CASE REPORTS

A near fatal arrhythmia in teenager with 2 year history of benign sounding syncope

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Key points
Benign sounding syncope may have a serious or life-threatening underlying cause.

Abstract
Benign sounding syncope may have a serious or life-threatening underlying cause. This 14 year old boy had recurrent benign syncopal attacks but subsequently had 2 syncopal attacks with exercise. He was then diagnosed to have a cardiac cause of syncope. This case highlights that warning signs in the history may not be present at the initial presentation and diligence in taking the history of all subsequent episodes will help identify the small percentage of patients with a potentially life threatening cause.

Introduction
Dizziness and syncope are extremely common. It is estimated that up to 20% of children will experience an episode of syncope by the age of 15 years (1). Thankfully the vast majority of such cases are benign but a small proportion is associated with potentially life threatening conditions. Clinicians must be aware of the warning signs that suggest a significant underlying cause and therefore require further investigation.

Case Report
A 14 year old boy was referred to the Paediatric out-patients clinic by his General Practitioner (GP) with an eleven month history of recurrent episodes of syncope. The episodes usually occurred on days when he had not eaten breakfast or when he stood up quickly from sitting or lying. He would feel dizzy and then "black-out". Unconsciousness would last for less than a minute and he would make an immediate recovery. The patient never experienced palpitations and none of these initial episodes were related to exercise. The GP made a diagnosis of "simple faints" and referred the patient to the Paediatric out patient clinic due to the frequency of the episodes.

The patient was reviewed by the General Paediatrician who thought the likely diagnosis was vasovagal syncope. A baseline 12 lead electrocardiogram (ECG) showed a sinus bradycardia (rate of 48beats per minute) but was otherwise normal. In view of the bradycardia a 24 hour ECG was booked; this was normal. The patient and his parents were reassured and routine follow-up was organized.

Follow-up revealed the episodes of syncope to continue at a rate of approximately one per month. After 12 months the patient described two recent episodes of syncope that had been associated with exercise. The description of the episodes was exactly the same as previously but this was the first time they had been associated with exercise. The Paediatrician was concerned that this may suggest there was an underlying arrhythmia and therefore referred the child to the Paediatric Cardiologists.

In the cardiology clinic a repeat ECG was normal, as was an echocardiogram. The history was again thought to be in keeping with vasovagal syncope but in view of the exercise induced symptoms, an exercise stress test
was organised. This showed exercise-induced ventricular bigeminy (*figure 1*). The arrhythmia subsided with rest. The QT / QTc intervals were normal throughout the test.

In view of this result it was planned to see the patient urgently in the cardiology clinic, unfortunately prior to this appointment the patient had a witnessed cardiac arrest whilst at school. Basic Life Support was administered on site by teachers and when the Paramedics arrived the rhythm strip showed him to be in ventricular fibrillation (VF) (*figure 2*). He was shocked with 200J twice which converted him to asystole. He was intubated and given Adrenaline and Atropine which re-established sinus rhythm.

He was transferred to the local Accident and Emergency Department where he was stabilized, 12 lead ECG was normal. He was subsequently transferred to the regional Paediatric Intensive Care Unit where he was extubated after 24 hours. He woke appropriately and subsequent follow-up has shown the patient to have made a full recovery with no neurological deficit.

Whilst on the PICU the patient was reviewed by the Cardiology team. They thought the likely diagnosis was catecholaminergic polymorphic ventricular tachycardia. It was decided he should have an automatic implanted cardioverter defibrillator; this was inserted under general anesthetic without complications. At the same time he had an adenosine test to exclude the possibility of a concealed accessory pathway; there was no evidence of this. He remains under the follow up of the cardiology team and has had no further episodes of collapse or arrhythmia.

