A combination therapy of 3 vasodilators may treat portopulmonary hypertension

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Combination therapy of Prostacyclin, Sildenafil, and Bosentan helped a young male patient with severe portopulmonary hypertension improve enough to receive a liver transplant. It was also used post-transplant to help him maintain his health. These findings are published in the March issue of *Liver Transplantation*, a journal by John Wiley & Sons.

Portopulmonary hypertension (PPHTN) is pulmonary arterial hypertension associated with liver disease or portal hypertension. Our knowledge about treatment options for this condition is limited by lack of randomized controlled trials, concerns about drug safety and the inability to reverse pulmonary disease. Liver transplant is an effective treatment for many patients with mild-to-moderate PPHTN characterized by a mean pulmonary artery pressure (mPAP) 50mmHg).

Researchers led by Mark Austin of King's College Hospital describe a case study of a 26-year-old male patient who had severe refractory PPHTN and non-cirrhotic portal hypertension. He was treated with sequential, then combination, therapy of the vasodilators Iloprost, Sildenafil, and Bosentan. His condition improved dramatically, with his mPAP falling from 70 to 32mmHg. He underwent liver transplantation, and continued on the therapy for two years post-transplant, when his mPAP fell even further to 28mmHg. He currently continues on dual combination therapy of Sildenafil and Bosentan, and his doctors hope to eventually withdraw both agents.

"This is the first reported patient with severe PPHTN using this

combination of vasodilator therapy as a bridge to liver transplantation and then as maintenance in the post-transplant phase. This regimen may enable liver transplantation in similar patients in the future, without long-term consequences," the authors conclude.

An accompanying editorial by Karen Swanson of the Mayo Clinic writes, "It is easy to see why combination therapy may make sense as we do not yet have a means of determining which predominant mechanism is influencing which patient and, which medications might be most beneficial specifically in POPH."

She laments the fact that POPH is excluded from every clinical trial involving pulmonary hypertension, especially given its prevalence, and worries about the cost of triple-drug therapy when we don't even have good prospective data on individual drug therapy.

"Only by following multi-center prospective protocols in POPH will we ever have more meaningful answers to the ideal course of treatment in these patients," she concludes. "Until then, we continue to extrapolate our thoughts from single center case reports and case series. Can we move on?"

Source: John Wiley & Sons