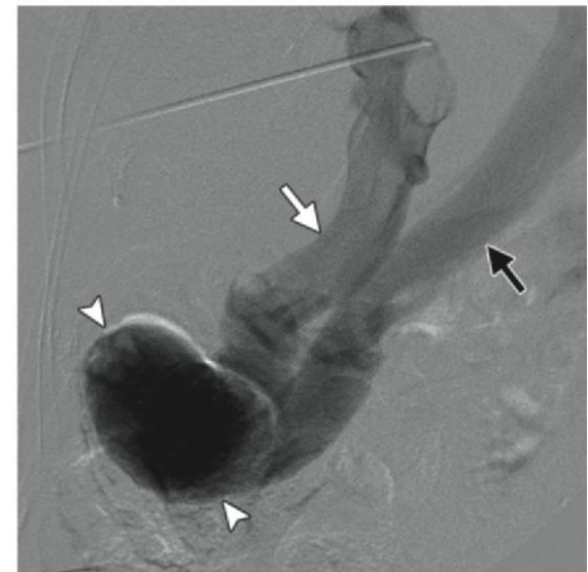
门脉右支细小

肝右叶占位

# 门脉发育及结构异常-肝内门体分流



**Figure 9.** Intrahepatic portosystemic shunt and focal varix in a 71-year-old woman with hepatic encephalopathy and no history of abdominal trauma or liver biopsy. **(a, b)** Oblique gray-scale **(a)** and color Doppler **(b)** sonograms show a 4-cm-diameter, peripheral anechoic mass (area between the cursors in **a**) with turbulent flow (arrowhead in **b**) in hepatic segment VI. The mass is supplied by an enlarged posterior branch of the right portal vein (arrow in **b**). **(c, d)** Coronal contrast-enhanced maximum intensity projection CT image **(c)** and transhepatic right portal venogram **(d)** show a focal venous dilatation (arrowheads) that is contiguous with an enlarged posterior branch of the right portal vein (white arrow) and an early-enhancing, enlarged right hepatic vein (black arrow), findings indicative of an intrahepatic portosystemic shunt with varix formation. Coil embolization of the varix was successful, and the hepatic encephalopathy resolved.



# 门脉疾病症候群



- 动门脉分流
- 门体静脉分流，门静脉高压
- 门静脉血栓，门脉海绵样变
- 门静脉瘤
- 门脉积气
- 闭塞性门静脉病

# 动门脉分流

肝内动门脉分流（arterioportal shunt, APS）是最常见的肝内血管分流病因

- 出生即存在的肝内血管分流，即先天性（如Osler-Weber-Rendu综合征）
- 后天性或获得性，如继发于良性肿瘤（例如血管瘤）、恶性肿瘤（例如肝癌）、外伤、医源性损伤、肝硬化等

临床表现

- 变异较多，可以无症状，
- 可在致病因素的症状外，出现分流引起的症状和体征（如高输出性心力衰竭），及并发症（如门脉高压）的症状

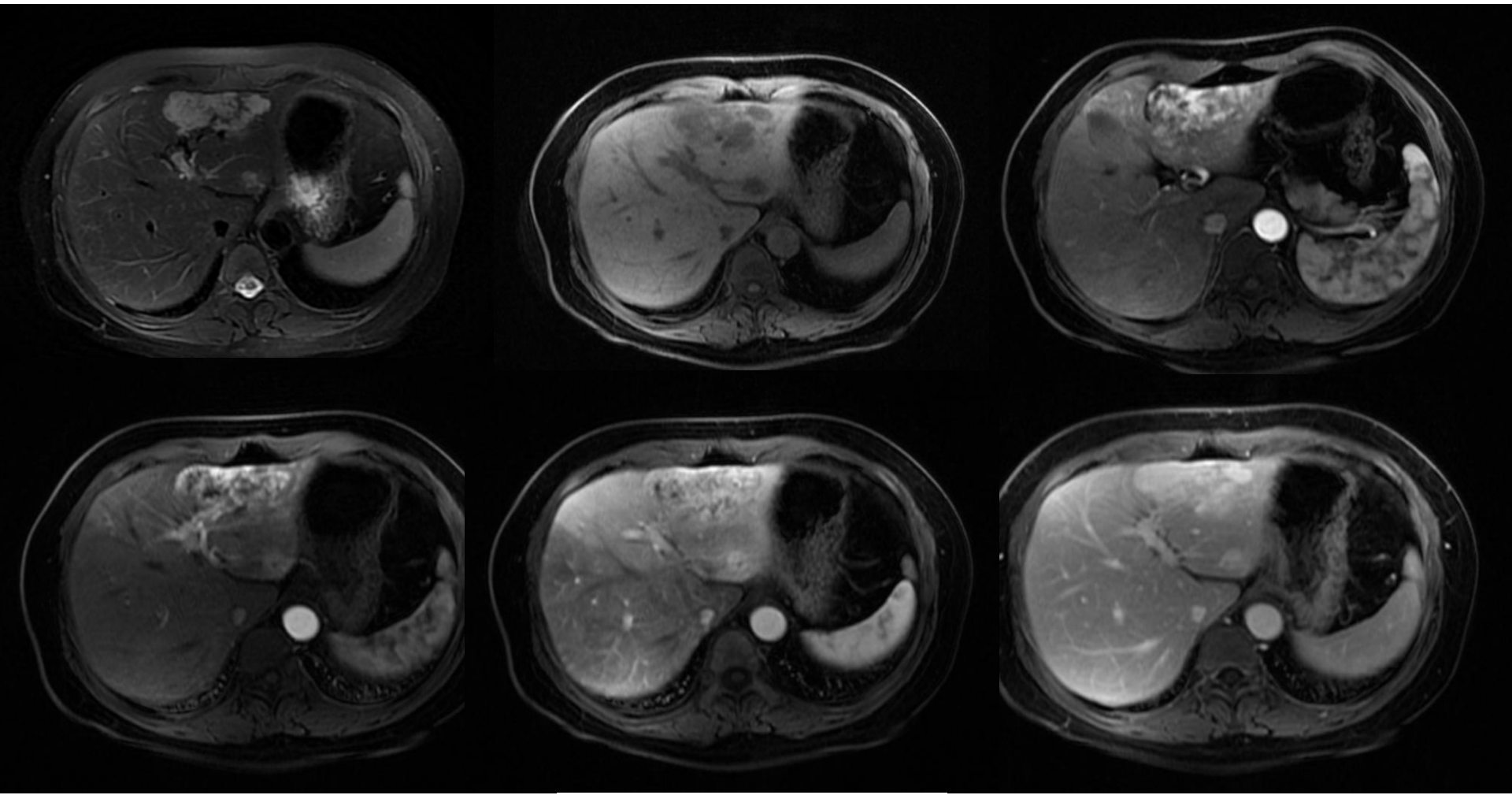
器质性分流

- 门静脉分支早期强化（增强早于门静脉主干），表现为与肝动脉伴行的强化血管影
- 动脉期肝实质一过性灌注异常，多呈边界清晰的高强化区，在门脉期呈等密度/等信号

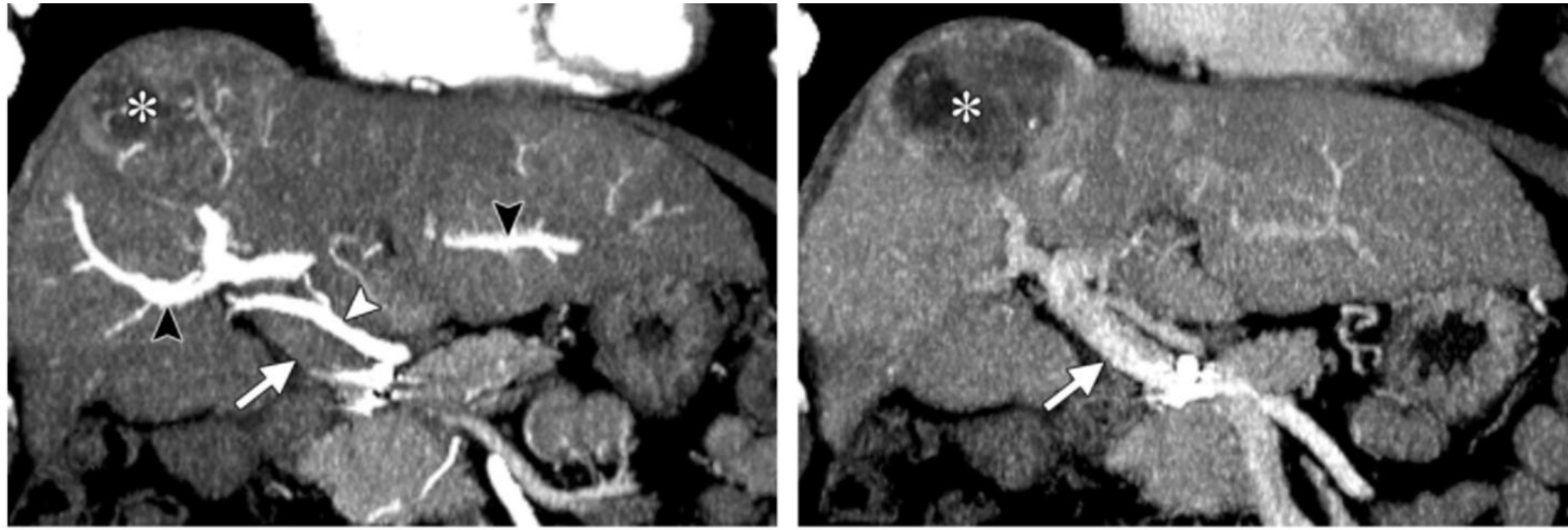
(功能性分流：动脉期肝实质区域灌注增强，但无门静脉早显)



