Research Article – Basic and Applied Anatomy

Congenital defects of pericardium: case reports and review of literature

Matteo Cuccuini[1,*], Francesca Lisi[1], Arturo Consoli[1], Sara Mancini[1], Valentina Bellino[1], Giorgio Galanti[2], Leonardo Capaccioli[1]

[1] Department of Radiology, Careggi University Hospital, Florence, Italy
[2] Sport Medicine Centre, Careggi University of Florence, Italy

Summary

Introduction: Pericardial defects are a rare condition, generally asymptomatic, due to failure in development of pericardial sac. They are difficult to detect, particularly the complete absence of the pericardium. At present magnetic resonance imaging (MRI) is considered the best performing exam and it could be the first choice in the diagnosis of agenesia of the pericardium.
Materials and Methods: A comprehensive review of dedicated books and PubMed literature was performed and three clinical cases have been analyzed.
Results: We report three cases , one with partial and two with total agenesia of the left pericardium, seen at our Institute and diagnosed at MRI as unexpected findings.
Discussion: The diagnosis of a congenital defect of the pericardium is challenging, even for expert radiologists, because this kind of deformity does not provide specific clinical nor conventional radiology findings. Nowadays, the most effective instrument for a certain diagnosis is MRI, interlocked with electrocardiography.

Key words

Pericardium; congenital defect; pericardial embryology; MRI; magnetic resonance imaging; heart.

Introduction

Congenital defects of the pericardium are an uncommon malformations of the pericardial sac with variable clinical features. Patients are usually asymptomatic and the diagnosis is often incidental during cardiac surgery, diagnostic exams made for other reasons or post-mortem investigations (Faridah and Julsrud, 2002).

When present, this condition is difficult to detect due to its infrequency and because it is not correlated with any specific finding on clinical examination. Magnetic Resonance Imaging (MRI) is considered actually the best performing imaging technique in the diagnosis of pericardial abnormalities and diseases (Abbas et al., 2005). We present a summary about pericardial anatomy and its embryological development, and a review of clinical features of pericardial defects followed by three case reports studied at our Institute.

*Corresponding author. E-mail: teo.cuccuini@gmail.com; Tel: +39 339 1454156; Fax: +39 055 431970.

Anatomy of the pericardium

The pericardium is a conical fibro-serous sac that envelops all four cardiac chambers and the root of the great vessels which arise from the heart. It consists of two layers intimately connected with each other: an outer sac, or fibrous pericardium, and an inner layer or serous pericardium (Gray et al., 2005).

The fibrous pericardium in situ has a truncated conic shape with apex directed upwards towards the sternal notch, surrounding the origin of the great vessels and extending in their adventitia; its basis is adherent to the central tendon and to the muscular fibres of the left side of the diaphragm. It is 12-14 cm high, with anteroposterior diameter ranging from 9-10 cm at the base and 6-7 cm in apex. Its basis extends transversely for 9-11 cm with an anteroposterior diameter of 5-6 cm (Ferrans et al., 1982; Ellis, 2005).

The front wall of pericardium is slightly convex transversely and is inclined from behind to front and from apex to base. It is divided from the anterior thoracic wall by the lungs and pleurae; only a small area, variable in extension and usually corresponding with the left part of the body of the sternum and the medial portion of the cartilages of the fourth and fifth left ribs, comes into direct contact with the chest wall. Its anterosuperior portion corresponds with the lower extremity of the thymus in children, while in adults the thymus is replaced by soft adipose tissue. Behind, the pericardium extends from the V to the IX-X vertebra, resting upon the tracheobronchial tree, the descending aorta, the esophagus, the inferior vena cava (on the right), and the posterior part of the mediastinal surface of each lung. Laterally, it is in contact with the mediastinal surface of the lungs through the interposition of the pleural coating (Ferrans et al., 1982; Ellis, 2005; Gray et al., 2005). The diaphragmatic attachment of the pericardium consists of loose fibrous tissue, except for a small area of the central tendon in which diaphragm and pericardium are completely fused. Short fibrous branches form the phreno pericardial ligament (anterior, right and left). Above and behind, the fibrous pericardium blends with the external coats of the great vessels and with the pretracheal layer of the deep cervical fascia, sending short fibrous branches up to vertebral bodies (vertebro-pericardial ligament). It is also attached to the posterior surface of the sternum by the superior and inferior sterno-pericardial ligaments; the upper one reaching the manubrium, and the lower one the xiphoid process. These connections provide a safe anchor within the thoracic cavity (Ellis, 2005; Gray et al., 2005).

The serous pericardium is a delicate membrane, of coelomic origin, which lies within the fibrous sac; it therefore consists of a parietal portion covering the inner surface of the fibrous pericardium and a visceral portion, or epicardium, covering the heart and the root of the great vessels. The portion surrounding the vessels is arranged in the form of two tubes. The aorta and pulmonary artery are enclosed in one tube, the arterial mesocardium. The superior and inferior vena cava, the cardiac atria and the pulmonary veins are enclosed in a second tube, the venous mesocardium and its attachment to the parietal layer is shaped as an inverted "U" (Ellis, 2005; Gray et al., 2005). The cul-de-sac enclosed between the limbs of the U lies behind the left atrium and is known as the "oblique sinus" while the passage between the venous and arterial mesocardia is termed the "transverse sinus". Furthermore the superior pericardial recess, with a curvilinear shape, lies in the anterior part of pericardium wrapping around the right wall of the ascending aorta (Groell et al., 1999; Breen, 2001; Ellis, 2005; Gray et al., 2005).

