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Commentary: Repair or replace—Potts shunt versus lung transplantation for refractory pediatric pulmonary hypertension

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Like killing a cylinder, Potts shunts restore balance while avoiding the underlying issue.

CENTRAL MESSAGE

Potts shunts equilibrate right and left ventricular pressures to improve the functional status in patients with refractory pulmonary hypertension, although long-term results remain largely unknown.

Patients presenting with medically refractory pulmonary hypertension are traditionally listed for lung transplantation. Historically, the availability of suitable donor lungs, particularly for pediatric patients, led to long waitlist times and increased morbidity and mortality. Lancaster and colleagues[1] review their success using Potts shunts as an alternative to lung transplantation in this population over the past 6 years, discuss key learning points, and demonstrate their technique for a valved Potts shunt.

Engines today are commonly built in a V-configuration (eg, V4, V6, V8), wherein an equal number of cylinders are present on either side of an engine block, orientated away from one another like a V. The configuration decreases the length of the engine block relative to an inline configuration, although it requires all cylinders to fire symmetrically for the engine to remain balanced. If a cylinder misfires, or becomes asymmetric, there is a reduction in power, increased noise, and a shutter due to the lack of balance within the engine. If left untreated, this can progress to engine failure. In a majority of cases, this issue can be fixed by replacing a spark plug or gasket. Although in other cases, the cylinder itself is warped and car owners are faced with the decision of repairing the engine block or replacing the engine entirely.

In a similar fashion, patients with refractory pulmonary hypertension develop cardiac dysfunction and activity restriction that can carry fatal long-term consequences unless managed through a Potts shunt or lung transplantation as described by Lancaster and colleagues[1] in this issue of the *Journal*. In their retrospective study, the authors compare outcomes for patients historically undergoing lung

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transplantation and those more recently receiving Potts shunts at their institution. Patients receiving Potts shunts experienced shorter hospitalizations, fewer complications, and similar survival without the need for immunosuppression when compared with those undergoing lung transplantation. At their latest follow-up of 1.8 years, patients with Potts shunts experienced a decrease in right-sided systolic pressures to systemic levels with an associated reduction in brain natriuretic peptide levels signaling decreased right heart strain. From a functional standpoint, 100% of patients were limited by their condition preoperatively, whereas only 17% of patients experienced limitations postoperatively. Although it is tempting to view Potts shunt as a repair for refractory pulmonary hypertension, with advantages to lung transplantation or a replace strategy, the long-term outcomes remain unknown.

Patients treated with Potts shunt in this study are being committed to Eisenmenger physiology with progressive underlying lung disease that will likely require future transplantation. Although the patient's functional status improves after Potts shunt, the right ventricle continues to have systemic pressures. Early patients in the series are anchored to systemic pressures given a bidirectional shunt, whereas later patients with a valved conduit in theory have the potential to develop sub-systemic right-sided pressures. Although Eisenmenger physiology is tolerated early in life, it continues to carry a 12- to 18-fold increase in early mortality with more than one-third of those dying of heart failure or sudden cardiac death.[2-4] Furthermore, Potts shunts only reduced the pulmonary arterial pressure by 10 mm Hg, which is not likely to affect the rate of pulmonary dysfunction and subsequent need for transplantation in this cohort. Transplantation after Potts shunt is rare and carries a significant perioperative risk for those undergoing the procedure. Although the authors report best practices for mitigating this future risk, it begs to question whether or not up-front lung transplantation may be safer, particularly in a modern era with lung allocation scores, improved waitlist mortality, and similar outcomes for risk-stratified patients after primary and redo transplantation.[5,6]

Potts shunt in the setting of pulmonary hypertension is not a repair or replace strategy, it is a palliative procedure. It is akin to disconnecting the fuel injector from both the misfiring cylinder and the opposite cylinder on the block. The engine is once again balanced and the shaking has stopped, but the check engine light continues to be on. It serves as a reminder that despite sounding better or feeling better in the case of Potts shunt, the underlying condition has not changed and the risk of future complications continues to exist.

CONG

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