Translational Perioperative and Pain Medicine

ISSN: 2330-4871

Case Report | Open Access Volume 11 | Issue 3

Anesthetic Management for Dental Procedures and Oral Surgery in a Patient with Cornelia de Lange Syndrome: A Case Report

Akkan Mahmud*, Dr. Ahmed Uslu and Prof. Nedim Çekmen

Department of Anesthesiology and Intensive Care, Faculty of Medicine, Baskent University, Ankara, Turkey

Abstract

Cornelia de Lange syndrome (CdLS) is a rare genetic disease of multiple malformations affecting the craniofacial, musculoskeletal, neurosensory, gastrointestinal, central nervous, cardiovascular, and genitourinary systems. CdLS patients may often require more than one high-risk anesthesia procedure, and due to the difficulties encountered in this syndrome, it is challenging for the anesthesiologist and important strategies are required in order to be successful in anesthesia management. We present the report on the successful anesthetic management of a 21-year-old woman with CdLS who underwent extensive dental procedures and oral surgery. The procedure was performed under general anesthesia by inhaling volatile gases due to the lack of intravenous (IV) access. We applied the rapid sequence induction and intubation (RSII) method with cricoid pressure to minimize the possibility of pulmonary aspiration due to the patient's difficulty mask ventilation and intubation, irritable airways, short neck, micrognathia, and poor cooperation, and she was intubated without complications. Due to developmental, mental, and motor delays in this syndrome, perioperative management requires comprehensive preoperative evaluation and a multidisciplinary team approach.

Keywords

Cornelia de lange syndrome, General anesthesia, Rapid sequence induction and intubation, Difficult airway, Videolaryngoscopy, Dental procedures

Introduction

Cornelia de Lange syndrome (CdLS) is a rare genetic disease of multiple malformations affecting the craniofacial, musculoskeletal, neurosensory, gastrointestinal, central nervous, cardiovascular (congenital heart disease), and genitourinary systems [1]. The etiology of CdLS is genetically linked to a cohesinopathy disorder caused by X-linked or autosomal heterozygous mutations in the cohesion core subunits of the SMCA1, SMC3, RAD21 genes or the cohesion-related factors NIPBL and HDAC8. The most common craniofacial and distinctive facial features include microcephaly, microbrachycephaly, short neck with receding hairline anteriorly and posteriorly, sinophris, long curly eyelashes, smooth and well-defined eyebrows, micrognathia, macroglossia, "carp-shaped" mouth, high-arched palate, downturned angles of the mouth, a long philtrum, low-set ears, thin lips, small and upturned nose, microdontia, delayed tooth eruption, partial anodontia growth, mental retardation, delayed skeletal maturation, upper extremity anomalies, and abnormal thoracic configuration [1-3]. Gastroesophageal dysfunction, along with reflux, esophagitis, and aspiration pneumonia, have also been described in CdLS [3-5]. It is especially difficult to anesthetize CdLS patients because it involves the risk of poor cooperation, difficult mask ventilation and intubation, irritable airways, gastroesophageal reflux, and aspiration pneumonia [6]. Mask ventilation and endotracheal intubation can be difficult due to cleft palate, micrognathia, short neck, collapse of the nasal bridge, and developmental disorders. Therefore, even during mild anesthesia, upper airway obstruction, bronchospasm, desaturation, and hypoxemia may occur in these patients [1-3,5]. Patients with CdLS often present with unique challenges in medical and dental care due to their physical, behavioral, cognitive, and emotional impairments [4].

Herein, we present the report on the successful anesthetic management of a 21-year-old woman with CdLS who underwent extensive dental procedures and oral surgery.

Case Presentation

Preoperative assessment

A 21-year-old female patient diagnosed with CdLS was scheduled for dental procedures and oral surgery under general anesthesia. The patient, who was diagnosed with CdLS at the age of 3, was evaluated by many disciplines before the surgical intervention. She had a history that included esophagitis, gastroesophageal reflux, recurrent aspiration pneumonia, and, in conjunction with findings of autonomic dysfunction, short neck, micrognathia, pronounced incisors, poor cooperation,

severe developmental delays, mental retardation, and motor dysfunction, presenting significant challenges for cooperation and management. Despite this, the patient was not under any kind of treatment. Her vital signs, laboratory results, electrocardiogram, and X-ray were within normal limits. Local ethics committee approval and the patient's caregiver written consent were obtained to publish this case report.

