Difficult airway management and suspected malignant hyperthermia in a Child with Cri Du Chat Syndrome

Sir,

Cri du Chat Syndrome (CdCs) also known as Cat Cry syndrome, is a genetic disorder caused by the partial or total deletion of genetic material from the short arm of chromosome 5 (5p-, 5p minus syndrome) and was first described in 1963 by Lejeune *et al*.[1] The phenotypic manifestations are variable and often affect multiple organ systems with the most common features being the characteristic high-pitched cat-like cry (hence the name), dysmorphic features, and mental retardation.[2,3] The reported incidence of CdCs is between 1 in 15,000 and 1 in 50,000 live births,[2,3] and the children often require surgery for correctable anomalies.[3] Difficult airway management is a recognized challenge because of dysmorphic changes such as microcephaly, micrognathia, short neck, and the anatomical anomalies of the larynx.[2,3] However, there have not been any reports of increased risk or susceptibility to malignant hyperthermia (MH) with this condition. I present a child that developed airway management challenge and possible MH during anesthetic induction.

A 2-year 5-month, 9.5 kg African male child with bilateral congenital talipes equinovarus congenital talipes equinovarus (CTEV) otherwise known as clubfoot, was scheduled for bilateral posteromedial release operation after minimal improvement with the conservative serial splinting, stretching, and bracing treatment known as the Ponseti method.

During the preoperative anesthetic assessment on the day before surgery, the child was noted to have an unusual facies, and he could not walk or talk. He had a peculiar high pitched cry that sounded cat-like that the mother said had been present since birth. He was the third in a family of three children, and he had been hospitalized twice in the past year for aspiration pneumonia. He was small for age and had a global developmental delay with significant hypotonia, gross motor and speech impairment, and severe bilateral CTEV. He had a microcephalic head with a round face, hypertelorism, depressed broad nasal bridge, low set ears, and micrognathia (head circumference 48 cm, height 79 cm). Clinical examination of the cardiorespiratory system was unremarkable. Hematologic examination indicated hypochromic microcytic anemia and thrombocytosis (hemoglobin - 10.5 g/dl, platelets - 513 $\times$ 10^9/L). Chest X-ray was unremarkable.

The anesthetic plan was to maintain the airway with a laryngeal mask airway (LMA) and perform a caudal epidural block for perioperative analgesia. On the operating table, routine monitors of noninvasive blood pressure (BP), electrocardiography, SpO_2, and temperature were applied, and anesthesia was induced inhalationally with sevoflurane and nitrous oxide in oxygen. An intravenous (iv) access was secured once the child was unconscious. After the adequate depth of anesthesia, a size 2.0 LMA was inserted but ventilation through it was ineffective and it was removed with a plan to reinsert it properly. Morphine 1 mg iv was given. When attempting to reinsert the LMA, the child was noticed to have suddenly developed generalized muscle rigidity including trismus and laryngospasm. Mask ventilation became very difficult and ineffective, and the mouth could not be opened to insert an oropharyngeal airway. Propofol 20 mg iv was given to facilitate jaw relaxation but had no effect. At this point, the patient's oxygen saturation had begun to rapidly decrease. Succinylcholine, 20 mg iv was given, but it also had no effect. Attempts at laryngoscopy were impossible as the teeth were tightly clenched. The pulse rate was now >195/min and the immediate preinduction temperature of 35.7°C had gone up to 37.7°C. The capnograph tracing that initially indicated hypercarbia had progressed to almost baseline because of inability to ventilate the lungs. At this stage, the sevoflurane and nitrous oxide were turned off, and the patient was maintained on 100% oxygen through bag and mask. We were about to perform an emergency cricothyroidotomy when we noticed that the child that had been apneic during this period started making some respiratory efforts that rapidly got better and the oxygen saturation progressively improved with assisted respiration. About 10 min after the commencement of the incident, the child was breathing adequately spontaneously with SpO_2 of 99%, and after close observation for about 15 min, he was transferred to the PACU for further monitoring. He was fully awake within 5 min of arrival in the PACU and was responding appropriately. The temperature and heart rate were still elevated (37.5°–377.7°C; 165–196/min) while his BP and oxygen saturation were normal during his 2 h stay

in PACU. He was stable enough to be sent back to the high dependency unit of the ward and was closely observed by the ward doctor and reviewed by the pediatrician. The heart rate and temperature returned to preinduction levels about 4 h after the incident, and he was discharged from the hospital without any adverse effects or sequelae 96 h after the incident.

Children with CdCs have variable multiple phenotypical manifestations that predispose them to a greater likelihood of surgical intervention and also that have significant anesthetic implications.[2,3] Diagnosis is clinical with confirmation by molecular cytogenetic tests such as fluorescent *in situ* hybridization. With increasing and widespread availability of cytogenetic studies, the anesthesia care provider is likely to see increasing numbers of patients with this relatively common chromosomal condition.[2,3] Up to two-thirds (75%) of patients with CdCs require general anesthesia for interventional procedures and parents felt that only about one-third (35%) of the anesthesiologists caring for their children were familiar with the condition.[3]

Previous studies have reported an increased risk of airway difficulty and inability to intubate the trachea.[2,3] This is not surprising considering the orofacial anomalies these patients possess. MH risk or susceptibility has however not been associated with CdCs to our knowledge. We have a strong impression that our patient developed intraoperative MH because of the constellation of signs and symptoms. We initially assumed that the muscle rigidity and masseter spasm in a hypotonic child may have been opioid induced and/or due to a "light" plane of anesthesia, but when administration of propofol and then succinylcholine and maximum concentration of sevoflurane had no effect, coupled with the rapid development of hyperthermia, tachycardia, and hypercarbia, MH seemed the most likely diagnosis. In addition, discontinuation of the presumptive trigger, sevoflurane, led to improvement.

As early diagnosis and prompt treatment are vital in minimizing the high morbidity and mortality associated with MH, the diagnosis is usually clinical and requires a high index of suspicion and good clinical acumen.[4] This is especially important in resource-poor environments where medical record keeping is poor and necessary monitoring and laboratory facilities are not readily available. A clinical standardized and validated grading scale has been developed to help determine if an MH event has occurred.[5] The score ranges from 0 (almost never/very unlikely) to 6 (almost certain). Our patient scored 5, which indicates a "very likely" MH episode just short of the maximum 6, that indicates "almost certain" event in spite of our lack of relevant blood tests.

We did not use dantrolene, a specific drug indicated for MH because of nonavailability. However, patients have been shown to survive MH episodes even in the absence of dantrolene provided the diagnosis is considered early enough, and aggressive symptomatic treatment is carried out.[6] We were unable to confirm a definite diagnosis of MH because of unavailability of the "gold standard" confirmatory *in vitro* contracture test[4] however, pending future confirmatory studies it would be prudent to consider patients with CdCs as susceptible to MH and treated as such.

Consent

The child's mother gave consent for publication of this.

Declaration of patient consent

The author certifies that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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