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## CASE REPORT

# Intravenous anaesthesia for adenoidectomy in a 3-year-old child with Kartagener syndrome and sleep disordered breathing

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## ABSTRACT

*Kartagener syndrome (KGS) is a rare inherited disorder characterized by situs inversus viscerum (including dextrocardia) and primary ciliary dyskinesia resulting in chronic respiratory tract infections, bronchiectasis and sinusitis. Possible anesthesiologic challenges are related to the respiratory system and increased susceptibility to infectious complications. There are several case reports of general anesthesia in these patients, but mainly in the adult population. Here, we report on a 3-year old female child with KGS, who underwent adenoidectomy because of sleep disordered breathing (SDB). Preoperative preparation consisted of intravenous antibiotics, steroids, as well as postural drainage and inhalations of bromhexine and salbutamol. Anesthesia was induced with propofol, fentanyl and vecuronium and maintained with a continuous infusion of propofol (150-200 µg·kg<sup>-1</sup>·min<sup>-1</sup>) and supplemental doses of fentanyl. The child was ventilated with oxygen/air mixture (50%:50%) in the pressure-controlled mode of ventilation to keep end-tidal CO<sub>2</sub> between 30 and 35 mmHg. During anesthesia the child's hemodynamic and respiratory parameters were stable. Extubation, after thorough endotracheal and oral suction, was uneventful. After two hours in the post-anaesthesia care unit (PACU), the child was transferred to the ward. To the best of our knowledge, this is the youngest reported child with KGS and SDB that underwent intravenous general anesthesia. We also stress here the importance of comprehensive pre-anesthetic preparation, i.e. postural drainage, inhalations, bronchodilators, i.v. antibiotics and steroids. Furthermore, the condition of the respiratory system in the patient with KGS is seldom appropriate at the time of surgery, so the decision to anesthetize or not, should be made on an individual basis.*

**Key words:** Kartagener syndrome (KGS), sleep disorder breathing (SDB), total intravenous anaesthesia (TIVA)

## Introduction

Kartagener syndrome (KGS) is rarely encountered in paediatric anaesthesia practice. It is an inherited disorder characterized by situs inversus viscerum and primary ciliary dyskinesia resulting

in chronic respiratory tract infection, bronchiectasis and sinusitis. (1) Possible anaesthetic concerns are related to the respiratory system and increased susceptibility to infectious complications. (2) There are several case reports of general anaesthesia in these patients, mainly in adults, (2) and only three case reports were found for children, ages 6 to 15. (1,3,4) Inhalational anaesthesia was used in all children. Here, we report

on a 3-year-old child with KGS and sleep disordered breathing (SDB), who underwent adenoidectomy under intravenous general anaesthesia (TIVA).

## Case report

A 3-year-old (38 months), 18.5 kg female child was scheduled for adenoidectomy. As a newborn she was diagnosed with KGS, including dextrocardia and sinistroposition of the liver.

Symptoms related to difficult breathing were manifested from birth, and the child suffered from significant respiratory problems, even during common colds. Besides, the child had a history of SDB. She slept restlessly and sweat profusely. Ear-nose-throat (ENT) exam revealed "facies adenoidea" (permanent oral breathing, small nose, opened mouth, drooling, greenish nasal discharge). Fibre endoscopy found enlarged adenoids with 90% choanal obstruction (grade IV). Otoloscopic finding showed retraction of the eardrums on both sides, and tympanometry was normal.

During the pre-anaesthetic visit, the child had no fever, was in good general condition, but occasionally was unable to expectorate whitish, thick and viscous respiratory secretions without her mother's help. Her laboratory findings included leukocytes  $11.5 \times 10^9 \cdot l^{-1}$  (neutrophils 33.7 %, lymphocytes 57.1%, eosinophils 2.6 %, monocytes 6.1%, basophils 0.5%) and CRP  $0.3 \text{ mg} \cdot l^{-1}$ . During lung auscultation, bronchial breath sounds were heard on the right side, with basal crackles anteriorly. Because of clinical suspicion of right lung lower lobe infiltration, preoperative chest X-ray was done, but without clear radiological findings of possible pneumonia (figure 1).

Nevertheless, after reviewing thoroughly the clinical situation and consulting with her paediatrician we decided not to postpone the surgery because the child had not only KGS, but also SDB and we considered adenoidectomy mandatory.

