Pectus excavatum: Right ventricular compromise with orthostatic syndrome and Brugada phenocopy

Eftychios Siniorakis [a], Spyridon Arvanitakis [a,*], Panagiotis Tzevelekos [a], Stamatia Panta [a], Apostolos Balanis [a], Fotini Aivalioti [a], Sotiria Limberi [a]

[a] Cardiology Department, Sotiria Chest Diseases Hospital, Athens

[a] Greece

Pectus excavatum (PEXT) consists of an overgrowth of the chondral region with posterior displacement of the inferior part of the sternum, resulting in a concave chest deformity. Characteristic clinical and imaging findings may occur, depending on the compression that right cardiac chambers suffer, when squeezed between the sternum and the column vertebrae.

Keywords: Brugada phenocopy, Orthostatic syndrome, Pectus excavatum, Ravitch procedure, Right ventricular compression

Pectus excavatum (PEXT) or funnel chest is an anterior chest congenital deformity, predominantly manifesting in male patients, with an incidence of 1 in every 300–400 births. It consists of an overgrowth of the chondral region with posterior displacement of the inferior part of the sternum, resulting in a concave chest deformity [1,2]. Although PEXT is viewed as a benign condition with mainly cosmetic interest, characteristic clinical and imaging findings may occur, depending on the compression that right cardiac chambers suffer, when squeezed between the sternum and the column vertebrae [3,4].

A 23-year-old man (height 1.83 cm, weight 68 kg, body surface area 1.90 m^2) presented for a routine cardiac examination, prior to undergoing surgical correction of his PEXT (Fig. 1A). Although his main intention was to get relief from cosmetic embarrassment, he was not completely free of symptoms. He reported exertional dyspnea, atypical chest discomfort, reduced endurance, and frequent orthostatic hypotension episodes. His past medical history and family history were clear. In particular, there was no record of syncope or sudden cardiac death.

Clinical examination in the supine position was unremarkable, apart from PEXT. On standing

Disclosure: Authors have nothing to disclose with regard to commercial support.

Received 6 November 2016; revised 6 December 2016; accepted 23 January 2017.
Available online 2 February 2017

* Corresponding author at: 10 Evrou Street, Ampelokipi, 11528 Athens, Greece.
E-mail address: sarvanitakis@doctors.net.uk (S. Arvanitakis).

P.O. Box 2925 Riyadh – 11461KSA
Tel: +966 1 2520088 ext 40151
Fax: +966 1 2520718
Email: sha@sha.org.sa
URL: www.sha.org.sa

Peer review under responsibility of King Saud University.
URL: www.ksu.edu.sa
http://dx.doi.org/10.1016/j.jsha.2017.01.003

