Head-Up Tilt Test in Complicated Neurocardiogenic Syncope in Children

To confirm the usefulness of head-up tilt test (HUT) in neurocardiogenic syncope (NCS) with complicating clinical features, retrospective analysis were done on 12 selected children. The age at onset was 12.7±1.9(mean±SD) years. Associated clinical features were postoperative congenital heart disease (PO CHD) in 3, coexistent arrhythmia in 8 (persistent ventricular arrhythmia during exercise in 3, premature ventricular contractions in 2, ventricular couplets in 1, sinoatrial exit block in 1 and resting sinus bradycardia in 1) and ST segment depression during exercise in 1. Four of them had a history of exercise-related syncope. All 3 patients with PO CHD had arrhythmia (ventricular tachycardia in 1, sinus bradycardia in 1 and atrioventricular block in 1). HUT provoked NCS in 8 (2 during baseline tilt, 6 during isoproterenol infusion). In one each, ventricular tachycardia and loss of consciousness without hypotension and bradycardia were induced. Atenolol was tried in 5 with improvement of NCS in 4 and aggravation of dizziness in 1. During follow-up, 7 became asymptomatic (2 with atenolol) and 5 were stationary. In conclusion, HUT was valuable in diagnosing NCS even in children with complicating clinical features such as arrhythmias or PO CHD. HUT could be done as a part of initial diagnostic tests if the past history suggests NCS, regardless of associated clinical features. In some cases, the unexpected results of the test turned out useful in managing children with syncope or dizziness. *(JKMS 1997; 12 : 44~8)*

Key Words : *Syncope, Head-up tilt test, Children*

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INTRODUCTION

Syncope is a brief sudden loss of consciousness and postural tone due to decrease in cerebral blood flow or inadequate delivery of oxygen or glucose to the brain (1). In children, syncope is not uncommon and is possible from various causes, according to which the prognosis after syncopal episode is to be determined (2).

The major part of syncope in children with normal heart is neurocardiogenic syncope (3). Neurocardiogenic syncope is defined as the clinical syndrome of the syncope that results from inappropriate and excessive autonomic reflex from the cardiac ventricle. This type of syncope can be diagnosed by meticulous history taking and physical examination in most cases. However, in some cases of neurocardiogenic syncope, children may not complain of prodromal symptoms ; may drop suddenly causing physical trauma. Syncope may be associated with exercise or minor seizure (4). These aspects provoke difficulty in defining the cause of the syncope as a neurocardiogenic mechanism. Furthermore, if the syncope happens to occur in children with arrhythmia or abnormal hearts, it is almost impossible to define the cause of the syncope without reproducing syncope in the laboratory.

This study was aimed to evaluate the usefulness of the head-up tilt test in diagnosing the neurocardiogenic syncope in children with features causing confusion in diagnosis.

MATERIALS AND METHODS

Among children with possible neurocardiogenic syncope or near syncope presented to Seoul National University Children's Hospital since 1990, 12 children were selected for study because of complicating features (postoperative congenital cardiac defect in 3, normal heart with arrhythmia in 8 and ST segment depression during exercise in 1). Four children with normal heart (case 1~4) also had a history of exercise-related syncope or dizziness. Associated arrhythmias in children with normal heart were premature ventricular contractions not suppressed by exercise in 3 (case 4~6), frequent premature ventricular contractions in 2 (case 3, 7), ventricular couplets in 1 (case 8), sinoatrial exit block in 1 (case 9) and resting sinus bradycardia on routine 12-lead surface electrocardiogram (ECG) in 1 (case 2). The arrhythmias in 3 children with postoperative congenital heart defects were sustained monomorphic ventricular tachycardia (case 10),

Table 1. Profiles of the Patients

Case No.	Onset age	Sex	Symptoms	Cardiac defect	Associated arrhythmia
1	15	M	syncope with exercise	none	none*
2	13	F	syncope with exercise	none	sinus bradycardia
3	8	F	dizziness with exercise	none	frequent PVC's
4	13	F	syncope with exercise	none	PVC during exercise
5	15	M	syncope	none	PVC during exercise
6	12	M	syncope	none	PVC during exercise
7	13	F	syncope	none	frequent PVC's
8	12	M	syncope	none	PVC's, couplets
9	11	M	dizziness	none	sinoatrial exit block
10	13	M	dizziness	s/p TOF	ventricular tachycardia
11	12	F	dizziness	s/p ASD	1° & 2° AV block
12	15	M	syncope	s/p TOF	sinus bradycardia

