ventilator dependence. Preoperative airway evaluation is essential, and the authors advocate for a multidisciplinary team approach, including consultation with palliative care. Postoperative pulmonary complications should be monitored closely along with a higher expectancy for reintubation or tracheostomy before discharge. At our center, we currently require a preoperative airway evaluation and assent for postoperative tracheostomy (in the event it is

needed) before embarking on heart surgery in children with T13/18. This study appears to support these practices.

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Commentary: Cardiac surgery in children with trisomy 13 and trisomy 18: “Is it the quality of life or the quantity?”

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It is now well established that children with trisomy 13 and 18 have a very short life expectancy, with a median survival of less than 2 weeks.¹ However, for those children who survive past 6 months, 50% to 60% may survive 10 years.¹ In translocations that result in partial trisomy or mosaics, improved survival up to 27 years has been reported.² Some parents choose to terminate pregnancy, whereas others proceed to make decisions based on their beliefs, understanding, and physician recommendations. Physicians too are often divided regarding treatment of these children, as some feel that aggressive treatments are futile whereas others defer to the parents' wishes.

Recent advances in surgical technique and postoperative intensive care unit (ICU) management have resulted in

The quality of life outweighs the quantity.

CENTRAL MESSAGE

Children with trisomy 13 and trisomy 18 with congenital heart disease can undergo corrective surgical repair with reasonable hospital survival but questionable medium-term survival.

significantly improved hospital mortality.³ Centers have therefore extended cardiac surgery to greater-risk populations with genetic syndromes including trisomy 13 and 18.³ A study conducted by the Pediatric Cardiac Care Consortium found that patients who required prolonged preoperative ventilator support had worse outcomes.⁴ For patients who were ventilator dependent for more than 2 days preoperatively, only 33% were extubated before discharge, and 11% died. They demonstrated a 91% survival and concluded that most patients with trisomy 13 and 18 can survive cardiac surgery.⁴ A study from the Society of Thoracic Surgeons database confirmed that the hospital mortality was reasonable except for children who required preoperative mechanical ventilation, who had an 8 times greater hospital mortality.⁵

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Swanson and colleagues⁶ evaluated their experience over 20 years, which included 14 children with trisomy 13 and 18 who underwent cardiac surgery. The authors found that although the hospital mortality was low, the children had longer postoperative mechanical ventilation times and longer length of stay in the ICU and hospital. One of the 4 children who was on mechanical ventilation preoperatively died. Three additional children died within 1 year following discharge. Although this study was underpowered, with incomplete and short follow-up, the authors did have a well-matched comparison group, and they did demonstrate excellent hospital survival, which is testament to their surgical expertise and perioperative ICU management.

Therefore, based on the current, albeit sparse, data, it seems evident that despite the poor long-term survival, low-moderate risk cardiac surgery can be offered to children with trisomy 13 and 18 with a reasonable hospital mortality, provided they are not on a mechanical ventilator preoperatively. It is specifically when caring for chronically ventilated children with trisomy 13 and 18 with congenital heart disease that ethical challenges are encountered and opinions between providers and parents may differ.

In conclusion, these children should be offered surgical repair when indicated as part of the informed consent process. It is important to identify the mosaic and partial trisomy variants, as these sub-types have a better life expectancy. However, for those unfortunate children who remain chronically ventilator dependent, it is reasonable to offer palliative care, keeping in mind that what most parents want for their child is the quality of life, not the quantity.

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