Original article

Syncope in children and adolescents: a two-year experience at the Department of Paediatrics in Parma

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Abstract. *Background and aim of the work:* Syncope occurs frequently in childhood and adolescence. It generally is benign, but may be a sign of pathology. The purpose of this study is to determine the incidence of syncope in children and adolescents in the Parma area, analyse the frequency of syncope due to cardiac, neurological or metabolic diseases, to identify a diagnostic procedure based on careful review, clinical features, and results of investigations. *Methods:* The records of 156 children (age 1 to 18 years, mean age 10.5), who were referred to the Paediatric Department for syncope were reviewed. *Results:* The incidence of syncope in the paediatric population of Parma during a 2-year period (2005-2006) resulted in 86.5/100.000 per year. The cause of syncope was discovered in 154/156 cases. Cardiovascular syncope was diagnosed in 121/156 cases (77.5%). Neurological syncope was diagnosed in 31/156 cases (20%), while metabolic syncope was diagnosed in 2/156 cases (1.25%). The most frequent type was the (benign) neurocardiogenic syncope, but 3 cases were due to arrhythmia, 9 cases were due to epilepsy, 1 case was due to subarachnoid hemorrage and 2 cases were due to hypoglicemia. *Conclusions:* Syncope frequently requires medical examination or admission to paediatric departments and, in the majority of cases, does not subtend serious diseases. Nevertheless, syncope always requires a complete medical evaluation in order to exclude underlying pathology. An electrocardiogram must complete the initial evaluation of all patients with syncope. Other investigations are required if clinical data and the ECG do not provide an etiological explanation of the episode. (www.actabiomedica.it)

Key words: Syncope, neurocardiogenic syncope, head up tilt-test, epilepsy, arrhythmia, hysteria, hyperventilation, breath holding attacks

Introduction

Syncope is defined as a loss of consciousness and muscle tone, usually of brief duration. It has been estimated that about 15% of subjects will experience syncope between eight and eighteen years of age (1-3).

In the great majority of cases syncope is benign (4), but it can sometimes be the clinical manifestation of a cardiac, neurological, or metabolic disease. Therefore, in order to recognize the few cases in which syncope is the result of a disease, children and adolescents with syncope require a rational medical approach.

We studied all patients admitted to the Department of Paediatrics of the "Azienda Ospedaliero-Universitaria" of Parma in a two-year period.

The purpose of this study is 1) to determine the incidence of syncope in children and adolescents in the Parma area; 2) to analyse the characteristics of these episodes and the clinical presentation in order to identify possible correlations between the etiologic category of syncope and age, sex, type of patients, and clinical features, 3) to identify a diagnostic course.

Methods

Study setting

Population. The population of Parma and province on January 1st 2007, was 597,125 people, 288,060 males and 309,065 females. The population in paediatric age (0 to 18 years-old) was 91,315 (15.3% of the whole population); the number of males was 47,036 (51.5%) while the females were 44,479. On January 1st 2006 there were 592,621 people totally, 89,178 in paediatric age (15%), 45,797 males (51,3%) and 43,381 females.

Hospital. In the province of Parma the Department of Paediatrics of the " Azienda Ospedaliero-Universitaria" is the only first-aid paediatric unit and paediatric ward. For this reason we can reasonably assume that every paediatric patient that lives in the province of Parma and that has had a syncope, if evaluated in hospital for this episode, would have been in our Department, and this would also have included all pediatric patients seen first in the Parma Emergency Department.

Data analysis

The present study reviews the characteristics of syncope in children who presented to our hospital over a 2-year period.

We retrospectively identified the patients with syncope that had been evaluated in our department from January 1st 2005 to December 31st 2006.

Cases of syncope after head trauma or in patients with a previous diagnosis of epilepsy were not included in our study.

Based on the analysis of our patients' medical reports we classified syncope into three categories: cardiovascular, neurological, and metabolic. Every category was then further divided into subgroups (table 1).

