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EDITORIAL COMMENTARY

Does chance truly favor the prepared mind?

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See related article on pages 1516-22.

As mortality statistics for complex congenital heart surgery drift asymptotically toward zero, significant perioperative morbidities have taken a more prominent position as a target of focus for quality improvement. Indeed, the long-term consequences of a perioperative major adverse cardiac event (MACE) can sully a sense of triumph with a technically excellent repair, and heightened awareness of “bad actors” among the *dramatis personae* may help to identify those at risk and thereby reduce the failure-to-rescue rate. In this issue of the *Journal*, Hornik and colleagues¹ present an observational study of the Society of Thoracic Surgeons database, which contains the largest cohort to date of patients with Williams-Bueren syndrome undergoing cardiovascular surgery. Their results (that the

entire cohort had a MACE incidence of 9%, whereas those with combined left ventricular outflow tract (LVOT) and right ventricular outflow tract (RVOT) repair had a MACE incidence of 21%) apply sobering statistics to what perhaps has been an assumed truth among congenital surgeons; namely, that from initial anesthetic induction to ultimate hospital discharge, children with Williams syndrome are at high risk for an adverse cardiac event.

These demographic data demonstrate an overall mortality of 5% but a range of MACE outcomes, depending upon the type of procedure performed (isolated RVOT, 0%; complex LVOT, 9%; and combined LVOT and RVOT, 21%), age, the presence of other preoperative risk factors, and the need for coronary artery repair. As a large retrospective database study, more subtle cardiac morbidities were not discernable for better characterization of these events (eg, inotrope score, evidence of ischemia). Moreover, 46 of the patients (9%) were excluded because of missing information regarding postoperative complications, and it is not possible to determine from the Society of Thoracic Surgeons database whether the coronary interventions were due to intraoperative complications or primary coronary anatomy.

These shortcomings become so pronounced, however, largely because the compelling nature of the data leaves

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the surgeon wanting more detail. As a prognostic tool, the data would suggest that the more “complex” the case (eg, RVOT and LVOT vs isolated RVOT repair), the more likely the chance of a MACE. Although the trend was not statistically significant, the data also suggest that the higher the complexity, the higher the mortality (RVOT and LVOT, 6.9%; complex LVOT, 5.3%; isolated LVOT, 1.6%; and isolated RVOT, 0%). A “simple” explanation based solely on inadequate or altered coronary perfusion, individual preoperative risk factors, or additional operative procedures, however, does not fully explain all these findings.

Naturally, the diagnosis of “Williams syndrome” in the database may represent a spectrum of disease, and it is unclear whether these findings may be applicable to other patients undergoing RVOT and LVOT repair for presumed vasculopathy. Indeed, the findings may raise more questions than they answer. Do these results reflect a different disease pathophysiology or severity to account for the difference in MACE and mortality for certain patients with Williams syndrome? Does this severity of disease confound the effect of smaller size? Can emergency mechanical support mitigate mortality in this setting?

What is clear is that this population may represent some of our highest risk candidates for congenital cardiac surgery, surely rivaling the most complex cases of single-ventricle anatomy. The overall in-hospital mortality of the Williams cohort was 5%, and their rate of rescue

was notably low (29%-44%)—not to mention that it is uncertain for any given patient what long-term significance a MACE will have, itself complicating a major cardiovascular procedure being performed on a patient who may have considerable cognitive challenges at baseline. Certainly, because none of the variables analyzed are largely modifiable, the greatest value of the study may be the prognostic data it provides for preoperative discussions with patients and their families, as well as for planning postoperative surveillance and the possibility of emergency intervention.

The burden of these findings may then reiterate a truism: one must be “on one’s game” when managing the postoperative patient with Williams syndrome. The next level of inquiry perhaps requires a more granular prospective analysis through mechanisms such as the Congenital Heart Surgeons’ Society, in which actual operative and perioperative practice variation may disclose superior management strategies. Whether chance will favor the prepared mind in this arena remains to be seen, but a heightened awareness of those most at risk for a postoperative MACE is an important first step.

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