Obstructive Sleep Apnea in a 17-Year-Old With Profound Cognitive Impairment

For children with cognitive impairments who develop obesity and sleep apnea, the treatment options are not good. Adenotonsillectomy sometimes improves symptoms, but if it does not, then home continuous positive airway pressure (CPAP) is the only option. However, CPAP requires the patient’s understanding and cooperation. If the patient does not have the cognitive capacity to understand the need for CPAP, difficult ethical questions arise. We present a case in which decisions had to be made for such a patient, with commentary by 2 experts in pediatric pulmonary medicine. Dennis Rosen is an Instructor in Medicine at the Harvard Medical School and Associate Medical Director of the Sleep Laboratory at Children’s Hospital Boston. Ben Wilfond is a pediatric pulmonologist and Director of the Treuman Katz Center for Pediatric Bioethics at Seattle Children’s Hospital.

CASE REPORT: PART I

J.R., a 17-year-old girl with profound cognitive impairment secondary to hypoxic encephalopathy in infancy, was referred to the pediatric sleep clinic because of snoring, choking, and witnessed apnea during sleep. She had undergone adenotonsillectomy (AT) 4 years earlier because of similar symptoms and had improved. However, in the 2 years preceding her clinic visit, her symptoms had reappeared and gotten progressively worse. On examination, she was microcephalic, she had a short neck, her BMI was 46.6, and she was uncooperative and combative during the exam.

J.R. is nonverbal and gets the attention of family members by grunting toward objects of interest. She has had spastic quadriplegia since birth and moves around the house on her knees when out of her wheelchair. Behaviorally, she often has temper tantrums that consist of furious spells of kicking and head banging. She has frequent minor injuries from these tantrums. She is on risperidone and fluoxetine to “calm her.” She eats by mouth, with occasional episodes of choking. She has never had aspiration pneumonia. Her mother views eating as one of the few pleasures in her life and has not been willing to limit oral intake.

A sleep study demonstrated severe obstructive sleep apnea (OSA), with 82 obstructive apneas and 95 obstructive hypopneas, giving an apnea hypopnea index of 26 obstructions per hour. The obstructive events lasted 9 to 102 seconds and were frequently associated with desaturation into the 70% range, to a nadir of 63%. The obstructive events were seen in all sleep stages and positions.
**CPAP titration was attempted on 2 separate occasions, but the patient became combative and immediately removed the CPAP apparatus. The mother returned with her daughter to the clinic to discuss further treatment options.**

**Question: What Would You Recommend as the Treatment of Choice for J.R.?**

**Dennis Rosen Writes**

OSA is present in 2% to 3% of the general pediatric population,1 with a much higher prevalence in children with craniofacial abnormalities and central hypotonia. Its causes are multifactorial and include adenotonsillar hypertrophy, soft tissue hypertrophy, inflammation, and swelling; craniofacial structure; baseline muscle tone; overweight and obesity; and abnormalities in autonomic responses to changes in blood gas tension.3

In the general population, OSA is associated with hypertension4; metabolic syndrome5; excessive daytime sleepiness6; and cerebrovascular7 and cardiovascular8 disease, including pulmonary hypertension.9 Specifically in children and adolescents, OSA causes short- and long-term cognitive deficits,10 behavioral disturbances,11 symptoms of attention deficit and hyperactivity,12 and poor school performance.13 Given this patient’s profound developmental delay at baseline, it is hard to know how significantly she was being affected in these areas by her OSA.

AT is usually the first line of therapy in children with OSA. CPAP is generally the treatment of choice for OSA in adults and in children in whom AT is contra-indicated or was unsuccessful. In this child, CPAP would almost certainly require the use of restraints to prevent the removal of the mask, which would require her being restrained for several hours each day. Such restraint, in turn, would likely cause her a great deal of distress, which would be heightened by her inability to understand why this was being done.

