J Anesth (2009) 23:456–459
DOI 10.1007/s00540-009-0779-7

Journal of
Anesthesia

Anesthetic management of a child with Langer-Giedion (TRPS II) syndrome

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Abstract
We describe the anesthetic and perioperative management of a child with Langer-Giedion syndrome (trichorhinophalangeal syndrome type II). This is a very rare genetic syndrome caused by 8q chromosome deletion. The clinical features of this syndrome include craniofacial and urogenital abnormities, variable postnatal growth deficiency with mental retardation, multiple exostoses, hyperflexible joints, and recurrent respiratory tract infections. Potential perioperative problems are highlighted.

Key words TRPS · Langer-Giedion syndrome · Dental anesthesia

Introduction

Trichorhinophalangeal syndromes (TRPS) were first described by Giedion in 1967 [1]. They are very rare inherited multisystemic disorders caused by chromosome 8 deletion or microdeletion. TRPS are characterized by unique facial features and skeletal abnormities (see Table 1 for features associated with various types of TRPS). Individuals with TRPS have an increased susceptibility to recurrent respiratory and urinary infections [2]. In addition to these common features, type II (Langer-Giedion syndrome; LGS) is usually associated with microcephaly, various levels of mental retardation, laxity or hypermobility of all joints, multiple exostoses, and redundant or loose skin [3].

Because of the multiple clinical abnormalities, these patients present frequently for surgical and interventional procedures [4]. We have identified, through a search in the MEDLINE and SCOPUS databases, only one report describing the anesthetic management of a patient with TRPS type I [2]. This report represents the first describing the specific anesthetic management in a patient with LGS.

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Received: February 27, 2009 / Accepted: April 16, 2009

Case report

A 8-year-old boy with TRPS type II (Langer-Giedion) syndrome was scheduled for multiple dental extraction of deciduous teeth under general anesthesia as a day-surgery patient. His past medical history included asthma, with repeated upper respiratory tract infections and frequent urinary tract infections.

He was of small stature with height between the 2nd and 9th centiles on the centile chart, and weighed 16 kg (between the 0.4th and 2nd centiles). He had a prominent and bulbous nose, slightly low-set ears, small mandible, fine hair, and thin upper lip (Fig. 1). Other features included hypermobile joints, including the cervical spine; brittle nails; multiple exostoses; and redundant skin, mainly on his upper extremities.

The patient was extremely anxious and uncooperative, and full assessment of his airway was impossible. With the parents' consent the patient was given a premedication of 10 mg temazepam liquid. Within 30 min he arrived appropriately premedicated to the anesthetic room. After connection for noninvasive blood pressure, ECG, and pulse oximetry, anesthesia was induced by inhalation of a 6% concentration of sevoflurane in a mixture of O_2/N_2O. Venous access on the back of the hand was avoided because of redundant creased skin in that area. A 22-g cannula was inserted on his forefoot. After manual in line stabilization laryngoscopy was performed and a grade I view was achieved with a small epiglottis visualized, a flexible disposable laryngeal mask (FLMA) size 2.5 (Intavent Orthofix, Maidenhead, UK) was inserted easily, and after securing the FLMA, a gauze throat pack was inserted. A view obtained through the FLMA with a pediatric bronchoscope showed clearly visible vocal cords from the middle of

Table 1. Features associated with various types of TRPS and differential diagnosis

Clinical features	TPRS I	TPRS II	TPRS III
Craniofacial abnormalities			
— Sparse scalp hair	+	+	+
— Large, laterally protruding ears	+	+	–
— Bulbous nose	+	+	+
— Prominent elongated philtrum	+	+	+
— Thin upper lip	+	+	+
— Apparent mandibular micrognathia	+	+	–
— Microcephaly	–	+	–
— Aplasia of epiglottis	–	1	–
— High arched palate	+	+	+
— Cleft palate	–	1	–
— Dental abnormalities (malocclusion)	+	+	+
Orthopedic manifestations			
— Clino-brachydactyly	+	+	+
— Cone-shaped epiphyses (hands)	+	+	+
— Winged scapulae	+	+	+
— Short stature	+	+	+
— Multiple exostoses, bone fractures	–	+	–
— Laxity or hypermobility of joints	+	+	–
— Limited movements of joints	–	–	+
— Loose or redundant skin	–	+	–
Mental status			
— Mental retardation	–	+	–
— Delayed onset of speech	–	+	–
— Psychomotor lability	–	+	–
Other			
— Skin nevi	–	+	–
— Recurrent pulmonary infections	+	+	–
— Ureteral reflux, persistent cloaca, congenital nephrotic syndrome	+	+	–

+, features generally present; –, features generally absent; 1, features described in a single case report

