Clinical Features, Natural History, and Management of Pericardial Cysts

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With the increased use of medical imaging, there has been an increase in the numbers of pericardial cysts identified. However, there is a paucity of data regarding the clinical course for pericardial cysts. Hence, we aimed to study the clinical features and natural history of pericardial cysts. We retrospectively studied all patients with the diagnosis of pericardial cysts based on computed tomography (CT) chest or cardiac magnetic resonance imaging (CMR) between 2008 and 2014. The maximum diameter of the cyst was measured at the initial study (CT/CMR) and was compared with the most recent follow-up imaging modality of the same type if available. A change in the maximum diameter more than 10% was considered significant. We included 103 patients in the study; 89% were asymptomatic and 67% were females. Twenty-nine asymptomatic patients had repeat imaging with the same modality (CT/CMR) with a mean follow-up of 23 months. The maximum cyst diameter decreased by a mean of 25% in 34% of the patients and increased by a mean of 13% in 17% of the patients. The remaining patients (48%) had no significant change. All 29 patients remained asymptomatic. In conclusion, most pericardial cyst cases were asymptomatic. On repeat imaging, approximately 1/3 of pericardial cysts were found to decrease in size, whereas interval enlargement was infrequent and unlikely to be clinically relevant. Therefore, within the limitations of our study, serial imaging in asymptomatic patients with CT or CMR does not appear to impact management decisions. **(Am J Cardiol 2019;123:159−163)**

Pericardial cysts are uncommon masses with an incidence of 1 per 100,000.[1] It is the third most common cystic mass of the mediastinum[2] and constitutes between 4% and 7% of all mediastinal masses.[1] The majority is asymptomatic and usually detected incidentally by various imaging modalities. However, rarely they can be associated with serious complications such as cardiac tamponade, obstruction of the right main stem bronchus, or even sudden death.[3] The clinical course of pericardial cysts is poorly defined in the literature. The aim of this study is to define the clinical features and natural history of pericardial cysts to help guide management decisions.

Methods

We conducted a single center, retrospective, observational study of all patients at the Cleveland Clinic with an ICD-9 code reflecting the diagnosis of pericardial cyst between 2008 and 2014. Patients were included in the study if they had computed tomography (CT) or cardiac magnetic resonance imaging (CMR) with characteristic imaging findings of a pericardial cyst. This was defined for CT as a thin-walled, nonenhancing structure with near water attenuation values, and for CMR as low signal intensity on T1-weighted images and high signal intensity on T2-weighted images without contrast enhancement.[4,5] Pericardial cysts were distinguished from diverticula by the presence of a complete wall and the absence of an open communication with the pericardium.

The diameters of the pericardial cyst were measured in different planes (transverse, coronal, or sagittal views if available) in the initial imaging study (CT/MRI). The maximum diameter of the cyst was compared with the most recent repeat imaging modality of the same type if available. A change in the maximum diameter of the pericardial cyst more than 10% was considered significant. A single blinded investigator performed all measurements and assessments. The study was approved by the Institutional Review Board, informed consent was waived, and data were deidentified. Continuous variables were reported as mean ± SD. Comparison was done using the *t* test for continuous variables and chi-square test or Fisher's exact test for categorical variables. A multivariable logistic regression model with stepwise selection was used to identify significant independent risk factors that affect the growth of pericardial cysts. For all analyses, a p value of < 0.05 was considered statistically significant. All analysis was conducted using SPSS version 23.

Results

Table 1 shows baseline characteristics of the 103 patients who were included in the study; 26 patients were

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Table 1
Baseline characteristics and imaging features of patients with pericardial cysts (n = 103)

Variable	Total (n = 103)
Age, (years)	49 ± 16
Women	69 (67%)
Chest pain	8 (8%)
Dyspnea	2 (2%)
Palpitations	1 (1%)
Number of cysts	
Single	100 (97%)
Multiple	3 (3%)
Size (cm)	5.5 ± 2.8
Cyst type on CT or CMR	
Simple	94 (91%)
Complex	9 (9%)
Abnormalities seen on chest x-ray (n = 99)	31 (32%)
Pericardial cyst detected on TTE (n = 86)	34 (38%)
Right heart border (n = 68)	25 (37%)
Left heart border (n = 15)	9 (60%)
Anterior mediastinum (n = 3)	0 (0%)

Data presented as mean (standard deviation) or number (percentage) unless otherwise stated. CMR = cardiac magnetic resonance imaging; CT = computed tomography; TTE = transthoracic echocardiogram.

