Pericardial Disease

26

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Abstract

The investigation and management of pericardial disease remains difficult due to the limitations of echocardiography in visualising the entire pericardium. This chapter explores the role for CMR in pericardial disease, covering pericardial effusion, constrictive pericarditis, congenital pericardial abnormalities, and pericardial tumours. The ability of CMR to identify pericardial anatomy as well the the functional consequences of pericardial pathology is emphasised.

Keywords

Pericardial constriction • Ventricular coupling • Pericardial effusion • Pericardial defect • Pericardial tumour • Pericardial cyst • Pericarditis

Pericardial Effusion

CMR Protocol in Pericardial Effusion

1. Anatomy module including T1 & T2 weighting
2. LV function module
3. Targeted sequences depending on findings e.g. tumour protocol, valve imaging, real-time dynamic respiratory cine.
4. Late Gadolinium Enhancement module

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Introduction

The aetiology of pericardial effusions is varied and can be broadly divided into transudates, exudates, haemorrhage and chyle. A full review of the causes of pericardial fluid is beyond the scope of this text. The role of imaging is to identify the effusion, assess its physiological significance and, if possible, diagnose the cause. Exudative processes tend to be associated with more complex effusions and greater pericardial inflammation, transudates conversely are generally simpler. Cross-sectional imaging has a specific role in assessing the extracardiac structures to identify malignancy and other systemic diseases.

CMR Versus Other Imaging Modalities

Echocardiography remains the primary investigation for diagnosis of pericardial effusions but is susceptible to both false negative and positive results due to the presence of loculated fluid, poor acoustic windows or adjacent pleural fluid. CMR demonstrates the whole of the pericardium, as well as the surrounding structures, making it an ideal tool for secondary imaging.

The functional significance of a pericardial effusion is also primarily assessed by clinical and echocardiographic examination. CMR provides similar information to echo regarding compromise to RV and RA filling but is advantageous if acoustic windows are poor or difficult to interpret.

Findings on CMR

The pericardial space normally contains a trace of pericardial fluid (<30 ml) which may be observed on CMR. There are no absolute CMR criteria for differentiating a physiological effusion from a pathological one but as a guideline a pericardial width of >4 mm is considered abnormal. An effusion measuring >5 mm anterior to the RV is likely to be a moderate volume (100–500 ml). Most effusions show a gravitational distribution being deepest posterolateral to the left ventricle.

Black Blood Images (Spin Echo Sequence)

It is important to initially scan the whole thorax when imaging a pericardial effusion. This will usually be performed with a fast black blood sequence and the intention is to identify extra-cardiac pathology such as lung tumours, lymph nodes, pleural effusions etc. Echocardiography provides little or no information about the extra-cardiac structures and this is a major advantage for CMR. Computed tomography (CT) fulfils a similar role but provides more detail relating to the lungs as well

Table 26.1 Findings on CMR in pericardial effusion of different aetiology

	T1 signal (SE)	Cine appearances and signal intensity (b-SSFP)
Transudate	↓	Simple ↑
Exudates	↓↑	Complex ↓↑
Haemorrhage	↓↑	Complex ↓↑
Chylous	↑↑	Simple ↑

as the abdomen. CT and MRI are thus complementary and may well both be indicated (In practice CT is generally used if there is a strong suspicion of underlying malignancy whilst MRI is favoured if functional information is required)

In principle T1 characteristics of the fluid can give an indication of the nature of the effusion (see Table 26.1). In practice the signal return from the fluid is more influenced by flow voids and loculation than by pure T1 effects. A more useful finding is the complexity of the effusion and degree of pericardial thickening and enhancement – transudates tend to be simple with little pericardial inflammation while exudates and haemorrhage are complex with greater pericardial thickening.

CINE Imaging (b-SSFP)

Pericardial fluid generally appears as high signal on b-SSFP techniques, comparable to pleural effusions (if these should be present). Loculation and soft tissue stranding is well visualised as areas of mixed or reduced signal within the fluid effusion.

As with echocardiography the size of the effusion has a limited relationship with the physiological consequences, being more related to the duration and rate of accumulation of fluid. CMR criteria for functionally important effusion are similar to echocardiography; diastolic compression of the RV free wall, early systolic collapse of the RA, distortion of the LV and RV morphology and potentially paradoxical interventricular septal motion during inspiration (ventricular coupling – more commonly seen in constrictive pericarditis).

Targeted Sequences

Depending on the initial findings further sequences may be indicated:

1. Pericardial inflammation – linear high signal with T1 weighted gadolinium enhanced images.
2. Tumour – first pass perfusion, T1 weighted post-gadolinium images, myocardial delayed enhancement, myocardial tagging.
3. Myocardial disease – additional tissue characterisation sequences (T1/T2/STIR/T1 & 2 mapping), myocardial delayed enhancement

The Role of CMR in Pericardial Effusion

1. CMR is useful in confirming the presence and size of a pericardial effusion if doubt exists on Echo (particularly if loculated).
2. CMR may identify the cause of a pericardial effusion or give indications as to the aetiology.
3. CMR provides functional information about the haemodynamic consequences of the effusion.
4. CMR is complementary to other imaging modalities.

Case Example

A 65 years old lady presented with increasing breathlessness on exertion and swollen ankles. She had a long history of Rheumatoid arthritis. An Echo suggested a pericardial effusion but this was not well visualised so that CMR was requested.

Following pericardial drainage the effusion was found to be an exudate and was rheumatoid factor positive. She was diagnosed with constrictive-effusive pericarditis and required a surgical pericardial window followed by pericardectomy (Figs. 26.1, 26.2, and 26.3).

Tips and Tricks

1. Cine images usually show pericardial effusions as high signal, if a pleural effusion is present the signal will often be similar on cine images but differ on TSE sequences.

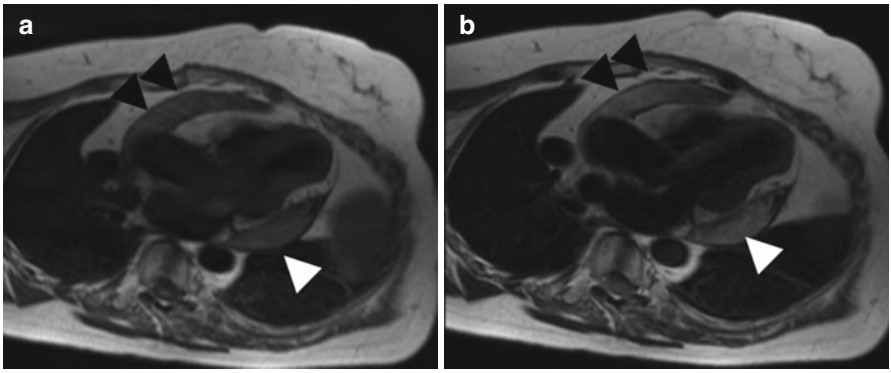

Fig. 26.1 (**a**) T1 TSE image in a 3-chamber view shows an intermediate signal pericardial effusion (*arrow heads*). (**b**) T2 weighted image in the same position again showing an intermediate signal (*arrow heads*). The signal intensity suggests a proteinaceous effusion/exudate rather than a simple transudate which should be lower signal on T1 images