* ST segment depression in inferior leads during exercise test and isoproterenol infusion
AV ; atrioventricular
ASD ; atrial septal defect
PVC ; premature ventricular contraction
TOF ; tetralogy of Fallot

atrioventricular block (first degree and Mobitz type I ; case 11) and sinus bradycardia in 1 (case 12) (Table 1). The laboratory evaluation of the syncope included 12-lead ECG, 24-hour ambulatory ECG monitoring, treadmill exercise test (with persistent standing for 10 minutes during the recovery period) and echocardiogram. In referred cases from neurologists, neurologic evaluations including electroencephalogram and brain computed tomogram or MRI had already been done. Electrophysiologic test was done in 2 children with postoperative tetralogy of Fallot. In one child with sustained monomorphic ventricular tachycardia, the episode of dizziness did not correlate well with the clinical ventricular tachycardia. In both of these children, ventricular tachycardia could not be induced by ventricular stimulation even during isoproterenol infusion. In a child (case 1) with ST segment depression in the inferior leads during exercise test and isoproterenol infusion, coronary angiogram was normal.

Head-up tilt tests were done according to the usual methods. Briefly, the test was performed in the morning after 4-hour fasting. The complete procedures were explained to the parents and the child to obtain informed consent and to relieve anxiety during the test. After attachment of the ECG electrodes and cannulation of the right radial artery for continuous blood pressure monitoring, the child was allowed to lie down in a quiet dark room for over 20 minutes. After recording the baseline heart rate and blood pressure in supine position, the child was positioned upright on the tilting table at the angle of 80 degrees for the maximum of 20 minutes with a footboard to bear the child's weight. In case of negative baseline tilt, the child was returned to the supine position for 10 minutes for stabilization. With continuous infusion of isoproterenol at a rate of 0.01 μg/kg/minute for 5 minutes in supine position, upright tilt was repeated as described above. If the result was negative, the child was returned to the supine position. The test was repeated with increased dose of isoproterenol (0.02 μg/kg/minute and 0.04 μg/kg/minute). A positive head-up tilt test was defined as a development of symptoms in association with bradycardia or hypotension or both, without regard to the loss of postural tone. A vasodepressor response was defined as a systolic blood pressure below 80 mmHg or reduction of systolic pressure more than 30 mmHg compared to initial head-up systolic pressure. A cardioinhibitory response was defined as a decrease in sinus rate below 60 beats per minute or persistent escape beats.

Head-up tilt test was discontinued if the result of the test was positive or the child became intolerant of isoproterenol with systolic blood pressure over 160 mmHg.

RESULTS

Head-up tilt tests were done on 12 children (7 boys and 5 girls). The age at the first episode of syncope was 12.7 ± 1.9 (mean $\pm$ SD) years (range, 8 to 15).

The result of the test showed typical neurocardiogenic syncope or near syncope in 8 children (two during baseline tilt - case 6, 9 ; six during isoproterenol infusion - case 1, 3, 5, 7, 11, 12), loss of consciousness without hypotension or bradycardia in 1 (case 2), ventricular tachycardia in 1 (case 10) and negative in 2 (case 4, 8) (table 2). The test was stopped prematurely because of severe palpitation and systolic hypertension over 160 mmHg during isoproterenol infusion in 1 with normal heart (case

Table 2. Results of Head-Up Tilt Test

Case No.	Response to Head-up tilt test	
	During Baseline Tilt	During Isoproterenol Infusion
1	−	+ (VD/CI)
2	LOC without VD/CI	not done
3	−	+ (VD/CI)
4	−	−
5	−	+ (VD)
6	+ (VD/CI)	not done
7	−	+ (VD)
8	−	−*
9	+ (VD)	not done
10	−	ventricular tachycardia
11	−	+ (VD)
12	−	+ (VD)

* Due to systolic hypertension and palpitation during isoproterenol infusion, test was stopped prematurely.
+ ; presyncopal or syncopal response
− ; no response
CI ; cardioinhibitory response
LOC ; loss of consciousness
VD ; vasodepressor response

8) and induction of ventricular tachycardia in 1 with postoperative tetralogy of Fallot and ventricular tachycardia (case 10) (fig. 1). In the latter, ventricular tachycardia was induced by tilting during isoproterenol infusion of 0.02 μg/kg/minute and subsided spontaneously on discontinuing isoproterenol infusion and lying down. In a child (case 11) with postoperative atrial septal defect and first and second degree atrioventricular block (Mobitz type I) on resting electrocardiogram, induced syncope was associated not with aggravated atrioventricular block but with sinus bradycardia.

