Taylor & Francis
Taylor & Francis Group

Acta Cardiologica

ISSN: (Print) (Online) Journal homepage: https://www.tandfonline.com/loi/tacd20

Freedom in the chest

Sébastien Piron, Julien Tridetti, Stella Marchetta & Patrizio Lancellotti

To cite this article: Sébastien Piron, Julien Tridetti, Stella Marchetta & Patrizio Lancellotti (2021): Freedom in the chest, Acta Cardiologica, DOI: 10.1080/00015385.2021.1991666

To link to this article: https://doi.org/10.1080/00015385.2021.1991666

Published online: 21 Oct 2021.

ACTA CARDIOLOGICA
https://doi.org/10.1080/00015385.2021.1991666

ORIGINAL SCIENTIFIC PAPER

Freedom in the chest

Sébastien Piron, Julien Tridetti, Stella Marchetta* and Patrizio Lancellotti

Department of Cardiology, University of Liège Hospital, GIGA Cardiovascular Sciences, Liege, Belgium

ABSTRACT

Pericardial agenesis is a rarely seen congenital defect characterised by the partial or, more rarely complete, absence of the pericardium. Most often asymptomatic, it is usually incidentally discovered following the demonstration of heart's laevorotation on imaging, in the operating room or at autopsy. In this article, we report the case of an 80-year-old patient with asymptomatic complete pericardial agenesis fortuitous discovered. Pericardial agenesis observations are extremely uncommon reported in the literature, which substantiate its original epidemiological character. In addition, this observation brings some clinical, electrical as well as iconographic elements to better understand the pathology and raises clinical suspicions. Finally, this case report confirms the exceptionally symptomatic nature of the pathology, illustrating the irrelevance of treatment or specific follow-up.

ARTICLE HISTORY
Received 27 June 2021
Revised 31 August 2021
Accepted 24 September 2021

KEYWORDS
Pericardial agenesis; heart's laevorotation; right axial deviation; medical imaging

Introduction

The first observation of a congenital absence of the pericardium (CAP) was made by the anatomist Colombus R in 1559 [1]. Epidemiologically, pericardial agenesis has an unknown prevalence, due to lack of scientific literature on the subject, given the rarity of the pathology. Since this first observation, more and more cases have been reported in the litterature. In this article, we report the case of an 80-year-old patient with a total pericardial agenesis. This observation brings some clinical, electrical as well as iconographic elements to better understand the pathophysiology that may raises clinical suspicions of CAP.

Case report

We report the case of an 80-year-old female patient hospitalised for assessing lipothymic-like discomfort with brief loss of consciousness associated with Grade IV dyspnoea (NYHA). Neither chest pain nor other associated cardiopulmonary symptom was percieved.

From her medical history, we will noted a history of pulmonary embolism following deep vein thrombosis in lower left limb that happened 2 months ago and was treated by new oral anticoagulants. Regarding cardiovascular risk factors, the patient suffered from hypertension stabilised by a beta-blocker, an angiotensin II receptor blocker and thiazide diuretic.

On clinical examination, a posterior axillary line deviated apical impulse, deafened heart tones and left basal hypoventilation at pulmonary auscultation were noticed. Electrocardiogram (ECG) revealed bradycardia, a right axis deviation (155° ventricular activation axis, aVR positive and aVL negative axis) as well as a strongly displaced QRS direction transition zone (incomplete right branch block with poor R-waves progression in precordial leads) (Figure 1).

Chest x-ray showed a projection of left middle arch as well as anomalies in position of aortic arch on profile incidences (Figure 2).

To investigate the cause of the dyspnoea, a computerised tomography (CT) scan was performed, showing a recanalization process of the left inferior pulmonary lobar artery embolism. Fortuitously, a laevorotation (left rotation) of the heart within the mediastinum without twist of the large vessels as well as an absence of parietal pericardial sheet was observed (Figure 3).

Transthoracic ultrasound (TTUS) did not show any right ventricular dysfunction, subject to technical difficulty of performing examination given atypical position of the heart, whose apex points towards dorsal column (Figure 4).

