Medical management of porto-pulmonary hypertension and right heart failure prior to living-related liver transplantation.

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Porto-pulmonary hypertension and right heart failure are relatively prevalent complications of end-stage liver disease and may increase mortality of patients undergoing cadaveric orthotopic liver transplantation. Even with extensive pre-transplant evaluation, these complications are frequently diagnosed unexpectedly in the operating room and transplant procedure may need to be aborted due to high perioperative mortality from both right and left ventricular failure. Living-related liver transplantation is a new surgical alternative to cadaveric liver transplantation, but presence of porto-pulmonary hypertension increases its postoperative mortality as well. Due to inherent elective nature, however, living-related liver transplantation may allow for preoperative hemodynamic optimization and treatment of right ventricular failure. To the authors' knowledge, this is the first reported case of an adult patient with porto-pulmonary hypertension who underwent successful living-related liver transplantation. Favorable transplantation outcome was obtained in this case through good hemodynamic control with long-term IV epoprostenol therapy, preoperative right heart calcification, and perioperative administration of pulmonary vasodilators and inotropic agents.

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