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Reaching Out, Providing Help, Giving Hope

Encounters with Anesthesia

By Bruce Cleinman, M.A., former Family Service Coordinator at the CdLS Foundation

The decision to have a child undergo surgery or medical procedures can provoke anxiety for both the parent and the child. The consideration of administering anesthesia to a child is often a contributing factor to the fear that can surround such events, especially given the contrary reaction individuals with CdLS can have to medication. From experience, parents learn what anesthetic agents are most effective for their child and which ones they should avoid. In an effort to broaden the understanding of how individuals with CdLS respond to anesthesia, the following families shared the lessons they have learned.

Will

Thirteen-year-old Will most recently had anesthesia to undergo an endoscopy and ear tube placement. His mother Beth recounted that, "Will has great difficulty with the pre-anesthesia medication called Versed used to relieve anxiety and impair memory. When Will is given Versed he gets loopy and very agitated prior to the anesthesia. After the operation he wakes extremely agitated and is very hard to control. Only allowing time for the effects of the medications to wear off has helped. In the interim, we made attempts to soothe him and keep him safe as he thrashed, yelled, and displayed out-of-control behavior. His doctors recently tried anesthesia without first using pre-anesthesia medication and that worked much better for Will. He came out of anesthesia with significantly less agitation, making the whole experience much easier on him and on us. Other than this issue we haven't had any difficulties with anesthesia beyond the typical issues for our kids, such as the small jaw/mouth making intubations a little more difficult or the difficulties with putting in an IV [due to their small veins]. Usually they put Will under with gas before trying the IV."

When asked what she would like other parents to learn from her experience Beth replied, "Each child is an individual and will react in their own way to any given medication. Let doctors know that our children can be sensitive to certain medications. Know which medications are absolutely necessary and which are not. Ask questions; take an active role in the process. We've learned to ask what drugs are given, know what the effects are, and how Will might or might not tolerate them. Note which medications to avoid in the future."

Brianne

Like Will, Brianne is 13 years old and has also had endoscopies and ear tube placement requiring the use of anesthesia. According to her mother Karen, Brianne has been given anesthesia 10-12 times and has never been administered any pre-anesthesia medication. Karen shared that "Brianne was given Versed by a nasal spray when she was very young for an endoscopy. Although it did not put her completely out, she was out of it enough for them to perform the procedure with no problems. Two or three years after that, when it was time for another endoscopy, they tried Versed nasal spray again, and it did not even phase her, so



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the appointment had to be rescheduled. That time, and with future procedures that required anesthesia, I was told that they just used straight gas (a mixture of Sevoflurane, Nitrous Oxide, and Oxygen) with a gas mask.”

Karen learned “that the anesthesiologist can make or break the whole procedure, particularly when you are dealing with fairly uncomplicated procedures like Brienne has had. The most important recommendation I would give to parents is to try to find out from other people, or even your child’s doctor, the name of some good anesthesiologists. Get at least two or three names and request them every time you have to have a procedure done. I don’t think all parents realize that you can request a particular anesthesiologist. Sometimes they are already scheduled for another surgery, that’s why I suggest getting at least two or three names. Not only will the parents feel more comfortable with having someone they trust, but by using the same anesthesiologist you are assured that they know your child’s particular case; they can look back on their records and see what worked best.”

Caitlin

Ten-month-old Caitlin was given general anesthesia (a mixture of Sevoflurane, Nitrous Oxide, and Oxygen) through an inhalation mask in order to put in an IV infusion of Propofol. Once on the Propofol, she was taken off the inhalation mask. Sara, Caitlin’s mother, noted that Caitlin responded favorably to both anesthetic agents and was able to have a Brainstem Auditory Evoked Response (BAER) hearing test and be fitted for hearing aide ear molds. Sara noted that, “Upon waking, Caitlin was initially a little fussy but was comforted by being held. She was then in a mellow mood.” Caitlin’s experience was positive overall. “We’ve been lucky, she’s quite a fighter,” said Sara.

Claudia

Claudia, age three, has had five procedures requiring anesthesia, including exploratory surgery of her intestinal tract, spinal cord surgery, cleft lip repairs, and eye surgery. Pre-anesthesia was unnecessary according to her mother Nicole as “Claudia was so young and unaware of the impending procedures. She was actually smiling as one surgeon carried her to the operating room!”

Sevoflurane and Fentanyl are two types of anesthesia that have been used for Claudia. According to Nicole, she had positive experiences with both. “There were no complications,” stated Nicole, “She came out of the anesthesia quickly, although slightly cranky, and recovered extremely well.”

“Her only difficult post-surgery experience came after her first cleft lip repair surgery. Claudia cried, arched her back, kicked her legs, and fussed for nearly six hours after the surgery. We eventually realized that it wasn’t the anesthesia that was bothering her, but the Morphine they had given her post-surgery to control the pain. Now we make sure she only gets Tylenol with Codeine after surgeries.”