CONTACT Patrizio Lancellotti plancellotti@chu.ulg.ac.be Department of Cardiology, University Hospital Liege Domaine universitaire du SartTilman, Liège, B35, B 4000, Belgium.
*Division of Cardiology, CHC MontLégia, Liège, Belgium.

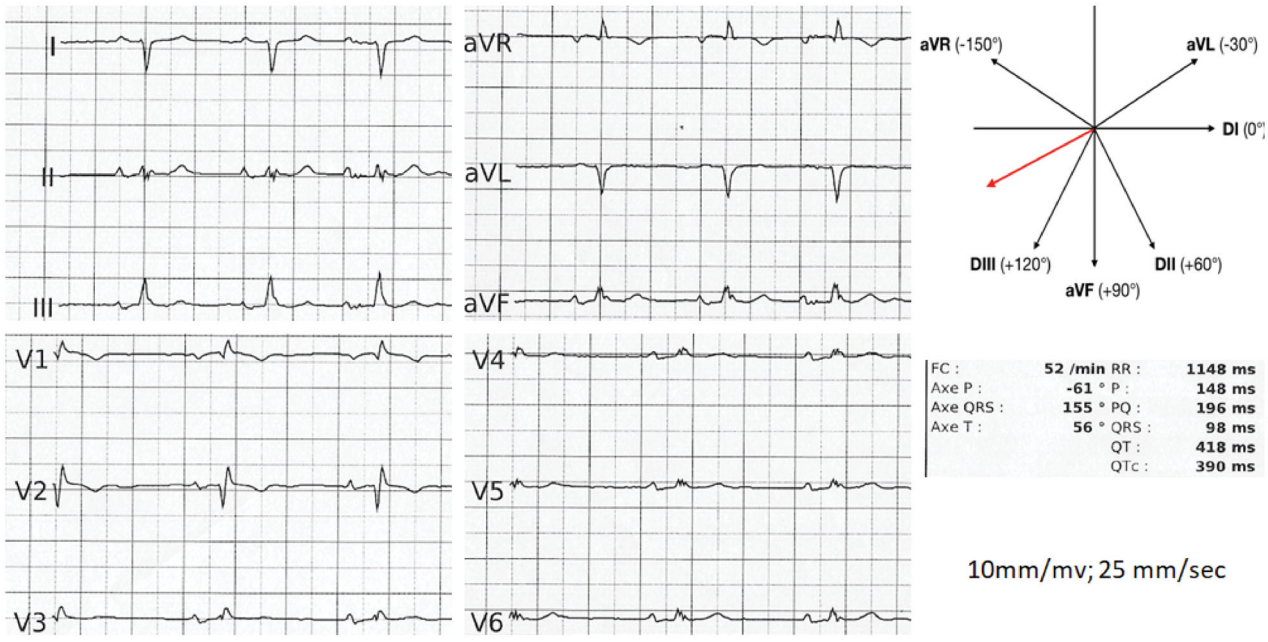

Figure 1. ECG showing a right axis deviation (155° ventricular activation axis, aVR positive and aVL negative axis) as well as a strongly displaced QRS direction transition zone (incomplete right branch block with poor R-waves progression in precordial leads.

