

Surgery Considerations for Adults and Children

Introduction

People who have osteogenesis imperfecta (OI) and their families need to plan ahead for hospitalizations. This planning process can include becoming familiar with services provided by the hospital, such as the availability of social workers and child life specialists, and sharing information about OI with the doctors and nurses. Hospitals usually have a Patient Advocate or Patient Representative on staff who can facilitate getting information to all staff members about OI and the special needs of the OI patient. A notice can be affixed above the hospital bed, at the nursing station, and on the medical chart identifying the patient as a person with OI and describing cautionary measures.

Preparing Children for Surgery

Carefully preparing a child prior to a hospitalization is important, especially for children with OI who may face multiple hospitalizations. Discussions should be age appropriate, use child-friendly language and attempt to reduce the stresses that lead to fear and anxiety. Parents are encouraged to answer the child's questions truthfully but without getting too complicated or confusing. Child life specialists recommend being honest with the child, even about the unpleasant parts. No one can promise there won't be pain or discomfort. Responding to questions in a straightforward manner will help parents keep their child's trust, which is also important. It is natural for children to fear the unknown, and hospitals look and smell very different from any other place in the child's experience. Children feel less anxiety if they have a chance to tour the hospital prior to admission. A child life specialist can help explain what will happen during the surgical procedure and any tests that the child will experience. Bringing a toy, stuffed animal, or other familiar object to the hospital can also help make a child feel more secure.

Child life specialists encourage parents to stay near their child as much as possible during a hospitalization. Parents may want to make arrangements to remain with their child as long as possible prior to surgery and to be present in the recovery room as the child wakes up. This step may help a child be calmer and more compliant with necessary medical procedures. Parents may prefer to move their child themselves to reduce the chance of fracture and to personally train members of the hospital staff who will be helping to care for their child. Parents can make arrangements to ensure that a parent or other familiar adult can stay overnight with the child if necessary.

Adult Concerns

Adults facing surgery should ensure that all relevant tests, including cardiac and pulmonary function tests, are performed ahead of time. Any adult with OI, but particularly those with OI Type III or those with short stature, may have compromised cardio-respiratory functions. This can cause serious complications during surgery and in some cases may prevent the individual from being a candidate for elective surgical procedures. Whenever possible, it is a good idea to make various legal arrangements ahead of time. These can include writing a will and having a trusted family member or friend hold durable power of attorney for medical issues. It can be very helpful to have someone who can explain OI accompany the adult patient to the hospital. Besides being knowledgeable about OI, this person should be fully briefed about the expected procedure and hospital policies so that he or she can serve as an advocate for the hospitalized adult if needed. Adults also benefit from a tour of the hospital facility prior to admission. Becoming fully informed about exactly what will occur during admission, getting to the room, pre-op and post-op routines, etc., will ensure that things go as smoothly as possible. Working with family members, friends, the primary care physician and hospital social services, a plan should be developed prior to surgery for any needed assistance during recovery and rehabilitation.

Coping with Hearing Loss

People who have hearing loss are advised to post this information on a sign above their hospital bed, at the nursing station, and to make sure it is included in their chart. All relevant medical personnel should be informed about the hearing loss and the fact that the patient might not hear over the intercom. Staff should know that they might need to communicate in person, up close, or in writing. The surgeon and anesthesiologist should be informed that the patient might not be able to understand surgical staff members who are wearing masks. Therefore any necessary information or instructions should be provided before the masks are in place. Recovery room staff should be alerted to have paper and pencil available and to return the hearing aid or other device as soon as possible. A small bag labeled with the owner's name in which to keep the hearing aid or assistive listening device will facilitate this process.

Hospitals often have brochures describing services and equipment that are available for people who are hearing impaired. Equipment could include captioned TV, amplified hearing-aid compatible phones, flashing phone alerts, TTY's and one-on-one communication devices, or a personal FM system. Notify the Patient Representative in advance so that the necessary equipment will be available on the date needed.

