CIRA Case of the Week

Case Courtesy of Drs. Sean A. Kennedy and Martin Simons

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Case Presentation

- 56 year old female presents to ER with right upper quadrant pain and flu-like symptoms, worsening over 2-3 weeks

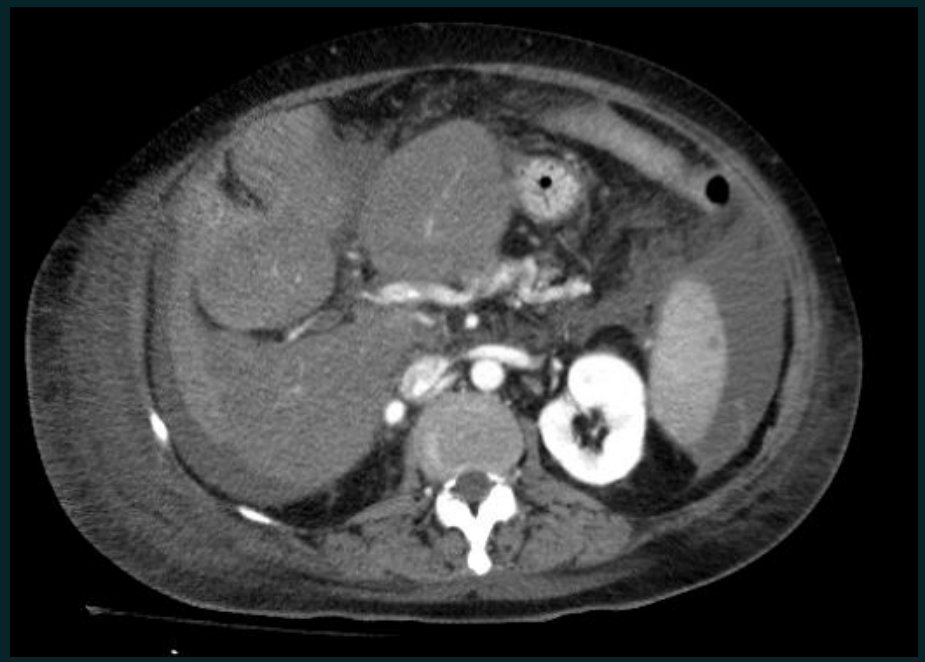
- Past medical history: Type 2 diabetes, hypertension, dyslipidemia, known polycythemia rubra vera with JAK2 mutation on rivaroxaban

- Past surgical history: Ventral hernia repair

Case Presentation

- Labwork (normal ranges in brackets):
  - Hemoglobin 173 (123-157 g/L), white blood cell count 12.2 (4-10 x 10^9/L), platelets 356 (130-400 x 10^9/L), hematocrit 50 (35-44%)
  - ALT 425 (3-36 U/L), AST 515 (35-100 U/L)
  - INR 3.03 (0.9-1.2), total bilirubin 83 (<26 umol/L), albumin 35 (35-50 g/L)
  - Creatinine 238 (60-110 umol/L)

- CT of the abdomen was performed



Case Presentation: CT Abdomen

- Complete thrombosis of hepatic veins
- Cirrhosis with caudate hypertrophy, ascites and splenomegaly
- Patent portal veins
- Caudate hypertrophy resulting in intrahepatic IVC compression

