Valvular heart disease is prevalent in systemic lupus erythematosus and is a common cause of morbidity and mortality. However, little is known about its relationship to other non-cardiac manifestations of systemic lupus erythematosus and its progression over time. Thus, Roldan et al. performed transoesophageal echocardiography in 69 patients with systemic lupus erythematosus and compared the findings to 56 healthy volunteers. A second transoesophageal echocardiogram was performed 29 ± 13 months later in 58 of the 69 patients (84%). The patients and controls were followed for 57 months to determine clinical outcome. Valvular abnormalities were detected in 61% of the systemic lupus erythematosus patients on the first transoesophageal echocardiogram vs 9% of the controls (P<0.001). Almost all of the abnormalities were on the mitral and aortic valves. Valve thickening was the most common (51% of systemic lupus erythematosus patients, 7% of controls, P<0.001). Vegetations were observed exclusively in the systemic lupus erythematosus patients (43%). Valve regurgitation was common (25% systemic lupus erythematosus vs 2% controls). On the second transoesophageal echocardiogram valve abnormalities often resolved, appeared de novo or persisted unchanged. These changes were not temporally related to the duration, activity, or severity of systemic lupus erythematosus or its treatment. In general, valvular regurgitation did not progress and new valve stenosis did not develop. The combined endpoints of embolic stroke, peripheral embolism, heart failure, infective endocarditis and valve surgery was 22% in the systemic lupus erythematosus patients and 8% of the controls. Death occurred in seven patients, almost all of which was due to valvular disease complications. The investigators concluded that valvular heart disease is common in patients with systemic lupus erythematosus, often changes over time, but appears temporally unrelated to other clinical features of systemic lupus erythematosus, and is associated with considerable morbidity and mortality.

The transoesophageal echocardiography findings in this study are of interest because they are consistent with pathological descriptions in systemic lupus erythematosus patients. The vegetations were usually located on the atrial side of the mitral valve and the vessel side of the aortic valve. Their size ranged from 0.006 to 1.25 cm². They were of heterogeneous echodensity and irregular in shape. These characteristics are consistent with the Libman–Sacks vegetation described in pathological studies. Valve thickening was present in two-thirds of patients with vegetations. The vegetations were not mobile as is often the case with infectious endocarditis vegetations and they were thicker than valve excrescences. Also, the changes observed in the vegetations over time are consistent with pathological descriptions of active and healed vegetations on the same valve suggesting a dynamic process.

The lack of relationship of these valve abnormalities with other clinical features of systemic lupus erythematosus means that therapy for systemic lupus erythematosus is not effective in preventing valvular disease and systemic lupus erythematosus disease actively cannot be used as a marker for valve disease. Also, the detection of valvular disease at one point is not necessarily predictive of progressive disease, since it may resolve spontaneously. Since most systemic lupus erythematosus patients with valve disease are asymptomatic, should echocardiography be serially performed in all systemic lupus erythematosus patients? If so, should transoesophageal echocardiography be performed since this study showed higher incidences of valve disease than previous transthoracic echocardiography studies? Echocardiography is probably too expensive to employ routinely and physical examination cannot reliably detect mild valve disease that may present a substrate for infectious endocarditis. Thus, all patients with systemic lupus erythematosus should probably receive antibiotic prophylaxis for dental and other non-sterile surgery. Systemic lupus erythematosus patients with fever or symptoms and signs of valve disease should have an echocardiogram, preferably transoesophageal echocardiography, to evaluate for possible endocarditis or surgical valve disease.

Strokes are frequent in systemic lupus erythematosus patients and can be due to cerebritis, vasculitis or cardioembolism. In systemic lupus erythematosus patients with stroke and little evidence of vasculitis or cerebritis, transoesophageal echocardiography can be useful to define a cardioembolic substrate which would suggest that anticoagulant
therapy is advisable rather than high dose anti-inflammatory agents. Also, prophylactic antiplatelet therapy should be considered in those with abnormal valves, since they can be a substrate for embolism, but there are no data to support this therapy. Finally, in patients with cardioembolism, surgical resection of valve vegetations may not prevent recurrences because of the unpredictable course of systemic lupus erythematosus valvular disease[8].

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References