After head-up tilt test, modification of life style such as dietary increase of sodium, no sudden upright posture, and limb exercise or lying down in case of dizziness were recommended in 7 patients. Beta blocker (atenolol) was tried in the other 5 patients with improvement in 4. In one with postoperative tetralogy of Fallot and sinus bradycardia, atenolol was discontinued after one month's trial because of the worsening of dizziness. Currently 7 patients are asymptomatic (2 with atenolol), 5 are stationary. There has been neither death nor worsening of syncope during a mean follow up period of 32.1 months (range, 8 to 73).

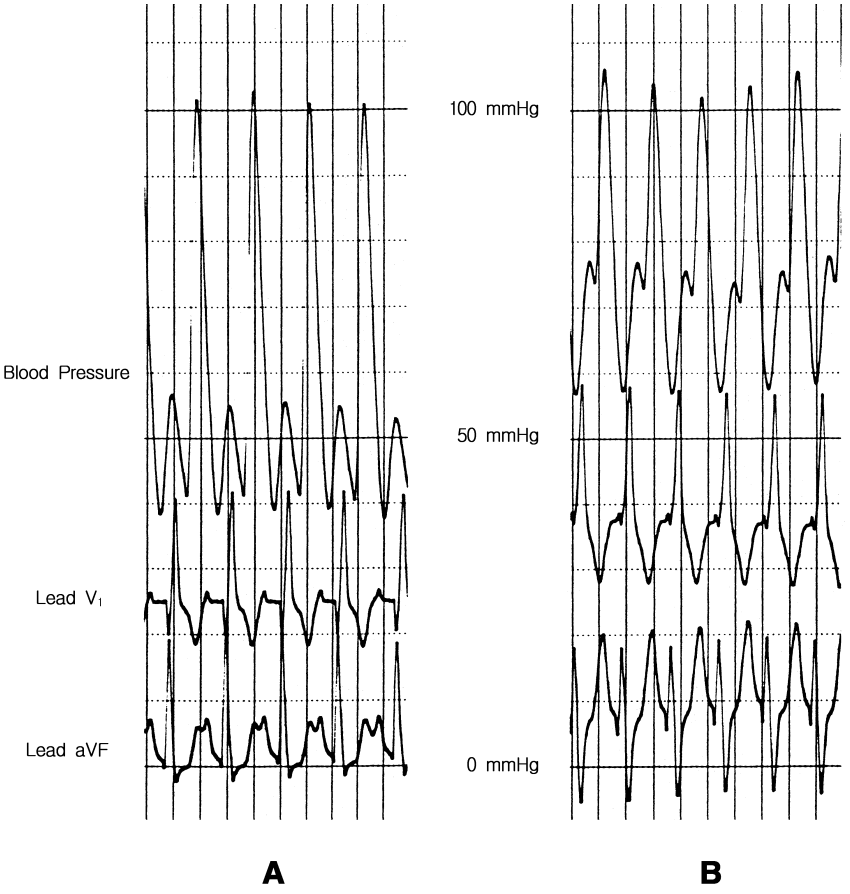

Fig. 1. Blood pressure and electrocardiogram (lead V1 and aVF) shortly before the induction of ventricular tachycardia (A) and during ventricular tachycardia (B) in case 10.