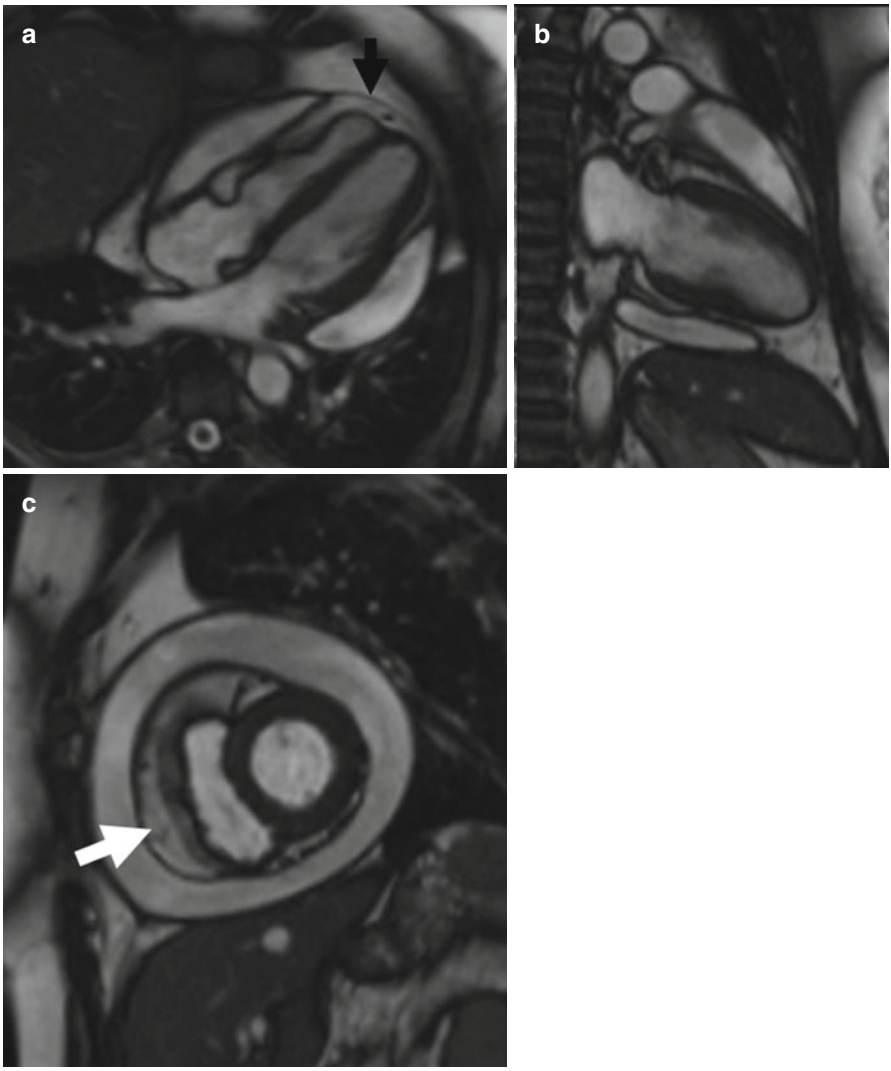

Fig. 26.2 (**a**) 4-Chamber cine image showing a loculated pericardial effusion anterior to the RV and posterior to the LV. The RV chamber is distorted and compressed suggesting that the effusion is haemodynamically significant. Normal pericardium is seen over the apex (*arrow*). (**b**) 2-Chamber cine image. The loculated effusion has a similar distribution to Fig. 26.3a. (**c**) Short axis cine. The effusion is circumferential and has a typical high signal on SSFP images. Epicardial fat (*arrow*) is easily visualised

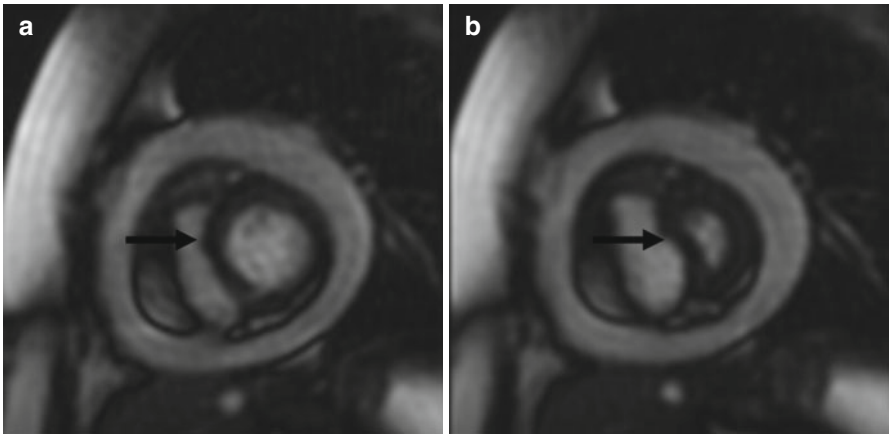

Fig. 26.3 (**a**) Real-time short axis cine image taken during expiration shows a normal interventricular septal position (*arrow*). (**b**) Real-time short axis cine image taken from the same cine loop during early inspiration. There is clear septal displacement to the left side (*arrows*) confirming a constrictive physiology

CMR Report in Pericardial Effusion

Morphology (descriptive)

1. Pericardial Thickness: describe as local or circumferential and list thickness measurements
2. Pericardial effusion : presence and extent

Ventricular parameters

1. LV volumes
2. Ventricular wall motion:
   Systolic wall motion
   Septal motion during normal respiration and breath holding.
3. Presence or absence of atrial inversion

Late Gadolinium Enhancement in RV, LV and pericardium

Key Points CMR in Pericardial Effusion

1. Useful adjunct to Echo
2. Gives functional and anatomical information
3. May identify or indicate the cause

Constrictive Pericarditis

CMR Protocol in Constrictive Pericarditis

1. Anatomy module including T1 & T2 weighting
2. LV function module
3. Targeted sequences depending on findings e.g. real-time dynamic respiratory cine, tagging, mitral valve flow.

Introduction

Constrictive pericardial disease is the end result of a number of inflammatory, infective, or malignant processes involving the pericardium causing a constrictive physiology. Usually this is associated with macroscopic thickening of the visceral and/or parietal pericardium which may become adherent to the myocardium and regularly calcifies (particularly if tuberculous in origin). Constrictive pericarditis may however be associated with a pericardial effusion (effusive-constrictive pericarditis) or a normal thickness pericardium. Generally it is a progressive and chronic condition but may be acute or rarely transient.

The physiological consequence of pericardial constriction is reduced ventricular diastolic filling. Ventricular filling pressures therefore increase in a similar way to restrictive cardiomyopathy, the main differential diagnosis. In constrictive pericarditis, however, the pericardial volume becomes fixed creating competition for diastolic filling between the ventricles – ventricular coupling or interdependence. The end result is preferential RV filling during inspiration (when negative intrathoracic pressure encourages systemic venous return), and preferential LV filling during expiration (when positive intrathoracic pressure encourages pulmonary venous return). The CMR sequelae of this is paradoxical displacement of the interventricular septum to the left side during early inspiration and normalisation during expiration. Ventricular coupling is not seen in restrictive cardiomyopathy.

The Role of CMR in Constrictive Pericarditis

The primary role of cross-sectional imaging in patients with signs and symptoms suggestive of constrictive pericarditis or restrictive cardiomyopathy is to identify pericardial thickening. The presence of thickened pericardium in the appropriate clinical context effectively differentiates constrictive pericarditis from restrictive cardiomyopathy and allows planning of pericardial stripping. Echocardiography is relatively poor at visualising the pericardium and hence cross-sectional imaging is superior. CT demonstrates the pericardium well and has the advantage of being very sensitive to pericardial calcification. CMR, as well as showing the pericardium, has a much greater ability to provide functional information to support the diagnosis of

constriction. CT and CMR are therefore often complementary particularly in the difficult diagnostic case.

Findings on CMR

Pericardial Thickening

The normal pericardium is seen over the RV free wall and the atrioventricular and interventricular grooves where there is abundant surrounding fat. The pericardium is usually pencil thin except over the diaphragmatic reflections. A thickness of >4 mm on spin-echo sequences is generally considered to be pathological. Pericardial thickening is often patchy and may in some cases be absent.

Cine images readily differentiate pericardial effusion from thickening but tend to overestimate the thickness.

