Partial absence of the pericardium: Only an incidental finding?☆

Ausencia parcial de pericardio: ¿únicamente un hallazgo incidental?

Dear Editor:

Absence of the pericardium (AoP) is a rare anomaly usually detected by chance. Its incidence is less than 1 case per 10 000 inhabitants.[1] It is classified based on its extension (complete or partial) and position (left, right or bilateral), and complete left-sided defects are the most frequent form (70%).[2–4] In 30%–50% of cases, AoP is associated with congenital heart defects such as tetralogy of Fallot, atrial septal defect, patent ductus arteriosus, mitral valve defects or sinus venosus defects with partial anomalous pulmonary venous drainage[3] as well as pulmonary malformations, Marfan syndrome, VATER syndrome and Palliester–Killian syndrome.[2] Although most cases are asymptomatic, patients with partial absence of the pericardium (PAoP) may experience chest pain, palpitations, syncope or even sudden death secondary to herniation of cardiac structures through the pericardial defect.

We present 2 cases of incidental finding of AoP. The first corresponded to a patient referred at the age of 13 years due to abnormal position of the heart in the chest radiography (CXR). He was symptomatic and did not have any relevant personal or family history. The physical examination was normal. The electrocardiogram (ECG) (Fig. 1A) showed sinus bradycardia with right axis deviation (+112 °), incomplete right bundle branch block (IRBBB), slow R wave progression, nonspecific intraventricular conduction delay (QRS duration of 96 ms) and abnormal repolarization (inverted T waves in V1–V4, transitioning in V5 and positive in V6). The CXR showed lung interposition between the aorta and pulmonary artery (PA), leftward shift of the heart with no visible right heart border and lung interposition between the left diaphragm and the base of the left heart (Fig. 2A–B). After performing an echocardiogram, PAoP was suspected on account of the posterior and leftward shift of the apex, paradoxical septal motion and apparent dilatation of the right ventricle due to leftward cardiac displacement (Fig. 1C; Supplemental material Appendix B, videos 1 and 2). Computed tomography (CT) and magnetic resonance imaging (MRI) findings confirmed the diagnosis (Fig. 2C–E). His exercise test was normal. As the patient was asymptomatic, a conservative approach was chosen. He has remained asmptomatic with no complications after 7 years follow-up.

