

Vasovagal syncope in monozygotic twins

Tek yumurta ikizlerinde vazovagal senkop

Erdoğan Arıkan, Murat Yeşil, Zeynep Apalı, Nursen Postacı, Serdar Bayata

Department of Cardiology Atatürk Education Hospital, İzmir Turkey

Introduction

Vasovagal syncope (VVS) is a transient loss of consciousness due to hypotension and/or bradycardia associated with abnormal autonomous activity. It is more common in the young adult population. Head-up tilt testing (HUT) is a gold standard tool to assess the etiology of syncope; albeit there are some drawbacks like low reproducibility and being affected by more than a few factors. The sensitivity of the test to show vasovagal syncope is reported to be between 32 to 85%, while the specificity is higher (1). Vasovagal syncope is classified according to VASIS as four types (2). Type I: mixed response. Type IIa: cardioinhibitory response without asystole. Type IIb: cardioinhibitory response with asystole. Type III: vasodepressor response. In spite of the fact that VVS is commonly a benign entity, some patients may experience recurrent life-threatening syncopal attacks. Familial VVS is even more uncommon based on a few reports. Here we present a familial form of VVS in monozygotic twins and their parents.

Case Report

A sixteen-year-old teenager was admitted to the hospital for having had sudden loss of consciousness while walking without any warning symptoms one week before. He had no preexisting health problems. He lived in a rural area where he worked as a shepherd with his monozygote twin brother. His Holter recording revealed an asystolic pause of 8 seconds, which caused syncope at midnight after getting up from sleep and just following micturition and 6 seconds of asystole during daytime while standing. A transient pacemaker was placed as his syncopal attacks were recurrent and traumatic. His physical examination was normal. His heart rate was 50bpm on resting electrocardiogram (ECG) which was otherwise normal. Routine biochemistry and hemogram were normal. His echocardiogram showed no anatomic or functional abnormality. A HUT was performed and 4-5 seconds after 80 degree tilting, the patient developed asystole and syncope for which we had to reactivate his turned-off temporary pacemaker. He was diagnosed as VVS type IIb. Beta-blocker therapy was avoided as he was already bradycardic at rest. Implantation of a permanent pacemaker with rate-drop-response was decided due to the recurrent, unexpected and traumatic nature of the syncopal attacks with a Class IIb indication

according to the guidelines (3). His monozygotic twin brother also deserved attention as he had had fainting episodes and mixed type syncope on a previously performed HUT. His Holter recording revealed no abnormality. The HUT revealed a 14 seconds of asystole (type IIb) at 80 degrees for which he was shortly resuscitated. However, device therapy was not planned due to lack of recent, recurrent or traumatic events. Their parents had no history of syncope or any other health problems at all. They had normal blood values, normal ECG and echocardiograms. The mother, age 35, showed a cardioinhibitory response (type IIb) on HUT, while the father, age 41, showed a cardioinhibitory response without asystole (type IIa). Six months after pacemaker implantation, our patient has not yet reported any syncope.

Discussion

Mathias et al. (4) observed a positive family history of VVS in 90% in subjects < 20 years but not adult-onset VVS. Serletis et al. (5) found that an individual with two fainting parents was more likely to faint. Offsprings of either sex whose mother faints are more likely to faint. Having a father who faints increases the risk of syncope in sons. While sociocultural, psychological, or environmental factors most commonly predispose to syncope, the role of genetics merits further molecular investigation (6). The efficacy of pacemaker therapy was questioned after two recent trials (7, 8); which failed to prove the superiority of pacing over placebo in 'unselected' patients with positive tilt table testing (mostly without cardioinhibition). In another study, DDI pacing significantly reduced the likelihood of syncope with asystole (9). The ISSUE 2 study (10) indicates that the patients with asystolic tilt response are likely to benefit more from pacing therapy because their spontaneous syncope is also asystolic. Hence, HUT limits greatly the number of candidates for pacemaker therapy since a cardioinhibitory response occurs in about 10% of patients.

Conclusion

Pacemaker therapy is not devoid of complications; therefore, in neurally mediated syncope, it seems prudent to limit pacemaker use to a few selected, recurrent, severely symptomatic patients with asystole who are particularly prone to injuries.

Address for Correspondence/Yazışma Adresi: Dr. Erdoğan Arıkan, Atatürk Education Hospital Department of Cardiology, İzmir Turkey
Phone: +90 232 243 43 43 / 2426 Fax: +90 232 244 91 15 E-mail: arikanmd@hotmail.com

©Telif Hakkı 2008 AVES Yayıncılık Ltd. Şti. - Makale metnine www.anakarder.com web sayfasından ulaşılabilir.
©Copyright 2008 by AVES Yayıncılık Ltd. - Available on-line at www.anakarder.com

