

# Ex Situ in vivo resection of leiomyosarcoma of the inferior vena cava: an anaesthetic challenge



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## INTRODUCTION

Leiomyosarcoma of the Inferior Vena Cava, although rare (380 cases in 2014 review), is the most common primary neoplasm of the IVC. They are more common in females (75%), and present most often in the fifth decade of life. Presentation is often late, with leg swelling, ascites or Budd-Chiari syndrome. Only 25% of tumours are intraluminal-significance for surgical resection. Aggressive surgical management as the only form of treatment to offer significant benefit in terms of five year survival. It is a very challenging surgery and should be done in a centre which caters for liver transplant anaesthesia.

## CASE REPORT

We present the case of a 37 year old male, ASA 2, BMI 24, who presented with bilateral lower limb swelling, ascites, shortness of breath and fatigue, due to acquired Budd-Chiari syndrome secondary to leiomyosarcoma of his IVC. Past medical history included craniotomy for medulloblastoma (age 3) with radiation to the brain and spine, and subsequent pan hypopituitarism. He also had a ventriculoperitoneal shunt at the time and multiple indwelling venous access devices. He was commenced on spironolactone and perindopril for blood pressure and oedema control and transferred to our liver transplant centre. Further investigations included a transfemoral endovascular biopsy (showing atypical smooth muscle proliferation, consistent with intravascular leiomyosarcoma), a Transthoracic Echo which showed a mass behind right atrium not moving with heart contractions, and a Transoesophageal echo which showed a visible margin between the right atrial wall and the mass. The planned surgery involved resection of the tumour including removal of a right atrial cuff, replacement of the IVC, hypothermic in-situ perfusion of the liver and left hepatectomy with reconstruction of hepatic veins. After induction the patient had a left internal jugular and right femoral venous bypass cannula inserted. We used the Vigileo(Edward Lifesciences) for intraoperative hemodynamic monitoring (Image 3). There was a prolonged anhepatic period of 117 minutes. Significant metabolic and coagulation abnormalities developed because of complete loss of hepatic function (Image 4-6). The predominant findings during the anhepatic period were hypoglycaemia and severe metabolic acidosis, with elevated levels of lactate.

Total blood loss was 10 litres. On day 2 post operatively he returned to theatre for removal of thrombus from right hepatic vein conduit and graft stent insertion. He was discharged home on day 36.

