

Recurrent Portal Vein Thrombosis after Liver Transplantation for Budd-Chiari Syndrome

Miriam Cortes Cerisuelo MD PhD
Consultant Liver Transplant Surgeon
Institute of Liver Studies
King's College Hospital
London

Case report

- 29 years-old male
- Ulcerative colitis (poor response to steroids) in 2006
- Budd-Chiari syndrome (ascites) in 2007

Abdominal CT:

- Heterogeneous big liver
- Partial occlusion of the hepatic veins



Full prothrombotic screening:

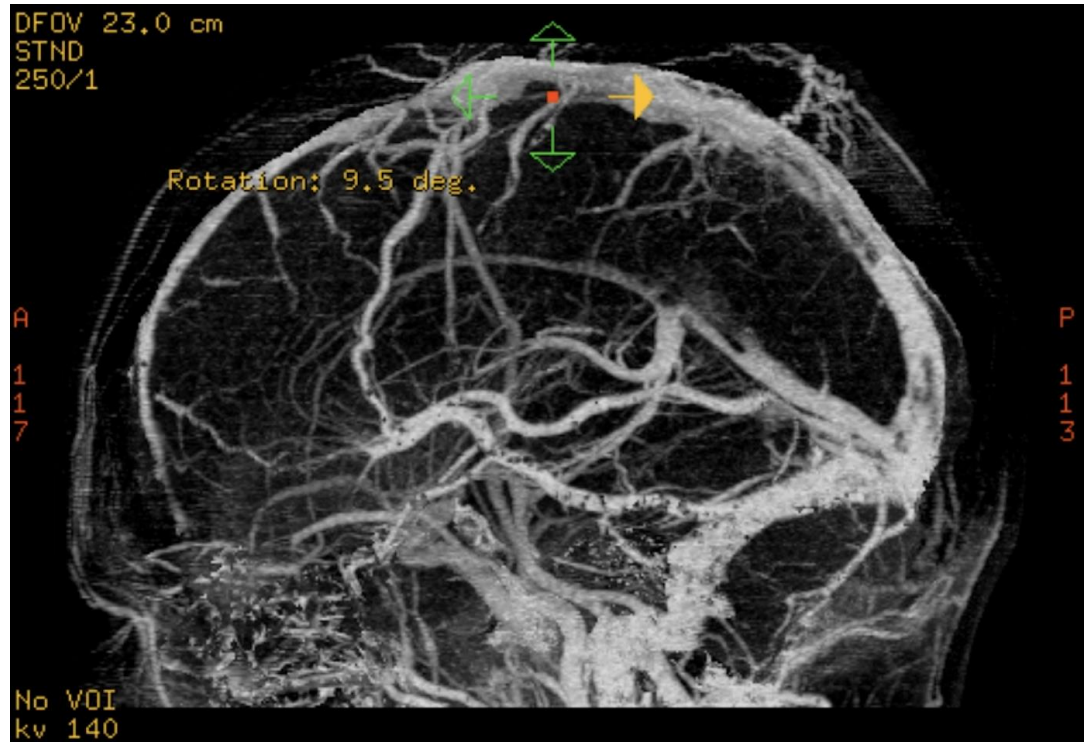
- Negative for JAK-2 mutation
 - Bone marrow normal
 - Antiphospholipid negative, Protein S normal..
 - Protein C deficiency**
- Myelodysplastic syndrome

Anticoagulated with Warfarin

Cerebral venous thrombosis in 2008 and PE

Cerebral CT venogram:

Venous thrombosis in the
sagittal sinus



-Initial improvement after TIPS in 2009

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-Progression of the symptoms:

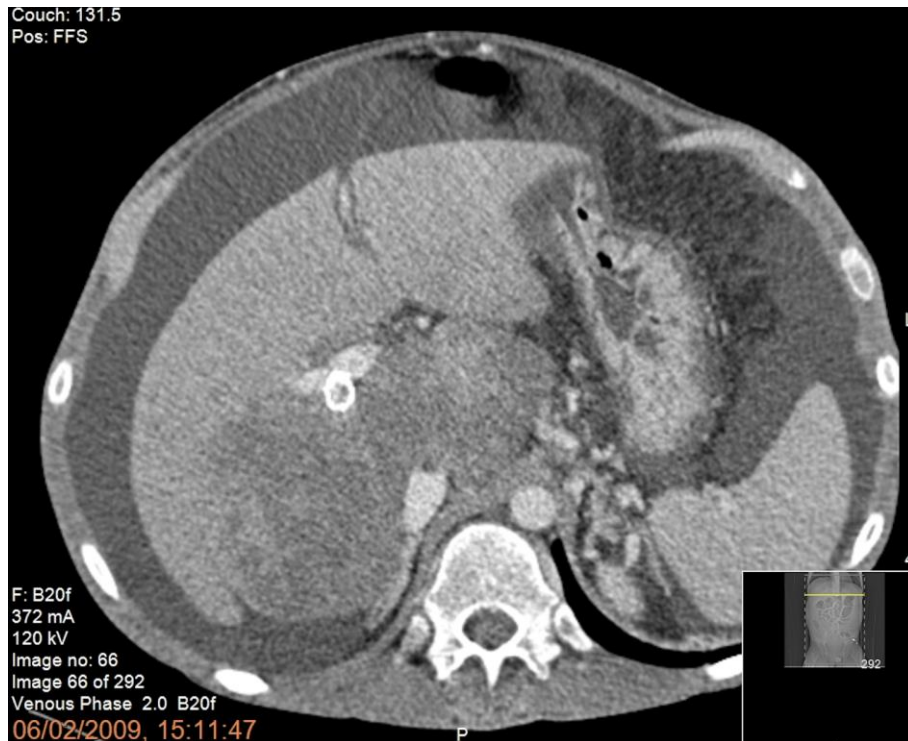
Ascites, abdominal pain, diarrhea, lethargy

