Echocardiography in Systemic Diseases

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Abstract

Cardiovascular disorders are a vital manifestation of systemic diseases and lead to substantial mortality and morbidity. Echocardiography has emerged as a useful noninvasive modality for the screening and evaluation of cardiac involvement in these cases. The technological developments in echocardiographic techniques and their concomitant advances in the recognition of subclinical diseases have ushered in a greater appreciation of the nature and prevalence of cardiac involvement in this group of patients. The present review explains cardiac involvement in patients with systemic lupus erythematosus, antiphospholipid antibody syndrome, Takayasu arteritis, and ankylosing spondylitis and introduces the role of different echocardiographic modalities in their evaluation.

Keywords

Systemic diseases · Echocardiography · Systemic lupus erythematosus · Antiphospholipid antibody syndrome · Takayasu arteritis · Ankylosing spondylitis

Abbreviations

APS	Antiphospholipid syndrome
AV	Aortic valve
CAD	Coronary artery disease
MR	Mitral regurgitation
MV	Mitral valve
PH	Pulmonary hypertension
RA	Rheumatoid arteritis
SLE	Systemic lupus erythematosus
TA	Takayasu arteritis
TEE	Transesophageal echocardiography
TTE	Transthoracic echocardiography

Rheumatologic diseases are chronic systemic inflammatory diseases with common involvement of cardiovascular system. Echocardiography provide a comprehensive cardiac assessment in these patients.

Systemic Lupus Erythematosus

- Cardiovascular system is involved in systemic lupus erythematosus (SLE) in different ways. There may be substantial crossover among many of the connective tissue diseases such as mixed connective tissue disease, SLE, Raynaud phenomenon, and scleroderma. Valvular heart disease from SLE may range from mild valvular thickening with normal function to overt valvulo-pathy with vegetations and severe regurgitation or stenosis [1, 2].

SLE affects all parts of the cardiovascular system including (Table 40.1):

- Libman-Sacks vegetations
- Leaflet fibrosis
- Valve regurgitation
- Pericarditis
- Asymptomatic effusion
- Myocarditis/cardiomyopathy
- Coronary artery disease (CAD)
- Atherosclerotic
- Arteritis,
- Pulmonary hypertension

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Table 40.1 Cardiac abnormalities in SLE

Site of involvement	Prevalence (%)
Pericarditis/effusion	11–54
Myocarditis	7–10
Valvular heart disease	15–75
Coronary artery disease	6–10

Modified from Doria A, et al. Cardiac Involvement in systemic lupus erythematosus

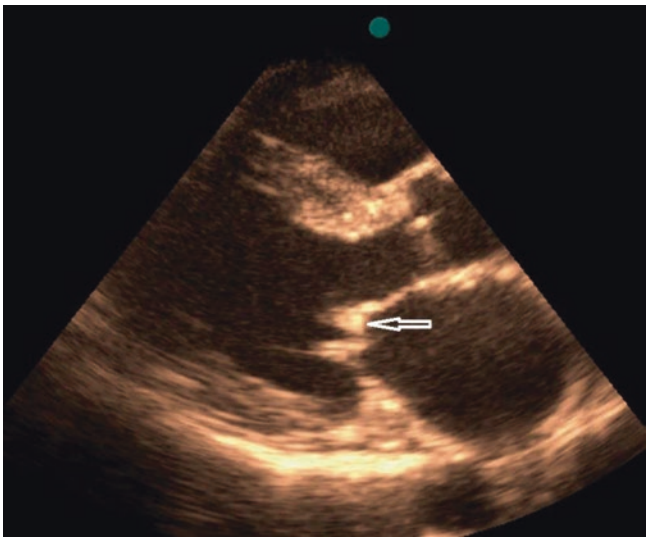

Fig. 40.1 LV long axis view showing thickened MV leaflets especially at their tips

- Echocardiography plays an essential role in the diagnosis, monitoring, and also management of SLE related cardiac disease [2–5].
- A classic lesion encountered in cases with SLE is non-infectious endocarditis with the so-called Libman-Sacks vegetation. Transthoracic echocardiography (TTE) is able to distinguish SLE related vegetations (Libman-Sacks endocarditis) with a sensitivity of 63% and a specificity of 58%. Transesophageal echocardiography (TEE) is superior to TTE in assessing SLE related valve disease. These are most frequently encountered on the mitral valve (MV) and more often are on the atrial side of the leaflet. They tend to be less mobile than infectious vegetations. They might have an inflammatory component that may result in leaflet deformity and variable degrees of valvular regurgitation (Fig. 40.1, Movies 40.1 and 40.2). When encountered on the aortic valve (AV), they are frequently on the arterial side. They might resolve with successful therapy of the underlying disease (Table 40.2).
- Another manifestations of SLE contain coronary vasculitis which may result in regional or global dysfunction and thereby mimic either an acute coronary syndrome or cardiomyopathy [2–4].
- The occurrence of SLE-related pulmonary hypertension (PH) is low; thus, screening echocardiography is not currently suggested for asymptomatic cases.