Anesthetic management

The patient was evaluated preoperatively with a comprehensive multidisciplinary approach. Because of the patient's mental state and poor cooperation, oropharyngeal examination and Mallampati airway scoring were unsuccessful, and the patient's American Society of Anesthesiologists (ASA) physical classification was class II.

In the preoperative evaluation, difficulty with ventilation and intubation was expected. Therefore, we planned to manage the airway as difficult intubation. Our routine airway management equipment included the supraglottic airway (SGA), ProSeal laryngeal mask airway (PLMA), the oesophageal tracheal combi-tube (OTC), the laryngeal tube with integrated suctioning tube (LTS), the laryngeal tube (LT), and tube stylet, gum bougie, direct laryngoscope, video laryngoscope, and fiberoptic bronchoscope, and we prepared an emergency tracheostomy and cricothyrotomy set in case of unsuccessful intubation.

Premedication was not administered due to poor cooperation. The patient underwent standard monitoring in the operating room, including electrocardiogram, noninvasive blood pressure (BP) cuff, pulse oximetry, respiratory rate, and bispectral index. We applied forced-air warming, heated blankets, and heated intravenous (IV) solutions to prevent hypothermia. Before induction, we pre-oxygenated the patient with a nasal cannula as a mixture of 80% inspired oxygen fraction (FiO_2) and 20% air at a rate of 8 L/min. Due to the patient's lack of cooperation, IV access could not be established preoperatively. Anesthesia induction was therefore performed using 8% sevoflurane and 80% FiO_2. After much difficulty and sedation was achieved, a 20-gauge cannula was inserted. During anesthesia induction, lidocaine 1 mg/kg, fentanyl 1 µg/kg, propofol 2 mg/kg, and rocuronium 0.6 mg/kg were administered intravenously. The patient was ventilated with 80% FiO_2 for 1 minute using a bag mask. We applied the RSII method with cricoid pressure to minimize the possibility of pulmonary aspiration due to the patient's difficulty masking ventilation and intubation, irritable airways, short neck, micrognathia, and poor cooperation. The patient was intubated with a 7.0 mm endotracheal tube (ETT) without complications in the first application using the Video-laryngoscope (Figure 1 and Figure 2). Video laryngoscopy revealed Cormack-Lehane Grade IV. Anesthesia maintenance was provided with 2.2% sevoflurane and 50% oxygen-50% air mixture, 0.01 to 0.05 µg/kg/min infusion of remifentanil.