-Worsening liver function: Bb 300 μ mol/L

AST 1835 IU/L

Albumin 25-35 g/L

Abdominal CT: ascites, heterogeneous liver, hypertrophy of the caudate lobe, thrombus in the TIPS and retro hepatic cava



-First liver transplant on March 2009

DBD whole graft 67 years-old

Venous-venous bypass

Caval replacement

Duct to duct

- Not fully compliant with Warfarin or Clexane
- Recurrent Budd-Chiari syndrome in 2013
- Worsening ascites, renal and synthetic dysfunction.

Warfarin/Clexane switched to Rivaroxaban

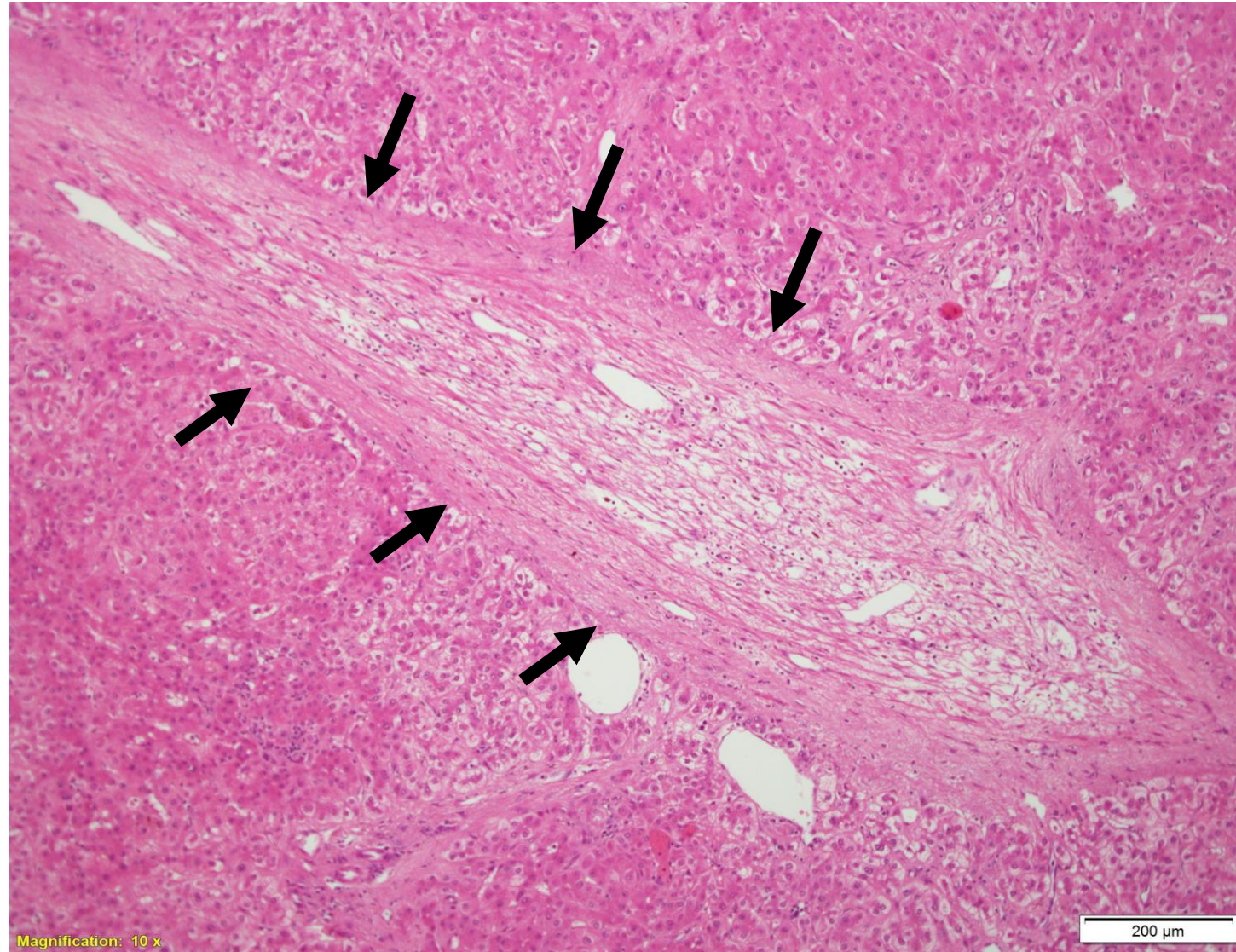
- Second LT on September 2014

DBD 54 years-old

Whole graft

Piggy-back 3 veins

Duct to duct



Hepatic vein
thrombosis

*Courtesy of Dr Alberto Quaglia, Consultant Histopathologist. Institute of Liver Studies

