General anaesthesia with and without intubation for patients with Cornelia de Lange syndrome

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Keywords: general anaesthesia; Cornelia de Lange syndrome; dental treatment

SAJAA 2009; 15(4): 32-34

ABSTRACT

We present the use of different methods of general anaesthesia in two patients with Cornelia de Lange syndrome and its contribution to the patients' oral health.

Case 1: The patient was a 22-year-old woman with Cornelia de Lange syndrome who underwent dental treatment under general anaesthesia. She exhibited the physical characteristics of Cornelia de Lange syndrome, including a small mouth, thin lips, short limbs, stiffness of joints and intellectual disability. General anaesthesia without intubation was performed safely eight times. No other complications except hypersensitivity to hypnotic agents were observed.

Case 2: The patient was a 10-year-old boy with Cornelia de Lange syndrome who underwent dental treatment under general anaesthesia. He had a history and symptoms of obstructive airway disorders in addition to showing physical characteristics of the syndrome similar to those seen in Case 1. General anaesthesia with nasal intubation was performed safely twice. Computed tomography (CT) of his head and neck produced unremarkable results.

These cases demonstrate that both general anaesthesia with and without nasal intubation can be safely used in managing individuals with Cornelia de Lange syndrome during dental treatment.

℗ Peer reviewed (Submitted: 2009-05-22, Accepted: 2009-08-13)

Introduction

Cornelia de Lange syndrome (CdLS) is a rare multiple congenital disorder. Patients with this syndrome are characterised by intellectual disorder, hirsutism and multiple anomalies in the cardiovascular and musculoskeletal systems.[1] Its incidence has been reported to be between one in 30 000 and one in 60 000 live births. Most children die within the first two years of life, primarily due to pneumonia along with cardiovascular, respiratory and gastrointestinal disorders.[2,3] Dental patients with this syndrome might require recurrent general anaesthesia because dental treatment must be performed repeatedly in many cases. Previous reports discuss the issues that arise when using general anaesthesia in patients with CdLS, such as intubation difficulty, pericarditis and oesophageal reflux.[4–8] However, recurrent anaesthesia including general anaesthesia without intubation[9] has not been discussed in these reports. The present article describes two patients with CdLS who received dental treatment under general anaesthesia with and without intubation. The results illustrate the effectiveness of repeated administration of general anaesthesia and the contribution of both methods of general anaesthesia to the patients' oral health.

Case reports

Case 1

A 22-year-old woman with CdLS was scheduled for dental treatment under general anaesthesia. The patient had been diagnosed with CdLS soon after birth after she had failed to progress normally and it became evident by her physical anomalies and intellectual disorder. She was first referred to our hospital in 2005 complaining of toothache caused by a cavity. Several attempts to familiarise the patient with various procedures used to facilitate dental treatment failed. Therefore, we opted for general anaesthesia to resolve the issue of her oral health. Her mother accepted our proposal and agreed to treatment under general anaesthesia.

The patient was 121 cm tall and weighed 35.1 kg. She had characteristics of CdLS, including a small mouth, thin lips, short limbs, stiffness of joints and intellectual disability. We succeeded in taking a blood sample, chest X-ray and electrocardiograph, whose results were within normal limits. There was no evidence of abnormality in her internal organs including the heart.

The patient was anaesthetised for the first time in August 2005. Before general anaesthesia we prepared for tracheostomy in case of an emergency. The patient cried in the waiting room and vehemently refused to enter the examination room. She became somnolent 10 min after 7.5 mg midazolam was orally administered in 50 ml clear fruit juice (apple) by a qualified dentist-anaesthetist*, after which he secured an intravenous route. (*A dentist-anaesthesiologist is registered to perform general anaesthesia for dental treatment and oral surgery in Japan.)