Table 40.2 Mechanisms of valvular heart disease in SLE

Mechanism	Clinical consequence
Healing of verrucous valvular lesions, leading to leaflet retraction	Valvular regurgitation, and rarely, stenosis
Large valvular verrucous lesion	Obstruction of valvular orifice leading to stenosis or malcoaptation of leaflets leading to regurgitation
Infective endocarditis	Due to underlying abnormal valve or immunosuppression
Chordal rupture in presence of verrucae	Valvular regurgitation
Papillary muscle dysfunction due to acute myocardial infraction	Valvular regurgitation
Mitral valve prolapse	Valvular regurgitation

Modified from Roberts WC, High ST. The heart in systematic lupus erythematosus. Curr Probl Cardio 1999; 24 (1):1–56

- The potential incremental diagnostic value of real time three dimensional TEE in this disease remains to be defined, cause there are only case reports of its use in this object.
- A final sign of SLE might be acute pericarditis. There are no typical features of the pericarditis or pericardial infusion seen in SLE. On rare circumstance, SLE has been related with PH, though this association is far more common with scleroderma.
- In summary, SLE affects all parts of the cardiovascular system. Echocardiography plays an essential role in the diagnosis, monitoring, and also management of SLE related cardiac disease [2–5] (Figs. 40.1 and 40.2; Movies 40.1–40.3).

Antiphospholipid Antibody Syndrome

- Antiphospholipid antibody syndrome is closely related to many connective tissue diseases and has been reported as an integral part of systemic lupus. This syndrome results in a variably hyper-coagulable state with a tendency toward both venous and arterial thrombosis. In addition, cases with the antiphospholipid antibody syndrome develop sterile valvular vegetations similar to those seen in systemic lupus. Though not basically destructive, they might result in valvular regurgitation. They might resolve with successful treatment of the underlying systemic illness and systemic anticoagulation. In all likelihood, some individuals formerly diagnosed with Libman-Sacks vegetative lesions might have had sterile vegetations related to the antiphospholipid antibody syndrome. Large, mobile valvular masses or vegetations may be seen in 10–40% of cases and might be difficult to distinguish from infective vegetations. In addition to valvular vegetations, focal or diffuse thickening of valve leaflets has been detected (Table 40.3).

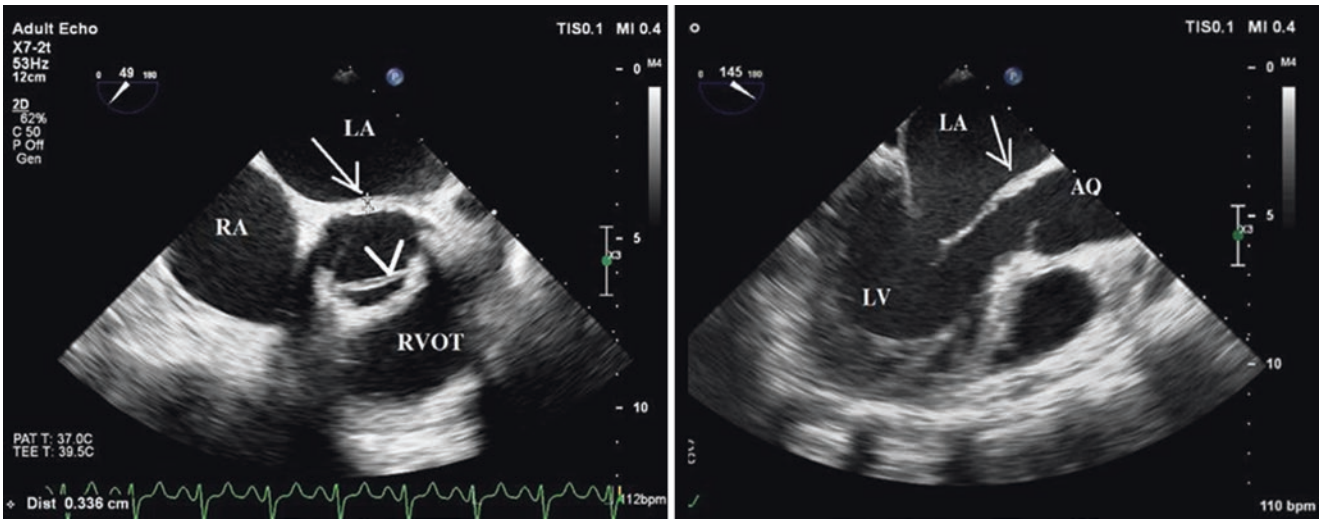

Fig. 40.2 Left: Thickened tricuspid aortic valve. Right: Thickened aortic wall suggestive of aortitis

Table 40.3 Valvular involvement in anti-phospholipid antibody syndrome

Valvular thickness > 3 mm
Localized thickening of proximal and middle portion of the valve leaflets
Nodules on the atrial surface of the mitral valve and/or the aortic surface of the aortic valve
Rheumatic valvular disease and infective endocarditis should be excluded

Modified from Miyakis S, Lockshin MD, Atsumi T, et al. international consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J Thromb Haemost 2006; 4(2):295–306

- On rare circumstances, a catastrophic antiphospholipid antibody syndrome develops with acute severe multi-organ system failure related to micro-thrombosis of arterial and venous circuits. Myocardial necrosis might be a part of this syndrome. From an echocardiographic perspective, it will present as acute vegetative lesions and-or myocardial necrosis with instances of isolated papillary muscle rupture having been described [2, 6, 7].