One month later, presented with tonic-clonic seizures.

Head CT

Intracranial fungal abscess

Voriconazol 6 months



- After stopping the antifungal, the dose of Tacrolimus was not increased resulting in severe ductopenic rejection
- Bilirubin 426 $\mu\text{mol/L}$ and no response to medical treatment

-Third re-transplantation 4 months later: January 2015

DBD whole graft 68 years-old

Venous-venous bypass

Caval replacement

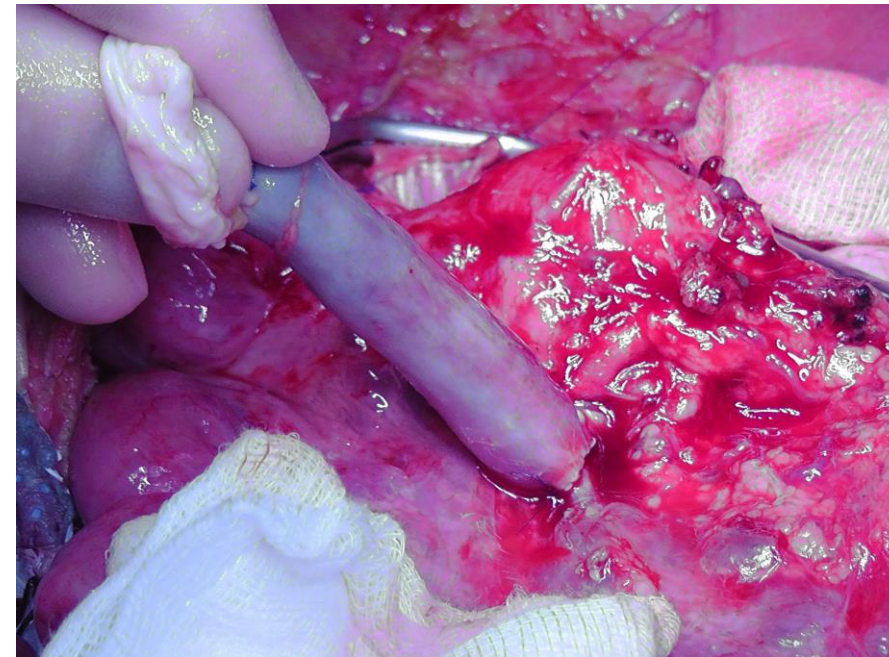
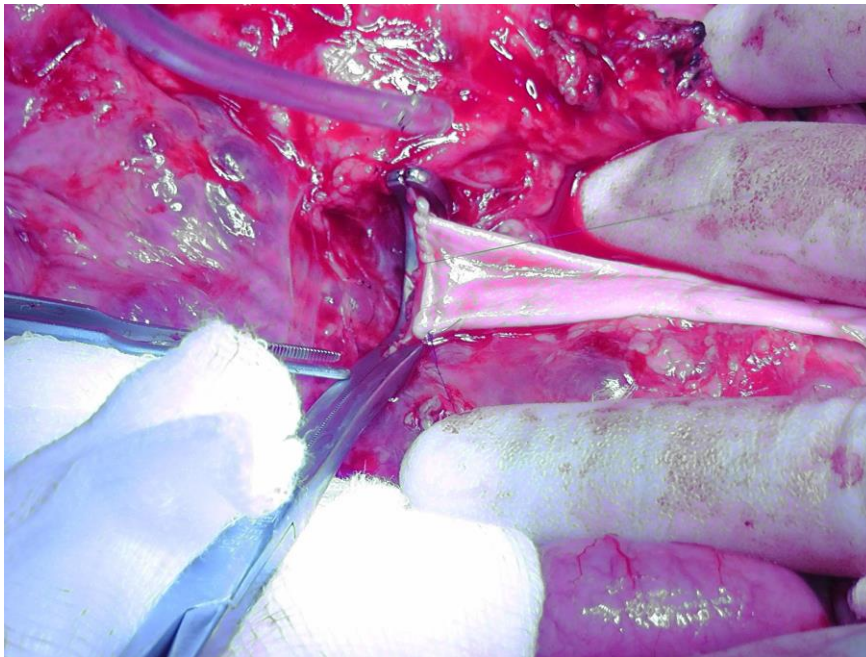
Jump graft to restore portal flow from SMV to donor PV

Duct to duct

Skin closure only

-Third re-transplantation 4 months later: January 2015

Jump graft between the SMV and donor portal vein with iliac vein from a deceased donor.