diagnosed by MRI, and 77 patients were diagnosed by CT. The majority were women (67%) with a mean age at diagnosis of 49 years. Eleven percent was symptomatic, with chest pain being the most common complaint followed by dyspnea and palpitations. The location of the pericardial cyst was most commonly seen in the right heart border (81%), less frequently in the left heart border (17%), and in the anterior mediastinum (3%). The mean maximum diameter of the pericardial cyst was 5.5 cm. One hundred patients (97%) had a single pericardial cyst, and 94 (91%) were simple cysts. A complex cyst (defined as the presence of solid elements within the cyst or the presence of internal septations) was seen in 9% of patients. Nonspecific abnormalities were detected on chest x-ray in 31 (32%) patients. The sensitivity of transthoracic echocardiogram (TTE) in detecting pericardial cyst was 38% with a nonsignificantly higher sensitivity in left-sided cysts compared with the right-sided cysts (60% vs 37%, p = 0.15).

In the 11 patients with symptomatic pericardial cysts, 6 patients underwent surgical resection and 1 patient underwent aspiration and sclerosis of the pericardial cyst but developed recurrence of the pericardial cyst.

Twenty-nine asymptomatic patients had a repeat imaging study of the same modality (CT or CMR) with a mean follow-up of 23 months ±14 (Figure 1). Fourteen patients (48%) had no significant change in the maximum cyst diameter. The maximum cyst diameter increased in 5 patients (17%) by a mean increase of 13% (range 11% to 17%), 0.6 cm (range 0.3 to 1 cm) whereas the maximum cyst diameter decreased in 10 patients (34%) by a mean of 25% (range 11% to 66%), 1.05 cm (range 0.6 to 2.2 cm). In the multivariate analysis of age, size, gender, and type of cyst, we did not identify any variable that significantly correlated with changes in pericardial cyst size.

Discussion

Despite the increase in the number of pericardial cysts identified in the general population, there is still a paucity of data regarding the clinical course and the appropriate follow-up of pericardial cysts. To the authors' knowledge, this is the largest study to evaluate the clinical characteristics of pericardial cysts and the first to report their natural history. Our study shows that the majority of pericardial cysts are asymptomatic, predominantly found in females and located on the right side of the heart. Following the natural history of pericardial cysts over an average of 2 years, approximately 1/3 of pericardial cyst were found to decrease in size on repeat imaging, and interval enlargement of pericardial cysts, when present, is infrequent and is unlikely to be clinically significant on intermediate length follow-up.

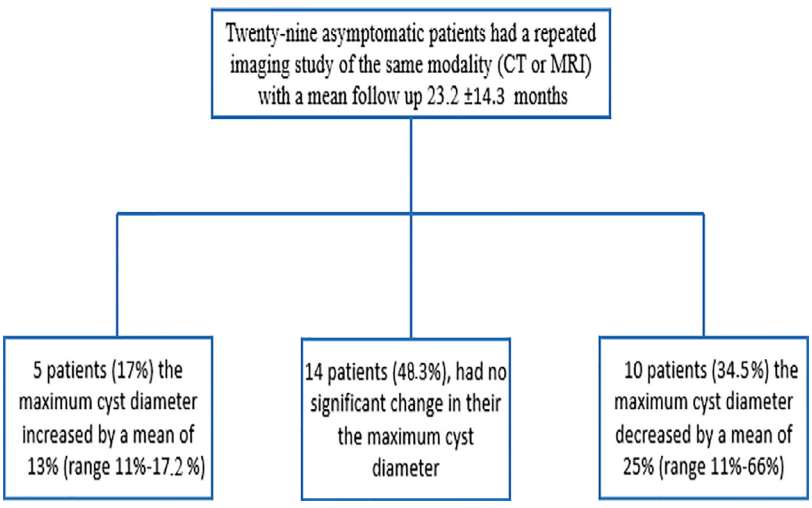

Figure 1. Changes in the maximum pericardial cyst diameter during the follow-up period. The maximum diameter of the cyst was compared to the most recent repeat imaging modality of the same type if available. A change in the maximum diameter of the pericardial cyst more than 10% was considered significant. The figure shows the change in the maximum diameter of the pericardial cyst during the follow-up of the 29 asymptomatic patients.