T2	T1	动脉早期（肝门层面）
动脉早期	动脉晚期	门脉期



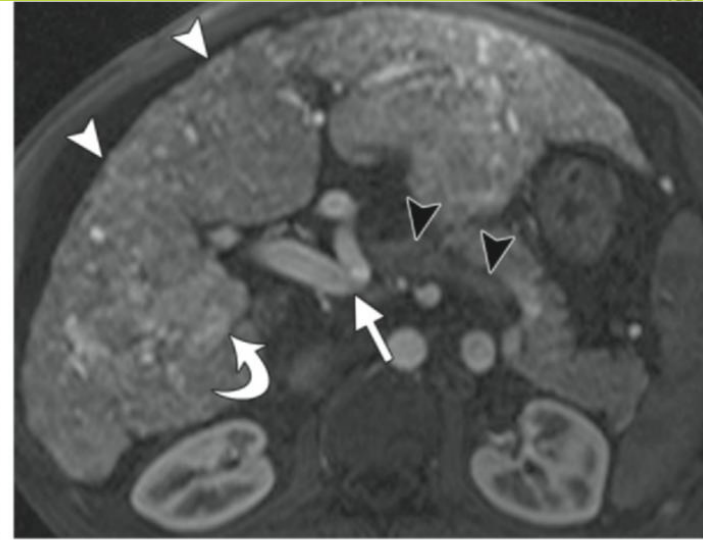
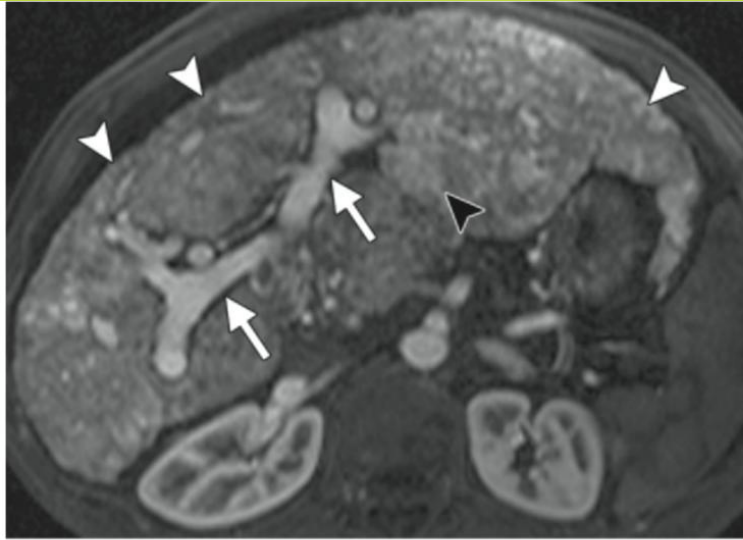
# 动门脉分流



**a.** **b.**

**Figure 10.** Malignant arteriportal shunt in a 70-year-old man with hepatocellular carcinoma in the right lobe of the liver and viral hepatitis–induced cirrhosis. Coronal contrast-enhanced CT images obtained during arterial (**a**) and portal venous (**b**) phases show a 7-cm heterogeneous tumor (\*) in hepatic segments VII and VIII. The right and left portal veins (black arrowheads in **a**) are isoattenuating relative to the right hepatic artery (white arrowhead in **a**) in the arterial phase, findings indicative of a tumor-related arteriportal shunt. Note the absence of enhancement of the main portal vein (arrow) in **a**, the arterial phase image, and its normal enhancement in **b**, the venous phase image.

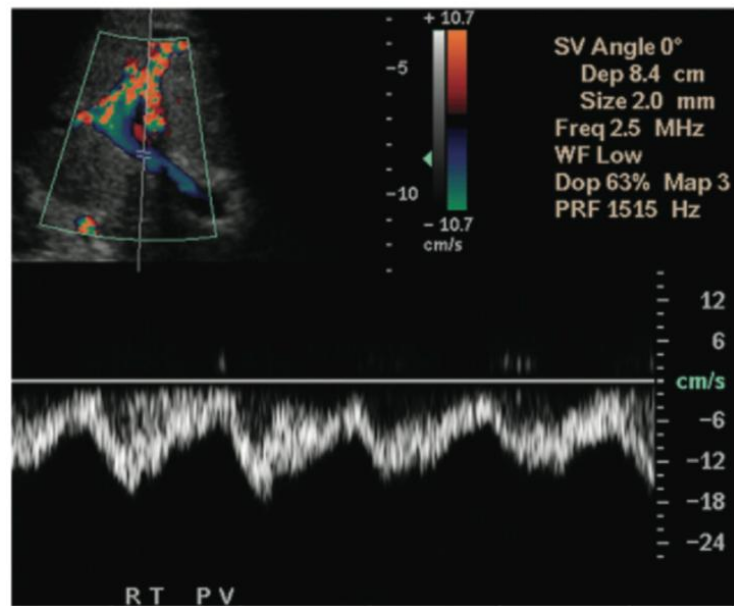
# 动门脉分流



a.

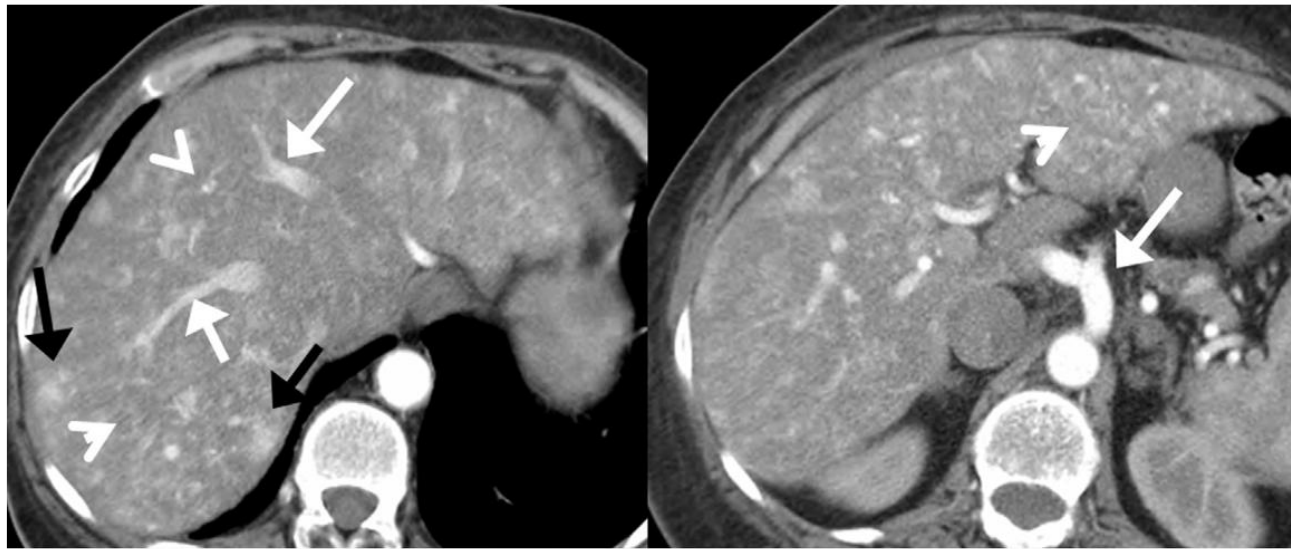
b.

**Figure 11.** Arterioportal shunts in a 51-year-old woman with hereditary hemorrhagic telangiectasia and decompensated cirrhosis. **(a, b)** Axial gadolinium-enhanced fat-suppressed T1-weighted MR images obtained in the early arterial phase at the levels of the portal vein bifurcation **(a)** and hepatic artery bifurcation **(b)** show innumerable tiny telangiectases (white arrowheads) throughout the liver with large confluent vascular masses (curved arrow in **b**), characteristic findings of hereditary hemorrhagic telangiectasia of the liver. The right and left hepatic arteries are enlarged because of intrahepatic vascular shunts (straight arrow in **b**). Early enhancement of the right and left portal veins (straight arrows in **a**) and absent enhancement of the splenic vein (black arrowheads) are indicative of arterioportal shunts. **(c)** Duplex Doppler image and waveform show reversed pulsatile flow in the right portal vein because of arterioportal shunts.



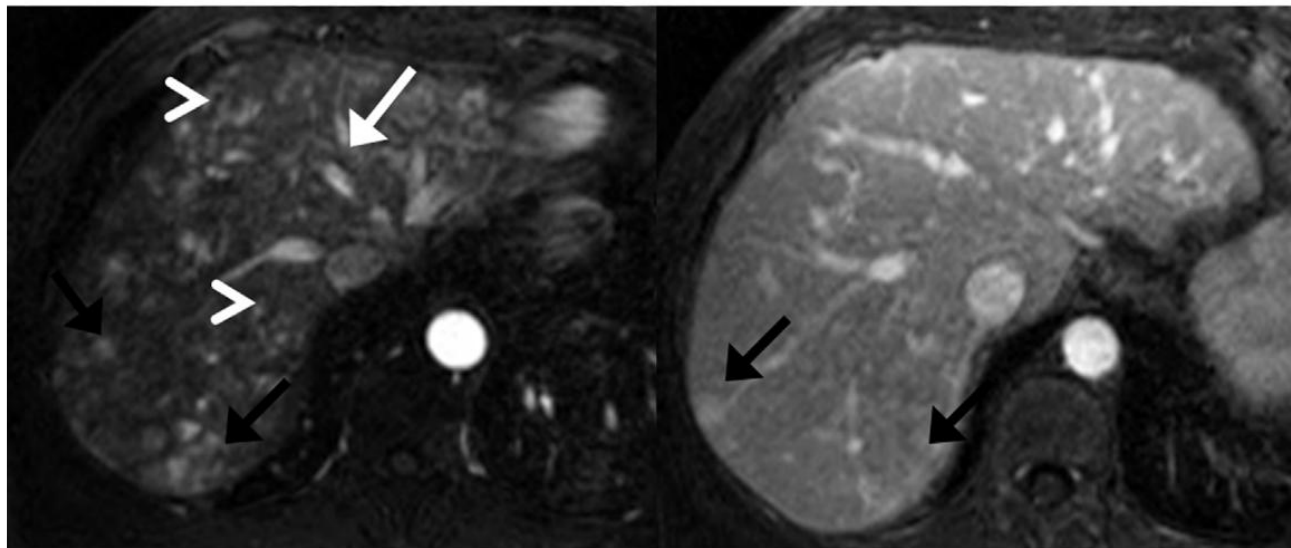


# ( 附：动脉 - 肝静脉分流 )



(a)

(b)



# 门体静脉分流

门体静脉分流（portosystemic shunt, PSS）

是门静脉和肝静脉或下腔静脉之间的交通，可分为肝内、肝外；先天性，获得性

获得性肝外PSS是最常见的门体静脉分流，主要继发于门脉高压

- 常见于胃食管周围、食管旁、脐旁、脾肾间、肠系膜下静脉侧支血管
- 表现为扩张、迂曲的管状结构

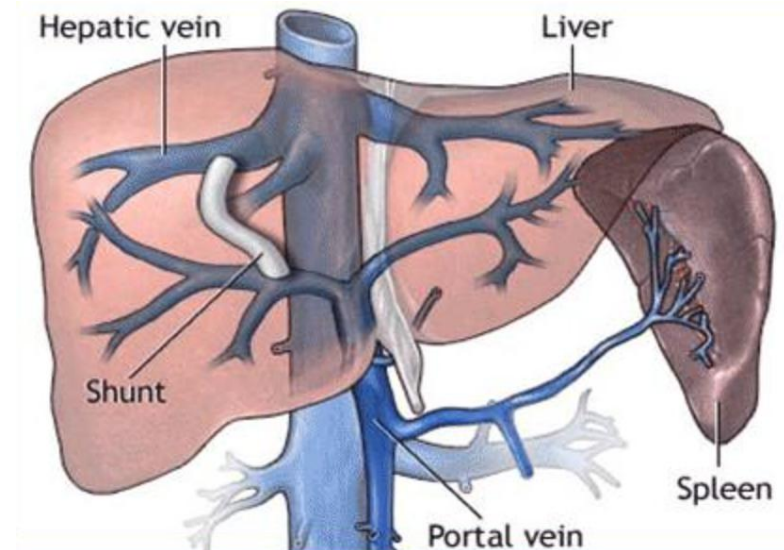
获得性肝内PSS可继发于肝硬化或外伤、医源性损伤等

- 肝硬化继发门脉高压后，常并发肝外的PSS，少数可并发肝内PSS
- 主要包括门脉左支与附脐静脉间分流、门脉右后支与下腔静脉间分流等
- 异常连通；门脉期不对称性肝静脉强化