Fourth abdominal surgery:

Biliary reconstruction with a hepatico-jejunostomy for anastomotic stricture for failed endoscopic treatment on October 2015
+ full muscle closure

One year later presented with abdominal pain, renal dysfunction and ascites.

- CT showed narrowing of the venous jump graft
- PTLD? Lymph node compressing the jump graft
- Dilatation declined by radiologist “High risk”

Few months later re-admitted in hospital with:

- Gastro-intestinal bleeding
- Persistent abdominal pain, distension
- Blood transfusion requirements.

Abdominal CT

Complete thrombosis of the
previous jump graft



Fifth laparotomy for meso-Rex shunt on November 2016

Findings:

Multiple varices and moderate portal hypertension

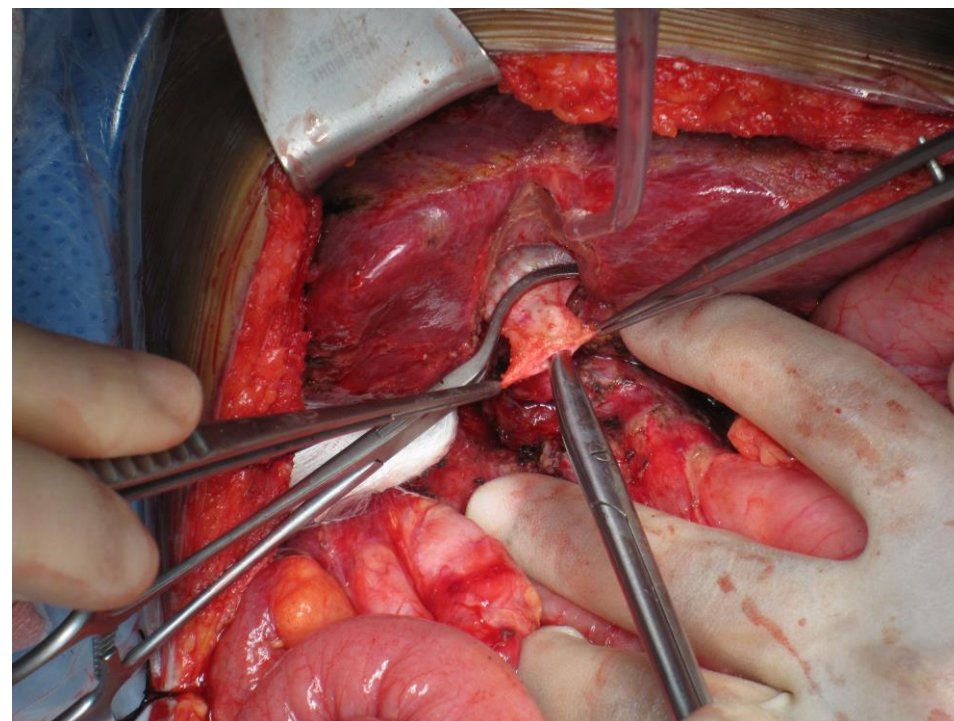
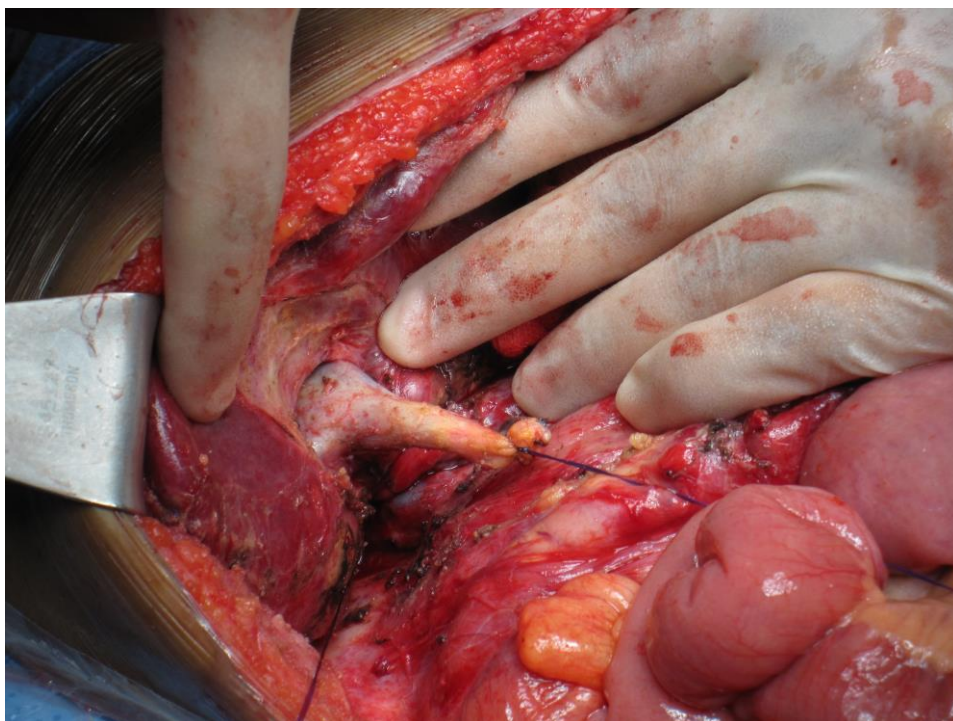
1.6L ascites

