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REPLY: NONELECTIVE CORRECTION OF PATIENTS WITH COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT FAILING MEDICAL MANAGEMENT IS A VIABLE OPTION EVEN IN VERY YOUNG INFANTS

Reply to the Editor:

I thank Miller and coauthors for their comments regarding our recently published paper.[1] The congenital cardiac community has continuously searched for the optimal surgical timing and treatment for complete atrioventricular septal defect (CAVSD). Since the regionalization of pediatric cardiac surgery in Sweden almost 3 decades ago, we have used elective surgical correction at 3 to 4 months of age as our clinical standard.[2] However, in patients who, despite optimized medical treatment, develop congestive heart failure, we have performed nonelective corrective surgery even before 3 months of age and avoided palliative measures.

In the young infants operated before 3 months (mean age, 1.6 months $\pm$ 0.6 months; median age, 2 months; interquartile range, 1 month), we found a greater disease burden and more complex atrioventricular valve disease. This group was prone to developing more preoperative severe atrioventricular regurgitation/heart failure, including the need for care in the intensive care unit, making nonelective early surgery the only viable option.

Concomitant coarctation of the aorta has preferably been managed by coarctation repair from the side, in the neonatal period, but in most patients without concomitant pulmonary artery banding to avoid pulmonary artery distortion. One patient in the present cohort was corrected with concomitant arch repair due to a severely hypoplastic arch needing selective cerebral perfusion during arch repair.

In the present patient cohort, the variable pulmonary atresia describes the whole obstructive spectrum from stenosis to atresia. The patient who needed a shunt neonatally was corrected with a concomitant conduit placement, whereas the other patients were corrected without the use of a conduit.

The discussion regarding durability of the left atrioventricular valve (LAVV) after early correction <3 months of age is important. In our study, 4 patients were reoperated within 30 days of CAVSD repair. Three of these patients (2 patients needed early LAVV re-repair; 1 patient had a closure of residual VSD) had their initial correction <3 months of age, showing the potential complexity of corrective surgery in the very young infant. However, we could not identify any significant difference in rate of LAVV-related reoperation between the groups.

"Why should we wait?" and "Do we need to operate now?" are pertinent questions. Surgical complexity and the fragility of atrioventricular valve tissue are still of utmost concern. In the majority of patients with CAVSD who are stable on medical management, surgical correction seems to be optimal at 3 to 4 months of age. Having followed our surgical strategy for the last quarter of a century, correction in the very first months of life is necessitated only in pharmacologically refractory congestive heart failure. However, when this situation appears, we believe our data support that primary correction is a valid option, and preferable, to palliative procedures.

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