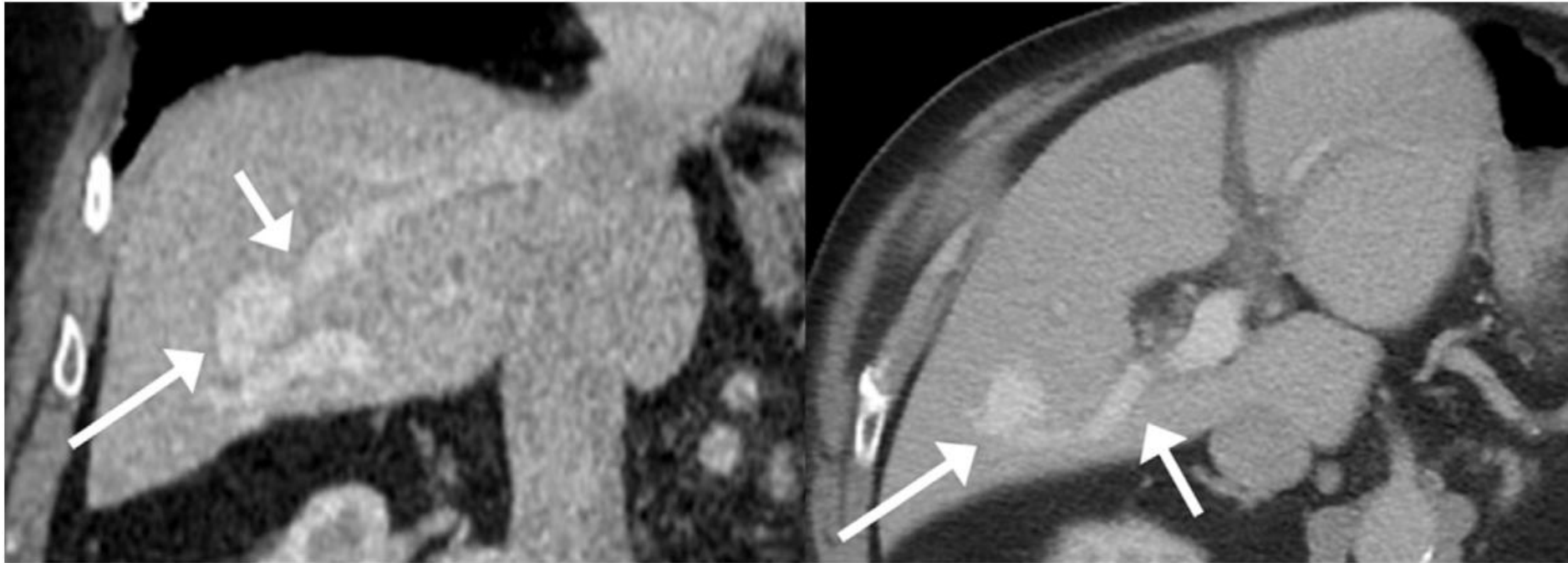
# 附：经颈静脉肝内门体静脉内支架分流术

经颈静脉肝内门体静脉内支架分流术（transjugular intrahepatic portosystemic stent-shunt, TIPSS）。

经颈静脉入路，建立肝内的位于肝静脉及门静脉主要分支之间的人工分流通道，并以内支架维持其永久性通畅，达到降低门脉高压后控制和预防食道胃底静脉曲张破裂出血，促进腹水吸收。

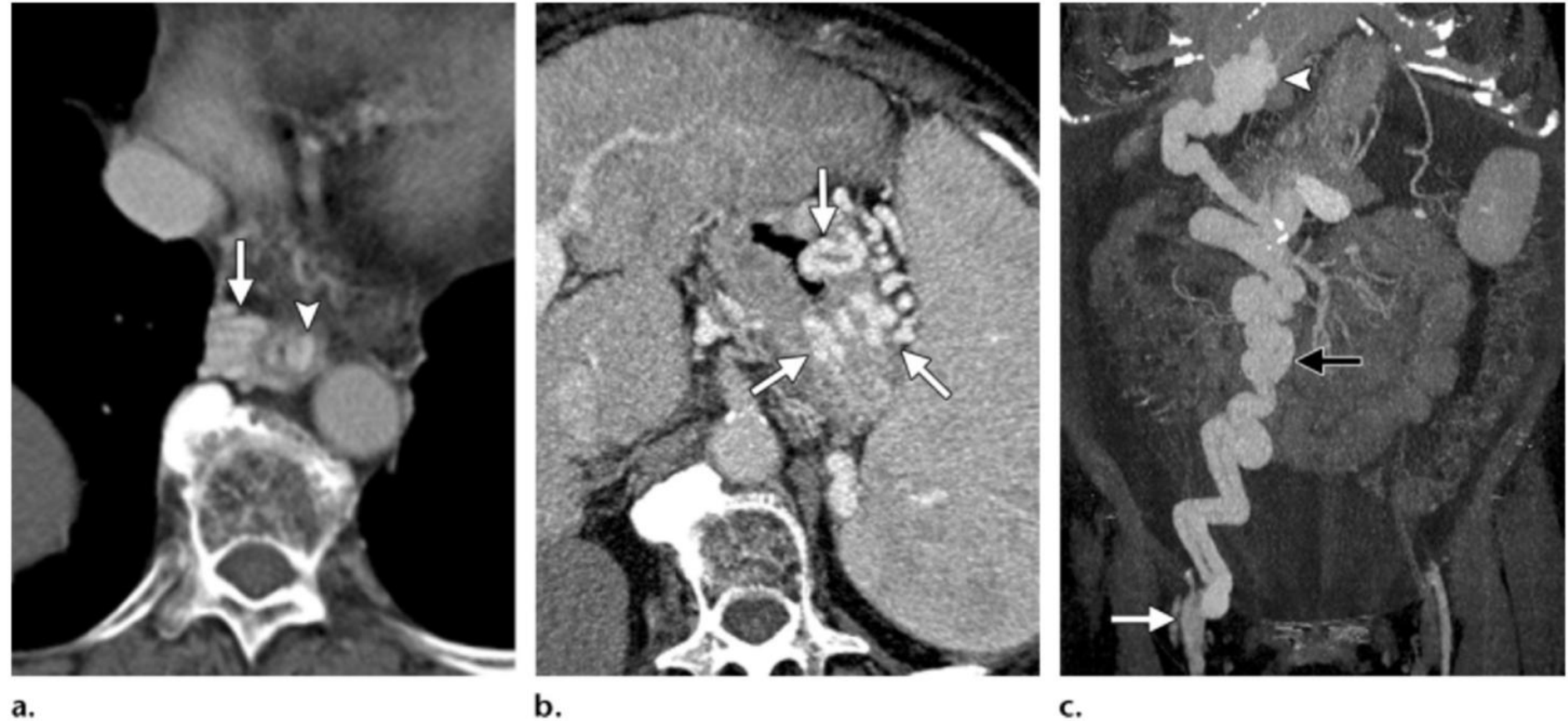


# 获得性肝内门体分流



M, 40y, 10年前肝脏扎伤

# 获得性肝外门体分流

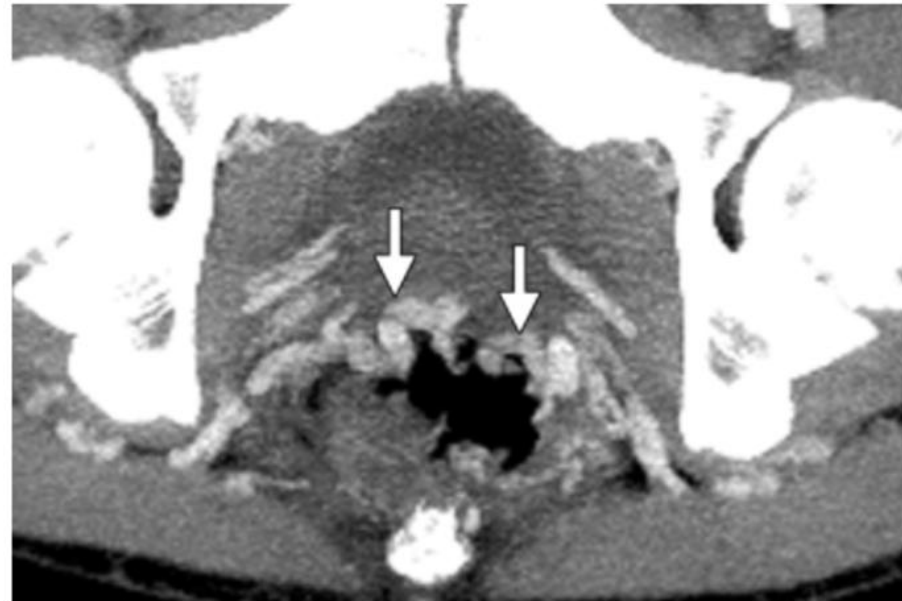


**Figure 6.** Extrahepatic portosystemic shunts in a 55-year-old man with alcohol-induced cirrhosis and portal hypertension. **(a, b)** Axial contrast-enhanced CT images show paraesophageal (arrow in **a**), esophageal (arrowhead in **a**), and gastric (arrows in **b**) varices. **(c)** Coronal contrast-enhanced maximum intensity projection CT image shows paraumbilical varices (arrowhead) draining into an epigastric vein (black arrow) and the right common femoral vein (white arrow).

# 获得性肝外门体分流



**Figure 7.** Splenorenal varices in a 68-year-old man with cirrhosis and portal hypertension. Coronal contrast-enhanced CT image shows multiple tortuous varices (arrows) medial to the spleen (S) that drain into an enlarged left renal vein (\*).



**Figure 8.** Asymptomatic rectal varices in a 60-year-old man with alcohol-induced cirrhosis and portal hypertension. Axial contrast-enhanced maximum intensity projection CT image shows numerous varices in the lower rectum (arrows) that shunt flow between the inferior mesenteric vein and the middle and inferior rectal veins.