Complex surgery:

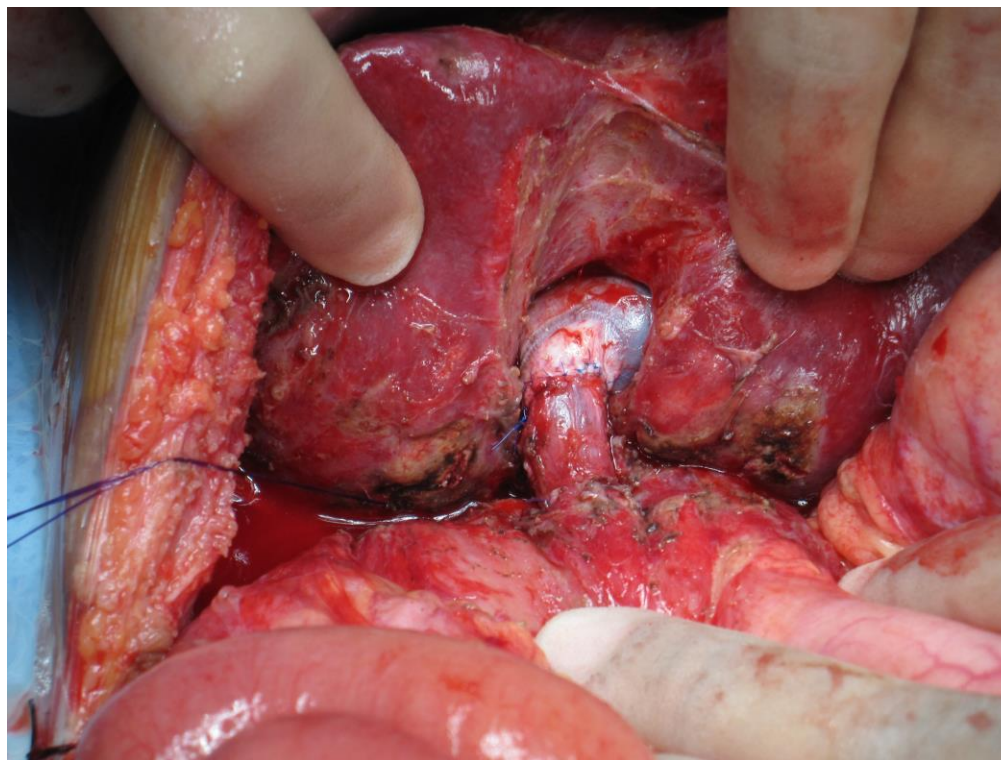
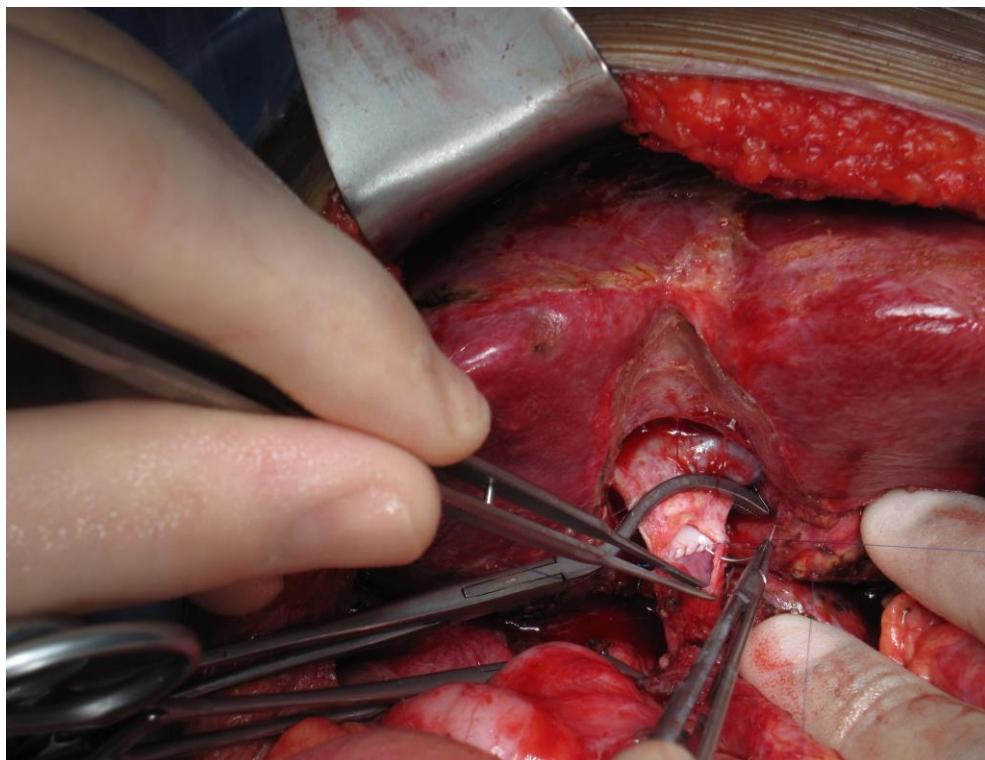
7 L blood loss

