Cardiovascular Magnetic Resonance Imaging

Textbook and Atlas

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HOCM patients LGE is most commonly located in the center of the hypertrophied septum. To identify high-risk individuals for SCD who are candidates for an ICD, risk factors including family history of SCD, syncope, nonsustained VT, abnormal blood pressure response to exercise, and severe LV hypertrophy have been proposed (Maron 2003). The presence of LGE seems to add new prognostic information, since it has been reported to be associated with progressive ventricular dilatation and markers of SCD (Moon 2003), with higher NYHA classes and prevalence of VT, impaired global LV function and asymmetrical hypertrophy, abnormal Q-waves, and giant negative T-waves (Satoh 2009). It has also been associated with greater likelihood and increased frequency of ventricular tachyarrhythmias, including nonsustained VT on ambulatory Holter ECG (Adabag 2008), and a specially designed LGE score proved to be a significant multi-variable predictor of both clinical VT/VF and of maximal LV wall thickness as markers of high-risk HCM patients (Leonardi 2009). In a mathematical model the 5-yrs probability of SCD in HCM has been calculated to be 0.11 in patients compared with 0.07 in patients without LGE, i. e. the approximate 5-yrs probability of SCD is 1.6-fold higher if scarring is noted on CMR of a patient with HCM (Nazarian and Lima 2008). A recent study (Bruder 2010) showed that in a cohort of 243 patients with largely low or asymptomatic HCM the presence of LGE indicating myocardial scar is a good independent predictor of all-cause and cardiac mortality.

CMR for Screening and Differential Diagnosis

Since first-degree relatives of affected HCM individuals have a 50% chance of being a gene carrier (Maron 2002), determining the presence of HCM in family members is essential and may be achieved most accurately by CMR. Disease expression in HCM patients with certain gene mutations may be very heterogenous, therefore precise phenotyping by CMR with demonstration of LVH is mandatory, providing incremental information to conventional ECG and echocardiographic screening. Particularly in young patients, repeat CMR examinations may be needed every 2–5 yrs for the early observation of disease progression with development of further LVH, ECG abnormalities, and later focal fibrosis (LGE). CMR is also important for differential diagnosis in patients with dyspnea on exertion and abnormal ECG, in whom, apart from HCM, LVH may be induced by underlying arterial hypertension, aortic stenosis, athlete's heart, amyloidosis, or metabolic abnormalities such as Fabry's disease. CMR criteria such as type, extent, and location of LVH, systolic and diastolic function, valvular function, and type, size, and location of LGE may aid differential diagnosis.

10.5 Restrictive Cardiomyopathy

Prevalence, Etiology and Pathology

The hallmark of restrictive cardiomyopathies is abnormal diastolic function due to increased stiffness of the ventricular walls, leading to heart failure. Idiopathic RCM strictly defined to include normal ventricular wall motion, normal wall thickness, and ventricular chamber dimension proved to be relatively rare, and has been observed in single families with sarcomere gene mutations. Approximately 50% of cases with RCM result from specific clinical disorders such as infiltrative (amyloidosis, sarcoidosis, Gaucher disease, Hurler disease, fatty infiltration), storage diseases (hemochromatosis, Fabry's disease, glycogen storage disease), and endomyocardial disorders (endomyocardial fibrosis, hypereosinophilic syndrome, carcinoid heart disease, metastatic cancers, and drugs causing fibrous endocarditis like serotonine, methysergide, ergotamine, mercurial agents, and busulfan). The remaining 50% of patients present with idiopathic RCM.

Clinical Presentation, Diagnosis and Therapy

In early stages of idiopathic RCM (IRC), systolic function may be normal together with impaired filling characteristics, although deterioration in systolic function is usually observed as the disease progresses. Clinically the patients frequently present with exercise intolerance that results from impaired ability to increase cardiac output during increasing heart rate due to the restriction of diastolic filling. Further common **symptoms** are dyspnea and/or chest pain on exertion, weakness, and edema. With advanced disease, profound peripheral edema with hepatomegaly, ascites, and later on anasarca may occur. Many patients report palpitations that are produced by atrial fibrillation. **Diagnosis** is made by history, clinical examination (positive Kussmaul sign, rales, S3–S4 gallop), chest X-ray, and echocardiography, including tissue Doppler (normal systolic function combined with diastolic dysfunction: myocardial relaxation with increased early left ventricular filling velocity, and decreased isovolumetric relaxation time). CMR imaging is a very valuable tool for determining structure, systolic and diastolic function, and tissue characteristics, and to aid in differentiating idiopathic from secondary forms of RCM. The most important differential diagnosis of IRC is constrictive pericarditis, which is also characterized by normal or nearly normal systolic function, but abnormal ventricular filling. Points frequently helpful in favoring constrictive pericarditis over restrictive cardiomyopathy are active pericarditis from history, paradoxical pulse on clinical examination, absence of intraventricular conduction defect within the ECG, calcification on chest X-ray, thickened pericardium

on CT/MR imaging, septal notch in the echocardiogram, respiratory variations indicating increased ventricular interdependence on Doppler, close equilibration of diastolic pressures, and absence of amyloid or other infiltrative disease on endomyocardial biopsy (Hanckock 2001). **Treatment** refers to classical drug therapy of heart failure.