Magnetic Resonance Imaging of the pericardium

Magnetic resonance imaging is actually considered the best performing imaging technique in the evaluation of pericardium and pericardial diseases, as it provides a good contrast resolution (Fig. 1), superior than echocardiography, between cardiac tissues and the surrounding soft-adipose mediastinal tissue, excellent delineation of the pericardial anatomy and gives some information even on its histology (Oyama et al., 2004; Francone et al., 2005). MRI may provide comprehensive depiction of the pericardium without the use of either iodinated contrast material or ionizing radiation. It provides a large field of view and a good spatial resolution allowing the examination of the entire chest and the detection of associated abnormalities in the mediastinum and lungs. Moreover MRI is a multiplanar imaging modality, devoid of limitations due to window of investigation (as in ultrasound examination). It can control the movement and minimize artifacts due to blood flow and breathing and provides functional evaluation of cardiac activity (Francone et al., 2005). MRI of the heart is usually performed during apnea by synchronizing with the R wave of electrocardiographic (ECG)-tracing (to avoid movement artifacts and to obtain the acquisition of static images), following the indications of the American Heart Association (Taylor et al., 2010), with scan planes similar to those used in echocardiography: long axis, perpendicular to the septum with longitudinal view of the four cardiac chambers or parallel to the septum with longitudinal view of two chambers (Fig. 2a-b-c), and short axis, with a transversal view of the atrial or ventricular chambers (Fig. 3a-b). The Turbo Spin-Echo (SE) T1-weighted, black blood, and Cine-MRI sequences provide an optimal anatomical resolution (Francone et al., 2005; Webb and Higgins, 2011). MRI shows the normal pericardial sac as a linear band, of low signal intensity due to its fibrous structure (Fig. 1), which separates the epicardial adipose tissue (of high intensity) or myocardial tissue (of medium intensity), from the high intensity of the adipose tissue surrounding the heart. It is best visualized during systole (Sechtem et al., 1986) above the right ventricle and may not be visualized over the lateral and posterior wall of the left ventricle, where there may be less epicardial and mediastinal adipose tissue to provide adjacent tissue contrast (Breen, 2001). The left atrium is only partially covered by the pericardium (Edwards, 1991; Webb W.R. and Higgins C.B., 2011). The thickness ranges from 1.7 ± 0.5 mm in systole to 1.2 ± 0.5 in diastole (Sechtem et al., 1986; Edwards, 1991; Webb and Higgins, 2011). Several pericardial recesses may be detected at MRI, as the transverse pericardial sinus, which is dorsal to the ascending aorta, or the oblique pericardial sinus, which is situated behind the left atrium; a small amounts of fluid may be present in these structures in healthy individuals (Levy-Ravetch et al., 1985; Bull et al., 1998; Webb and Higgins, 2011).

Physiology of the pericardium

Although the pericardium is not a vital organ, it may be considered much more than a sac containing liquid to reduce the friction between cardiac walls and contiguous structures (Nasser, 1970; Spodick, 1997). In fact the fluid that fills the pericardial cavity contains small amounts of complement, other immune factors, prostaglandins and cellular myocardial enzymes, with a biological role possibly wider than the sim-

ple function of lubricant. Furthermore, during development, it represents a source of undifferentiated progenitor cells, which participate in the formation of endothelial, smooth muscle and fibroblasts cells of the developing heart. It also produces cells forming the connective tissue of the heart valves (Di Meglio et al., 2010; Limana et al., 2011). Epicardial cells assume an instructive role providing soluble factors that stimulate coronary vessel development as well as cardiomyocytes proliferation and differ-
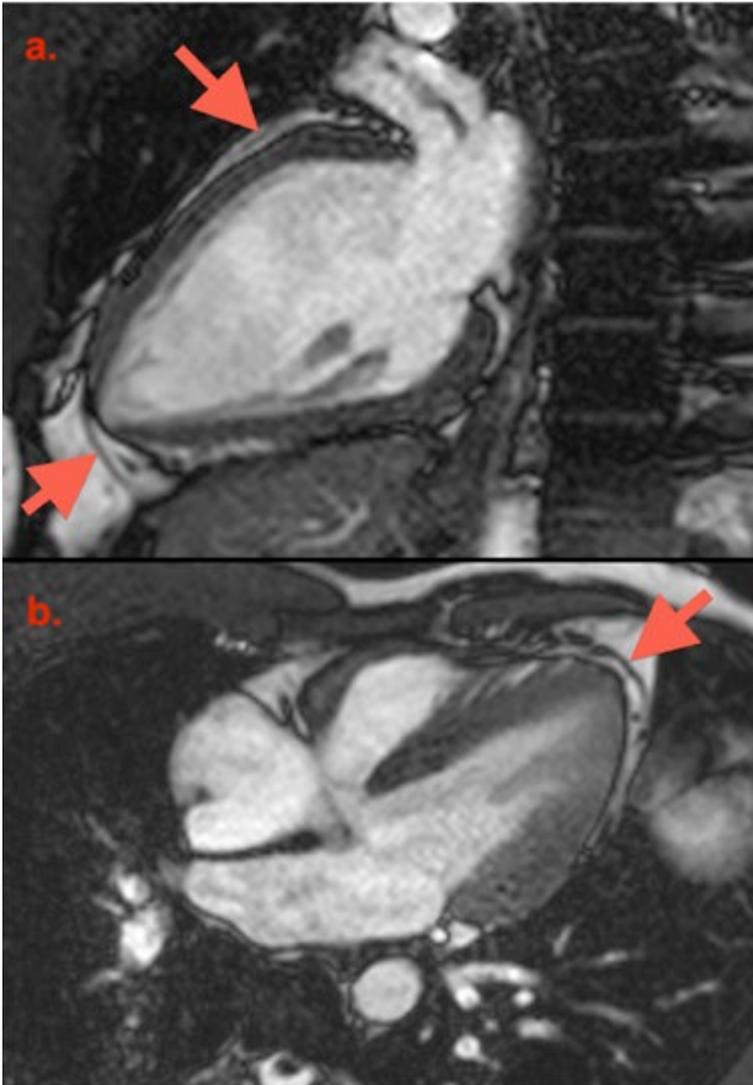

