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# Libman-Sacks Endocarditis “A Wolf in Sheep’s Skin”

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## Introduction

Libman-Sacks endocarditis (LSE) is a form of non-bacterial endocarditis seen in patients with systemic lupus erythematosus (SLE). It was first described by Emanuel Libman and Benjamin Sacks at Mount Sinai Hospital in New York City in 1924. Libman-Sacks valvular lesions are sterile fibrofibrous vegetations that favor the left-sided heart valves and usually form on the ventricular surface of the mitral valve. The pathogenesis is thought to involve the formation of fibrin-platelet thrombi, which organizes and leads to fibrosis and scarring with subsequent valve dysfunction. An association has been made between LSE and antiphospholipid antibodies (APAs), showing an increased risk of thromboembolic phenomena.<sup>1</sup> Here we report an anomalous case where the patient was found to have SLE, LSE, and antiphospholipid syndrome (APS) that likely presented with an acute peripheral arterial thromboembolic event accompanied by clinical signs and symptoms of congestive heart failure.

## Case Report

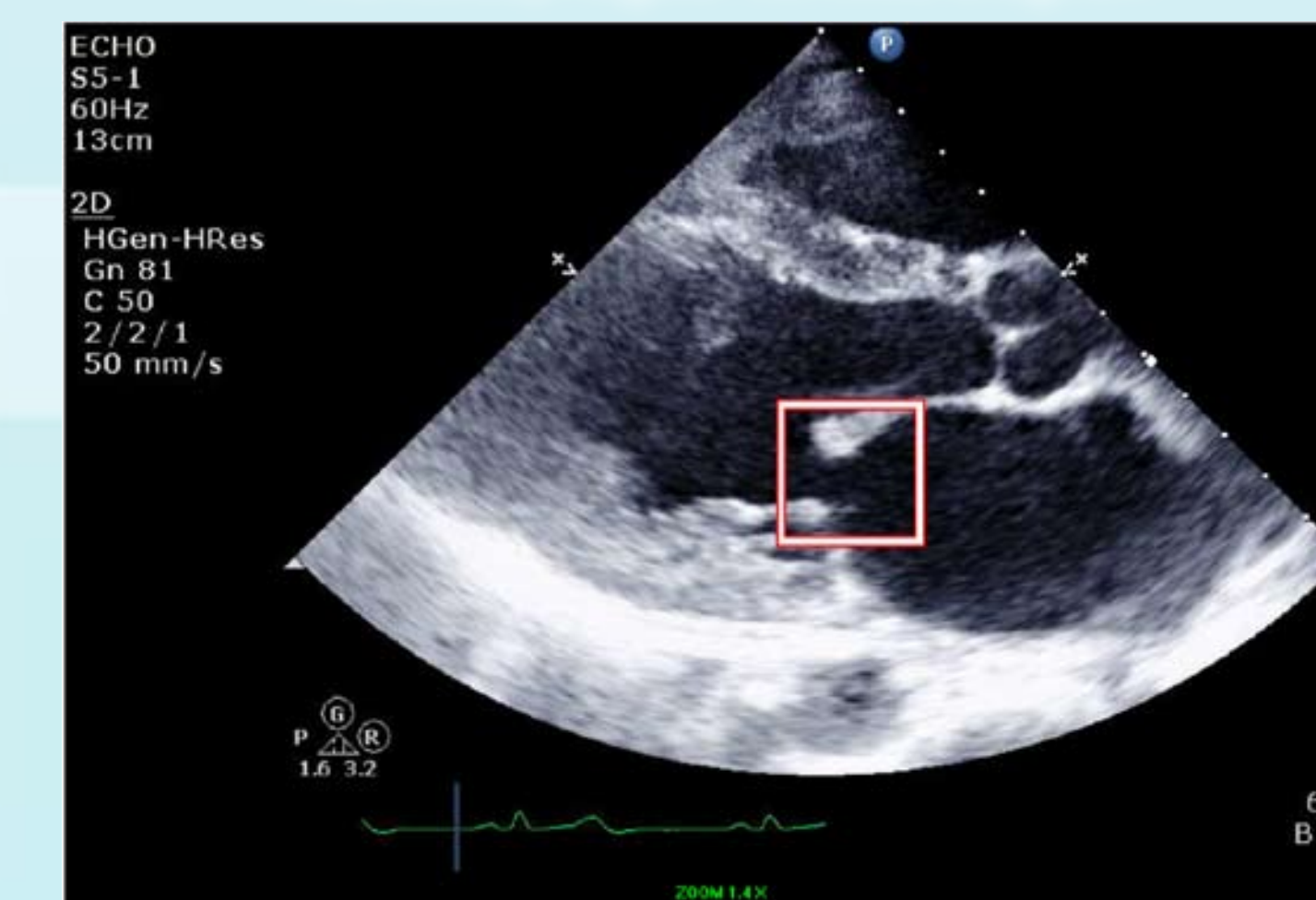
**History of Present Illness:** A 28-year-old African American female with past medical history of hypertension, iron deficiency anemia, tobacco abuse and depression presented to the emergency department with shortness of breath and fatigue worsening over the past three weeks. Over this time period, symptoms did not significantly impact her daily life until she noticed extreme shortness of breath on exertion to the point where she could not walk her child to school. The patient reported having orthopnea during these three weeks with increased swelling in both of her lower extremities; as well as darkening of her fingertips, which had increased in severity within the same time span. She denied fevers, chills, nausea, vomiting or diaphoresis. There was no history of cough, abdominal pain, bloody bowel movements or melena. She did, however, admit to occasional retrosternal chest discomfort, described as a dull ache, rated 3 out of 10, without any radiation to any particular site.