Indirect Signs of Constrictive Physiology

1. Distortion of RV and LV shape. The RV in particular may be flattened and tubular. This is best demonstrated with cine imaging.
2. Atrial dilatation (in the absence of ventricular dilatation).
3. IVC and SVC dilatation (reflecting elevated filling pressures).
4. Pericardial adhesions between the thickened pericardium and the epicardial surface of the myocardium. This may be highlighted by tagged cine imaging demonstrating loss of the normal slippage of the pericardium over the myocardium.
5. Ventricular coupling. Paradoxical diastolic interventricular septal motion during early inspiration. This is best seen with a real-time cine sequence during deep breathing.

Tips and Tricks

1. Use real-time dynamic respiratory sequence in several short axis views and a 4-chamber view.
2. The 4-chamber view demonstrates the whole length of the interventricular septum, the paradoxical septal motion often being limited to one part of the septum.
3. The short axis views show the diaphragmatic position clearly, allowing assessment of the respiratory phase.
4. Obtaining several short axis views samples the septum at a number of levels.

The Role of CMR in Constrictive Pericarditis

1. CMR identifies pericardial thickening.
2. CMR provides functional information that may support or indicate constrictive physiology.
3. CMR helps exclude restrictive cardiomyopathy.
4. CMR is complementary to other imaging modalities.

Case

This 80 year old man presented with a history of increasing exertional breathlessness and upper abdominal pain. An Echocardiogram suggested reduced RV function and diastolic dysfunction. He was referred for a CMR scan which diagnosed constrictive pericarditis. He subsequently underwent pericardiectomy and has made a full recovery (Figs. 26.4, 26.5, 26.6, 26.7, and 26.8).

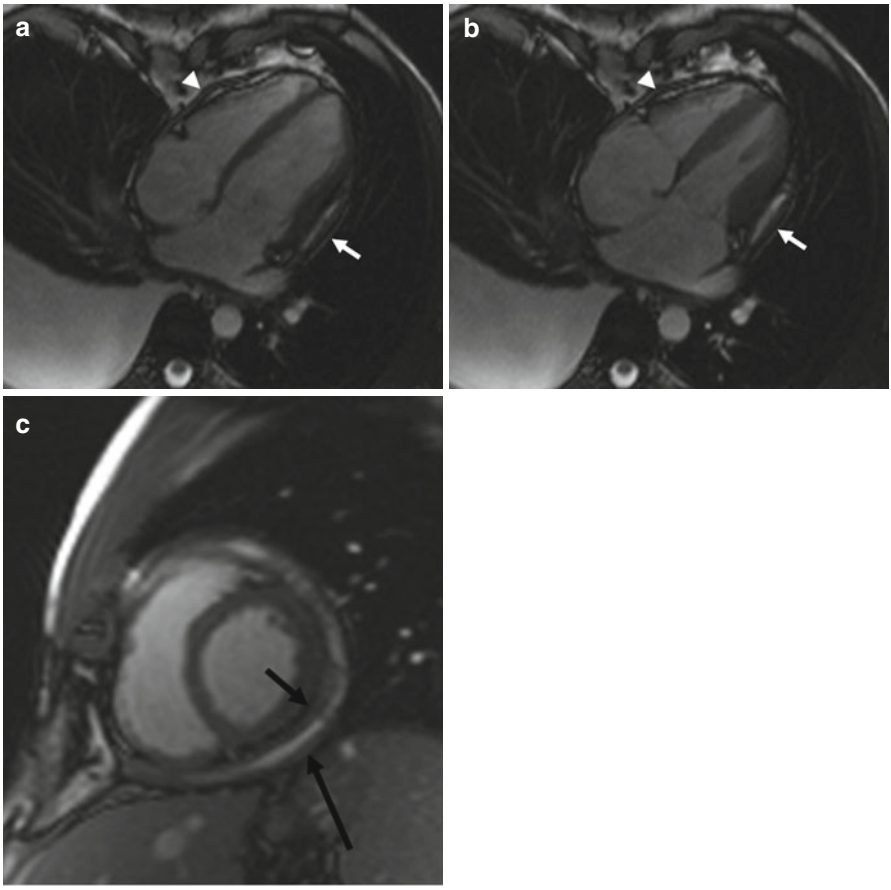

Fig. 26.4 (**a**–**c**) 4 chamber cine image in diastole (**a**) and systole (**b**). There is pericardial thickening over the lateral wall with a trace of pericardial fluid (*white arrow*). There is further pericardial thickening anterior to the RV (*arrow head*). There is subtle distortion of the LV free wall during diastole and bi-atrial dilatation – features suggesting a constrictive physiology. Note the substantial right pleural effusion. (**c**) Short axis cine image showing thickened parietal (*long black arrow*) and visceral (*short black arrow*) pericardium separated by a thin pericardial effusion

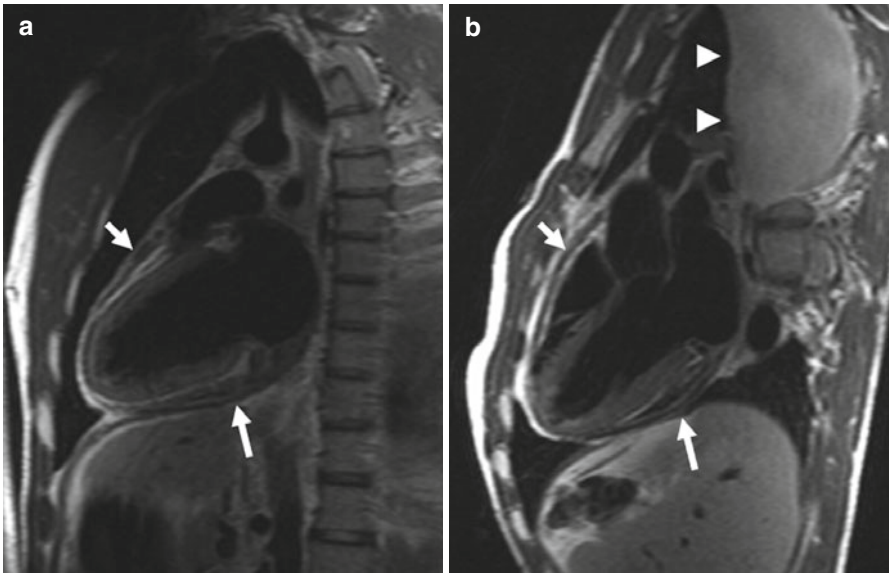

Fig. 26.5 (**a**, **b**) T1 weighted TSE image in a 2-chamber plane (**a**) and T2 weighted image in the 3-chamber plane (**b**) showing diffuse pericardial thickening (*arrows*) sparing the apex. This measures up to 1 cm in thickness. Note is made of the pleural effusion (*arrowhead*)

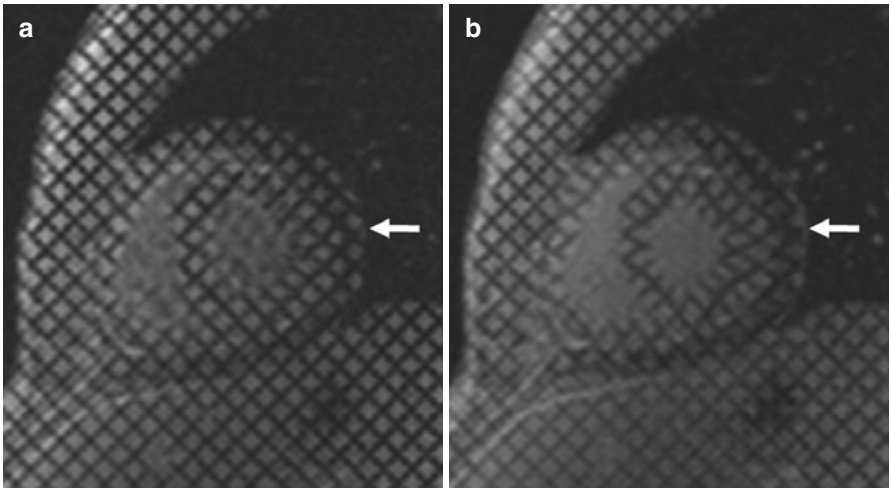

Fig. 26.6 (**a**, **b**) Myocardial tagged image in the short axis plane at end-diastole (**a**) and end-systole (**b**). There are adhesions between the pericardium and the myocardium over the lateral wall demonstrated on the images as failure of 'slippage' of the tag lines between the myocardium and pericardium during the cardiac cycle (in the region of the *arrows*)

