



KEYS TO HELP RARE DISEASE PATIENTS ENSURE A SUCCESSFUL SURGERY

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Introduction

Numerous patients in the rare disease community are among the more than 50 million people who undergo surgical procedures in U.S. hospitals each year. If you or your loved one is contemplating a surgical procedure, this toolkit is designed to provide information and resources that will help ensure a safe and successful experience before, during and after an operation. It also will share steps to help individuals decide if surgery is the best option, the questions to ask the surgeon and care team before and after the surgery, a simple surgical checklist, and direct advice from other patient advocates.

Outpatient surgery has grown in the past decade, and now there are more surgeries performed in outpatient locations, such as in a surgical center, compared to in hospitals. While most patients and families prefer to return home the same day, this means families are often managing the recovery process without the help and support of hospital staff.

Modern day surgery is a group effort and patients are vital members of the surgical team, especially when they have the information and basic skills to ask the right questions and work in partnership with their health care providers.

Introduction

What Does it Mean to be an Active Participant in the Surgical Process?

“I am definitely an active participant in my son’s surgeries,” says Lori Davis, mother of a child with Osteopetrosis. “After his first surgery, it became apparent that our surgeries were not going to follow the normal surgical protocol and that our involvement as parents would be necessary. Sometimes, the medical professionals forget that the disease you are dealing with is ‘rare’ therefore the complications, side effects, and recovery may be quite different from anything they have seen in previous patients. And we, as parents, need to be the reminder for them to slow down and look at the situation from a rare disease perspective.”

Often, the first step in being an active participant is deciding if surgery is in fact the best option for you. The American College of Surgeons recommends asking the following questions first:

- Why do I need this operation?
- Are there other treatment options, and is this the best option for me?
- What are the risks, benefits, and possible complications for this operation?
- Will my health history and the medications I currently am taking mean that the possible risks, complications, and benefits will be different for me?

Access the **American College of Surgeons** for the complete list of questions to consider when contemplating surgery. <https://www.facs.org/education/patient-education/patient-resources/prepare/10-questions>.



SECTION 1: PREPARING FOR SURGERY

Lenore Alexander, the parent of a child with a rare condition, believes that being an active participant in the surgical process, while it may seem simple, can make a significant difference.

“I believe it is our responsibility to be informed and proactive when facing a surgical procedure.” “I lost my 11 year-old daughter, Leah, to an adverse event after surgery for a rare condition. If I had known my daughter needed to be on a continuous electronic monitor, because of the strength and the amount of narcotics she was receiving, I would have made sure it happened. In doing that, I may have been able to save her life. There are so many ways to be prepared for either an unexpected or planned visit to a hospital.”

- Lenore Alexander, Executive Director of Leah's Legacy (www.leahslegacy.org).

The decision to undergo a surgical procedure is a process that requires understanding basic information about you or your child's condition and knowing the important questions to ask.

All surgical procedures, even those that are brief and considered routine, carry some degree of risk to a patient. But the risk can be reduced if you know what questions to ask and how to prepare in advance to participate in care and recovery.

Deciding to Have Surgery

You, as a patient or caregiver, need to be very clear about why surgery is needed, what the procedure is expected to correct or accomplish, and how your surgical team can help ensure a safe and successful outcome. You must also be sure to completely walk through the rare condition as it is never safe to assume the surgeon is intimately familiar with the disease. Discussing the special considerations of a rare condition with the surgeon is an important safety strategy.

“I can think of numerous times I've mentioned a symptom I had been experiencing and hearing the doctor thank me and explain how important it is that he knows about it,” says Tom Christie, who has Relapsing Polychondritis. “So my best advice is to leave nothing out when talking to your doctors, and letting them decide if it's important or not. When you are suffering from a rare disease, it's your responsibility to make sure that every healthcare professional you deal with knows about your history.”

Questions to ask include:

- Can you write down the medical name of my procedure so I can research it online?
- Are there any alternatives to having surgery? Is “waiting and watching” an option?
- Is there medical evidence that supports having surgery over other options?
- How often is this surgery successful in correcting the problem?
- Could you explain the risks and benefits of this surgery?
- Is this a new or experimental procedure?
- If there is an increased risk of complications, what can be done to minimize the risk?
- Do you recommend obtaining a second opinion? Why or why not?