CMR Findings

CMR imaging allows precise assessment of atrial and ventricular volumes, systolic and diastolic function, as well as of associated valvular insufficiencies, and provides parameters to differentiate RCM from constrictive pericarditis (CP). In CP a leftward inversion or flattening of the interventricular septum during early ventricular filling is seen, which is not present in patients with RCM. The amount of ventricular coupling can be further evaluated by quantifying the difference in the maximal interventricular septal excursion between inspiration and expiration. This parameter normalized to the end-diastolic biventricular dimension has been shown to be significantly greater with CP (about 20 %) than with RCM, where a cut-off value of 11.8 % enables such a differentiation (Francone 2006). CMR imaging also provides a detailed description of the morphology of the pericardium in CP vs. RCM (in CP irregular thickening, signal void due to calcifications), an assessment of pericardial motion patterns with cine-SSFP sequences (in CP abolished motion), as well as the impact on ventricular filling by PC flow analysis, i. e. mitral and pulmonary vein inflow patterns.

10.6 Arrhythmogenic Right Ventricular Cardiomyopathy or Dysplasia (ARVD)

Prevalence, Etiology and Pathology

ARVD is a progressive cardiomyopathy primarily affecting the right ventricle (RV) and is characterized by fibro-fatty replacement with myocyte loss, right heart failure, and ventricular (tachy-) arrhythmias with left bundle branch block (LBBB) pattern. Increasing evidence suggests that ARVD is a disease of desmosomal dysfunction. Its prevalence is estimated to be 1 : 5,000 in the US and 1 : 10,000 in Europe up to 1 : 1,000 in the region of Venezia, and it can exist in both sporadic and familial form with a predominantly autosomal dominant inheritance pattern with variable penetrance. ARVD accounts for 5–20 % of SCD in young individuals less than 35 yrs, and most SCDs occur during strenuous exercise. There is a 6-fold male predominance over females. The estimated death rate per year is 2.5 % (Jain 2008). Morphological features include myocyte loss with fibro-fatty replacement of the ventricular myocardium, focal thinning of the RV wall in diastole, RVOT enlargement, RV dilatation, RV aneurysms and trabecular disarray, and hypertophy. Functionally global or regional contraction abnormalities and RV systolic and/or diastolic dysfunction may be found at least in later stages of the disease.

ARVD is considered a disease of desmosomal dysfunction due to mutations affecting components of the cardiac desmosome, with consecutive disruption of cell-cell adhesion, provoking detachment of myocytes particularly under conditions of mechanical stress, leading in turn to apoptosis and cell necrosis, secondary inflammation, and repair by fatty substitution.

Clinical Presentation, Diagnosis and Therapy

This generally considered progressive disease may present with **four different clinical phases**:

- a concealed phase (subtle RV structural changes, minor ventricular arrhythmias, rarely sudden cardiac death during intense sports or physical exercise in very young individuals),
- an overt clinical disorder (symptomatic RV arrhythmias including VTs associated with overt RV structural and functional abnormalities),
- RV failure due to disease progression, and finally
- biventricular pump failure from significant LV involvement.

Patients most commonly present with ventricular tachycardias (VTs) with LBBB configuration at stage two. **Symptoms** of right-sided heart failure are rare at least in the early stages of disease. **Diagnosis** is confirmed by a set of six major and minor criteria (McKenna 1994) including global and/or regional RV dysfunction and structural abnormalities, tissue characterization of the RV wall, repolarization and depolarization/conduction abnormalities, arrhythmias, and family history (Table 10-1). Patients must have either two major criteria, one major and two minor criteria, or four minor criteria to be labeled as affected with ARVD (Jain 2008). Patients should be **treated** with ACE-inhibitors and β-blockers, and those with high risk or proven ventricular tachyarrhythmias should receive an ICD in order to reduce the major cause of mortality in affected individuals. In patients progressing to overt heart failure, management involves the same principles for the treatment of other forms of cardiomyopathy e. g. DCM. Prognosis of patients with ARVD is moderate, Hulot et al. (2004) reported 21 deaths with an annual death rate of 2.3 % in a cohort of 130 patients with ARVD over a follow-up period of 8.1 ± 7.8 yrs, and death from heart failure occurred in 59 % of patients compared with 29 % SCDs. In a cohort of 61 patients with ARVD, in multivariate analysis Lemola et al.

