



Anesthetic Management of Cornelia de Lange Syndrome: A Case Report

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LETTER TO THE
EDITOR

Dear Editor,

Cornelia de Lange syndrome (CdLS) is described as a congenital disease characterized by delayed growth, mental-motor retardation, typical face appearance, thick eyebrows meeting at the midline, micrognathia, thin lips, micromelia, dental problems, and malformations in the systems such as cardiac, gastrointestinal, genitourinary, and musculoskeletal systems. It is rarely seen, with a reported incidence of 1 in 10,000 to 40,000. It is generally sporadic, but can also be dominant and recessive (1). Problems such as difficult intubation, gastroesophageal reflux, aspiration, and hyperthermia make the airway safety more difficult in general anesthesia practices (2). We aimed to present our anesthesia management experience with a patient having CdLS who was operated because of undescended testis and circumcision after taking the consent of his family.

The patient with a diagnosis of CdLS was 4 years of age, 12 kg in weight, and 85 cm in height. The operation was planned for undescended testis and circumcision. The preoperative examination revealed a history of retarded development, speech impediment, atrial septal defect (ASD), frequent upper respiratory tract infection, conjunctivitis, and food allergy. He had the findings of small face size, restricted mouth opening, short nose, and micrognathia, which could make intubation difficult in general anesthesia. No finding was available in the cardiovascular system, except ASD. The history of infections recurring frequently in the respiratory system constituted a perioperative risk for the patient. The patient who would be operated for circumcision and undescended testis was decided to undergo caudal anesthesia and sedation. However, alternative instruments were kept available in order to maintain airway safety in case of an unsuccessful block. After administering endocarditis prophylaxis, he was taken to the operation room, 1 mg of intravenous midazolam was injected, and he was monitored in the standard manner. Then, sedation was deepened using 10 µg of fentanyl and 20 mg of propofol, and the patient was given lateral decubitus position. After providing aseptic conditions, 12 cc of 2% levobupivacaine was given to the caudal space. The operation was initiated when no pain was observed with the pinprick test after waiting for about 30 min. Anesthetic gases were not used in order to avoid upper airway irritation. The patient was monitored for a while after the 30-min operation and transferred to the clinic because no complication developed.

In CdLS, typical facial appearance is characteristic. The findings, including thick eyebrows that often meet at the midline, microcephalia, micrognathia, long eyelashes, small face, high-arched palate, and short nose, are also indicators of difficult intubation (1).

In patients with CdLS, the risk for gastroesophageal reflux and associated aspiration pneumonia is high (3). In our case, the existence of food allergy and recurring upper respiratory tract infections were signs of an irritable airway. We avoided the application of general anesthesia and volatile anesthetics to prevent respiratory depression and ensure protective reflexes. During the maintenance of anesthesia, cardiovascular findings in the patient can cause hypoxemia and hypercarbia. In our case, the ASD diameter was decreased compared with that in the postpartum period. Endocarditis prophylaxis was administered and no perioperative complication was encountered hemodynamically.

Although CdLS is rarely seen, it is an important syndrome with regard to a difficult airway, organ anomalies, and dysfunctions during preoperative evaluation because of its typical findings during physical examination. Anesthetists should be careful while administering anesthesia and alternatives during preoperative evaluation and intraoperative close follow-up. We believe that regional anesthesia is a good alternative to general anesthesia in surgeries of the lower abdomen and extremities, as in our case.

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Submitted
29.10.2013

Accepted
31.01.2014

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Informed Consent: Written informed consent was obtained from parent who participated in this study.

Peer-review: Externally peer-reviewed.

Authors' contributions: Conceived and designed the experiments or case: AŞ. Performed the experiments or case: ASC. Analyzed the data: HŞ. Wrote the paper: AŞ. All authors have read and approved the final manuscript.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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