DALHOUSIE UNIVERSITY

CIRA Case of the Week
January 2017

Case Courtesy of Dr. Gillian Shiau

Chief Complaint & HPI

- CC:
  - 73 year old male with suspected hepatic encephalopathy
- HPI:
  - Recurrent hospital visits over the course of a year with vague neurological symptoms (weakness, confusion) originally thought to be related to TIAs
  - One ER visit, patient also c/o abdominal “cramp” pain diffusely throughout abdomen with no relation to meals, position, or bowel movements without N/V or change in BMs – further investigation revealed disturbance of his liver enzymes and elevated ammonia levels (> 200)
  - Treated with lactulose with symptoms clearing but treatment found to be difficult for the patient

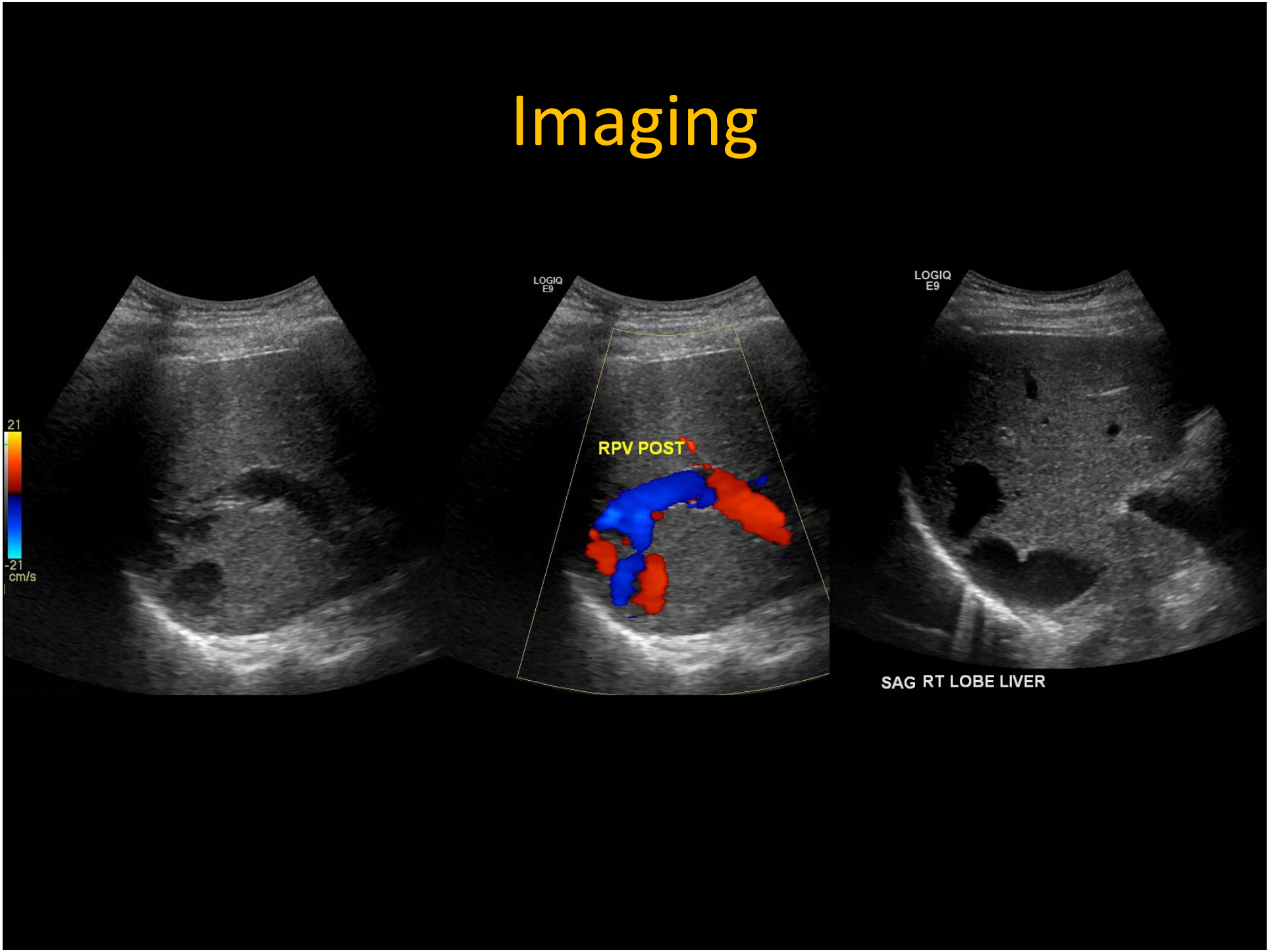
Medical History

- Past Medical History:
  - Significant cardiac history (hypertension; 3 MI's; atrial fibrillation with pacemaker/defibrillator insertion and subsequent lead infection requiring laser lead extraction; CHF with orthopnea)
  - NIDDM
  - Parkinson's Disease
  - No significant history of trauma
  - No history of liver interventions including biopsy
- Social History:
  - Social drinker
  - Non-smoker
- Family History:
  - Non-contributory
- Allergies:
  - Bactrim
  - Morphine
- Home Medications:
  - Furosemide 80 mg po OD
  - Warfarin 5 mg po OD
  - Nitroglycerin patch transdermal 0.6 mg daily
  - Pantoprazole 40 mg po OD
  - Glicazide 30 mg po OD (in AM)
  - ECASA 81 mg po OD
  - Potassium chloride 8 mEq po BID
  - Levocarb 100/25 one tab po TID
  - Tamsulosin 0.4 mg po OD
  - Lactulose 60 cc po TID – patient's symptoms rapidly recur if he misses a dose

Diagnostic Workup – Labs

AST: 27
ALT: 22
ALP: 120
Bilirubin (total): 50.0
Bilirubin (direct): 19.5
Ammonia: 176 (previously > 200)