- Findings in keeping with Budd-Chiari Syndrome

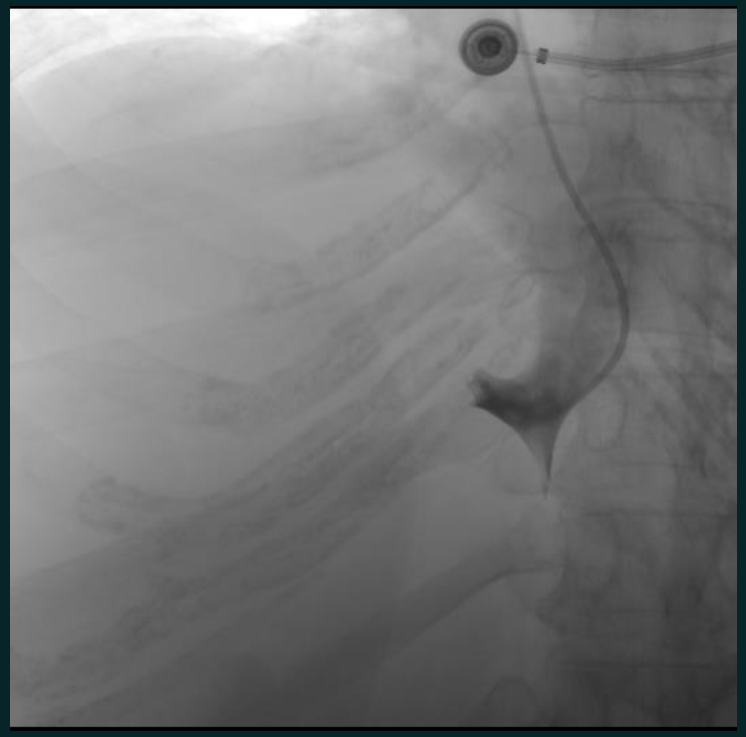
Case Presentation: Plan

- Hepatic failure, hepatorenal syndrome, pain, and ascites secondary to Budd-Chiari Syndrome
- Interdisciplinary discussion between IR, hematology, and hepatology for consideration of portosystemic shunt
- Standard transjugular intrahepatic portosystemic shunt (TIPS) not feasible due to extent of hepatic vein thrombosis

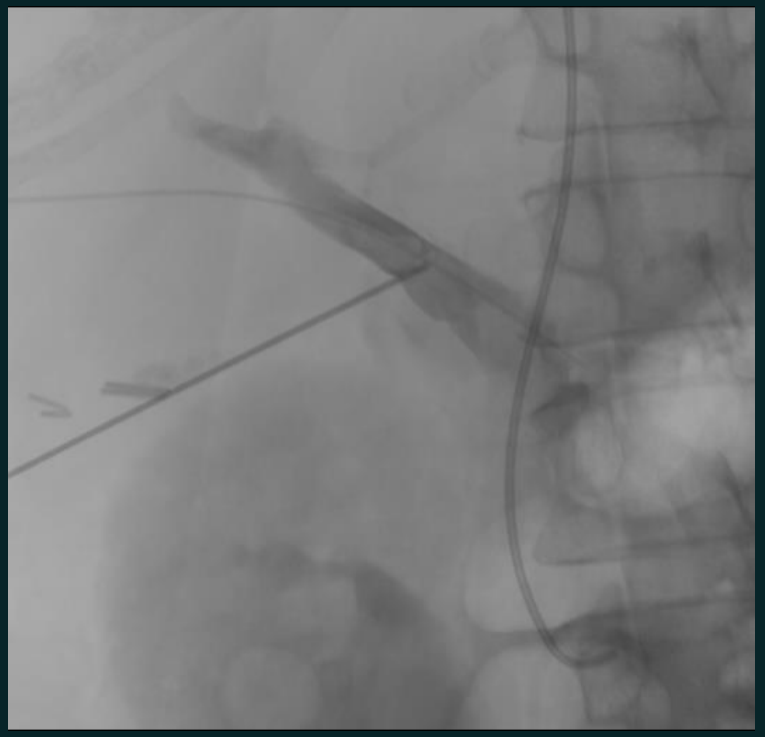
Case Presentation: Plan

- Decision made to attempt direct intrahepatic portosystemic shunt (DIPS)
- INR reversed with 4 units FFP
- Total paracentesis performed

