



Diagnosed with Friedreich's Ataxia: Questions for your Cardiologist

Virtually all individuals with FA have a cardiomyopathy or some cardiac differences at some level, e.g., most individuals with FA will have flattened T waves on their EKG even when they don't have symptoms of heart problems or cardiomyopathy. Because FA is a rare disease some doctors are less familiar with the significant variability in the cardiac disease associated with FA. For example, some individuals have minor changes that are stable for long periods of time, and other individuals develop cardiac changes that progress quickly and can be life threatening. Unfortunately there is not much guidance for advice or treatment in the medical literature as this aspect of FA has not yet been adequately studied.

Below are some suggested questions for consideration for initial and follow-up visits with a cardiologist, related to the evaluation of the heart. Some of these questions may not be appropriate as they assume different types of symptoms which you or your child may or may not have. Use these questions to open a dialogue with your cardiologist and to become better informed about your/your child's cardiac function.

1. Based on your examination and studies, is there any evidence of obvious heart disease?
2. Is the heart abnormally thick?
3. Is the heart's ability to pump (systolic function) good? What is the Ejection Fraction (measure of the heart's ability to pump) or shortening fraction? How do these measurements for the Ejection Fraction or Shortening Fraction compare to "normal" measurements?
4. Is the blood flow into the left ventricle normal? Is there any evidence of diastolic dysfunction (declining function of one or both ventricles)?
5. Is there any evidence of arrhythmia (abnormal electrical activity of the heart) or other EKG abnormalities, like flattened T waves in the precordial leads?
6. I/My child frequently have/has symptoms of skipping heart beats, or beating funny. Is that cause for concern?
7. I/My child frequently have/has a fast heart rate. Is that important? When should I call you about a fast (or irregular) heart rate?
8. I/My child sometimes have/has chest pain. Is that important and when should I call you?

9. If I/my child have/has an episode of passing out, should I call you? Is that an important issue with cardiomyopathy?
10. Should I be seen on a yearly basis for an examination, echo, EKG, and holter monitor exam?
11. Can I/we get a copy of the EKG and Echocardiogram report? (It is important for you to maintain your own copies of these records because they could be very important to have on hand in case of an emergency, if you have to go to the ER and the doctors are not familiar with FA or if you need to quickly determine if your/your child's heart function has changed.)
12. Are there any symptoms or signs of heart problems in me/our child that you would want us to contact you?
13. From a heart standpoint, are there any activities you would recommend for or against in my/my child's case?
E.g., Any activity that includes a warning for "heart patients" such as hot tubs or amusement park rides does that apply to me/my child.

Be sure to tell the cardiologist if your child is on any prescription medications OR over-the-counter medications. Some medications, including anti-depressants or cold medicines, can interfere with heart medications.

For follow up visits and evaluations of the heart (select as indicated):

1. Has there been any thickening of the heart since I/we last saw you? Is this a change?
2. Is there any evidence that the pumping ability of the heart is decreased or altered since I/we last saw you? Is this a change?
3. Is there any evidence that blood flow into the left ventricle has changed? Is there any evidence of diastolic dysfunction (declining function of one or both ventricles)?
4. Is there any evidence of arrhythmias? Has this changed from my/my child's previous visit? Is the EKG the same as last time or has there been a change?
5. Is there any evidence of heart failure?
6. If my/my child's heart seems to be getting worse, should we see you more often? Should I/we be getting an EKG, Echo, and Holter monitor more often?

Should I/my child be started on a medication?
7. I/My child have/has started noticing that the heart is skipping beats, (or beating fast, or pounding in the chest, etc). Is that important? Do we need to monitor it by an event recorder or holter monitor?
8. Since I/we last saw you, I/my child seems to have less energy and endurance, and is more tired after work/school than a year ago. Is that important and could the heart function explain this?
9. Since I/we last saw you, I/my child get/s short of breath doing things that I/he /she used to do all the time. Is that important and can it be due to the heart?
10. I/My child am/is scheduled for a big surgery next month (e.g., scoliosis surgery). Is my/his/her heart strong enough to withstand this? Can you be available for consultation with the orthopedic and ICU doctors if they need a cardiologist?
11. I/My child sometimes wake/s up in the middle of the night with chest pain that goes away quickly. Should I call you with this?
12. If I/ my child have/has to go to the emergency room can we contact you? Can the ER doctor call you?

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