

## CASE AND COMMENTARY: PEER-REVIEWED ARTICLE

### How Should Refusal of Tracheostomy as Part of an Adolescent's Perioperative Planned Intubation Be Regarded?

Case and Commentary by Katherine Gentry, MD, MA and Aaron Wightman, MD, MA

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#### Abstract

Here we present a case of a patient in terminal respiratory failure refusing to consent to emergent tracheostomy in the setting of an anticipated difficult intubation. We examine ethical concerns that arise from deviations from the standard of care in the operative setting and the anesthesiologist's sense of culpability. Finally, we will review the ethical arguments and guidelines that support anesthesiologists' participation in palliative operative procedures when limitations on resuscitation are in place.

#### Case

Kelly is a 16-year-old girl with spinal muscular atrophy type 1. Her weakness made it difficult to perform needed pulmonary clearance treatments, leading her to develop pneumonia and progressive air hunger that required her to be hospitalized. After discussion with her pulmonologist, she and her parents requested intubation to facilitate aggressive attempts at improving pulmonary toilet. If these attempts proved to be unsuccessful, Kelly's parents, with her assent, requested that the endotracheal tube and ventilator be discontinued and that goals of care be shifted to focus on comfort only.

As Kelly's weakness and contractures prevented her from fully opening her mouth, oral intubation was predicted to be difficult; therefore, anesthesiology and otolaryngology services were consulted to consider performing a fiberoptic nasal intubation in the operating room. During discussion of the plan, the anesthesiologist explained that in the case of a failed intubation attempt, her next step would be to secure the airway surgically (ie, via a tracheostomy). This troubled Kelly and her family, as they considered life with a tracheostomy to be an unacceptable outcome. Over the years, they had consistently refused tracheostomy and chronic ventilation as a potentially life-sustaining treatment. For Kelly, a life worth living included retaining some ability to speak. Given her

degree of weakness, a tracheostomy would render her unable to vocalize, and she would require mechanical ventilation without respite.<sup>1,2</sup>

Kelly's acceptance of short-term intubation but refusal of an emergency tracheostomy was difficult for the anesthesiologist to accept. She rejected the notion that a tracheostomy could be refused in the setting of an operating room intubation, as the provision of anesthesia could directly precipitate respiratory insufficiency, and a tracheostomy could immediately treat this iatrogenic complication. Prohibiting a tracheostomy would limit the anesthesiologist's ability to secure Kelly's airway successfully. Furthermore, Kelly could die of respiratory failure in the operating room if the intubation attempts were unsuccessful. The anesthesiologist expressed moral concerns that she had a duty to rescue a patient under her care and that by honoring the patient's wish for no tracheostomy she could be playing a role in "killing" or "euthanizing" the patient.

### **Commentary**

Spinal muscular atrophy (SMA) is considered the most common lethal disease of children younger than 2 years of age in the United States.<sup>2</sup> SMA results in weakness and wasting of voluntary muscles due to degeneration of anterior horn cells. Intellect is normal and sensation is intact.<sup>2</sup> In SMA1, also known as Werdnig-Hoffman disease, symptoms of hypotonia and diffuse motor weakness present before the age of 6 months; children with SMA1 typically are never able to sit without support.<sup>3</sup> Most children with SMA1 die by the age of 2 due to respiratory failure.<sup>3</sup> Survival rates for children with SMA1 have improved for patients born after 1994, likely due to increased use of noninvasive ventilation, invasive ventilation, feeding via gastrostomy, and nutritional supplementation.<sup>4</sup> The recent introduction of nusinersen can significantly improve functional status and survival in SMA patients who receive this therapy.<sup>5,6</sup> Nevertheless, for Kelly to have survived to the age of 16 without the need for chronic invasive ventilation is quite unusual.

It is generally accepted that parents or guardians of children with SMA1 may refuse tracheostomy. In a 2012 multinational survey of pediatric pulmonologists and intensivists, 95% felt that parents should be able to refuse tracheostomy in children with SMA1.<sup>7</sup> In the same survey, 78% felt that intubation and ventilation would be acceptable in the setting of acute respiratory failure, but only 60% felt that it would be an acceptable therapy for chronic respiratory failure.<sup>7</sup> In Kelly's case, intubation was intended to be a short-term intervention to enable her to recover from an acute pulmonary infection and thus would likely be viewed as appropriate by many physicians, based upon the responses in the survey cited above.