Figure 1 – MRI, sBTFE_BH sequence. Long axis, 2 chambers (a) and 4 chambers (b). Normal anatomy of the pericardium appearing as a linear hypointense band (arrows) between epicardial and pericardial adipose tissue.
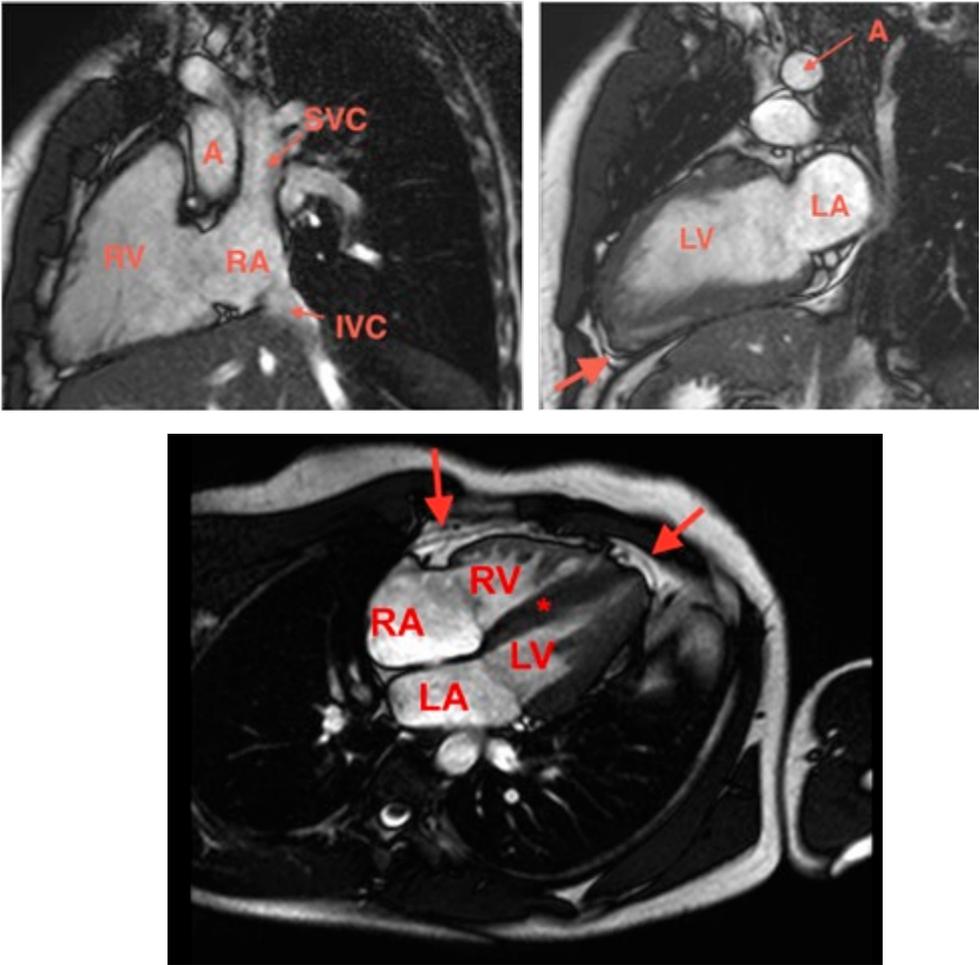
Figure 2 – MRI, sBTFE_BH sequence. Long axis 2 chambers right side (a), long axis 2 chambers left side (b), long axis 4 chambers (c). RA: right atrium; RV: right ventricle; LA: left atrium; LV left ventricle; A: aorta; SVC: superior vena cava; IVC: inferior vena cava; star: interventricular septum. The pericardium is detectable as a linear band of low intensity (arrows) between epicardial and pericardial adipose tissue.

