Introduction:
Cornelia de Lange Syndrome is a rare congenital disorder, affecting 1 out of 60,000 live births. Dr. Cornelia de Lange reported this syndrome in 1933 as a multisystem syndrome involving congenital malformations, growth retardation, and neurodevelopmental delay. The etiology of this syndrome remains unclear, however it has been suggested that the duplication or partial trisomy of chromosome 3 is a possible cause. Although this syndrome is rare, its multiple clinical features can make the perioperative management of such a patient a challenge for the anesthesiologist. We report the management of an adult patient with chronically decayed and abscessed teeth, cared for by both pediatric anesthesia and dental teams.

Case:
A 32-year-old female 21 kg and 121cm with history of Cornelia de Lange syndrome presented for dental rehabilitation and trans-thoracic echocardiogram (TTE) under general anesthesia. Her past medical history was significant for mild aortic stenosis, gastroesophageal reflux disease (GERD), and unrepaired cleft palate. In addition, the patient had several clinical features typical of Cornelia de Lange syndrome that included microcephaly, macrognathia, micrognathia, short neck, severe mental retardation, and hyperactivity.

Preoperative sedation was avoided in this patient despite her hyperactive behavior, because of the possibility of upper airway obstruction. Anesthesia preparation included having a variety of airway devices and equipment on standby in the operating room, as other alternatives for securing this patients airway.

On arrival to the operating room, standard ASA monitors were placed on the patient. An inhalation induction was performed consisting of nitrous oxide/oxygen (70% : 30%) and sevoflurane. An intravenous (IV) catheter was inserted. Ketamine infusion was started at 75 mcg/kg/min and titrated to effect to maintain the patient spontaneously breathing. A few minutes later, an IV bolus of Propofol 30 mg was administered to blunt the response of laryngoscopy. Video laryngoscopy was performed with the GlideScope ® Cobalt blade # 2 and a grade 1 view was obtained, while at the same time a large cleft palate was observed. An atraumatic intubation was performed using a # 4.5 cuffed endotracheal tube (ETT). Ventilation of the patient proceeded without difficulty. Anesthesia maintenance consisted of isoflurane, fentanyl and vecuronium.

Prior to the start of dental surgery, a pediatric cardiologist performed a TTE as a follow-up examination to one performed several years ago. The patients aortic stenosis remained unchanged. The intra-operative course was uneventful and the patient was extubated at the completion of surgery. Her stay in the post anesthesia care unit (PACU) was also uneventful and she was discharged home with her parents later the same day. Follow-up the next day was also uncomplicated.