

Surface electrocardiographic features and prevalence of arrhythmias in paediatric Friedreich's ataxia

*Montañes M. E., Toledano M., Field E., Walsh H., Maguire K., Cervi E., Kaski J.P.
Great Ormond Street hospital, London, UK*

Introduction:

Friedreich's ataxia (FA) is a progressive neurodegenerative disease inherited as an autosomal recessive trait. The diagnosis is usually made on the basis of neurological symptoms with progressive ataxia. Cardiac involvement, in the form of concentric non obstructive hypertrophic cardiomyopathy (HCM), is very common (up to 90%). Heart failure in the 3rd and 4th decades is the most common cause of death. However, data on the prevalence of arrhythmias in this population are scarce.

Methods:

Clinical and electrocardiographic data from a cohort of 26 consecutive paediatric patients with FA evaluated between 2007 and 2016 were reviewed. Fourteen patients were female (54%) and mean age at diagnosis was 10 ± 3 years. Patients were followed up for a median of 3 years (interquartile range 2 to 6 years).

Results:

24 (92%) patients had echocardiographic features of HCM. Twenty five (96%) showed abnormalities on their baseline ECGs, with only one patient, without cardiac involvement, having a normal ECG. The most common abnormalities were: inferolateral repolarisation abnormalities [n= 24 (92%)]; voltage criteria for left ventricular hypertrophy [n= 19 (73%)] and pathological Q waves [n= 4 (15%)]. Three patients (11%) showed relatively small QRS voltages throughout and five (19%) had an abnormal QRS axis (4 with right axis deviation and 1 with left axis deviation). None had evidence of preexcitation or conduction disease.

20 patients (77%) underwent at least one 24 hour Holter monitor during follow up, including 8 patients (40%) who were symptomatic (chest pain, n= 4; palpitations, n=3; syncope, n= 1). Four patients (20%) were found to have tachyarrhythmias at follow-up: 2 (10%) had isolated short runs of nonsustained supraventricular tachycardia (SVT); 1 had an 8 beat run of non-sustained ventricular tachycardia; and 1 had an episode of paroxysmal atrial fibrillation (AF). There were no sudden arrhythmic deaths.

Conclusions:

ECG abnormalities are very common in FA, and 20% had non-sustained arrhythmias at follow-up. These results suggest that 24 hour Holter monitoring should be performed routinely during follow up to detect underlying arrhythmias in FA.