Production and hosting by Elsevier

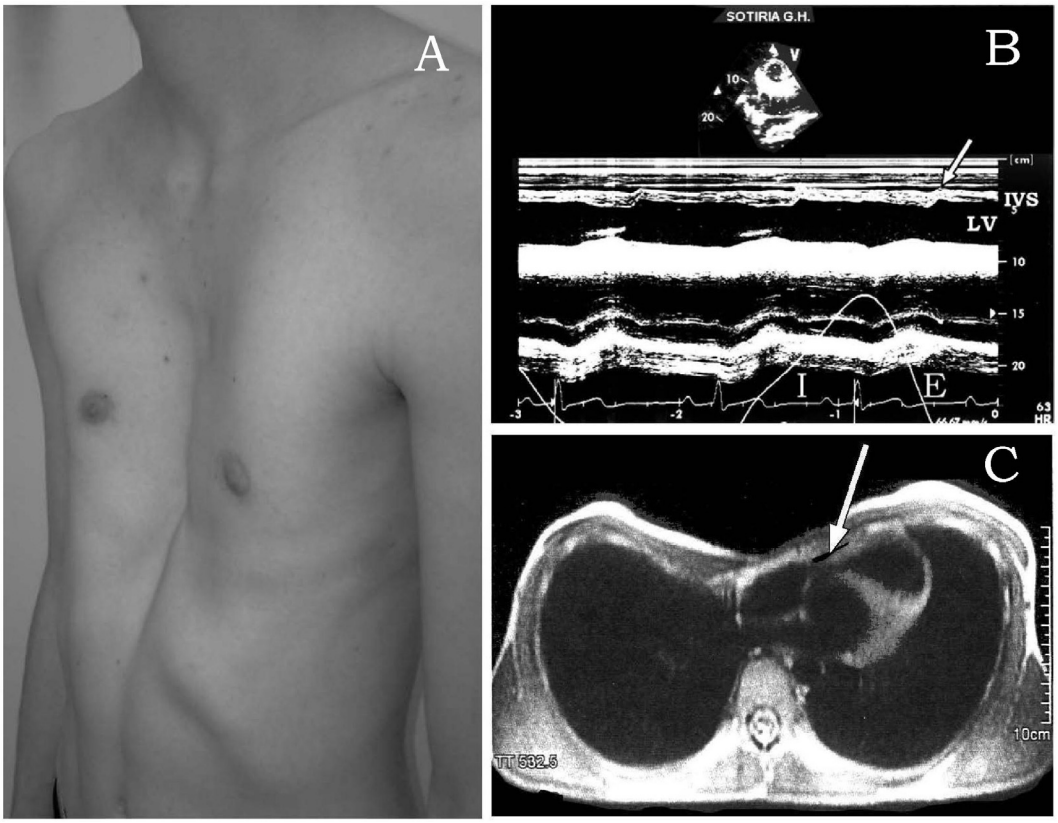

Figure 1. Constrictive pathophysiology of pectus excavatum on imaging. (A) Pectus excavatum provoking a squeezing of the right ventricle and abnormal motion of the interventricular septum, compatible with a constrictive pathophysiology, as seen on (B) echocardiography (arrow). (C) Magnetic resonance shows a filiform right ventricular outflow tract (arrow). LV = left ventricle; IVS = interventricular septum; I = inspiration; E = expiration.

upright symptomatic orthostatic syndrome was noted, consisting of a fall in systolic blood pressure by 50 mmHg, an increase of his heart rate by 30% and concomitant dizziness and near-fainting.

An enlarged cardiac silhouette was evident in the chest X-ray with leftward displacement of the heart. The usual laboratory blood tests including electrolytes and arterial blood gases were normal. His electrocardiogram (ECG) showed sinus rhythm with right axis deviation and leads V1–V2 depicted an rSR pattern with down-sloping ST segment and negative T waves (Fig. 2A). In lead V3, he had bifid T waves with a prominent late component. P waves in V1 were negative, a common finding in many PEXT cases. Given that his ECG picture was mimicking Brugada phenocopy, intravenous procainamide was administered to unmask a possible true Brugada substrate. This did not provoke any ECG changes.

Transthoracic echocardiography showed compression of the right atrium and right ventricle (RV), with reduced tricuspid annular plane systolic excursion. Short axis M-mode imaging (Fig. 1B) revealed a sudden pulling of the interventricular septum towards the anterior chest wall at the onset of diastole, causing almost total obliteration of the RV cavity. This was evident in inspiration as well as expiration, resulting in a constrictive pathophysiology. Potential intracavitary pressure gradients were not recorded due to positional deformities of the RV. Imaging and function of the great vessels, left heart cavities, and valves was normal.

Magnetic resonance imaging (MRI) revealed squeezing of the RV outflow tract with reduced RV ejection fraction at 48% (Fig. 1C).

The sternovertebral distance was 2.40 cm, resulting in a Haller index (transverse diameter of the chest divided by the anteroposterior diameter) of 3.80 (normal values around 2.50). Six months after the uneventful surgical correction of his chest deformity, by a Ravitch procedure, he was reassessed. His symptoms were no longer present and his orthostatic syndrome could not be reproduced any more. His ECG findings in leads V3 and V4 were attenuated (Fig. 2B) although negative P waves in leads V1-V2 persisted. RV was

