Delivery of Anesthesia for Children with Trisomy 18 (Edward's Syndrome): A Review of 80 Anesthetics

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Introduction

Trisomy 18 or Edwards syndrome is the second most common autosomal anomaly and multiple malformation syndrome (1) with a prevalence between 1/3,000 and 1/10,000 live births (2). The syndrome encompasses malformations of the central nervous, cardiac, respiratory, gastrointestinal, and genitourinary systems. Cardiac defects are present in >95% of patients (1). Historically, only 50% of patients survive beyond one week and 90% of patients do not survive beyond one year of age (2). Common outcomes for children that survive beyond the neonatal period include severe developmental delay, inability to communicate, G-tube dependence, and chronic lung disease. There is some indication that more aggressive supportive care has increased survival beyond infancy(2). This means that anesthesia providers may have more frequent exposure to this patient population in the future. We performed a comprehensive review of all patients with Trisomy 18 who were treated at our institution in the past five years.

Methods

After obtaining IRB approval, we performed a retrospective chart review of all Trisomy 18 patients treated at Children's Hospital of Wisconsin between 2012 and 2017. For patients born before 2012 all prior anesthesia records were also included in the analysis. Anesthesia records were searched for comorbidities, ASA status, code status, types of procedures performed, airway exam, airway management, perioperative complications, and causes of death. Timing of surgical procedures was analyzed with respect to timing of other procedures, involvement of palliative care, and patient death.

Results

A total of twelve Trisomy 18 patients were treated at our institution between 2012 and 2017. The date of birth ranged from 2001 to 2016. Nine patients required anesthesia on 80 occasions for 115 different procedures. Comorbidities are displayed in Figure 1. Eight of the surgical patients were considered "full code." One patient was considered "do-not-resuscitate", however was made "full code" for surgery. Eleven patients were classified as ASA 4, and one patient was ASA 3. Figure 3 shows the timing of the procedures for each patient relative to patient age. Four patients died during the study period. Palliative care was involved starting in early infancy in all patients born after 2012.

Serious perioperative complications were reported for 28 anesthetics, most involving difficult airway management (n=25), at times requiring fiberoptic intubation or tracheostomy (n=6). Five instances of cardiopulmonary resuscitation were required in four patients. In all cases cardiac arrest was preceded by loss of airway patency/ ventilation, either during induction or after extubation in the early postoperative period. Three patients were marked "difficult access."

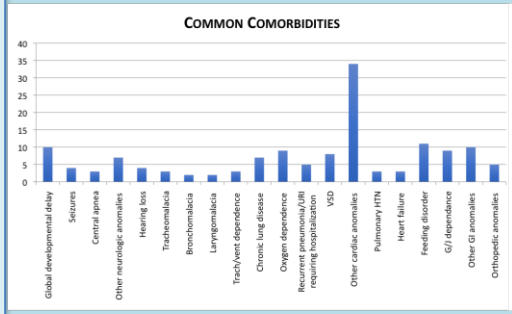

Figure 1: Summary of comorbidities for all 12 patients.

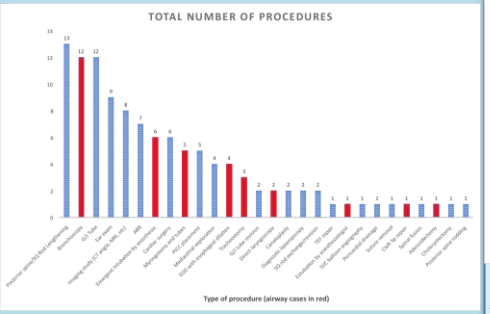

Figure 2: Total number of procedures performed in 12 patients. Procedures Involving the airway are highlighted in red.

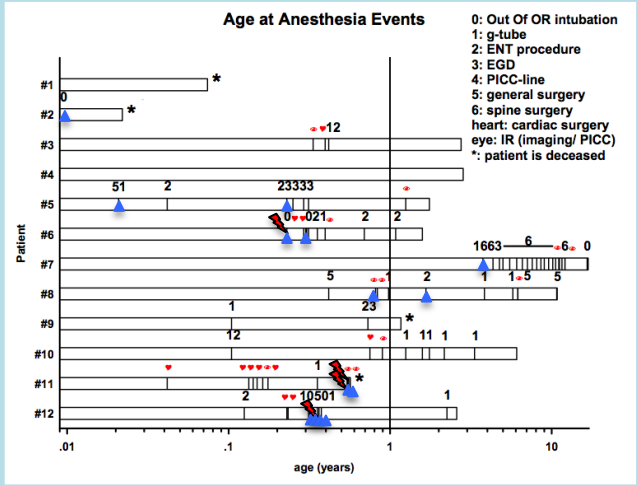

Figure 3: Timing and types of procedure for each patient. Three patients did not undergo any surgeries (#1, #2, #4). Patient #2 was ultimately intubated by ENT. Airway evaluations and g-tube placement were generally performed within the first months of life. Five patients underwent heart surgery within the first months of life. Patients living beyond infancy continued to require surgical procedures and anesthesia care. Four patients required anesthesia assistance with intubation on the wards, most during infancy.
Please note logarithmic scale for patient age.
Flash: Cardiopulmonary resuscitation.
Blue triangle: Difficult intubation.

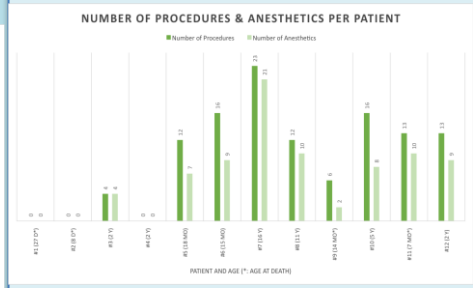

Figure 4: Three patients did not undergo any procedures. In most patients, more than one procedure was performed in at least some of the anesthetics. #: patient number, matches Figure 3. Patient age is per March 2017 or age at death (*).

Conclusion

This review of our institutional experience represents the largest series of anesthetics performed in Trisomy 18 patients to date. Anesthesia care was required for a significant number of airway emergencies, surgical and imaging procedures. We found that airway management and respiratory compromise were the overarching concerns during the perioperative period, All of our patients had an abnormal external airway exam. While published case reports describe that airways were secured with relative ease (3), we found that inability to ventilate was a significant risk factor and led to perioperative hypoxic cardiac arrest in five cases. Even in patients where the airway was successfully secured, this often required additional resources such as glidescopes and fiberoptic intubation, and in some cases a surgical airway. Growth did not necessarily improve the airway anatomy, requiring repeated fiberoptic intubations in two and tracheostomies in three patients.

This information is highly relevant for anesthesiologists, parents and other care providers to better weigh the risks and benefits of each procedure, in particular in light of the patient's individual prognosis. Since the most critical part of the anesthetic appears to be the induction, combining multiple procedures during one anesthetic should always be considered.

References

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