Successful Liver Transplantation in a Patient with Alstrom Syndrome and Cardiomyopathy: A Case Report

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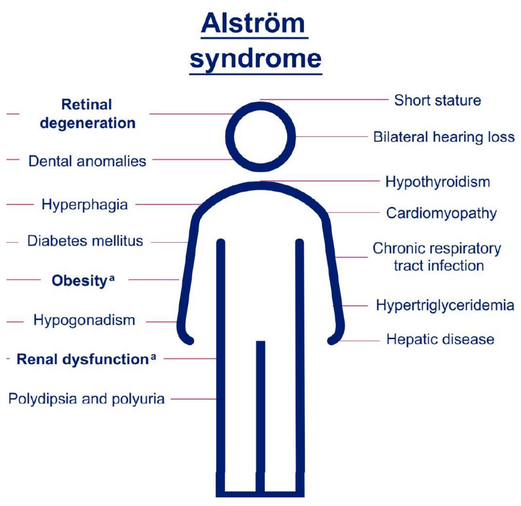
BACKGROUND

- Alström syndrome (AS) is a rare autosomal recessive disorder due to *Alström syndrome 1* (*ALMS1*) mutation
- Characteristics: progressive multiorgan failure with early onset coronary artery disease, cardiomyopathy, fatty liver disease, vision and sensorineural hearing loss.
- Average age expectancy ~50 years.
- Treatment is supportive.
- Only 1 case in literature to date of patient undergoing successful liver transplant. We present a case of successful liver transplant in an Alström patient with pre-existing cardiomyopathy



CASE

- 33 year-old female patient with AS and the following complications:
  - blindness
  - hearing impairment
  - Cardiomyopathy: low-normal left ejection fraction, severe biatrial enlargement, moderate mitral and tricuspid regurgitation, paroxysmal atrial fibrillation
  - NASH cirrhosis
- Presented for transplant evaluation after developing decompensated hepatic failure after variceal banding and TIPS.
- Multidisciplinary discussions resulted in a recommendation for liver transplantation only despite previous cardiomyopathy.
- MELD 40 at time of transplant.

INTRAOPERATIVE

- The operation was performed via piggyback technique with portal bypass and continuous renal replacement therapy.
- Catheters were placed preoperatively for rapid initiation of VA ECMO if needed, and cardiac surgery was available on standby.
- Intraoperative transesophageal echocardiography was notable for significantly depressed systolic function with mild hypocalcemia, resolved after repletion.
- Reperfusion was notable for significant bradycardia, but was otherwise uneventful.

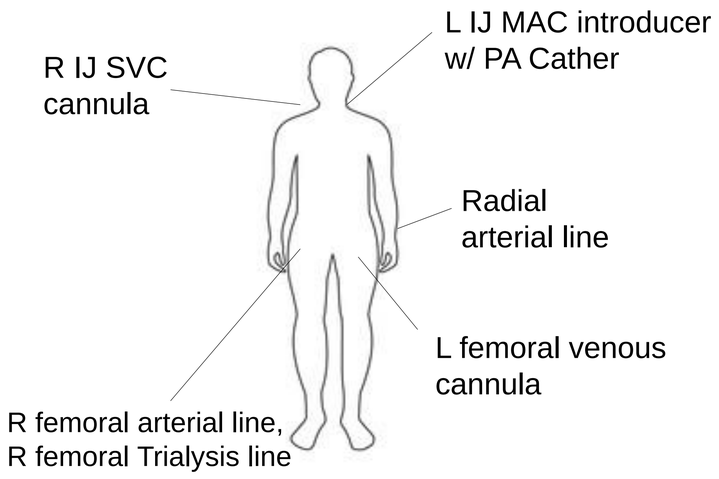
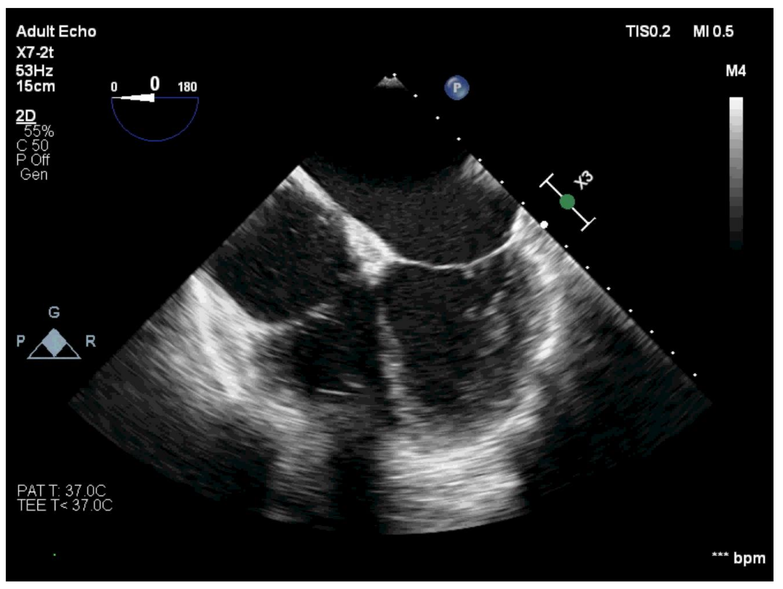


Diagram of vascular access for the case. Bilateral groins accessed to facilitate cannulation to VA ECMO if needed



Intraoperative TEE: Dilated atria, impaired biventricular function

POSTOPERATIVE

- ICU course complicated by ongoing arrythmia and prolonged ventilator weaning.
- Delayed renal recovery with ongoing need for dialysis.
- Discharged post-op day #73.

- 1 year update:
  - Hepatic congestion requiring IVC stenting, but otherwise excellent liver function.
  - Paroxysmal atrial fibrillation on amiodarone – ablation planned in the future.
  - Renal recovery no longer needing hemodialysis, but CKD IV.
  - TTE 10 months postop: normal left and right ventricular size and systolic function. Moderate mitral regurgitation, mild tricuspid regurgitation.

DISCUSSION

- Liver transplantation is safe in patients with Alström disease.
- Multidisciplinary role in liver transplantation for patients with additional organ dysfunction.
- Transesophageal echocardiography is a safe and useful tool for monitoring dynamic changes in cardiac function.
- Further long-term observation needed to understand impact of liver transplant on progressive diseases.
- What considerations are needed for transplantation in patients with progressive comorbid diseases.

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