Bifid epiglottis in a patient with Pallister-Hall syndrome

Molly Kraus, MD · Michelle Diu, MD

Received: 13 May 2016/Revised: 8 June 2016/Accepted: 14 June 2016/Published online: 23 June 2016
© Canadian Anesthesiologists' Society 2016

A 14 yr-old girl with Pallister-Hall syndrome was admitted for osteotomy and fusion of the right fifth metatarsal, tenotomy of the flexor tendon, curly toe release of the right fourth toe, and removal of hardware from the left fifth metatarsal. General anesthesia was induced. During intubation using a video laryngoscope and a 6-mm cuffed endotracheal tube, a bifid epiglottis was noted (Figure).

Pallister-Hall syndrome is a rare, autosomal dominant, genetic condition involving a mutation in the *GLI3* gene, which is located on the short arm of chromosome 7.¹ This gene encodes zinc finger protein GLI3, a transcription repressor that is a mediator of sonic hedgehog signaling. The latter plays a key role in a wide variety of embryonic developmental processes.² Most patients with this condition have polydactyly and cutaneous syndactyly. Hypothalamic hamartoma, which is characteristic of this disorder, may lead to seizures or neuroendocrine abnormalities. Other features of the syndrome include bifid epiglottis, laryngeal cleft, imperforate anus, and kidney anomalies. Bifid epiglottis is reported in 40% of patients with Pallister-Hall syndrome.³ It is typically asymptomatic but can be associated with dysphagia, stridor, chronic aspiration, and, rarely, airway obstruction.³ The presented patient denied any associated symptoms.

Figure Both images show the bifid nature of the epiglottis in this patient with Pallister-Hall syndrome. The images were captured during intubation with a video laryngoscope

M. Kraus, MD (✉) · M. Diu, MD
Division of Pediatric Anesthesiology, Omaha Children's
Hospital, University of Nebraska Medical Center, Omaha, NE,
USA
e-mail: mokraus@childrensomaha.org

Conflicts of interest None declared.

Editorial responsibility This submission was handled by Dr. Hilary P. Grocott, Editor-in-Chief, *Canadian Journal of Anesthesia*.

References

1. *Hall JG*. The early history of Pallister-Hall syndrome—buried treasure of a sort. *Gene* 2016. DOI:[10.1016/j.gene.2016.01.003](https://doi.org/10.1016/j.gene.2016.01.003).
2. *Hill P, Wang B, Ruther U*. The molecular basis of Pallister-Hall associated polydactyly. *Hum Mol Genet* 2007; 16: 2089-96.
3. *Stevens CA, Ledbetter JC*. Significance of bifid epiglottis. *Am J Med Genet A* 2005; 134: 447-9.