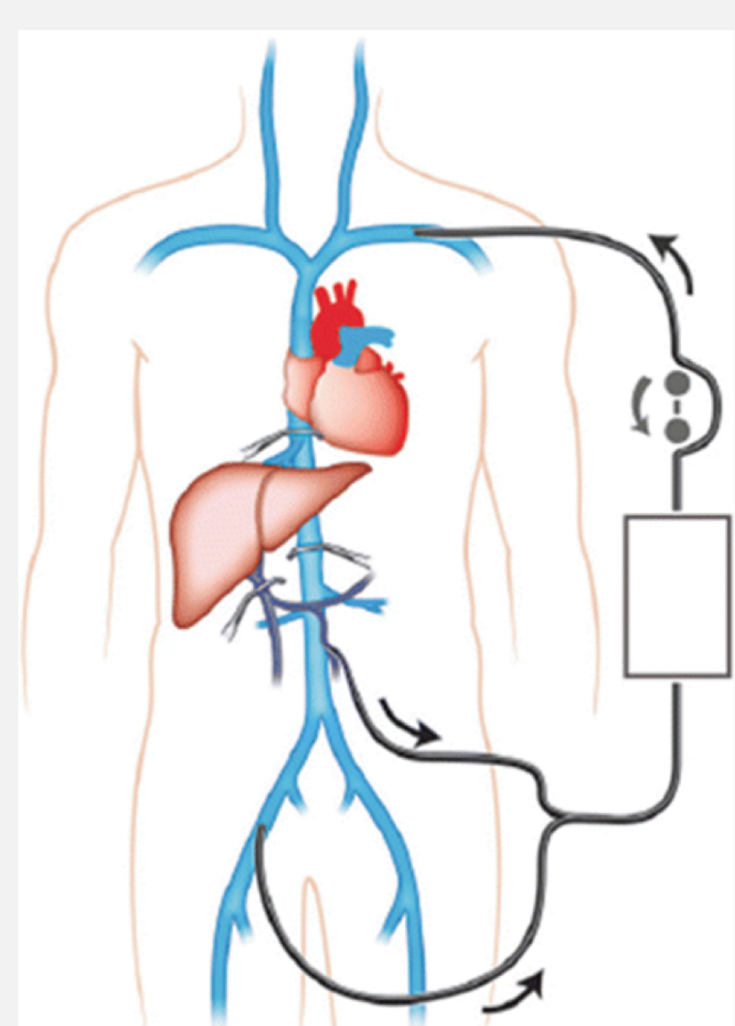


Image 1: Ex Vivo liver resection set up

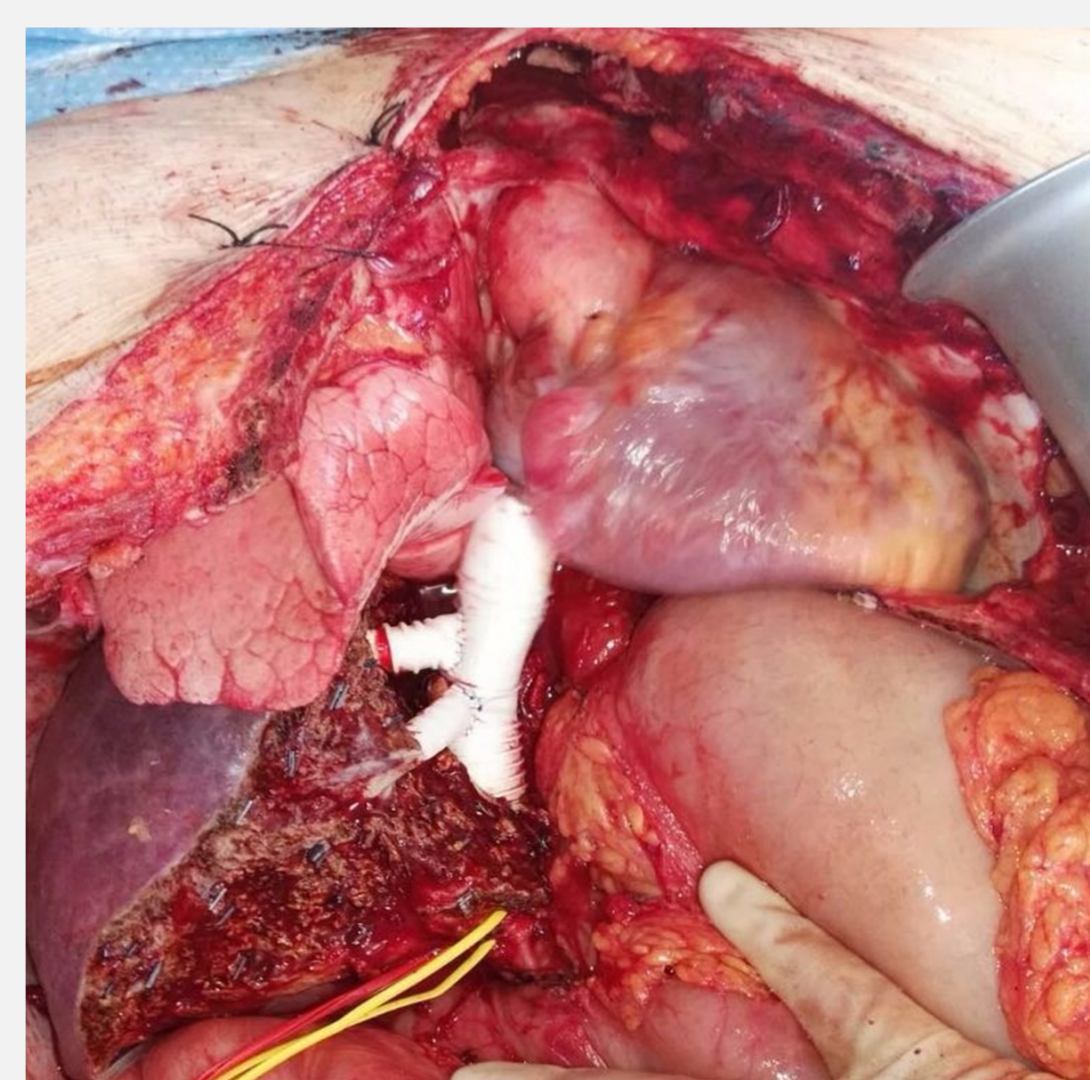


Image 2: Final operative result with graft and hepatic vein reconstruction

## DISCUSSION

Anaesthetic challenges included

1. Difficult central venous access including for large bore endogenous bypass (VV bypass) line secondary to the patient's short stature and multiple previous childhood lines. There was only a tiny right internal jugular vein on ultrasound. Left internal jugular was cannulated for VV bypass and right subclavian vein for 5 lumen oximetry line.
2. Hypothermia down to 33C secondary to VV bypass and in situ cold perfusion of liver.
3. Clamping of the right atrium with hemodynamic compromise.
4. Prolonged an hepatic phase with warm ischaemia, and associated reperfusion injury including massive coagulopathy, fibrinolysis and hyperkalaemia.
5. Hypoglycaemia and hyperlactatemia