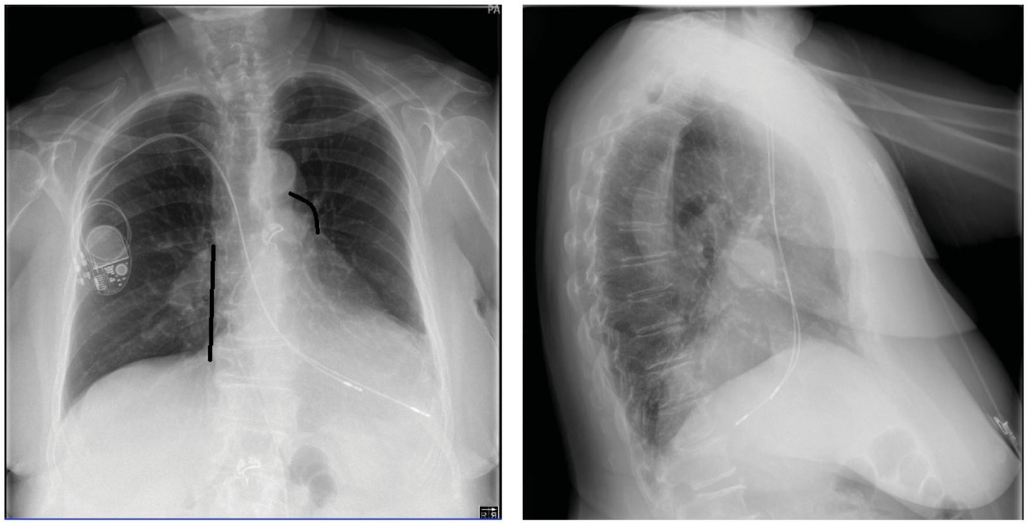

Figure 2. Chest X-ray showing a projection of left middle arch due to a prominent pulmonary artery and anomalies in position of the aortic arch on profile incidences.

Telemetric monitoring during hospitalisation displays sinus bradycardia associated with supraventricular tachycardia bursts and paroxysmal atrial fibrillation. Treatment proposed was implantation of a double chamber pacemaker, which one improved her symptomatology. Pacemaker's probes implantation was complicated given the lack of usual point of reference for radioscopic control.

Discussion

The pathology results from early atrophy of common cardinal vein (Cuvier's canal) between 3-4rd and 6th embryonic week [2], where anterior and posterior cardinal veins join from both side, and which are subject to major changes during development of the circulatory system. This atrophy would alter vascularisation of pleuro-pericardial membrane, explaining its developmental arrest in the partial form up to agenesis in the total form.

Pericardial agenesis is classified into six forms according to defect importance and localisation. Thus, we distinguish a total form (the rarest, estimated at 9% of pericardial agenesis cases), 2 left forms (complete or partial), 2 right forms (complete or partial) and a