DIPS

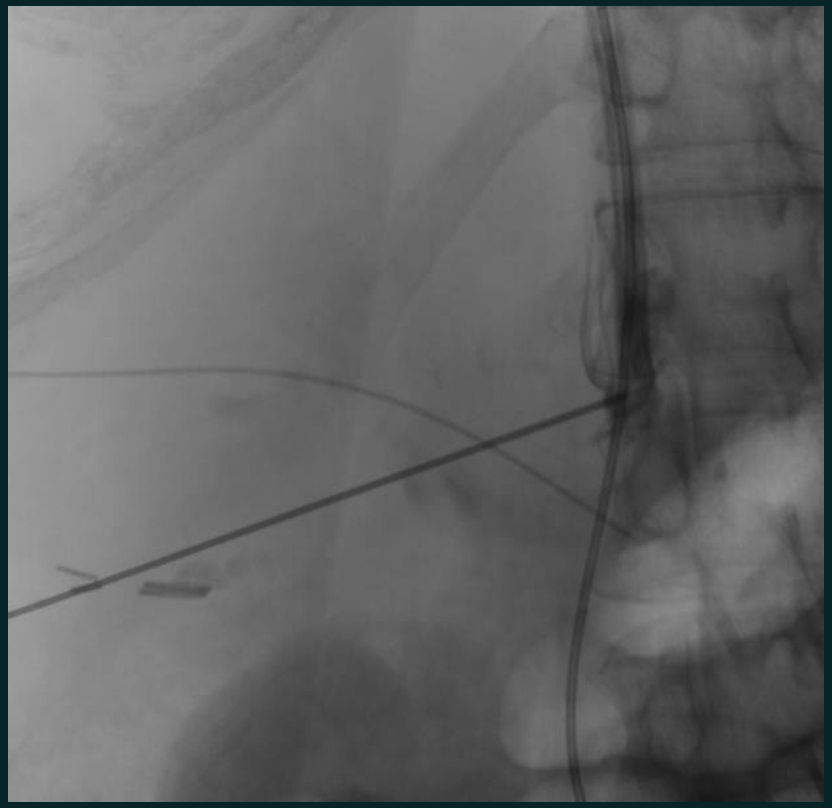


- Transjugular attempts to access hepatic veins unsuccessful
- Meniscal-like occlusion at hepatic vein-IVC confluence

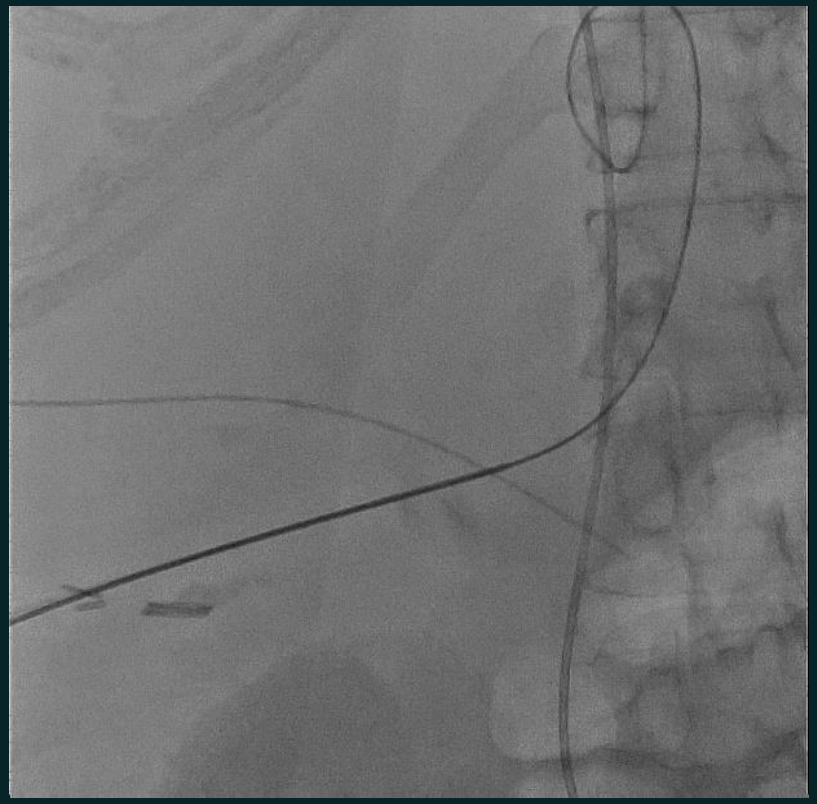


- Transhepatic portal access gained via an 18G 20 cm Chiba needle
- Transjugular liver access sheath placed in IVC