When CPAP cannot be used, supplemental oxygen may be beneficial. Although intermittent hypoxia is not the only cause of negative outcomes of OSA in children (recurrent arousal and sleep fragmentation play an important role as well),14 attenuating or eliminating the recurrent desaturation could provide at least partial treatment and protect against the development of pulmonary hypertension. However, the patient’s intolerance of anything on her face as seen with the CPAP makes delivering supplemental oxygen by nasal cannula or face mask unlikely to succeed.

Tracheostomy is the definitive cure for OSA. However, patients with self-injurious behavior such as this patient are at risk for yanking out a tracheostomy tube and causing themselves severe harm.

Given all these complexities, I would recommend a trial of supplemental oxygen at home and then reassessment of the patient and family in a few weeks.

**Benjamin Wilfond Writes**

Treatment of J.R.’s severe OSA is important because of the risk of secondary pulmonary hypertension and heart failure. Some of her behavioral problems may also be exacerbated by her chronically disrupted sleep. It will be important to convey these assessments to the family and ascertain their concurrence with a goal of care to avoid heart failure and death.

The direct approach is to improve her sleep-disordered breathing with CPAP. Trying a variety of CPAP masks at home during the day may help J.R. become accustomed to using a mask. Applying the mask to other family members or favorite stuffed animals may also help J.R. become more tolerant. The CPAP itself could be initiated at home as well, before evaluation in the sleep laboratory. Alternatively, if this fails, supplemental oxygen may be better tolerated.

Although the supplemental oxygen may not improve her ventilation, it may reduce cardiopulmonary complications. The family could get her accustomed to wearing a nasal cannula and then have her return to the sleep laboratory to assess the impact of supplemental oxygen. She should also be monitored with electrolytes and capillary blood gases to assess her ventilation and with an echocardiogram to estimate her pulmonary artery pressure and right ventricular size.

J.R.’s behavioral issues are not simply an obstacle to her OSA treatment. Rather, the efforts to address her behavior contribute to her OSA. The risperidone and fluoxetine may be contributing factors, and her BMI is most certainly a contributing factor. However, simplistic suggestions to stop the medications and place her on a diet are not likely to work. The key approach will be to develop a therapeutic alliance with the family and explore together how to allow J.R. to maintain a good quality of life while consuming fewer calories and to explore alternative approaches to managing her behavior. These management strategies will not be simple and will depend on the family’s willingness to try new approaches. The family may benefit from the involvement of people with special expertise in behavioral and eating issues for children with profound disabilities. For example, the effective approaches developed for children with Prader-Willi syndrome,15 whose obsessive eating traditionally resulted in severe OSA, illustrate that it is not always necessary to make trade-offs between physical health and quality of life.16

**CASE REPORT: PART II**

Supplemental oxygen was attempted at home. As feared, the patient was unable to tolerate either a mask or nasal cannula. The mother was concerned
with subjecting her daughter to the discomfort of major surgery, especially as an improved outcome could not be guaranteed. The mother found the idea of using restraints for several hours a day in perpetuity to be intolerable.

After discussing the various options and their risks, as well as the risks from untreated OSA, the mother decided to forgo all of the treatment options described above. Instead, she wanted to shift to a palliative mode of treatment. She understood that it might lead to an earlier death for her daughter than would more aggressive approaches to treatment, but she thought that to be appropriate given the options.

**Question: Is Palliative Care an Acceptable Treatment Choice for J.R. at This Point?**

Benjamin Wilfond Writes

This parental request is ethically problematic. The mother now indicates that her primary goal is to maintain J.R.’s quality of life by feeding her even if it means that her life will be shorter. We must ask the following: is it ethically appropriate to allow parents of children with profound disabilities to make medical decisions that would be generally considered ethically unacceptable in a child without profound disabilities?

The case of Philip Becker illustrates our social ambivalence about this question. Philip was a 10 year old with Down syndrome who lived a facility for disabled children. His physicians advocated on his behalf in court after his parents refused surgery for his ventricular septal defect in 1977. His parents did not believe he would have an acceptable quality of life if he was to outlive them. The court upheld the parents’ right to refuse surgery. Next, a couple that volunteered in the facility and who had become emotionally attached to Philip successfully sued to be authorized to make his medical decisions.