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Through Claudia's experiences with anesthesia, Nicole has learned that "Claudia is an extremely strong and resilient little girl. I've also learned to trust my own instincts about my child. When Claudia was acting strangely and the hospital staff kept saying it was just the normal effects of anesthesia, I refused to believe them. Luckily for Claudia, we figured out that the culprit was Morphine."

The family was able to schedule multiple procedures for Claudia's surgery dates requiring her to receive anesthesia only once for many procedures. Her doctors were able to successfully repair her cleft lip, lift her nose, correct her strabismus, and unclog her tear ducts at the same time.

David

David, age 22, has undergone general anesthesia for endoscopies, major dental work, and to release the muscles in his feet. His mother Cindy shared that "Propofol worked wonderfully for David. His anxiety level is so high that he needs to be put out immediately once they insert the IV. Propofol does just that. Then he wakes up and is ready to go home very quickly. He had his dental work and endoscopies done using this. I have been very fortunate that all of his medical providers allowed me to stay with David until he was out and again right there with him as soon as they stopped the Propofol, so I was there when he woke up.

David seems to get manic with moderate to high doses of Benzodiazepines (Valium, Ativan, Versed). I will only allow Benzodiazepines to be used in conjunction with other medications and only at very minimal doses. We now use Zyprexa Zydis disc [which dissolves rapidly on the tongue] to relax David quickly in order to transition him from his group home to any doctor appointments and as a pre-surgery medication. It takes about two hours for the medication to render him completely unconscious. The only problem with it is that once the Zypreza takes effect he sleeps for 12 hours.

I think it's important to remember each child is different. It is important for each parent to become knowledgeable about medical issues and how anesthesia affects their child. I would suggest that parents try Propofol knowing that it has worked well for other children with CdLS. It's trial and error with medications; what works for one doesn't necessarily work for all."

Nicole

Karen noted that her daughter Nicole, age 11, has had annual endoscopies for some time now. Karen observed that "Nicole seems fine with the anesthesia. She is initially given a relaxing gas agent and then put to sleep with Propofol through an IV. This has worked well for a number of years. She comes out of it and just wants to be held. Once we get her home and I put her in a bath she plays as if nothing happened. I think it helps that we have used the same doctor for all the endoscopy procedures. He probably has kept notes and feels that he knows her."



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This year, Karen had Nicole's ptosis surgery and dental work done at the same time. Karen gave the surgeons several months to schedule it since there was no urgency for the procedures. The medical team understood her request to combine procedures and accomplished it willingly. Karen stated, "I always try to stack my procedures provided that the doctors also agree that Nicole would benefit. Sometimes the hospital has the last say."

Karen has learned that "Nicole's veins are difficult to find so I have always asked that she be put under [with a relaxing agent] while they search for the proper veins. Otherwise, she is too strong and will keep up the fight. I'd rather her doctors concentrated on the procedure than be stressed out from the IV."

Karen urges parents to "Always ask questions such as how long it will take to come out of anesthesia, and what side effects they can expect. It gives you an idea of what to expect and what to look for afterwards. A good anesthesiologist will sit down with you and give you an overview and time to ask questions." Nicole has not had any adverse reactions to anesthesia.

Stephanie

Stephanie, now age 13, was given Versed with her first endoscope at 2 1/2 years old and it was "absolutely awful" according to her mother Shirley. "It did not relax her at all and she had a terrible side effect of hallucinations that lasted for about 45 minutes. She came out of the procedure room trying to get off of the gurney and kicking, crawling, and standing – just delirious – which was not her normal demeanor. She is a sweet, quiet child. So we refuse to have any doctor or dentist give her Versed. She was given Demerol once for pre-anesthesia but it caused her to vomit, so we also avoid that drug."

Fortunately, her family also found Propofol to be an effective and safe alternative for her future endoscopies and surgeries. Shirley explained, "It knocked her out but there were no side effects, no hallucinations, nausea, or vomiting. She came out of the surgeries drowsy, but very calm and knew who we were right away."

Through the family's experience with anesthesia they have learned that "There are some medications that Stephanie is extremely sensitive to and may react with adverse behaviors. We have learned that no matter what pre-anesthesia or anesthesia is used, she does get nervous and anxious about the procedure and can't verbalize that to us. Because she does get nervous (like anyone without a disability) she needs to have her IV put in right away. We have found that her nervousness and the coolness of the rooms make it very difficult to find a vein in which to put the IV. This in turn added to her nervousness and her pain."

Shirley advises parents to "Stand firm on what medication you will and will not let your child have once there is a known adverse reaction. Insist on having the IV done as soon as possible. Children with CdLS tend to have small veins and when they are nervous and



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dehydrated from not having fluids after midnight it can upset them further to try an IV after waiting at the hospital.”

If you have questions about anesthesia you may contact the Foundation or use the on-line Ask the Doctor feature of our website. Concerns will be forwarded to Dr. Aaron Zuckerberg, our Clinical Advisory Board anesthesiologist. We also welcome feedback on what anesthetics worked and did not work well for your child. A special thank-you is extended to each of the families who contributed to this article.