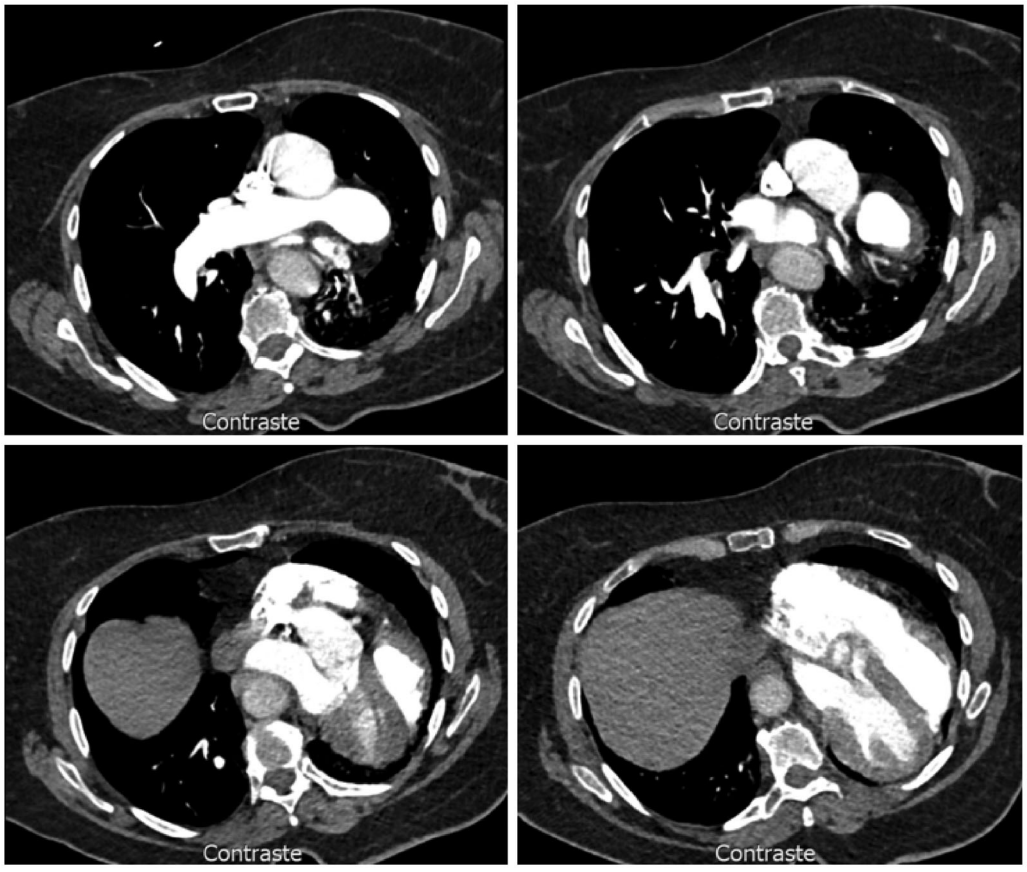

Figure 3. CT scan showing a laevorotation of the heart within the mediastinum without twist of the large vessels and an absence of parietal pericardial sheet.

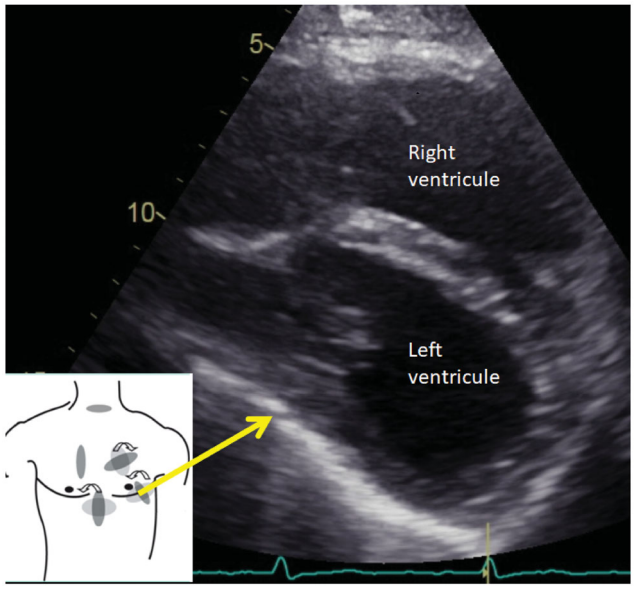

Figure 4. TTUS showing a atypical position of the heart, whose apex points towards the dorsal column.

diaphragmatic form (17% of cases) [3]. More often, the left form is observed (70–80% of pericardial agenesis) [3]. Different entities are to be distinguished because of differences in clinical presentation and impact.

Due to the aetiology, associated congenital anomalies have been described. Among these, the most common are heart disease (30% of cases), including inter-auricular communication, persistent ductus arteriosus and tetralogy of Fallot [3] or bronchogenic cysts [4]. Diaphragmatic partial form is frequently associated with diaphragmatic hernia [5]. These abnormalities can sometimes be the object of an antenatal diagnosis and require a treatment.

Total pericardial agenesis is less common but also better tolerated than other forms [3]. This is explained by the fact that there is no risk of herniation (responsible for disorders of myocardial kinetics, arrhythmia or sudden death) or of extrinsic coronary compression at the level of the fibrous hernia collar, resulting in dynamic coronary stenosis, increased during diastole, visible in coronarography [6].

Pericardium being totally absent, positional anomalies of the heart can however be more important. So, some cases of twist of major blood vessels due to cardiac malposition have been described. Tricuspid insufficiencies can also occur [7], explained by an abnormal ventricular geometry favoured by heart's laevorotation, causing anterior pillar's chordae distension and ultimately

weakening subvalvular apparatus [8,9]. This disability leads to more tricuspid insufficiencies, especially traumatic ones although spontaneous tricuspid insufficiencies have also been described [8]. Apart from theses rares complications, total ageneses are asymptomatic and have, as only consequence, possible technical difficulties during cardiac auscultation and certain complementary examinations. One case of delayed diagnosis of rupture of ascending aorta has been described, which has been evidenced by a severe progressive left haemothorax and the absence of haemopericardium due to absence of pericardium [10].

Regarding the diagnosis, the main sign to look for is a laevorotation of the heart. This is clinically reflected by a shift of apical impulse and an attenuation of cardiac tones [11].

Laevorotation can induce a right branch block, which can lead to asynchronism, responsible for a B2 splitting at auscultation [12].