The Becker case articulates some limits to parents’ authority to make decisions based on their concerns related to disability. Thus, there is a professional consensus that withdrawal of support can be justified at some threshold of disability when the quality of life is ambiguous. When facing a request by parents to not provide life-saving care for a child with a disability, the clinicians must ask whether the parents’ goals of care are reasonable, given the specific context.

Context does matter, and I would not consider palliative care an acceptable option for pulmonary hypertension secondary to obesity-related OSA, other than in the context of profound disability or imminent death. However, I would not agree to palliative care at this point for J.R., even though I agree that the family’s concerns about tracheotomy and long-term restraints are reasonable. My concern is that the family may be acting against J.R.’s interest by requesting palliative care because her obesity-related OSA might be reasonably treatable, even in the context of her disabilities.

At this point, further conversations with the family about their hopes and expectations for J.R. and the family are necessary. The providers and/or family should engage with palliative care and/or bioethics consultants as part of this assessment, as well as providers experienced with developmental issues, psychiatric and behavioral issues, and weight management. It sounds like J.R.’s care has taken a toll on this family, and I would look for additional ways to support this family who is doing their best to care for a child in challenging circumstances. I would actively explore weight loss programs and adjusting her psychoactive drugs and consider a short-term voluntary placement in either a foster home or a facility equipped to provide behavioral management and weight control for adolescents such as J.R. Placement of the child in such a program could give both providers and the family more data about the long-term benefits and burdens of these interventions.

I could imagine working hard to persuade the family to allow such measures and would anticipate some resistance. I would assure them that ultimately we would respect their wishes to not pursue a tracheotomy but that additional alternatives that could improve her quality of life need to be explored. I might point out that it may turn out to be either much easier or much harder than we imagine to address these issues, but we will not know until we try.

I would engage this discussion with an open mind, however, and I would request the family do so also. As our multidisciplinary team learns more about the family and as the family learns more about the options, it is hard to predict the direction of a consensus between us. At the very least, it would be important to work hard to reach a consensus over time. However, at some point, if the family insisted on providing unrestricted feeding and if we were not persuaded that this was an appropriate decision, I would reluctantly acquiesce and acknowledge that this was a tragic and unfortunate situation. Although I may believe that this family’s decision is ethically wrong, a gap exists between ethical determinations and the legal threshold for medical neglect. I would seek advice from the ethics committee and the child protection team, but I would anticipate a group consensus that states that intervention should not be pursued for weight management for J.R., given diverse professional attitudes about children with profound disabilities and the ambiguity and subjectivity of this assessment.

Dennis Rosen Writes

Although OSA is a treatable medical condition in most children, it can be very
difficult to manage in those with developmental and/or behavioral impairment. While striving to do the best for all their patients, it is important that physicians weigh carefully the risks and benefits of treatment on an individual basis and present these to parents or guardians so that an appropriate patient-specific treatment plan can be developed. Such individualized treatment plans are especially important when some or all of the treatment options may be associated with significant discomfort or even risk to the patient, and the patient is unable to provide consent or assent or even to understand their purpose. Just because something can be done does not mean that it should be done for all patients under all circumstances.

For some terminally ill patients, the aggressive pursuit of radical therapy may result in a short-term extension of life at the cost of significant discomfort or suffering. In such cases, it is easy to defend a shift to palliative care.

This patient’s situation, however, is very different. The end of her life is not imminent or inevitable. The patient’s mother and physician had to choose between subjecting her to significant discomfort and even the risk of possible injury to treat the OSA or leaving it untreated, knowing that this would likely lead to her death.

This situation is more like that faced by the doctors and family of Joseph Saikewicz. Saikewicz was a 67-year-old profoundly developmentally and intellectually delayed, nonverbal man with an estimated IQ of 10 who had lived in residential facilities his entire life. He was diagnosed in April 1976 with acute myeloblastic leukemia. The standard of treatment at the time was prolonged chemotherapy, which could at best provide a 30% to 50% remission rate lasting 2 to 13 months. Saikewicz would have suffered from the side effects of chemotherapy without understanding why it was being given and would have needed to have been physically restrained to be treated.

