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Commentary: Regionalization refines surgical dogma regarding trisomy 21 and atrioventricular septal defect

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It is common surgical “dogma” that nonsyndromic patients with a complete atrioventricular septal defect (AVSD) have a poorer prognosis than patients with trisomy 21. This is thought to be based on the observation that patients with trisomy 21 tend to have exuberant and excessive AV-valve tissue versus nonsyndromic patients who tend to have a dearth of AV valve tissue. An analysis of these patients by the group from Lund, Sweden, has helped to clarify and refine this dogma.¹

I want to begin by noting that I have enjoyed reviewing the surgical outcomes of this truly regionalized congenital heart center. In 1992 Sweden (population of 10 million) centralized congenital heart surgery from four centers to two.² The outcome of patients operated on at one of the centers (Lund) has been remarkable. They recently reported 300 patients with an AVSD with an operative mortality of 1% and freedom from left AV valve related reoperation of 93% at 20 years.³ These are some of the finest results ever reported for an AVSD repair, and I believe are attributable to in some measure to their regionalization of care.

This most recent subanalysis by the Lund group reviewed 62 nonsyndromic patients who underwent complete AVSD repair. One-fourth of the patients were labeled as complex having concomitant cardiac anomalies including tetralogy of Fallot, double-outlet right ventricle, coarctation, and additional ventricular septal defects. The analysis revealed that at 10 years nonsyndromic patients with noncomplex complete AVSD had a 100% survival and the patients

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CENTRAL MESSAGE

Regionalization of care for patients with atrioventricular septal defect (AVSD) has allowed this Swedish heart center to tease out the factors that complicate AVSD repair in nonsyndromic patients.

with complex lesions in contrast had only a 66% survival. Their analysis concluded that it is the combination of complex anatomy in a nonsyndromic patient where the poor outcomes are clustered. The surgical dogma that a poor prognosis and difficult surgical repair is associated with all patients with AVSD without trisomy 21 is not exactly correct. The nonsyndromic patients with complete AVSD and no other cardiac diagnosis (noncomplex patients) actually had very good outcomes. It is the complex nonsyndromic AVSD group that has the more difficult surgical repair and worse outcome. The regionalization of care in Sweden has allowed the surgeons to amass the number of patients in the various subgroups of AVSD to make this type of subanalysis possible. I look forward to the Lund, Sweden, group's analysis of other congenital cardiac lesions and the excellent outcomes that have occurred with their conscientious regionalization of care.

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References

1. Ramgren JJ, Zindovic I, Nozohoor S, Gustafsson R, Hakacova N, Sjögren J. Impact of concomitant complex cardiac anatomy in nonsyndromic patients with complete atrioventricular septal defect. *J Thorac Cardiovasc Surg*. 2022;163:1437-44.
2. Lundström NR, Berggren H, Björkhem G, Jögi P, Sunnegårdh J. Centralization of pediatric heart surgery in Sweden. *Pediatr Cardiol*. 2000;21:353-7.
3. Ramgren JJ, Nozohoor S, Zindovic I, Gustafsson R, Hakacova N, Sjögren J. Long-term outcome after early repair of complete atrioventricular septal defect in young infants. *J Thorac Cardiovasc Surg*. 2021;161:2145-53.