## INTRA OP VARIABLES

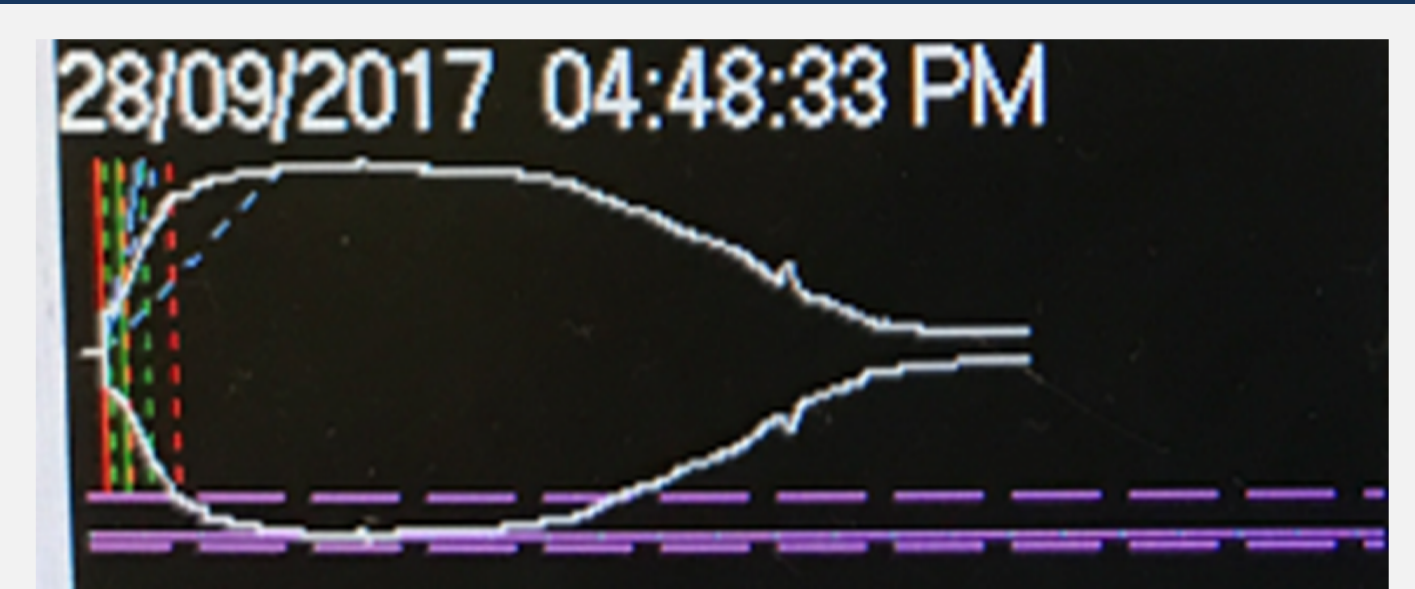


Image 4: An hepatic phase

Image 3: Hemodynamic changes intra op

