

Issues to Discuss with Your Doctor Prior to Surgery in Friedreich's Ataxia Patients

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Preface

This paper documents the things I have learned since losing my son, Benj, following scoliosis surgery. I am not a doctor, but a parent who is trying to prevent the same tragedy from affecting other families. This paper is a result of my own experience, research and interviews with many members of the medical profession, including surgeons, neurologists, cardiologists, anesthesiologists and pathologists. It is intended to give the Friedreich's Ataxia patient and parents (the patient family) some basic information to help facilitate a discussion with your medical team prior to major surgery to prevent adverse outcomes related to fluid management and heart function. This article is not a comprehensive guide to all aspects of surgery.

Abstract

People with Friedreich's Ataxia (FA) are considered fragile patients and require additional pre-op planning and careful post-op care. Fluid management combined with good communication between the patient family and the entire medical team is critical to the surgical outcome. This is true for any person affected by FA. Even if the heart looks normal on echocardiogram and EKG, and presents no symptoms, the FA heart still has the genetic abnormality that makes it harder to generate energy, and produces more damaging free radicals due to higher iron levels. Under stress, the FA heart may not react as well as a non-FA heart.

Discussion

It takes a coordinated team of physicians to take care of complex or fragile patients. Thus, for FA patients undergoing surgery, it requires good communication and understanding between the surgeon, anesthesiologist, cardiologist (who needs to understand FA), and the Intensive Care Unit (ICU) doctors. One of the most important, proactive things that patients/parents can do for themselves/their FA children is to insist on good team management, and to insist on a group meeting between the surgeon, anesthesiologist, cardiologist, and themselves prior to surgery. This is often called a family meeting and serves to clarify who is responsible for what, and what the possible outcomes and treatments are after surgery. This is difficult to arrange but well worth it in the long run.

Pre-surgery

1. Since FA is degenerative, you are never healthier than you are today, so if surgery is going to be required for treatment you should consider it being done as soon as possible. Speak to your surgeon and cardiologist about treating the issue in an aggressive manner.
2. Prior to surgery, there should ALWAYS be an in-depth conversation between the physicians and the patient / parents about the expected outcomes, and potential adverse effects. This family meeting needs to occur between the surgeon, patient/parent, anesthesiologist and probably the neurologist.
3. Check with the surgeon to see if it is reasonable to have 2 surgeons working simultaneously so the length of time under anesthesia is decreased.

4. FA patients will receive the most comprehensively coordinated care in an academic center that has professional collaborations between surgery, anesthesia, cardiology, and ICU already established and on-going. Also, academic medical centers usually specialize in treating patients with complex conditions, such as FA.

5. The cardiologist's most important role is to make certain that the patient is in the best possible condition prior to surgery, and to communicate the patients' condition to the anesthesiologist and the surgeon before the operation so they can determine the best plan of action. Virtually all pediatric cardiologists today are well trained in Operating Room (O.R.) procedures, and pre- and post-op care (ICU). Centers with academic pediatric cardiology programs (including surgeons) usually have a pediatric cardiologist in the O.R. all day long or on call for the OR. They typically work very closely with the surgeons and ICU staff, and there is always one pediatric cardiologist on call for ICU or O.R. duty.

6. If possible, find an anesthesiologist that specializes in pediatric cardiology (anesthesiologists who are trained and board certified in both pediatric cardiology and anesthesia are rare and usually employed by academic medical centers). Speak to the anesthesiologist before the surgery. An anesthesiologist trained in pediatrics will usually have a greater understanding of fluid management in children relative to inherited defects. Again, make sure the anesthesiologist and the cardiologist are in agreement for the surgical care.

7. Ask about the use of a Trans Esophageal Echocardiogram (TEE) during surgery. The TEE is placed inside the esophagus so that they can image the heart throughout the surgery. A cardiologist can be in the O.R. or at a monitor watching the echo in real time with communication to the surgeon.