entiation (Winter et al., 2007; Cai et al., 2008). Moreover the adult epicardium may play a role in the physiologic process of myocardial regeneration (Di Meglio et al., 2010; Limana et al., 2011). The pericardium is also a physical barrier against infection, by preventing pathogen spread from lungs and pleural cavity to the heart. It maintains the interdependence between the two ventricles by controlling the expansion of the heart chambers during the periods of volume overload and pressure. The pericardium protects the lungs from trauma due to the beating heart. It is also responsible for fixing the heart in a functional position within the mediastinum and to avoid torsion of the great vessels (Nasser, 1970; Spodick, 1997).
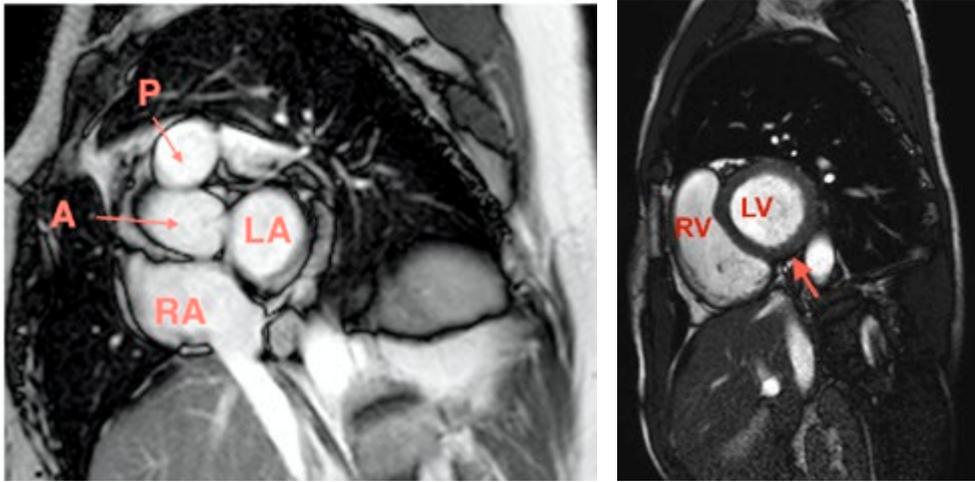
Figure 3 – MRI, sBTFE_BH sequence. Short axis, atria (a) and ventricles (b). RA: right atrium; LA: left atrium; RV: right ventricle; LV left ventricle; A: aorta; P: pulmonary artery. The pericardium is detectable as an hypointense linear band around the posterior wall of the left ventricle (arrow).

Embryology of the pericardium

The pericardium begins to develop in the third week of embryonic life, when small irregular cavities appear in the lateral mesoderm plate. These spaces enlarge and coalesce to form the primitive coelomatic cavity, which will then divide into the pericardial, peritoneal and pleural cavities. As a result of embryonic growth and folding, the pericardial cavity moves to a position ventral to the anterior intestine. By the fourth week, a septum of mesodermal tissue (septum transversum) starts to grow and separates the primitive pericardial from the peritoneal cavity. It contains the liver primordium and the terminal portion of the umbilical and vitelline veins. The division, however, is not complete: pericardio-peritoneal canals remain on both sides, medial to the common cardinal veins (ducts of Cuvier). At the end of the fourth and respectively the fifth week, two folds begin to grow out from the ducts of Cuvier, on each side: the ventral fold forms the pleuro-pericardial membrane while the dorsal one gives origin to the pleuro-peritoneal membrane. The pleuro-pericardial membranes arise along the walls of the trunk on a coronal plane, ventral to the lung buds. At the end of the fifth week these membranes reach the medial wall of the pleuroperitoneal canals, fusing with the ventral mesoderm around the primitive esophagus and eventually obliterate the pleuro-pericardial openings with complete separation of the cavities. Within the seventh week also the separation between the pleural and peritoneal cavities will be completed by the growth of the pleuro-peritoneal membranes. The lung buds expand into pleuro-pericardial canals that become primitive pleural cavities. Each lung extends anteriorly to the front of the primitive heart, surrounding it and infiltrating the space between heart and chest wall (Abbas et al., 2005; Ellis, 2005; Gray et al., 2005).

Pericardial deficiencies are the result of a failure of the pleuropericardial membranes to fuse completely on one or both sides. It is commonly assumed that these defects are linked to a persistence of the pleuropericardial foramen and two different theories on the causative factors that interfere with this closure have been proposed.

In the 1909 Perna (Perna, 1909) suggested that this defect of closure is due to an early and premature atrophy of the left duct of Cuvier. It normally atrophies after forming the innominate left vein (its distal portion persisting as the coronary sinus) reducing blood supply to the pleuro-pericardial membrane; when the extension of the membrane is not yet complete, or the atrophy process is premature, defects of closure may be observed. On the right side the cardinal vein persists developing into the superior vena cava and providing a sufficient blood supply for the closure of the pericardium on the right. This hypothesis may explain why the majority of these defects involve the left pericardium and is the commonly accepted hypothesis. An alternative theory proposed by Sunderland et al. (1944) is a correlation between the heart expansion and the development of pericardium: the pleuropericardial opening has to close before the heart enlarges because the increased volume of the heart stretches the enveloping primitive pericardium. Failure of the membranes to grow and fuse in time would result in a persistent defect. The size of the defect may depend on the rate of growth of the heart and of the enveloping membrane. This assumption may explain both the predominance of the left defects (since the heart develops in volume mainly on the left) and the cases in which there is an absence of pericardial membrane with a normal pleural membrane.

Congenital defects of the pericardium

A defect was first observed by Columbus in 1559 and described in detail by Baillie in 1793. However, it was not until 1959 that Ellis et al. reported the first case recognized with radiological imaging (Ellis et al., 1959). Since this time, an increasing number of cases have been reported. Although rare (only two cases described by Versè, 1909, in 13.000 post-mortem examinations and only one by Southworth and Stevenson, 1938, in 14.000 autopsies), its prevalence may be underestimated since most patients are asymptomatic and the diagnosis is incidental during investigations made for other reasons, during cardiac surgery or at autopsy (Faridah and Julsrud, 2002). It is more common in males and familial occurrence is rare (Southworth and Stevenson, 1938; Abbas et al., 2005). Associated congenital anomalies (such as patent ductus arteriosus, bronchogenic cysts, tricuspid insufficiency, atrial septal defects, left diaphragmatic hernia and pulmonary sequestration) occur in approximately 30% cases (Nasser, 1970).

Pericardial defects include a wide range of congenital pericardial abnormalities usually classified as partial (a foramen in the pericardium sac) or complete, i.e. absence of the entire pericardium (Nasser, 1970). Right side defects are uncommon (17%), while bilateral partial absence of the pericardium are even more rare (13%). Complete congenital absence of the left pericardium is the most frequent defect, representing about 70% of all pericardial defects (Versè, 1909; Tabakin et al., 1965; Nasser, 1970; Abbas et al., 2005).

