

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

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Considerations for Families

Most children with Cornelia de Lange Syndrome will have to undergo sedation and/or anesthesia at some point in their lifetime. Whether it's for a dental cleaning, imaging study or surgical procedure, this page should help give you an overview of what to expect for your child's anesthetic/airway management.

My child's doctors tell me that he/she will be undergoing anaesthesia for a surgery/procedure. What is anaesthesia and what exactly will happen?

We will explain by dividing the process into preoperative, intraoperative, and postoperative.

Preoperative

Sedation medicine will be given to prepare/calm child before the procedure. These may be given orally or intravenously.

- Let the anesthesiologist know who your geneticist is, or any other doctor that knows the medical needs of your child.
- Let the doctor know if your child has cognitive and/or behavioural issues that may lead to a lack of cooperation
- Some hospitals may allow you to accompany your child into the operating room to calm him/her down until he/she is put to sleep if you think that would be beneficial.
- Notify the anesthesiologist if your child has had previous complications when sedated and/or put under anesthesia, and what those complications were.
- If you are aware of any specific medications that your child is sensitive to, let the anesthesiologist know.
- If you are aware of any specific medications that you know your child benefits from, such as something to reduce postoperative nausea and vomiting, let the anesthesiologist know.
- Notify the anesthesiologist if your child has GERD (gastroesophageal reflux disease), the movement of contents from the stomach back to the esophagus.
- Notify the anesthesiologist if your child has a history of pneumonia.
- Notify the anesthesiologist if your child has very small facial features and/or cannot open his/her mouth easily
- Let the anesthesiologist know if your child has a challenging airway and/or a history of difficult intubation (the placement of a breathing tube).

Intraoperative

What happens during your child's procedure under anesthesia.

- Some children are sensitive to certain medications. Ask the anesthesiologist what medications he/she plans on using.
- If your child has challenging airway anatomy, it may be difficult to visualize the voice box. Ask the anesthesiologist which instruments he/she plans on using.
- Ask the anesthesiologist if he/she thinks it will be difficult to place a breathing tube and the reasons why.
- Often times, a smaller breathing tube is needed to intubate children with CdLS
- Ask the anesthesiologist if he/she plans on using a smaller than expected tube.
- Ask the anesthesiologist if he/she plans on using any specialized devices, such as a laryngeal mask airway, or a flexible fiberoptic bronchoscopy.
- Ask the anesthesiologist if he/she expects any complications while intubating, such as a decrease in oxygen levels or an increase in carbon dioxide levels.
- Ask if he/she expects any major complications during the procedure, such as a respiratory arrest and/or a cardiac arrest.
- Aspiration is a high risk for children with CdLS, especially those with GERD, so ask your anesthesiologist what precautions he/she plans on taking to prevent it.
- If the anesthetic technique your anesthesiologist uses seems to work for your child, take note of it so that you can inform anesthesiologists in the future.
- Likewise, if anything did not go well with your child's airway and anesthetic management, take note of it as well so that you can ensure it does not happen again.

Postoperative

The emergence (waking up) from anesthesia and what happens after the procedure is over.

- Compare your child's general temperament (how they generally act everyday) to their temperament upon waking up.
- Children with CdLS seem to be unpredictable in how they wake up from sedation or general anesthesia. Note what medications were used and how your child woke up from them.
- In addition, some children wake up slower than expected or some wake up quicker than expected when coming out of sedation or general anesthesia.
- Note if your child woke up calm and aware, or if your child woke up disoriented, agitated, crying, etc.
- Your child may be treated with medication if the wake up did not go well.
- Look for breathing problems after the procedure, which would include problems with oxygen levels, problems with carbon dioxide levels, respiratory depression, or delayed extubation (the removal of the breathing tube).
- Most of these problems happen soon after surgery if they are going to occur; therefore, overnight stay is not always needed.

- Make sure the nurses in the room that your child is taken to after the surgery know that your child has CdLS and that there is an increased risk of there being a breathing problem postoperatively.
- Keep track of your child's airway and anesthetic management for every procedure. This will give you an idea of what works and what doesn't work for your child.

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