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REPLY: SALVAGING A PATIENT WITH A FAILING BIDIRECTIONAL CAVOPULMONARY SHUNT REQUIRES CAREFUL ANALYSIS AND TIMELY CORRECTIVE ACTION

Reply to the Editor:

The bidirectional cavopulmonary shunt (BCPS) is the cornerstone of the staged palliation of a functional single ventricle. When a BCPS "fails," the causation may be anatomic, physiologic, or a combination of both. Anatomic abnormalities that are associated with hypoxemia following a properly constructed BCPS could be related to stenosis, distortion, or hypoplasia of one or both pulmonary arteries. Physiologic causes of BCPS failure include increased pulmonary vascular resistance (which can be due to reversible or permanent pulmonary pathology), pulmonary venous obstruction, atrioventricular valve regurgitation, and ventricular dysfunction.

The selection of patients for BCPS palliation is being continually refined in the modern era. Luo and colleagues[1] reported a retrospective analysis of a group of patients in which BCPS failure was predicted by low preoperative superior vena cava (SVC) blood flow detected by cardiac magnetic resonance imaging (CMRI). In their model, low preoperative CMRI-derived SVC blood flow is probably a surrogate for ventricular dysfunction, systemic atrioventricular valve regurgitation, or maldistribution of systemic blood flow.

Turkoz and Dogan[2] successfully salvaged 2 patients suffering from hypoxemia immediately following a BCPS procedure. In both cases, a systemic to pulmonary artery shunt was added to a BCPS in the setting of documented acute elevation of the pulmonary artery pressure. To prevent SVC syndrome, the authors banded the proximal shunted pulmonary artery. The technique used by Turkoz and Dogan can salvage a difficult situation, thereby creating reasonable palliation at the cost of proximal pulmonary artery stenosis. We do not know the long-term outcomes for their patients. Ideally, anatomic abnormalities are corrected, not created, at the time of the BCPS connection.

Luo and colleagues' report demonstrates that sophisticated preoperative analyses might not always predict ideal outcomes. The 65 consecutive patients they selected were deemed to have appropriate anatomy and physiology for a BCPS. Yet, 7 patients had BCPS takedown, with 5 late deaths. In addition, 4 other patients died with an intact BCPS. Eleven BCPS failures in 65 patients who had been carefully selected for BCPS indicates that the process of preoperative analysis requires additional refinement. In contradistinction to the report of Turkoz and Dogan, none of Luo and colleagues' patients were immediate/intraoperative failures. Whether any of these patients were candidates for the addition of accessory pulmonary blood flow (APBF) is unclear from their report. Adding APBF can salvage a failing BCPS, but it is unknown whether the addition of APBF makes the patient a better long-term candidate for the Fontan–Kreutzer circulation.[3,4] Some patients may do well for a prolonged period after palliation with a combination of BCPS and APBF. When the cause of acute hypoxemia following BCPS is due to reversible pulmonary pathophysiology, adding APBF may not only be lifesaving, but also relatively harmless. Long-term follow-up will ultimately determine whether adding APBF to a failing BCPS has a negative impact on the early, mid-term, or late Fontan–Kreutzer outcome. In the meantime, further analyses are needed to determine how low SVC blood flow at CMRI reveals a propensity for BCPS failure.[5]

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