Clinical features

Most of the reported cases in literature are asymptomatic, discovered incidentally during researches made for other reasons or in the course of an autopsy (Abbas et al., 2005). The most common symptom of complete absence of pericardium is vague chest pain occurring in up to one third of patients; it might result from torsion of the great vessel with increased stress on the anchoring structures of the cardiac base due to absence of the stabilizing effect of the left pericardium (Glover et al., 1969; Faridah and Julsrud, 2002; Abbas et al., 2005). Dyspnea, dizziness, bradycardia and syncope have also been reported (Nasser, 1970; Faridah and Julsrud, 2002). Herniation and strangulation of a portion of heart through the pericardial foramen may be observed in partial pericardial defects, resulting in myocardial acute ischemia and possible death; this never occurs in the presence of a large or complete pericardial defect (Nasser, 1970; Brulotte et al., 2007). Prominent shift of the apical impulse towards the mid-axillary line is frequently observed in patients with complete absence of pericardium. The heart seems enlarged on physical examination and on chest X-ray due to its slight rotation to the left (Schad and Stark, 1992).

There are no specific features at cardiac auscultation: in some cases an ejective systolic murmur may be heard in the second left intercostal space or along the sternal border (Nasser 1970). ECG alterations are commonly observed in cases of complete left pericardial defect and consist of right axial deviation, incomplete right bundle branch block pattern and leftward displacement of the transitional zone in precordial leads (Abbas et al., 2005).

Magnetic Resonance Imaging of defects of the pericardium

In complete defect of pericardium, MRI shows leftward and posterior rotation of the heart, even more evident when the patient is in the left lateral position. The bulging of the main pulmonary artery is also evident as well as the herniation of the lung between the lower border of the heart and left hemidiaphragm and between the pulmonary trunk and aorta. It allows to study the pre-aortic pericardial recess, commonly present in a normal heart, but absent in some cases of agenesis of the pericardium (Sechtem et al., 1986; Spodick, 1997; Oyama et al., 2004; Francone et al., 2005).

In addition, providing a direct image of the pericardial sac with high sensitivity, MRI may detect the possible herniation of the heart through the defect of the pericardial sac, which appears as a notch or a fold of myocardium. Basal MRI shows a prominent convexity on the left side of the cardiac silhouette, in the area of the left atrium, or a groove, if the the pericardial defect extends beyond the basal portion (Spodick, 1997; Faridah and Julsrud, 2002; Abbas et al., 2005). In 10% patients, agenesis of the pericardium may be difficult to detect for the limited presence of pericardial adipose tissue, as commonly observed in children. In these cases, the diagnosis is suggested by indirect signs of complete or partial absence of the pericardium as described above, but must then must be confirmed by thoracoscopy (Faridah and Julsrud, 2002).

Case reports

We report a case of partial defect of the left pericardium and two cases of total agenesis of the left pericardium, detected occasionally and recognized by MRI.

Case 1

A 15-year-old, asymptomatic sportsboy underwent routine ECG exam by a sport medicine physician, in which several extra-systoles were detected; no significant findings were seen at echocardiography, therefore he was referred for cardiac magnetic resonance.

MRI showed partial absence of the pericardium sac on the left side. Axial images revealed normal rotation of the heart and the pericardial coat extending over the inferior and anterior walls of the left ventricle; in sagittal images (Fig. 4a-b) it was observed that the medium-apical portion of the posterior-lateral wall of the left ventricle was not covered by pericardium and was in direct contact with pulmonary parenchyma. Both ventricles were normal for morphology and function, particularly the left ventricular wall did not show any alteration suspicious for herniation of cardiac structures through the defect.

Case 2

A 40-year-old male presented with dyspnoea for some months. Echocardiogram showed the displacement of the cardiac apical window from the left emi-clavear line to the left mid-axillary line, on the fifth intercostal space. The right atrium had a more anterior position than normal, the apical portion of the left ventricle appeared enlarged as for a ventricular diverticulum. The ventricular wall had normal motility, while the inter-ventricular septum presented an anterior paradoxical systolic movement. The patient therefore underwent heart MRI.

Axial MRI demonstrated (Fig. 5) leftward displacement of the heart with the apex extending to the left lateral chest wall and a posterior rotation of the cardiac axis. One could see, on the anterior wall of the right ventricle and atrium, a black hypointense line (1-2 mm thick) that was the parietal pericardial lamina, which was interposed between the hyperintense pericardial adipose tissue and the intermediately dense myocardium. The hypointense line of the pericardium was not detectable around the lateral and posterior walls of the left ventricle, indicating complete defect of the left pericardium.

Case 3

A 24-year-old, healthy sportsboy was checked by ECG during a routine physical examination, which led to detect deviation of the cardiac axis to the right, slight bradycardia (55 bpm), incomplete right bundle branch block, poor progression of the R wave in V1-V5 andprominent P-wave in V3-V4. The patient underwent echocardiography that revealed backward rotation of the cardiac axis with normal ventricular function and slight mitral insufficiency (+/++++) for a mild prolapse.