SECTION 1: PREPARING FOR SURGERY

After consulting with the health care team and agreeing that proceeding with surgery is the best option, there are several key decisions that require attention. Choosing the best surgeon and deciding where to have surgery are important steps to ensuring a successful recovery. Patients assume that their primary care provider will recommend a surgeon, but feel free to get involved in the process as well. Select a surgeon who makes you feel comfortable, inspires confidence, and meets your needs. This may be accomplished by seeking a second opinion.

“I would definitely recommend a second opinion especially if you aren’t seeing an ‘expert.’ There are often multiple ways to treat something and with a procedure as serious and invasive as surgery, it doesn’t hurt to make sure it is 100 percent necessary,” says Maija Rannikko, a young adult with Chiari Malformation.

“In my case, the very first surgeon I spoke with was going to do a procedure that wasn’t recommended for pediatrics. With that procedure, I would have most likely had a failed surgery that would cause more symptoms and likely require more surgeries later on in life. A second opinion probably saved my life. By the time I did have surgery, I had seen multiple surgeons, neurologists, and done my own research on Chiari Malformation and felt that the procedure I had would be right for me.”

Choosing a Surgeon

Choosing a specialist to perform the surgery is a vital part of the decision-making process. The primary care physician can make a recommendation as a starting point, but it is important to meet with the surgeon to assess whether he or she meets your needs by asking the following questions:

- Are you board-certified in the specific specialty I need?
- Are you a member of the **American College of Surgeons** <https://www.facs.org/search/find-a-surgeon> and the **American Board of Medical Specialties** <http://www.certificationmatters.org/>?
- How many times have you performed this exact surgical procedure?
- Will you be performing my entire surgery? Will you be assisted by other surgeons, residents, or students? If so, will you be present in the operating room and provide direct supervision?
- What is your complication rate? Infection rate?
- What is the recovery time?
- Do you accept my insurance plan? Are the other specialists you intend to consult or work with also part of my plan?

Jamie Holmes, a young adult with V.A.T.E.R. Syndrome and Founder of Jamie’s Dream Team, has undergone many hospitalizations and surgeries. She also stresses the importance of asking questions before a surgery. “It is very important to ask all of the right questions. Like, what is the success rate with this surgery? Have you ever performed it? How many times? What are the pros and cons? But also make sure you look at all of your options. Don’t be scared to look for an alternative.”

SECTION 1: PREPARING FOR SURGERY

Deciding Where to Have Surgery

During the consultation appointment with the surgeon, discuss location options for the procedure. Surgery may be scheduled in a hospital, a surgery center, or a doctor's office, and it is important to understand why the surgeon recommends a certain setting.

Once you and your surgeon decide where to have surgery, research the hospital online or review the Empowered Patient® Safety in Outpatient (Ambulatory) Surgery Centers fact sheet for same day surgeries in the Supplemental Materials section. Other steps include:

1. Check if the hospital is accredited by the **Joint Commission** <http://www.qualitycheck.org/consumer/searchQCR.aspx>;
2. Access hospital quality ratings and results of patient surveys on the **Medicare website** <http://www.medicare.gov/hospitalcompare/search.html>; and
3. Most importantly, be sure to confirm that the hospital accepts your insurance plan and that any pre-authorizations are obtained in advance.

If the hospital is a teaching institution you should expect to receive care from residents and students. Ask if the hospital has an intensive care unit (ICU) staffed with specially trained physicians called intensivists in case you need more advanced care. Also, note if the hospital has a rapid response team (RRT) to assess patients in distress, and if patients and family members can call the rapid response team directly.

Access the Empowered Patient® one page fact sheet Be Prepared For Your Surgery and Surgical Patient Checklist in the **Resource Guide** section to help you stay on track before, during and after surgery. If you are scheduled for same-day surgery, please access the Empowered Patient® one page fact sheet Safety in Outpatient (Ambulatory) Surgery Centers in the Supplemental Materials section.

Anesthesia and Other Considerations

Ask to meet with the anesthesiologist prior to surgery to review your medical history, allergies, the type of anesthesia that will be used, and any additional risk factors. Issues you may want to discuss include:

- If the sedation will be performed by a board-certified anesthesiologist or a certified nurse anesthetist. If the patient is a child, make sure the provider has specialized training in pediatric anesthesia.
- Any previous problems with anesthesia including difficulty placing a breathing tube, nausea, or vomiting.
- If the patient has sleep apnea or a restricted airway.