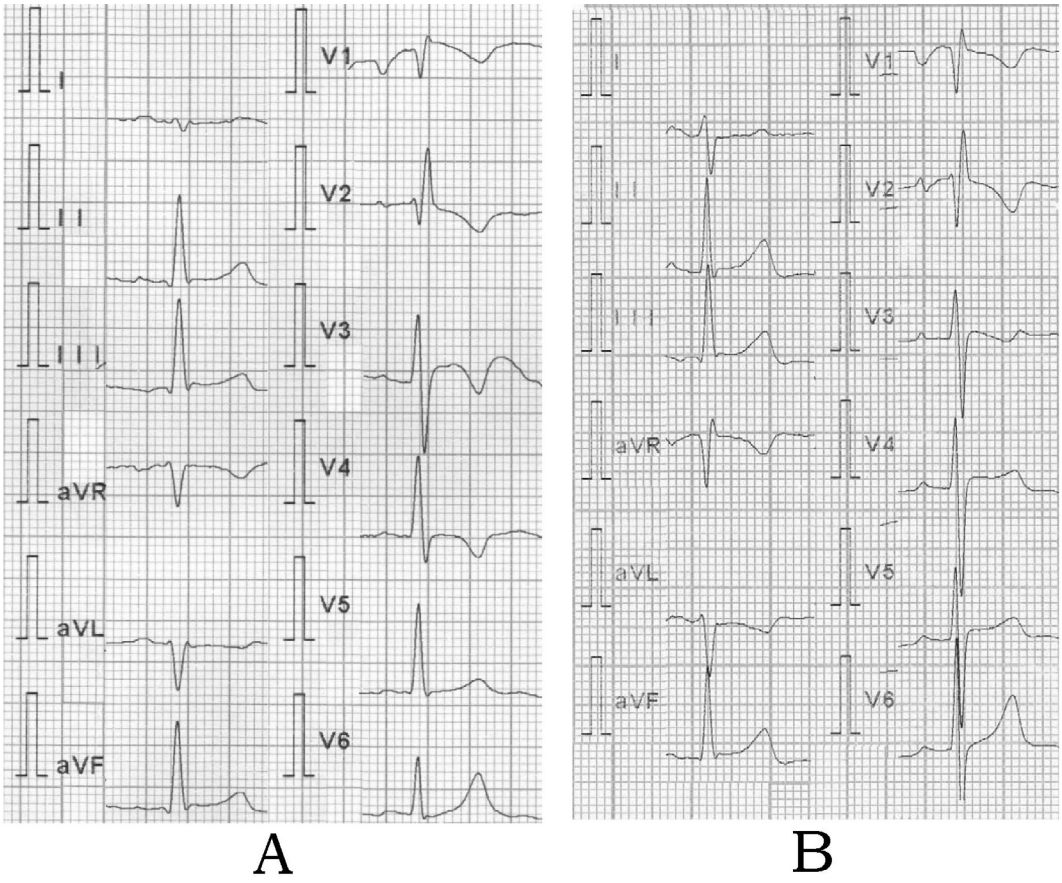

Figure 2. Brugada phenocopy of pectus excavatum patient. Electrocardiograms (A) before surgical correction, mimicking a Brugada phenocopy, and (B) attenuated after operation.

decompressed on ECG, with normal tricuspid annular plane systolic excursion, and RV ejection fraction had risen to 56% on MRI. The sternovertebral distance was increased to 5.10 cm, resulting in a Haller index of 2.0.

Several clinical implications can be deduced from the case described here. PEXT is not of esthetic concern only. Potential hemodynamic consequences related to the compression of cardiac chambers should be estimated cautiously, starting with a thorough clinical examination and proceeding with echocardiography and MRI if necessary [5–7]. Symptoms like orthostatic hypotension can be related to a reduced venous return in an already compromised RV, especially in several positional changes of the patient [8]. Common vasovagal episodes should be differentiated from syncopal episodes, especially if the ECG displays a Brugada phenocopy [9,10]. In this respect, provocation tests with sodium channel blockers, as well as the absence of a family history of sudden cardiac death can be useful in excluding Brugada syndrome [11,12]. Cardiac imaging by echocardiography is mandatory, to exclude hemodynamic derangement.