Table 1. Classification of syncope

Cardiovascular:
Neurocardiogenic
Reflex
Mechanical
Arrhythmic
Vascular
- Orthostatic hypotension
- Cerebrovascular occlusive disease
- Subclavian steal syndrome
Neurologic:
Epilepsy
Breath holding spells
- pale
- cyanotic
Hyperventilation
Hysteria
Basilar artery migraine
Metabolic:
Hypoglicemia

Results

Incidence

We found 156 patients with syncope, 85 in the year 2005 and 71 in the year 2006.

Seventy-one were males and 85 were females.

The range of age went from 1 to 18 years (mean age: 10 years and 6 months).

The incidence of syncope in the paediatric population of Parma resulted in 86.5/100.000 per year.

Eighty-nine patients (57%) were evaluated as outpatients (0.3% of all ambulatory visits) and 67 patients were admitted to the hospital (1.2% of the admissions) in the period of time considered.

Types of syncope

In 154/156 cases we were able to make an aetiological diagnosis of syncope, while in 2 cases this was not possible (table 1).

Cardiovascular: One hundred twenty one syncope were diagnosed as cardiovascular (77.5%). The great majority of cardiovascular syncope were neurocardiogenic in type (107/121, 88%); 6.5% of neurocardiogenic syncope presented with brief myoclonus and 2.8% presented with brief muscular rigidity. Seven cases of cardiovascular syncope were reflex in type (5.8%), 4 were secondary to orthostatic hypotension (3.3%), and 3 cases (2.5%) were due to arrhythmias (one ventricular tachycardia in long QT syndrome, one ventricular tachycardia in left ventricular fibroma,

one atrial flutter in a Fontan patient with tricuspid atresia). No cardiovascular mechanical syncope was diagnosed in our study.

Neurological: Thirty-one out of 156 cases of syncope were classified as neurological (20%). Twelve cases of neurological syncope were classified as breath holding spells (38.7%), seven were pale and five were cyanotic. Nine out of 31 (29%) of neurological syncope were due to epilepsy. Seven out of 31 (22.6%) were due to hysteria, 2/31(6.4%) were due to hyperventilation and one case (3.2%) was secondary to subarachnoidal hemorrage. Epilepsy was diagnosed in 9 (5.7%) out of 156 cases in our study.

Metabolic: Metabolic aetiology was diagnosed in 2/156 cases of syncope (1.25% of all cases). Both of them were due to hypoglicemia (one was a patient with type I diabetes mellitus in insulinic treatment).

Age, circumstances of the episode

Table 2 summarizes the ages (range and mean) in aetiological groups of syncope.

Table 3 lists the circumstances in which each type of syncope occurred.

Table 2. Causes of syncope in our study

	Total	M	F	Mean age (range)
Total	156	71	85	10 yrs 6 mos (1 -18 yrs)
Cardiovascular	121			
1) Neurocardiogenic	107	50	57	11 yrs (2 yrs 6 mos -16 yrs)
2) Reflex	7	5	2	11 yrs 6 mos (8 -16 yrs)
3) Mechanical	0			
4) Arrhythmic	3	1	2	12 yrs 7 mos (5-18 yrs)
5) Vascular Orthostatic hypotension	4	2	2	13 yrs 3 mos (8 -15 yrs)
Neurologic	31			
1) Epilepsy	9	6	3	8 yrs 9 mos (17 mos -14 yrs)
2) Breath holding spells	12	4	8	2 yrs (1 -4 yrs)
Pale	7	1	6	1 yrs 5 mos (1 -2 yrs 6 mos)
Cyanotic	5	3	2	2 yrs 9 mos (1 yrs 4 mos -4 yrs)
3) Hyperventilation	2	1	1	15 yrs and 6 mos (15-16 yrs)
4) Hysteria	7	1	6	14 yrs 9 mos (13-17 yrs)
6) Subarachnoid Hemorrhage	1		1	11 yrs
Metabolic	2			
1) Hypoglicemia	2		2	14 yrs (14 yrs)
Undetermined	2	1	1	11 yrs and 4 mos (8 yrs 8 mos-14 yrs)

Discussion

Incidence

The incidence of syncope in this study resulted in 86.5/100.000 per year. Nevertheless we believe that the actual incidence of syncope in childhood may be higher, considering that some patients with syncope do not come to medical attention.

Driscoll et al. collected cases of syncope in two 4 year periods of time (1950-1954 and 1987-1991) and found that the incidence of syncope was 71.9/100.000 per year relatively to the first period of time and 125.8/100.000 per year in the second period (5).