Pericardial cysts can be classified into 2 categories: congenital and acquired. Embryologically, the pericardium arises from lacunas that gradually merge and form the pericardial cavity; if they fail to unite a pericardial cyst occurs.[6] Acquired pericardial cysts can occur due to infections, with the leading cause being a rare complication of Echinococcosis hydatid cysts.[7] Tuberculosis causing pericardial cyst has also been described.[8] Other acquired causes include malignant metastasis,[9] post-traumatic,[10] pericarditis,[11] cardiac surgery, and rheumatic heart disease.[12] In our study, 1 patient's pericardial cyst was clearly attributed to post cardiac surgery and the etiology of the remaining 102 patients was attributed to be congenital and/or idiopathic.

The majority of the pericardial cysts are asymptomatic.[1] The clinical manifestations vary depending on the location

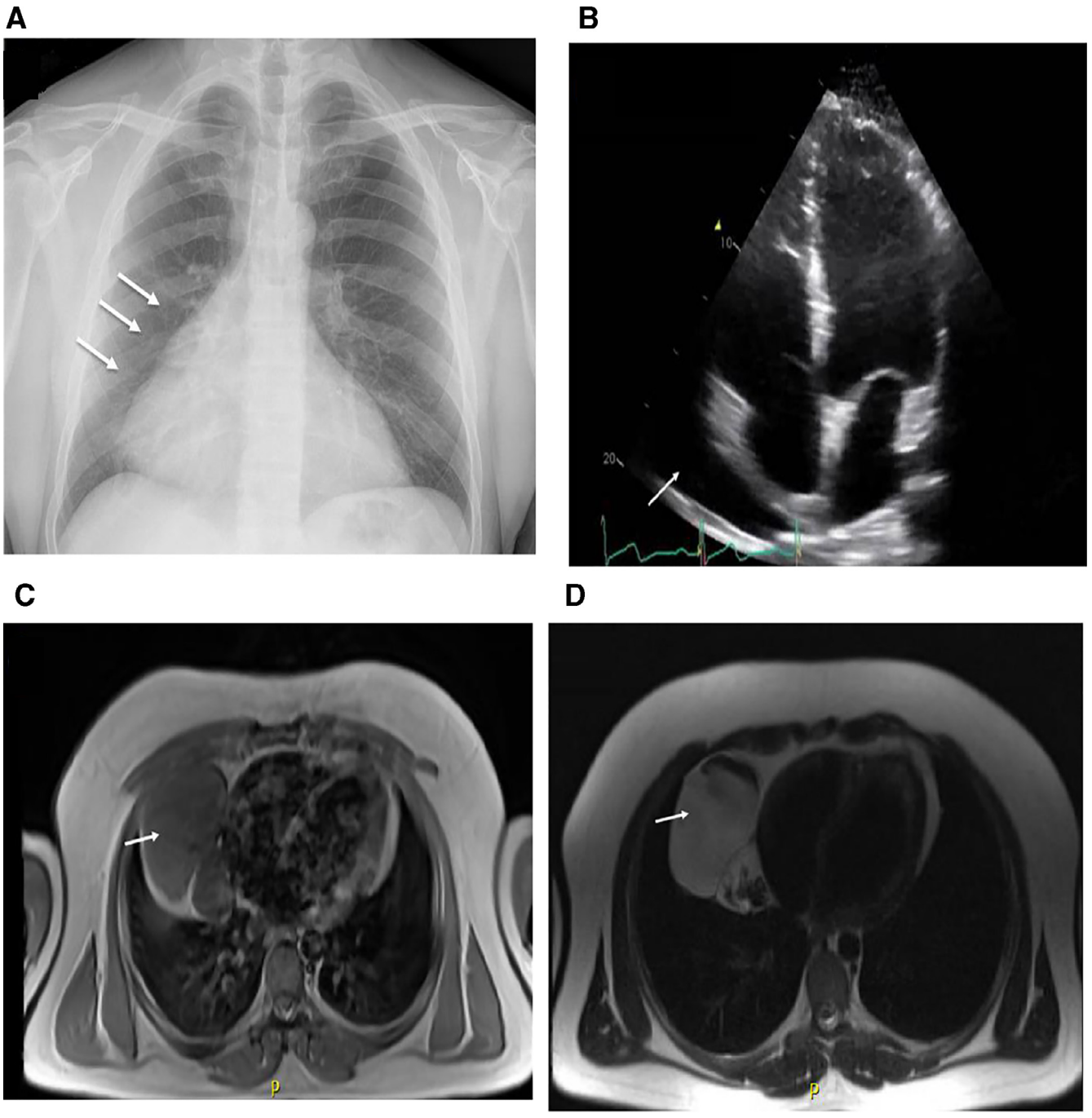

Figure 2. **(*A*)** Chest radiograph showing a well-circumscribed density (*white arrows*) along the right heart cardiophrenic angle **(*B*)** TTE showing the presence of an echo-lucent space (*white arrow*) adjacent to the right atrium suggesting a pericardial cyst. **(*C*)** CMR T1-weighted image showing a lobulated, low-intensity signal, cystic mass (*white arrow*) within the pericardium to the right of the right heart border. **(*D*)** CMR T2-weighted image showing a high-intensity signal of the cystic mass. CMR = cardiac magnetic resonance imaging, TTE = transthoracic echocardiography.

