

Anesthesia for People with CdLS

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One of the guiding principles of pediatric anesthesia is to utilize an individualized approach for each patient. As emphasized in the accompanying family stories, 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 anesthesia providers what the anesthetic 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 with a mask induction of anesthesia. Others 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 than medications like Benzodiazepines Midazolam (Versed) and Diazepam (Valium). Depending on the institution, parents can bring their child's comfort items with them for both induction and the recovery period. As a child wakes up in an unfamiliar environment they become agitated and upset. The remaining effects of the anesthetic confound the child's discomfort by clouding their perceptions and sensations. The presence of a parent and the child's favorite blanket, music tape, or video can be very effective at easing children through these periods, by making things more familiar. Find out ahead what you can bring with you on the day of the procedure.

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), even agitated, emotional, excited, or demonstrate violent behaviors. It is estimated that 1% of healthy adults and up to 5% of healthy children will develop these paradoxical reactions to Midazolam. Interestingly, Ketamine has been shown to be an effective rescue medication for those who develop a paradoxical reaction to Midazolam. The exact cause of this paradoxical 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.

Unfortunately, much of what we know about the interaction of anesthesia with those who have CdLS is based on anecdotal experience. In pediatric 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 contractures at the elbows, which limit the areas available

for IV placement when they are awake. Often, following sedation, these contractures are modestly relaxed and IV placement is facilitated. One text states that “Patients with Cornelia de Lange Syndrome may have decreased anesthetic requirements.” This certainly is not my 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.

The common risks of an anesthetic 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 only areas of increased risk for the patient with CdLS are airway and emergence injury. As described, *some* children with CdLS have an abnormal structure to their airway which increases the chances of developing some form of airway obstruction either during or after the procedure, as well the increased difficulty with breathing tube placement. This should be reviewed with your anesthesiologist prior to the procedure. 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 past history is usually very predictive, alerting the care team of this will allow them to minimize the chances of injury by the use of pads on the hard surfaces of the bed.

One of my goals in working with those who have CdLS is to collect data on their experiences with anesthesia, anesthetic drugs, and airway difficulties. Only by collecting a large database of information from patients with CdLS who have experience with anesthesia will we be able to clearly define those issues which are specific to CdLS. Through education, we will be able to improve the anesthetic management of these patients. We are presently evaluating the best way to organize this effort. We will notify families and professionals once the anesthesia registry is up and going. Until that time, please feel free to document your child’s experiences (good or bad) with anesthesia at sleepingcdls@netscape.net.