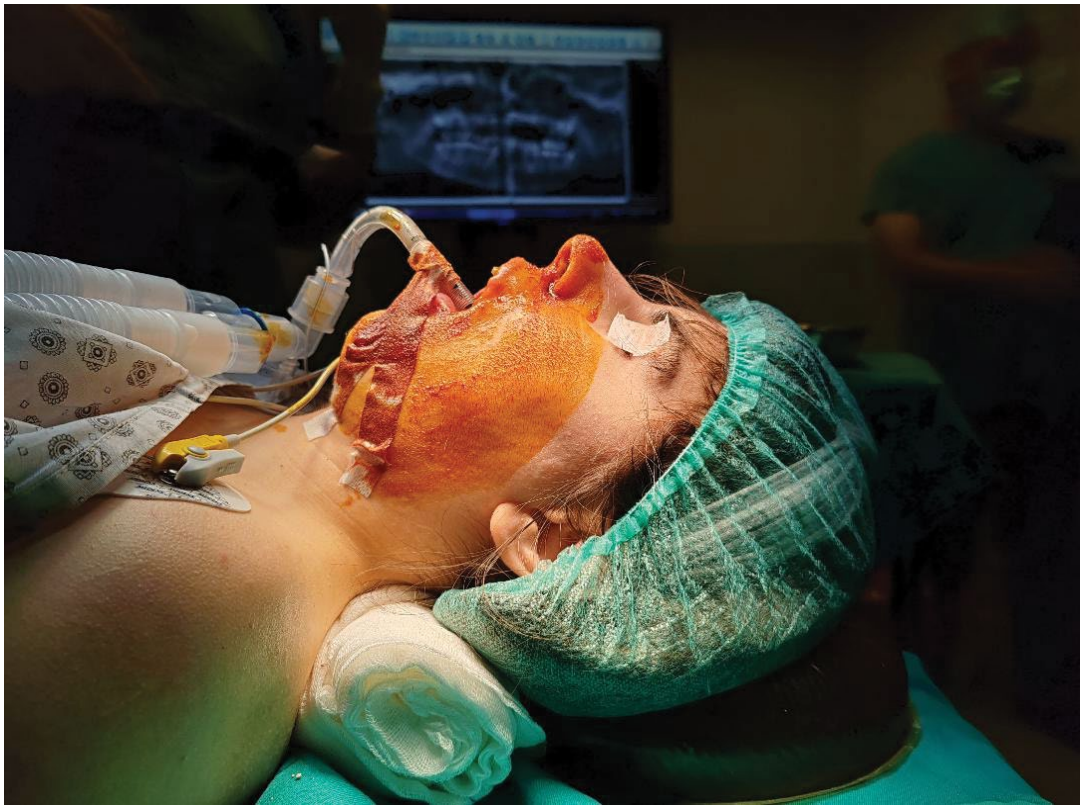

Figure 1: Intubated lateral view of the patient with CdLS.

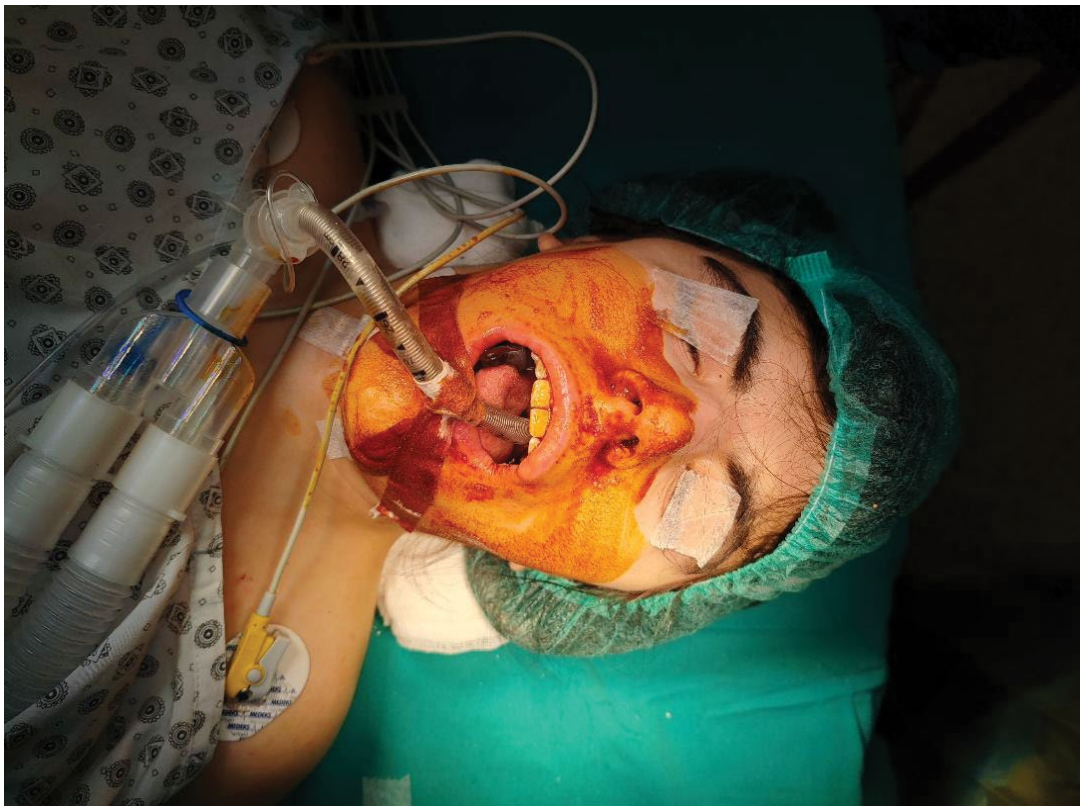

Figure 2: Intubated frontal view of the patient with CdLS.