Anesthesiologists are responsible for the "support of life functions under the stress of anesthetic, surgical, obstetrical and radiological manipulations."<sup>8</sup> This support is

necessary for the safe provision of anesthesia (eg, securing the airway) and for mitigating the undesirable effects or complications of anesthesia and surgery (eg, administering fluids to treat hypotension). In Kelly's case, the administration of sedatives for intubation could cause respiratory depression and airway obstruction; if face mask ventilation and intubation both became impossible, a tracheostomy would be necessary to prevent death. Thus, Kelly and her family's request constrains the usual practice for difficult airway management and could result in Kelly's death under the anesthesiologist's care.

### **Ethical Analysis**

The ethical rationale for allowing patients to request limitations on resuscitation is respect for autonomy—individuals' "right to hold views, make choices, and to take actions based upon their values and beliefs."<sup>9</sup> This respect is granted to adults in part because they are presumed to have the capacity to understand the decision at hand and to freely accept or reject proposed treatment options. Adolescents, on the other hand, are not considered to be fully autonomous due to a lack of decision-making capacity. In general, the assent of adolescents is sought, but their parents have ultimate authority to make medical decisions.<sup>10</sup> However, several empirical studies have demonstrated that adolescents do not differ from adults in their ability to make rational health care decisions.<sup>11-13</sup> Additionally, it has been suggested that the experience of chronic illness furthers the development of decision-making capacity in adolescents.<sup>14</sup> Capacity assessments employ a series of questions that probe patients' understanding, their ability to express a choice, their appreciation of their particular situation, and their reasoning.<sup>15</sup> In Kelly's case, given her long-standing experience of SMA and consistently demonstrated preferences, we suspect that she has capacity to make the decision to refuse tracheostomy. For this reason, Kelly's assent or refusal should be taken seriously by her parents, as they provide the ultimate authorization for her medical care.

The issue of [adolescent refusal of life-saving treatment](#) remains controversial.<sup>16</sup> Adolescents might focus on short-term outcomes and be overly influenced by socioemotional concerns.<sup>16</sup> Ross has proposed that agreement between adolescent and parent preferences may justify refusal of experimental or low-efficacy treatments.<sup>17</sup> In this case, Kelly's views are concordant with those of her parents. The tracheostomy can be considered a low-efficacy treatment given that it would not alter her prognosis and in light of her goals of retaining the ability to speak and survive independently of long-term mechanical ventilation. Given Kelly's apparent capacity, the concordance between her wishes and those of her parents, and the understanding that a tracheostomy would not change her prognosis, we argue that Kelly's preferences should be respected.

The anesthesiologist's moral quandary stems from concerns about deviating from the standard of care and her perceived potential culpability in a patient's death. Some physicians have argued that placing [limitations on resuscitation in the operating room](#)

demands that anesthesiologists act as if “one hand [were] tied behind the[ir] back” and are “an unreasonable intrusion and distortion of practices that form the very core” of their professional identity.<sup>18</sup> From the perspective of the anesthesiologist in Kelly’s case, intubation and tracheostomy are bundled because the intubation is predicted to be difficult, and the ultimate rescue maneuver for a failed intubation is an emergent tracheostomy.<sup>19</sup> Thus, she sees refraining from a tracheostomy to be an unacceptable deviation from the standard of care.

However, intubation and tracheostomy are distinct concepts for Kelly. The intubation is a temporary intervention aimed at helping her recover from pneumonia. She is willing to accept the incapacity associated with being intubated for a short time if it allows her to return to her prior level of functioning. After much consideration and with her parents’ support, Kelly has decided that life with a tracheostomy would be untenable. To her, life with a tracheostomy would be worse than death. The goals of care established by Kelly and her parents render the typical standard of care less pertinent.