Table 10-1 Major and minor criteria for the diagnosis of ARVD (modified from Mc Kenna et al. 1994; the CMR criteria have been added to the original concept).

Criterion	Major	Minor
global/regional dysfunction, structural abnormalities of RV	severe RV dilatation, reduced RV-EF, minimal involvement of LV, severe segmental RV dilatation, regional RV aneurysms	mild global RV dilatation, mildly reduced RV-EF, mild segmental RV dilatation, regional RV hypokinesis
tissue characteristics of ventricular myocardium	fibro-fatty degeneration at endocardial biopsy	none
CMR criteria	severe global or segmental RV dilatation, global systolic and diastolic RV dysfunction, severe trabecular disarray of RV, localized RV aneurysms or excavations, presence of LGE in RV wall	intramyocardial fatty infiltration, focal wall thinning of RV, regional wall hypertrophy of RV, moderator band hypertrophy, RVOT enlargement
abnormalities in depolarization and AV conduction	epsilon-wave of the QRS complex in V1 to V3, localized QRS prolongation ≥ 110 ms in V1 to V3	ventricular late potentials in the signal averaged surface ECG
abnormalities in repolarization	none	inverted T-waves in V2 and V3 in patients older than 12 yrs, absent RBBB
cardiac arrhythmias	none	LBBB type of ventricular tachycardias (non-sustained or sustained), frequent ventricular ectopic beats (≥ 1,000/24 h)
family history	familial disease of ARVD confirmed by autopsy or surgery	sudden death victims in the family (victims < 35 yrs) due to suspected ARVD, positive family history by the appropriate criteria of ARVD

(2004) found a history of congestive heart failure and presence of LV involvement as independent predictors of an adverse outcome during a follow-up period of 55 ± 47 mths, with a total death rate of 10 patients (16%).

CMR Findings

CMR abnormalities in ARVD may be divided into structural abnormalities (intramyocardial fatty infiltration, focal wall thinning, regional wall hypertrophy, trabecular disarray and hypertrophy, moderator band hypertrophy, RVOT enlargement), and into functional abnormalities (regional wall motion abnormalities, focal aneurysms and microaneurysms or excavations, RV dilatation, RV diastolic and systolic dysfunction) (Figure 10-8). Of these severe global or segmental dilatation of the RV, global systolic and diastolic dysfunction, and localized aneurysms are considered major CMR criteria, while mild global or regional dilatation of the RV, regional contraction abnormalities, and global diastolic dysfunction are considered minor criteria according to the Task Force (McKenna 1994). Lesions in the RV wall such as fibro-fatty replacement or late-stage fibrosis may be detected by gadolinium-enhanced CMR (LGE). Tandri et al. (2005) found 12 of 30 consecutive patients prospectively evaluated for ARVD to meet the criteria of the Task Force, and 8 of these 10 showed LGE compared with none of 18 patients not meeting the Task Force criteria for ARVD. These LGE areas correlated well with histological findings and with inducibility of VT during electrophysiologic testing. Sen-Chowdrhy et al. (2006) performed comprehensive CMR studies in 232 patients undergoing evaluation of suspected ARVD, and CMR studies were positive in all 64 patients who fulfilled the Task Force criteria (100% sensitivity), while specificity was low (29%) for the whole group. In the group of 26 genotyped individuals (gene-positive) sensitivity of CMR was 96%, specificity 78%, PPV 93% and NPV 88%. Among other diagnostic criteria such as LV or RV dilatation, abnormal trabeculation, and severe RV segmental dilatation/wall motion abnormalities/aneurysms, LGE in the LV had a sensitivity of 100% and specificity of 67%, compared with LGE in the RV with a sensitivity of 59% and a specificity of 100%. The conclusion of this large study on 64 ARVD patients was that CMR including LGE is a valuable component of the diagnostic workup for ARVD when performed with a dedicated protocol by specialists in CMR imaging. In summary, according to the literature and our own experiences severe global or segmental dilatation of RV, global systolic and diastolic RV dysfunction, severe RV trabecular disarray, localized aneurysms, and the presence of LGE within the RV wall should be considered major criteria for ARVD, compared with the other features of CMR imaging considered as minor criteria.

