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Commentary: Down or non-Down: It all comes down to the details

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Many studies have found that patients with trisomy 21 (T21) tend to have better outcomes after atrioventricular septal defect (AVSD) repair than those without T21.¹⁻³ This comes to no surprise to congenital heart surgeons, who in general find working with atrioventricular valves more challenging in patients without T21.

In this issue of the *Journal*, Ramgren and colleagues⁴ report on 62 nonsyndromic patients who underwent complex or noncomplex AVSD repair at their institution. The authors included mostly patients with complete AVSD, although 19 (28%) patients had a restrictive ventricular septal defect, consistent with a transitional AVSD. The authors found that nonsyndromic patients without complex cardiac anatomy had short- and long-term outcomes comparable with those previously reported for patients with T21 but that mortality was significantly greater in those with complex anatomy. The authors concluded that anatomical factors were more important in predicting prognosis than the mere presence or absence of T21. The study specifically excluded patients with T21, thus making it impossible to have a direct comparison of outcomes.

The study demonstrates that good perioperative outcomes can be achieved in nonsyndromic patients.⁴ However, the incidence of reintervention is high. The

Atrioventricular septal defect in a nonsyndromic child with significant regurgitation.

CENTRAL MESSAGE

Good perioperative outcomes can be achieved in nonsyndromic patients with atrioventricular septal defect, but reintervention rates are high.

10-year incidence of reoperation in this cohort was almost 30%, regardless of anatomy. This is greater than the 9% 10-year reoperation rate we reported after repair of noncomplex AVSD (20% after complex AVSD) in a combined cohort where the majority of patients had T21.⁵ Some of these differences could potentially be explained by anatomic differences on the left atrioventricular valve that may make repair more challenging and reintervention more frequent (Figure 1). For example, in the current study with nonsyndromic patients, 20% to 25% of them (depending on complexity) had a deficient mural leaflet and 7% to 13% had a single papillary muscle.⁴ In our study with combined T21 and non-T21 patients, only 5% to 11% had a deficient mural leaflet and 2% to 4% a single papillary muscle.⁵

As suggested by Ramgren and colleagues,⁴ it is time to reframe the discussion surrounding AVSDs by placing more emphasis on the effect that anatomic complexity, rather than genetics alone, has on outcomes. Only by developing a good understanding of the anatomic nuances that impact outcomes in these patients will we be able to develop strategies to improve those outcomes. After all, it all comes down to the details.

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FIGURE 1. Four-chamber color-compare view of a transesophageal echocardiogram of a nonsyndromic infant with an atrioventricular septal defect demonstrating a dysplastic left atrioventricular valve with significant regurgitation.

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