# 门静脉高压 ( Portal hypertension )

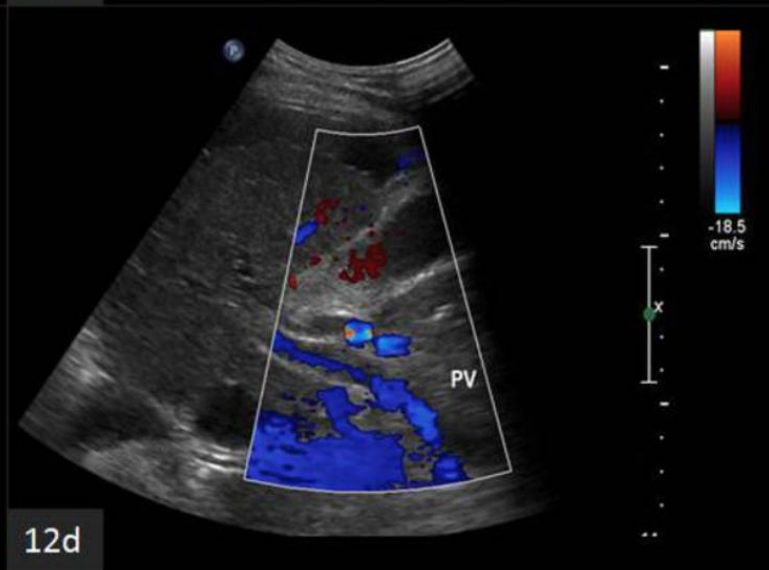
由门静脉压力持久增高引起的症候群

- 定义: 门静脉压力大于12 mmHg或肝静脉压力梯度大于5mmHg
- 按照病理生理学可分为原发性**血流量增加型**及原发性**血流阻力增加型**
- 大多数由**肝硬化**引起（90%以上），少数继发于门静脉主干或肝静脉梗阻以及原因不明的其他因素

影像学表现:

- 门脉管径大于13mm
- 胃左静脉大于4mm
- 附脐静脉开放，大于3mm
- 门体分流开放
- 门静脉管径小于10mm提示门脉离肝性血流（特异性高）
- 脾大
- 腹水

# 门静脉高压 ( Portal hypertension )





# 门静脉血栓

- 血栓形成可分为急性、亚急性和慢性，以**慢性**最为多见
- 急性期血栓为**凝血块**，随后形成较新鲜的纤维性血栓，继而血栓机化，并逐步发生纤维化，最终表现为**纤维条索**，伴周围侧枝静脉影，即仅仅存留**继发性门静脉海绵样变**
- 最常见原因是**肝硬化（门静脉高压引起门脉血流速度缓慢）**
- 还包括感染及炎性疾病（如胰腺炎、胆管炎、继发于肠道感染的化脓性门静脉炎等）、肿瘤（肝癌、胰腺癌等）、血液高凝状态、手术后（肝移植后、食管胃底静脉曲张硬化治疗后）等
- 肠系膜上静脉、脾静脉血栓也可延续到门静脉

- 门静脉血栓常为偶然发现
- 急性门静脉血栓常表现为腹痛，并可能引起肠梗死（血栓延续至肠系膜上静脉）及肝脏缺血性梗死
- 没有肝脏基础疾病的患者，发生门静脉血栓后可继发肝内、外血管分流、侧枝静脉开放以及门静脉高压
- 具有肝硬化等基础疾病者，发生门静脉血栓后上述血管异常表现会加重

# 门静脉血栓

## CT

- 急性门静脉血栓为高密度或软组织密度，门静脉管腔增粗，直径常大于20mm，增强后表现为腔内充盈缺损
- 门静脉远端的肝实质内，门脉期可见缺血所致的强化减低区，动脉期缺血区可显示为一过性相对高强度（肝动脉代偿性供血增加）
- 慢性门静脉血栓可发生钙化，表现为壳状或点、片状钙化密度，门脉期可显示血栓的再通（闭塞管腔或管壁内纤细条状强化）和继发门静脉海绵样变
- 一般情况下血栓不强化

## MRI

- 管腔的形态和肝脏实质的灌注异常与CT相仿
- 常规序列上表现为流空现象消失，血栓信号会高于无血栓区域的血流信号，呈软组织信号
- 在亮血序列上，流动血液为高信号，血栓为相对低信号
- 动态增强MRI进行诊断更为准确

# 门静脉血栓



鉴别诊断:

•门静脉癌栓:

- 门静脉明显增粗 ( $>23\text{mm}$ )
- 动脉期条纹状强化
- 邻近或远处肝实质内肿瘤病变

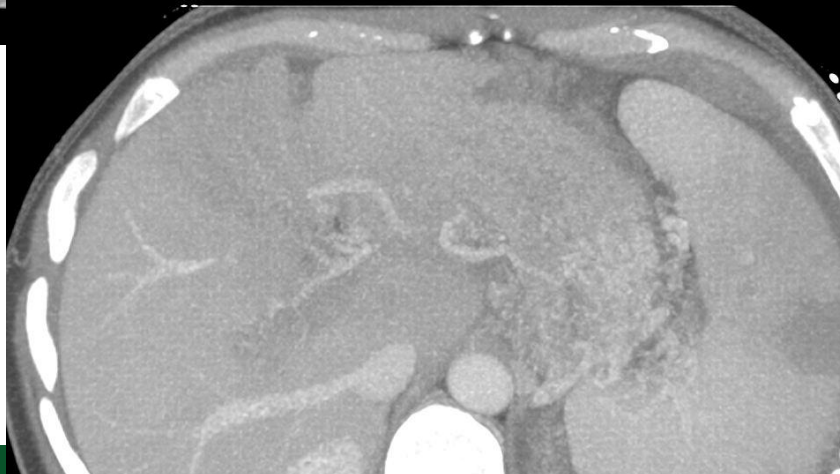
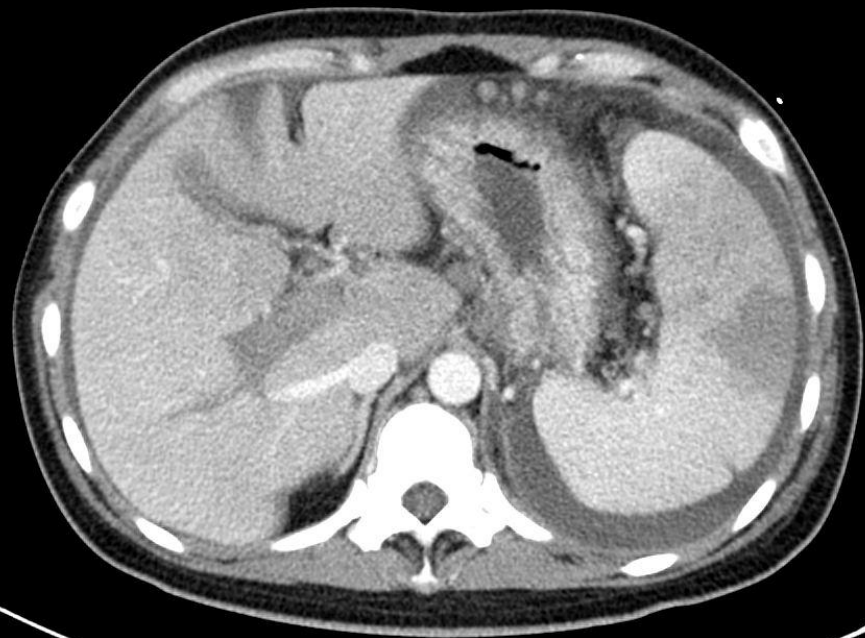
•良恶性肿瘤压迫门静脉

