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LETTER TO THE EDITOR

Postoperative cyanotic breath-holding spells in a child with Worster–Drought syndrome

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To the Editor:

We report multiple cyanotic spells in the postoperative period in a child with Worster–Drought syndrome.

A male child, 3.5 years old, 12 kg, who had experienced drooling of saliva since birth, was accepted for bilateral submandibular duct translocation and sublingual gland excision. He had delayed milestones (sat at 15 months and walked at 3 years). Difficulty in swallowing resulted in frequent vomiting during feeding and recurrent upper respiratory tract infections (URI). He was fed by his parents and could mainly swallow liquids or semi-solids. He did not require a nasogastric tube or gastrostomy for feeding. There was no history of epigastric pain or discomfort following feeds that would suggest a gastroesophageal reflux disorder. He was unable to protrude his tongue despite there being no tongue-tie. He could not speak despite normal hearing (normal brainstem auditory evoked responses) and communicated with signs. There was no history of seizures. Head noncontrast computerized tomography (NCCT) showed a paucity of periventricular white matter with prominent ventricles, gliotic areas, and bilateral frontoparietal volume loss. Other systems were normal. Before surgery, the patient did not have an URI, and no adventitious sounds were heard on auscultation.

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In the operating room, after instituting routine monitoring, anesthesia was induced with sevoflurane in oxygen and an IV access was secured. Following the administration of fentanyl and vecuronium, nasotracheal intubation was accomplished using a 4-mm-ID uncuffed endotracheal tube and the oropharynx packed. Paracetamol suppositories (250 + 170 mg) were inserted rectally. Anesthesia was maintained with oxygen, nitrous oxide, isoflurane, and supplemental doses of fentanyl and vecuronium. The intraoperative period was uneventful, and at the end of surgery, which lasted for 2 h, a nasogastric tube was inserted to facilitate feeding the child after the oral surgery. Neuromuscular blockade was reversed and the trachea extubated once the patient was fully awake and breathing regularly. As he was being shifted out, he sat up and started crying. He held his breath, desaturated, and became cyanosed. Saturation improved on bag-mask ventilation with 100% oxygen. Two similar episodes of desaturation occurred. After the third, a conscious effort was made to avoid any stimulation that could precipitate crying, and he was gently shifted to his mother in the post anesthesia care unit (PACU). On retrospective questioning, the parents admitted that if they force-fed the child, he would have prolonged crying with breath-holding spells; however, there was no history suggestive of cyanosis during these episodes.

Worster–Drought syndrome (WDS) is a phenotypically distinct but underdiagnosed form of cerebral palsy first described in 1956 [1]. Patients share features with bilateral congenital perisylvian syndrome, a type of childhood-onset epilepsy [1].

Worster–Drought syndrome is characterized by weakness of the orofacial and bulbar muscles, leading to difficulty in feeding, swallowing, and voluntary lip, tongue, and palate movements. Attempts at feeding can cause choking and

aspiration, leading to frequent chest and middle-ear infections. Because of bulbar involvement, White [2] used conscious sedation instead of general anesthesia for a 29-year-old patient undergoing bilateral intraarticular injections.

Another major problem is speech with delayed onset or abnormal voice or articulation.

Mild tetraplegia presents as delayed milestones. These children are, however, ambulant, in contrast to children with commoner forms of cerebral palsy. Cognitive impairment is not severe, but learning difficulties are observed in 81%, congenital defects (palatal defects, contractures, micrognathia, arythrogryposis) in 60%, and neuropsychiatric abnormalities (attention deficit, hyperactivity disorders, mood problems, autism) in 41%; 28% may have associated epilepsy. [1]

The breath-holding spells in this child could have many causes. The child had recovered well from anesthesia and neuromuscular blockade before these spells occurred. Surgery in the oral cavity combined with a nasogastric tube in the pharynx could have resulted in lack of coordination in breathing and swallowing in the crying child. The retrospective history of such episodes occurring during feeding suggests this as a major cause of the breath-holding cyanotic spell.

He received a total of 4 μg/kg fentanyl in titrated doses intraoperatively in addition to the paracetamol suppositories. Pain was not a cause of crying and cyanosis, as was evidenced by the fact that on reaching his mother's lap he became quiet and looked comfortable. The presence of strangers [the operating room (OR) personnel] trying to pacify him could have distressed the child. Descending limbic connections to the ventrolateral medulla may modulate the somatic respiratory musculature, and breathing may be affected by emotional outbursts [3].

In conclusion, children with WDS may not have the obvious motor deficit observed with commoner forms of cerebral palsy; however, bulbar involvement, difficulty in articulation, and behavioral problems may make them more likely to experience respiratory complications in the perioperative period.

References

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