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Editorial

Truncus Arteriosus: An Updated Benchmark for Clinical Management of an Important Congenital Cardiac Malformation

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See article by Morgan et al., pages 446–452 of this issue.

In pediatric cardiac surgery, there are a host of variables that affect outcome. Those variables, both modifiable and not, change at different rates, so it is our practice, as a group, to re-examine a cohort of a particular diagnosis from time to time to understand what has improved, on the whole, and what management challenges remain. In this issue of the *Canadian Journal of Cardiology*, the authors of "Contemporary Outcomes and Factors Associated With Mortality After a Fetal or Postnatal Diagnosis of Common Arterial Trunk"[1] have given us this new "line in the sand" with their single-centre (Toronto Sick Children's Hospital), 25-year experience of 165 subjects with common arterial trunk (CAT), diagnosed either *in utero* or postnatally. They have shown us that patients with complex CAT continue to have significantly higher (although better over time) mortality risks vs simple CAT. They have also shown us that prenatal diagnosis is not associated with improved survival, that a small proportion of patients will receive a truncal valve repair during the index hospitalization, and that there is an ongoing hazard for reintervention, driven largely by the right ventricle (RV)-pulmonary artery (PA) conduit.

The series begins with fetal outcomes and follows the cohort of 153 patients through a median of 8.6 years. Of the total birth cohort, 12 fetuses did not survive, and 16 live-born infants died without undergoing surgery, with the latter group having a very high incidence of significant truncal valve dysfunction. Among patients who underwent surgery, the authors dichotomized the cohort by an era (before and after January 2000), based on an institutional change in the surgical approach, and secondarily dichotomized patients by the presence or absence of particular complicating factors (aortic arch obstruction, truncal valve dysfunction, and PA hypoplasia). Although overall operative mortality has improved significantly over time (36% in the first era to 7% since 2000), 1-year survival continues to be worse in the patients with "complex" CAT than in those with "simple" CAT. Long-term survival is relatively stable, and reintervention rates are high (85%) and reasonably consistent over time.

To put these findings in context, we will consider them one at a time.

Complex CAT Continues to Carry a Significant Operative Mortality Risk

Total CAT operative mortality is reported as 7% in the 2000-2014 era in this report, similar to the 2018 Society of Thoracic Surgery (STS) report for this STAT 4 category procedure. However, the early mortality rate of simple CAT in this series is 5%, whereas complicated CAT carries a 6.46 odds ratio risk of 1-year mortality. Russell et al.[2] reported the STS Congenital Heart Surgery Database cohort of CAT with an in-hospital mortality of 11% for the entire cohort 2000-2009, significantly higher for the patients requiring aortic arch repair ± truncal valve repair, in whom early mortality was 18/61 (30%). In the STS cohort, patients with "complex" CAT only accounted for 61/572 (10.2%) of the entire surgical CAT population, whereas in the Toronto series, "complex" CAT (defined as significant pathology of the aortic arch/truncal valve and/or branch PA) comprised 50% of the cohort.[1] This incidence gap may lie in the difference of definition of "complex" CAT, which in the Toronto series includes PA hypoplasia, a variable that is not included in the Russell et al. definition of complex CAT.

The amount that can be said definitively about the prenatal course of patients with CAT remains limited. As a result of not knowing the true denominator (ie, the number of fetuses with the diagnosis of CAT that were spontaneously or therapeutically terminated), the course of a gestation with CAT cannot be precisely defined. That being said, the number of fetal diagnoses of CAT has increased to a higher proportion of those patients with a fetal diagnosis of any congenital heart anomaly. In contrast to the hope that higher rates of prenatal

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diagnosis and enhanced perinatal care would be associated with improved outcomes, fetal diagnosis was not associated with improved postnatal survival. Prenatal diagnosis was in fact associated with younger gestational age at birth, lower birth weight, and increased odds of preoperative death, leading the authors to conclude that "considering the low risk of spontaneous *in utero* demise, most fetuses with CAT will not benefit from earlier induction of labor or delivery." This is similar to the findings with prenatal diagnosis of obstructed total anomalous pulmonary venous connection[3] and raises questions about why prenatal diagnosis is not making the predicted positive impact on outcome. A possible explanation is that early outcome is most impacted by the dysfunction of the truncal valve, the incidence of which will not be influenced by prenatal diagnosis, and for which there are few satisfactory solutions.