DIPS

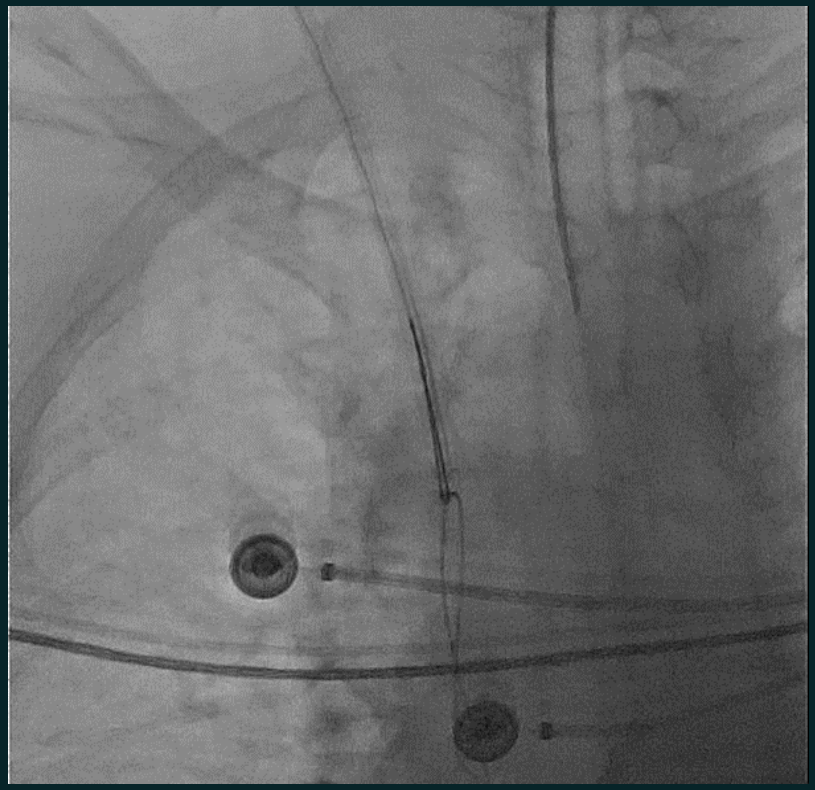


- Chiba needle advanced directly from portal vein into the IVC

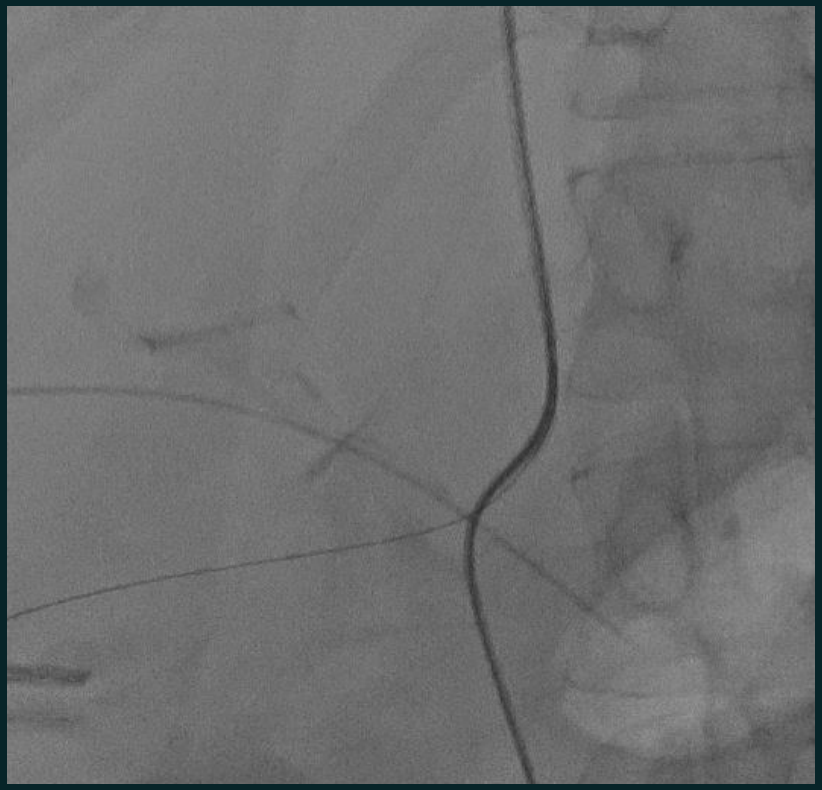


- 0.035” guidewire advanced directly through Chiba needle into the IVC

DIPS

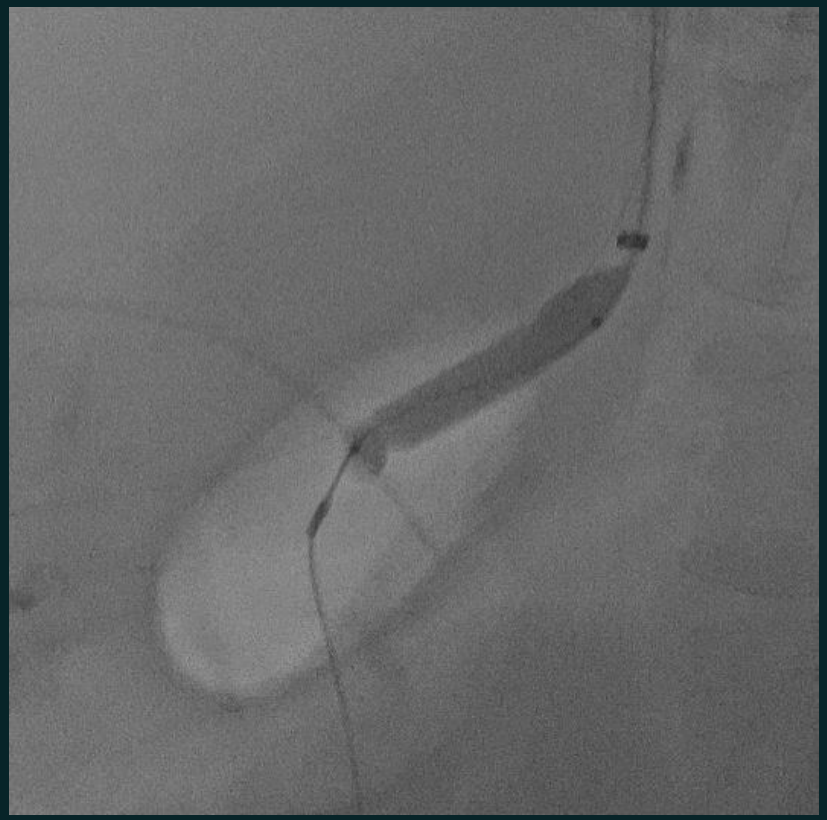


- Body-floss technique