References

1. Beneditt DG, Ferguson DW, Grubb BP, Kapoor WN, Kugler J, Lerman BB, et al. Tilt-table testing for assessing syncope. An American College of Cardiology expert consensus document. J Am Coll Cardiol 1996; 28: 263-75.
2. Brignole M, Menozzi C, Del Rosso A, Costa S, Gaggioli G, Bottoni N, et al. New classification of haemodynamics of vasovagal syncope: beyond the VASIS classification. Europace 2000; 2: 66-76.
3. Vardas PE, Auricchio A, Blanc JJ, Daubert JC, Drexler H, Ector H, et al. Guidelines for cardiac pacing and cardiac resynchronization therapy. The Task Force for Cardiac Pacing and Cardiac Resynchronization Therapy of the European Society of Cardiology. Eur Heart J 2007; 28: 2256-95.
4. Mathias CJ, Deguchi K, Bleasdale- Barr K, Smith S. Familial vasovagal syncope and pseudosyncope: observations in a case with both natural and adopted sibilings. Clin Auton Res 2000; 10: 43-5.
5. Serletis A, Rose S, Sheldon AG, Sheldon RS. Vasovagal syncope in medical students and their first degree relatives. Eur Heart J 2006; 27: 1965-70.
6. Márquez MF, Urias KI, Hermosillo AG, Jardón JL, Iturralde P, Colín L, et al. Familial vasovagal syncope. Europace 2005; 7: 472-4.
7. Connolly SJ, Sheldon R, Thorpe KE, Roberts RS, Ellenbogen KA, Wilkoff BL, et al. Pacemaker therapy for prevention of syncope in patients with recurrent severe vasovagal syncope: second vasovagal pacemaker study (VPS II). JAMA 2003; 289: 2224-9.
8. Raviele A, Giada F, Menozzi C, Speca G, Orazi S, Gasparini G, et al. The vasovagal syncope and pacing trial (Synpace). A randomized placebo-controlled study of permanent pacing for treatment of recurrent vasovagal syncope. Eur Heart J 2004; 25: 1741-8.
9. Sutton R, Brignole M, Menozzi C, Raviele A, Alboni P, Giani P, et al. Dual-chamber pacing in treatment of neurally-mediated tilt-positive cardioinhibitory syncope. Pacemaker versus no therapy: a multicentre randomized study. Circulation 2000; 102: 294-9.
10. Brignole M, Sutton R, Menozzi C, Garcia-Civera R, Moya A, Wieling W, et al. Lack of correlation between the responses to tilt testing and adenosine triphosphate test and the mechanism of spontaneous neurally-mediated syncope. Eur Heart J 2006; 27: 2232-9.

Ventricular septal diverticule and ventricular septal defect after penetrating cardiac trauma

Penetran kardiyak travmadan sonra gelişen ventriküler septal divertikül ve ventriküler septal defekt

F. Ayşenur Paç, Deniz N. Çağdaş

Section of Pediatric Cardiology, Yüksek İhtisas Education and Research Hospital, Ankara, Turkey

Introduction

Traumatic ventricular septal defects are rarely encountered. The incidence of ventricular septal defect (VSD) is about 4.5% among the cardiac traumas (1). We report a case of traumatic ventricular septal defect after a penetrating cardiac trauma causing pericardial tamponade and cardiac rupture.

Case Report

A 14-year-old boy was recalled to the emergency department because of a penetrating chest trauma due to stab in the 4th intercostal space. After 1 hour, he was admitted to the emergency service. Initial examination revealed dyspnea, tachycardia (125/min), and hypotension (60/30 mm Hg). Urgent echocardiographic examination revealed pericardial effusion and thrombus-like appearance in the pericardial space. The patient underwent surgery via a left anterior thoracotomy. A perforation was noted in the right ventricle, and a small amount of blood was seen in the pericardial cavity. The perforation was repaired with direct suture closure using Teflon pledgets. No other cardiac injury was noted at the time of that operation.

Several months after the operation, control physical examination revealed a new systolic murmur. By two-dimensional transthoracic echocardiography (TTE) a very small muscular ventricular septal defect was seen. So, cardiac catheterization and angiography were performed 5 months after the cardiac trauma. It revealed a saccular lesion at the upper interventricular septum, elongating to the right ventricle. Very little contrast media was crossing to the right ventricle from the centre of this lesion (Fig. 1).

The murmur of the ventricular septal defect was still present on physical examination one year after the trauma. Ventricular septal defect was also confirmed by echocardiography.

Discussion

Penetrating cardiac trauma in children is rarely reported in the literature. It is life-threatening and often requires urgent surgical intervention. It is not always limited to the free wall of the heart or the great arteries; it can cause damage in more than one of the cardiac structures. It may also involve the interventricular and interatrial septa, cardiac valves, coronary arteries, and conduction system (2). Traumatic injuries of heart reported before are atrioventricular valve insufficiency, aortic

Address for Correspondence/Yazışma Adresi: Prof. Dr. F. Ayşenur Paç, Yüksek İhtisas Education and Research Hospital, Section of Pediatric Cardiology, Ankara, Turkey Phone: +90 312 306 17 24 Fax: +90 312 312 41 20 E-mail: aysepac@gmail.com

©Telif Hakkı 2008 AVES Yayıncılık Ltd. Şti. - Makale metnine www.anakarder.com web sayfasından ulaşılabilir.
©Copyright 2008 by AVES Yayıncılık Ltd. - Available on-line at www.anakarder.com