The Massachusetts Supreme Court weighed the duty toward preservation of life against the right of an individual to decline treatment. In its decision, the court found that the effect of the treatment would be for him to “experience fear without the understanding from which other patients draw strength” and that “to presume that the incompetent person must always be subjected to what many rational and intelligent persons may decline is to downgrade the status of the incompetent person by placing a lesser value on his intrinsic human worth and vitality.” They ruled that Mr. Saikewicz did not need to undergo chemotherapy. He died 5 months after the initial diagnosis was made.

In J.R.’s case, her severe developmental and behavioral impairment makes treating her residual OSA possible only by causing her substantial discomfort and/or by placing her at risk for serious injury. In my opinion, these risks outweigh the benefits of treatment. Her unique risk of complications from the untreated OSA is a consequence of her underlying profound developmental and intellectual disabilities. Thus, although treatment of OSA would be considered the standard of care in most cases, the unique circumstances of this case justify withholding life-prolonging treatment and shifting to palliative care.

Treatment dilemmas of this kind are not uncommon when caring for children with severe and complex disabilities. Successfully resolving them necessitates placing the well being of the specific patient ahead of what might be considered standard of practice for the general population. In the final analysis, first doing no harm remains as important a principal in guiding patient care today as it was in the days of Hippocrates.

**John D. Lantos Comments**

Patients should not be denied beneficial medical treatments simply because they have cognitive disabilities. It seems like that is happening in this case. CPAP is clearly the standard of care for OSA. J. R. would likely benefit from CPAP. If J.R. was not cognitively impaired, CPAP would certainly be provided. It doesn’t seem fair that JR should die. On the other hand, patients should not be forced to undergo treatments that they find noxious. Families should not be required to make extraordinary sacrifices to comply with burdensome treatment regimens. The only response to such a case is to strike a balance between 2 ethically problematic choices. When every option is bad, none can be considered obligatory. In such cases, doctors must ensure that the family is counseled and that they understand the options and the implications of their choices. Then they should be supported in whatever choice they make.

**REFERENCES**


Obstructive Sleep Apnea in a 17-Year-Old With Profound Cognitive Impairment

Dennis Rosen, Ben Wilfond and John D. Lantos

Pediatrics 2013;131;581; originally published online February 4, 2013;
DOI: 10.1542/peds.2012-1489

Updated Information & Services
including high resolution figures, can be found at:
/content/131/3/581.full.html

References
This article cites 18 articles, 4 of which can be accessed free at:
/content/131/3/581.full.html#ref-list-1

Post-Publication Peer Reviews (P3Rs)
One P3R has been posted to this article:
/cgi/eletters/131/3/581

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
Developmental/Behavioral Pediatrics
/cgi/collection/development:behavioral_issues_sub
Cognition/Language/Learning Disorders
/cgi/collection/cognition:language:learning_disorders_sub

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
/site/misc/Permissions.xhtml

Reprints
Information about ordering reprints can be found online:
/site/misc/reprints.xhtml

PEDIATRICS is the official journal of the American Academy of Pediatrics. A monthly publication, it has been published continuously since 1948. PEDIATRICS is owned, published, and trademarked by the American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, Illinois, 60007. Copyright © 2013 by the American Academy of Pediatrics. All rights reserved. Print ISSN: 0031-4005. Online ISSN: 1098-4275.
Obstructive Sleep Apnea in a 17-Year-Old With Profound Cognitive Impairment

Dennis Rosen, Ben Wilfond and John D. Lantos

*Pediatrics* 2013;131;581; originally published online February 4, 2013;
DOI: 10.1542/peds.2012-1489

The online version of this article, along with updated information and services, is located on the World Wide Web at:
/content/131/3/581.full.html