DISCUSSION

Neurocardiogenic syncope is defined as the clinical syndrome of the syncope that results from inappropriate and excessive autonomic reflex from the cardiac ventricle. It is regarded as benign and unrelated to sudden death in most cases. This study also confirmed the benignancy of the neurocardiogenic syncope regardless of the treatment. However, syncope may not always be benign from other point of view and can cause trauma or accident at the time of episode (5). Typical neurocardiogenic syncope can be defined by history and physical examination and differentiated from cardiogenic syncope which has a worse prognosis. However, atypical presentation of neurocardiogenic syncope mimics cardiogenic syncope, especially arrhythmia-related syncope, and the cause of symptoms can not be defined even after extensive diagnostic tests. Other features against the typical neurocardiogenic syncope are sudden syncope without prodromal symptoms, syncope related to exercise, syncope causing physical trauma or syncope associated with convulsion or recurrent seizures (4).

In children with long QT syndrome, catecholamine-sensitive ventricular tachycardia or a concealed form of arrhythmogenic right ventricular tachycardia, recurrent syncopes are usually due to ventricular tachycardia and occur during excited state. However, exercise-related syncope can also occur as a part of neurocardiogenic syncope (6). This episode is related to bradycardia or asystole and/or hypotension and has clinical features which are similar to that of ventricular tachycardia-related syncope, besides the fact that syncope usually occurs after exercise rather than during exercise. Because of concealed abnormalities in children with exercise-related ventricular tachycardia and difficulty in defining neurocardiogenic syncope in children with syncope-unrelated cardiac abnormalities such as coexisting ventricular arrhythmia, it is very important to reproduce syncope in the laboratory. In exercise-related neurocardiogenic syncope, the head-up tilt test is a very useful diagnostic tool (7). The other advantage of the head-up tilt test is that concomitant infusion of isoproterenol with tilting can induce ventricular tachycardia or TU complex changes in long QT syndrome and some exercise-related ventricular tachycardia (8).

Syncope can occur in patients with abnormal heart and this leads to considering cardiac causes of syncope firstly even if history suggests neurocardiogenic syncope. If significant residual cardiac defects can be excluded, syncope in children with postoperative congenital cardiac defects is usually regarded as having originated from tachycardia such as ventricular tachycardia or atrial flutter with 1:1 atrioventricular conduction, or from bradycardia such as sinus arrest, atrioventricular block or pause after termination of tachycardia. These facts put emphasis on the electrophysiologic test in children with syncope with postoperative congenital cardiac defect. However, considering the facts that syncope in these children may not be an absolute marker of sudden death and can be neurocardiogenic syncope, the head-up tilt test is very important as a screening test in these children before electrophysiologic test.

The head-up tilt test has been regarded as the test provoking the clinical neurocardiogenic syncope. The simultaneous infusion of isoproterenol during tilting increases the sensitivity of the test (9). Although there are reports about the statistical significance according to various protocols, the head-up tilt test is considered specific if the tilting degree is less than 80 degrees and the dose of isoproterenol is not high (10, 11). In our study, we used steep angle of 80 degrees to increase sensitivity. With this tilting degree the usual tilting duration was reported between 10 and 30 minutes (12). Therefore we selected modest duration of 20 minutes. Although recent analysis of head-up tilt test in normal children without a history of syncope indicates relatively high false-positive results, the statistical significance of head-up tilt test is quite acceptable even in children (13, 14). Another usefulness of the head-up tilt test exists when the outcome of the test is not typical of neurocardiogenic syncope. Whether this type of result indicates true neurocardiogenic syncope is not clear (15). In our study, two patients had positive but atypical results of the test : loss of consciousness without hypotension and bradycardia in one and induction of sustained monomorphic ventricular tachycardia which was not induced during electrophysiologic test in the other. The former case implies the occurrence of significant cerebral vasoconstriction prior to the systemic hypotension or bradycardia and uncommon pattern of neurocardiogenic syncope (16). The latter case indicates that disturbed autonomic equilibrium can create the optimal environment for tachycardia (17). Tilt test in a child with PR prolongation and Mobitz type I atrioventricular block revealed only sinus bradycardia at the time of near syncope during the test. These results gave useful information in evaluating the cause of the dizziness and planning the management.

In conclusion, the head-up tilt test could be an effective method in diagnosis of neurocardiogenic syncope in children with complicating clinical features such as abnormal heart, arrhythmias or exercise-related syncope. By performing head-up tilt test in symptomatic children, the cause of the syncope can be defined easily without a complicated test in a cost-effective way. In some cases, the unexpected results of the test turned out very useful

in managing these children with syncope or dizziness.

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