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GRAND ROUND
Other

Case report: ventricular fibrillation and cardiac arrest provoked by forward bending in adolescent with severe pectus excavatum

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Background Life-threatening arrhythmias have been reported in patients with severe pectus excavatum in absence of other cardiac abnormalities. Literature is scarce regarding diagnosis, cause and management of this problem, particularly regarding the question as to whether the placement of an implantable cardioverter-defibrillator (ICD) is necessary.

Case summary A 19-year-old male patient with severe pectus excavatum was scheduled for elective surgical correction. During forward bending for epidural catheter placement, syncope and ventricular fibrillation (VF) occurred resulting in cardiac arrest. After successful cardiopulmonary resuscitation, extensive analysis was performed and showed no cause for VF other than cardiac compression (particularly of the left atrium, right atrium, and ventricle to a lesser degree) due to severe pectus excavatum. Postponed correction by modified Ravitch was performed without ICD placement, with an uneventful post-operative recovery. Eighteen months after surgery, the patient remains well. Upon specific request, he did remember dizzy spells when tying shoelaces. He always considered this unremarkable.

Discussion In severe pectus excavatum with cardiac compression, forward bending can decrease central venous return and cardiac output, causing hypotension, arrhythmia, and cardiac arrest. In absence of structural or electric abnormalities, cardiac compression by severe pectus excavatum was considered a reversible cause of VF and ICD placement unnecessary. Patients with cardiac compression due to severe pectus excavatum may report pre-existing postural symptoms upon specific request. When these postural symptoms are present, extreme and prolonged forward bending postures should be avoided.

Keywords Case report • Pectus excavatum • Cardiac arrest • Ventricular fibrillation • Cardiac compression • Adolescent

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Learning points

- Cardiac compression resulting from severe pectus excavatum is usually well tolerated but may cause posture dependent symptoms, such as lightheadedness when tying shoelaces. Patients may not spontaneously mention these symptoms. If they are present, patients should avoid maintaining the particular position.
- In extreme postures, such as for epidural placement, cardiac compression may become so severe that it decreases cardiac preload and output, resulting in hypotension, arrhythmias, and potentially cardiac arrest.
- In this patient, ventricular fibrillation was considered secondary to cardiac compression due to severe pectus excavatum. The patient was managed with pectus correcting surgery without implantable cardioverter-defibrillator placement.

Primary specialities involved other than cardiology

General thoracic surgery, anaesthesiology, cardiothoracic surgery.

Introduction

Pectus excavatum is the most common congenital deformity of the chest wall. Symptoms include cardiopulmonary symptoms such as palpitations, fatigue, and exercise intolerance. Moreover, patients often suffer from poor body image and lower quality of life.[1] The anatomic severity of the defect is often expressed using the Haller index, which was first described in 1987.[2] This index is often based on chest computed tomography (CT) scan and is calculated by dividing the transverse diameter (the widest horizontal distance inside of the ribcage) by the anteroposterior diameter (the shortest distance between the vertebrae and the sternum). It is frequently used in publications, although it does have some disadvantages. These include absence of clear normal values, although 3.25 is most commonly used as a cut-off point with values above 3.25 indicating pectus excavatum. Determining a cut-off point to select patients for surgery is difficult, as a result of variability of the Haller index depending on age, gender, thoracic shape, and phase of respiration at the moment of scanning.[3] The most frequently used surgical correction techniques are the modified Nuss and the Ravitch procedure.

In patients with pectus excavatum, the electrocardiogram (ECG) may be normal. There have been descriptions of ECG changes associated with a shifted position of the heart in patients with pectus excavatum. These include S1S3 or S1Q3 pattern, negative P-wave in V1, and incomplete right bundle branch block (RBBB) rSr'-pattern.[4] Life-threatening arrhythmia such as ventricular fibrillation (VF) in patients with severe pectus excavatum have been described but are extremely rare. With respect to VF resulting in cardiac arrest in the presence of severe pectus excavatum, literature is scarce regarding aetiology and management, particularly whether an implantable cardioverter-defibrillator (ICD) should be placed.[5,6]

This case report describes a 19-year-old male with a severe pectus excavatum, who experienced syncope, VF, and cardiac arrest while bending over in sitting position for epidural catheter placement. This case teaches us first how a careful history could have revealed pre-existing postural symptoms. Secondly, it teaches that in absence of structural or electric disease combined with the well-documented sequence of events, cardiac compression due to severe pectus excavatum—and subsequent decreased preload and cardiac output, hypotension, and ischaemia—was considered the cause of VF. Finally, it describes the management of this episode of VF with pectus correcting surgery without ICD placement, as the cause was considered reversible.