**Hospital Course:** Patient was admitted to the medical/surgical floor. She had an initial hemoglobin of 6.4, WBC of 8.0 and platelets 162,000. On physical exam, she was afebrile, heart rate was 57 per minute and regular, blood pressure was 155/100 mmHg and oxygen saturation of 100% on room air. Patient had mildly elevated JVD as well as a loud S2. There was a 4 out of 6 systolic murmur heard at the left sternal border, which increased with inspiration and a 3 out of 6 systolic murmur at the mitral area, radiating to her axilla. There was trace bilateral pitting edema of lower extremities, halfway up her tibial surface with normal pedal pulses. There were ischemic changes noted on her right fifth digit with some duskiness of the fourth digit. She also had some mild slight changes on the left hand, which were not as pronounced. No oropharyngeal lesions, arthritis or joint deformities were observed.

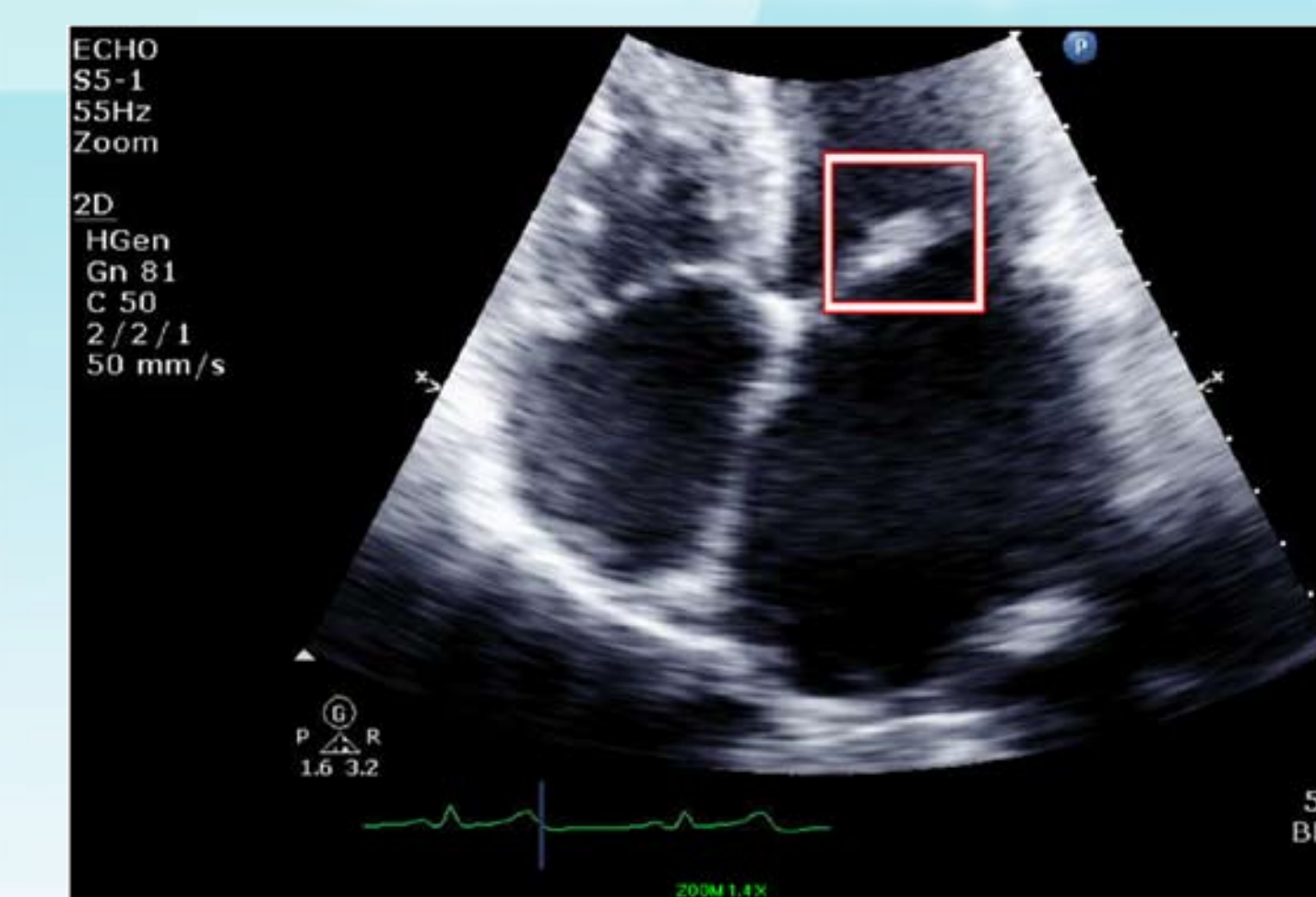
Initially, she was thought to have symptomatic iron deficiency anemia and the GI service was consulted. She was subsequently transfused two units of packed-red blood cells and underwent esophagogastroduodenoscopy revealing erosive gastritis and placed on a proton pump inhibitor. CT scan of the chest showed moderate bilateral pleural effusions as well as a pericardial effusion. The patient underwent a transthoracic echocardiogram showing severe mitral and tricuspid regurgitation with vegetations on the anterior, as well as posterior valve with possible perforation of the valve leaflets. She had pulmonary artery systolic pressure in the 70s, and a positive ANA of 1:5120 in a speckled pattern. For these reasons, both Rheumatology and Cardiothoracic Surgery services were consulted. Transesophageal echocardiogram confirmed a teardrop vegetation located on mitral valve with thickened valve leaflets and severe tricuspid and mitral regurgitation with a moderate pericardial effusion. Further rheumatologic workup revealed positive lupus anticoagulant and anticardiolipin antibodies. She was found to have elevated IgG, IgA, ESR, CRP, RNP, positive dsDNA and anti-Smith antibodies as well as positive APL antibodies. Blood cultures were obtained on four different occasions, all found to have no growth and the patient remained afebrile. Using modified Duke’s criteria the diagnosis of Libman Sacks endocarditis was made. Anticoagulation therapy with unfractionated heparin was started, which was later maintained with warfarin (INR- 2.5-3.5). The patient also received prednisone (60 mg/day) and Plaquenil (400mg/day). She ultimately underwent mechanical valve replacement and repair of her tricuspid valve 3 weeks later.