Therefore we performed a heart MRI to investigate for cardiac abnormalities; the images acquired on an oblique plane (Fig. 6a), parallel to the heart axis, showed pos-
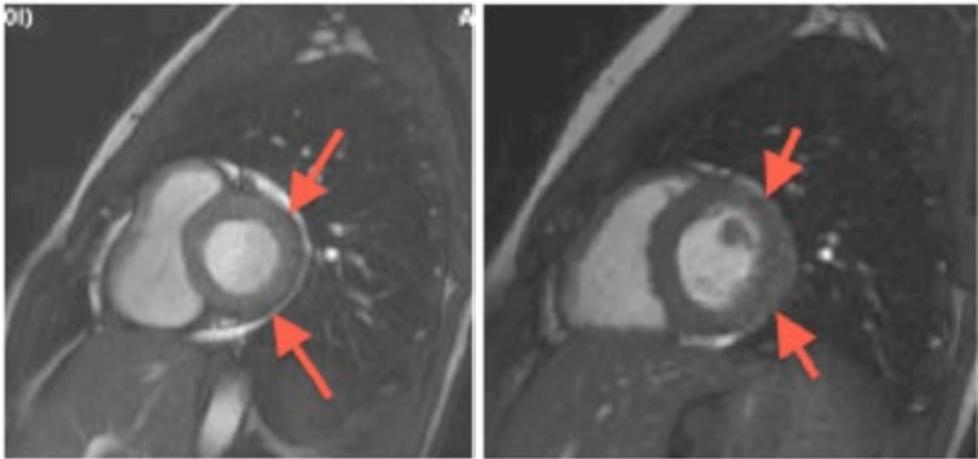
Figure 4 – MRI, sagittal scan: partial agenesis of the left pericardium. (a) The basal portion of the posterior-lateral wall of the left ventricle is covered by pericardium (arrows), which appears as an hypointense line between the myocardium (of medium intensity) and the adipose pericardial tissue (hyperintense). (b) The pericardial coating disappears in the medium-apical portion (arrows). No signs of herniation or ischemia are detected.

Figure 5 – MRI, transversal section: complete agenesis of the pericardium. The left ventricle (arrow) is totally lacking pericardial coating and is rotated in a leftward and posterior position.
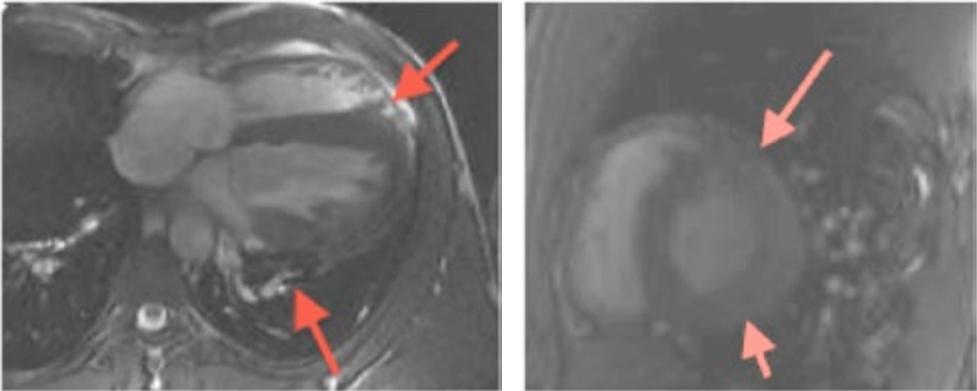
Figure 6 – MRI, transversal (a) and sagittal (b) sections: complete agenesis of the left pericardium. The pericardium is totally lacking over the whole surface of the left ventricle (arrows); there are posterior displacement of the cardiac apex and partial rotation of cardiac axis.

terior displacement of the cardiac apex and partial rotation of cardiac axis. The pericardium was detectable around the free anterior wall of the right atrium and ventricle, but disappeared on the posterior wall of the left ventricle (recognized by the intermediate signal intensity), that was in direct contact with the left lung (of low intensity) through the interposition of only the visceral pleura; lung tissue was partially interposed between the heart and the left hemidiaphragm. Images in the sagittal plane confirmed the complete absence of the left pericardium over the posterior wall of the left ventricle (Fig. 6b).

Discussion

Pericardial defects are a rare condition and have generally been classified for their extension as complete (when heart and lung are in the same cavity) or partial (when pericardial sac and pleural cavity communicate through a defect of the parietal pericardium) and for their position as left, right or bilateral (Nasser, 1970). Complete absence of the left pericardium is the most common defect accounting for 70% cases; right sided defects are uncommon (17%), while bilateral partial absence of the pericardium is even more rare (13%) (Tabakin et al., 1965; Faridah and Julsrud, 2002; Abbas et al., 2005). Most patients are asymptomatic and are detected incidentally during exams made for other reasons or in the course of autopsy. Partial absence of the pericardium may present symptoms as chest pain or dyspnea (Nasser, 1970; Abbas et al., 2005) and may lead to mechanical complications due to entrapment of parts of the heart through the pericardial defect and strangulation of the atria or of a portion of the ventricles (Brullotte et al., 2007). The prognosis of complete absence of the left pericardium and of total lack of pericardium is good; surgery is needed only in case of complications or debilitating symptoms, In partial defects, even if asymptomatic, surgery should be considered due to the possible risk of severe mechanical com-

plications (Van Son et al., 1993; Barcin et al., 2006). Prophylactic intervention is not always required even in complicated partial defects, but patients should be carefully monitored to prevent the risk of herniation, strangulation of the heart and myocardial ischemia (Alcaide et al., 1999; Barcin et al., 2006).