We administered methylprednisolone 1 mg/kg IV before surgery as an anti-edema premedication. We administered cefazolin 1000 mg IV prophylactically. The patient underwent multiple dental fillings, dental detartrate, tooth extractions, and surgical removal of a mandibular dental cyst. During the surgery, adequate hydration was maintained with a balanced electrolyte solution (1000 mL), lactated ringer solution (1000 mL), and hydroxyethyl starch 250 mL. The procedure lasted for two hours, and after hemostasis control, the surgery was completed without any complications, such as bleeding or respiratory problems. The patient's vital signs remained stable under close monitoring. Analgesia was achieved with paracetamol 10 mg/kg IV infusion at the beginning and end of the procedure, supplemented by 1 mg/kg IV morphine. Neuromuscular blockade was reversed by administering sugammadex 2 mg/kg, after which spontaneous breathing returned 4 minutes after discontinuing sevoflurane. The patient was extubated and transferred to the postanaesthesia care unit (PACU). The patient was discharged from the hospital one day later.

Discussion

Clinical and physical abnormalities and associated problems in patients with CdLS are extremely challenging for anesthesiologists, especially in uncooperative patients. Managing patients with CdLS undergoing surgery requires meticulous preoperative planning and preparation, and a comprehensive multidisciplinary approach is crucial. The RSII method with a video laryngoscope was useful in minimizing potential complications. It is important to examine these patients carefully and recognize the difficult airway features in the preoperative period as they may present with various physical abnormalities that lead to difficult intubation [2,3,6]. Careful monitoring of possible intraoperative blood loss and anesthesia management is crucial in ensuring hemodynamic stability. It is important to manage pain and try to prevent complications such as tissue edema in postoperative care.

Anesthesia management in patients with CdLS can pose serious challenges, mostly due to difficult airway and tracheal intubation and aspiration complications [1,3,4]. Gastroesophageal dysfunction is also common in this syndrome, with risks of reflux, esophagitis, and aspiration pneumonia [5]. We placed a nasogastric tube before induction and applied cricoid pressure by administering a high dose of rocuronium due to the risk of aspiration. Therefore, by applying the RSII method to minimize the risk of aspiration and appropriate preoperative preparation for the difficult airway, the correct intraoperative management was adapted to address these special concerns, and a smooth perioperative process was achieved. In the preoperative assessment, due to the patient's short neck and micrognathia, we prepared an intubation cart containing masks of different size, PLMA, SGAs, LT, LTS,

OTC, a tube stylet, a gum bougie, a video laryngoscope and fiberoptic bronchoscope to avoid the possibility of difficult intubation and we prepared an emergency tracheostomy and cricothyrotomy set in case of unsuccessful intubation. We intubated our patient without any problems.

When the literature on airway management in CdLS is examined, it is seen that there are cases with difficult airway and intubation, and they reported that the cause is the characteristic facial appearance found in CdLS, limited mouth opening, short, hard neck, micrognathia, high arch, and cleft palate. Due to the coexistence of mental retardation, poor cooperation, hyperactive or autistic behavior, and neurosensory auditory disorders in patients with CdLS, many difficulties may be experienced in pre-anesthesia evaluation [4,6]. Therefore, routine airway examination is often impossible, and we cannot perform an airway examination on our patients. In these patients, it often causes crowding due to missing teeth, and the upper central incisors are usually prominent, making it difficult to place the laryngeal mask because tooth decay and bruxism are common. A safe airway may not always be provided with a laryngeal mask in these patients due to gastroesophageal reflux and aspiration risk [1,3,5,6]. As this was a dental procedure we did not use an LMA.