- Guidewire snared via transjugular approach

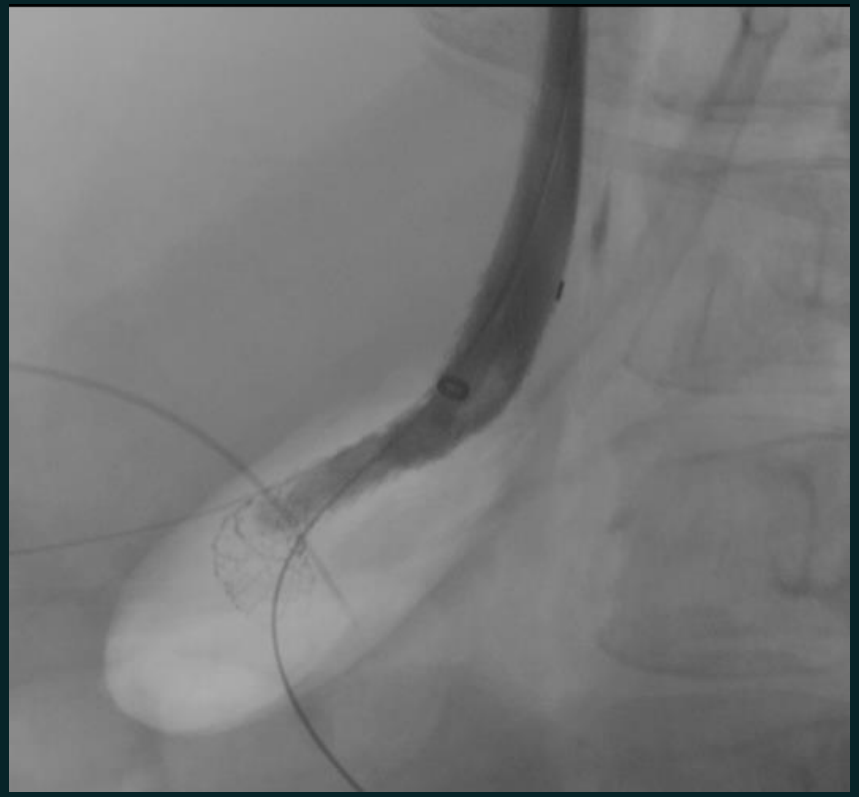


- Guidewire and 5F C2 catheter advanced through new portosystemic tract

DIPS



- Parenchymal balloon dilatation of tract using 8x40 mm Dorado



- 10x60 mm Viatorr TIPS stent graft deployed within portosystemic tract completing DIPS