门静脉会受压狭窄，但不表现为充盈缺损

•动脉期门静脉强化不均

观察门脉期图像可以鉴别

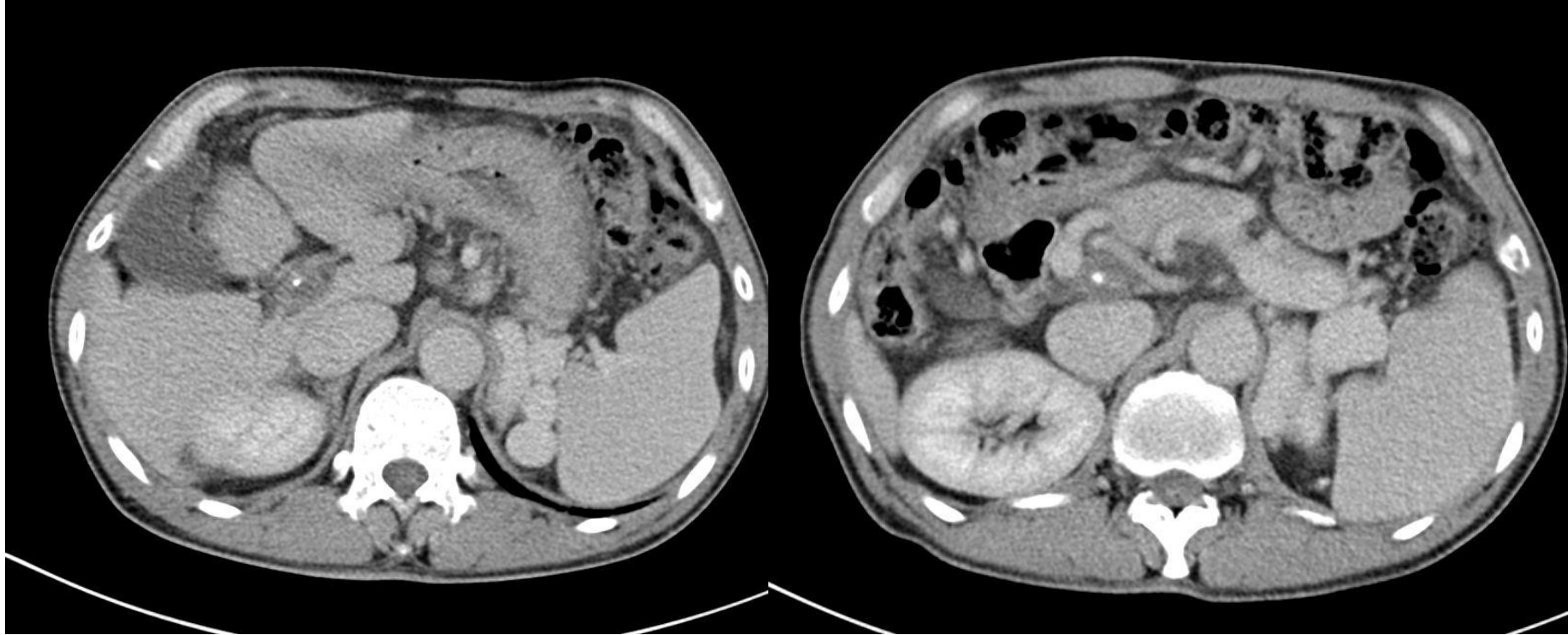
# 门静脉血栓



F, 39y, 腹痛2周

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# 门静脉血栓

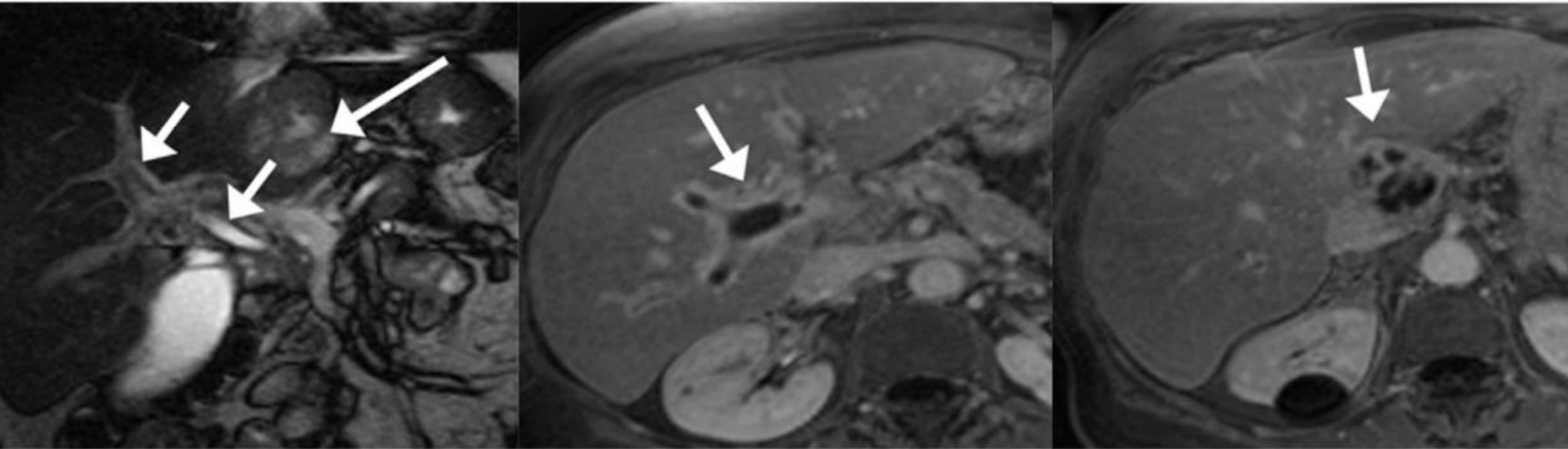


M, 52y, 诊断肝硬化10余年

门脉内低密度充盈缺损伴点状钙化, 诊断为慢性门静脉血栓

另可见肝脏比例失调, 胆囊窝增宽, 脾大, 脾周迂曲增宽侧支静脉影

# 门静脉血栓



M, 60y, 急性门静脉血栓, 肝脓肿

# 门脉海绵样变

门静脉海绵样变性（cavernous transformation of the portal vein, CTPV）

肝门及邻近区域（包括肝十二指肠韧带）海绵状、蜂窝状的细小血管结构，用于替代或补偿门静脉向肝脏供血

- 病因大多为获得性，即门静脉血栓、癌栓或其他原因导致的门静脉主干和（或）分支完全或部分闭塞
- 少数可为先天性（常伴有先天性肝纤维化等其它疾病）
- 部分无明确的原因，称为特发性
  
- 海绵样变性常合并门静脉高压
- 原有门静脉高压的患者，继发门静脉血栓和海绵样变性后，门静脉高压的症状会加重

# 门脉海绵样变

## 门脉海绵样变性的特征性表现

- 肝门部正常门静脉血管结构狭窄、中断或闭锁
  - 门静脉腔内、壁内及周围出现多发细小迂曲的强化血管影
  - 血管网还可出现于门脉左、右支旁及肝胃韧带、肝十二指肠韧带区域
  - 血管网可向肝内延伸并累及肝内门脉分支，但门脉主干形态正常
- 
- 肝实质动脉期强化不均匀，周边可见弥漫的斑片状高灌注区（中央区较周边区血运丰富，周边区的动脉血运代偿性增加）
  - 非肝硬化患者发生门脉血栓继发海绵样变性时，肝脏形态变化，出现中央区（肝左内叶和 / 或尾状叶）肥大，周边区（肝左外叶和 / 或右叶）萎缩
  - 由于门脉海绵样变易继发或加重门脉高压，患者还可出现肝外型门静脉高压的一系列改变（脾大、腹腔积液、侧枝形成等）



# 门脉海绵样变



肝右叶占位伴门脉主干及右支内癌栓形成，肝门区多发迂曲交织血管影

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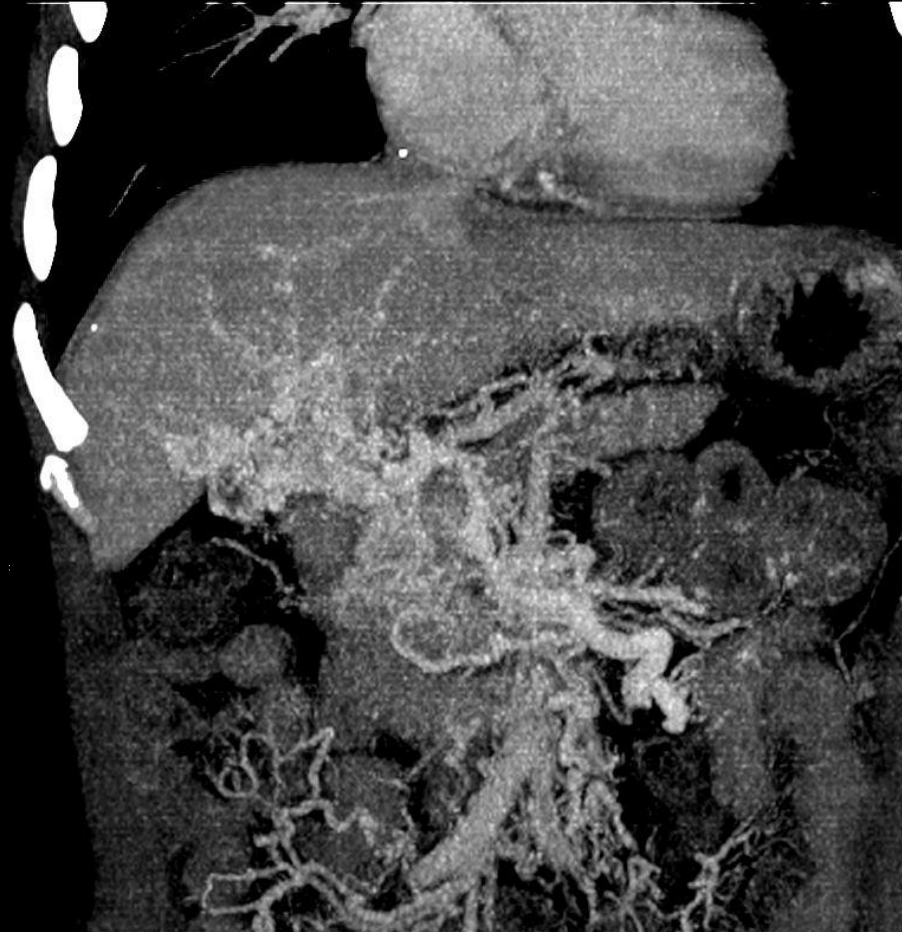
# 门脉海绵样变



门脉主干及肝内分支、肝胃韧带、脾周多发迂曲侧支静脉影；患者的基础病为易栓症

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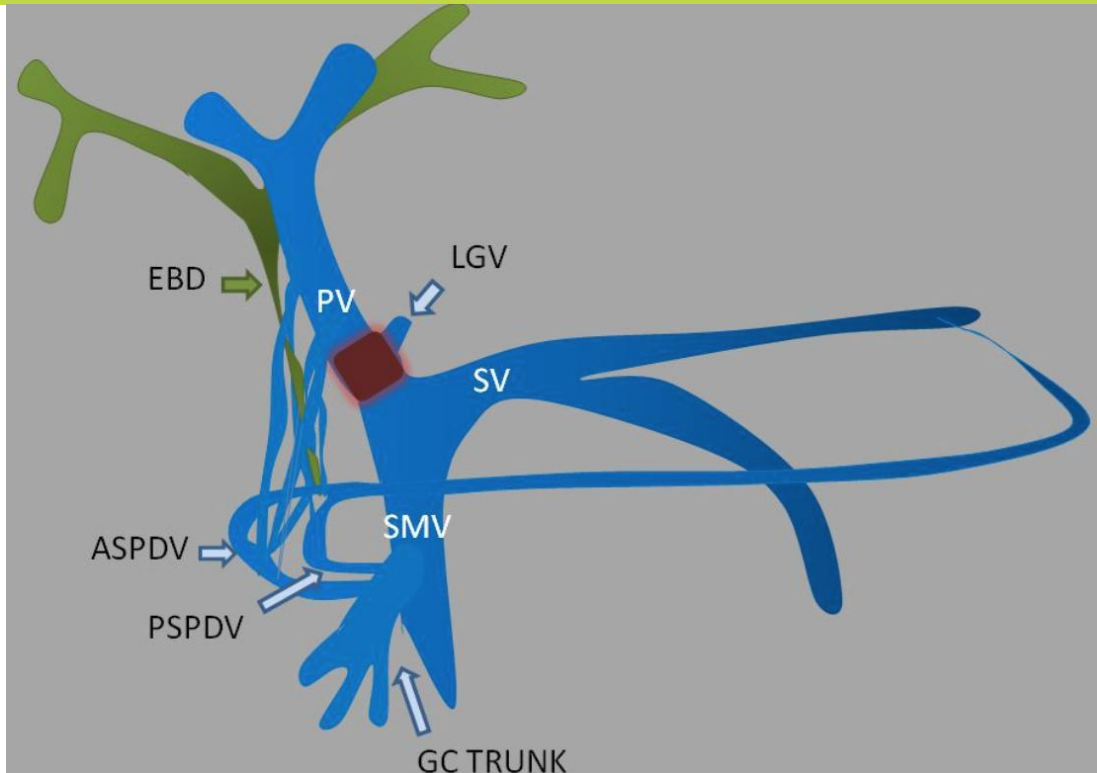
# 门脉海绵样变



F, 43y, 间断呕血黑便1y

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## 附：门脉性胆道病（ Portal biliopathy ）