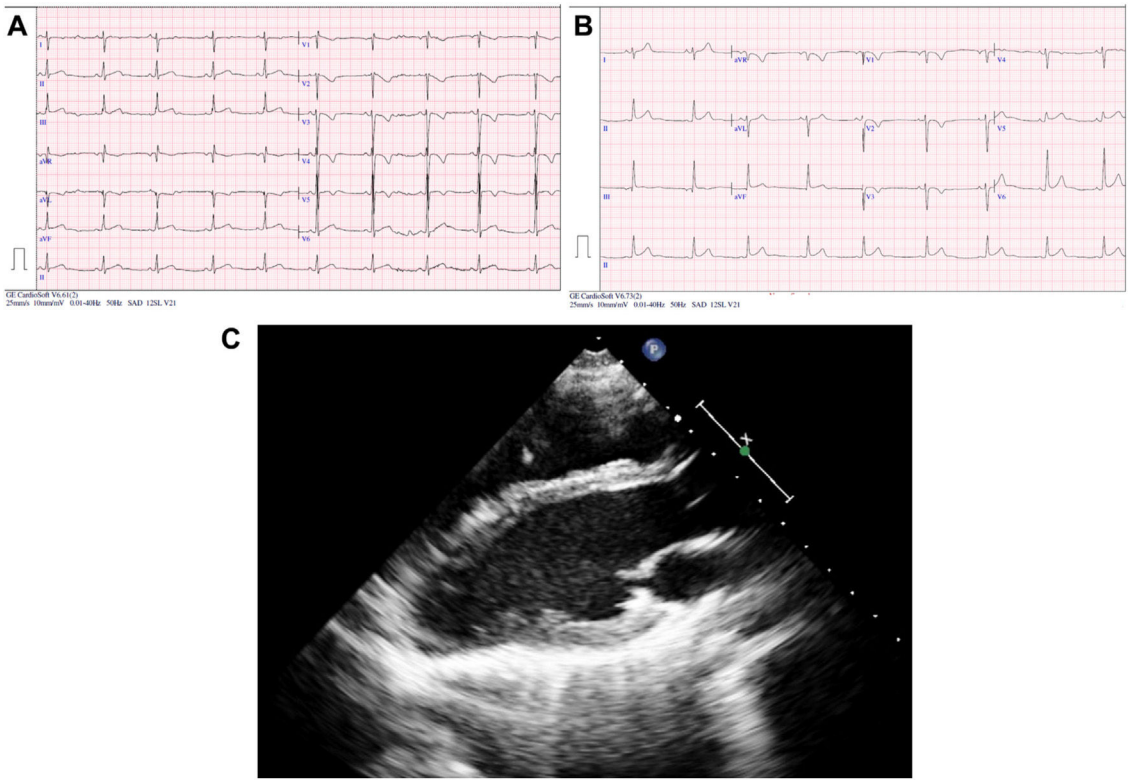

Figure 1 (A) Resting 12-lead ECG, patient 1. (B) Resting 12-lead ECG, patient 2. (C) Echocardiogram, patient 1. Long axis view showing posterior and rightward deviation of the apex of the left ventricle.

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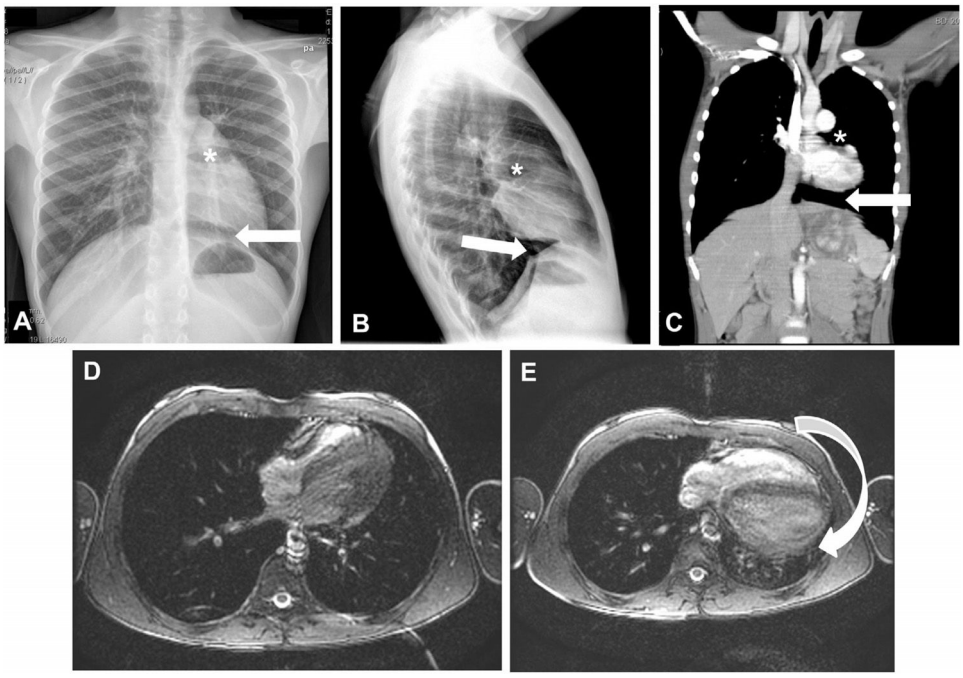

Figure 2 (A–C) Chest radiography, posteroanterior (A) and lateral (B); cardiac CT (C); leftward shift of the heart with lung interposition between the aorta and the pulmonary artery (*) and interposition of lung parenchyma between the left diaphragm and the border of the base of the left heart (arrow). (D) Cardiac MRI with the patient in the right lateral decubitus position. (E) Cardiac MRI with patient in supine position showing posterior shift of the cardiac silhouette (arrow).

The second patient was referred at the age of 5 years due to deviation of the heart in the CXR. During surveillance, she was diagnosed with Behcet disease. She was asymptomatic from the cardiac point of view and she did not have any other relevant personal or family history. She had normal physical examination. The ECG (at the age of 15 years, Fig. 1B) revealed mild right axis deviation (+94°), IRBBB, poor R wave progression, nonspecific intraventricular conduction delay (QRS duration of 98 ms) and abnormal repolarization (inverted T waves in V1–V3, transitioning in V4 and positive in V5–V6). The CXR and echocardiogram (supplementary material Appendix B, videos 3 and 4) revealed the same features described in patient 1. Her diagnosis was confirmed by MRI. She had a normal exercise test. A conservative management was chosen in view of the absence of symptoms. She has remained asymptomatic with no evidence of complications after 13 years of follow-up.