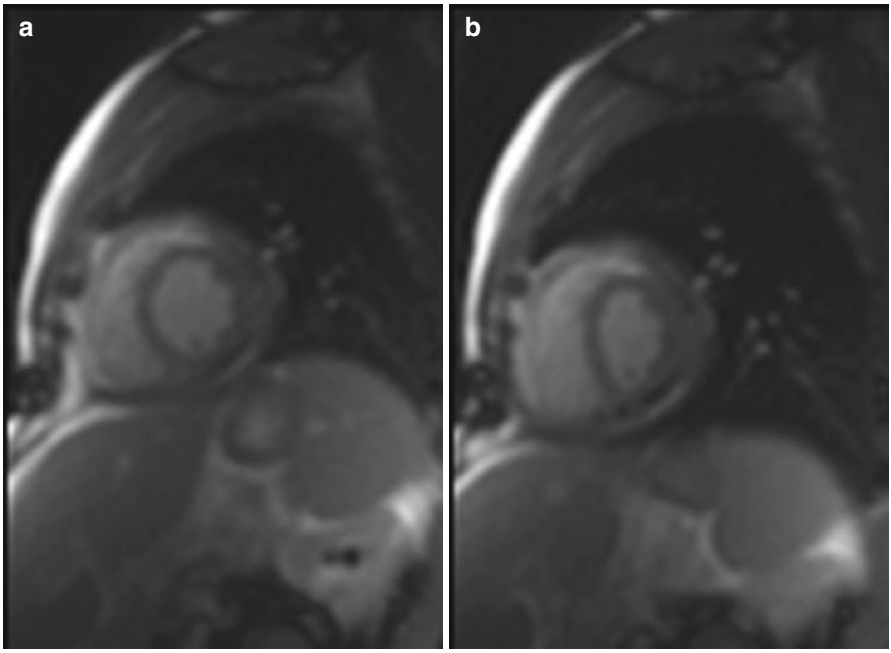

Fig. 26.7 (**a**, **b**) Two diastolic images taken from a real-time dynamic respiratory sequence. During expiration (**a**) the interventricular septal contour is normal, during inspiration there is subtle flattening of the septum indicating abnormal ventricular coupling. This finding is useful for differentiation between restrictive cardiomyopathy and constrictive pericarditis (although in this case is not marked)

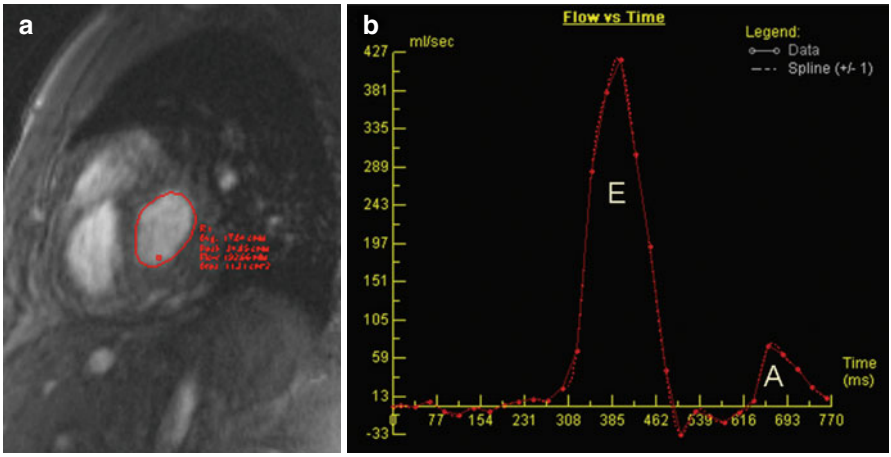

Fig. 26.8 (**a**, **b**) Phase contrast flow mapping of the mitral in-flow shows a pronounced E wave with rapid deceleration time and a small A wave. This indicates a restrictive or constrictive physiology. Respiratory variation would be seen in constriction but CMR does not have the ability to demonstrate this due to the need for breath-hold acquisition or respiratory averaged images

Case 2

This patient demonstrates transient pericardial constriction. The initial CMR clearly demonstrates pericardial thickening and constrictive physiology which resolves on a follow-up scan 3 months later (Figs. 26.9 and 26.10).

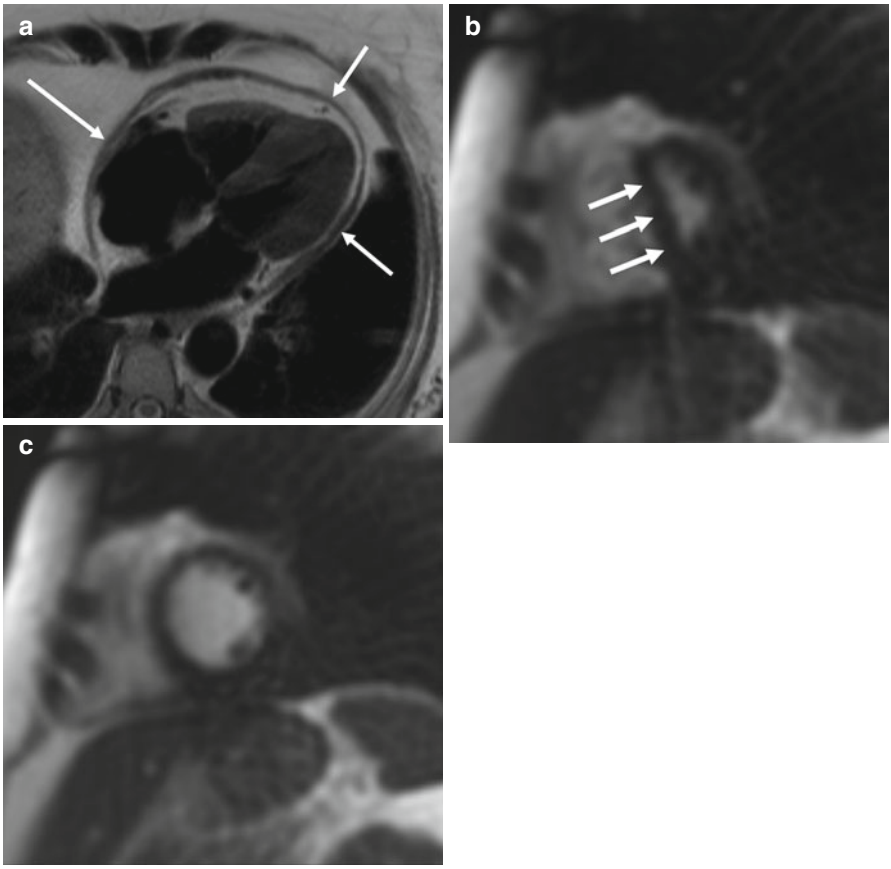

Fig. 26.9 (**a**–**c**) Axial T1 weighted TSE image (**a**) demonstrating pericardial thickening (*white arrows*). Real-time dynamic respiratory sequence shows a diastolic image during inspiration (**b**) and expiration (**c**). The inversion of the interventricular septum during inspiration (*white arrows*) indicates pathological ventricular coupling and a constrictive physiology