Surgery

8. Fluids must be carefully regulated during the surgery. This includes both blood and saline. Fluid management is tough because some types of surgery like scoliosis surgery cause a tremendous loss of blood. Additionally, there is the problem called 3rd spacing. When tissue is injured it swells. In order to do this it pulls the fluid out of the circulatory system. Scoliosis surgery causes a lot of tissue injury, so there is a large amount of fluid (liters and liters) that disappears from the circulatory system to the injured tissue (3rd space). The longer the surgery lasts the more fluid and blood is lost. Both the blood loss and the 3rd space temporary loss have to be replaced during surgery in order to maintain a good blood pressure.

9. When cardiac problems are known to exist prior to surgery, ask the cardiologist about the anticipated plan for evaluating the heart following the surgery.

Post-surgery

10. After surgery, the blood volume must be removed as the tissue recovers from the 3rd space effect. (See #11 below.) Most patients tolerate this, but FA patients cannot accommodate the fluid so easily. Thus, it is more likely for them to enter congestive heart failure after surgery, especially if there is cardiomyopathy present before surgery. The anesthesiologist is usually quite good about managing fluids during surgery. The surgeon, however, must be made aware of the

critical nature of the FA heart and how intolerant it can be to fluid shifts. Frequently, the surgeon will want to consult the pediatric cardiologist for help with management of the heart function in the postoperative care. (See #11 below.)

11. The 3rd postoperative day is the most critical! All the fluid that was hidden in the injured tissue (3rd space) begins to return to the circulatory system. A normal heart can easily handle this load because it delivers all the excess fluid to the kidneys where it is urinated away. An FA heart may be ill equipped for the extra strain. As a cautionary note, even a heart in an FA patient that looks normal on echocardiogram and causes no symptoms, still has the genetic abnormality that makes it harder to generate energy, control free radical production, and keep iron levels normal. Thus, under stress, the heart may not be as strong as or react in the same way as a heart in a non-FA patient. FA patients undergoing ANY major surgical procedure need close monitoring, even if the heart appears normal on pre-op screening. This is why many anesthesiologists and cardiologists may consider the prophylactic use of dialysis (ultra filtration) in FA patients to help prevent heart failure.

12. In addition to pre-operative consultation, postoperative care is also where the input of the cardiologist will be very helpful. The ICU doctors and the cardiologist should be working closely together. Make sure the cardiologist and anesthesiologist speak with the ICU doctor about the fragile nature of the heart in FA. Also, be sure you have requested that past echocardiograms, EKGs, and medical records be available for the ICU doctors. Since FA is a somewhat rare disease, the ICU doctors and nurses who will handle the postoperative care may never have seen a case of FA in the ICU before and will appreciate the information. Most academic centers have very close involvement of cardiologists with the ICU doctors (they are frequently the same person), and anesthesia doctors (again, anesthesiologists frequently run the ICU). The necessity for collaboration between different specialties cannot be over stated and is a must for FA patients.

13. Early mobilization after any surgery is critical. It is important to get the patient up and on their feet as soon as is physically possible and safe to do so. In the 1970's and 1980's many FA patients who had had scoliosis or foot surgery were emerging from the surgery with a significant worsening of their strength and ataxia. It is hard to get back without aggressive post-op rehab.

Conclusions

Good communication between the patient family and the medical team is critical to a successful surgical outcome in people with FA. Fluid management consultation and monitoring is very important in all persons with FA. This is especially true where cardiac problems are manifest. Aggressive rehabilitation after surgery is paramount to regain pre-surgical function.

Reviewed By: Drs. Mark Payne, Susan Perlman, Arnulf Koeppen, David Lynch and Lance Clyde

FARA Disclaimer: The information provided here should NOT be used as a substitute for seeking professional medical diagnosis, treatment, or care. You should NOT rely on any information in these pages to replace consultations with qualified health professionals. As stated

above, the information presented in this document results primarily from the experiences of one FA parent now trying to be helpful to other FA families that might be facing similar procedures. Although this information has been reviewed by a number of physicians expert in FA, FARA does not offer this information as medical advice but, rather, as the thoughtful reflections of an FA parent that you might want to consider, along with your own team of medical professionals, if your FA family is approaching a similar medical procedure.