In our study the type of syncope most frequently found was the neurocardiogenic one (68.5% of all cases); Massin et al. (6, 7), from a study of 226 cases of syncope in paediatric patients, concluded that the neurocardiogenic mechanism was responsible for 80% of cases; as in our study Massin et al. diagnosed cardiac arrhythmic syncope in 2% of patients and did not find mechanical aetiology.

Age

The mean age of the patients that were evaluated in this study was 10 years and 6 months (range from 1

Table 3. Circumstances in which syncope occurred

	Cardiovascular				Neurologic						Metabolic	Indeterminated
	NC	R	A	OH	E	cBHA	pBHA	HV	HYS	SaH	HY	
Circumstances												
Using hair-dryer	2											
Pain/ blood sample	11											
During or after effort			2						2			
Hot bath or shower	6											
Quarrel						1	1		1			
Defecation		1										
Miction		4										
Cough/Sneeze		1										
Fasting	6											
Menstruation	3				1							
Infection	31	2		2	1		1					
Consequent trauma	8				1							

NC: neurocardiogenic; HV: hyperventilation; R: reflex; HYS: hysteria; A: arrhythmic; SaH: subarachnoid hemorrhage; OH: orthostatic hypotension; HY: hypoglicemia; cBHA: cyanotic breath holding attack; E: epilepsy; pBHA: pale breath holding attack

to 18 years old), and is slightly lower than the mean age reported in the literature.

Gordon et al. (8) analyzed 73 patients with syncope from 1981 to 1986 with a mean age of 13 years (range from 2 to 20 years old).

Pratt and Fleisher (1) evaluated 77 children and adolescents who presented with syncope. These patients were 22 months to 21.7 years old, with a mean age of 12.7 years.

When classifying patients based on aetiology we found that patients with hysterical syncope were very homogeneous for age, which ranged between 13 and 17 years (mean 14 years and 9/12).

Patients with breath holding spells also show homogeneity for age which ranged between 1 and 4 years.

Sex

We only found a sharp difference between males and females in the hysteric type of syncope with a higher prevalence in females (86%). Although the number of our patients with hysteric syncope is low, our percentage is very close to that reported in the literature (9).

Clinical presentation

In the majority of cases, a carefully review with the description of the episode obtained from the patient and/or from a witness, and the patient's history, which may be positive for epilepsy, cardiac diseases, arrhythmias, or cardiac surgery, followed by a complete physical examination permitted the diagnosis or suggested other appropriate investigations (10).

Diagnosis of neurocardiogenic syncope is based on careful reconstruction of the episode, normal physical examination and normal ECG.

Neurocardiogenic syncope is characterized by a prodrome that last only a few seconds (pallor, palpitations, light-headedness, nausea, sweating, mental confusion). Neurocardiogenic syncope may occur after prolonged standing, fear, fright, pain, blood sample, sight of blood, hot and crowded places, emotion, or surprise. Some of these conditions such as fear, fright, emotion or surprise may also trigger the arrhythmic syncope of the long QT syndrome. Therefore, the characteristics of the episode cannot clarify all cases. The presence of a prodrome such as vertigo, visual symptoms, nausea with or without vomiting, paleness, and sweating is very frequent in neurocardiogenic syncope, but are not exclusive of this (11).

Some of these symptoms can also be present before an epileptic seizure, during reflex syncope, in the orthostatic syncope, in the hysteric syncope, and in syncope due to hypoglicemia. Similarly the lack of a prodrome and the sudden loss of consciousness are

typical of the cardiac type of syncope, but are not exclusive of these diseases.

In breath holding spells a careful reconstruction of the sequence of events (quarrel, pain, anger or frustration followed by apnea, cyanosis or pallor, and loss of consciousness in a child between one and four years of age) is suggestive for the diagnosis.

Hysterical syncope is defined as fainting due to, or to avoid, emotional stress. The diagnosis is often difficult; even if some characteristics such as the occurence of the episode in presence of other people, typically in adolescent females, the negativity of the medical examinations, the description of the event from people that were present, and the precise description provided with indifference by the patients often helps in making a correct diagnosis.