Although AoP is a rare disease, it is important to keep a high level of suspicion, mainly because patients with PAoP, in addition to being symptomatic more frequently, are at risk of herniation and strangulation of the left ventricle through the pericardial defect and, therefore, of sudden death.[2,3,5]

CXR, ECG, echocardiogram and CT are useful for diagnosis of AoP. However, the gold standard is MRI, which in black blood fast spin echo and steady-state free precession sequences evinces cardiac indentation at the location of the defect, interposition of lung tissue between the aorta and the PA or between the diaphragm and the base of the heart and leftward and posterior displacement of the heart. A rotation angle greater than 60° between the anterior-posterior line passing through the vertebral body and left ventricle main axis in the end-diastolic phase of the axial cine stack offers a good sensitivity for detection of left ventricular AoP.[6]

Although there is no global consensus on the management of these patients, close follow-up and monitoring for complications is recommended in asymptomatic patients in whom AoP is an incidental finding and in the case of complete bilateral or left-sided AoP. Surgery is reserved for patients with PAoP or at risk of complications.[2,5]

In conclusion, we have presented 2 cases of incidental diagnosis of AoP, a disease that is difficult to diagnose due to its rarity and usually asymptomatic, whose early detection is vital due to its potential association with sudden death. Although MRI is the gold standard of diagnosis, the abovementioned imaging investigations can also be useful. Despite the absence of a global consensus, the approach to treatment is conservative, reserving surgery for patients with symptoms or with PAoP at risk of complications.

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Conflicts of interest

The authors have no conflicts of interest to declare.

Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.anpedi.2021.05.005.

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Unicentric experience in percutaneous stent treatment of aortic coarctation in children and teenagers☆

Experiencia unicéntrica en el tratamiento percutáneo con stent de la coartación de aorta en niños y adolescentes

To the editor:

The advantages of stenting for treatment of coarctation of the aorta compared to balloon angioplasty is that it avoids excessive dilation of the adjacent aorta and elastic recoil of the vessel, in addition to sealing potential acute dissections, thereby reducing the incidence of aneurysms, recurrence of coarctation and aortic rupture. Most hospitals consider it the procedure of choice for native coarctation and for recurrent coarctation in patients with weights greater than 25 to 30 kg, in whom the calibre of femoral arteries is usually adequate.[1] Below this weight, although there is not sufficient evidence to establish the optimal therapeutic approach, angioplasty or surgery are used in most cases.[2,3] We conducted a retrospective analysis of all the patients aged less than 18 years managed with percutaneous stent implantation between 1996 and 2020 and followed up for at least 1 year. The resulting consecutive sample included 25 patients with a mean age of 10.6 years and a mean weight of 33.9 kg, of who 16% had weights of less than 30 kg. Thirteen patients were treated for native coarctation and 12 for recurrence of coarctation treated with surgery. Three of the cases of recurrent coarctation had required angioplasty due to a previous recurrence before placement of the stent. The most frequent associated cardiac defects were bicuspid aortic valve and ventricular septal defect (VSD) (Table 1).

In 20 cases (80%) the coarctation was at the level of the aortic isthmus, in 2 (12%) at the level of the aortic arch (8%) at the level of the diaphragmatic aorta.

Table 1 Clinical and haemodynamic characteristics of the sample.

Clinical variables	
Mean age ± SD (years)	10.6 ± 4.6
Mean weight ± SD (kg)	33.9 ± 18.9
Male sex	18 (72%)
Associated heart defects (other than aortic valve defect)	5 (20%)
VSD	
Anomalous pulmonary venous return	1 (4%)
Bicuspid aortic valve	14 (56%)
Significant aortic valve disease	2 (8%)
Type of coarctation	
Native	13 (52%)
Recurrent	12 (48%)
Previous surgical treatment	
End-to-end anastomosis	8 (32%)
Subclavian flap	3 (12%)
Patch aortoplasty	1 (4%)
Previous angioplasty	3 (12%)
Haemodynamic variables	
Aortic arch diameter ± SD (mm)	13.2 ± 3.8
Diaphragmatic aorta diameter ± SD (mm)	15.12 ± 4.4
Coarctation diameter ± SD (mm)	
Previous procedure ± SD	5.1 ± 2.2
After stenting ± SD	14.6 ± 3.7
Transaortic gradient (invasive)	
Previous ± SD	33.6 ± 14.3
After stenting ± SD	2.8 ± 4.1
Stent diameter ± SD (mm)	15 ± 4.2
Stent length ± SD (mm)	38.8 ± 18.2
Need of more than 1 stent	2 (8%)
ePTFE-covered stent	6 (24%)

ePTFE: expanded polytetrafluoroethylene; SD, standard deviation; VSD, ventricular septal defect.

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