< VGH Guidelines for Management of Liver Transplantation (LT) Candidates with Portopulmonary Hypertension (POPH) >

* Abbreviations
  + LT: liver transplantation
  + PH: pulmonary hypertension
  + POPH: portopulmonary hypertension
  + RHC: right heart cath
  + mPAP: mean pulmonary artery pressure
  + PVR: pulmonary vascular resistance
  + PCWP: pulmonary capillary wedge pressure
  + TPG: transpulmonary gradient
  + CO: cardiac output
  + TTE: transthoracic echocardiogram
  + TEE: transesophageal echocardiogram
  + ECMO: extracorporeal membrane oxygenation
  + PAC: pulmonary artery catheter
  + RVSP: right ventricular systolic pressure
* Definition & Clinical Diagnosis of POPH
  + Presence of PH in a patient with portal hypertension, in whom other causes of PH have been ruled out.
  + Pulmonary hemodynamics must be evaluated by a RHC.
    - mPAP > 25 mmHg
    - PVR > 240 dynes/s per cm-5
      * PVR = (mPAP – PCWP)/CO \* 80
      * Normal PVR = 20-160 dynes/s per cm-5, or 0.25-2 Woods units
    - PCWP < 15 mmHg
    - TPG = mPAP – PCWP
      * TPG > 12 mmHg reflects obstruction to flow, and distinguishes the contribution of volume and PVR to the increased mPAP.
    - CO (L/min)
  + Severity (assuming PVR is increased)
    - Mild: 25 ≤ mPAP (mean PAP) < 35 mmHg
    - Moderate: 35 ≤ mPAP < 45
    - Severe: 45 ≤ mPAP
* Clinical implications
  + 5-6% of LT candidates have hemodynamic criteria meeting diagnosis of POPH.
  + Presence or severity of POPH does not correlate with the severity of liver disease or the degree of portal hypertension.
  + Mild POPH is not associated with increased mortality or graft failure following LT.
  + Moderate to severe POPH is associated with increased perioperative morbidity and mortality.
  + Many LT centres consider mPAP > 50 mmHg (or > 45 mmHg in some) an absolute contraindication to LT due to prohibitively high risk of mortality.
  + The RV function should be taken into an account when determining the likelihood of a successful transplantation.
    - Any degree of RV dysfunction increase perioperative risk
  + There is no data demonstrating predictable resolution of POPH following a successful LT. As such, POPH itself is not an indication for LT.
* Screening and Initial Evaluation
  + All LT candidates should be screened for POPH with a TTE.
    - TTEs done in smaller none cardiac centres should be repeated at VGH with a focus on POPH
  + LT candidates with RVSP > 45 mmHg on TTE and/or significant RV hypertrophy, dilation, or dysfunction should be referred for RHC.
    - Patients referred for RHC should have Dr. Brunner requested to perform procedure
  + LT candidates with RHC criteria of POPH should be referred to the PH clinic for consideration of pulmonary vasodilator therapy.
  + For patients with borderline RVSP (35-45 mmHg) repeat TTE should be performed every 6 months.
    - Earlier repeat TTE should be considered if there is a change in clinical status
  + For patients treated for POPH repeat TTE will be performed regularly at the discretion of the PH clinic.
* Preoperative Management
  + Every effort should be made to perform these transplants during daytime hours when personnel and services are readily available.
  + A backup recipient candidate should be admitted to hospital if a patient with POPH is the primary recipient candidate.
  + In-hospital ECMO service should be notified of the booked LT and the potential need for rescue ECMO in setting of pulmonary hypertensive crisis with RV failure.
    - The booking surgeon will notify the OR desk that the LT candidate has POPH and the anesthesiologist can then notify the ECMO service.
* Intraoperative Management
  + PAC monitoring should be conducted prior to and during LT in all adult cases of known POPH.
  + TEE monitoring should be conducted in the setting of POPH to monitor RV function, unless absolutely contraindicated
  + Intraoperative, post-induction, pre-abdominal incision mPAP, CO, PCWP, and PVR should be measured with PAC.
  + If mPAP < 35 mmHg,
    - Proceed with LT if RV function is preserved.
  + If 35 ≤ mPAP < 45 mmHg,
    - PVR < 240 and TPG <12
      * Proceed with LT if RV function is preserved.
    - PVR > 240 or TPG >12
      * Refer to PH clinic for optimization prior to LT
    - Consider intraoperative pulmonary vasodilatory therapy including inhaled nitric oxide and/or intravenous milrinone, especially during reperfusion.
  + If mPAP > 45-50 mmHg
    - Abort LT.
    - Etiology and reversibility of elevated PAP should be assessed.
  + ECMO should be considered for acute rise in mPAP with RV dysfunction.
* References
  + Krowka MJ, Fallon MB, Kawut SM, et al. International Liver Transplant Society Practice Guidelines: Diagnosis and Management of Hepatopulmonary Syndrome and Portopulmonary Hypertension. *Transplantation* 2016;100(7):1440-52.
  + Ramsay M. Portopulmonary Hypertension and Right Heart Failure in Patients with Cirrhosis. *Curr Opin Anaesthesiol* 2010;23:145-50.