Congenital absence of the pericardium is difficult to detect (Faridah and Julsrud, 2002). Chest X-ray shows poor signs that may lead to suspect complete absence of the pericardium: rotation of the heart to the left side of the chest with the right cardiac border projected over the midline; prominence of the left cardiac border with accentuation of the convexity of the aortic knob and of the main pulmonary artery; widening of the space between the aorta and the main pulmonary artery due to the presence of lung parenchyma; interposition of lung tissue between the left hemidiaphragm and the inferior cardiac border. Partial pericardial defect usually presents with a normal chest radiograph (Ellis et al., 1959; Glover et al., 1969; Schad and Stark, 1992). Even if echocardiographic M-mode and B-mode studies help to detect agenesis of the pericardium, the images may not be considered diagnostic because they do not directly show the pericardium and pericardial defects, although they may show indirect signs. In congenital absence of the pericardium, echocardiogram detects a lateral movement of the apical window from its normal position; the heart seems to be rotated to the left and assumes a teardrop shape. In fact, it is suspended and sustained by only the root of the great vessels and presents tilting and fluctuating movements; interventricular septum in systole moves precociously (paradox movement) (Payvandi and Kerber, 1976; Alcaide et al., 1999; Yamano et al., 2004). Spiral computerized tomography may detect the absence of the pericardium and of the attached adipose tissue, providing non-invasive diagnosis of agenesis of the pericardium. Computerized tomography directly shows parietal pericardium, like a thin structure of medium density between epicardial and mediastinal adipose tissue (Bull et al., 1998; Oyama et al., 2004). This method has a good temporal resolution and provides a large field of view allowing the examination of the entire chest, including lung parenchyma, and the detection of associated pathological conditions (Bull et al., 1998; Breen, 2001; Oyama et al., 2004). However computerized tomography has a lower tissue contrast resolution than MRI and requires the use of ionizing radiations.

Magnetic resonance imaging is now considered the best performing exam in the non-invasive diagnosis of pericardial defects and may be considered the gold standard imaging technique (Breen, 2001; Oyama et al., 2004; Francone et al., 2005). At MRI the pericardial sac appears as a black line (Fig. 1) since it is a fibrous tissue of low signal intensity which divides the epicardial adipose tissue (of high intensity) or the myocardium (of medium intensity) from the high intensity of the adipose tissue of the mediastinum. This hypointense line is usually 1-2 mm tick (Sechtem et al., 1986; Edwards, 1991). In the complete forms of agenesis of the pericardium, MRI, performed with the patient supine, shows the left displacement and the posterior rotation of the heart, which is even more evident when the patient is set in a left sidelong position. Bulging of the pulmonary artery and the herniation of the lung between the inferior border of the heart and the left hemidiaphragm and between the pulmonary trunk and the aorta may also be appreciated. Ventricles usually have normal morphology and function. By offering a direct depiction of the pericardial sac, with good contrast and spatial resolution and a large field of view, this diagnostic investigation

may detect a possible herniation of the heart through the defect of the pericardium, that appears as an indentation or a bulge on the myocardial profile. Furthermore, in the partial defects of the pericardium MRI shows a prominent convexity along the left side of the cardiac silhouette in the area of the left auricle, or a furrow, if the extension of the pericardial defect crosses the basal portion (Sechtem et al., 1986; Spodick, 1997; Faridah and Julsrud, 2002).

Conclusions

Agenesis of the left pericardium is a rare congenital defect, mostly without symptoms; in most cases, it has a good prognosis so it is sufficient to follow the patient with periodic observations. The diagnosis is challenging, even for expert radiologists, because this kind of deformity does not cause specific clinical or imaging findings. Although clinical examination, X-ray chest, ECG, echocardiography and computerized tomography may be of aid in the diagnosis of pericardial defects, nowadays the most effective instrument for certain diagnosis and proper follow up is MRI made free of breathing artefacts through interlock with ECG.

References

Abbas A.E., Appleton C.P., Liu P.T., Sweeney J.P. (2005) Congenital absence of the pericardium: case presentation and review of literature. Int. J. Cardiol. 98: 21–25.

Alcaide A., Platero C., Calle J., Martin M., Barakat S., et al. (1999) Diagnostico de imagen de la agenesia de pericardio. Rev. Esp. Cardiol. 52: 211-214.

Baillie M. (1793) On the want of a pericardium in the human body. Trans. Soc. Improve Med. Chir. Knowl. 1: 91-102.

Barcin C., Ayhan O., Murat K. (2006) Asintomatic congenital pericardial defect: an aspect of modalities and treatment. Anadolu Kardiyol. Derg. 6: 387-389.

Breen J.F. (2001) Imaging of the pericardium. J. Thorac. Imaging 16: 47-54.

Brulotte S., Roy L., Larose E. (2007) Congenital absence of the Pericardium presenting as acute myocardial necrosis. Can. J. Cardiol. 23: 909-912.

Bull R.K., Edwards P.D., Dixon A.K. (1998) CT dimensions of the normal pericardium. Br. J. Radiol. 71: 923-925.

Cai C.L., Martin J.C., Sun Y., Cui L., Wang L., Ouyang K., Yang L., Bu L., Liang X., Zhang X., Stallcup W.B., Denton C.P., McCulloch A., Chen J., Evans S.M. (2008) A myocardial lineage derives from Tbx18 epicardial cells. Nature 454: 104-108.

Columbus M.R. (1559) De re anatomica, Vol. XV; N. Beurlaquae, Venice. P. 265.

Di Meglio F., Castaldo C., Nurzynska D., Miraglia R., Romano V., Russolillo V., Giuseppina L., Vosa C., Montagnani S. (2010) Localization and origin of cardiac CD117-positive cells: identification of a population of epicardially-derived cells in adult human heart. It. J. Anat. Embryol. 115: 71-78.

Edwards E.D. (1991) Applied Anatomy of the Heart. In: Giuliani E.R., Fuster V., eds. Cardiology: Fundamentals and Practice; Mosby-Year Book, St Louis. Pp. 47-51.

