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**ARTICLE OF THE MONTH**

Koulava A, Sannani A, Levine A, Gupta CA, Khanal S, Frishman W, Bodin R, Wolf DC, Aronow WS, Lanier GM. Diagnosis, Treatment, and Management of Orthotopic Liver Transplant Candidates with Portopulmonary Hypertension. *Cardiology in Review* 2018; 26 (4): 169-176

**Abstract:**

“Portopulmonary hypertension (POPH) is seen in 5–8% of orthotopic liver transplantation (OLT) candidates and has significant implications for clinical outcomes. POPH is characterized by vasoconstriction and remodeling of the pulmonary vasculature. It is exacerbated by the hyperdynamic circulation that is common in advanced liver disease. Screening all OLT candidates with transthoracic echocardiography to assess pulmonary pressures and right ventricular function is crucial, as clinical symptoms alone are not reliable. Any significant right ventricular dysfunction or dilatation along with an elevation in estimated pulmonary pressures usually triggers further investigation with right heart catheterization. The mainstays of therapy of POPH are vasodilators that are used in pulmonary arterial hypertension. They include monotherapy or combination therapy with prostanoids, endothelin receptor antagonists, and phosphodiesterase-5 inhibitors/guanylate cyclase stimulator. Limited evidence from smaller studies and case series suggests that a timely diagnosis of POPH and the early initiation of treatment improve patient out- comes, whether or not OLT is ultimately undertaken. Given the historically high perioperative mortality rate of more than 35%, POPH remains a contra- indication to OLT unless it is treated and responsive to vasodilator therapy. We review the current literature and International Liver Transplant Society practice guidelines (2016) for the latest in understanding POPH, its pathogenesis, diagnosis, modern pharmacological treatment, indications, and contraindications for OLT, as well as perioperative management.”

COMMENTS MADE BY CROUCH, CARA MD

**Summary:**

This article was selected from the July/August 2018 issue of *Cardiology in Review* because it serves as an excellent review of the management of portopulmonary hypertension (POPH) in patients who undergo liver transplantation. This article begins with a review of the definition, epidemiology and pathophysiology of POPH. The authors quote a prevalence rate of POPH in 5.3-8.5% of liver transplant candidates1. Given the relatively high mortality rate of these patients, the management of this diagnosis is always an important topic for us to review. With the routine use of transthoracic echocardiography screening of transplant candidates, these patients are often identified sooner and can begin treatment. The authors discuss how confounders such as high cardiac output and volume overload can appear similar to POPH when utilizing echocardiographic screening. The authors then review the different parameters that will differentiate these conditions when these patients undergo right heart catheterization, which is used for confirmation of the diagnosis.

The article also provides a review of current treatment options, including prostanoids, endothelin receptor antagonists, phosphodiesterase-5 inhibitors and combination therapy. The authors also present an algorithm for the evaluation of transplant candidacy in patients who may have POPH that is based on the 2016 consensus statement from the International Liver Transplant Society Practice Guidelines. This review concludes with recommendations for perioperative and postoperative management. Overall, this article does an excellent job reviewing the diagnosis and management of this complex diagnosis and offers several suggestions for the development of management guidelines.

**References:**

1. Koulava A, Sannani A, Levine A, Gupta CA, Khanal S, Frishman W, Bodin R, Wolf DC, Aronow WS, Lanier GM. Diagnosis, Treatment, and Management of Orthotopic Liver Transplant Candidates with Portopulmonary Hypertension. *Cardiology in Review* 2018; 26 (4): 169-176

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