海绵样变性可引起胆总管旁、胆总管上静脉丛扩张，造成肝外和（或）肝内胆管受压，以及胆管与胆管周围纤维化，继发胆管狭窄

# 门静脉瘤

门静脉系统的局限性囊状或梭形扩张，即门静脉瘤（aneurysm of the portal venous system, varix）

- 较多发生于肝外，如脾静脉、肠系膜上静脉汇合部、门静脉主
- 也可发生于肝内，常见于门静脉肝内分叉处

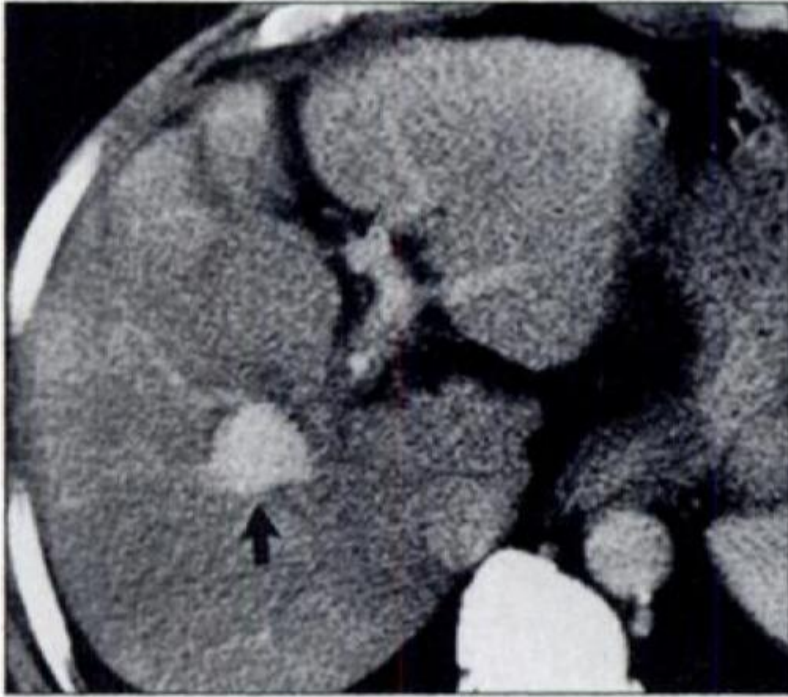
临床表现

- 小的门静脉瘤没有症状
- 较大者可有压迫症状（非特异腹痛等）
- 并发其它病变（如门静脉高压、门静脉血栓形成），会有相应症状

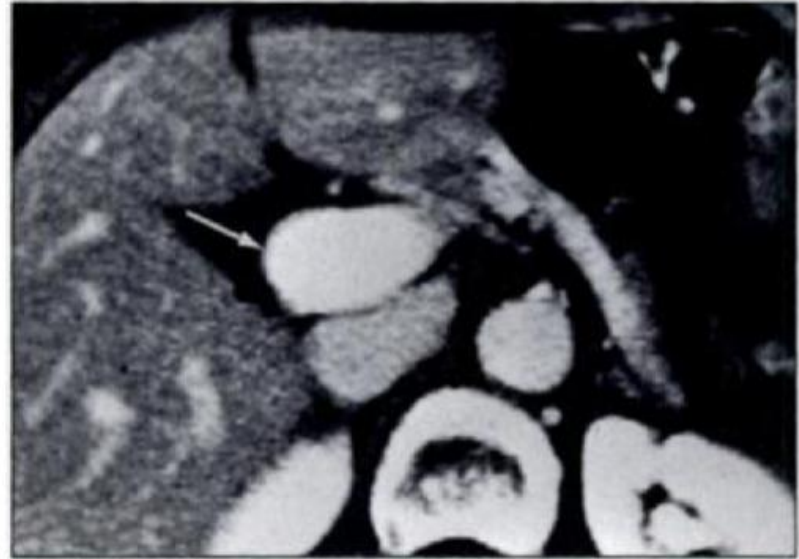
门静脉某点血管直径较邻近血管直径显著增宽，尤其呈囊状或梭形扩张，即可以诊断为静脉瘤

出现并发症时CT和MR可显示相关的改变，如血栓形成、门静脉高压（由于血流波动或门静脉受压），破裂，胆总管受压等

# 门静脉瘤

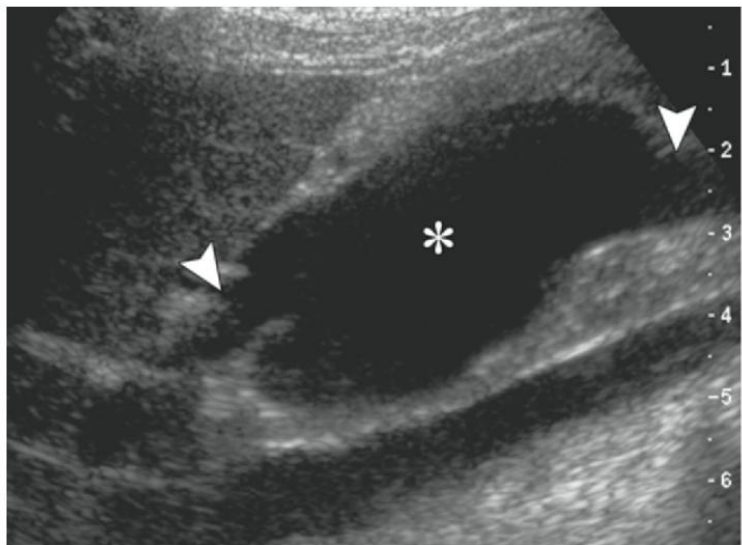


**Fig. 4.**—Intrahepatic portal vein aneurysm in 58-year-old man who had no clinical symptoms. Contrast-enhanced CT scan reveals well-enhanced, dilated vascular structure indicating aneurysm of right portal vein (*arrow*).

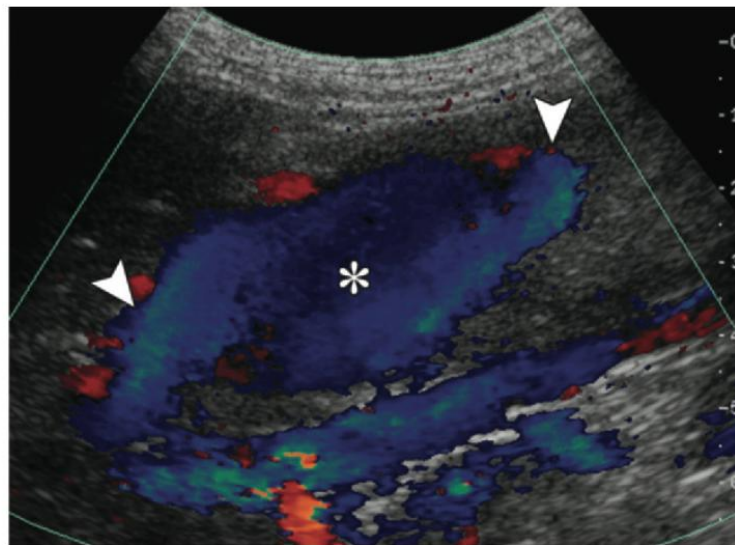


**Fig. 5.**—Extrahepatic portal vein aneurysm in 79-year-old woman who had no clinical symptoms. Contrast-enhanced CT scan reveals well-enhanced, dilated vascular structure indicating aneurysm of main portal vein (*arrow*).

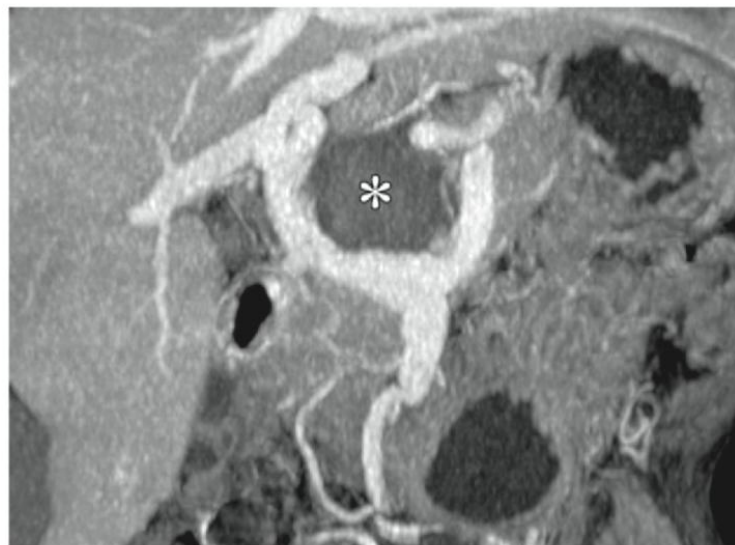
# 门静脉瘤



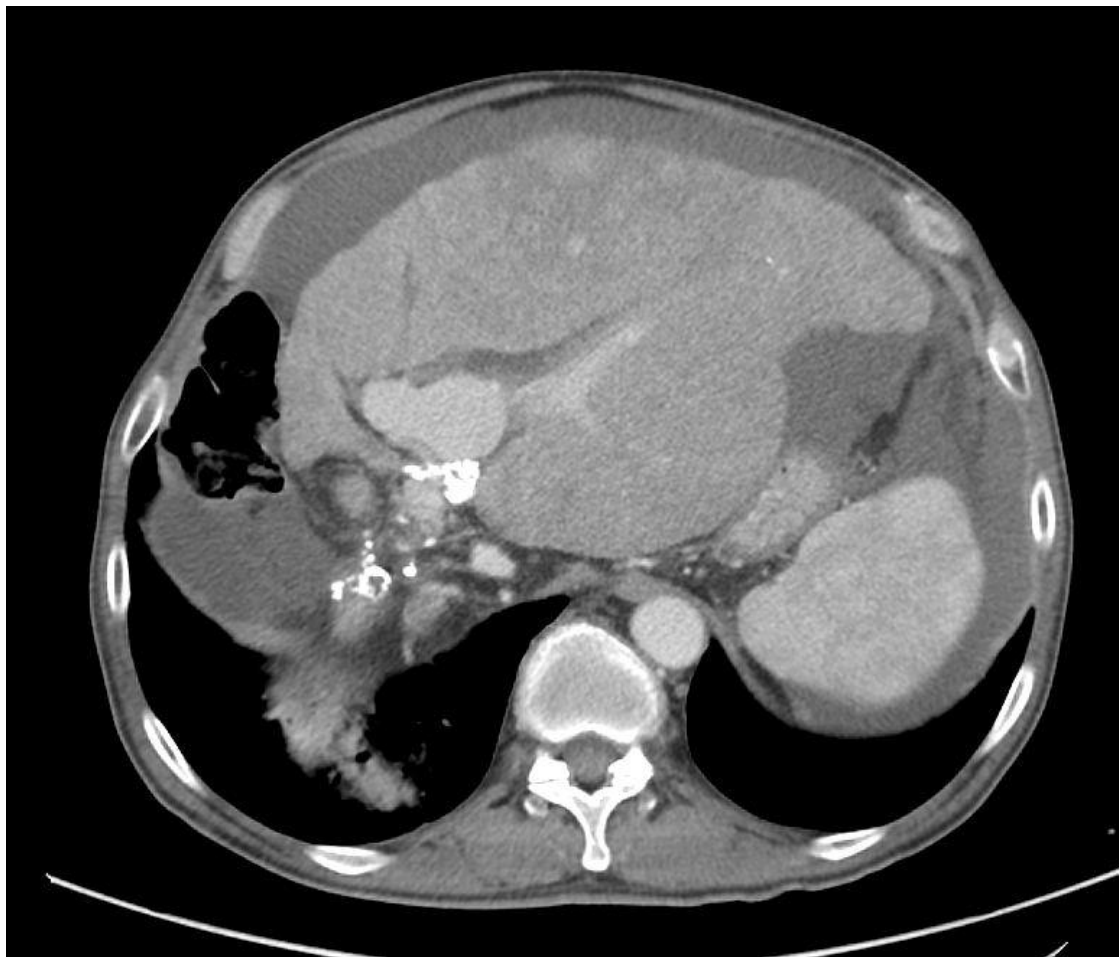
a.



b.



