



A Case Study of Congenital Ciliary Dyskinesia: Navigating Difficult Ethical Considerations for Pediatric Anesthesia in an Amish, Rural Community

James Kim BA^[1]; Anjana Kundu, MD^[2] ; Alexander Mandych, MD^[3]; Robert Williams, MSNA, Ph.D, MD^[4]

^[1]= University of Rochester School of Medicine and Dentistry ; ^[2]= University of Rochester Medical Center, Golisano Children's Hospital;

^[3]= University of Rochester Medical Center, Department of Otolaryngology ; ^[4] = University of Rochester Medical Center, Department of Anesthesiology



UNIVERSITY of
ROCHESTER
MEDICAL CENTER

Introduction

Pediatric anesthesia in rural settings presents unique challenges, often requiring a balance between limited resources and the complex health needs of vulnerable populations. Social determinants of health, such as access to specialized facilities, can create ethical dilemmas when families face barriers to optimal care. In such cases, healthcare providers must navigate parental preferences, cultural considerations, and safety risks, making difficult decisions about when and where to proceed with care. This case highlights the ethical complexities of providing anesthesia to a young child with congenital ciliary dyskinesia in a rural, Amish community, where both medical risk and healthcare access play pivotal roles.

Case Report

A two-year-old Amish patient presented to a rural outpatient surgery center for a scheduled bilateral tympanostomy tube insertion for chronic otitis. Owing to lack of phone access and automobile transportation, routine screening had not occurred, and no known prior medical history was available for comparison. Day-of-surgery preoperative interview revealed that the patient had chronic cough secondary to a suspected case of congenital ciliary dyskinesia, a rare disorder associated with impaired mucociliary clearance and higher incidence of respiratory complications under general anesthesia, due to the presenting symptoms and a family history of the condition. However, this was neither confirmed nor ruled out owing to a lack of confirmatory ciliary biopsy needed for diagnosis.

The surgery center was staffed by a single Board-Certified anesthesiologist working alone without any pediatric specialists on site. The consent discussion considered whether to proceed, or to reschedule at the university's pediatric hospital. Given the demographics, there was risk of losing the patient to follow-up or, at best, delaying care with potential worsening of hearing loss. While the patient did not have any significant history for developmental delay, prolonged hearing deficient has been shown to lead to speech delay, and this patient was noted to speak only a few words by their parents.

Ultimately, the parents were well informed about the diagnosis and wished to proceed with general anesthesia using inhalational anesthetics. The surgery was uneventful, and the patient recovered without complication.



Figure 1: Common Pathologies and Radiographic Findings in Primary Ciliary Dyskinesia. This figure illustrates characteristic radiographic findings in primary ciliary dyskinesia (PCD). The first two images highlight bronchiectasis, a common manifestation of PCD due to impaired mucociliary clearance, resulting in widened bronchi and recurrent respiratory infections. The third image displays maxillary sinus opacification, a sign of chronic sinusitis, which frequently accompanies PCD as a consequence of persistent mucus buildup in the sinuses.



Figure 2: Geographic Distribution of University of Rochester Medical Center Hospitals. This map displays the locations of Strong Memorial Hospital, Noyes Memorial Hospital, St. James Mercy Hospital, and Jones Memorial Hospital within the University of Rochester Medical Center (URMC) healthcare network. Strong Memorial, located in Rochester, serves as the primary tertiary care facility with specialized pediatric resources, including advanced anesthesia and critical care capabilities. However, the rural placement of affiliate hospitals like Noyes, St. James, and Jones Memorial highlights a geographic barrier to accessing comprehensive pediatric care. For patients with complex conditions requiring specialized intervention, the significant distance from Strong Memorial presents logistical and safety challenges, particularly when emergency transport or continuity of care is needed.

Discussion and Ethical Considerations

Primary ciliary dyskinesia (PCD), also known as Siewert's Syndrome, is an autosomal-recessive disorder that leads to structural and/or functional abnormalities of motile cilia lining the epithelium of the respiratory tract, sinuses, and Eustachian tube (prevalence estimated at 1/15000). Decreased mucociliary clearance leads to clinical presentation commonly characterized by chronic sino-pulmonary dysfunction, including respiratory tract infections, bronchiectasis, and otitis media^[1]. Literature and case studies have suggested an increased risk of airway obstruction due to viscous mucous plugs and difficulty maintaining adequate oxygenation^[2].

Thus, regional anesthesia would be ideal to reduce risk for anesthesia-related respiratory complications if that can meet the surgical needs. However, the syndrome is often contributing to otolaryngological issues for which regional anesthesia is not appropriate. For these respiratory and otolaryngological issues, anesthesia is the only reasonable option^[3]. The heightened risk for post-operative pneumonia and other respiratory complications, such as bronchospasm and atelectasis, should also be considered within the context of the patient's presentation and the resources available to manage these complications should general anesthesia be performed.

This case underscores critical ethical challenges in rural pediatric anesthesia, particularly balancing patient safety with access to care. Limited resources in rural settings can compromise the ability to safely manage complex conditions, especially when specialized facilities are far from reach. Here, the immediate risk of respiratory complications due to suspected congenital ciliary dyskinesia must be weighed against the potential harms of delaying care or losing the patient to follow-up, both of which could hinder timely treatment and developmental outcomes. Informed consent is central to ethical care, as parents must understand and weigh these risks when choosing between accessible but less-resourced care and distant, specialized facilities. Additionally, rural healthcare equity comes into play, as patients in isolated areas may not receive the same standard of care due to staffing and equipment limitations, raising questions of justice and fairness. Ultimately, healthcare providers must strive to minimize harm while respecting patient autonomy, all within the constraints imposed by the rural setting and cultural factors. This balance requires careful risk stratification and a commitment to both patient safety and equitable access to care.

References

- [1] Zariwala MA, Knowles MR, Leigh MW. Primary Ciliary Dyskinesia. 2007 Jan 24 [Updated 2019 Dec 5]. In: Adam MP, Feldman J, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2024
- [2] Dylan M, Gothard, J.W. (2006). Sudden hypoxia during anesthesia in a patient with Kartagener's syndrome. Pediatric Anesthesia, 16: 977-980.
- [3] Cheng L, Dong Y, Liu S. Anesthetic Management of Patients With Kartagener Syndrome: A Systematic Review of 99 Cases. J Cardiothorac Vasc Anesth. 2023 Jun;37(6):1021-1025. doi: 10.1053/j.jvca.2023.01.033. Epub 2023 Feb 3. PMID: 36849313.