Figure 1: Exercise stress test ECG showing development of ventricular bigeminy

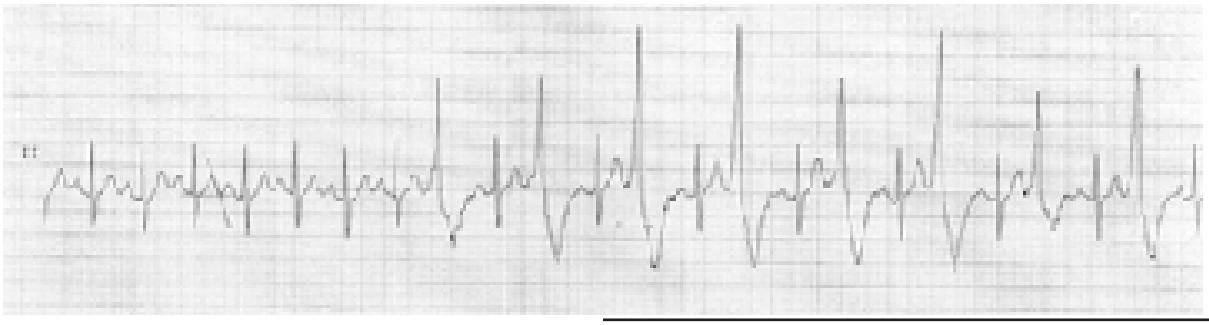
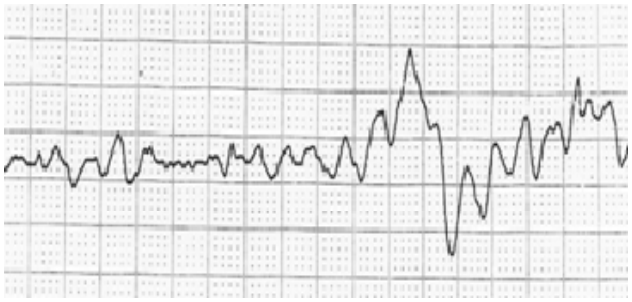


Figure 2: Initial paramedic rhythm strip showing ventricular fibrillation



Discussion

Childhood syncope can be classified in to three main groups: neurally mediated, cardiovascular and non-cardiovascular pseudosyncope (2). The vast majority of cases are benign neurocardiogenic or vasovagal syncope and therefore fall within the neurally mediated group. These episodes are caused by a transient disturbance in the autonomic control of blood pressure and heart rate leading to cerebral hypoperfusion and hypoxia. Depending on the degree of cerebral hypoxia the collapse may be associated with urinary incontinence and stiffening or fine twitching movements (1).

Arrhythmias or structural cardiac lesions account for the cardiovascular causes and it is these that although rare, can be life threatening. The structural cardiac lesions include aortic stenosis, hypertrophic obstructive cardiomyopathy and primary pulmonary hypertension. The arrhythmias that can cause syncope include supraventricular tachycardia (SVT), ventricular tachycardia (VT) (often in association with long QT syndrome) and heart block. Other cardiac causes of syncope include Tetralogy of Fallot spells and pump dysfunction (3).

The non-cardiovascular pseudosyncope group comprises of children whose episodes of syncope are actually seizures and children who present with psychogenic syncope.

The key to the diagnosis in syncope is a detailed history, during which the clinician must look for certain "warning signs" that may indicate a more serious underlying cause. As in this case one of the warning signs is syncope induced by exercise, others include: syncope in response to a fright / emotional stress, syncope when supine and a family history of sudden death in an adult < 30yrs (1).

The most important investigation in a child presenting with recurrent syncope is a 12 lead ECG. The main reason for this is to exclude long QT syndrome but also to look for heart block and ventricular hypertrophy. As in this case, it must always be remembered that a normal ECG does not exclude an underlying arrhythmia and if there are warning signs in the history further investigations may be warranted.

The majority of children with recurrent syncope have a benign cause. This case highlights that warning signs in the history may not be present at the initial presentation and diligence in taking the history of all subsequent episodes will help identify the small percentage of patients with a potentially life threatening cause.

References

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