# 门静脉瘤



肝内外胆管多发结石伴肝右叶萎缩

门脉期轴位CT示门脉左支起始部窄，以远瘤样扩张

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# 门脉系统积气

## 门脉肠系膜静脉积气（Portomesenteric Vein Gas）

- 最常见于肠系膜缺血（常合并肠梗阻）
- 也可见于消化道穿孔（溃疡或肿瘤所致）、炎性肠病、腹部感染（如腹腔脓肿、憩室炎）、创伤等
- 医源性因素（如使用灌肠剂、结肠镜后、胃造口术后留置引流管及肝移植术后）也可引起
- 肠壁通透性增加、肠壁结构破坏、肠腔上皮脱落、肠腔扩张或压力变化等多种因素，均可使肠腔内气体穿过血管壁，进入肠系膜静脉系统，从而回流进入肝脏门脉系统

如果门脉系统积气合并肠系膜缺血，必须手术处理  
单纯门脉系统积气大多预后良好

# 门脉系统积气



门脉系统积气CT表现为管状、分枝状充盈缺损

肝内门静脉积气：

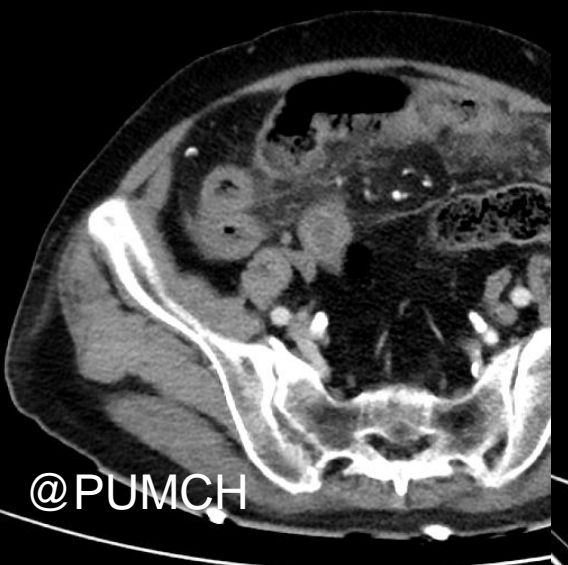
应与胆道积气鉴别

- 门静脉积气为周边分布，形态细小，分支较多
- 胆道积气在肝内呈中央性分布，距离肝脏包膜2cm以上，二者均多见于肝左叶

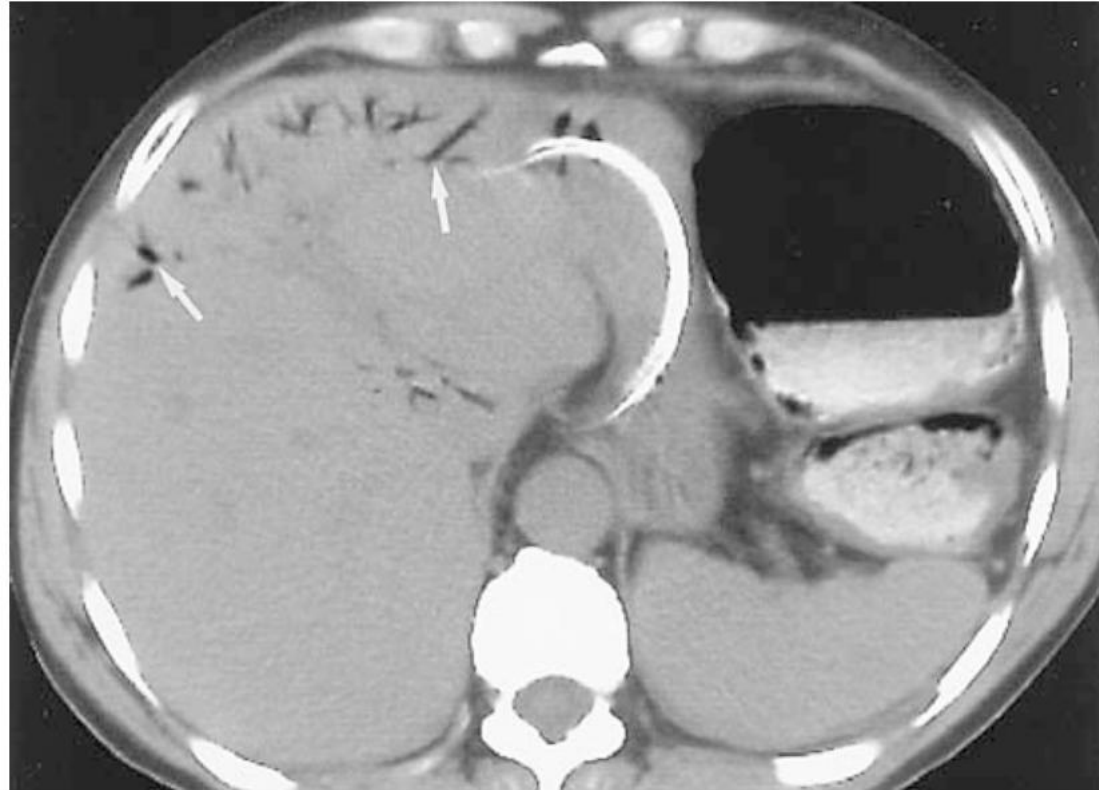
肠系膜静脉分支积气：

应与气腹鉴别

- 气腹不呈管状或分支状，且可在小肠肠管的系膜缘、腹膜腔内出现



@PUMCH



患者腹痛，腹胀，10小时前接受结肠镜检查

# 附：门静脉炎



M, 45y, 发热黄疸4天

A computed tomographic scan showed acute perforated sigmoid diverticulitis (Panels A and B, arrow) and gas in the mesentery (Panels A and B, white arrowheads). In addition, there was gas in the inferior mesenteric vein extending to the splenic vein up to the origin of the portal vein (Panel A, red arrowheads), a finding that was consistent with pylephlebitis.

Lindsey et al, NEJM 2015

# 闭塞性门静脉病

闭塞性门静脉病 (literative portal venopathy, OPV)是临床上罕见的肝脏血管疾病

- 目前临床最新命名为**特发性非肝硬化门静脉高压症**(idiopathic non-cirrhotic portal hypertension, INCPH)
- 既往命名有**Banti综合征**、**特发性门静脉高压** (idiopathic portal hypertension)、**良性肝内门静脉高压症**、**肝门静脉硬化症** (hepatoportal sclerosis)、**非肝硬化性门静脉纤维化**(non-cirrhotic portal fibrosis) 等

病因和发病机制不明，可能与接触毒物、感染、免疫、遗传等因素有关  
组织学上显示肝脏闭塞性门静脉病，**肝内门静脉大、中型分支呈现明显的内皮下增厚，胆管周围纤维化**

临床上常见隐匿起病，多以上腹肿块为主诉就诊，也出现消化道出血、贫血、水肿等，体检可见脾大，贫血，肝不大，少数可见腹壁静脉怒张，黄疸及腹水少见，**肝性脑病罕见，肝功能多正常或轻度异常**

# 闭塞性门静脉病



**Table 2**

**Main CT Findings Observed in Patients with OPV and Patients with Cirrhosis**

Parameter	OPV Group (n = 42)	Cirrhotic Group (n = 42)	PValue*
Intra- and/or extrahepatic portal vein abnormalities	30 (71)	7 (17)	<.001
Extrahepatic portal vein abnormalities	18 (43)	5 (12)	.003
Intrahepatic portal branches abnormalities	18/31 (58)	1 (2)	<.001
Mural calcifications of the portal veins	7 (17)	2 (5)	.16
Morphologic changes of the liver	35 (83)	41 (98)	.057
Hypertrophy of the caudate lobe and atrophy of segment IV	10 (24)	27 (64)	<.001
Nodular liver surface	7 (17)	37 (88)	<.001
FNH-like nodules	6 (14)	0	.03

Note.—Data are numbers of patients, with percentages in parentheses.

\* P value represents the difference between observed findings in the two groups. The difference is statistically significant when P is less than .05.

5 特发性非肝硬化门静脉高压  
指南推荐了 INCPH 的诊断

表 3 INCP

门静脉高压症临床表现

- 脾大/脾亢
- 食管静脉曲张
- 非肿瘤性腹水
- 肝静脉压力梯度轻度升高
- 门体侧支循环

肝活组织检查排除肝硬化可能

排除导致肝硬化或非肝硬化门静脉高压症的慢性肝病可能

- 慢性乙型/丙型肝炎
- 非酒精性脂肪性肝炎/酒精性脂肪性肝炎
- 免疫性肝炎
- 遗传性血色素沉着症
- Wilson 病
- 原发性胆汁性肝硬化

排除导致非肝硬化门静脉高压症的可能

- 遗传性肝纤维化
- 结节病
- 血吸虫病

多普勒超声或 CT 证实门静脉及肝静脉开通

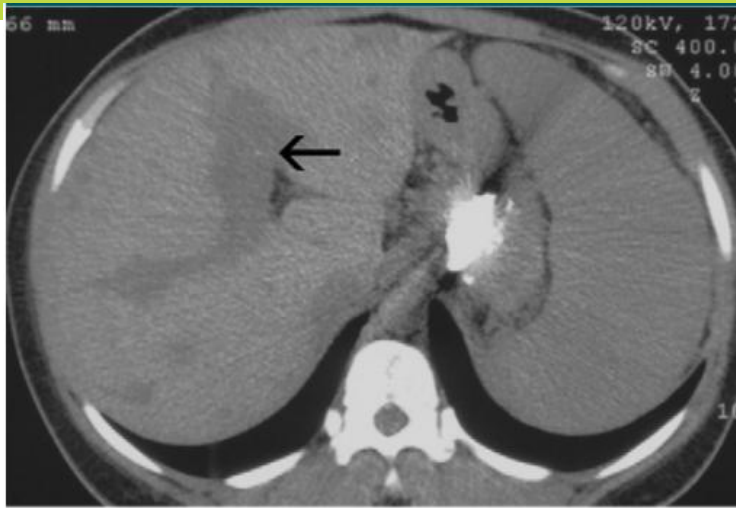
(1) 伴有门静脉高压、但无其他肝病原因的患者应考虑 INCPH 的可能 (B1)。