A second concern the anesthesiologist expressed is her perceived potential culpability in a patient’s death. The active nature of care in the operating room has led some to argue that deaths that occur in the operating room are acts of commission, since anesthesia and surgery actively change the patient’s state and can often be said to be the proximate cause of death, while deaths that occur on the medical ward are perceived to be acts of omission, as the patient’s underlying disease is presumed to have prevailed.<sup>20</sup> When a patient dies in the operating room, anesthesiologists and surgeons are not asked, “What happened?” They are asked, “What did you do?”<sup>20</sup> While acts of commission and omission may feel emotionally different, it is generally accepted that there is no ethical distinction between the two; what makes either act—commission or omission—ethically justifiable are the physician’s obligations to the patient.<sup>9</sup> If there is no clear duty to provide an intervention (such as a tracheostomy), then withholding or withdrawing treatments could be permissible.<sup>9</sup> Therefore, key to allaying the anesthesiologist’s fears of culpability would be a clarification of her obligations to Kelly in the setting of this procedure.

Although adult patients have a legal right to refuse medical treatment, as established by the Patient Self-Determination Act of 1990,<sup>21</sup> questions about how to handle advance directives often come up in the setting of anesthesia and surgery. In a seminal paper on the topic, Robert Truog stated, “With the increasing recognition of the autonomy of the competent patient in medical decision-making, it would be inappropriate not to seek the patient’s guidance and provide as much latitude as possible within the constraints of the physician’s own ethical standards.”<sup>22</sup> Subsequently, the American Society of Anesthesiologists (ASA), the American College of Surgeons, and the American Academy of Pediatrics have recommended a process of “required reconsideration” of advance directives in the perioperative period.<sup>23-25</sup> This process entails a thoughtful discussion

involving the patient, family, and treating physicians to identify and develop plans to support the patient's goals of care while allowing the team enough latitude to perform the indicated procedures. If such a discussion occurred with Kelly and her family, it would likely become clear to the medical team that Kelly has the capacity to assent or dissent, and that she and her parents concur on the goals of care. Ideally, the team would develop a consensus about the method for the planned intubation, the limits framing the attempt, a range of back-up plans that exclude tracheostomy, and a communication strategy if all attempts are failing and Kelly's death is imminent. The team should be challenged to consider the entire range of management options available for this patient including a completely awake fiberoptic intubation with topical anesthesia, which carries a very low risk of airway compromise or death, or placement of a laryngeal mask airway if intubation is impossible in order to provide ventilation until sedation drugs wear off.

The anesthesiologist could also recommend that Kelly accept a tracheostomy for the short term and defer the decision to continue or withdraw respiratory support until she is back in the ICU supported by her parents. This alternative should only be offered alongside the option of no tracheostomy in order to allow Kelly and her family the opportunity to make a decision consistent with their goals of care.

### **Honoring Limitations on Resuscitation**

Anesthesiologists strive simultaneously to ensure patient comfort while maintaining normal circulation and respiration, and, in most crisis situations, acts of rescue to maintain life take precedence over comfort. However, for patients with terminal conditions who request limitations on resuscitation, the physician's obligations may shift towards ensuring comfort at the expense of sustaining life. Being asked to refrain from rescuing a patient is understandably difficult, but we would urge anesthesiologists to view this scenario similarly to situations in which terminally ill patients request discontinuation of life-sustaining treatments. Honoring an adult patient's or guardian's request for limitations on resuscitation in an operative setting supports the patient's right of self-determination and respects her ability to understand and consider the risks of mortality in a manner consistent with her version of the good. The cause underlying a need for resuscitation, physician induced or otherwise, might be irrelevant to the patient if the patient has considered the potential outcomes and explicitly stated a goal of care.

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**Katherine Gentry, MD, MA** is a bioethicist and pediatric anesthesiologist at the University of Washington School of Medicine and Seattle Children's Hospital in Seattle. Her research focuses on informed consent in the perioperative setting.

**Aaron Wightman, MD, MA** is a bioethicist and pediatric nephrologist at the University of Washington School of Medicine and Seattle Children's Hospital in Seattle. His research focuses on decision making for children with complex, chronic medical conditions.

#### **Citation**

*AMA J Ethics*. 2018;20(3):E683-689.

#### **DOI**

10.1001/amajethics.2018.683.

#### **Conflict of Interest Disclosure**

The author(s) had no conflicts of interest to disclose.

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ISSN 2376-6980**