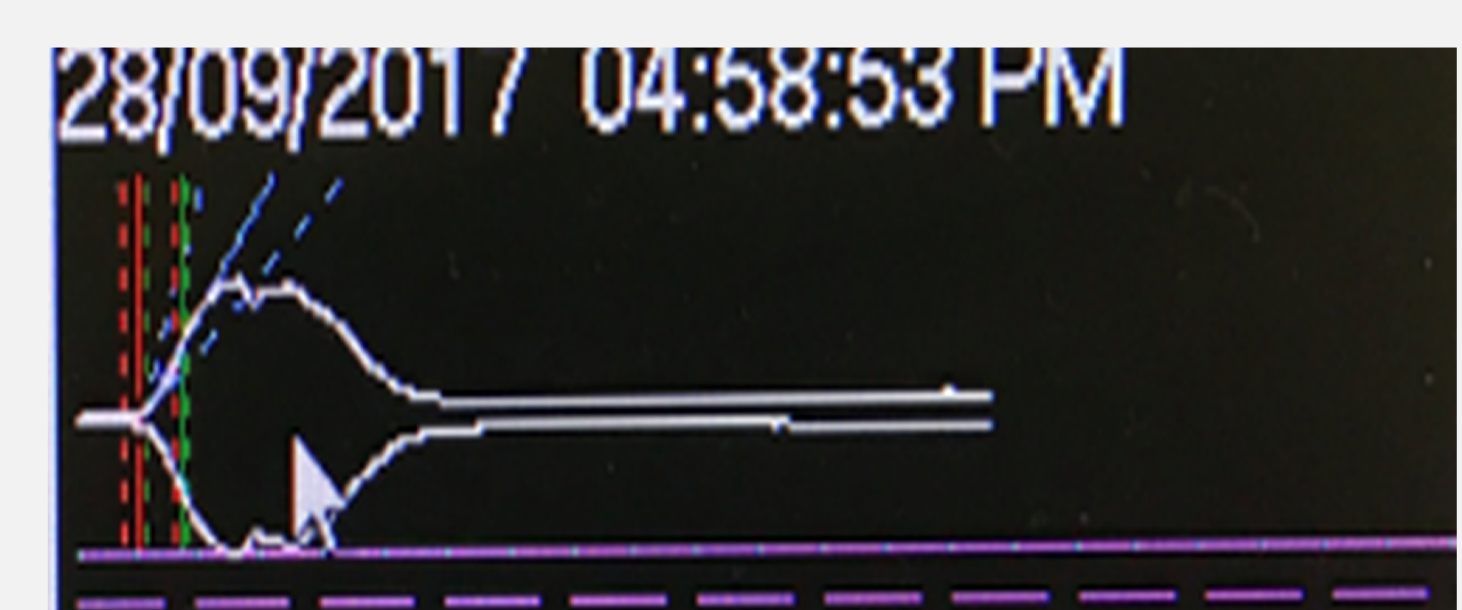
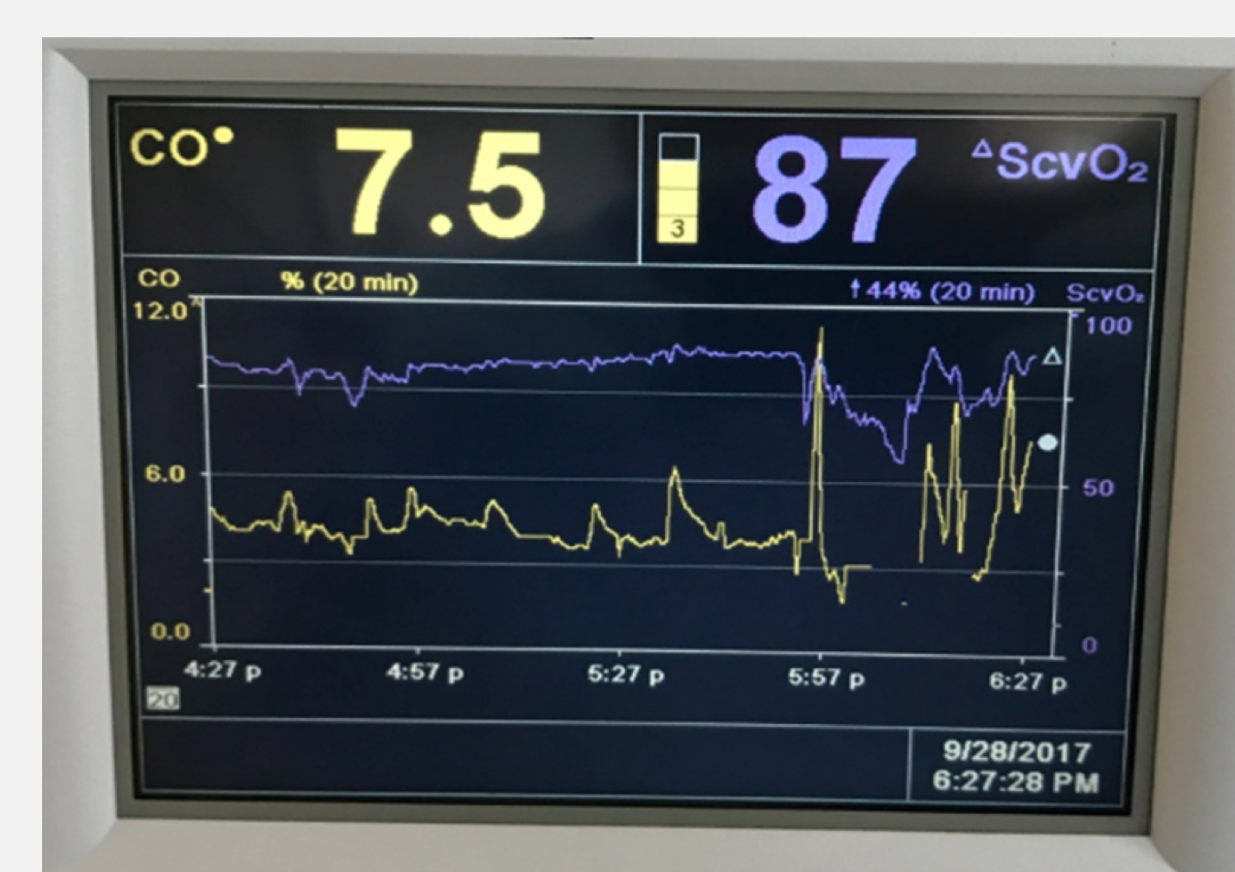


Image 5: Post reperfusion Thromboelastogram(hyper fibrinolysis)