Imaging



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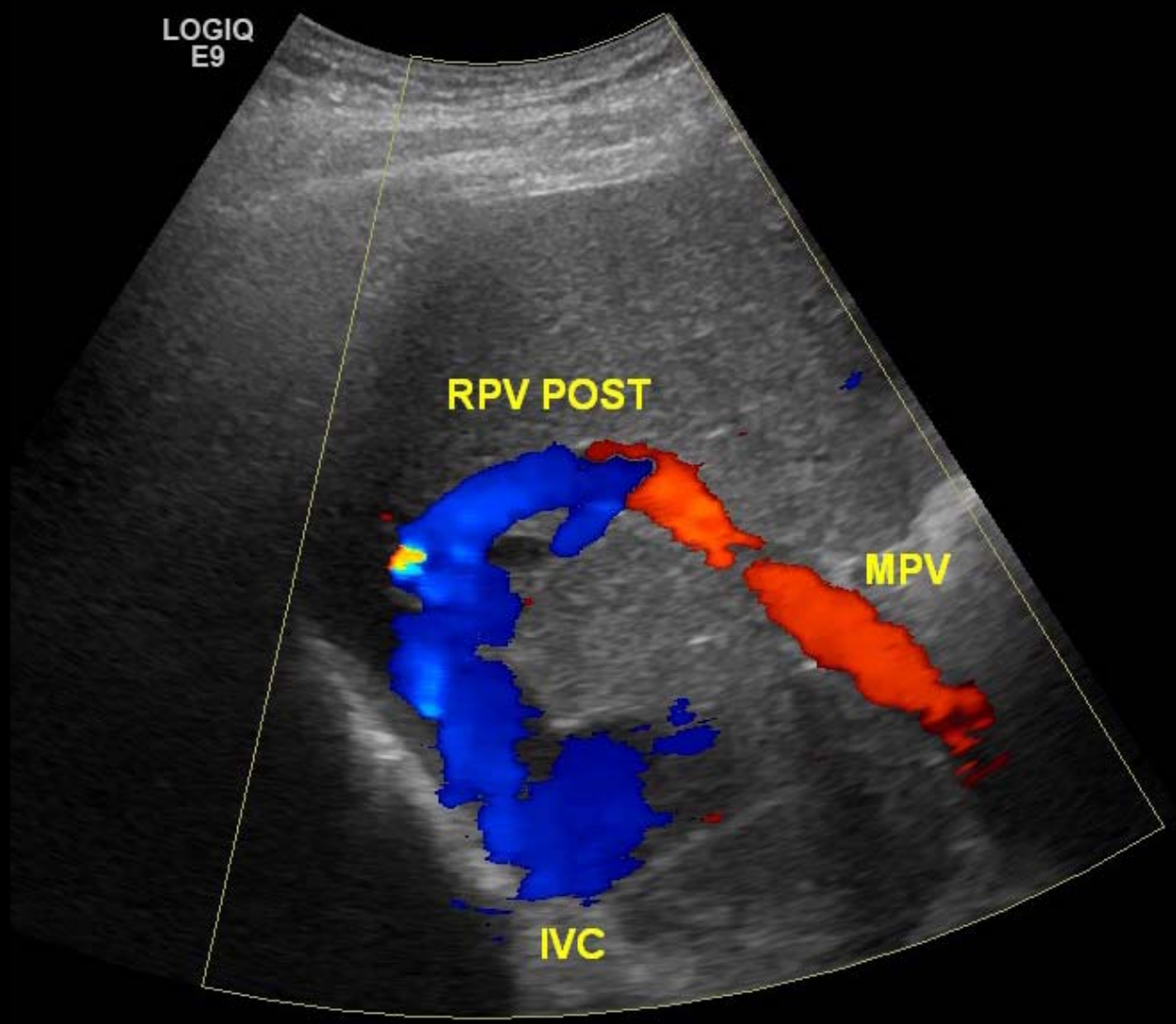