Pericardium opened during the surgery

Fifth laparotomy for meso-Rex shunt on Novemeber 2016

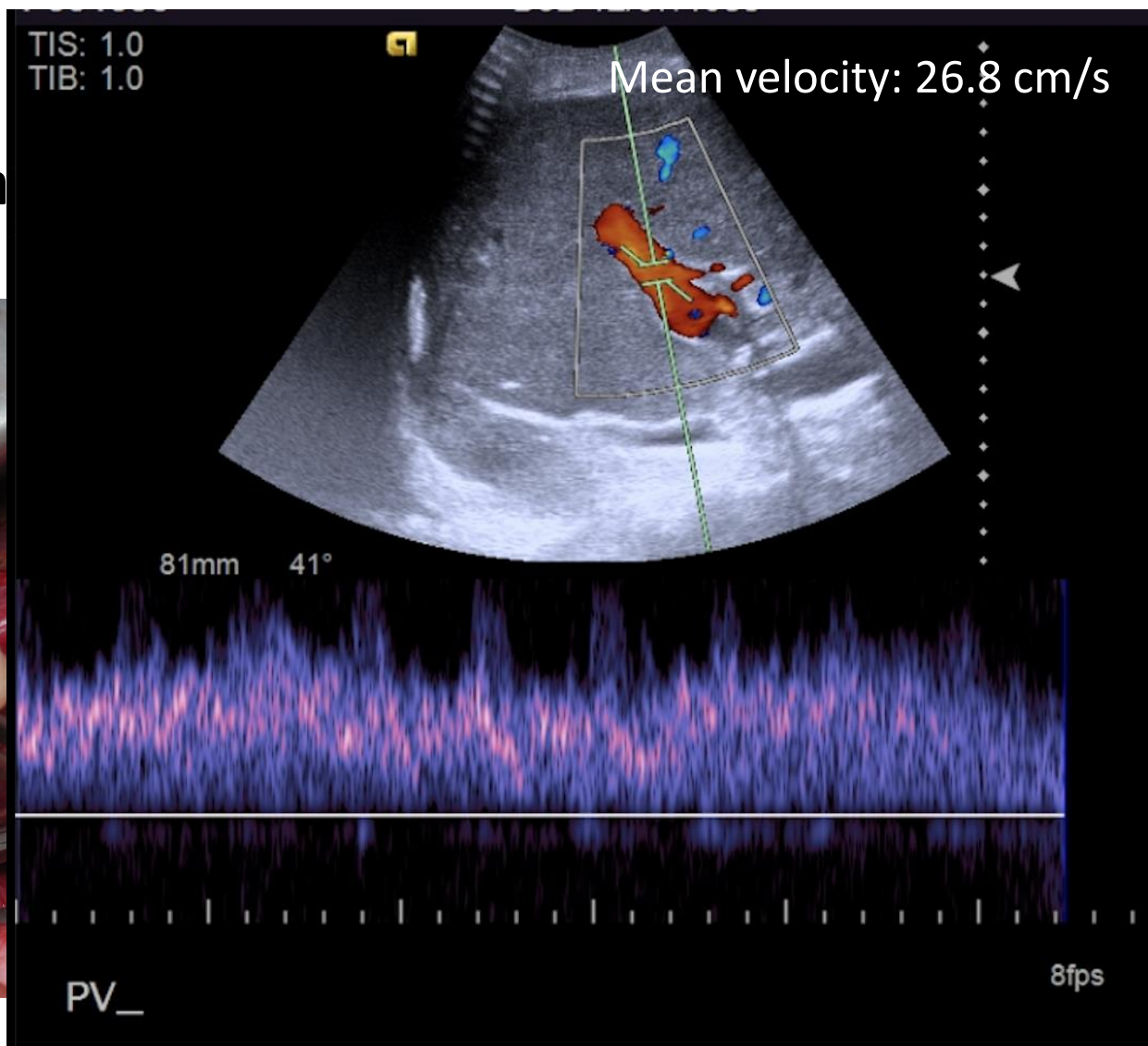


Fifth laparotomy for meso-Rex shunt on November 2016

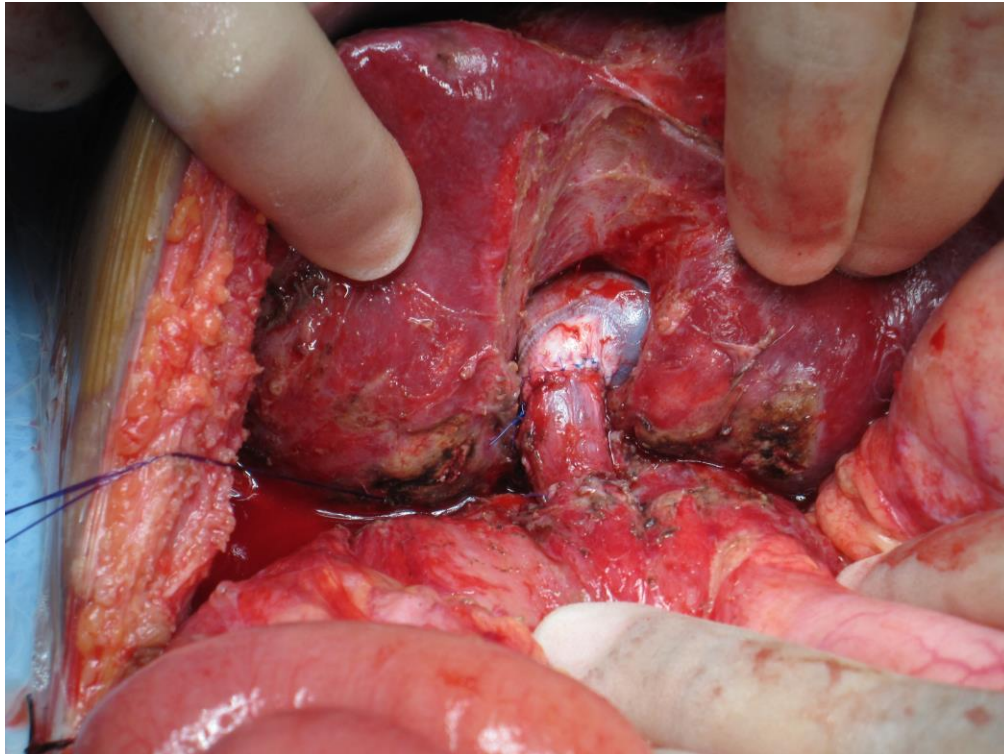


Fifth lapa

er 2016



Fifth laparotomy for meso-Rex shunt on November 2016



Pulp pressure

Before: 40 mmHg

After: 23 mmHg

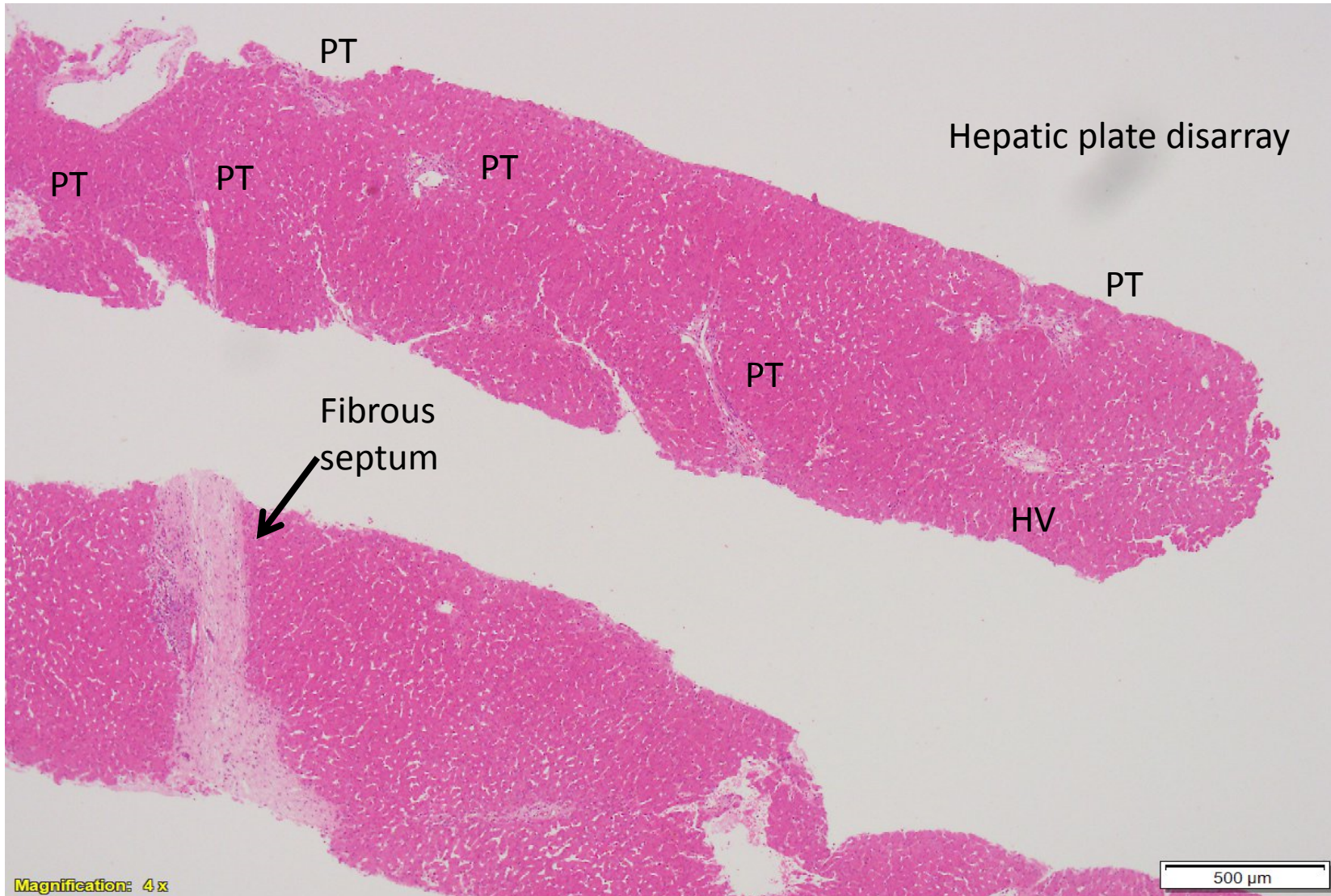
Pressure in the SMV

Before: 23 mmHg

After: 5 mmHg

Complications:

- re-laparotomy for bleeding 2 days later.
- Pericardiocentesis by cardiologist 25/11/2016, 900mls aspirated
- Chyle leak
- CMV viremia
- Pulmonary emboli



Courtesy of Dr Alberto Quaglia, Consultant Histopathologist, Institute of Liver Studies

Discharged home 6 weeks later but...

-After 8 months, admitted in his local hospital with:

abdominal pain and distension

-Abdominal CT

Thrombosis of the new jump graft

-Normal LFT:

Bilirubin 3 $\mu\text{mol/L}$

ALP 197 IU/L

AST 27 IU/L

GGT 87 IU/L

Abdominal CT:

Thrombosis of the new jump graft

Several non-occlusive thrombi

within the portal vein branches



What to do next?

What to do next?

- Wait and see...
- Refashioning of the Rex shunt +tying all possible spontaneous shunts?
- Liver re-transplantation + tying all possible shunts (left gastric...)
- Combined liver and small bowel transplant+/- colon
+/- colectomy

What to do next?

Why is he still procoagulant?

Why is he still procoagulant despite Liver transplantation?

-Protein C polymorphism in the donor liver:

development of antibodies by the recipient

-Is Rivaroxaban enough?

-Thrombotic tendency related to UC +/- episodes of dehydration?

-Donor acquired pro-thrombotic status

Thrombotic tendency in UC

Table 1 Acquired risks factors for thrombosis in IBD

1	Fluid depletion
2	Surgery
3	Central venous catheters
4	Immobilization
5	Steroid therapy
6	Oral contraceptive/hormone replacement therapy
7	Vitamine deficiency
8	Hyperhomocystenemia
9	Cigarette smoking

Table 2 Abnormalities in coagulation, anticoagulation and fibrinolytic system in IBD

Coagulation factors	Fibrinolytic factors	Plasma coagulation inhibitors
↑ Fibrinogen	↓ tPA	↓ AT III
↑ Prothrombin	↑ PAI-1	↓ TFPI
↑ Factors: Va, VIIa, VIIIa, Xa, XIa, XIIa	↑ TAFI	Conflicting data about PS and PC
↑ Prothrombin factors 1+2		
↑ Thrombin-antithrombin III complex (TAT)		
↑ Fibrinopeptide A and B		
↑ Microparticles		
↓ Factor XIII		

Thank you