Surgical correction of PEXT performed in centers with accrued experience is expected to ameliorate the clinical and imaging characteristics of the disease, although this is not always possible [13,14]. ECG abnormalities may persist, probably due to the chronicity of the disease.

PEXT is an intriguing congenital condition. A multidisciplinary approach including cardiologists, pneumonologists, and thoracic surgeons is bound to uncover further aspects of the deformity, streamlining a holistic and individualized therapeutic strategy.

References

[1] Kelly Jr RE. Pectus excavatum: historical background, clinical picture, preoperative evaluation and criteria for operation. Semin Pediatr Surg 2008;17:181–93.

[2] Lesbo M, Tang M, Nielsen HH, Frøkiær J, Lundorf E, Pilegaard HK, et al.. Compromised cardiac function in exercising teenagers with pectus excavatum. Interact Cardiovasc Thorac Surg 2011;13:377–80.

[3] Kragten HA, Siebenga J, Höppener PF, Verburg R, Visker N. Symptomatic pectus excavatum in seniors (SPES): a cardiovascular problem?: A prospective cardiological study of 42 seniors with a symptomatic pectus excavatum. Neth Heart 2011;19:73–8.
[4] Tardy MM, Filaire M, Patoir A, Gautier-Pignonblanc P, Galvaing G, Kwiatkowski F, et al.. Exercise cardiac output limitation in pectus excavatum. J Am Coll Cardiol 2015;66:976–7.
[5] Saleh RS, Finn JP, Fenchel M, Moghadam AN, Krishnam M, Abrazado M, et al.. Cardiovascular magnetic resonance in patients with pectus excavatum compared with normal controls. J Cardiovasc Magn Res 2010;12:73.
[6] Oezcan S, Attenhofer Jost CH, Pfyffer M, Kellenberger C, Jenni R, Binggeli C, et al.. Pectus excavatum: echocardiography and cardiac MRI reveal frequent pericardial effusion and right-sided heart anomalies. Eur Heart J Cardiovasc Imaging 2012;13:673–9.
[7] Gürkan U, Aydemir B, Aksoy S, Akgöz H, Tosu AR, Öz D, et al.. Echocardiographic assessment of right ventricular function before and after surgery in patients with pectus excavatum and right ventricular compression. Thorac Cardiovasc Surg 2014;62:231–5.
[8] Bafus BT, Chiravuri D, van der Velde ME, Chu BI, Hirshl R, Farley FA. Severe hypotension associated with the prone position in a child with scoliosis and pectus excavatum undergoing posterior spinal fusion. J Spinal Disord Tech 2008;21:451–4.
[9] Awad SF, Barbosa-Barros R, Belem Lde S, Cavalcante CP, Riera AR, Garcia-Niebla J, et al.. Brugada phenocopy in a patient with pectus excavatum: systematic review of the ECG manifestations associated with pectus excavatum. Ann Noninvasive Electrocardiol 2013;18:415–20.
[10] Borrhomée S, Lenoir M, Gouton M, Fadel E. Syncope caused by right ventricular obstruction by pectus excavatum. J Thorac Cardiovasc Surg 2016;151:e67–9.
[11] Baranchuk A, Enriquez A, Garcia-Niebla J, Bayés-Genis A, Villuendas R, Bayés de Luna A. Differential diagnosis of rSŕ pattern in leads V1–V2. Comprehensive review and proposed algorithm. Ann Noninvasive Electrocardiol 2015;20:7–17.
[12] Dendramis G. Brugada syndrome and Brugada phenocopy. The importance of a differential diagnosis. Int J Cardiol 2016;210:25–7.
[13] Guntheroth W, Spiers PS. Cardiac function before and after surgery for pectus excavatum. Am J Cardiol 2007;99:1762–4.
[14] Töpper A, Polleichtner S, Zagrosek A, Prothmann M, Traber J, Schwenke C, et al.. Impact of surgical correction of pectus excavatum on cardiac function: insights on the right ventricle. A cardiovascular magnetic resonance study. Interact Cardiovasc Thorac Surg 2016;22:38–46.