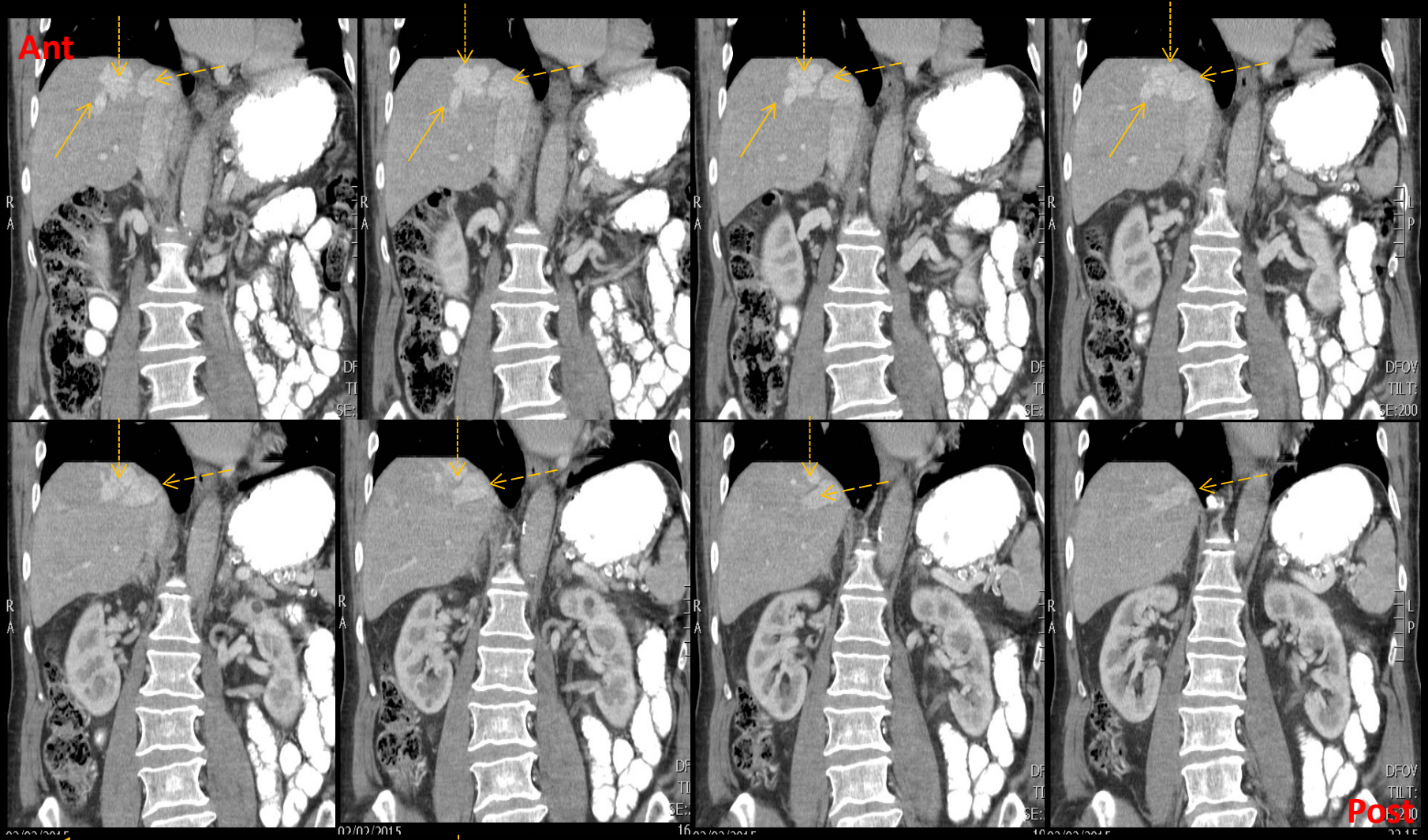
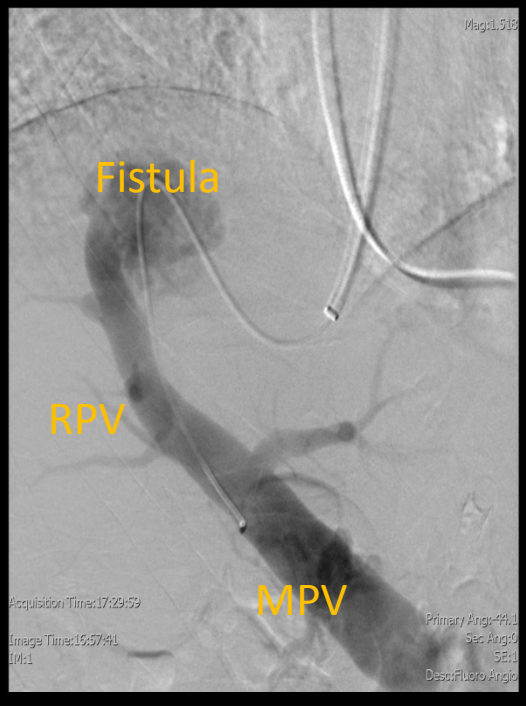