Torres MD, et al. [1] reported that a 34-year-old patient with CdLS underwent an orthopedic procedure due to severe scoliosis, and joint contractures. Moreover, the airway was not evaluated properly before surgery, and intubation was performed with an awake fiber optic bronchoscope under light sedation after three unsuccessful attempts. Sugiyama, et al. [7] reported that they could not intubate a 7-year-old boy undergoing bilateral orchidopexy with direct laryngoscopy due to limited neck extension after slow induction of anesthesia with sevoflurane inhalation and IV rocuronium administration, after which he was intubated using an Airway Scope equipped with a pediatric introducer blade or Intlock. Tsusaki B, et al. [8] reported that a 17-year-old female patient with CdLS undergoing tympanomastoidectomy was intubated with direct laryngoscopy by applying cricoid pressure to reduce the risk of aspiration. We applied the RSII method with cricoid pressure to minimize the possibility of pulmonary aspiration due to the patient's difficult masking ventilation and intubation, irritable airways, short neck, micrognathia, and poor cooperation. She was intubated without complications on the first attempt with a video-laryngoscope. It is critical to develop personalized anesthesia plans that address potential challenges in anesthesia management.

In conclusion, CdLS is a highly complex disease affecting systems, and the anesthetic management of these patients is challenging for the anesthesiologist. The anesthesiologist should know all the features and difficulties of the disease and its relative complications and be knowledgeable and equipped to treat these patients. We emphasize that comprehensive preoperative evaluation, a multidisciplinary approach, individualized patient management, careful postoperative care, and early detection and treatment of possible complications are important in the anesthetic management of patients with CdLS.

Keypoints of this article include comprehensive perioperative management of syndromic pantients. Extensive preoperative physical examination and multidisciplinary approach would be of utmost importance in the management of any complications that can arise during the induction, surgery and the postoperative period.

References

1. Torres MD, Calvo E, Fernandez Espla F, Gilsanz F. Anesthetic management of an adult patient with Cornelia de Lange syndrome. Minerva Anestesiol 2010; 76:229-231.
2. August DA, Sorhabi S. Is a difficult airway predictable in Cornelia de Lange syndrome? Paediatr Anaesth 2009; 19(7):707-709.
3. Kline AD, Moss JF, Selicorni A, Bisgaard AM, Deardorff MA, et al. Diagnosis and management of Cornelia de Lange syndrome: First international consensus statement. Nat Rev Genet 2018; 19:649-666.
4. Moretto A, Scaravilli V, Ciceri V, Bosatra M, Giannatelli F, et al. Sedation and general anesthesia for patients with Cornelia De Lange syndrome: A case series. Am J Med Genet C Semin Med Genet 2016; 172:222-228.
5. Corsini LM, De Stefano G, Porras MC, Galindo S, Palencia J. Anaesthetic implications of Cornelia de Lange syndrome. Paediatr Anaesth 1998; 8:159-161.
6. Vincent MV, Duncan ND. Gastroesophageal reflux and reflux oesophageal strictures in children with Cornelia de Lange syndrome. West Indian Med J 2013; 62:161-164.
7. Sugiyama T, Okutani R. Difficult tracheal intubation in a child with Cornelia de Lange syndrome using a paediatric Intlock installed in a Pentax Airway Scope. Anaestheisa 2012; 67(12):411-412.
8. Tsusaki B, Mayhew JF. Anaesthetic implications of Cornelia de Lange syndrome. Paediatric Anaesthesia 1998; 8:181-184.

Citation: Mahmud A, Uslu A, Çekmen N. Anesthetic Management for Dental Procedures and Oral Surgery in a Patient with Cornelia de Lange Syndrome: A Case Report; Transl Perioper Pain Med 2024; 11(3):678-682

***Corresponding Author:** *Akkan Mahmud, Department of Anesthesiology and Intensive Care, Faculty of Medicine, Baskent University, Fevzi Cakmak Caddesi 10, Sokak No: 45 Bahcelievler, 06490 Ankara, Turkey, Tel: +90-312-203-6868-4867; GSM: +90-555-474-18-22, E-mail: akkan.mahmud@gmail.com*

Editor: *Renyu Liu, MD; PhD; Professor, Department of Anesthesiology and Critical Care, Perelman School of Medicine at the University of Pennsylvania, Center of Penn Global Health Scholar, 336 John Morgan building, 3620 Hamilton Walk, Philadelphia, PA 19104, USA, Fax: 2153495078, E-mail: RenYu.Liu@pennmedicine.upenn.edu*

Additional publication details

Journal short name: Transl Perioper Pain Med

Received Date: December 01, 2024

Accepted Date: December 18, 2024

Published Date: December 19, 2024