of the cyst which usually includes chest pain, dyspnea, or palpitations.[3] More serious and rare complications can occur including cardiac tamponade, obstruction of the right main stem bronchus, or sudden death.[3] In our study, 11 patients (11%) were symptomatic on initial presentation.

Chest x-rays occasionally show an abnormal soft tissue density along the heart border, mainly in the cardiophrenic angles (Figures 2, 3). Such nonspecific findings were present in 32% of our cohort. Transthoracic echocardiography is a modality that can exclude other diagnoses (e.g., fat pad) and support the diagnosis of pericardial cyst by demonstrating a thin walled, echo-free space adjacent to the cardiac border filled with fluid that lacks any flow by color Doppler (Figure 2).[13] A major limitation of transthoracic echocardiography is its low sensitivity for detection of a pericardial cyst. In our cohort, 86 patients had a TTE, with a mean time difference between obtaining the TTE and CT/CMR of 63 days. Its sensitivity was 38% with a nonsignificantly higher sensitivity in left-sided cysts of 60%. One possible explanation for this discrepancy is the predominant location of the pericardial cyst on the right heart border where image quality can be limited with traditional echocardiographic views. Hence, if a pericardial cyst is suspected, off-axis views of the right heart border, e.g., superiorly oriented transthoracic subcostal views,[14] can be used to increase the sensitivity for detection. The use of transesophageal echocardiography may better visualize pericardial cysts, especially if they are in an atypical location.[15]

The American Society of Echocardiography recommends obtaining a CT or CMR when a pericardial cyst is detected by chest x-ray or echocardiography to confirm the diagnosis.[13] The typical appearance of a pericardial cyst on CT is a nonenhancing cyst with homogeneous water attenuation (Figure 3).[16] With CMR, the pericardial cyst appears as a nonenhancing thin-walled mass with gadolinium administration, and the fluid content of the cyst will demonstrate a low T1 signal intensity on T1-weighted images and high T2 signal intensity on T2-weighted images (Figure 2).[13] CMR is superior compared with CT in defining the fluid nature of the cyst, since some cysts may contain nonserous fluid that has high attenuation at CT, which can be mistaken as a solid mass. In contrast with CT, these cysts continue to have characteristically high signal intensity when imaged with T2-weighted sequences, regardless of the nature of the cyst's contents.[17]

To date, no study has described the natural history of pericardial cysts, and the literature is limited to case reports describing the complete resolution of the pericardial cyst[18–20] with the mechanism thought to be cyst rupture or resorption. In our study, 29 asymptomatic patients had a repeated imaging study of the same modality with a mean follow-up of 23 months. Based on the maximum cyst diameter, 48% of the pericardial cysts did not have any significant change, approximately 1/3 of the patients had a significant decrease, and 17% had a significant increase in the maximum diameter. Although a small percentage did increase in size, these patients remained asymptomatic. The current American Society of Echocardiography recommendations for pericardial disease support serial monitoring with CT or CMR every 1 to 2 years.[13] However, our data may suggest that a reasonable follow-up approach could be to decrease the frequency of serial imaging to at least 3 to 5 years unless indicated by a clinical change in the patient, hence minimizing unnecessary costs and/or radiation exposure. Further research is necessary to make formal changes to serial frequency imaging of the pericardial cysts.