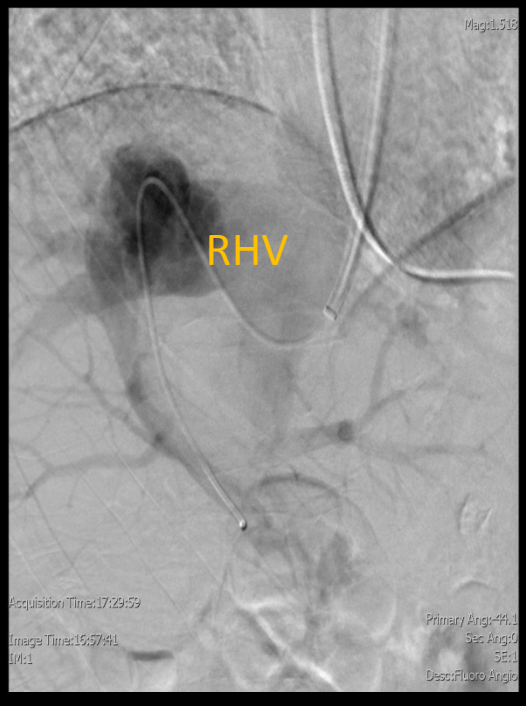
CTA – Coronals (ant to post)