Anaesthesia was induced with a bolus administration of 0.4 mg atropine sulphate and 35 mg propofol, and was then maintained by a continuous infusion of propofol. Blood pressures were stabilised in 110-80/70-50 mmHg range, and heart rates between 100 and 90 beats.min^{-1} throughout anaesthesia. Signs of airway obstruction were not observed during induction. The dentist-anaesthetist inserted a tracheal tube via the left nostril and set its tip at a point two cm from the tip of the epiglottis. When the dentist-anaesthetist attempted to open her mouth for a laryngoscopy to ascertain the tip of the tube, the maximum distance between the upper and lower incisors was 26 mm. The dental hygienist maintained proper head tilt and forward jaw thrust to ensure an open airway throughout the period of anaesthesia. Thoracic movement, pulse oximetry and laryngeal auscultation were monitored to observe respiration. Starting with an initial propofol infusion rate of 8 mg.kg^{-1}.h^{-1}, the dentist-anaesthetist titrated the anaesthetic agent to provide an adequate level of anaesthesia. The infusion rates of propofol were 6-8 mg.kg^{-1}.h^{-1}. The patient stopped breathing and ventilation with a mask was provided for about 30 s when 20 mg of propofol was administered in a bolus during the steady state of anaesthesia. Another dentist performed the tooth extraction and filling of composite resin. The patient started responding to eyelash stimulation 15 min after the end of propofol infusion and spent two hours in the ward under the observation of her mother and nurses until discharge from the hospital after lunch. Since her first treatment under general anaesthesia, we were able to anaesthetise her successfully seven times. After anaesthetising her for the fourth time, we reverted to nitrous oxide gas instead of midazolam because of the effectiveness of the gas. Using nitrous oxide gas as a sedative agent, the dentist-anaesthetist could secure an intravenous route.

Case 2

A 10-year-old boy with CdLS was scheduled for dental treatment under general anaesthesia. In addition to the usual characteristics of CdLS as observed in Case 1, he had micrognathia with obstructive sleep apnoea and cleft palate. He had received a push-back operation to close the cleft in the palate in a hospital when he was three years old. After the operation, the plastic surgeon in charge informed the patient's parents that intubation had been very difficult because of micromandible and deformity of the trachea during anaesthetic induction. However, they did not understand the reason behind the deformity in their son's jaws and trachea and why anaesthetic management was difficult. Thereafter, his growth had been followed-up by the plastic surgeon, who indicated the possibility of sleep apnoea syndrome. The patient had refused oral care by his parents and was referred to our hospital in 2008 complaining of gingival abscess in the mandible caused by cavities. He refused to open his mouth so aggressively that the dentist in charge decided to opt for dental treatment under general anaesthesia.

The patient was 84 cm tall and weighed 17 kg. He had characteristics of CdLS similar to those in Case 1. Results of a blood sample, chest X-ray and electrocardiography were within normal limits. The maximum distance between the upper and lower incisors was estimated to be more than 30 mm when he was crying. The airway was graded as Mallampati class 2. He had several cavities that might have caused gingival abscess. The plastic surgeon showed us the anaesthetic record of the push-back operation, but a detailed description of the difficult intubation had not been documented.