Outcome

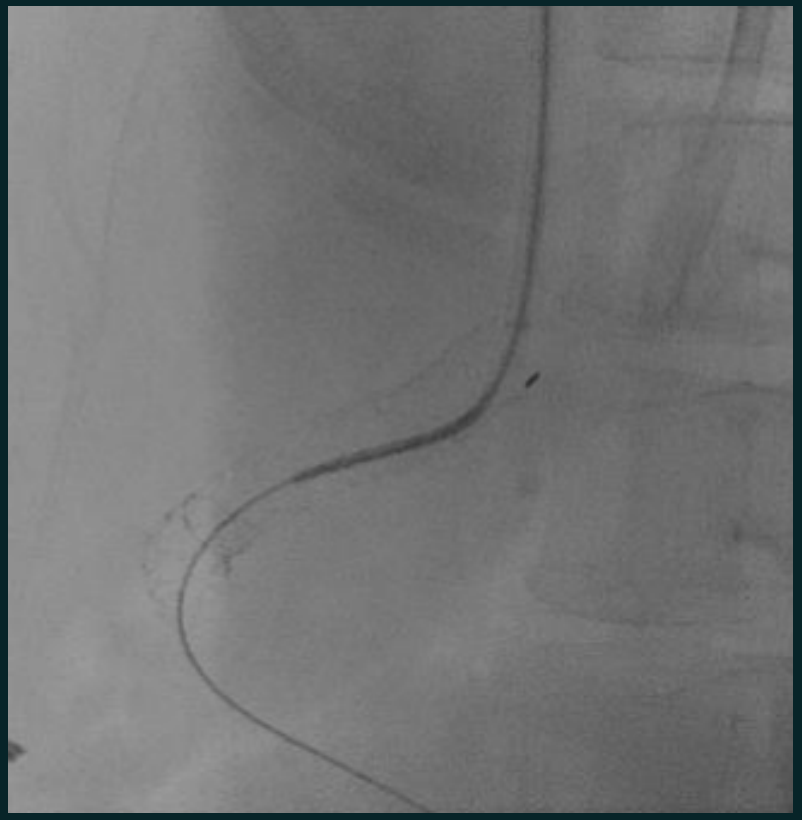
- Portosystemic gradient: Pre 25 mmHg, Post 7 mmHg
- Patient's liver/renal function, abdominal pain, and ascites rapidly improved
- Anticoagulation with low molecular weight heparin in light of extensive hepatic vein thrombosis
- Follow-up ultrasounds demonstrated shunt patency

Outcome

- Until 51 months later…
- Patient presented with recurrent ascites and abdominal pain