## Images

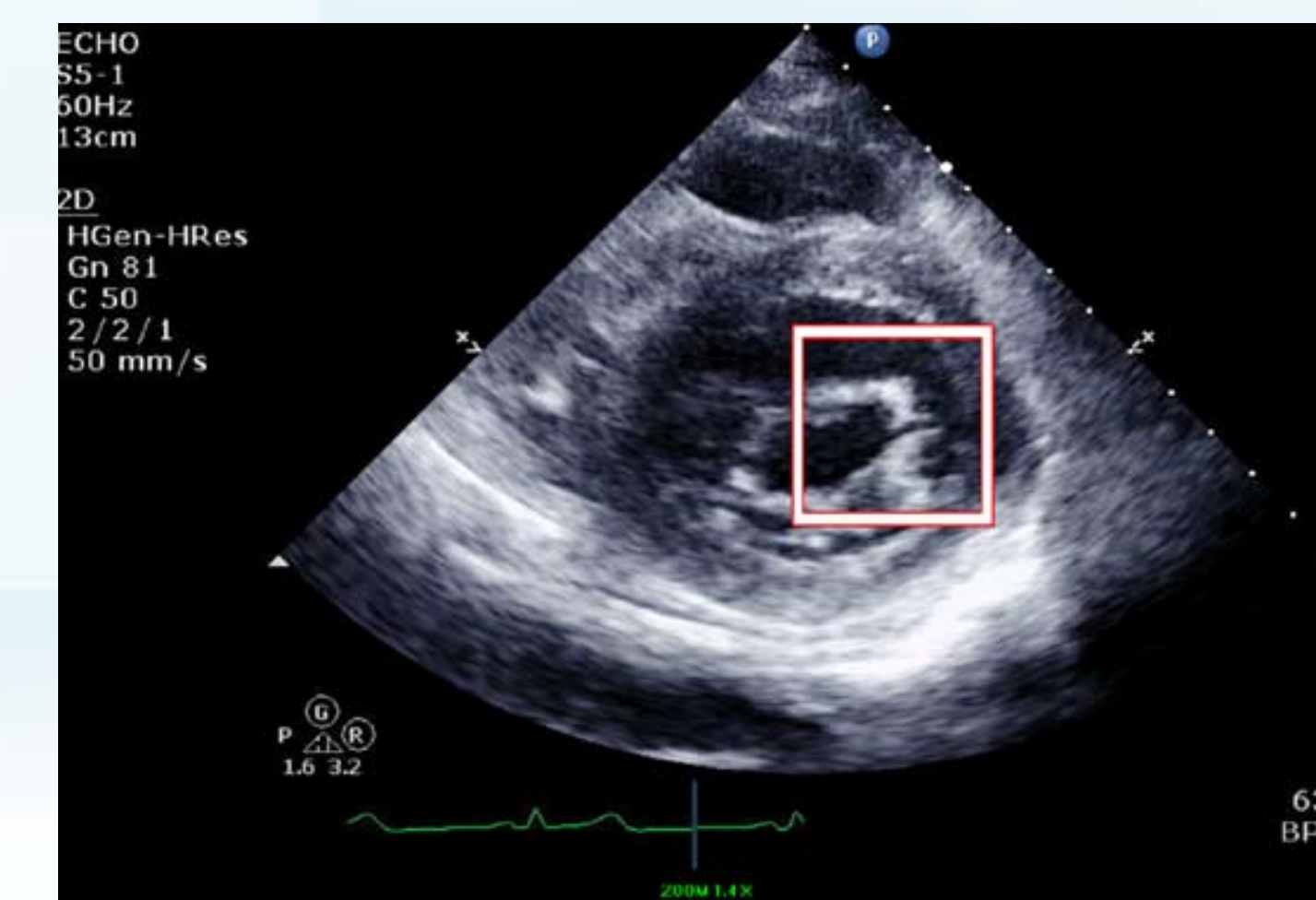
**Mitral Valve Vegetation**  
Transesophageal Echocardiogram