ECG can reveal a right axis deviation of the QRS, a right branch block, and a R- transition zone shifted to the left in precordial leads due to horizontal rotation [3]. The case discussed here presents all of these signs (Figure 1).

Concerning imaging, chest X-ray may show a marked levoposition of cardiac silhouette with a loss of right heart border and a projection of left middle arch due to a prominent pulmonary artery [10], as well as anomalies in aortic arch position on the profile incidences, as illustrated in our clinical case (Figure 2). Finally, abnormal position and the absence of pericardium can be pointed out by ultrasound, CT scan or nuclear magnetic resonance. Ultrasound can also reveal cardiac hypermobility [3] and a paradoxical movement of the septum.

Given its asymptomatic nature, total pericardial agenesis does not, most often, require treatment or follow-up, as evidenced by our 80-year-old patient, asymptomatic until then. Thus, surgical treatment such as pericardioplasty should not be proposed in cases of total pericardial agenesis [13]. This treatment is also not indicated in case of severe agenesis such as the two complete left and right forms due to the low risk of hernia [3]. However, pericardioplasty associated with valve replacement in the particular case of tricuspid insufficiencies allowed a good final result in the few described cases [8,9,14].

Conclusion

Pericardial agenesis is a unfrequent condition, most often fortuitously diagnosed. Total agenesis is the rarest form and results in a laevorotation of the heart. Laevorotation and the absence of pericardium are both noticeable by imaging. Apart from a twist of the major blood vessels, it is asymptomatic and does not require treatment or follow-up.

Disclosure statement

No potential conflict of interest was reported by the author(s).

References

[1] Columbus MR. De re anatomica. Venise: N. Beurlaquae; 1559.
[2] Southworth H, Stevenson CS. Congenital defects of the pericardium. Arch Intern Med. 1938;61(2):223–240. [Internet].
[3] Shah AB, Kronzon I. Congenital defects of the pericardium: a review. Eur Heart J Cardiovasc Imaging. 2015; 16(8):821–827.
[4] Mukerjee S. Congenital partial left pericardial defect with a bronchogenic cyst. Thorax. 1964;19:176–179.
[5] Gonzales M, Dekoster G, Perrin L, et al. Congenital absence of the left pericardium and diaphragmatic defect in sibs. Eur J Med Genet. 2010;53(3):133–135.
[6] Nguyen DQ, Wilson RF, Bolman RM, et al. Congenital pericardial defect and concomitant coronary artery disease. Ann Thorac Surg. 2001;72(4):1371–1373.
[7] Higashikawa M, Nishio M, Kanamori T, et al. Traumatic tricuspid regurgitation associated with congenital partial pericardial defect. Jpn Circ J. 1997;61(4):358–360.
[8] Goetz WA, Liebold A, Vogt F, et al. Tricuspid valve repair in a case with congenital absence of left thoracic pericardium. Eur J Cardiothorac Surg. 2004;26(4): 848–849.
[9] Harskamp RE, McNeil JD, van Ginkel MW, et al. Postoperative internal thoracic artery spasm after coronary artery bypass grafting. Ann Thorac Surg. 2008; 85(2):647–649.
[10] Nakajima M, Tsuchiya K, Naito Y, et al. Partial pericardial defect associated with ruptured aortic dissection of the ascending aorta: a rare feature presenting severe left hemothorax without cardiac tamponade. Ann Thorac Surg. 2004;77(3):1066–1068.
[11] Gatzoulis MA, Munk M-D, Merchant N, et al. Isolated congenital absence of the pericardium: clinical presentation, diagnosis, and management. Ann Thorac Surg. 2000;69(4):1209–1215.
[12] Bueno Palomino A, Palomar Estrada A, Crespín Crespín M, et al. Congenital complete absence of pericardium in a young woman with non-specific symptoms. Rev Port Cardiol. 2014;33(4):249.e1–249.e5.
[13] Van Son JA, Danielson GK, Schaff HV, et al. Congenital partial and complete absence of the pericardium. Mayo Clin Proc. 1993;68(8):743–747.
[14] Fracasso A, Pothen P, Gallucci V. Tricuspid regurgitation caused by blunt chest trauma in association with pericardial agenesis: surgical correction after eight years. Thorax. 1982;37(1):75–76.