DIPS Occlusion

- Inferior vena cavogram demonstrates DIPS stent occlusion

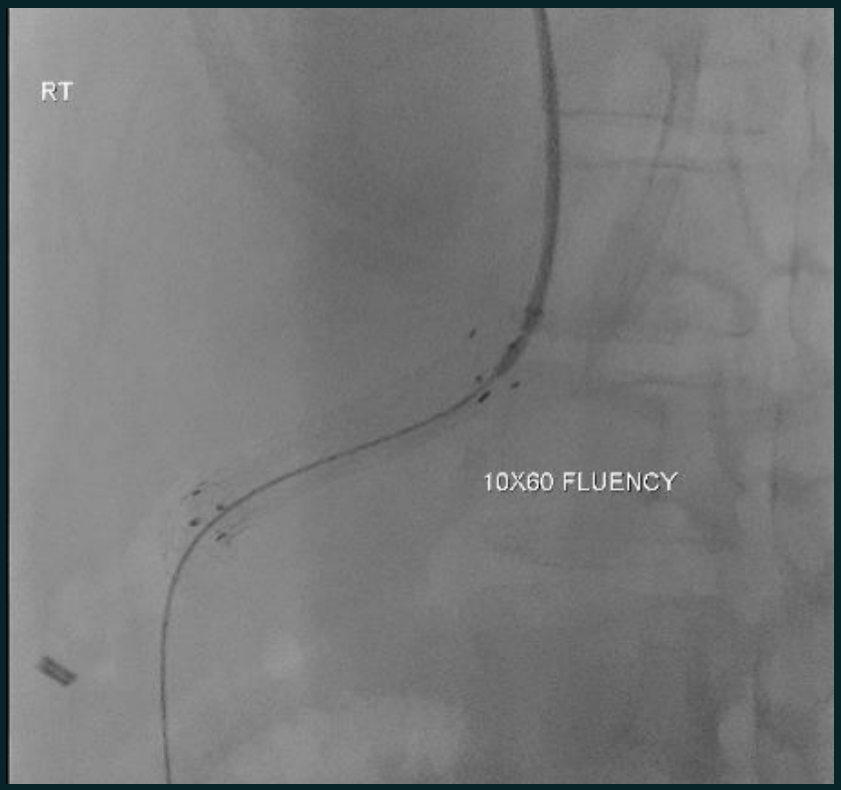


- Occlusion crossed with 5F MPA catheter and 0.035” guidewire
- 4 mg t-PA laced into thrombus

DIPS Occlusion

- 10x40 Mustang balloon angioplasty and 6F Fogarty balloon-assisted thrombectomy performed at level of DIPS occlusion

