Anesthesia and Ullrich Congenital Muscular Dystrophy: Comment

DOI: 10.1097/ALN.0000000000003698

To the Editor:

I read with great interest the short report published in the Images in Anesthesiology section about difficult intubation in a 2-yr-old patient with Ullrich congenital muscular dystrophy.[1] This disease is well known to carry a risk of difficult intubation.[2–6] The authors nicely described how they used a nasopharyngeal airway to administer a volatile anesthetic and oxygen through one nostril while performing nasotracheal fiberoptic intubation *via* the other. I am however surprised that the choice of the anesthetic agent(s) used was not discussed. Ullrich congenital muscular dystrophy indeed belongs to the subgroup of the collagen type 6–related myopathies occurring after a mutation of the COL6A1, COL6A2, or COL6A3 gene. Collagen type 6 is part of the large complex that anchors the basal lamina and the interstitium in muscle cells. The myopathy is probably caused by the muscle membrane fragility and an associated mitochondrial dysfunction, which can be decreased with cyclosporine A. As collagen 6 is close to the dystrophin-glycoprotein complex, this muscle disease could be at risk of anesthesia-induced rhabdomyolysis in the presence of halogenated agents or succinylcholine, as are children with Duchenne or Becker progressive muscular dystrophy. Very few reports on the anesthetic management of patients with this type of myopathy have been published so far and all except two[5,6] report using total intravenous anesthesia.[2–4] Carefully titrated intravenous anesthesia to maintain spontaneous ventilation and using either dexmedetomidine, propofol, and/or ketamine could therefore be a safe alternative.

Competing Interests

Dr. Veyckemans reports a financial relationship with *European Journal of Anaesthesiology*'s Editorial Board and *Pediatric Anesthesia*'s Editorial Board.

Francis Veyckemans, M.D., Jeanne de Flandre Hospital, Academic Hospital of Lille, Lille, France. francis.veyckemans@chru-lille.fr

References

1. Duong KW, Reddy SC, Buchanan EP, Chen JH: Mandibular and maxillary cysts in a pediatric patient with Pierre Robin sequence and Ullrich congenital muscular dystrophy. Anesthesiology 2020; 133:919–20
2. Puangsuvan N, Mester RA, Ramachandran V, Tobias JD: Perioperative care of a child with Ullrich congenital muscular dystrophy. Middle East J Anaesthesiol 2009; 20:319–23
3. Vandenberghe W, Jacobs TF, Plasschaert FS, Willems J, Den Blauwen NM, Vereecke HE, Wouters P: Anesthesia and perioperative management for a patient with Ullrich syndrome undergoing surgery for scoliosis. Acta Anaesthesiol Belg 2010; 61:43–7
4. Erbabacan E, Köksal GM, Şeker TB, Ekici B, Özcan R, Altindaş F: Anaesthesia management and use of sugammadex in a patient with Ullrich's disease. Turk J Anaesthesiol Reanim 2015; 43:356–9
5. Grosu I, Truong D, Teodorescu S, Mousny M, Veyckemans F: Anesthetic management of a child with Ullrich myopathy. J Anesth 2012; 26:636–7
6. Martin DP, Tobias JD, Warhadpande S, Beebe A, Klamar J: Perioperative care of a child with Ullrich congenital muscular dystrophy during posterior spinal fusion. South Afr J Anaesth Analg 2013; 19:73–6

(Accepted for publication January 6, 2021. Published online first on February 2, 2021.)

This letter was sent to the author of the original article referenced above, who declined to respond.—Evan D. Kharasch, M.D., Ph.D., Editor-in-Chief.

Goal-directed Therapy and Postcystectomy Ileus: Comment

To the Editor:

We read with interest the recently published study by Arslan-Carlon *et al.* in Anesthesiology.[1] This primary objective of this randomized, controlled trial (N = 283) was to determine whether a goal-directed fluid therapy approach would result in a lower incidence of postoperative ileus compared with a standard fluid therapy approach