Anaesthetic and Airway Management of Patients with Cornelia de Lange Syndrome (CdLS)

By Yvon Bryan, M.D., Anaesthesiology-Paediatric 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 anaesthetic 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 behavioural 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 Gastro-esophageal Reflux Disease (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 supra-glottic 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 supra-glottic 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 supra-glottic 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 supra-glottic 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 behaviour in response to being sedated and/or put under anesthesia.
- Beware of breathing problems postoperatively as there is a higher incidence of lost airway/re-intubation and subsequent cardiac arrest in this population
- Re-intubation 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.

CdLS and Anesthesia

By Aaron Zuckerberg, M.D., Paediatric Anesthesiologist and Critical Care Medicine, Sinai Hospital and member of CdLS Foundation Clinical Advisory Board and Antonie Kline, M.D., Medical Director, CdLS Foundation

One of the guiding principles of paediatric anesthesia is to utilize an individualized approach for each patient. As has been emphasized previously, the anesthetic approach to children and young adults with CdLS is and must be very individualized.

The most important elements for a positive experience are meaningful communication with the anesthesiologist before the procedure and parental presence during induction and emergence from anesthesia. Parents should be clear as to their child's previous experience with anesthetic drugs and actively participate in choosing what approach would be best for their child. Parents should find out from their anaesthesia providers what the options are available for their child.

Parents know their children and will often guide the anesthesiologist into choosing the best option. As an example, some children who have asthma have experience with breathing treatments and are usually more comfortable than others are with a mask induction of anesthesia.

Some may not be fearful of needles, and others will only do well with a rapid shot of sedating medicine. The challenge is to successfully obtain IV access in the least traumatic fashion possible. Parental presence is often much more effective at relieving a child's anxiety during these difficult periods with medications. Depending on the institution, parents can bring their child's comfort items with them for both induction and the recovery period.

As children awaken in an unfamiliar environment, they may become agitated and upset. The remaining effects of the anesthetic may worsen the child's discomfort by clouding their perceptions and sensations.The presence of a parent and the child's favourite blanket, music, or video can be very effective at easing children through these periods. Parents should find out ahead of time what to bring on the day of the procedure. Unfortunately, much of what we know about the interaction of anesthesia with individuals who have CdLS is based on anecdotal experience. In paediatric anesthesia textbooks, the anesthetic consideration for those with CdLS is limited to concerns about their airways and difficulty with IV access. Children with CdLS often have short jaws and necks, which increase the difficulty of maintaining an open airway and placement of a breathing tube. Not infrequently, a bronchoscope (an airway telescope) is required to safely place the breathing tube.Many patients with CdLS have shortened upper extremities and limitation of movement at the elbows, which limit the areas available for IV placement when they are awake. Often, following sedation, these contractures may be modestly relaxed and IV placement is easier. One textbook has stated that "patients with Cornelia de Lange Syndrome may have decreased anesthetic requirements." This certainly has not been our experience. Although individualized, children with CdLS seem to require more anesthetic per body weight than average in order to maintain an adequate plane of anesthesia.

Several medications often used in paediatric anesthesia are the Benzodiazepines: Midazolam (Versed) and Diazepam (Valium). Benzodiazepines are commonly used as a preoperative sedation in an effort to minimize anxiety and ease IV insertion. Depending on a child's age, these drugs can be given orally, nasally, as a suppository, or as an injection. These drugs work by binding to certain specific areas, called GABA receptors, which decrease the general activity of the brain, usually producing a state of calmness, sedation and amnesia.

Unfortunately, some people respond to Benzodiazepines in a paradoxical manner, becoming disinhibited (free of inhibitions), agitated, emotional, excited, or violent. It is estimated that one percent of healthy adults and up to five percent of healthy children will develop these paradoxical reactions to Midazolam. The exact cause of this reaction is unknown. There may be subgroups of patients who have abnormal GABA receptors that may predispose them to this abnormal response. To date, there is no data to determine whether the GABA receptor in CdLS is similarly affected.

Some recent information has been obtained, however, from a Foundation- supported study carried out by two dentists,DouglasClemens,D.M.D,a CdLS Foundation Clinical Advisory Board member in private practice, and Ellen Alpano,D.D.S., from the University of Maryland Dental School.

Dr. Alpano collected records and hospital charts on patients with CdLS who had undergone anesthesia for dental procedures. Based on hospital records, of those individuals with CdLS who received a pre-medication for anesthesia, 80 percent (four of five patients) who received Midazolam had an adverse post-operative event, compared to 28 percent (two of seven patients) who received a different medication as a pre-medication.

Thus, an alternative medication to Midazolam for anesthesia should be considered in patients with CdLS, but further studies need to be carried out since these numbers were very small [Kline AD, et al. Cornelia de Lange syndrome 4th biennial scientific and educational symposia abstracts. Am J Med Genet Part A 152A:2683-94, 2010].
The common risks of an anesthetic can involve complications of breathing, heart function, recall, and allergic reactions to the medications. These risks are usually far less than the risks of the car ride to the hospital. The major areas of increased risk for the patient with CdLS are airway and injury when emerging from anesthesia. As mentioned, some children with CdLS have an abnormal structure of their airway which increases their chance of developing some form of airway obstruction either during or after the procedure, as well producing an increased difficulty when placing the breathing tube.

Some children with CdLS awaken from their anesthetic in an aggressive state, which puts them at risk for injury from contact with bed rails and other medical equipment.

Since history is usually very predictive, alerting the care team if this has happened previously will allow them to minimize the chances of injury by the use of pads on the hard surfaces of the bed.

All of this information should be reviewed with the anesthesiologist prior to any procedure performed under anesthesia. We are always happy to speak directly with the professionals, if indicated, as well.

Contact the Canadian CdLS Foundation at support@canadiancdlsfoundation.com with any questions or concerns.