Timeline

Day 1	Admission for elective modified Ravitch procedure. Epidural placement on OR holding, eventually leading to ventricular fibrillation and cardiac arrest, and performance of cardiopulmonary resuscitation. After return of spontaneous circulation transfer to intensive care unit (ICU). Coronary angiogram shows normal coronaries.
Day 2	Extubation, haemodynamically normal without support.
Day 3	Transfer from ICU to cardiac care unit.
Day 3–38	Continuous rhythm monitoring on cardiology ward (no events occurred). Several investigations were performed including chest computed tomography, cardiac magnetic resonance imaging, and Ajmaline challenge in university hospital.
Day 38	Transfer to university hospital (with full facilities for cardiothoracic surgery including extracorporeal circulation and dedicated anaesthesiologists) for surgical correction.
Day 41	Modified Ravitch procedure by general thoracic surgeon and cardiothoracic surgeon, with extracorporeal circulation and full cardioanaesthesiologic team at hand.
Day 45	Discharge home.
Day 450	18 months after surgery, patient remains well, no cardiac events, postural symptoms have decreased.

Case presentation

A 19-year-old male was scheduled for elective modified Ravitch procedure for severe pectus excavatum (*Figure 1*) in a large teaching hospital that functions as a tertiary referral centre for chest wall and pectus surgery. His medical history reported right-sided video-assisted thoracoscopic surgery bullectomy and pleurectomy for spontaneous pneumothorax. The preoperative ECG (*Figure 2A*) showed a sinus rhythm 76 b.p.m., with subtle RBBB with a small r' in the anterior chest leads, which is consistent with earlier descriptions of ECG appearances in pectus excavatum.[4] Epidural catheter

placement for perioperative analgesia is a standard procedure in our hospital. While in sitting position and bending forward, the patient became hypotensive and announced feeling lightheaded and nauseous (heart rate 80 b.p.m., mean arterial pressure 85 mmHg). This was initially considered as a vasovagal response. Ephedrine (2× 5 mg) was administered while continuing epidural placement. Blood pressure recovered after administration of ephedrine, with a sinus rhythm of 100/min. Nevertheless, the patient's symptoms were progressive, and he collapsed. Epidural placement was discontinued, and phenylephrine (0.2 mg), atropine (0.5 mg), dexamethasone (4 mg), and ondansetrone (4 mg) were administered intravenously. Despite this, the patient deteriorated, developed progressive tachycardia, haemodynamic and respiratory failure, and eventually went into VF. Advanced life support was initiated. After eight cycles of cardiopulmonary resuscitation (including defibrillation) spontaneous circulation occurred. The ECG directly after return of spontaneous circulation demonstrated a sinus tachycardia with broadening of the QRS-complex and ST-elevations most pronounced in the inferior leads that resolved within the next minutes (*Figure 2B and C*). Cardiac ultrasound in the acute setting showed global hypokinesia without local wall motion abnormalities and no pericardial effusion or other abnormalities. Ionized electrolytes in the arterial blood gas were normal. A coronary angiography showed normal coronaries. There were no signs of anaphylaxis. The patient was admitted to the intensive care unit. He recovered quickly without any neurological sequelae.

An additional history regarding postural symptoms and syncope was taken. The patient reported experiencing lightheadedness and dyspnoea when bending over, for example when tying shoelaces. He always considered this unremarkable.

In addition to the treating thoracic surgeon, cardiologist and anaesthesiologist, a cardiologist specialized in electrophysiology and a cardiothoracic surgeon from a university hospital were involved. Several investigations were performed to evaluate conduction and structural cardiac causes of the arrhythmia. Cine cardiac magnetic resonance imaging (MRI) (*Figure 3*, *Video 1*) and chest CT (*Figure 4*) showed a severe pectus excavatum (Haller index 4.72: 274/58) and an extensive compression of the heart (in particular of the left atrium, and to a lesser degree of the right atrium and ventricle) between the depressed sternum and the thoracic spine. Cardiac MRI further showed optically normal right and left ventricle contractility and a normal right outflow tract. There were no signs of arrhythmogenic right ventricular cardiomyopathy and no delayed enhancement. Ajmaline challenge was negative for Brugada syndrome.