Intervention

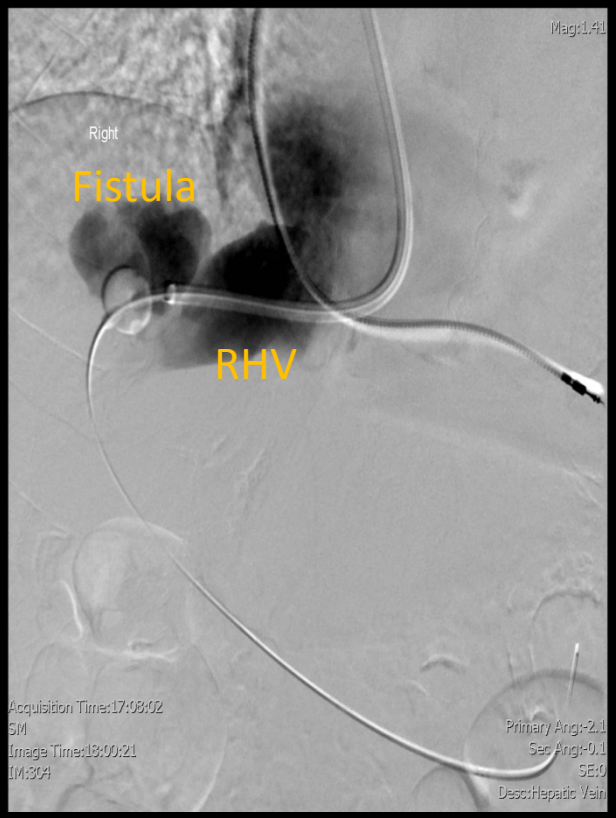
- R IJV access
- Catheterization of shunt and across into the portal system
- Venogram