LV involvement in ARVD has been described in 16–76% of cases with subnormal LV ejection fraction, regional wall motion abnormalities, and evolution of LV involvement has

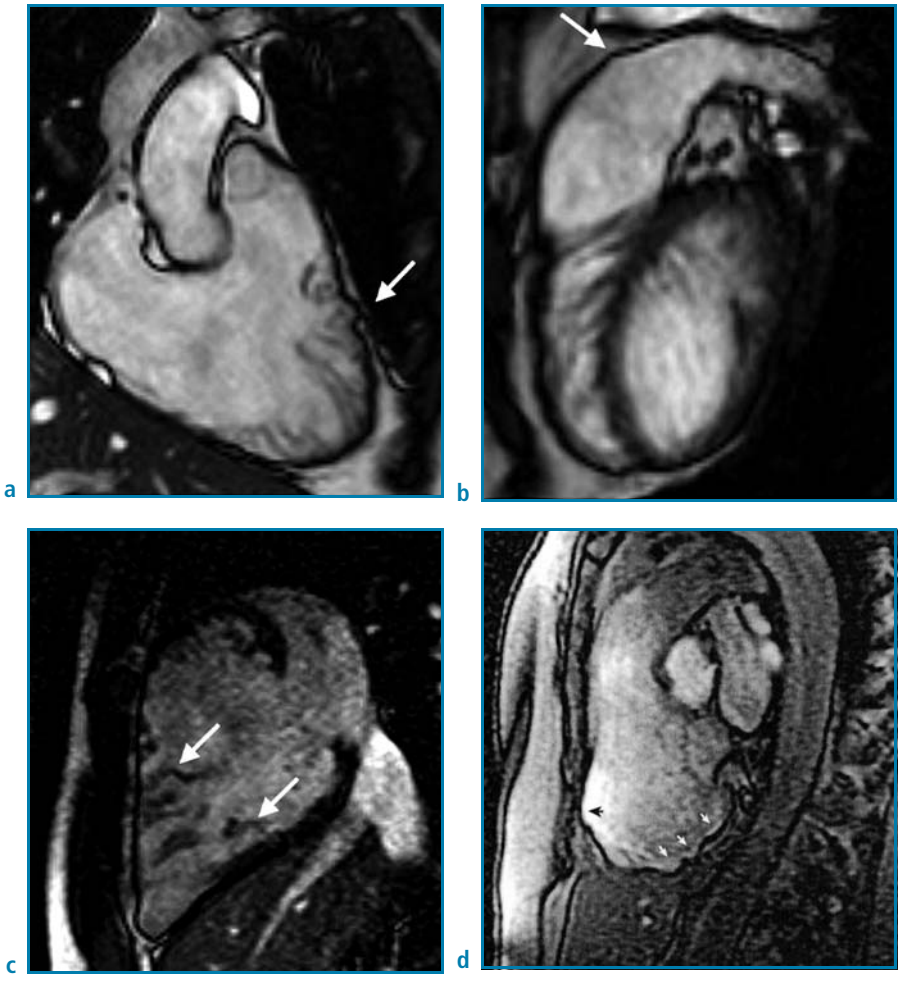

Figure 10-8 Different patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy with circumscribed aneurysm in the anterior right ventricular (RV) wall (arrow) and trabecular disarray (a), circumscribed aneurysm in the RVOT (arrow in (b)), severe trabecular disarray (arrows in (c)), and multiple small aneurysms/excavations at the basal and anterior part of RV musculature (small arrows in (d)) (Figure d by courtesy of Dr. J. Schwab, Nuremberg).

been demonstrated in terms of appearance of new abnormalities or worsening of existing abnormalities or both, with progression of RV disease on long-term follow-up (Jain 2008). The most common variant of ARVD is considered a biventricular form with LV involvement from its early stages, and LV involvement is considered to be part of a larger disease process extending across the entire heart, since ARVD is considered a desmosomal disease and desmosomal genes are expressed in both ventricles. Fibro-fatty changes of the LV located in the subepicardial or mid-ventricular areas can extend across varying thickness of myocardial wall. They can affect both the septum and LV free-wall with a predilection for the posteroseptal and posterolateral areas. CMR imaging provides precise information on LV dilatation and regional wall motion abnormalities including aneurysms, mostly located at the LV apex, by cine-SSFP sequences, and fibro-fatty areas by delayed-enhancement on gadolinium administration. LV involvement is associated with increased myocardial mass, inflammatory infiltrates (i.e. LGE in CMR), clinical arrhythmic events, and more severe RV wall thinning and heart failure, but frank left-sided heart failure is unusual (Jain 2008).