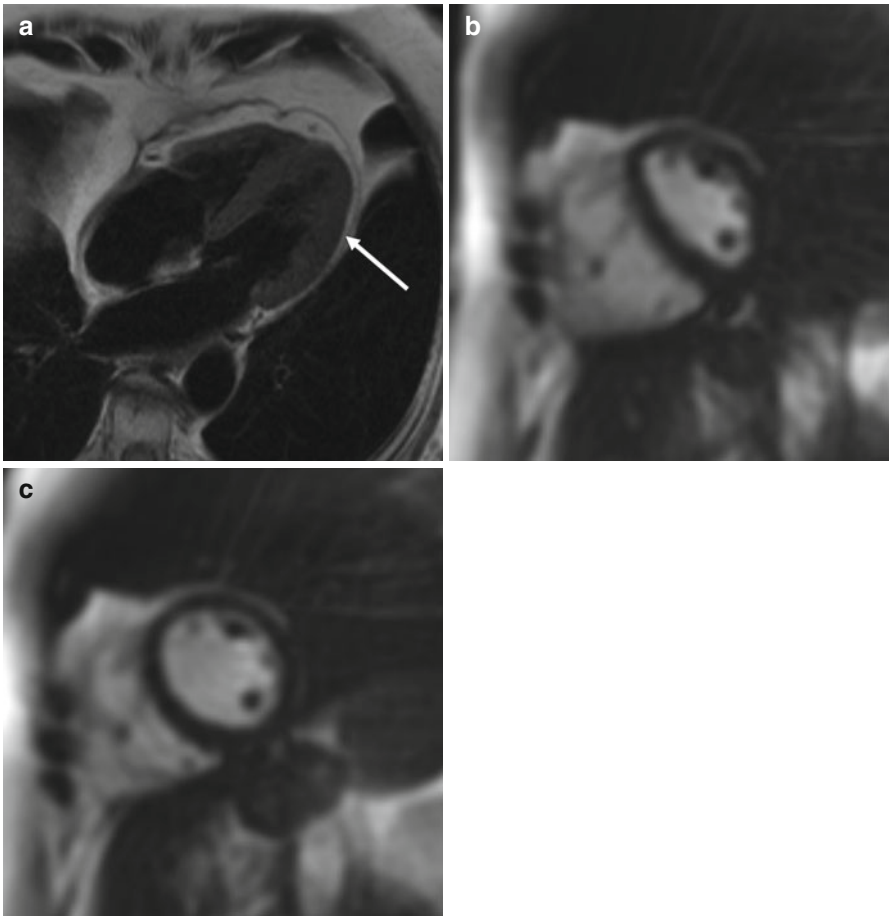

Fig. 26.10 (**a**–**c**) Axial T1 weighted TSE image (**a**) 3 months later shows partial resolution of the pericardial thickening, particularly over the LV (*white arrow*). Real-time dynamic respiratory images (**b**, **c**) now show a normal interventricular septal position confirming physiological recovery

CMR Report in Pericardial Constriction

Morphology (descriptive)

1. Pericardial Thickness: describe as local or circumferential and list thickness measurements
2. Pericardial effusion : presence and extent

Ventricular parameters
1. LV volumes
2. Ventricular wall motion:
   Systolic wall motion
   Septal motion during normal respiration and breath holding.
3. Presence or absence of atrial inversion

Late Gadolinium Enhancement in RV, LV and pericardium

Key Points CMR in Constrictive Pericarditis
1. Identification of thickened pericardium in the appropriate clinical context is diagnostic of constriction
2. CMR gives indirect physiological information that may support the diagnosis
3. Pericardial constriction may be present with a normal pericardial thickness or patchy thickening
4. CMR is complementary to CT, Echo and cardiac catheterisation

Pericardial Tumours

CMR Protocol in Pericardial Tumours
1. Anatomy module including T1 & T2 weighting. Images should cover the whole thorax.
2. LV function module.
3. Myocardial Tagging.
4. First pass perfusion imaging.
5. T1 weighted post contrast images.
6. Late Gadolinium Enhancement module.

Introduction

Pericardial tumours are divided into primary and secondary lesions. The commonest are secondaries with breast cancer, lung cancer or haematological tumours.

Primary pericardial tumours are rare, the commonest being mesothelioma (not associated with asbestos exposure) followed by sarcomas and intrapericardial teratomas. Pericardial tumours tend to cause complex pericardial effusions but may encase the pericardium and result in constrictive physiology. The volume of tumour in pericardial metastasis may be small and not macroscopically visible.

Tumours in the lung and mediastinum may directly invade the pericardium and CMR has a useful role in identifying such invasion and staging thoracic malignancies.

CMR Versus Other Imaging Modalities

Echocardiography remains the initial investigation for most patients with pericardial tumours as the presentation is often that of pericardial effusion. If there is a suspicion of tumour clinically or on other imaging modalities a CMR is likely to be the next imaging modality. Its excellent soft tissue differentiation and large field of view make it an ideal tool to identify pericardial tumour and delineate its origin and extent. CT has a complementary role although is less versatile at dynamic imaging or multiplanar imaging. CT is, however, more robust in identifying other thoracic lesions including lung primaries, pulmonary metastases and mediastinal nodes, all of which may be instrumental in making a definitive diagnosis.

Having identified a pericardial tumour attention turns to delineating its extent and the presence of local invasion. CMR is ideal for this and using the full range of imaging sequences can also provide useful information about the nature of the lesion and its physiological consequences. Coronary angiography may be needed if surgical resection is needed to identify distortion/invasion of the coronaries although CT and CMR will provide adequate information in most cases.

Findings on CMR

The whole of the thorax should be imaged in patients with suspected pericardial tumours to identify primary lesions, other secondaries and mediastinal nodes.

CINE Imaging (b-SSFP)

Standard cine images will identify a pericardial effusion if present and also show areas of thickened pericardium. This alone may be enough to confirm a pericardial mass or pericardial deposits as well as extra-pericardial tumours invading the pericardium.

Most pericardial tumours will show similar signal to the myocardium but greater heterogeneity of the tumour and slight increase in signal will usually allow differentiation. Tethering and/or invasion of the adjacent myocardium or great vessels are generally easily identified and are an indicator of malignancy. Well defined or encapsulated lesions are more likely to be benign.

Cine images provide additional direct or indirect information about the physiological consequences of the tumour (see the sections on "Pericardial effusion" and "Constrictive pericarditis").

Myocardial Tagging

Tagging may help to differentiate tumour (which is non-contractile) from myocardium and subsequently indicate the extent of myocardial invasion.

Black Blood Images (TSE)

Characterisation of tumours is difficult although differentiation of benign and malignant lesions can be indicated by well defined/encapsulated margins versus soft tissue invasion respectively. Lipomas can be positively identified by their fat content on T1/T2 weighted TSE images and fat suppressed sequences. Most soft tissue tumours will have similar signal characteristics to myocardium although increased tissue oedema may be manifest by slight increase in signal on T2 weighting.

First-Pass Perfusion Imaging

Both primary and secondary pericardial tumours will show some degree of enhancement on first pass perfusion imaging. Enhancement on first pass perfusion imaging helps to confirm the presence and extent of the tumour and differentiate it from haematoma or complex effusion.

Post Contrast T1 TSE and Delayed Enhanced Images

As indicated above most soft tissue tumours will display some contrast enhancement and delayed contrast wash-out. T1 TSE images early following contrast may show heterogenous enhancement in the tumour as will delayed inversion recovery sequences. These findings help confirm the presence and extent of tumour but give little information regarding the pathological diagnosis.

The Role of CMR in Pericardial Tumours

1. CMR identifies and delineates focal pericardial masses.
2. CMR demonstrates extracardiac tumours invading the pericardium and is useful in staging mediastinal and thoracic tumours.
3. Excellent soft tissue differentiation allows accurate assessment of extent of pericardial tumours and invasion of adjacent structures.
4. CMR may provide useful tissue characterisation e.g. lipoma.
5. CMR provides additional functional information.