Additionally, here are some other things to consider and complete before the surgery:

- Complete all pre-operative tests including physical exams, blood tests, or x-rays.
- Do not shave the area that is being operated on and report any cuts or wounds on or near the surgical site. Breaks or cuts in the skin can be a portal for bacteria to enter and cause infection.
- Ask if you should shower with an antibacterial body wash the night before and/or the morning of surgery to help reduce the amount of bacteria on the skin.
- Ask if antibiotics need to be taken before surgery, which is called antibiotic prophylaxis or premedication.
- Inform the surgeon if the patient has ever had a methicillin-resistant staph infection called **MRSA** (Mer-suh). If so, ask if the patient should be screened for carrying MRSA before having surgery.
- For adults, bring copies of your signed Medical Power of Attorney and Advance Directive documents to the hospital.

SECTION 2: THE DAY OF SURGERY



Before, during and after the moment when the patient is wheeled into the operating room, there are steps that you, as the patient and/or caregiver, can take to be an active participant in the surgery. The hours before surgery often are spent supporting and reassuring the patient, but it is also important to focus on the important actions that will help the patient obtain the best possible outcome, including:

Remind the staff of the scheduled meeting with the anesthesiologist, and make sure to have all of the important documents available. The surgeon should personally sign the surgical site in marking pen. Patients or parents may also be involved in this process and can add their initials to confirm the exact location of the surgery.

Take the time to confirm the specific surgical procedure one more time with the surgeon. Be sure to ask the surgeon and staff members if they will be taking a “time-out” in the operating room before starting the procedure and using a surgical checklist. Studies have shown that patients are safer when the team takes a “time-out” before starting the surgery to confirm that they have:

- The right patient
- The right surgical site
- The right procedure
- The presence of x-rays or scans
- The presence of any special tools or implants

If premedication with antibiotics is prescribed by the surgeon, be sure the medication has been ordered and confirm when it will be given. Caregivers, family and advocates should ask where to wait to receive information and updates. Confirm that you or your caregiver will meet with the surgeon, after the procedure to discuss the details of the surgery, while they are still fresh in the doctor’s mind.

Access the **World Health Organization’s Surgical Safety Checklist** for a list of steps the surgical team should take before and after surgery.

SECTION 3: AFTER SURGERY



“Patients need to advocate for themselves or for their family during the entire process of surgery,” says Dr. Soham Roy, MD, FACS, FAAP, Director of Pediatric Otolaryngology - Head and Neck Surgery at University of Texas Medical School at Houston and Children’s Memorial Hermann Hospital. “Postoperatively, this means being prepared as much as possible, which is not always an option with emergency surgeries. But with many elective cases, patients need a special diet postop, so you will need to get all that ready before you leave for the hospital. Or if you know you’ll be in a cast or a wheelchair for a week after surgery, have the necessary things at home. That kind of preparation makes your postop recovery a lot smoother and a lot less stressful for everyone.”

Once surgery is over, there still is a need for careful observation and communication with your healthcare team. Caregivers know best when you may be uncomfortable. Caregivers who are able to recognize subtle changes in the patient’s condition and report observations to the staff are a vital part of the post-surgical plan. Caregivers should consider the following steps:

- Talking to the surgeon after the procedure and asking if there were any complications or additional procedures needed that could impact or delay the recovery process.
- Confirming ways to reach the surgeon if there are any serious complications, and writing down any contact information given.
- Asking about who will be managing post-operative issues such as pain control and nausea as it is important to know which provider will handle these potential challenges.
- If the patient is receiving narcotic pain medications, be sure that vital signs are being monitored continuously.
- Being aware that narcotic pain medications often cause constipation, so asking if stool softeners or diet modification is needed to prevent this uncomfortable and potentially risky situation.
- If the patient was given antibiotics before surgery, be sure to ask when they should be stopped. Generally, antibiotics given as premedication before surgery are given as a one-time dose or stopped within 24 hours.
- Knowing that patients should be kept warm after surgery, controlling blood sugar if the patient is a diabetic, and using oxygen may help prevent post-surgical infections. Ask if these actions would be helpful.

Access the Empowered Patient® fact sheets **Important Questions for Your Hospital Care Team** and **Questions to Ask a Surgeon after a Procedure** for additional helpful information in the **Resource Guide** section.