10.7 Tako-Tsubo Cardiomyopathy

Prevalence and Pathogenesis

Tako-Tsubo cardiomyopathy or apical ballooning syndrome (ABS) is a novel acute coronary syndrome that mimics acute myocardial infarction. Its prevalence is very low and was first described by Japanese colleagues, but is now detected worldwide. It is most commonly observed in women secondary to heavy emotional or physiological stress. Its pathogenesis is unclear, severe coronary artery spasms are suspected to produce the transient and reversible severe transmural ischemia within the apical region of the left ventricle.

Clinical Presentation, Diagnosis and Therapy

Patients present with the clinical syndrome of acute myocardial infarction with severe acute chest pain, ST segment elevation, and/or elevated cardiac markers (troponin T/I). Typically these patients immediately undergo coronary angiography and show normal coronary arteries together with a typical apical ballooning, i.e. akinesis of the apex up

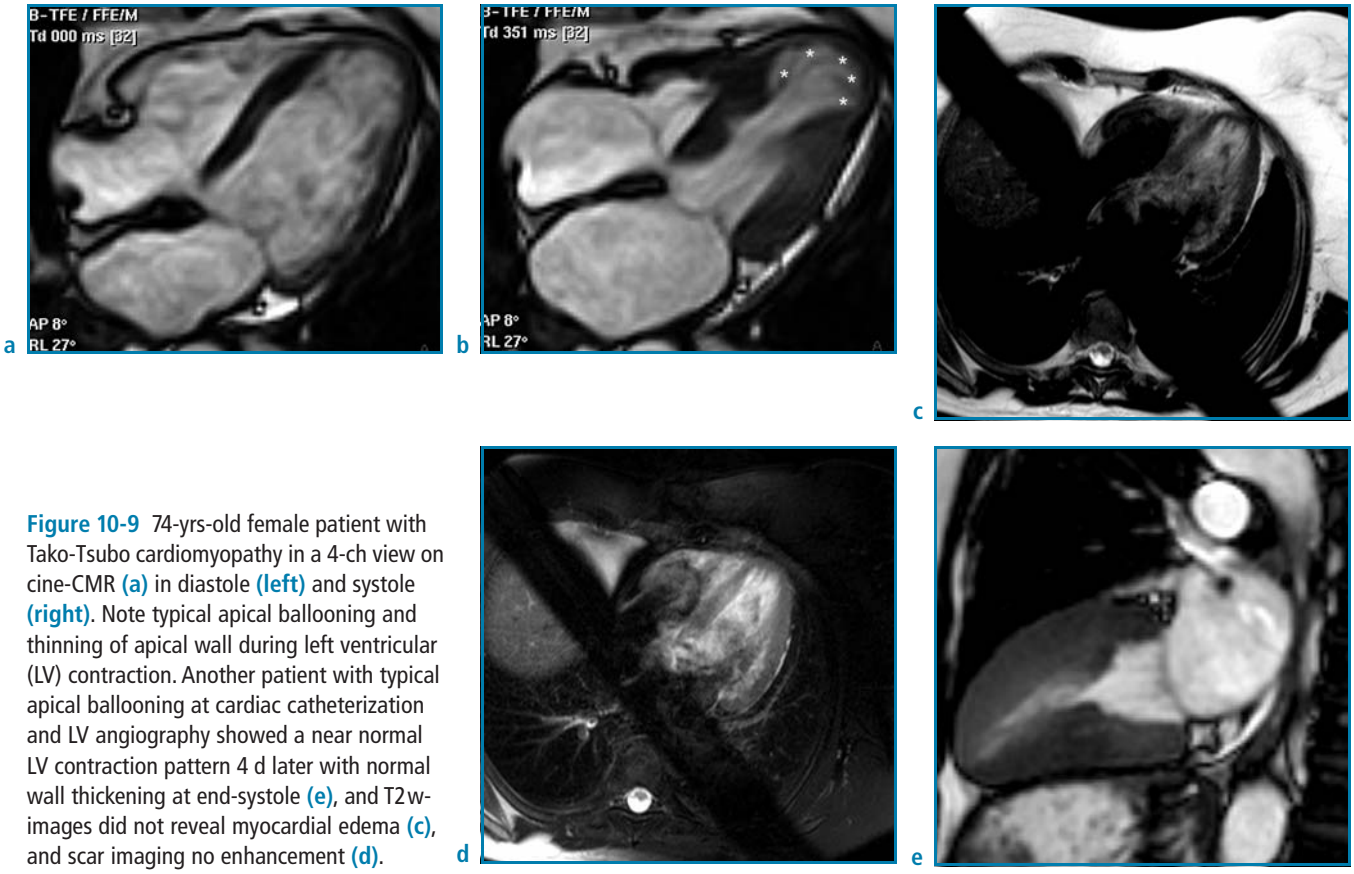

Figure 10-9 74-yrs-old female patient with Tako-Tsubo cardiomyopathy in a 4-ch view on cine-CMR **(a)** in diastole **(left)** and systole **(right)**. Note typical apical ballooning and thinning of apical wall during left ventricular (LV) contraction. Another patient with typical apical ballooning at cardiac catheterization and LV angiography showed a near normal LV contraction pattern 4 d later with normal wall thickening at end-systole **(e)**, and T2w-images did not reveal myocardial edema **(c)**, and scar imaging no enhancement **(d)**.

to the mid-ventricular portion of the left ventricle on left ventricular angiography or echocardiography. Typically wall motion abnormalities disappear within days or weeks on regular treatment with β-blockers and ACE-inhibitors.

CMR Findings

Cine-SSFP imaging shows the typical apical ballooning, and in some patients myocardial edema may be present on T2-weighted imaging (Joshi 2009). In most reports LGE has not been found in ABS (Haghi 2007, Mitchell 2007, Hombach 2008, Koeth 2008, Syed 2008). Myocardial edema disappears and LV dysfunction returns to normal or near normal on follow-up CMR imaging, typically after 3–6 mths a completely normal CMR scan may be present (Figure 10-9).