Scleroderma and Raynaud Phenomenon

- Many other connective tissue diseases may have cardiovascular manifestations. Diseases closely related to SLE such as mixed connective tissue disease characterize a crossover category for' which all the different manifestations of SLE might be seen. Cases with Raynaud phenomenon or with the full complex of scleroderma have a greater than natural prevalence of pulmonary arterial hypertension. In cases with scleroderma, PH anatomically and physiologically is similar to primary PH with an increase in pulmonary vascular resistance at the arteriolar level. Concurrent pericardial effusion might be more common in scleroderma than in PH of other etiologies and is not necessarily an indicator of end stage disease [2, 8].

Takayasu Arteritis (TA)

- TA, a granulomatous panarteritis, affects the aorta and its major branches, characteristically before the age of 40 years. The disease predominates in women, with a female-to-male ratio of up to 10:1. Cause the diagnosis is often delayed, considerable arterial injury accrues. The current diagnostic criteria depend on detection of established stenotic disease and do not yet reflect the increasing sensitivity of noninvasive imaging.
- By Echo-Doppler study aortic arch branch vessels are often affected and the carotid artery can have diffuse, homogeneous wall thickening, known as the 'macaroni sign'. The descending aorta can be stenotic with 'pseudo-coarctation'.
- So, echocardiography, a noninvasive technique, shows a great utility in detection and follow-up of cardiovascular manifestations in cases with TA. New techniques, more sensitive toward detecting the early stages of left ventricular dysfunction, are promising to limit left ventricular hypertrophy development. Also, aortic valve involvement may cause aortic regurgitation in some patients with Takayasu arteritis and that aortic regurgitation is more common than previously believed.
- Additionally, high-resolution ultrasound, CMR, MRA, CTA, and PET have all been studied. Though the potential of these techniques is not in doubt, their specificity and sensitivity in the management of TA remain to be determined. 18F-FDG-PET-CT may reveal evidence of active arteritis and lead to early detection of pre-stenotic disease. A current consensus review has suggested that this technique is particularly useful for the detection of active arteritis in cases

not receiving immunosuppressive therapy. Demonstration of arterial wall enhancement, edema, or thickening on MRA and CTA may facilitate the diagnosis of pre-stenotic disease, and stenoses and aneurysms can be readily identified and monitored. Color duplex ultrasound is of specific use in assessing the common carotid and proximal subclavian arteries in TA. Homogeneous, bright concentric arterial wall thickening is a typical finding in affected common carotid arteries [9–11].

Rheumatoid Arteritis (RA)

- By echocardiography, Mitral regurgitation (MR) is evidenced in 80% of RA. Aortic regurgitation is found in 1/3 of cases. Near 1/4 of RA patients have tricuspid valve abnormalities. Pericarditis is found in about 10% of cases. 1/3 of RA patients has evidence of cardiomyopathy and about 40% has atheroma of the aorta. Rarely, echogenerating nodules are seen on a mitral or aortic valve [12].

Ankylosing Spondylitis

- Cases with ankylosing spondylitis and cardiac abnormalities were older, had a longer disease duration, and more peripheral joint disease than those without cardiac abnormalities. Doppler echocardiography is a useful method in the assessment of cardiac disease in ankylosing spondylitis and might detect aortic valve disease at an early preclinical stage [13].

Questions

1. Which Sentence is wrong in SLE:
   A. SLE mimics either an acute coronary syndrome or cardiomyopathy.
   B. The occurrence of SLE-related pulmonary hypertension is high.
   C. A final sign of SLE might be acute pericarditis.
   D. There are no typical features of the pericarditis or pericardial infusion seen in SLE.

2. Valvular involvement in Anti-phospholipid Antibody Syndrome are except for:
   A. Valvular thickness >3 mm
   B. Localized thickening of proximal and middle portion of the valve leaflets.
   C. Nodules on the ventricular surface of the mitral valve and the aortic valve.
   D. Rheumatic valvular disease and infective endocarditis should be excluded.

3. Which Sentence is wrong in RA:
   A. Mitral regurgitation (MR) is evidenced in 80% of RA.
   B. Aortic regurgitation is found in 1/3 of cases.
   C. Near 1/4 of RA patients have tricuspid valve abnormalities.
   D. Pericarditis is found in about 50% of cases.

Answers

1. B
2. C
3. D

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