We concluded that in this case of severe pectus excavatum, compression of both atria and the right ventricle between sternum and spine was usually well tolerated. However, bending forward increased cardiac compression and caused cardiac inflow obstruction resulting in symptomatic hypotension. This likely occurred during epidural placement as well. It was potentially exacerbated by relative hypovolaemia due to preoperative fasting and vasovagal reaction to epidural placement. These circumstances combined decreased cardiac preload and output to such an extent that it caused prolonged hypotension, resulting in haemodynamic instability and respiratory insufficiency which most likely caused myocardial ischaemia and VF.

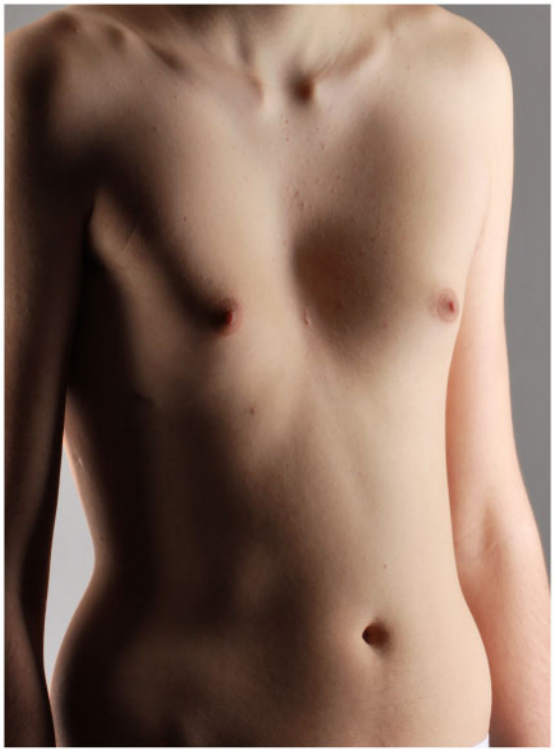


Figure 1 Severe pectus excavatum in 19-year-old male (preoperative status).

Thus, in the absence of other abnormalities, cardiac compression resulting from severe pectus excavatum was considered as the underlying cause of the arrhythmia. As VF was considered secondary to pectus excavatum, the cause would be reversible by pectus correcting surgery. Therefore, an ICD was not placed and a modified Ravitch procedure was scheduled. The patient was under continuous rhythm monitoring in the weeks between event and surgery, no further cardiac events or symptoms occurred. As a precaution, surgery was performed in a university hospital with a full cardiothoracic and cardioanaesthesiologic team present and extracorporeal circulation at hand. Both the procedure and perioperative period were uneventful. The patient made a full recovery. Follow-up consisted of 6 monthly outpatient checkups by his cardiologist (including ergometry and ECG, *Figure 2E*) and thoracic surgeon. Eighteen months after surgery, he remains well. His postural symptoms have decreased.

Discussion

In conclusion, in this 19-year-old male patient, cardiac compression as a result of a deep pectus excavatum caused posture dependent symptoms. They were usually well tolerated and the patient considered them unremarkable, although a careful history with specific questions regarding postural symptoms would have revealed them. In this case, pectus-induced cardiac compression was most likely exacerbated during epidural catheter placement by extreme posture

