Ethics: End-of-life decision-making in a pediatric patient with SMA type 2

The influence of the media

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ABSTRACT

Objective: Spinal muscular atrophy (SMA) is a group of progressive and fatal neurodegenerative disorders that are characterized by destruction of the anterior horn cells of the spinal cord. In this case report we outline the medical and ethical issues involved in a 7-year-old boy with SMA type 2 who experienced acute respiratory failure.

Methods: A review of the literature was conducted focusing particularly on the pathology, presentation, and outcomes of SMA and end-of-life decision-making in pediatrics.

Results: In a world where 40%–60% of deaths in pediatric intensive care units are a result of withdrawal or limitation of life-sustaining treatment, end-of-life decision-making has become an integral and difficult part of pediatric practice.

Conclusion: Limitation or withdrawal of life-sustaining treatment in a cognitively normal child with SMA poses a significant medical and ethical dilemma. This difficult decision is influenced by confluence of parental, doctor, social, cultural, moral, religious, legal, and economic factors and more recently the media. *Neurology*® 2012;78:e143–e145

GLOSSARY

SMA = spinal muscular atrophy.

In a general sense, worldwide mortality rates in children have fallen from 12.4 million per year in 1990 to 8.1 million per year in 2009.¹ The highest mortality rates continue to be in developing countries, where the etiology of child death remains predominantly due to poverty and lack of adequate medical care and education surrounding transmission of disease. This is in stark contrast to developed countries, where modern medicine has significantly improved survival. In fact, we have become so good at prolonging life, that up to 40%–60% of deaths in pediatric intensive care units are a result of the withdrawal of life-sustaining treatment.² Limitation or withdrawal of life-sustaining treatment in a cognitively normal child with spinal muscular atrophy (SMA) and its acknowledged respiratory complications poses a significant medical and ethical dilemma. In this case report, we outline the medical and ethical issues involved in a 7-year-old boy who experienced acute respiratory failure.

SMA II CASE A 7-year-old boy with SMA type II presented with a 10-day history of progressive cough, shortness of breath, lethargy, and feeding intolerance, complicated by desaturation and hypercarbia during sleep. This was on a background history of profound weakness of both upper and lower extremities and nocturnal hypoventilation requiring BiPAP (noninvasive ventilation). After admission, he became increasingly tachypneic, hypopneic, and bradycardic, then unresponsive. Following bag mask resuscitation, on BiPAP he continued to be in respiratory distress; however, he was alert, orientated, and conversing at an age-appropriate level. Progressive deterioration over a period of hours with increasing drowsiness required endotracheal intubation and mechanical ventilation. Intubation was extremely difficult due to a grade IV airway and multiple unsuccessful attempts at intubation were performed prior to securing the airway with a glidescope. It was recognized at this point that future emergency airway access would be a challenge and tracheostomy the only long-term solution. His parents saw his ability to express himself and communicate as the source of his quality of life and, understanding that worsening respiratory compromise would lead eventually to respiratory failure and death, opted to forgo tracheos-

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tomy insertion. While they utilized social media for health care information, they made this decision with full disclosure by the physicians and kept the decision private.

DISCUSSION This case highlights the ethically loaded question of when does treatment become excessive or even futile? SMA is a group of progressive neurodegenerative disorders that are characterized by destruction of the anterior horn cells of the spinal cord leading to skeletal muscle atrophy and weakness.^{3,4} It is autosomal recessive with 95% of children demonstrating a defect in the spinal motor neuron 1 (*SMN1*) gene.³ The disease manifests with symmetric proximal muscle weakness and wasting, tongue fasciculation, and absent deep tendon reflexes. Respiratory compromise is caused by very weak intercostal muscles, with a relatively strong diaphragm, leading to impaired cough, poor secretion clearance, hypoventilation during sleep, and underdevelopment of the chest wall and lungs. Recurrent infections exacerbate the weakness and further damage the lungs, leading to eventual respiratory failure and death. Sensation remains intact and in the majority of cases children have normal intelligence and cognitive function. SMA is classified into 4 types.³ SMA type I is the most severe, with death normally occurring within 2 years. SMA type II is the intermediate form, with death commonly in adolescence. SMA types III and IV are the mildest forms of the disease, with death occurring in adulthood. With advances in medical care, the life expectancy for SMA has improved significantly; however, more than 70% of patients with SMA type I + II are always dependent on others for self care.⁴ In SMA type I, continuous noninvasive ventilation can lead to survival >5 years and tracheostomy to up to 20 years.⁵ Ultimately, however, patients may become locked in on a ventilator.^{4,5}

Life-prolonging medical interventions, including mechanical ventilation, have allowed medical practitioners to keep children alive who in the past would have certainly died. However, with this great benefit has come the widespread desire to maintain life at all costs. But at what point does this desire extend beyond benefit to the patient and even do harm or prolonging suffering? In a survey of 7 hospitals in the United States, 85% of critical care physicians stated, "Sometimes I feel we are saving children who should not be saved."⁶ But who decides when to withdraw medical treatment in a severely ill or disabled child?