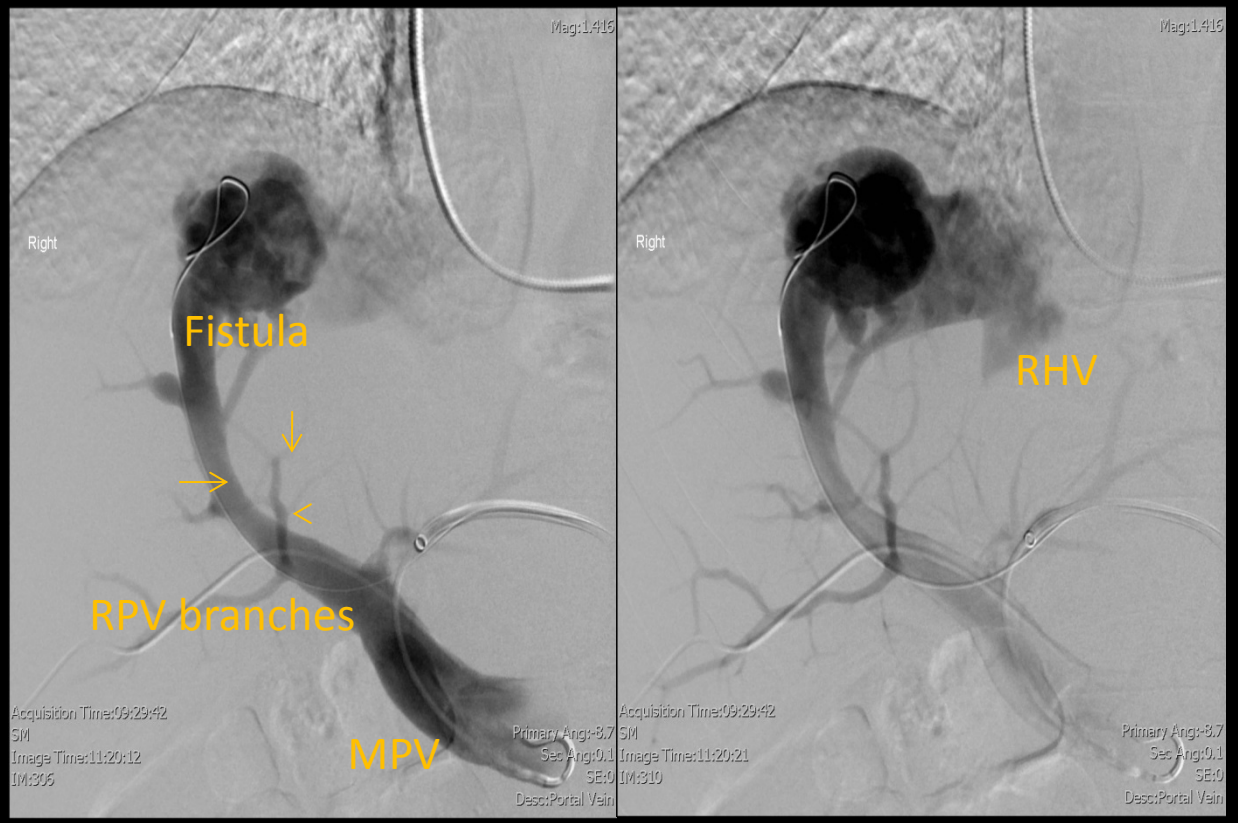
Intervention

- Pressures were measured
- Attempted occlusion balloon inflation (balloon would not stay seated)
- Procedure terminated with plans for alternative approach



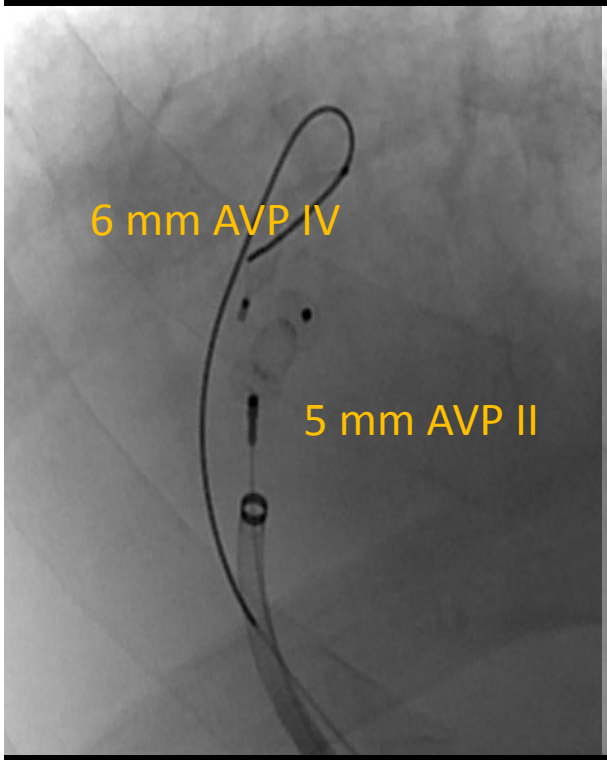
Intervention

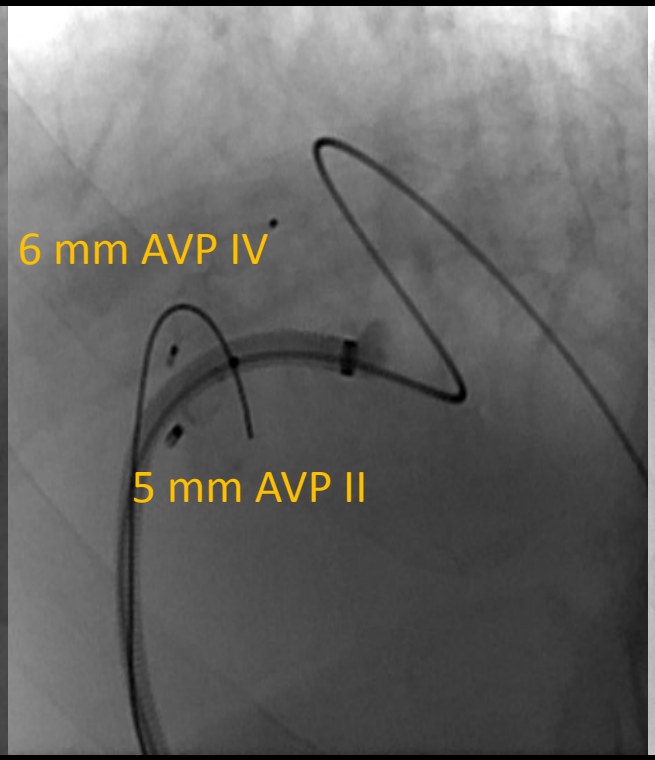
- Percutaneous access into a left peripheral portal venous branch was obtained
- Venogram showed rapid filling of the venous fistula