A consistently small proportion of the truncus cohort will require early truncal valve repair/replacement, and this remains a high-risk cohort. This is in contradistinction to some reports indicating that they have reduced the additional risk associated with truncal valve dysfunction.[4,5] The STS cohort reported 29 of 572 patients (5.1%) who required 33 truncal valve operations during the index admission. None of the patients who had truncal valve replacements at the time of reoperation survived, and the overall in-hospital mortality for any patient requiring truncal valve repair or replacement (regardless of aortic arch intervention) was 12/29 (41%).[2] The risk of having both truncal valve and aortic arch intervention was shown to be incremental, with an operative mortality in this group of 60%. This is also reported in the Congenital Heart Surgeons Society cohort of truncus arteriosus with interrupted aortic arch, in which the early mortality is 34/50 (68%).[6] Perhaps the biggest question arising from reflecting on truncal valve function and outcome is what precisely the threshold for surgical intervention is, as this appears to be different among series and very likely is different between individual surgeons. Ten percent of the preoperative deaths in this series were associated with either significant truncal valve stenosis or regurgitation. Although moderate-to-severe truncal valve regurgitation or stenosis was diagnosed in 61 patients (34 patients in era 1 and 27 patients in era 2), truncal valves were repaired or replaced in only 17 patients (13% of the surgical cohort). Until there are better options for truncal valve repair or replacement, the threshold for intervening will likely remain high, with the surgeon often opting to leave a truncal valve that is felt to be adequate (even if not optimal), with hopes that the patient will survive to an age/size where there are more valve replacement/repair options.

Finally, there is an ongoing hazard for reintervention. As for any pathology that requires an RV-PA conduit or valve replacement, reoperations and catheter-based reinterventions will be required. Seventy-six of 93 one-year survivors (82%) required at least 1 reintervention including truncal valve surgery (either primary or repeat, 19%), RV-PA conduit replacement (57%), pulmonary valve replacement (8%), and catheter interventions for conduit/PA stenosis (73%, which accounted for 121/224 reinterventions).[1] Clearly, the dominant driver for reintervention is the RV-PA conduit.

What Does This Series Add to Our Understanding of Truncus Arteriosus?

A contemporary single-centre report that is pathology/diagnosis based such as this one provides the important denominator information that is lacking in procedure-based reports. The report is a "real world" view of how patients with CAT fare in the current era and will be valuable to family and medical personnel alike, to appreciate what the likelihood of survival and reintervention for patients with CAT in general, but the report also strongly emphasizes the very different outlook for complex vs simple CAT variants. In contrast to prior reports, the authors chose to include PA hypoplasia in their definition of complex lesions, a diagnosis for which the definition is not clearly delineated in the article. To support the use of PA hypoplasia in the definition of complex CAT, the authors reference the finding of Brown et al.[7] that discontinuous PAs (in association with a group of cardiac pathologies that included but was not limited to CAT) were associated with increased mortality risk. This may be a defensible definition of "complex," given that although the main early outcomes relate to the mortality risk increment added by neonatal aortic arch reconstruction and/or truncal valve repair/replacement, the late management issues are mainly related to the status of the truncal valve and the RV-PA conduit. It will be a significant help to the literature of CAT to have an empirically derived, standardized definition of what constitutes complex CAT.

The fact that the authors chose to use 30-day mortality rather than the STS definition of operative mortality creates a challenge to understanding and comparing early outcomes in this series with that in other series. Besides mortality and reintervention, there is no new information regarding the complication burden that patients bear in the acute phase. The authors suggest that the Contegra valved conduit, used in era 2, was associated with fewer interventions, but there is no report of the incidence of bacterial endocarditis, a significant concern with this conduit.[8] Likewise, an accounting of complications with important long-term sequelae for patients and their families (such as neurologic complications) is notably absent, and will no doubt be included in the next update of this and other cohorts.

Conclusions

The authors have shown a significant improvement in mortality over time for all cases, but patients with simple CAT lesions continue to have significantly better 1-year survival than patients with complex CAT lesions. As we continue to uncover preventable or modifiable risk factors, routinely collect and report complication outcomes, and develop better options for valve repairs, valve and conduit replacements, both large academic registry data collections and detailed institutional reports such as this one will continue to have important roles in informing practices that will positively impact outcome, for this and many of the congenital cardiac pathologies that we treat.

Disclosures

The authors have no conflicts of interest to disclose.

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