SECTION 3: AFTER SURGERY

Discharge

Discharge is considered a “transition in care” and carries additional responsibilities for staff, patients, and caregivers. As you prepare to leave the hospital or surgery center, you need to understand what is expected of them and how they can help ensure that important information is shared and reviewed. The following will help you stay on track:

- Hospitals have discharge planners who can assist if help is needed at home such as additional caregivers, special equipment (oxygen, wheelchair, hospital bed, commode, etc.), or visiting nurses.
- Be sure you have a copy of the discharge instructions and that you understand them. Take notes on the page if you need to clarify any points. Be sure the instructions are clear about what to do and whom to call if you experience any complications at home.
- Review the discharge medications and dosages. Will medications need to be picked up from a pharmacy? How soon do they need to be started?
- Does the patient need any medications to prevent blood clots? Be aware that clots can happen even weeks after a surgery. Should the patient be up and walking several times a day to help reduce the odds of a blood clot?
- Plan to keep a log at home to help you keep track of all medications. Access a free patient journal to help track medications [here](#).

- Be sure you understand how to provide any wound care or deal with surgical drains.
- Will the patient need any physical or occupational therapy? When should therapy start, does insurance authorization need to be obtained, and who will schedule it?
- When can the patient remove bandages, drive, or return to work?
- When should the patient schedule a follow-up visit and which physician should they see?

Please access the **Empowered Patient® one-page fact sheet Preparing for Discharge** in the **Resource Guide** section and **Your Discharge Planning Checklist** www.medicare.gov/Pubs/pdf/11376.pdf from **Medicare.gov** www.medicare.gov.

SECTION 3: AFTER SURGERY

Care at Home

“In the case of certain surgeries your post-surgical care is just as, if not more, important than what you received in the hospital,” says Tom Christie, Chair/CEO of Relapsing Polychondritis Awareness and Support Foundation, Inc. “Probably the most important items in your discharge are your limitations. If there are things listed that you simply cannot do, tell them and they will make arrangements for someone to come help you with these tasks. For example, I live alone and they told me not to lift anything over five pounds. Now let’s be realistic, a small bag of groceries weighs more than that. They called the county and gave me a number to call to schedule someone to meet me and help me get my groceries put away.”

A significant part of the recovery process often happens in the patient’s home, which shifts several duties from trained medical staff members to the patient and/or their loved ones. Leaving the hospital or surgery center is often the beginning of new duties such as caring for the surgical site, managing medications at home, and monitoring the patient’s progress. Here are some points to consider when caring for yourself or someone who has undergone surgery:

- Should I track any vital signs at home including blood pressure, temperature, or weight?
- Are there any dietary restrictions or need nutritional supplements?
- Ask if any of the pain medicines contain acetaminophen or ibuprofen, which are the ingredients in Tylenol and Advil. If so, do not take additional Tylenol or Advil at home as it can cause serious side effects. Check the ingredients of any over-the-counter medications such as cold medication or sleep aids to be sure it does not contain acetaminophen or ibuprofen.
- Report to the physician any type of infection that develops at home, even if it seems unrelated to the surgery including infection in the mouth or teeth, skin, sinus, etc.
- Notify the physician if you notice any worrisome signs such as shortness of breath, redness or discharge from the surgical site, fever or excessive pain.
- Call the doctor as well if you notice loss of appetite, weight loss, depression, anxiety or insomnia as all of these issues can slow down the healing process.

Please access the **Empowered Patient® one-page fact sheet Care at Home** after Surgery fact sheet in the **Resource Guide** section and the **“Taking Care of Myself Guide”** from the Agency for Healthcare Research and Quality (AHRQ) for additional helpful information.

PERSPECTIVES



Lori Davis, Mother of a Child with Osteopetrosis

When my son was a baby, an eye doctor noticed that he had a “lazy eye.” We underwent four surgeries to correct the “lazy eye” before it became evident it was not just lazy but instead a result of the rare bone disease, Osteopetrosis, my son has. It was compressing the optic nerve, causing blindness.

In hindsight, those initial four surgeries were not needed and should not have been performed. As “uninformed” parents, we were unaware of the visual complications our disease could cause.

After understanding that the optic nerve was compressed and we would need to undergo two “optic canal decompressions,” I had started researching the surgery and realized that with our disease, the neurosurgeon would most likely break multiple carbide tip drill bits during the surgery. I explained this to the neurosurgeon who looked at me like I was from a different planet.