Meet with the Surgical Team

When surgery is required, it may be necessary to provide the surgeon and other members of the surgical team with information about the special needs of a person who has OI. Fragile bones, delicate tissues, anesthesia, and in some cases, short stature, are important topics for discussion. Surgical staff needs to be prepared to take extra precautions when moving or positioning the OI patient. Special padding may need to be added to stretchers and restraints to help minimize stress on bones. Automatic blood pressure cuffs must be carefully monitored so that the arm is not fractured. Adult patients with OI may be small in stature, and the use of some materials sized for children may be appropriate. On the other hand, medical staff should be aware that stature is not a useful indicator of cognitive development. The patient's age is the best guide for medical staff to use in determining how to interact appropriately with a person with OI.

Anesthesia and Hyperthermia

People with OI have some special risks that distinguish them from the general population regarding the use of anesthesia. Because of physical deformity, the mobility of the neck and the jaw may be reduced. Chest and rib deformities and scoliosis may affect breathing. Other hazards may be posed by complicating factors such as dental problems (skeletal fragility makes dentition prone to fracture), cleft palate, joint stiffness or heart valve disease. Anesthesia personnel also need to be aware of the susceptibility of fragile bones to fracture from movement, impact, or stress, and the possibility that alternative procedures for intubation, such as fiber optics through the nose, may be necessary when treating some individuals.

For those with moderate and severe OI, the small stature of the patient can also be confusing to anesthesia personnel. Tube size should be determined by the size of the head instead of the size of the body.

Thermal instability is also a problem for patients with OI, and a pseudo-malignant hyperthermia with elevated temperature, but without the enzymatic or system changes, has been described.

Patients should also tell their doctor if they are taking any nutritional supplements, or herbal medications. It may be necessary to stop these supplements two to three weeks prior to surgery. A number of anesthesiologists have reported serious problems with heart rate or blood pressure when patients did not reveal that they were taking supplements before their surgery. Included in the list of supplements to mention are St. John's Wort, ginkgo biloba, feverfew and ginseng. Any history of postanesthesia vomiting should also be brought to the attention of the anesthesiologist.

After general anesthesia, it is very important to optimize respiratory function. Simple steps like taking deep breaths or more complicated respiratory therapy treatments prevent fevers and breathing problems after surgery. Early mobilization out of bed also decreases the risk of pulmonary complications. It helps to discuss ahead of time with the doctors and nurses what accommodations or assistance will be needed to meet these goals.

Latex Allergy

Latex allergy affects some people with OI because of their frequent exposure to latex. Latex allergy is a reaction to products made from natural rubber latex, the cloudy white sap of the rubber tree. These products contain a variety of proteins that may cause an allergic reaction when either absorbed through the skin or inhaled. In most cases, the allergy develops after repeated exposures.

Blood Donation

If possible, surgery should be accomplished using tourniquet hemostasis; meticulous hemostasis should be maintained. Blood volume in patients with OI is body size and weight dependent, not age dependent. Blood loss should be carefully monitored, and necessary blood should be available. During a normal femoral IM rodding, loss of 120 cubic centimeters could occur, and in the case of a patient with an intravascular volume of 1200 to 1600 cubic centimeters, this constitutes a 10% loss of blood volume. If a transfusion is likely during surgery, patients may have available the following three options:

1. Autologous pre-deposit: storing the patient's own blood for use during the surgery. This is the safest form of transfusion, but the patient's health needs to be such that this form of blood donation is allowed.
2. Blood from the community supply. This blood has been donated by healthy donors, with a known medical history. Each pint of blood is thoroughly tested.
3. Directed donations: blood donors are selected from among the patient's family and friends. This option is not deemed to be safer than blood transfusion from the community supply, but some patients are more comfortable with this option. Blood can be donated up to 42 days prior to the surgery, but must be donated at least 72 hours prior to the surgery for testing and processing, so this option is not available in emergencies.

For any form of blood donation, donors must generally be in good health and weigh at least 100 pounds. Certain medical conditions or medications could disqualify a particular potential donor.

Those who require frequent surgery may want to make a list of friends and relatives who are consenting donors with matching blood types. The patient's doctor can place an order for directed blood donations at the relevant blood center, including the amount that needs to be available at the time of surgery. Donors can give blood every eight weeks, provided their red blood cells remain within the normal range and they meet all the other donor requirements.