The boy was anaesthetised for the first time by an anaesthesiologist and a dentist-anaesthetist in the operating theatre in October 2008. An intravenous route was secured in the right hand by a paediatrician. General anaesthesia was induced by a bolus administration of 100 mg thiamylal sodium. Immediately after mask ventilation had been established, two mg of vecuronium bromide was infused and the airway was easily secured. We prepared a fibre-optic bronchoscope and optical laryngoscopes. However, nasal tracheal intubation was smoothly performed when two per cent sevoflurane and remifentanil 0.2 mg.h^{-1} (0.196 mcg kg^{-1}.min^{-1}) were administered. The maximum distance between the upper and lower incisors was 31 mm. Anaesthesia was maintained at a rate of 0.2–0.3 mg.h^{-1} (0.196 -0.294 mcg kg^{-1}.min^{-1}) of remifentanil, two per cent sevoflurane and 50% nitrous oxide gas mixed with oxygen. Ventilation was controlled throughout the operation. Blood pressures were controlled in 130-90/100-80 mmHg during induction and were stabilised in 100-80/60-50 mmHg range, and heart rates between 70 and 80 beats.min^{-1} intraoperatively. An oral surgeon extracted two teeth. After the treatment, the boy was sent to the X-ray room where a computed tomography (CT) scan was performed under general anaesthesia with propofol 80–40 mg.h^{-1} and remifentanil 0.15 mg.h^{-1} (0.147 mcg.kg^{-1}.min^{-1}). A radiologist diagnosed that the patient had no abnormality from the temporomandibular joints to the trachea. The patient was then returned to the operating theatre and the administration of propofol and remifentanil was discontinued when 0.5 mg atropine sulphate and 1.0 mg neostigmine were administered. Approximately 20 min later, he awoke almost perfectly and the anaesthesiologist extracted the tracheal tube after checking for sufficient recovery. The patient spent two hours in the ward under the observation of his parents and nurses before he was discharged from the hospital.

Dental treatment under general anaesthesia was performed safely one month later. Spontaneous respiration under sedation was established when anaesthesia was induced with a bolus administration of 20 mg of propopfol. Should the patient require dental treatment in future, general anaesthesia without intubation will be performed.

Discussion

Anomalies in the head and neck of patients with CdLS, such as micrognathia, small mouth, a high arched palate, a short neck and stiffness of the temporomandibular joints and neck, have made laryngoscopy for tracheal intubation difficult. Some anaesthetists have overcome these difficulties with special techniques or instruments such as blind nasotracheal intubation and tracheal intubation through a laryngeal mask airway with a fibre-optic bronchoscope.[4,10] In Case 2, we also prepared these instruments for the first anaesthesia and investigated anomalies of the patient's head and neck using CT scan to prepare for the next anaesthesia. In Case 1, general anaesthesia without intubation was successfully performed eight times over a period of approximately three years. However, we were concerned that tracheal intubation might prove difficult in the case of an emergency as a result of the physical anomalies present in individuals with CdLS. Therefore, we prepared for a possible tracheostomy. If possible, general anaesthesia should be non-invasive and effective, particularly in dental treatment for individuals with challenging disorders. The use of general anaesthesia without intubation for dental treatment has the advantages that the patients are free from the risk of anaesthetic problems, such as intubation difficulties, laryngeal oedema and injuries to teeth. We have also used it in our examination room since 2001[11] but have limited its use in our hospital to patients who do not suffer from serious heart diseases or respiratory disorders because of the uncertainty of airway management. The patients described in the present report had no heart disorders and hence we could perform pre-anaesthetic tests despite their intellectual disability. Moreover, in Case 1, the patient did not exhibit any symptoms of obstructive airway disorders and we had a successful experience using general anaesthesia without intubation, a case similar to that of a woman with Rett syndrome.[12] In Case 2, because the patient had a history and symptoms of obstructive airway disorders we were worried that tracheal intubation might prove difficult. Therefore, general anaesthesia with intubation was chosen.

Excluding anomalies of the airway, anaesthetic problems in patients with CdLS include heart disease, aspiration, poor communication and hypersensitivity to drugs.[13] In Case 1, the woman also exhibited sensitivity to midazolam, propofol and nitrous oxide gas. The amounts of midazolam and propofol administered did not exceed those given to other handicapped patients in our hospital. Moreover, we have not encountered any other patients who exhibited the same level of sensitivity to nitrous oxide gas as observed in Case 1. The cause of hypersensitiveness to hypnotic agents is unknown. Therefore, anaesthetists must also be cautious to the amounts of drugs administered when treating patients with challenging disorders.

In conclusion, we safely performed dental treatment repeatedly in two patients with CdLS under general anaesthesia with and without intubation. Anaesthetists can use both methods of anaesthesia for handicapped patients because of its simplicity, effectiveness and optimal health benefits. SAJAA

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