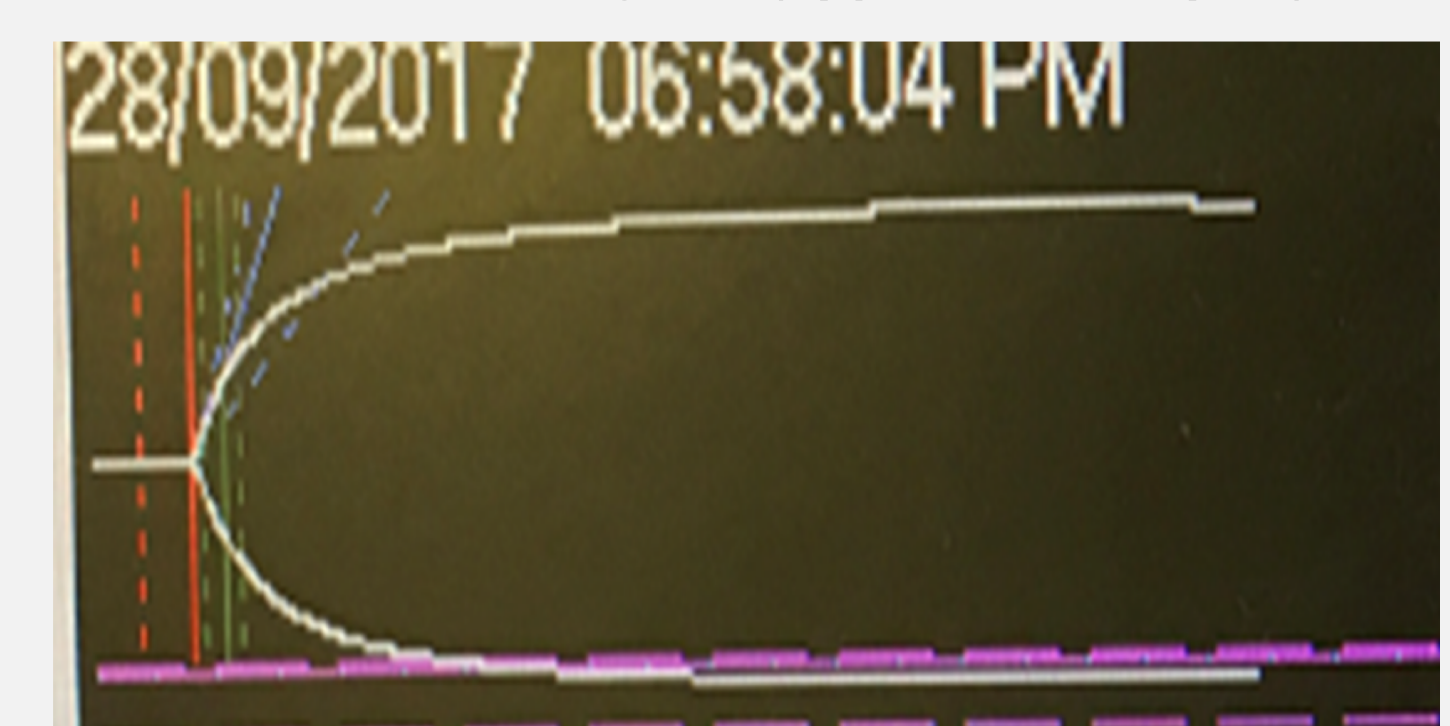


Image 6: Thromboelastogram at the end of the surgery

## CONCLUSION

This case highlights the unique challenges of anaesthesia provision for ex-situ in-vivo liver surgery. These unique cases require multidisciplinary pre-operative planning and should only be undertaken in centres performing liver transplants. The participation of a multidisciplinary team in ex vivo liver surgery is of utmost importance. Optimal anaesthetic management of hemodynamic and coagulation is essential. (Fujun Cheng et al.)

## REFERENCES

- ♦ Gallagher TK, Udupa KV, Geoghegan JG et al. Techniques and outcomes of combined inferior vena cava and visceral resection for benign and malignant disease. Int J Surg. 2014; 12(8): 864-7
- ♦ Fujun Cheng, Zhiyong Yang, Jing Zeng et al. Anaesthesia management of Modified Ex Vivo Liver Resection and Auto transplantation. Ann Transplant 2018; 23:274-284 DOI: 10.12659/AOT.907796