Learning objectives

1. Explain embryologic origins, imaging findings and their clinical implications of congenital absence of the right pericardium.

2. Review the importance of imaging in guiding diagnosis and management.

Background

Isolated congenital pericardial absence is rare with a reported frequency of 0.01-0.04% [1, 2]. Right pericardial absence is extremely uncommon, with fewer than 30 cases reported [2, 3]. The prevalence of these entities may be underestimated due to a lack of standardized diagnostic criteria [4].

Many cases of congenital pericardial absence are incidentally detected, however herniation and strangulation of the heart or lungs through the pericardial defect can be life threatening. To date, the majority of literature focuses on the more prevalent left-sided absence. We discuss the embryology of right pericardial absence and review its characteristic radiologic findings.

Images for this section:
Fig. 1: Cases of complete bilateral absence, partial left absence and right pericardial absence comprise one quarter of cases with complete left pericardial absence accounting for the remainder.
Findings and procedure details

Embryology

At the end of the 4th week of embryo development, 2 pleuropericardial folds develop and grow medially toward the midline (Fig. 2). By the end of the 5th week, the pleuropericardial folds fuse, partitioning the thorax and two partially formed pleural cavities.

Congenital defects of the pericardium are generally considered to be a consequence of failure of pleuropericardial membrane closure during development. One theory hypothesizes that the developing heart stretches the enveloping pericardium during development [3]. If the pleuropericardial membranes fail to fuse before the heart enlarges, a pericardial defect is generated, with defect size depending upon the differing growth rates of the heart and the pericardial membrane. An alternative theory proposes that a traction-induced tear may develop in the pleuropericardial membrane, with a resultant pericardial defect. Kaneko et al's [4] case of pericardial absence associated with a divided phrenic nerve, passing both ventrally and dorsally to the defect, illustrates this conjecture. Potentially, a certain percentage of pericardial defects are attributable to each of the above mechanisms.

Imaging

Radiography

If the superior right aspect of the right pericardium is absent, interposed lung between the right pulmonary artery and the aorta produces a characteristic lucency and right pulmonary artery margin prominence (Fig. 3). If the inferior aspect of the right pericardium is absent, interposed lung may create lucency between the right inferior cardiac border and the right hemidiaphragm [6]. In instances of cardiac tissue herniation through a right pericardial defect, a prominent bulge of the right heart border may be seen (Fig. 4).

Computed tomography

Characteristic CT findings of right pericardial absence include visualizing herniation of right heart structures, such as the right atrial appendage (Fig. 5), through a defect in the pericardium. Interposed lung parenchyma herniating through the defect can also be seen (Fig. 6), producing "retroaortic air" anterior to the pulmonary artery in a characteristic configuration.
In the era prior to cross-sectional imaging, diagnostic pneumothorax or diagnostic thoracotomy was recommended to confirm suspected cases of pericardial absence [6]. With advances in imaging techniques, cross-sectional imaging can now confirm suspicion of pericardial absence raised on radiographs, obviating invasive diagnostic procedures.

Magnetic resonance imaging

Cardiac magnetic resonance imaging (MRI) with T1-weighted morphological sequences can demonstrate pericardial defects by the absence of the preaortic pericardial recess or the pericardium itself [7]. Although some propose MRI to be the preferred method to diagnose pericardial defects [8-10], patients with a normal pericardium but decreased pericardial fat may be inaccurately diagnosed as having pericardial absence [8].

Echocardiography

Pericardial defects are not typically directly visualized with echocardiography. Atypical orientation and position of the heart within the chest as well as abnormal wall motion are suggestive of the diagnosis. Echocardiography is also useful in detecting associated congenital cardiac anomalies (Fig. 7).

Clinical consequences

Generally, individuals with pericardial absence are asymptomatic. Symptoms typically result from mechanical complications arising from smaller defects. Herniation of cardiac structures can lead to strangulation [7, 8]. Paroxysmal non-exertional chest pain is the most commonly described symptom, thought to be related to herniation of the right atrial appendage, right atrium and/or ventricle [9].

In addition to herniation-related ischemia, other symptoms such as dyspnea and syncope may be related to compression of great vessels, traction on pleuropericardial adhesions or postural distention of vessels [10]. Symptomatic cardiac tissue herniation or strangulation typically requires surgical liberation of the fibrous ring and pericardioplasty [8]. Xenograft pericardioplasty (9) and defect enlargement by direct excision [12] have also been performed to treat symptomatic, partial defects of the right pericardium. The role of prophylactic surgical repair of incidentally detected defects in asymptomatic individuals remains controversial [7].

Images for this section:
**Fig. 2:** Illustration of normal pericardial development from the fourth to sixth week of gestation. A, At 4 weeks of gestation, the laterally positioned pleuropericardial folds are developed. B, During the 5th week of gestation, the pleuropericardial folds grow toward the midline while the root of each fold migrates ventrally. C, At the end of the 5th week, the pleuropericardial folds fuse, partitioning the thoracic cavity into a pericardial cavity and two partially formed pleural cavities. Note that union of the pleuropericardial folds and the root of the lungs also occur during this time. D, The lungs continue to extend anteriorly to the front of the heart.
Fig. 3: PA chest radiograph revealed an abnormal right mediastinal contour due to partial herniation of the ascending thoracic aorta through a pericardial defect (arrow). Note the ascending aorta is slightly more horizontal due to the herniation.
Fig. 4: Scout view demonstrates a bulging right heart border.
**Fig. 5:** Transaxial CT images confirm partial right pericardial absence. Part of the right atrial appendage extends laterally through the partial defect (arrow).

**Fig. 6:** Lateral radiograph demonstrated abnormal lucency between the ascending aorta and the right pulmonary artery resulting in an unusually dense-appearing and prominent right pulmonary artery (arrow).
Fig. 7: Echocardiography demonstrated a secundum type atrial septal defect with left to right shunting (arrow).
Conclusion

Advancements in diagnostic imaging will improve detection of congenital pericardial absence. Increased awareness and recognition of characteristic radiologic findings by diagnostic imagers is crucial in guiding management and preventing complications in this population.

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References