10.8 Cardiac Non-Compaction (NC)

Prevalence, Etiology and Pathogenesis

This specific (unclassified) cardiomyopathy develops due to a pathological arrest of the compaction process of the loose network of fibers with sinusoids, which are in continuity with the ventricular cavity present during the embryologic period up to the 5th week of intrauterine life. This arrest of compaction leads to the persistence of ventricular hypertrabeculation, which is called spongy myocardium or left ventricular non-compaction (NC). The childhood form of NC was first described in association with other congenital or neuromuscular abnormalities, but NC may be observed in adults without further congenital anomalies. Although NC has generally been regarded as a familial cardiomyopathy, a family history is not always present in adults or children. The prevalence of NC has been estimated at 0.05 % in the general population (Ritter 1997). The finding of a ratio of > 2.0 between the thickness of the non-compacted and compacted myocardial layers in systole is considered diagnostic at echocardiographic examination, though using CMR due to its high resolution properties a ratio of > 3.0 has been reported (Fazio 2008).

Clinical Presentation, Natural History and Therapy

Patients may be **asymptomatic** for a longer period, but later on present with dyspnea on exertion, chest pain, palpitations, systemic emboli including stroke, and symptoms of overt heart failure. In a study by Yousef et al. (2009) on 42 patients with non-compaction, the distribution of symptoms was dyspnea in 50 %, chest pain in 19 %, palpitations in

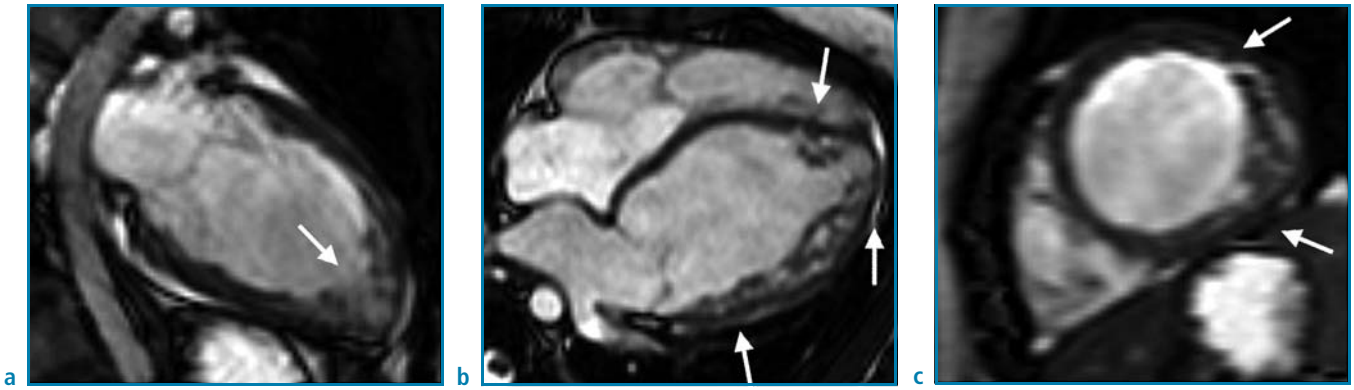

Figure 10-10 13-yrs-old patient with cardiac non-compaction cardiomyopathy and only slight impairment of physical fitness. Cine-CMR images in the 2-ch (a), 4-ch (b) and short-axis view (c) show the typical spongy myocardium with apical (arrows in (a) and (b)) and lateral wall recessus (arrows in (c)). Left ventricular ejection fraction was 50 % using CMR volumetry.