In medicine, autonomy—the right to accept or decline treatment—is determined by capacity. In Canada, under a physician's judgment, any child with full understanding and comprehension can de-

termine his or her own medical treatment. However, the majority of patients about whom the decision to limit or withhold life-sustaining treatment are made are unable to consent for themselves. This leaves the difficult decision in the hands of the parents and doctors of these children.

The factors associated with the decision to limit or withdraw life-sustaining treatment are many and individual to each case. The decision is made through a confluence of parental, doctor, social, cultural, moral, religious, legal, and economic factors, and more recently, the increasing role of the media. This article focuses specifically on the role of the media in end-of-life decision-making.

The media, particularly social media, has become a massive cornerstone in the shaping of today's culture and public opinion. Examples of this are President Obama's successful election as President in 2008; the wave of revolution in the Middle East in spring 2011; and identification of faceless rioting hockey fans in Vancouver in June 2011. It is therefore no surprise that the media has begun to influence the practice of medicine. As early as the 1970s, medicine and media collided with the infamous case of Karen Ann Quinlan,⁷ a previously healthy 21-year-old woman who was left in a permanent vegetative state after taking diazepam and drinking recreationally with friends. Her parents, based on her previously stated wishes and prognosis, wanted to withdraw life support; however, her doctors refused. The case made international headlines and it became the first widely publicized debate on the sanctity of life vs the right to die. The parents lost the first trial, and had to appeal to the New Jersey Supreme Court to win the right to let their daughter die, invoking the first media frenzy for this previously private and sensitive decision. Conversely, mass media campaigns have affected smoking cessation and improved road safety.⁸

In a world where up to 60% of adults use the Internet per day, and an unknown, but substantial number of children surf the Web, Facebook and other forms of social media are becoming forefront in the meshing of the medical world and the media.⁹ In 2010, 89 million adults used social media resources—predominantly blogs, chat rooms, online committees, and personal testimonies—for health-related purposes.¹⁰ Free access to information has educated and empowered patients; however, the limited control of accuracy or validity can make the Internet a misleading and a hazardous resource.

Furthermore, with free access to personal information, Facebook has ethical dilemmas of its own. To quote Erin Martz from the American Counseling Association: "Facebook can be quite dangerous."¹¹ In the case of Baby Joseph with Leigh syndrome, a pro-

gressive and fatal neurodegenerative syndrome, all physicians involved with his care at the London Ontario Health Science Center, supported by pediatric specialists in Canada, the United States, and Europe, agreed that a life-prolonging tracheostomy should not be performed.¹¹ The parents were unsatisfied with this decision and took the case into the media. Once again, the sensitive debate on end-of-life care was challenged and thrown into the spotlight. It quickly went “viral” on the Internet, including the “Save Baby Joseph” Facebook page with over 12,000 members. The turmoil was such that the hospital heightened security. In the end, with the help of an American priest, Baby Joseph was airlifted to the United States for a tracheostomy. While being an excellent resource for expression of personal opinion and support, Facebook and other unregulated social media sites lack the medical and economic resources required to make a fully informed decision of this magnitude. For example, it is terrible to consider cost-benefit in the context of a child’s life; however, it is the reality. The cost of home ventilation for children with SMA type 1 is at least \$100,000 USD per year. With 80% of the world population lacking essential medication and care, there is a point where medical interventions not only become burdensome for the patient and family, but for society.¹²

End-of-life decision-making in pediatrics has become an essential although difficult and sometimes controversial feature of practice and SMA is just one of many conditions where parents and doctors are faced with the toughest of decisions: when should life-saving measures cease?

AUTHOR CONTRIBUTIONS

Madeline Drake: conducted research, wrote the paper. Peter Cox: editor and intellectual contributor.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. **Go to Neurology.org for full disclosures.**

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