

Anesthesiology
78:408, 1993
© 1993 American Society of Anesthesiologists, Inc.
J. B. Lippincott Company, Philadelphia

Anesthesia for Children with Freeman-Sheldon Syndrome

To the Editor:—I read with interest the report by Jones and Dolcourt¹ on muscle rigidity following halothane anesthesia in two patients with Freeman-Sheldon syndrome. Other case reports²⁻⁴ describing anesthesia in such patients have not reported malignant hyperthermia. I recently anesthetized a child with Freeman-Sheldon syndrome. She had undergone multiple surgical and anesthetic procedures at another medical center without incident. The parents stated that she had experienced difficulties with tracheal intubation and that her trachea was deviated to the left. The parents expressed concern that malignant hyperthermia had been reported in other children with this syndrome, although their child had not experienced any temperature instability. A smooth inhalation induction using halothane nitrous oxide and oxygen was accomplished. Temperature was monitored per rectum. An electrocardiogram, a pulse oximeter, and end-tidal CO₂ were used to monitor for early signs of malignant hyperthermia.

The hypopharyngeal area was sprayed with 2% xylocaine, 4 mg/kg. With the child breathing spontaneously and with the knowledge that the trachea was deviated to the left, the glottis was found on the left side of the hypopharynx. Laryngoscopy and intubation were easier due to the movements of the arytenoid cartilages (because of spontaneous ventilation). There were no signs of malignant hyperthermia and the strabismus surgery was completed without problems.

This is a case of a child with Freeman-Sheldon syndrome who has undergone multiple anesthetics without signs of malignant hyperthermia. Whether or not children with Freeman-Sheldon syndrome are prone to malignant hyperthermia is open to question; more experience is needed. My conclusion differs from the authors': one should consider these children as "potentially susceptible" to malignant hyperthermia and monitor them appropriately. However, to

deny the patient and the anesthesiologist the use of spontaneous ventilation under deep halothane anesthesia is not warranted on the basis of one or two case reports of suspicious, but unconfirmed malignant hyperthermia episodes.

James F. Mayhew, M.D.
Director, Pediatric Anesthesiology
Acting Chairman, Anesthesiology
Pediatric Anesthesiology
Children's Hospital of the King's Daughters
601 Children's Lane
Norfolk, Virginia 23507

References

1. Jones R, Dolcourt JL: Muscle rigidity following halothane anesthesia in two patients with Freeman-Sheldon syndrome. *ANESTHESIOLOGY* 77:599-600, 1992
2. Laishley RS, Roy WL: Freeman-Sheldon syndrome: Report of three cases and the anaesthetic implications. *Can J Anaesth* 33:388-393, 1986
3. Duggar RG, DeMars PD, Bolton VE: Whistling face syndrome: General anesthesia and early postoperative caudal analgesia. *ANESTHESIOLOGY* 70:545-547, 1989
4. Tateishi M, Imazumi H, Namiki A, Katsuno M, Kawana S, Ujike Y: Anesthetic management of a patient with Freeman-Sheldon ("whistling face") syndrome. *Masui* 35:1114-1118, 1986

(Accepted for publication November 18, 1992.)

Anesthesiology
78:408, 1993
© 1993 American Society of Anesthesiologists, Inc.
J. B. Lippincott Company, Philadelphia

In Reply:—From the 51 families (59 affected individuals) in the Freeman-Sheldon Syndrome (FSS) Parent Support Group, we identified the two FSS patients with muscle rigidity as described in our report.¹ These two children had received recognized triggering agents for malignant hyperthermia (MH). Not included in our report is one patient who had muscle rigidity occurring during anesthetic induction. This child had two other family members affected with MH.

There is recognized heterogeneity in the physical findings of FSS. There similarly may be an inconsistent association of MH in FSS. We continue to think that in the absence of results from a caffeine-halothane muscle contracture test, nontriggering anesthetic agents should be considered for all patients with FSS.

Roger Jones, M.D.
Department of Anesthesiology

Jack Dolcourt, M.D.
Department of Pediatrics
Primary Children's Medical Center
100 North Medical Drive
Salt Lake City, Utah 84113

Reference

1. Jones R, Dolcourt JL: Muscle rigidity following halothane anesthesia in two patients with Freeman-Sheldon syndrome. *ANESTHESIOLOGY* 77:599-600, 1992

(Accepted for publication November 18, 1992.)