14 %, and stroke in 5 % of cases. Diagnosis is made by echocardiography, using a ratio of > 2.0 of the non-compacted (NC) and compacted (C) myocardium as diagnostic, though echocardiography poses inherent problems in assessing the LV apex, known to be the most commonly non-compacted area. Therefore, **diagnosis** should be confirmed by CMR imaging using an NC/C ratio of > 2.3 (Petersen 2005) or > 3.0 (Fazio 2008), and by demonstrating the typical non-compacted myocardium at LV apex, anterior and lateral LV wall with open recessus at diastole and obliteration during systole. Patients with overt heart failure are **treated** with ACE-inhibitors, diuretics, and β-blockers. Some patients at high risk of life-threatening ventricular tachyarrhythmias need an ICD implantation, and some may finally undergo heart transplantation. Prognosis of patients with NC is poor. In a study by Oechslin et al. (2000) on 34 patients after a follow-up of 44 ± 40 mths, major complications such as heart failure (18 patients = 53 %), thromboembolic events (8 patients = 24 %), and ventricular tachycardias (14 patients = 41 %) occurred, and 12 patients died (8 suddenly, 4 from end-stage heart failure, and 2 from other causes). 4 patients underwent heart transplantation, and 4 patients received an ICD. Therefore, patients with NC should be carefully observed in shorter time intervals, using CMR imaging as the gold-standard for assessing changes in LV volumes and function over time, and to detect developing thrombi within the non-compacted myocardium prior to systemic embolization.

CMR Findings

In the study by Petersen et al. (2005) on 7 patients, non-compacted myocardial segments were most commonly found at the LV apex (91 % of subjects), followed by mid-cavity levels (78 %), and the basal segments (21 %), and NC was most common in the anterior segment, becoming less frequent in successive segments in a clockwise direction. Yousef et al. (2009) found a slightly different distribution of NC segments with a more or less equal frequency at the apex, apical-inferior, apical-septum, and mid-septal segments between 35 and 40 %. The second CMR criterion to confirm the diagnosis is an NC/C ratio of > 2.3, and the demonstration of open recessus at diastole and obliteration during systole (Figure 10-10). In addition, in NC patients presenting with dyspnea LV volumes may be greater and LV ejection fraction lower than in age-matched healthy controls (Yousef 2009). CMR imaging is the ideal tool to follow NC patients for changes in the myocardial structural and functional status.

10.9 Secondary Types of Restrictive Cardiomyopathy

10.9.1 Amyloidosis

Etiology and Pathogenesis

Infiltrative types of RCM include amyloidosis, sarcoidosis, Gaucher disease, Hurler disease, and fatty infiltration of the myocardium. Amyloidosis results from tissue deposition of special proteins (twisted beta-pleated sheet fibrils) that may be found in almost all organs, but only produce symptoms when tissue infiltration is extensive. **Primary amyloidosis** results from the deposition of portions of immunoglobulin light chain (AL) that result in most cases from a monoclonal expansion of plasma cells in multiple myeloma or Morbus Waldenström. **Secondary amyloidosis** (reactive amyloidosis) results from the excess production of non-immunoglobulin protein known as AA in the setting of rheumatoid arthritis, Morbus Crohn, chronic osteomyelitis,

or mucoviscidosis. In rare cases various familial diseases may lead to amyloidosis, most commonly induced by the production of a variant form of prealbumin termed transthyretin. Cardiac amyloidosis is a progressive infiltrative cardiomyopathy that carries a grave prognosis; it shows male predominance, and is rare before age 40. About 25 % of patients with familial (transthyretin-induced) amyloidosis experience clinically significant cardiac involvement, and neurological and renal involvement may also predominate, patients typically present with clinical symptoms after age 35. The amyloid infiltrated heart appears tan and waxy on pathological examination, and is rubbery in consistency. Amyloid deposits may be found within the ventricular and atrial myocardium between the myocytes, atria may be enlarged, and focal thickening of cardiac valves due to amyloid deposition may be present; also amyloid may deposit in the media and adventitia of intramural coronary arteries and may cause impairment of coronary perfusion (Hare 2012).