Ellis H. (2005) Heart and Mediastinum: Heart and Great Vessels. Elsevier Churchill Livingstone, New York.

Ellis K., Leeds N.E., Himmelstein A. (1959) Congenital deficiencies in parietal pericardium: review with two new cases including successful diagnosis by plain roentgenography. Am. J. Roentgenol. Radium Ther. Nucl. Med. 82: 125-137.

Faridah Y., Julsrud P.R. (2002) Congenital absence of pericardium revisited. Int. J. Cardiovasc. Imaging 18: 67-73.

Ferrans V.J., Ishihara T., Roberts W.C. (1982) Anatomy of the Pericardium. Raven Press, New York.

Francone M., Dymarkowsky S., Kalantzi M., Bogaert J. (2005) Magnetic resonance imaging in the evaluation of the pericardium. A pictorial essay. Radiol. Med. 109: 64-74 quiz 75-76.

Glover L.B., Barcia A., Reeves T.J. (1969) Congenital absence of pericardium: a review of the literature with demonstration of a previously unreported fluoroscopy finding . Am. J. Roentgen. 106: 542-549.

Gray H., Standring S., Ellis H., Berkovitz B.K.B. (2005) Gray's Anatomy: the Anatomical Basis of Clinical Practice. Elsevier Churchill Livingstone, New York.

Groell R., Schaffler G.J., Reinmueller R. (1999) Pericardial sinuses and recesses: findings at electrocardiographically triggered electron-beam CT. Radiology 212: 69-73.

Levy-Ravetch M., Auh Y.H., Rubenstein W.A., Whalen J.P., Kazam E. (1985) CT of the pericardial recesses. AJR Am. J. Roentgenol. 144: 707-714.

Limana F., Capogrossi M.C.,Germani A. (2011) The epicardium in cardiac repair: from the stem cell view. Pharmacol. Ther. 129: 82-96.

Moore R.L. (1925) Congenital deficiency of the pericardium. Arch. Surg. 11: 765–777.

Nasser W.K. (1970) Congenital absence of the left pericardium. Am. J. Cardiol. 26: 466–470.

Oyama N., Oyama N., Komuro K., Nambu T., Manning W.J., Miyasaka K. (2004) Computed tomography and magnetic resonance imaging of the pericardium: anatomy and pathology. Magn. Reson. Med. Sci. 3: 145-152.

Payvandi M.N., Kerber R.E. (1976) Echocardiography in congenital and acquired absence of the pericardium. An echocardiographic mimic of right ventricular volume overload. Circulation 53: 86-92.

Perna G. (1909) Sopra un arresto di sviluppo della sierosa pericardica nell' uomo. Anat. Anz. 35: 323.

Schad N., Stark P. (1992) The radiologic features of cardiac rotation: a pictorial essay. J. Thorac. Imaging 7: 81-87.

Sechtem U., Tscholakoff D., Higgins C.B. (1986) MRI of the normal pericardium. AJR Am. J. Roentgenol. 147: 239-244.

Southworth H., Stevenson C.S. (1938) Congenital defects of the pericardium. Arch. Intern. Med. 61: 223–240.

Spodick D. (1997) The Pericardium: A Comprehensive Textbook. Marcel Dekker, New York.

Sunderland S., Wright-Smith R.J. (1944) Congenital pericardial defects. Br. Heart J. 6: 167–175.

Tabakin B.S., Hanson J.S., Tampas J.P., Caldwell E.J. (1965) Congenital absence of the left pericardium. Am. J. Roentgen. 94: 122–128.

Taylor A.J., Cerqueira M., Hodgson J.M., Mark D., Min J., O'Gara P., Rubin G.D., American College of Cardiology Foundation Appropriate Use Criteria Task Force; Society of Cardiovascular Computed Tomography, American College of Radiology; American Heart Association, American Society of Echocardiography, American

Society of Nuclear Cardiology, North American Society for Cardiovascular Imaging, Society for Cardiovascular Angiography and Interventions, Society for Cardiovascular Magnetic Resonance (2010) ACCF/SCCT/ACR/AHA/ASE/ASNC/ NASCI/SCAI/SCMR 2010 Appropriate Use Criteria for Cardiac Computed Tomography. A Report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, the Society of Cardiovascular Computed Tomography, the American College of Radiology, the American Heart Association, the American Society of Echocardiography, the American Society of Nuclear Cardiology, the North American Society for Cardiovascular Imaging, the Society for Cardiovascular Angiography and Interventions, and the Society for Cardiovascular Magnetic Resonance. J. Cardiovasc. Comput. Tomogr. 404: e1-e33.

Van Son J.A.M., Danielson G.K., Schaff H.V. (1993) Congenital partial and complete absence of the pericardium. Mayo Clin. Proc. 68: 743-747.

Versè M. (1909) Fall von kongenitalem Defekt des Herzbeutels. Muenchen Med. Wschr. 56: 2665.

Webb W.R., Higgins C.B. (2011) Thoracic Imaging – Pulmonary Cardiovascular Radiology. Lippincott Williams & Wilkins, New York.

Winter E.M., Gittenberger-de Groot A.C. (2007) Epicardium-derived cells in cardiogenesis and cardiac regeneration. Cell. Mol. Life Sci. 64: 692-703.

Yamano T., Sawada T., Sakamoto K., Nakamura T., Azuma A., Nakagawa M. (2004) Magnetic resonance imaging differentiated partial from complete absence of the left pericardium in a case of leftward displacement of the heart. Circ. J. 68: 385-388.