

Anaesthesia for a child with cleidocranial dysplasia

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Key points

Cleidocranial dysplasia (CCD) is a rare autosomal dominant disorder with generalized skeletal dysplasia resulting in abnormal development of skull, clavicles, teeth and pelvis (1). We present a case report of an 11-year-old girl undergoing multiple teeth extractions under general anaesthesia.

Abstract

We present a case of a child with cleidocranial dysplasia undergoing multiple teeth extractions. Characteristics of this genetic disorder and implications for the paediatric anaesthetist are discussed.

Keywords: cleidocranial dysplasia; Marie-Sainton Syndrome

Case presentation

The patient presented with a short stature and dysmorphic features. Past medical history was unremarkable, there were no previous anaesthetics. Diagnosis of CCD was made at birth. Family history of the maternal and paternal side was negative for any genetic disorder. Physical examination revealed brachycephaly with frontal bossing, large nasal bridge, hypertelorism, high-arched palate, micrognathia, presence of supernumerary teeth and brachydactyly with cup-shaped distal phalanges and down-curving nails. Hypoplasia of both clavicles was described by chest x-ray and the girl was able to touch her shoulders together in front of the chest. X-ray of the spinal column revealed schisis of thoracic vertebra T1 to T4. Radiologic findings of the skull included agenesis of the

frontal sinus and lack of pneumatization of mastoid cells. A dental scan showed delayed eruption of permanent teeth, presence of supernumerary deciduous and permanent teeth within the dental arch and malocclusion. Height and weight were below the fifth percentile. Assessment of respiratory function by auscultation and chest x-ray was normal, saturation was 98% in room air. Clinical evaluation of cardiac function by ECG and auscultation showed no abnormalities. Neurological assessment revealed generalized joint laxity and hypermobility, but normal muscle tone and a mental status appropriate for age. A full blood count and coagulation profile were within normal ranges. Biochemistry revealed normal electrolytes, normal renal and liver function, but an elevated CPK value of 238 U/L.

Surgery for multiple teeth extraction was scheduled and a difficult airway was anticipated. The child was starved according hospital guidelines and Emla® cream was applied on the dorsum of both hands. A 22 G iv cannula was positioned on the dorsum of the right hand. Routine monitoring was applied and propofol 2 mg/kg were slowly injected. Anaesthesia was deepened with the

child spontaneously breathing sevoflurane 8% in 100% oxygen. A Guedel airway was inserted to facilitate mask ventilation. Once anaesthesia was sufficiently deepened, laryngoscopy was performed with a Miller blade size 2 and a Cormack / Lehane grade III was diagnosed. With application of cricoid pressure a Cormack / Lehane view grade II could be obtained and topical anaesthesia (1 ml of 2% lidocaine) was nebulized with a laryngo-tracheal atomizer (MADgic®, Wolfe Tory Medical Inc) on the vocal cords. At the same time topical anaesthesia with 2% mepivacaine and epinephrine 1:200000 was applied as nasal packing to the right nostril. A lubricated preformed 5.5 mm nasal-endotracheal tube was gently pushed from the right nostril into the hypopharynx and then with help of a Magill forceps directed through the glottis into the trachea. Anaesthesia was maintained with sevoflurane 2% in 50% oxygen and nitrous oxide. An infusion of remifentanyl 0.2 mcg/kg was started and prophylaxis for PONV with ondansetron 0.1 mg/kg and dexamethasone 0.1 mg/kg was given. Local anaesthesia was performed with 2 mg/kg of 2% mepivacaine and adrenaline 1:200000. Postoperative analgesia was provided by intravenous paracetamol 15 mg/kg, ketoprofen 1 mg/kg and morphine 1 mg. Duration of surgery was 60 minutes and emergence from anaesthesia was smooth. The postoperative course was uneventful and the child was discharged home after 24 hours.

Discussion

Cleidocranial dysplasia has also been called cleidocranial dysostosis or Marie-Sainton Syndrome and was first described in 1765 by Martin (2) in a patient with absent clavicle. More than 500 cases have been reported so far worldwide and there is a considerable variability of genetic expression. The responsible gene has been mapped to 6p21(3). Initially it was believed that the disease affects only membranous bones such as neurocranium, clavicle and some facial bones, but it has now been classified as a general skeletal dysplasia (1).

Diagnosis is often made at birth or shortly thereafter when permanence of a widely open anterior fontanel and sutures or short stature cause parental concern. Radiological findings of skull, dental, pelvic and clavicular malformations together with genetic mapping confirm the diagnosis. Other than the abovementioned characteristic findings include cleft palate, deafness, a narrow, bell-shaped thorax, narrow pelvis with wide symphysis pubis and spina bifida occulta (4).

Anaesthetic considerations: the narrow thorax may cause restrictive lung disease and lead to respiratory failure in early infancy and development of right heart failure. Preoperative assessment of respiratory and cardiac function is mandatory. Facial dysmorphism together with micrognathia, abnormal shape of the palate and fragile teeth should anticipate a difficult airway management. The patient should be positioned very carefully with regard to joint laxity and tendency for joint dislocations. Spinal and vertebral abnormalities should be evaluated before considering neuraxial blockade (1). A recent case report on a woman with CDC undergoing four caesarian sections reported a safe and effective use of neuraxial anaesthesia (5).

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