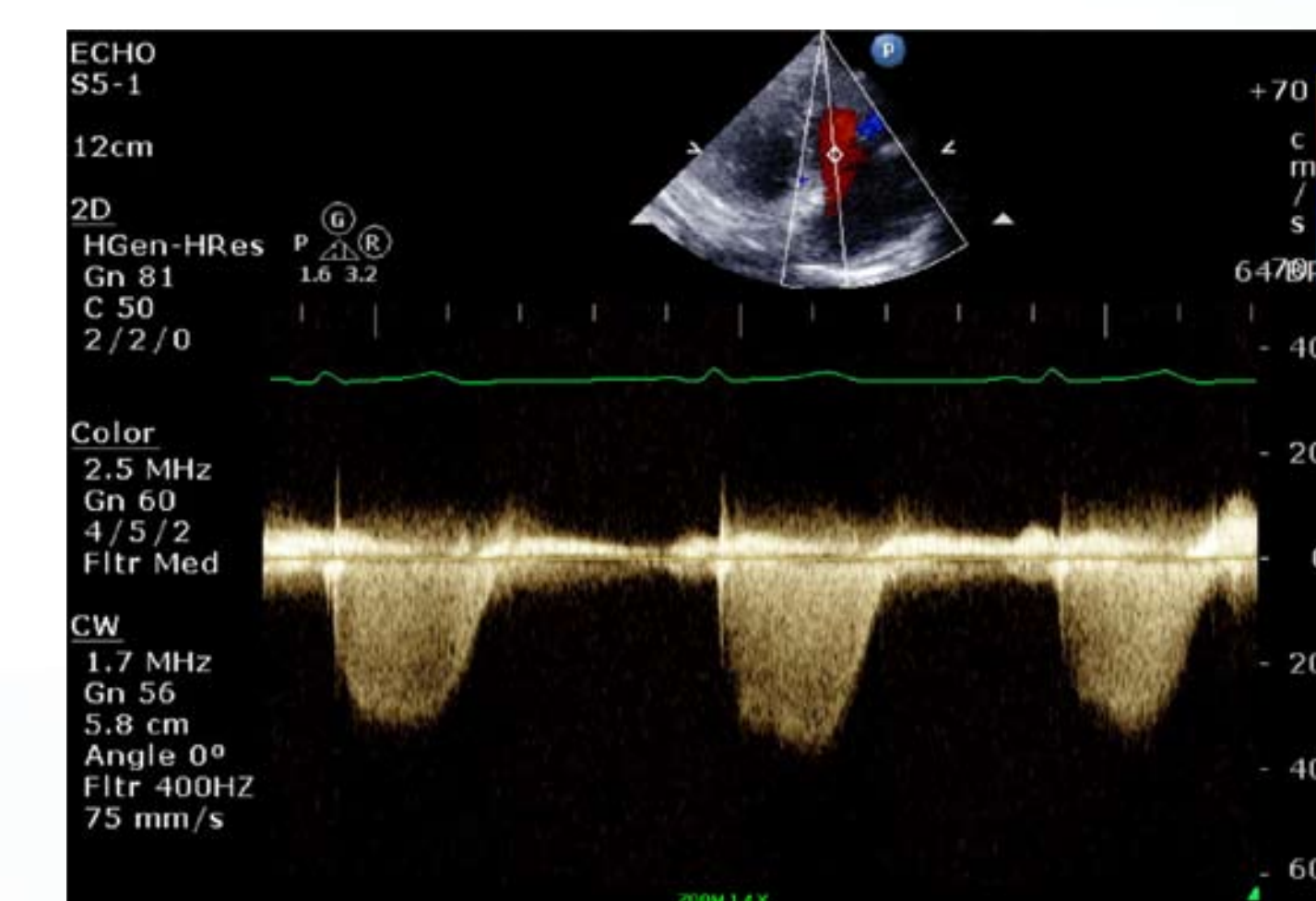
**Mitral Valve 4-Chamber Apical View**  
Transthoracic Echocardiogram