After EMLA (Eutectic Mixture of Local Anesthetics) cream application, intravenous access was established on the ward. Ampicillin 1 gram and dexamethasone 4 mg i.v. were administered. The mother performed postural drainage, and the child inhaled a pre-mixed solution consisting of bromhexine 4 mg and salbutamol 2 mg, dissolved in 3 millilitres of normal saline for 20 minutes.

Anaesthesia was induced with propofol

$3 \text{ mg} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$ , fentanyl  $2 \mu\text{g} \cdot \text{kg}^{-1}$  and vecuronium  $0.1 \text{ mg} \cdot \text{kg}^{-1}$ . Paracetamol  $15 \text{ mg} \cdot \text{kg}^{-1}$  was given as a slow intravenous infusion immediately after induction. Anaesthesia was maintained with continuous infusion of propofol ( $150\text{-}200 \mu\text{g} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$ ) and a supplemental dose of fentanyl  $1 \mu\text{g} \cdot \text{kg}^{-1}$ . The child was ventilated with a mixture of oxygen and air (50%:50%) in the pressure-controlled mode of ventilation (inspiratory pressure  $20 \text{ cmH}_2\text{O}$ , respiratory rate  $17 \text{ min}^{-1}$ ) to keep end-tidal  $\text{CO}_2$  between 30 and 35 mmHg. No additional muscle relaxant was added. On two occasions during surgery, gentle endotracheal catheter aspirations had to be performed, since there were decreases in achieved tidal volumes, due to accumulated secretions. During anaesthesia the child was stable. Reversal of neuromuscular blockade was performed with atropine/neostigmine ( $0.02/0.07 \text{ mg} \cdot \text{kg}^{-1}$ ). Extubation, after thorough endotracheal and oral suction, was smooth. The child was transferred to the post-anaesthesia care unit (PACU). The postoperative course in PACU was uneventful in regard to bleeding and excessive secretions. The child needed just one more postural drainage. After 2 hours in PACU, the child was transferred to the ward. Postoperative analgesia was achieved with paracetamol syrup 240 mg every 6-8 hours. Immediately upon arrival in her room, the child began to drink liquids. No bronchodilators were needed. She was discharged the next day.

## Discussion

To the best of our knowledge, this is the youngest reported child with KGS that underwent general anaesthesia. Besides, TIVA was used here with a good result and volatile anaesthetics were deliberately omitted. Although volatile anaesthetics have been the mainstay of paediatric anaesthesia for a long time, and have salutary bronchodilatory action, we considered the possible laryngospasm and/or bron-

chospasm at induction or emergence from anaesthesia, in a child with KGS and SDB, to be potentially disastrous. Sevoflurane-related emergence agitation, with a reported incidence of 10-100%, is also a significant post-anaesthetic problem. (5)

Moreover, intravenous anaesthesia nowadays has become a very attractive option for a large part of routine paediatric anaesthesia practice, (6,7) because of its rapid onset of action, improved quality of emergence, reliable administration in patients undergoing airway procedures, as well as its reduction of postoperative nausea and vomiting (PONV), and all these items were very important in this child with KGS and SDB. According to some authors, the i.v. technique should be the technique of choice for tonsillectomy and squint surgery. TIVA helps to keep the incidence of PONV low, allowing a peaceful and rapid recovery, thus significantly increasing parental satisfaction and improving quality of care. (7)

In conclusion, intravenous anaesthesia for adenoidectomy could be safely and routinely performed in small children, even in situations that could jeopardize airway maintenance, i.e. KGS and SDB. Comprehensive pre-anaesthetic preparation is vital in a child with KGS, i.e. postural drainage, bronchodilators, i.v. antibiotics and steroids. Adequate parental education in performing postural drainage manoeuvres is of paramount importance. The condition of the respiratory system in the patient with KGS is seldom appropriate at the time of surgery. Preoperative clinical, laboratory and radiology findings are often borderline and the decision to anaesthetize or not, should be made on an individual basis. Therefore, we recommend more flexible criteria for patient-readiness in KGS, especially if combined with SDB. Adenoidectomy provides great benefit and should be performed in these patients as needed, perhaps in an urgent fashion.

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