To Extubate or Not to Extubate: Ethical Implications in a Rare Genetic Disorder with Craniofacial Abnormalities

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Goals

1. Identify the characteristic features of Trisomy 14 Syndrome
2. Understand the role of the anesthesiologist in extraoperative airway management
3. Identify resources and alternatives when conflicts of interest arise
4. Appreciate different perspectives when approaching an ethical dilemma

Case

A 3 year-old male with bilateral clubbed feet presents for a routine scheduled heel cord release procedure. On review of the patient’s chart, it is revealed that the child has a history significant for Trisomy 14, developmental delays, feeding difficulties, and reactive airways disease. He displays microcephaly, micrognathia and dysmorphic facies on physical examination. Further investigation yields a cardiac history involving cor triatriatum and total anomalous pulmonary venous return with patent foramen ovale, repaired in infancy.

General anesthesia is induced with Sevoflurane. Intravenous access proves to be difficult. After injection of muscle relaxant, laryngoscopy reveals a grade IV Cormack and Lehane score. Fortunately, the airway is secured with a blind intubation technique. Anesthesia is maintained with Sevoflurane, delivered with air and oxygen. Once the procedure is completed, spontaneous ventilation resumes. At emergence, the endotracheal tube is removed. However, the patient can not be mask-ventilated. Laryngospasm is quickly diagnosed and succinylcholine is administered. Mask ventilation is still unsuccessful.
Another attempt at blind intubation fortuitously establishes a secure airway. Otorhinolaryngology is consulted for further airway management and a tracheostomy is their ultimate recommendation. At this time, the patient’s mother is updated as to her child’s condition. She intimates concern over such invasive interventions and is considering assigning the child a DNR status. The patient is then taken intubated to the pediatric ICU. Three days later, the PICU attending is uncomfortable extubating the patient and has advised the primary orthopedic service that the patient be extubated in the OR by an anesthesiologist. When faced with this request, what is your next move?

Questions

1. What physical findings comprise the Trisomy 14 phenotype? What are the major anesthetic challenges in this population?
2. What is the life expectancy of these individuals and what concerns might quality-of-life issues present for the parents?
3. If a DNI order is instated, when would extubation be appropriate?
4. Is another visit to the operating room a viable place for extubation? Is the PACU? What venue would be most comfortable for you?
5. Pressure is on from the ICU to extubate; ENT does not want to participate except to be called for difficult reintubation; the mother’s wishes for advance directive are unclear; what are your options?
6. When and how would it be appropriate to approach a hospital’s ethics committee? What is the protocol for doing so?

Discussion

Mosaic trisomy 14 syndrome is a rare genetic disorder, of which only 18 cases have been described in the literature outside of anesthesia. Growth and psychomotor retardation, micrognathia with short neck, dysplastic ears and broad nose, cryptorchidism and congenital heart disease are present in more than 90% of reported cases. Less frequent findings include: frontal bossing, ocular hypertelorism, narrow palpebral fissure, cleft palate, body asymmetry, and abnormal skin pigmentation. Epidemiological gender ratio is 3 female: 2 male and the oldest surviving patient known is 29 years of age. The most important anesthetic challenges in this population are difficult airway and cardiac abnormalities including TOF, ASD/VSD, and PDA.

In the case of this young boy, the mother is confronted with difficult quality of life decisions. If she has not had sufficient time for discussion and contemplation with the medical care team, family, and others involved to make sound conclusions, extubation would be inappropriate as this may force reintubation and/or the child’s demise. If the final decision is made to not attempt resuscitation/intubation of the child and this is legally documented, the patient may remain in the PICU for extubation as no further action is required. However, if the mother decides against a DNR status, you must choose the most appropriate venue for extubation. While the most comfortable location
for us as anesthesiologists in conducting airway management may be the operating room, that is not viable without a surgical procedure. Hospital policy does not allow one to be both anesthesia provider and proceduralist. Certainly we would like to have alternate airway equipment close at hand, such as LMA, fiberoptic or glidescope, emergent tracheostomy kit, and various blades and endotracheal tubes. The availability of auxiliary personnel would also be prudent. Whether the PACU or PICU is a more comfortable place may be a more individual preference.

In the extraoperative setting, anesthesiology performs the role of a consulting service and is not obligated to adhere to the demands of another department or staff member. However, it is our professional duty to provide the best assistance possible. If the patient’s mother (i.e. primary caregiver or proxy) has not yet defined a clear advance directive, more time for that decision should be requested. If an inordinate amount of time has passed and a decision has not been reached or there are disparate wishes among those involved, a hospital ethics committee may indeed be the next resource of choice. These panels typically consist of appointed physicians, nurses, attorneys and clergy. A consultation by the committee may be requested by health care workers or family members. The structure and function of each hospital’s committee may differ somewhat but the general intent is to assist in resolving complex ethical ambiguities or disagreements by providing a forum for discussion and professional guidance. Most committees are available 24hrs/7 days a week and are open to informal discussion or to address specific questions. In situations where legal proxy decisions are not deemed to be in the best interest of the patient, they may provide direction in taking legal actions as first-line patient advocates.

References