**Mitral Valve Parasternal View**  
Transthoracic Echocardiogram



**Tricuspid Regurgitant Jet**  
Transesophageal Echocardiogram



## Discussion

This case report describes a patient with SLE (established according to the criteria of the American College of Rheumatology),<sup>1</sup> who presented with signs and symptoms of congestive heart failure as well as multiple areas of suspected ischemia possibly secondary to either LSE or APS. In fact, the patient had aseptic vegetations on the mitral valve with associated positive lupus anticoagulant antibodies characterizing a diagnosis of APS.<sup>2</sup>

More than half of the patients with SLE, when assessed with transesophageal echocardiogram have clinically silent valvular alterations. One complication of SLE seen in approximately 9% of patients is a thromboembolic phenomenon with the brain being most affected.<sup>3</sup> In most cases, embolic episodes are subclinical, but sometimes may manifest as signs and symptoms of ischemia. Approximately 33% of SLE patients are found to have positive APL antibodies. An increased risk of thromboembolic events is observed in LSE, which might be in part due to the presence of the APAs.<sup>4,5</sup> Patients with moderate to high titers of anticardiolipin IgG antibodies have a higher incidence of valve lesions, whereas there are patients with valve disease in whom lupus anticoagulant are the only APAs detected.<sup>2</sup>

It should be considered that infectious endocarditis is not uncommonly seen in SLE patients with LSE as patients already have valvular dysfunctions. Thus a broad differential diagnosis is mandatory. In this aspect, three laboratory data are important: leukocyte count, CRP levels, and APA levels.<sup>5</sup> Leukocytes tend to decrease during lupus activity and the opposite occurs in infectious endocarditis. As for the APAs, it is unlikely that elevated levels would be caused by an infectious disease, thus suggesting the presence of SLE. Very high CRP levels suggest an infectious cause, as lupus patients are less capable of presenting a large response of this protein; however, for a definitive differential diagnosis, blood cultures are imperative.<sup>5</sup> In the present case, a diagnosis of LSE was attained, as the leukocyte count was normal, the CRP was not very elevated, APA was positive and 4 pairs of blood cultures had no growth.

There is scarce information in the literature regarding treatment of LSE. It is known that the use of corticosteroids and immunosuppressive drugs seem to have no effect on valve lesions; however, anticoagulation therapy must be used for treatment of patients with thromboembolic events.<sup>5</sup>

In the present case, the patient initially received Lovenox and ultimately Warfarin therapy to maintain an INR of 2.5-3.5. In this aspect, it should be considered that prospective, controlled, and randomized studies have shown that moderate (INR 2.0-3.0), as well as a more intensive (INR 3.0-4.0) anticoagulation with warfarin were similarly effective in preventing new thromboembolic events in patients with APS after the first thrombosis.<sup>6</sup>

Hydroxychloroquine and prednisone (60 mg/day) were also added to her treatment regime as she had signs of disease activity (pericardial effusion, increased ESR and decreased C3). The potential usefulness of hydroxychloroquine in preventing events related to the APS remains in the case of SLE patients, particularly those with APAs, as there is evidence of fewer thrombotic events with its use.<sup>7</sup>

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