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

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A 23-year-old man (height 1.83 cm, weight 68 kg, body surface area 1.90 m²) 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
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
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.

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