< 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.