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LETTERS TO THE EDITOR

Anesthesia for intestinal obstruction in a six years old child with cerebro-oculo-facio-skeletal syndrome



Anestesia para obstrução intestinal em uma criança de seis anos de idade com síndrome COFS

Dear Editor,

We report our anesthesia management of a 6 years old girl with Cerebro-Oculo-Facio-Skeletal (COFS) Syndrome who was consulted as intestinal obstruction in emergency department of our hospital. She was already a follow-up patient of pediatric department and diagnosed as COFS syndrome by them previously.

She had characteristic signs of the syndrome including growth failure, weighing 6 kg, cachexia, micrognathism, microcephaly. She had small mouth opening and dental abnormalities, flexure contractures at her elbow and knees. Her preoperative blood tests were within normal limits except C-reactive protein (40 mg.dL⁻¹) and White Blood Cell (18 × 10³). Plevral effusion and abdominal volvulus was found in her radiological images. Her initial vital signs were in normal range. Intravenous access was established by 24 gauge catheters. Anesthesia was induced with sevoflurane 8% in air–oxygen mixture and 12 mcg fentanyl intravenously. Intubation was done with a cuffed 4.0 mm endotracheal tube successfully. Anesthesia maintenance was provided with sevoflurane 2% in air–oxygen mixture, and fentanyl. She was ventilated in pressure control ventilation. There was no anesthetic problems during the operation that continued for 135 min. After adequate breathing spontaneously, she was extubated smoothly.

COFS syndrome is initially described by Pena and Shokeir in 1974 and has been recognized as a rare, autosomal recessive disorder characterized by DNA repair defect.¹ Cockayne syndrome Type II is known as COFS syndrome. Degenerative problem of the brain and spinal cord is usually seen before birth. Clinical findings are psychomotor development delay, neurological dysfunction, peripheral neuropathy, microcephaly, micrognathism, hypotonia,

hyporeflexia, convulsions, and congenital cataract. And is associated with feeding difficulties, sensorineural hearing loss, coxa valga, knee flexion contracture.¹ Progressive neurological degeneration can lead to a risk of hyperkalemia by using succinylcholine in these patients. Reports can be seen describing the anesthesia management of Cockayne syndrome in the literature but there was no about the anesthesia management of COFS syndrome. Difficult airway management was one of the most important problem in anesthesia procedure. Also, due to the contractions and osteoporosis, adequate care should be considered while giving position during the surgery.

That is difficult to predict the effects of muscle relaxants' in these patients for they are cachectic, almost lack of muscle with insufficient development of the neuromuscular junction for this, they are need to be avoided. We saw that only one case that muscle relaxant was not used during anesthesia in the literature.² Endotracheal intubation was done via sevoflurane combined with nitrous oxide in this patient who was scheduled for liver biopsy, and the procedure lasted for 13 min. We did not use muscle relaxants in induction and maintenance of anesthesia, too. Our procedure continued 135 min. We used weight-appropriate endotracheal tube than age-appropriate one but we preferred cuffed – tube to avoid the risk of aspiration.

In conclusion; all of these associated anomalies and dysfunctions requiring attention and experience. Using smaller endotracheal tubes and avoiding muscle relaxants during intubation can be kept in mind during anesthesia management of COFS syndrome.

Conflicts of interest

The authors declare no conflicts of interest.

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Severe hypoxemia follows hypoxic pulmonary vasoconstriction and/or hypoxic pulmonary vasoconstriction inhibition by inhaled anesthetics: prognostic potential of 100% shunt fractions



Hipoxemia grave após vasoconstrição hipóxica pulmonar e/ou inibição da vasoconstrição hipóxica pulmonar por anestésicos inalatórios: potencial prognóstico de frações de shunt de 100%

Dear Editor,

Hypoxic Pulmonary Vasoconstriction (HPV) was first reported by Bindsley et al. in adult patients and was managed using double lumen catheters to ventilate one lung with 100% oxygen and the other with 95% N₂ and 5% oxygen during intravenous barbiturate and fentanyl anesthesia.¹ However, "HPV inhibition" has predominantly been attributed to the use of inhalation agents and is considered as a cause of hypoxia during anesthesia. Moreover, both in vitro and in vivo studies have demonstrated that inhalation agents inhibit HPV under a range of conditions,²⁻⁵ and sevoflurane has been shown to decrease HPV in a dose-dependent manner.^{4,6,7}

Case description

Here we report a case of severe hypoxemia during sevoflurane induction in a 6 year-old boy who was scheduled for adenotonsillectomy. The patient was treated according to American Society of Anesthesiology (ASA) class I and weighed 22 kg. Pre-operative assessments, physical examinations, and laboratory investigations were unremarkable, and the patient had a hemoglobin level of 12 mg.dL⁻¹ and a hematocrit of 36%. Following transfer to the operating room with no pre-medication, routine Electrocardiography (ECG), non-invasive blood pressure, and SpO₂ levels were monitored. Anesthesia was introduced via a facemask and a pediatric circle system providing 8% sevoflurane in 100% oxygen at a flow rate of 6 L.min⁻¹. Subsequently, sevoflurane concentrations were reduced to 5% within the first minute and to

2% on loss of eyelash reflexes. Following intravenous cannulation, rocuronium (0.6 mg.kg⁻¹) was administered, and SpO₂ levels rapidly and progressively decreased from 98% to 38% at 10 and 15 min of induction, respectively, with no clinical explanation. As a consequence, the patients' Heart Rate (HR) suddenly decreased from 109 to 90 bpm in response to hypoxia, and sevoflurane concentrations were reduced to 2% and subsequently discontinued prior to tracheal intubation and ventilation with 100% oxygen. Efficient lung ventilation was possible throughout the period. A clinical improvement was observed within seconds of intubation and ventilation, and SpO₂ levels and HR returned to 100% and 118 bpm, respectively. No blood pressure abnormalities were observed during the procedure, and the rapid restoration of SpO₂ and HR with 100% oxygen was considered symptomatic of an adverse drug reaction. Thus, anesthesia was maintained with 1%–1.5% sevoflurane and oxygen in 50% N₂O.

Discussion

HPV is considered as a protective mechanism that optimizes systemic oxygen delivery. Thus, the inhibition of HPV by inhalational agents is believed to cause hypoxia during anesthesia. However, the protective effects of HPV against hypoxia may depend on the size of affected lung regions. Lung tissues are globally affected by all pulmonary anesthetics, and sevoflurane has been shown to induce pulmonary vessel dilatation in normoxia.⁷ Therefore, HPV should be considered as a multifactorial response to local or global pulmonary hypoxia during acute hypoxia. Thus, the present observations suggest that the degree of acute local shunting can be used to determine whether HPV mechanism is useful or not. The type of anesthetics can be significant with regard to this reaction.

It is widely accepted that inhalation agents have global effects on pulmonary vessels during either induction or maintenance of anesthesia. Thus, hypoxemia because of global HPV is likely in the presence of 100% intrapulmonary shunt fractions, and the protective effects of HPV-influenced perfusion to the better ventilated lung regions to improve oxygenation may be abolished under these conditions. In contrast, inhibition of HPV by inhalation agents in commonly affected lungs leads to rapid onset of hypoxemia because of normal effects on perfusion and alveolar hypoxia and direct effects of anesthesia. Thus, the effect of HPV inhibition by inhalation agents or hypoxia may depend on global effects on lungs and the ensuing shunt fractions. Therefore, we suggest that both HPV and HPV inhibition may cause