Commentary: Assessing risk factors after truncus arteriosus repair—The devil is in the details

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In the current issue of the Journal, Mastropietro and colleagues retrospectively reviewed the results of 216 children who had undergone surgical repair of truncus arteriosus at 15 institutions in the United States between 2009 and 2016. They focused on early outcomes and assessed risk factors associated with major adverse cardiac events (MACE), an outcome variable that combined in-hospital mortality with the requirement for postoperative cardiopulmonary resuscitation or extracorporeal membrane oxygenation support. They found that MACE occurred in 44 patients (20%), including 26 (12%) who received cardiopulmonary resuscitation, 21 (10%) who received extracorporeal membrane oxygenation support, and 15 (7%) who died during the index admission. They examined a fairly comprehensive list of demographic, anatomic, surgical, and clinical variables for potential association with MACE and identified the following 3 significant factors that predicted poor outcome: failure to diagnose truncus arteriosus before discharge from the nursery, cardiopulmonary bypass (CPB) duration longer than 150 minutes, and a right ventricle–to–pulmonary artery (RVPA) conduit diameter ratio greater than 50 mm/m². Mastropietro and colleagues concluded that those factors were potentially modifiable and that tackling them could subsequently lead to improved repair outcomes in this challenging population.

Surgical repair of truncus arteriosus remains challenging. Despite major improvements in the perioperative care of these patients, reported in-hospital death remains high (9%-11%). Although the report of Mastropietro and colleagues shows a slightly lower operative mortality, the limited number of experienced centers that have participated in this study may have influenced this finding. Moreover, despite the somewhat lower operative mortality risk, the study of Mastropietro and colleagues still shows significant postoperative morbidity. Multiple series have explored variables associated with poor outcomes; in most of these series, the risk factors were often demographic or anatomic variables that were not modifiable (eg, prematurity, low weight, genetic syndromes, preoperative shock, truncal valve regurgitation, interrupted aortic arch). Interestingly, those traditional variables were not found to be significant in this current study by Mastropietro and colleagues. The identification of any modifiable risk factor should naturally stimulate a change in the treatment approach that would alleviate that risk factor and consequently improve outcomes. The devil is usually in the details, however, and the answer of how to use new findings to alter the management strategy for the best is usually not as clear and instead is hidden in little specifics that are commonly not available to the readers. Although Mastropietro and colleagues have used a fairly comprehensive data set in this retrospective review, important information about the adequacy of repair, degree of preoperative illness, severity of complications, and mode of death are not well defined, and these deficits seem to limit our ability to use this study to modify the way that we treat our patients.

Mastropietro and colleagues identified failure to recognize patients born with truncus arteriosus before discharge from the nursery as a risk factor for MACE. In their series, 63% of patients had prenatal diagnosis; however, 21% did not have the condition diagnosed by the time that they were discharged from the nursery, with 6% having the diagnosis...
made longer than 30 days after birth. Given the established role of routine pulse oximetry screening in detecting undiagnosed congenital heart disease before discharge from the nursery, this seems to be a surprising high figure that may reflect lack of adherence to the routine screening policy, especially in the early experience. In that case, delayed diagnosis could potentially be a modifiable factor. Nonetheless, it is unclear from the study why delayed diagnosis was associated with MACE risk. It does not appear that those patients were more likely to have preoperative shock or poor clinical condition; it is also not clear whether pulmonary hypertension or poor diastolic function as a result of the late intervention played a role in the occurrence of those adverse events. To develop a proper clinical judgment that would improve outcomes of those patients, it would be more helpful to learn the characteristics of those who had MACE develop and also to discover more details surrounding the adverse events. The one clear conclusion from the study is that patients with unrepaired truncus arteriosus have an unstable physiology that warrants repair after birth, and that delays in the treatment of those patients, whether intentional or unintentional, would be associated with significant preoperative morbidity and potential mortality.

Mastropietro and colleagues’ also found a diameter ratio of the RVPA conduit greater than 50 mm/m² to be associated with MACE and believe that this is a modifiable factor. To address this issue, however, more details are again needed for a better understanding of the type of interaction between larger conduits and adverse events. On the one hand, larger conduits might be associated with technical complications, such as distortion of the branch pulmonary arteries leading to stenosis, and they might also be more prone to compression by the sternum, with subsequent development of obstruction or regurgitation. In that case, surgical modifications, which might or might not involve placement of a smaller RVPA conduit, could be needed to guarantee a surgical repair with minimal residual lesions. On the other hand, a diameter ratio of the RVPA conduit greater than 50 mm/m² might be inevitable in smaller patients; for example, a 2.5-kg neonate will very likely have a body surface area smaller than 0.18 m², and any conduit equal or larger than 9 mm in diameter (the vast majority of cases) would therefore be associated with a diameter ratio of the RVPA conduit greater than 50 mm/m². The conduit diameter ratio risk factor thus might be a surrogate for patients with very small weight and associated prematurity, genetic anomalies, and extracardiac problems—factors that are hardly modifiable and at the same time are well associated with prolonged recovery, postoperative complications, and in-hospital death. Inarguably, meticulous surgery with minimal residual lesions and vigilant perioperative care are essential elements for successful outcomes after truncus arteriosus repair in general, and in high-risk patients in particular. The idea that significant improvement in the outlook of those patients could be achieved by choosing smaller conduits seems to be an oversimplification of a complicated problem.

Finally, CPB duration longer than 150 minutes was associated with MACE, and Mastropietro and colleagues argue that this is potentially a modifiable factor. The effect of CPB on postoperative recovery and possibly survival is well established, and many of the recent developments in the field of perfusion have focused on mitigating CPB effects to enhance recovery and decrease morbidity. Limiting CPB duration is certainly one way to do so, although the value of this action in the current era of improved perfusion technology may be debatable. Although CPB duration can be surgeon dependent, in most cases, excessive variations in CPB duration is related to the necessity of performing additional work, revisions of residual lesions, or resuscitation of a dysfunctional myocardium. I believe that adequacy of care, rather than CPB duration, is a more important factor in the development of MACE. There are surgeons and institutions that are currently achieving the best surgical results despite long CPB durations, and in those cases it seems that improved perfusion strategy, meticulous perioperative care, and excellent repairs have all mitigated any effect of prolonged CPB on outcomes. I therefore believe that CPB as a risk factor is in fact a proxy for intraoperative complications, revisions, or residual lesions, and in my mind, addressing those should be the priority of the surgeons to achieve superior results.

In summary, there continues to be a room for improvement in the management outcomes of children born with truncus arteriosus. Numerous single-institution and multi-institution studies have examined the outcomes of truncus arteriosus repair; however, these studies are commonly limited by the lack of data granularity and of important fine details that would enable us to generate knowledge that could lead to better outcomes. A carefully developed, prospectively collected multi-institutional database might be necessary if we are to make noticeable progress in the management of this challenging disease.

References