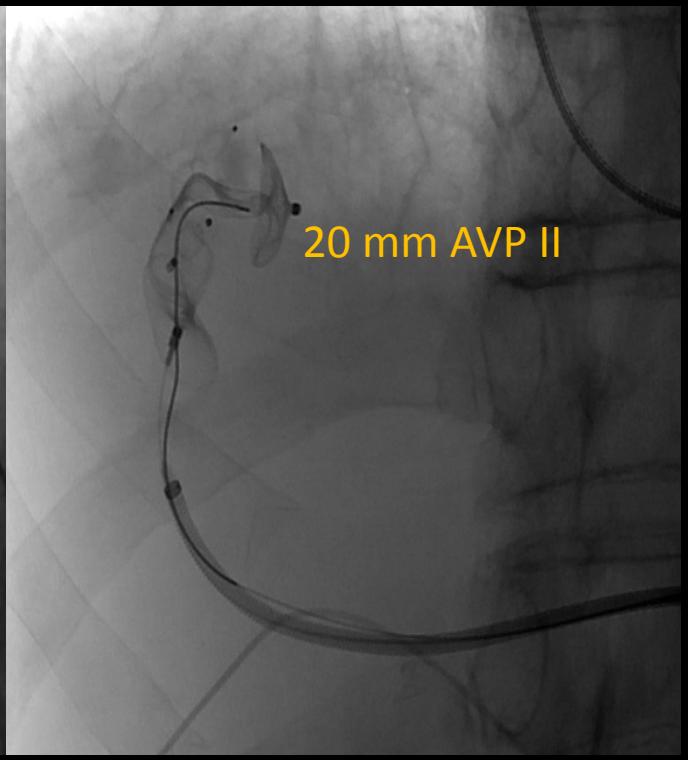


Intervention

- Multiple feeding vessels were identified, progressively selected and embolized using 4th generation Amplatzer vascular plugs







Intervention

- Completion venogram confirmed absent flow through the venous fistula

Clinical Follow-up

- Post-embolization in-hospital course complicated with bouts of shortness of breath at night attributed to worsening CHF – resolved with increased Lasix dose
- Patient transferred to home hospital
- Patient doing well – hepatic encephalopathy symptoms have resolved 3 months out

Discussion

- Etiology/pathogenesis is controversial with multiple theories:
  - Congenital (multiple pediatric cases) due to a persistent embryonic venous anastomosis or in patients without history of liver disease or abdominal trauma
  - Acquired due to portal hypertension from cirrhosis/hepatitis; iatrogenic; post-traumatic; rupture of a portal venous aneurysm into a hepatic vein
- Internal type thought more likely congenital as low prevalence of coexisting cirrhosis
- Clinical significance of this entity: potential for hepatic encephalopathy (can consider it as a “natural” TIPS)
  - Postulated that there is increased risk correlated with increasing age (brain may be more susceptible to toxic metabolites) and larger shunts

Management

- Asymptomatic – watchful waiting
- Symptomatic – first step is dietary management limiting protein intake and lactulose supplementation; if fail dietary modifications, consider intervention to shut down the shunt
- Multiple case studies reporting successful embolization of IPSVS have been detailed in the literature
- Only about 50 cases had been reported in the literature as of 2003
- 47 publications in the literature describe types II-IV (Chevalier classification – those usually not associated with portal hypertension)
- Multiple different approaches including transjugular; transileocolic; percutaneous transhepatic; and retrograde transcaval

References

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