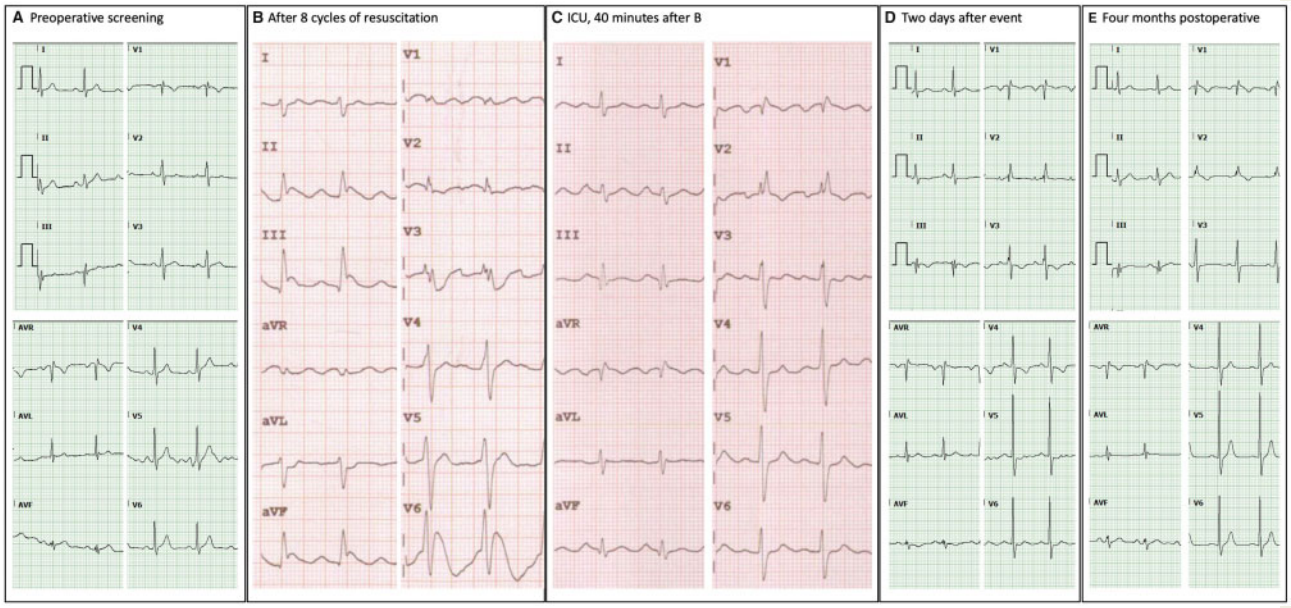


Figure 2 Electrocardiogram. (*A*) At baseline, preoperative screening. (*B*) Return of spontaneous circulation after eight cycles of advanced life support. (*C*) At the intensive care unit, 40 min after panel *B*. (*D*) Two days after the event. (*E*) Four months after modified Ravitch procedure.

(bending forward in sitting position). Potentially aggravated by hypovolaemia due to preoperative fasting and a vasovagal response during epidural placement, this resulted in decreased preload and cardiac output, prolonged hypotension, and myocardial ischaemia, which triggered VF and lead to cardiac arrest. Given the well-documented sequence of events and after exclusion of underlying structural and electric cardiac disease, pectus correcting surgery was considered to relieve cardiac compression and thereby reverse the cause of VF. Hence, the patient was managed with a modified Ravitch procedure without ICD placement or other (invasive) monitoring. One year after surgery, the patient remains well.

This case has several strengths and limitations. The fact that VF and circulatory arrest were witnessed is an advantage in determining the circumstances leading to the event. Furthermore, extensive investigations were performed to exclude structural and electrical disease. The relatively short follow-up period of 1.5 years can be considered a limitation. A long-term follow-up study regarding survivors of out-of-hospital cardiac arrest due to idiopathic VF showed a median time to first arrhythmic recurrence of 29 months (25–75th percentile: 12–70 months) and an incidence of 21% in 5 years.[7]

Reports on the management of comparable cases of arrhythmia or cardiac arrest in the context of pectus excavatum are scarce. A search revealed two case reports on sudden cardiac arrest based on VF in patients with severe pectus excavatum.[5,6] Both authors describe management with subcutaneous ICD placement, although Rachwan *et al.*[6] discuss that this is not supported by evidence. Less severe symptoms (such as lightheadedness when bending over, such as the current patient experienced before the event) are also described. They include recurrent syncope resulting of right ventricular compression by pectus excavatum in a patient with previous heart surgery.[8] The authors suggest that previous surgery, which caused adhesions and scar tissue, could aggravate cardiac compression by preventing shifting of the heart to the pleural cavity. This is interesting in connection to the current case, as this patient also had a history of thoracoscopy. A second case report describes positional orthostasis and hypotension due to right ventricular obstruction caused by pectus excavatum in seated but not in supine position,[9] and another reports hypotension resulting from right heart compression after placing a patient with pectus excavatum in prone position for scoliosis surgery.[10]

