Neonates and isomerism: Are the rules different?

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Bhaskar and colleagues¹ report long-term outcomes for 182 patients with isomerism. Survival at 18 years was 74% and 50% for left atrial isomerism and right atrial isomerism (RAI), respectively. Obstructive pulmonary venous drainage was a significant risk factor for mortality in RAI (odds ratio, 4.2). Dextrocardia was an independent risk factor for mortality in left atrial isomerism (hazard ratio, 3.0).

In contrast to trends for other complex diagnoses in the congenital cardiac population, survival did not improve over eras. Implicit is that strategies successful for other diagnoses did not work for isomerism. The Kaplan-Meier survival curves shed light on why this might be. There is a disproportionately high mortality in early infancy (particularly for RAI) with a subsequent flattening of the curve to resemble the survival slope for other complex congenital cardiac diseases. One explanation could be that despite appropriate anatomic surgical intervention, the physiologic stress is simply less well tolerated in subsets of neonates/early infancy patients with isomerism. In attempts to lessen this stress, less-invasive catheter-based interventions or hybrid surgical intervention techniques, such as relief of obstructed pulmonary venous return, for RAI in neonates/infants have been reported.^{2,3} These novel temporizing strategies can postpone the timing of more definitive palliation or repair until such a time that physiologic tolerance is greater. Further exploration of less-invasive initial interventions appears to be warranted.

It is known that pushing the limits of complex biventricular repair can yield a higher early mortality rate than single-ventricle palliation.^{4,5} Although the outcomes for biventricular repair of RAI in the study by Bhaskar and colleagues¹ were disappointing, there are small cohort reports of success in the literature.^{6,7} The mixed findings simply indicate that the ability to achieve an appropriate anatomic biventricular repair must also be

wedded to an ability to tolerate the physiologic stress associated with repair. Interestingly, patients with RAI, who underwent a successful biventricular repair, tended to have it performed after the early infancy window—a time when the tolerance of patients with isomerism for physiologic stress seems to be much more analogous to other complex congenital cardiac diseases.

Survival outcomes for atrial isomerism, particularly RAI, have not yet reached what has become the standard for other complex problems. This long-term study by Bhaskar and colleagues¹ points to where we as a professional community need to find improved solutions. The focus needs to be on understanding the mechanisms of and improving the physiologic intolerance of procedures in neonates and infants.

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