Syncope during exercise occurred in 4 patients of our study: in two patients the mechanism of syncope was arrhythmic and in two patients the cause was hysteria. Therefore syncope occurring during exercise is often due to cardiac disease and needs accurate cardiovascular investigation.

In our study the evaluation of the patient with syncope included an electrocardiogram (ECG) in 127 patients (81.5% of the cases). The ECG in our study allowed recognition of three causes of syncope due to arrhythmia (long QT syndrome, ventricular tachycardia in cardiac fibroma and atrial flutter in a Fontan patient).

There are diseases such as the long QT syndrome which cannot be diagnosed without an ECG, and can be provoked by fright, fear, or emotion like neurocardiogenic syncope.

The differential diagnosis between epilepsy and other types of syncope can also be difficult. A hasty review, a family history of epilepsy or a personal history of seizures during fever are causes of diagnostic confusion (12). Other misleading data may be a pathologic electroencephalogram (EEG) or an erroneous interpretation of this exam (13). In fact abnormal EEGs have been observed in normal subjects (14) and in subjects after syncope, especially in the recovery period or in syncope secondary to migraine. Similarly, the EEG can be normal in 50% of subjects with epilepsy. Furthermore, the presence of myoclonus or rigidity during the episode (15, 16) may generate more confusion: in fact, since cerebral hypoxia can occur in every case of syncope that lasts for more than 15 seconds, seizures can also appear in each type of syncope. All of these reasons can lead to an erroneous diagnosis of epilepsy. Jeavons evaluated 200 patients with a diagnosis of epilepsy and concluded that in 44% of these the episode could be defined as a syncope (15).

Punal et al. used the tilt table test in 9 patients (mean age of 11.7 yrs, range from 9 to 16 yrs) with a previous diagnosis of epilepsy, but whose clinical records suggested syncope rather than epilepsy on revaluation. This test was positive in all of their patients (16). The head upright tilt test is a useful and reliable diagnostic technique, allowing syncopal events to be induced and evaluated under controlled conditions and, in a subset of patients, it may help in distinguish epilepsy from syncope. Since tilt table test can result positive also in patients with epilepsy or in subjects that didn't experience syncope, this test could not be considered as a primary screening procedure. We agree with other authors (13,16) that it should be used only when the etiology of syncope remains uncertain.

In our study tilt table test was performed in 2 cases of syncope with uncertain etiology and was useful to prove neurocardiogenic syncope.

In cases of syncope with an unclear aetiology after careful cardiac and neurological investigation, an implantable event recorder may allow detection of an arrhythmic cause of the episode (17)

Conclusions

Syncope is a frequent event which requires medical examination or admission to paediatric departments and, in the majority of cases, is benign.

Neurocardiogenic syncope is the most frequent type and has a benign significance.

Nevertheless, initially a complete medical evaluation is necessary in order to exclude serious pathology that may cause syncope. In our series of 156 cases we diagnosed 15 cases with clinically significant etiology: 3 serious arrhythmias, 1 subarachnoid hemorrage, 9 cases of epilepsy and 2 cases of hypoglicemia, which represents 9.6% of the cases in this study.

An ECG must complete the initial evaluation of all patients who come to medical attention for syncope since it may reveal arrhythmias, pro-arrhythmic conditions, or other anomalies.

Other investigations should be performed in patients with abnormal ECG, abnormal cardiac auscultation, history of congenital heart diseases, operated or not, syncope occurring during effort, syncope preceded by thoracic pain or palpitations, family history of sudden cardiac death or long QT syndrome, episodes that last longer than two minutes, recurrent syncope, evidence of seizures, loss of sphincteric control, or any presentation that cannot be explained by a neurocardiogenic etiology.

EEG and neuroradiologic imaging procedures are recommended if neurologic aetiology is suspected.

If a cardiac aetiology is suspected cardiology consultation, echocardiography, holter-ECG and exercise-test are mandatory.

Tilt table test may be very useful in cooperative patients when, despite adequate neurological and cardiac studies, the aetiology of syncope remains obscure.

An implantable event recorder may help to clarify the aetiology of recurrent syncope when an arrhythmic cause is suspected but not documented.

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