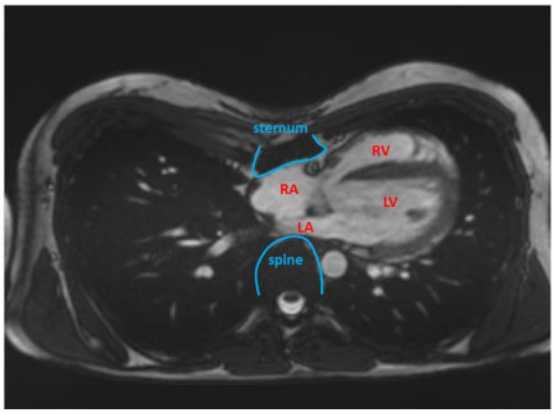
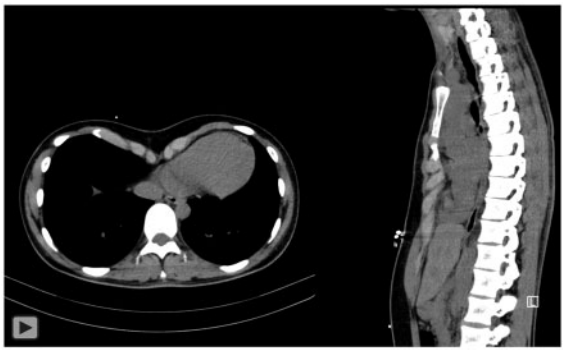


Figure 3 Preoperative cardiac cine magnetic resonance imaging (screenshot) showing compression of left and right atrium and right ventricle caused by pectus excavatum. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



Video 1 Preoperative computed tomography scan with Haller index of 4.72, preoperative cardiac cine magnetic resonance imaging showing compression of left and right atrium and right ventricle caused by pectus excavatum.

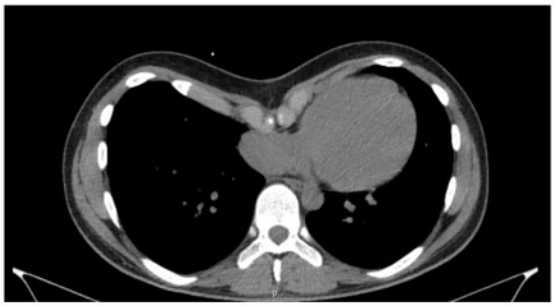


Figure 4 Preoperative chest computed tomography showing severe pectus excavatum (Haller index 4.72: 274/58).

These previous cases illustrate the haemodynamic effects of a depressed sternum in patients with pectus excavatum and discuss that there is little evidence on how to manage these patients. The current case firstly adds insight regarding the relationship between pectus excavatum, cardiac compression, and arrhythmia. Secondly, it illustrates that patients may have pre-existing postural symptoms. Finally, it describes successful management with pectus correcting surgery without ICD placement.

Routine assessment of postural symptoms is, at the moment, not part of standard clinical practice. There are, however, several options that could improve preoperative assessment. Relatively simple investigations that clinicians could implement immediately include taking a specific history regarding postural symptoms (such as dizziness when tying shoelaces), as well as blood pressure and heart rate and rhythm in different positions. Normal values, cut-off points, and implications of these measurements have not been established and could be the subject of future research. Additional options for imaging include stress echocardiography or postural echocardiography, and cardiac MRI. Stress echocardiography may reveal important circulatory changes which are not present at rest.[11] Postural echocardiography may be of value as it has been described that compromise of the right ventricular outflow tract in pectus excavatum is exacerbated in a sitting upright,[9] or sitting and forward bending position,[12] which was likely relevant in our patient as well. Cardiac MRI may be performed during end expiration, as the degree of cardiac compression worsens with expiration.[13] These imaging modalities may increase our understanding of the haemodynamic consequences of pectus excavatum in a specific patient. Cut-off points and normal values will have to be established, but they may provide the treating physicians with a more comprehensive assessment of cardiac function in a specific patient, which could be used in preoperative decision-making and aid the anaesthesiologist in their perioperative management.

In conclusion, patients with cardiac compression due to severe pectus excavatum may report pre-existing postural symptoms. Cardiac compression in pectus excavatum may be exacerbated by extreme posture and can be so severe that it causes syncope or even arrhythmia and cardiac arrest. We consider the cause of VF reversible by pectus correcting surgery. When postural symptoms do occur, patients should avoid maintaining this position.

Lead author biography

Erik R. de Loos is a general thoracic and trauma surgeon with a specific interest in the treatment of congenital and acquired chest wall disorders. Since 2011, he works as a consultant at Zuyderland Medical Center (Heerlen, the Netherlands), a tertiary referral centre for minimally invasive thoracic surgery and chest wall pathology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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