Case Example

A 61-year-old man presented with a 3 week history of shortness of breath, dry cough, chest tightness, and orthopnoea. Having been previously fit he was now breathless on climbing one flight of stairs. Examination primarily showed evidence of right-sided failure with raised JVP and swollen legs. Echocardiogram showed a large pericardial effusion and on pericardiocentesis two litres of blood stained fluid was drained.

The patient was transferred to our institution with the drain in-situ and a repeat echocardiogram suggested a pericardial clot. CMR imaging helped to clarify the anatomical findings. The patient underwent surgical resection of the lesion which on histology was a synovial sarcoma (Figs. 26.11, 26.12, 26.13, 26.14, and 26.15).

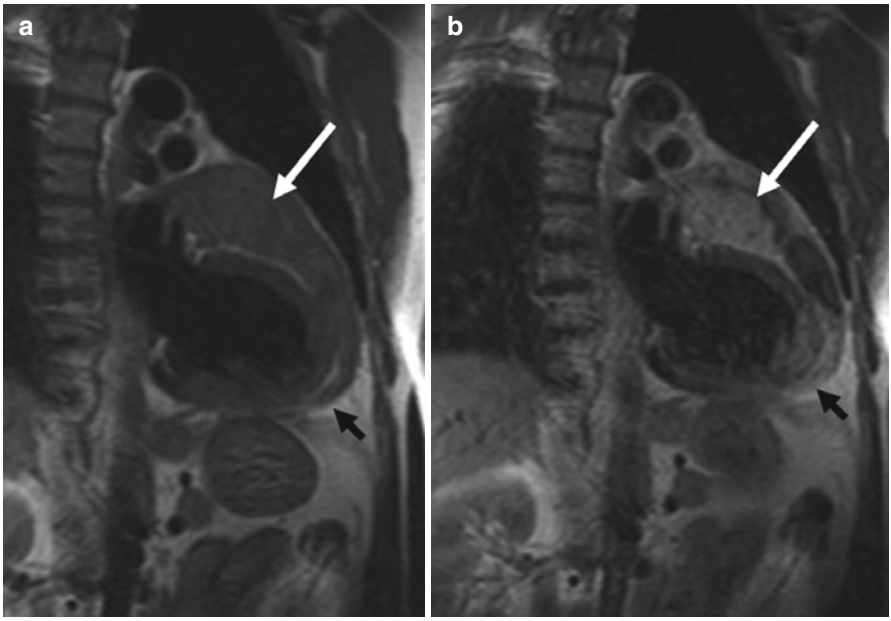

Fig. 26.11 (**a**, **b**) T1 weighted TSE pre and post gadolinium in a 2 chamber view. Pre-contrast (**a**) there is 4×6 cm soft tissue mass (*white arrow*) arising from the pericardium with more diffuse pericardial thickening elsewhere (*black arrow*). The mass appears not to involve the myocardium with preservation of the epicardial fat. Post-contrast (**b**) the mass enhances (*white arrow*) and becomes differentiated from the adjacent pericardial thickening

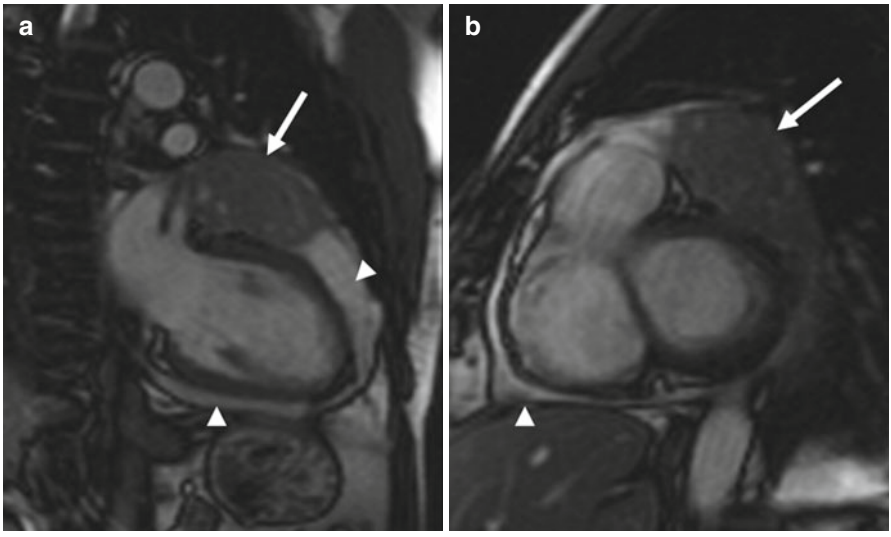

Fig. 26.12 (**a**, **b**) Cine SSFP 2 chamber and short axis views. The soft tissue mass (*white arrows*) seems to arise from the pericardium with normal myocardial signal and motion. The fat around the interventricular groove and LAD is preserved. The pericardial effusion has homogeneous high signal (*arrow heads*)

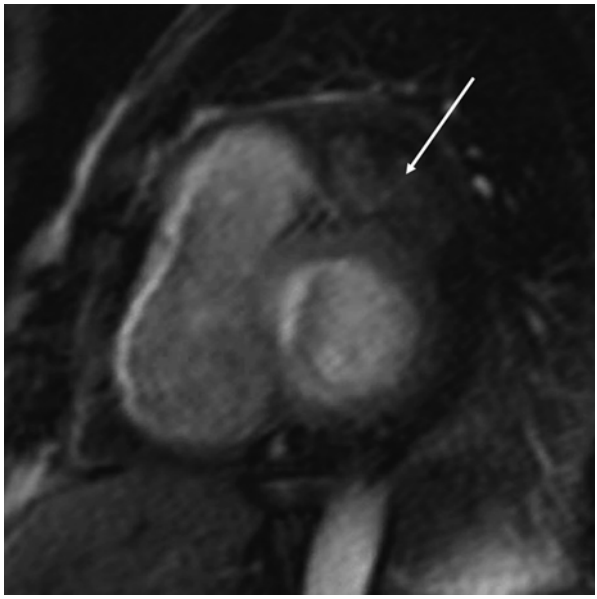

Fig. 26.13 1st pass perfusion study. An arterial phase image from a first-pass perfusion study shows heterogenous enhancement of the mass (*arrow*). This effectively precludes a pericardial haematoma and makes a soft tissue tumour the most likely diagnosis

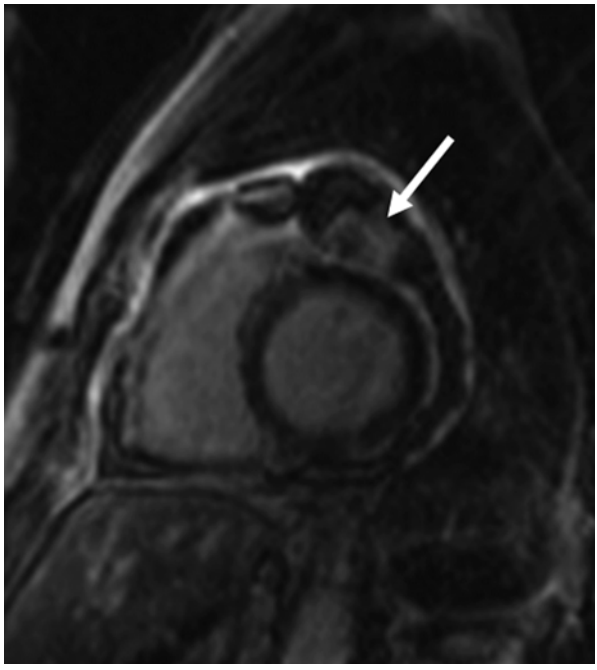

Fig. 26.14 TSE inversion recovery late gadolinium enhanced image. There is patchy high signal within the lesion. The myocardium remains of normal low signal. This is a non-specific finding but again helps to exclude an haematoma