Clinical Presentation, Diagnosis and Therapy

Most patients with cardiac amyloidosis present with **symptoms** of congestive heart failure, i. e. dyspnea on exertion, reduced exercise capacity, edema etc., blood pressure may be normal or reduced, and pulse pressure may be quite narrow as a result of a low cardiac output. The ECG often reveals low QRS voltage, bundle branch block, atrioventricular blocks of varying degree, and atrial fibrillation with amyloid infiltration of the atria. **Diagnosis** is made by echocardiography revealing increased ventricular wall thickness with small intracavitary chambers, enlarged atria, and a thickened interatrial septum. Systolic function may be normal, but Doppler examination may show a restrictive filling pattern indicating diastolic dysfunction. Diagnosis can be confirmed by endomyocardial biopsy and histologic examination using Sirius red or Congo red staining for amyloid deposition, and immunohistochemistry for identification of specific amyloid proteins. **Therapeutic intervention** includes classic heart failure treatment (digitalis particularly for rate control in atrial fibrillation, diuretics, β-blockers, ACE-inhibitors), pacemaker implantation in AV block, or ICD implantation in severely depressed systolic function, and in AL amyloidosis chemotherapy with alkylating agents alone, or in combination with autologous bone marrow transplantation with a 30–39 % survival rate after 4–5 yrs. The prognosis is rather poor (for details see Hare 2012).

CMR Findings

CMR imaging may reveal the typical homogenous increased thickness of atrial and ventricular walls and the interatrial septum, together with a normal or reduced size (Figure 10-11). Occasionally thickening of papillary muscles or valve leaflets may be seen with CMR. The atria are usually enlarged and pleural and pericardial effusions are commonly observed. Systolic function can be exactly quantified, and diastolic dysfunction can be assessed by PC diastolic mitral and pulmonary vein flow patterns showing a restrictive filling pattern. LGE may also be present and aid in confirming the diagnosis by its type, size and location (Maceira 2005, 2008, Perugini 2005).

T1 and T2 mapping has been tested for confirming amyloid deposition within the myocardium. In the study of Maceira et al. (2005) subendocardial T1 in 30 patients with cardiac amyloidosis was significantly shorter than subepicardial T1, and was correlated with markers of increased myocardial amyloid load such as LV mass, wall thickness, interatrial septal thickness, and diastolic function. In contrast, Sparrow et al. (2009) could not find any difference in T2 relaxation times between the 49 patients with histologically confirmed cardiac amyloidosis and normal controls. LGE has also been found in both studies, in the study of

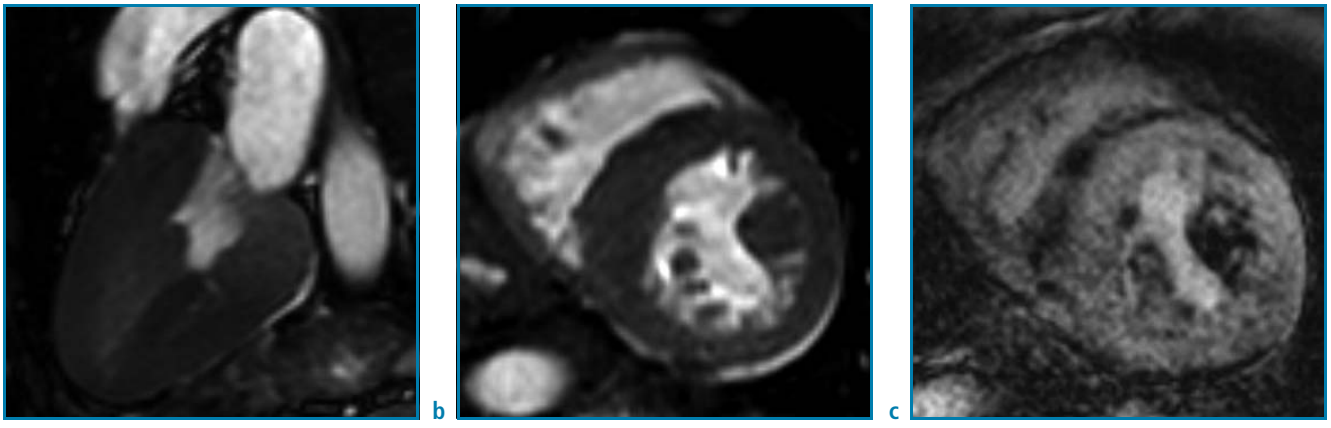

Figure 10-11 32-yrs-old patient with dyspnea on exertion and NYHA class II–III due to cardiac amyloidosis. Note thickened left ventricular myocardium in a 2-ch **(a)** and short-axis view **(b)**. After gadolinium administration there was a diffuse patchy myocardial enhancement (short-axis view) **(c)**.