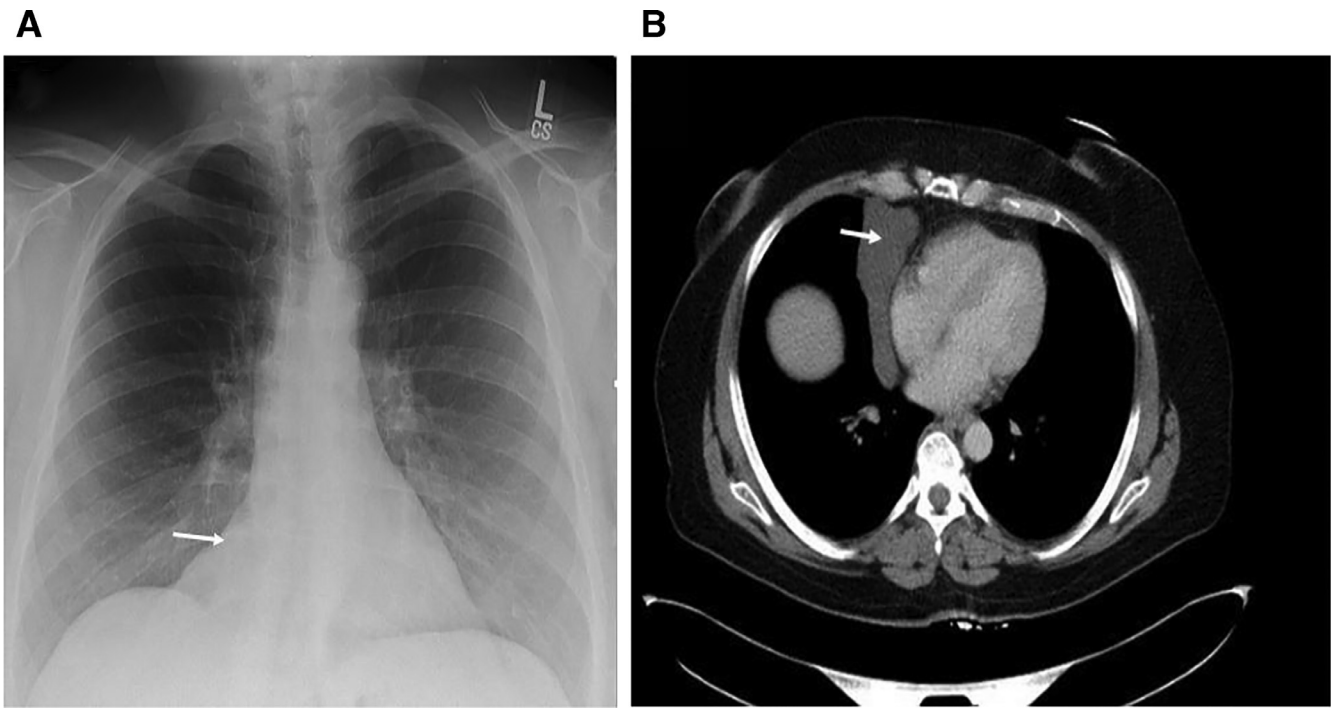

Figure 3. **(*A*)** Chest radiograph showing a triangular density (*white arrow*) at the right cardiophrenic angle **(*B*)** CT chest with contrast showing a thin-walled, nonenhancing pericardial mass (*white arrow*) compatible with a pericardial cyst. CT = Chest tomography.

Treatment options in patients with severe symptoms include: percutaneous aspiration,[21−23] ethanol sclerosis,[24,25] or resection through video-assisted thoracoscopic surgery,[3,26,27] thoracotomy,[3,27,28] or median sternotomy.[29] The 2015 European Society of Cardiology guidelines recommend percutaneous aspiration and ethanol sclerosis as a first line if feasible.[12] Despite it being less invasive and the ability of percutaneous aspiration to be done emergently if needed,[23] this approach lacks the ability to make a histopathological diagnosis, may be associated with a higher recurrence due to fluid reaccumulation,[3] reaching up to 33%,[30] with the possibility of injury to nearby structures. Seven of the 11 symptomatic patients in our cohort had severe enough symptoms to warrant intervention. The one patient who underwent aspiration and sclerosis experienced a symptomatic recurrence of his pericardial cyst that was managed conservatively.

This study has several following limitations: (1) retrospective single-center design; (2) pericardial cyst diagnosis was based on imaging findings rather than histological diagnosis; (3) the measurement of changes in maximum diameter of the cysts may not accurately reflect the overall changes of the size given the irregular shape of the cyst; (4) only 29 patients had a serial imaging follow-up; and (5) finally, mean follow-up imaging was limited to approximately 2 years and thus only provides an intermediate duration of follow-up.

In conclusion, majority of the pericardial cysts are asymptomatic. Most of the asymptomatic cysts remain stable or decrease in size with repeat imaging on an intermediate range follow up. Although small percentage of cysts increase in size, this change remains clinically insignificant. Severe symptomatic pericardial cysts require intervention, and given the higher chance of pericardial cyst recurrence after sclerosis and aspiration, surgical approach may be considered as the first line of therapy.

Disclosures

The authors have no conflicts of interest to disclose.

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