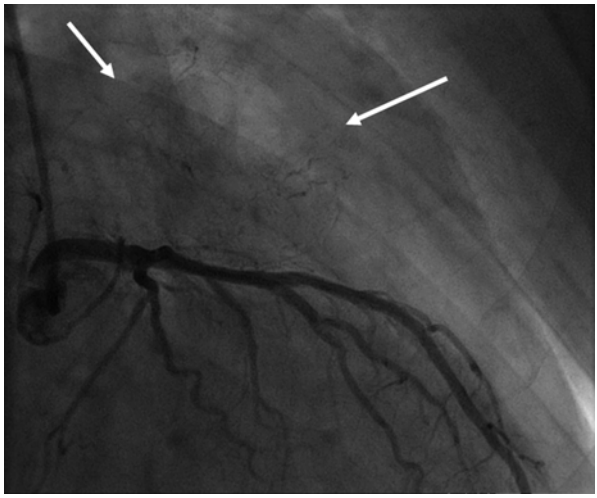

Fig. 26.15 Angiogram. The coronary angiogram shows normal coronaries with capillary filling of the soft tissue lesion (*arrows*)

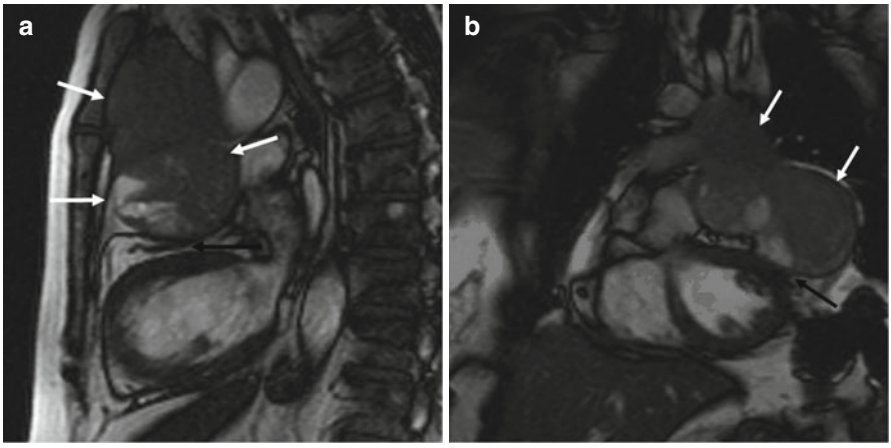

Fig. 26.16 (**a**) Coronal Cine image. The large heterogenous anterior mediastinal mass is well demonstrated (*white arrows*) and appears to invade the pericardium (*black arrows*). Although the epicardial fat is effaced the myocardium is not obviously involved. (**b**) Sagittal Cine image. In this plane the tumour abuts the pericardium and distorts the Left anterior descending coronary artery although does not clearly invade it. There is again no tumour invasion of the myocardium

Case 2

A 35 year old man presented with a large anterior mediastinal mass closely applied to the pericardium. CMR was used to assess myocardial invasion. The tumour was a thymic carcinoma and was resected. As the CMR demonstrates the tumour invaded the pericardium but could be removed from the myocardium and coronary arteries. CMR is a powerful tool for staging mediastinal tumours prior to surgery (Fig. 26.16).

CMR Report in Pericardial Tumours

1. Location (pericardial, myocardial, valve relationship, chamber relationship)
2. Size (cross-sectional dimensions)
3. T1 signal intensity (homogeneous, heterogeneous, hyper, iso or hypo intense to myocardium/or chest wall (specify reference tissue))
4. T1 fat sat images signal intensity (if performed) (homogeneous, heterogeneous, hyper, iso or hypo intense to myocardium/or chest wall (specify reference tissue))
5. T2 signal intensity (homogeneous, heterogeneous, hyper, iso or hypo intense to myocardium/or chest wall (specify reference tissue))
6. STIR signal intensity
7. Perfusion pattern (if perfusion performed)
8. Late gadolinium enhancement pattern on static/delayed images (if gadolinium administered)
9. Relationship to myocardium/pericardium, mediastinum
10. Margins (e.g., smooth, irregular, infiltrating, pediculated)
11. Cine CMR appearance (pedunculated, motion with myocardium/pericardium)
12. Myocardial function (if performed, qualitative or quantitative as appropriate)
13. Pericardial abnormalities if present (pericardial thickness should be reported along with determination of the presence or absence of a pericardial effusion)

Key Points CMR in Pericardial Tumours

1. Useful for identifying primary and secondary pericardial tumours
2. Useful for assessing pericardial involvement from other mediastinal or lung tumours
3. Accurate tumour characterisation is usually not possible and requires histological confirmation

Congenital Abnormalities of the Pericardium

CMR Protocol in Congenital Abnormalities of the Pericardium

1. Anatomy module including T1 & T2 weighting (page X)
2. LV function module (page X)
3. Targeted sequences depending on findings e.g. first pass perfusion, tagging,

Pericardial Cysts and Pericardial Defects

Introduction

Pericardial cysts are caused by embryonic remnants of pericardium which become fluid filled. They generally lie adjacent to the pericardium but may rarely be intrapericardial. If they communicate with the pericardium they are termed diverticulae but usually they are separate. Approximately 70 % are located in the right cardiophrenic angle and 20 % on the left. The majority of patients are asymptomatic and present coincidentally following a chest X-ray with a right paracardiac mass. Occasionally patients present with symptoms from complications such as pressure effects or secondary infection.

Pericardial defects or agenesis are congenital abnormalities that often pass unnoticed. Part of or all of the pericardium fails to develop, possibly as a result of vascular insufficiency during pericardial embryogenesis. One third of cases are associated with other cardiac malformations and the condition is more common in males. Complete agenesis is rare (9 %) and is considered benign as symptoms are uncommon. Partial defects are more common and are generally over the left heart (70 %). They may result in herniation of cardiac structures through the defect with potential strangulation or compromise of the herniated structure e.g. the left atrial appendage. Cases of coronary compromise have been described. Repair of partial pericardial defects has therefore been recommended.

The Role of CMR in Congenital Pericardial Abnormalities

CMR is used to confirm the position and nature of cystic lesions around the heart which have been identified by CXR or Echocardiography. Likewise pericardial defects can be demonstrated on CMR if there is a radiological or clinical suspicion, but are more commonly incidental findings.

Additional physiological information can be gained if there is compression or distortion of an adjacent cardiac chamber or vessel.

Findings on CMR

Pericardial cysts are fluid filled thin walled lesions usually abutting the pericardium. The fluid is a transudate and as such appears simple on MRI with low T1 signal, high T2 signal and high signal on SSFP sequences. Occasionally the cyst will contain proteinaceous fluid and show higher T1 signal. They are generally round and well defined but may conform to surrounding structures as they are under low pressure, indeed they may change shape during respiration. Occasionally they become tense and distort or compress the adjacent intrapericardial structures with significant haemodynamic consequences particularly if the cyst itself is intrapericardial. The main differential is from other paracardiac masses and this is usually straight forward given the simple cystic nature of the lesion and the location.

Pericardial defects are identified by absence of the normal pericardium and a subsequent change in the normal cardiac contour. The heart may shift leftward or specific chambers may herniated through the defect. Likewise

paracardiac structures that are normally separated from the heart may lie in direct contact with heart and create abnormal contours, such as the lung extending into the aorto-pulmonary window. In clinical practice these defects are rarely seen.

The Role of CMR in Congenital Abnormalities of the Pericardium

1. CMR identifies the normal pericardium and pericardial defects.
2. CMR demonstrates and characterises cystic lesions relating to the pericardium.
3. CMR is complementary to other imaging modalities.

Case Example

This patient presented following an abnormal CXR as part of a health screening assessment. He has no symptoms of cardiac disease (Figs. 26.17, 26.18, and 26.19).

Case Example 2

This patient presented following a CT scan of the chest for atypical chest discomfort, the CT showed an unusual cardiac orientation. He had a long history of intermittent breathlessness and chest discomfort that was positional (Figs. 26.20 and 26.21).