(2) 诊断 INCPH 时,需排除肝硬化及其他非肝硬化门静脉高压症的可能 (B1)。

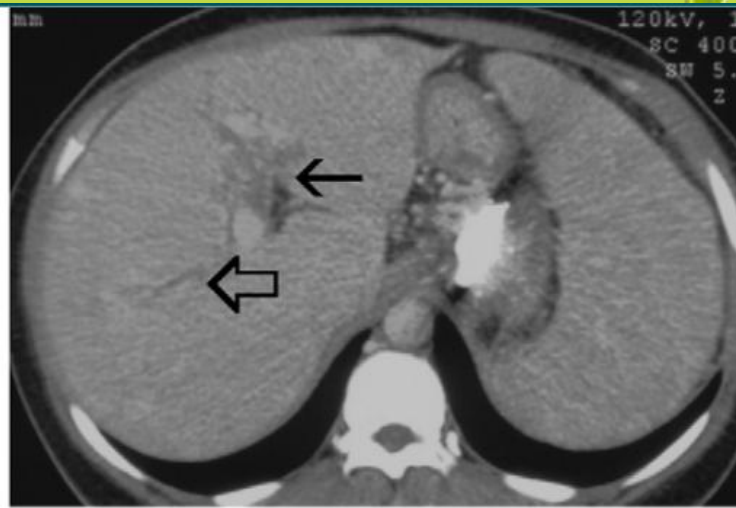
(3) 诊断 INCPH 时,需行肝活组织检查 (A1)。

- 影像上, 门静脉二级分支截然变细, 肝脏轮廓较少结节样改变, 尾状叶增大及左叶内侧段萎缩发生率不高, 可伴有FNH样的结节
- 需与肝硬化门静脉高压相鉴别
- 确诊需肝组织病理学检查发现没有弥漫性再生结节、并排除各种原因肝硬化、门静脉阻塞等

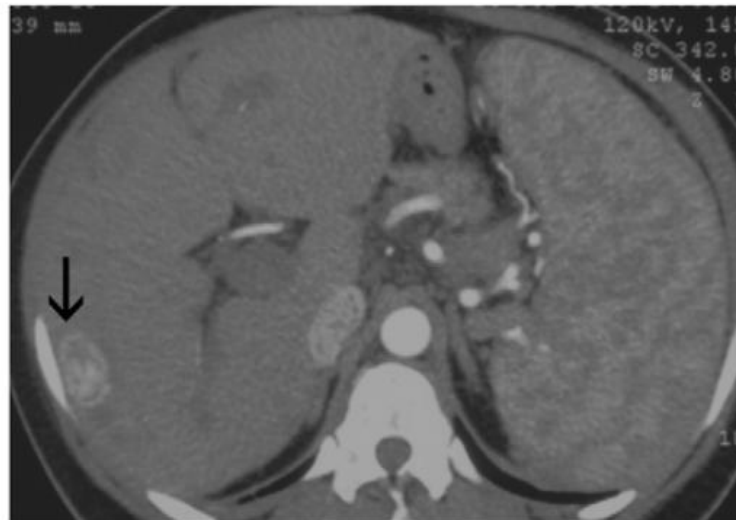
# 闭塞性门静脉病



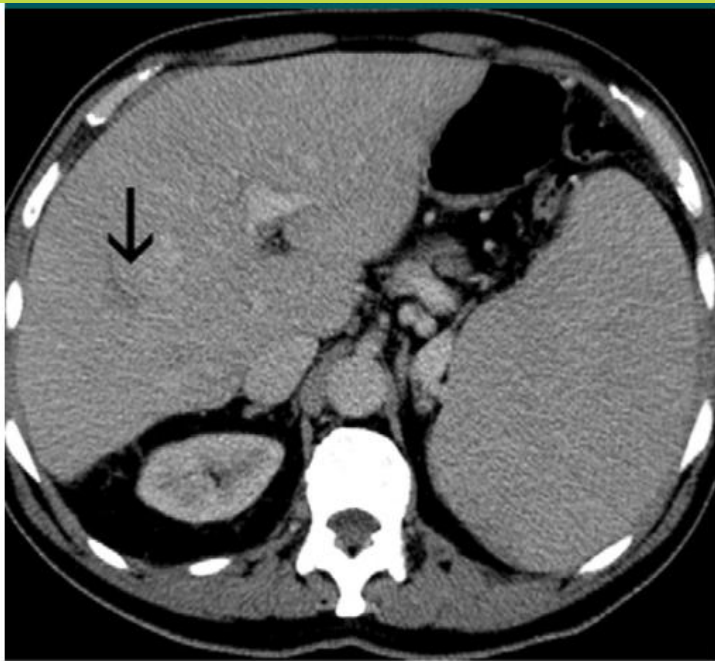
a.



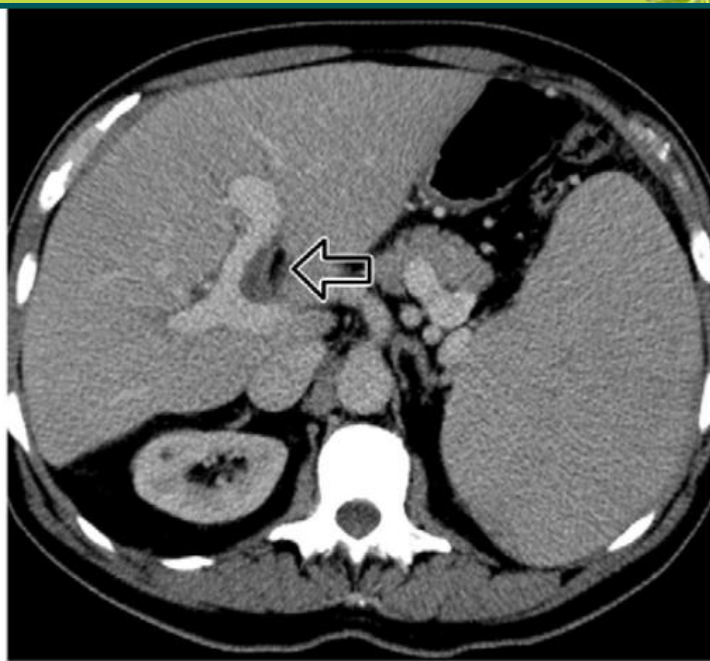
b.



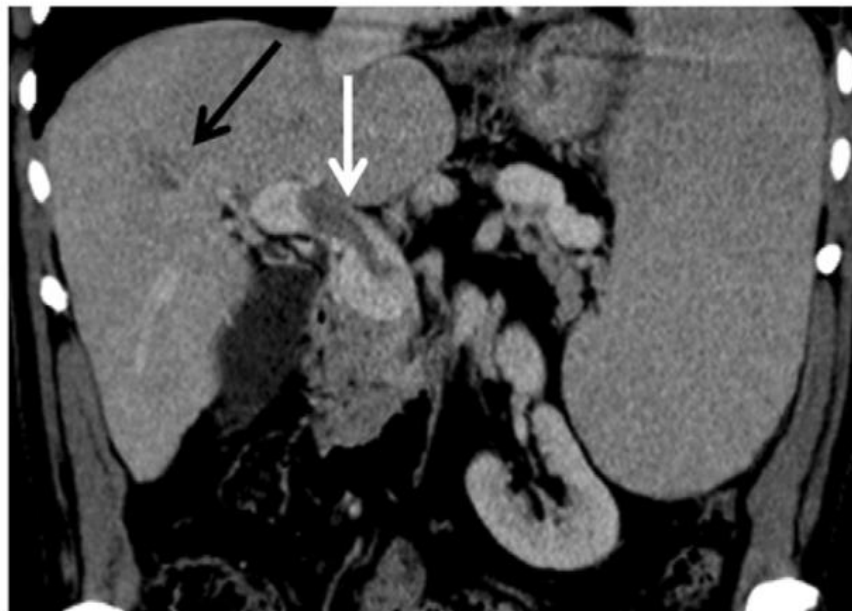




a.

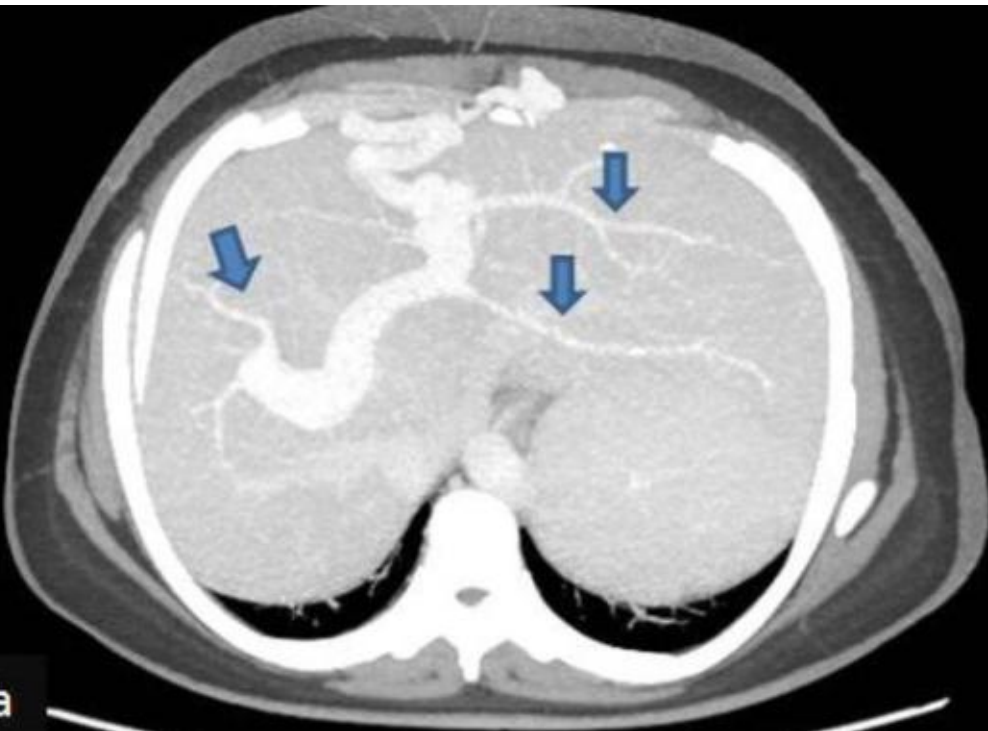


b.



c.





# 小结

- 胰腺前方门静脉，双门静脉，先天性门体分流
- 肝内动门脉分流；门静脉高压、获得性门体分流；门静脉血栓、门脉海绵样变、门静脉瘤、门静脉积气、闭塞性门静脉病

掌握门静脉的组织解剖与病理生理有助于更好地理解病变  
注意患者发病年龄、临床症状体征、病程演变、既往病史

门静脉高压胆管病  
肝硬化 → 门静脉高压  
→ 门静脉血栓 → 门脉海绵样变 → 门  
→ □ 获得性门体分流

肝细胞癌  
→ 门静脉癌栓 → 门脉海绵样变  
→ 肝内动门脉分流

# 小结



## 门脉相关疾病注意观察：

门脉的位置、形态、直径、密度、强化（方式、顺序）

门脉系统的分支、属支

肝动脉、肝静脉、下腔静脉（异常连通、早显、管径变化）

肝脏：轮廓、比例、实质灌注异常、占位  
胆管系统

脾脏、肠道、其它腹盆腔结构

心脏、肺动脉、上腔静脉等

**Thanks!**

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