After the nine hour surgery was complete, he came out “excited” that he had in fact broken multiple drill bits (eight to be exact) which was something he had never encountered before. That was the turning point in our relationship with the neurosurgeon who in the future listened to the facts I had learned from my own research that related specifically to our rare disease.

Advice from Lori on Strengthening a Relationship with Your Surgeon:

Sometimes surgeons have a lot of patients they see, so it is always good to perform little tasks that set you apart and help your surgeon remember you. This could be by bringing in a written outline of your child and their disease, having a wallet sized card that explains their symptoms, or showing your appreciation for their time. After major appointments, I have brought in candy bouquets with written cards of appreciation. This shows my appreciation of their time and dedication to my son.

RESOURCE GUIDE



Governmental and Special Society Information

Agency for Healthcare Research and Quality's "**Having Surgery? What you need to know**" <http://archive.ahrq.gov/patients-consumers/diagnosis-treatment/surgery/questions/surgery.pdf>: This booklet was developed by Agency for Healthcare Research and Quality, part of the U.S. Department of Health and Human Services, and is intended for patients who are facing surgery that is not an emergency. The questions in this booklet may help you understand more about your surgery and whether it has to be done right away.

Agency for Healthcare Research and Quality's "**Taking Care of Myself: A Guide for When I leave the Hospital**" <http://www.ahrq.gov/patients-consumers/diagnosis-treatment/hospitals-clinics/goinghome/goinghomeguide.pdf>: When patients leave the hospital, there are many things they need to do to take care of themselves. They need to see their doctor, take medication, exercise, eat healthy, and know whom to call with questions or problems arise. This guide will help you stay on track when you leave the hospital or surgery center.

Agency for Healthcare Research and Quality's "**Quick Tips - When Planning for Surgery**" <http://www.ahrq.gov/patients-consumers/diagnosis-treatment/surgery/tips/tipsurgery.pdf>, **Spanish** <http://www.ahrq.gov/health-care-information/informacion-en-espanol/index.html>: The single most important way you can stay healthy is to be an active member of your own healthcare team. This tip sheet will help you take an active role in your care when planning for surgery.

American College of Surgeons' "**10 Questions to Ask Before Having an Operation**" <https://www.facs.org/education/patient-education/patient-resources/prepare/10-questions>: In addition to asking questions to the physician, there are many questions patients should be asking themselves; this list covers some of these.

Medicare's "**Your Discharge Planning Checklist**" <http://www.medicare.gov/Publications/Pubs/Pdf/11376.pdf>: This discharge planning tool is designed for patients who are preparing to leave a hospital, nursing home, or other care setting. The guide includes action items for patients and their caregivers, a medication tracking form and additional agencies who may be able to provide assistance.

The Joint Commission's "**Speak Up: Help Avoid Mistakes in Your Surgery**" http://www.jointcommission.org/assets/1/18/wrong_site_brochure.pdf: The Joint Commission is the largest healthcare accrediting body in the United States that promotes quality and safety in healthcare settings. As a patient, you can make your care safer by being an active, involved and informed member of your healthcare team by speaking up and asking questions before and after your surgery.

RESOURCE GUIDE



Empowered Patient Resources and Checklists

Empowered Patient Be Prepared for Your Surgery

http://engagedpatients.org/wp-content/uploads/2014/09/EP_be_prepared_for_surgery.pdf

Empowered Patient Care at Home After Surgery

<http://engagedpatients.org/care-home-surgery-fact-sheet/>

Empowered Patient Important Questions for Your Hospital Care Team

http://engagedpatients.org/wp-content/uploads/2014/09/EP_Important_Questions_For_Your_Hospital_Care_Team.pdf

Empowered Patient Preparing for Discharge

<http://engagedpatients.org/epc-preparing-discharge-fact-sheet/>

Empowered Patient Safety in Outpatient (Ambulatory) Surgery Centers

http://engagedpatients.org/wp-content/uploads/2015/04/Ten-Things_Outpatient_Surgery_Center.pdf

Empowered Patient Surgical Patient Checklist

http://engagedpatients.org/wp-content/uploads/2014/09/EP_surgical_patient_checklist.pdf

Empowered Patient Questions to Ask a Surgeon After a Procedure

<http://engagedpatients.org/questions-ask-surgeon-procedure/>

World Health Organization's "Surgical Safety Checklist"

http://www.who.int/patientsafety/safesurgery/tools_resources/SSSL_Checklist_finalJun08.pdf

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