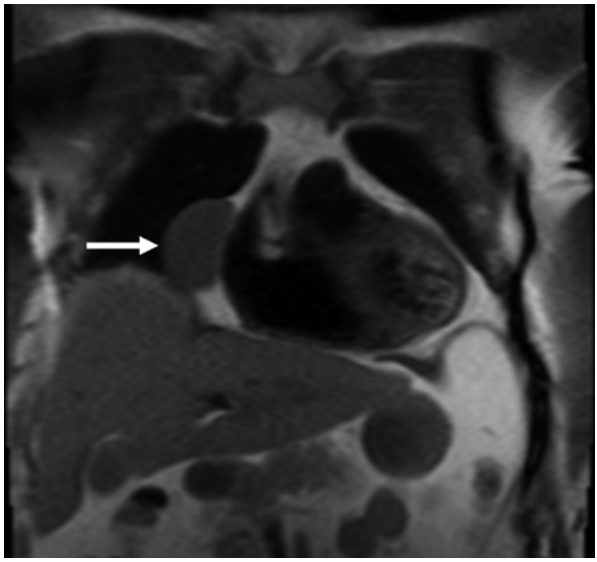

Fig. 26.17 Coronal HASTE image. A pericardial cyst is demonstrated in the typical position (*white arrow*)

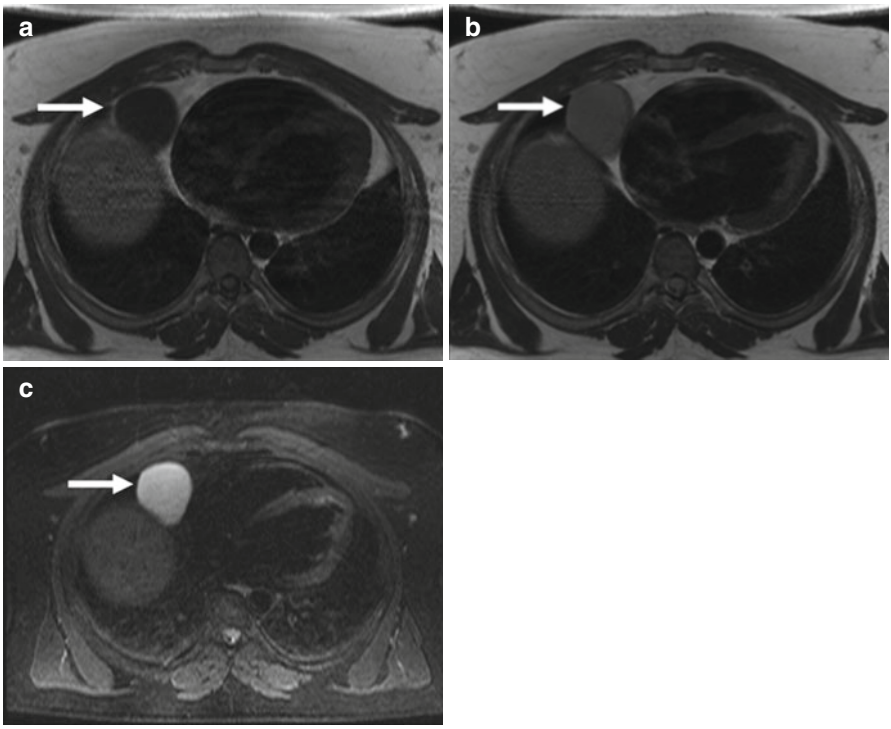

Fig. 26.18 (**a**–**c**) TSE images in an axial plane with T1, T2 and Fat suppressed weighting respectively. The cyst (*white arrow*) is well defined and abuts the pericardium. It is low signal on T1 weighted images, intermediate signal on T2 weighting and high signal on T2 weighted fat suppressed images consistent with a simple transudate

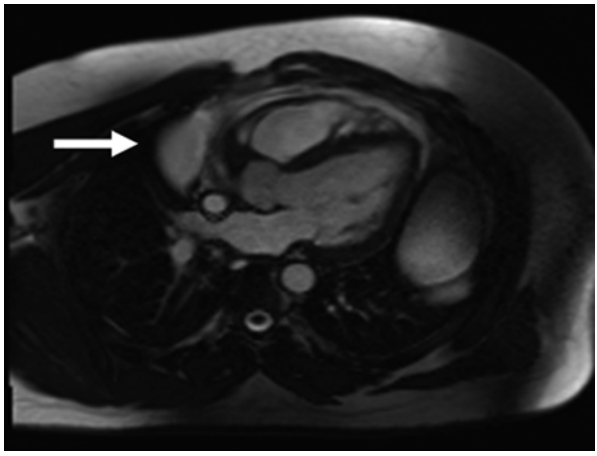

Fig. 26.19 3-chamber cine image (SSFP). The pericardial cyst (*arrow*) shows high signal without septations. There is no distortion of the adjacent cardiac structures

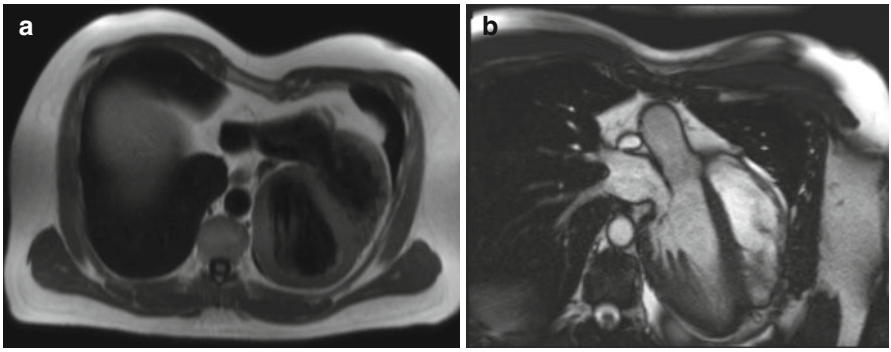

Fig. 26.20 (**a**, **b**) Axial HASTE image and axial cine (SSFP) image respectively. There is complete absence of the pericardium allowing the heart to lie in a dependent position in the left hemithorax with the apex against the posterior chest wall and the left ventricle adjacent to the spine

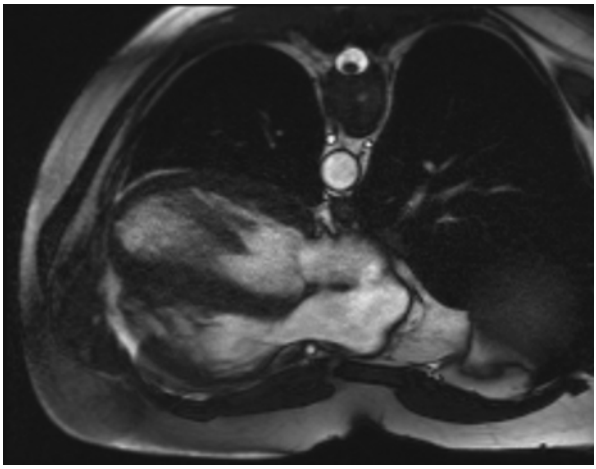

Fig. 26.21 Axial cine image in a prone position. The heart now lies anteriorly against the chest wall demonstrating abnormal mobility within the thorax

Tips and Tricks

1. Imaging with the patient in supine and prone positions may help highlight absence of the pericardium.

CMR Report in Congenital Pericardial Abnormalities

1. Morphology of the abnormality
2. Appearance on different CMR image types
3. Associated pathologies

Key Points: CMR in Congenital Pericardial Abnormalities

1. Used to confirm the typical cystic nature and location of pericardial cysts.
2. Excludes other causes of paracardiac mass lesions.
3. Identifies congenital pericardial absence or pericardial defect.
4. Demonstrates any functional consequence of pericardial abnormality.