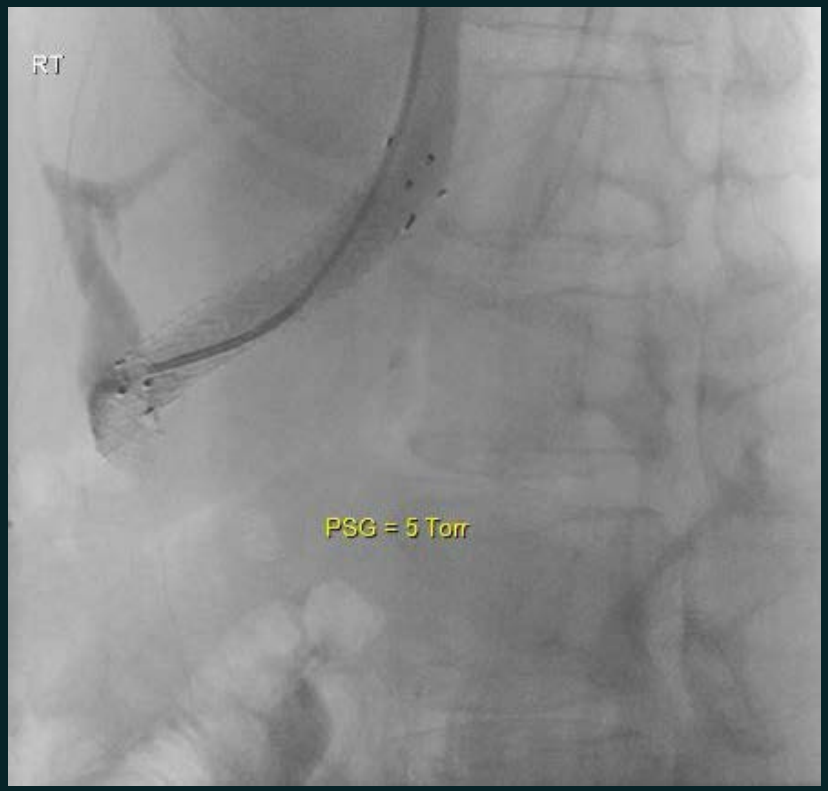
DIPS Occlusion



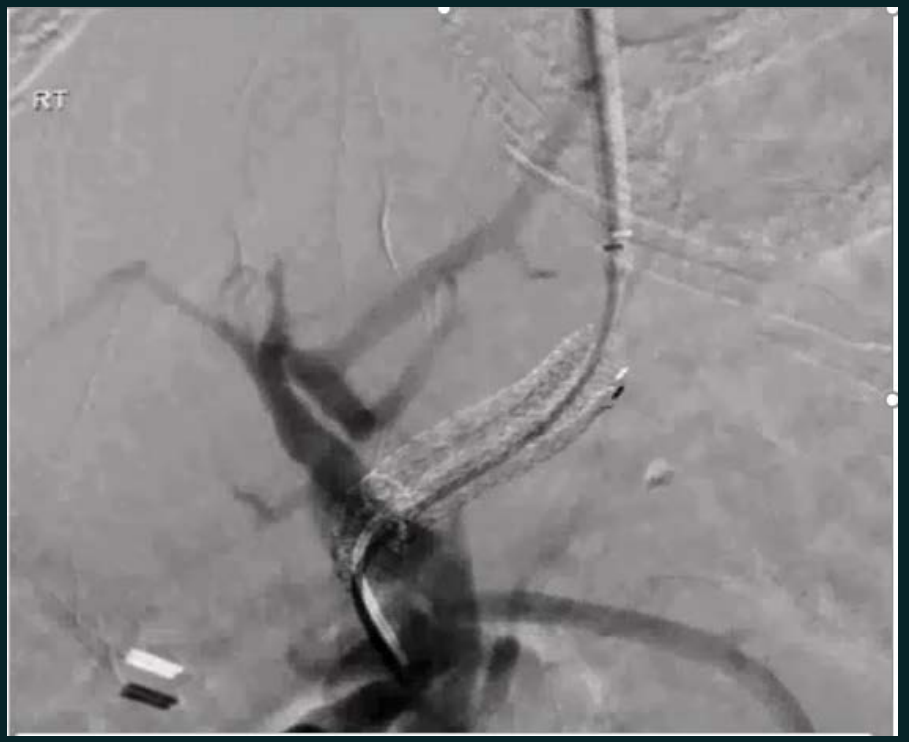
- Post-thrombectomy residual irregularity of DIPS lumen and persistent high pressure gradient of 21 mmHg

- 10x60 Fluency covered stent placed within DIPS in attempt to resolve pressure gradient
- Balloon angioplasty with 10x40 Mustang

DIPS Occlusion



- Post-Fluency stent placement and angioplasty
- Successful resolution of high portosystemic pressure gradient (5 mmHg)



- Select image of final angiographic run demonstrating patent DIPS revision

Budd-Chiari Syndrome

- Defined as hepatic venous outflow obstruction
- Primary = obstruction due to venous process
- Secondary = obstruction due to venous compression/invasion by lesion external to veins
- Etiology: Myeloproliferative disorder (up to 49% of BCS patients), malignancy, oral contraceptives, hypercoagulable states, Behcet's syndrome, membranous webs, idiopathic

Management

- Medical therapy:
  - Anticoagulation
  - Standard cirrhosis/portal hypertension management
- Interventional:
  - Recanalization of hepatic veins (catheter-directed thrombolysis, angioplasty +/- stenting)
  - Portosystemic shunt (transjugular, direct)
- Surgical:
  - Surgical portosystemic shunts
  - Transplantation

Portosystemic Shunts

- Primarily retrospective data on shunts in BCS
- Large series of 124 patients (mean 36.7 month followup) with BCS treated with TIPS:
  - 93% technical success
  - 5-year transplant free survival of 78%
  - During followup, 41% had evidence of TIPS dysfunction
  - PTFE-covered stents had significantly less shunt dysfunction than bare metal stents
- Smaller series on DIPS demonstrate high technical success rates

Back To Our Case

- Patient required hepatic decompression
- Neither recanalization nor TIPS were feasible due to the extent of hepatic vein thrombosis
- DIPS represents a potential alternative in such BCS populations having a high technical success rate and transplant free-survival
- Shunt dysfunction is common and intervention often required to regain shunt patency

References

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Tripathi D, Macnicholas R, Kothari C, Sunderraj L, Al-Hilou H, Rangarajan B, Chen F, Mangat K, Elias E, Olliff S. Good clinical outcomes following transjugular intrahepatic portosystemic stent-shunts in Budd-Chiari syndrome. Aliment Pharmacol Ther. 2014 Apr;39(8):864-72.