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Letters to the Editor

The Author Replies

J. N. Graziano

Division of Pediatric Cardiology, University of Michigan, L1242 Women's, Box 0204 1500 East Medical Center Drive, Ann Arbor, MI 48109-0204, USA

Dr. Chintala appropriately emphasizes that data regarding the survival of infants with hypoplastic left heart syndrome (HLHS) and restrictive atrial septal defects (rASDs) following transplantation are quite limited. The author is also correct in emphasizing that the "pulmonary venous hypertension and pulmonary vascular changes associated with rASD are long-standing, occurring during the intrauterine period, and therefore may not be influenced by postnatal intervention." These changes and their effect on pulmonary vascular resistance are the basis for concern regarding cardiac transplantation and potential right ventricular failure postoperatively in these patients.

Addonizio and colleagues [1] described patients with elevated pulmonary vascular resistance indicies to be at higher risk for right ventricular failure and death than patients with normal pulmonary vascular resistance. Additionally, their study suggested that the responsiveness to preoperative pulmonary vasodilators may be a more useful prognostic indicator for survival; this was later corroborated by Gajarski et al. [2]. Unfortunately, such hemodynamic data regarding pulmonary vascular relaxation to vasodilators in patients with HLHS and rASD are lacking. Furthermore, Rychik et al. [4] proposed that patients with HLHS and rASD may have a labile pulmonary vasculature with episodes of pulmonary vasospasm based on findings in similar animal models of in utero pulmonary venous hypertension. This lability may not be clearly evident by conventional hemodynamic evaluation. The precise impact of such lability on posttransplantation right ventricular function is also not known.

Clearly, more data are needed to better understand the influence of a rASD on the outcome of neonatal transplantation in patients with HLHS. However, the limited availability of donor hearts and the increased theoretical risk of posttransplant right ventricular failure based on pathologic analyses of the pulmonary vasculature in these patients [3, 4], and their potential physiologic implications as described previously, mandate careful consideration of each patient's risk of transplantation when rationing these valuable, limited organs.

References

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