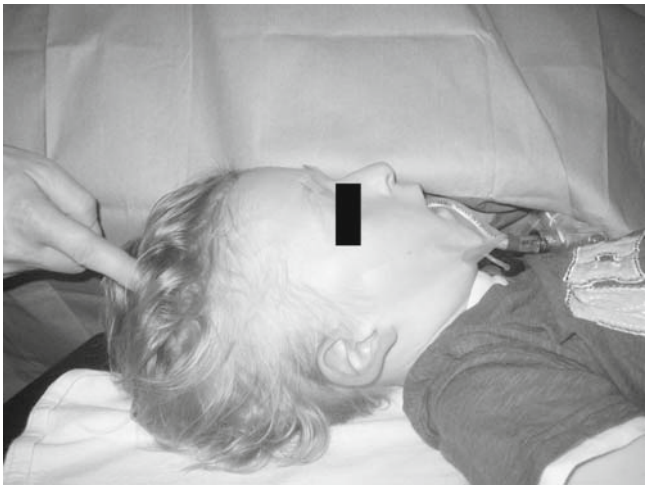

Fig. 1. The patient with Langer-Giedion syndrome. Craniofacial features—bulbous nose, small mandible, low-set ears, and fine hair—are present

the distal end of the FLMA. Anesthesia was maintained with 2.0% end-tidal sevoflurane with the patient breathing spontaneously. The dental procedure continued with eight extractions and took 30 min. After the throat pack was removed the patient's oral cavity was examined with the laryngoscope and all blood and saliva was suctioned out. The patient emerged from anesthesia within 5 min and the laryngeal mask was removed. After a brief period of agitation during emergence the patient recovered uneventfully and after fulfilling the necessary criteria was discharged from the day surgical unit later that evening.

Discussion

Trichorhinophalangeal syndromes are subclassified into three types depending on the pattern of mutation of the *TRPS 1* gene on chromosome 8 [5]. The severity and pattern of gene deletion in LGS accounts for the additional clinical manifestations described below. Inheritance appears to be autosomal dominant; however, various patterns have been described [6]. As there have been only a few hundred cases identified, the incidence of LGS is difficult to describe [7,8].

Building on the previous work of Graybeal et al. [2], who described perioperative care for a teenage patient with TRPS I, we would like to emphasise that patients

with LGS may present many additional challenges for anesthesia. In comparison to patients with other TRPS subtypes, patients with LGS usually have various levels of mental deficiency. They are often withdrawn and can be uncooperative or may present with aberrant behavior. Behavior modification through desensitization or cognitive-behavioral therapy allays anxiety and is important before the child presents to hospital. Our patient presented with a short period of agitation in recovery which could be attenuated by sedatives or analgesics. On the other hand, patients after dental procedures should be fully alert and able to protect their airway against aspiration of blood. This is usually a reason why postoperative sedation is avoided [9].

Hearing impairment and delayed speech development can also complicate preoperative assessment and the development of a rapport with these patients [8]. It is important that these patients are made as comfortable as possible, and generous premedication is usually necessary.

Although full preoperative airway assessment may be difficult in LGS patients, a difficult airway scenario should be preempted. These patients commonly have craniofacial abnormalities and they usually have some degree of microcephaly. Common facial features include protruding upper lip, atypically prominent maxilla, dental anomalies, recessive mandible, and bulbous nose. This can make mask ventilation and/or tracheal intubation difficult. Additionally the alae nasi may be hypoplastic, complicating or even contraindicating nasotracheal intubation [7]. Other airway anomalies that have been associated with the syndrome include epiglottic aplasia and submucous cleft palate [10]. These anomalies may influence the mode of induction and airway establishment; so that difficult airway plans should be made. For short procedures, a flexible laryngeal mask airway (FLMA) may be an appropriate airway option [11]. There may be some arguments against the use of a laryngeal mask in dental anesthesia—risk of malposition, obstruction, or aspiration—but many authors support its insertion in these procedures [11,12]. We have had experience with laryngeal masks in more than 1100 pediatric dental patients, without any serious complications and with a high satisfaction rate on the side of dental surgeons. For longer procedures with a difficult laryngoscopic view, even fiberoptic intubation through a supraglottic airway may be indicated [13].

The skin in LGS patients has been described as elastic and similar to that in individuals with Ehlers-Danlos syndrome. There may be redundant skin overlying the backs of the hands and feet, which can produce difficulties for intravenous cannulation [8].

Because of the hypermobility of the cervical spine in LGS patients there is a theoretical risk of luxation or subluxation and subsequent spinal cord injury. It is imperative that the cervical spine should be stabilized during intubation, and excessive neck extension should be avoided.

These patients have an increased susceptibility to respiratory tract infections and it is important that these infections be excluded in the preoperative assessment [8]. Surgery may need to be postponed to allow for optimization of respiratory function. Perioperative prophylactic antibiotics may be required to cover respiratory or urogenital infections.

There is also a high incidence of vertebral segmentation defects and scoliosis. Additionally the joints are lax and there are multiple exostoses. Spontaneous bone fractures have been described in these patients [14,15]. It is very important that attention be paid to careful positioning and padding of pressure areas.

After extensive literature review we cannot identify whether these patients have any cardiac abnormities, but one must have a high index of suspicion when there are concerns about cardiac function.

Although the case we have reported was uncomplicated from an anesthetic point of view, we have pointed out potential problems that may be encountered when managing any patient with Langer-Giedion Syndrome (LGS). When managing these patients it is important that good perioperative plans are in place to deal with potential problems. In particular, the patient and family should be informed of the increased risk and they should be involved in any decision-making processes.

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