

Anesthetic and Airway Management of Patients with Cornelia de Lange Syndrome

By Yvon Bryan, M.D., Anesthesiology-Pediatric Anesthesia at Wake Forest Baptist Medical Center

Considerations for Physicians

Children with CdLS tend to have a variety of procedures performed during their lifetime. Depending on the procedure being performed, different anesthetic techniques may be chosen. These recommendations are based on responses to a survey on the airway and anesthetic management of children with CdLS. The survey primarily dealt with response to medications, intubation, aspiration, ventilation, oxygenation, and emergence.

Preoperative

- It is important to discuss the medications that the child is already taking, if any.
- Certain medications, such as benzodiazepines (midazolam), and/or psychiatric medications, can cause excitation and/or aggression, so beware of paradoxical responses.
- The responses to medications are unpredictable among children with CdLS. Certain patients may be sensitive to narcotics.
- The cognitive function of children with CdLS ranges from age-appropriate to almost complete cognitive dysfunction. Therefore, expect a spectrum of behavioral issues in spite of their age.
- Be careful with the limbs of the child due to deformities of the upper and lower extremities leading to difficult IV access.
- Be careful of aspiration due to the high risk of GERD.
- Due to the high incidence of a high-arched palate, small mouth opening, and micrognathia expect difficult intubation.
- Use a smaller sized endotracheal tube or supraglottic airway device.
- Expect difficulty with the insertion of the airway device.
- There is a high risk of problems with conventional devices; therefore, consider using an alternate device, such as a flexible fiberoptic bronchoscopy (FFB), or supraglottic airway device.

Intraoperative

- A choice of intravenous or inhalational anesthetics is possible.
- Beware of increasing CPAP during bag mask ventilation due to an increase in the incidence of GERD.



- Consider using flexible fiberoptic bronchoscopy (FFB), with or without a supraglottic airway, due to the greater maneuverability leading to an easier insertion of the endotracheal tube.
- Be careful of the risk of aspiration.
- Consider using alternate supraglottic airway devices for a better seal.
- There is a higher risk of desaturation which may lead to bradycardia or cardiac arrest.
- Standard anesthetic agents and medications may be used; however, expect variability in the response to the medication.
- If multiple procedures are being done, it is important to decide whether all procedures should be done under one anesthetic or if different anesthetics should be given for each procedure.
- There is a lack of evidence for there being one safest anesthetic technique.

Postoperative

- There is variability in how children with CdLS emerge from sedation and/or general anesthesia.
- Delayed emergence can occur and may last up to one week.
- There is sometimes a change in the diet patterns of the child.
- Amnesia has been known to occur after sedation and/or anesthesia in some children with CdLS.
- Watch for self-mutilating behavior in response to being sedated and/or put under anesthesia.
- Beware of breathing problems postoperatively as there is a higher incidence of lost airway/reintubation and subsequent cardiac arrest in this population
- Reintubation and/or postoperative ventilation is sometimes required.
- Consider bridging the extubation with an airway exchange catheter.
- Most of the postoperative problems occur shortly after the procedure; therefore, overnight stay is not always necessary.

Date: February 2016

