Thrombotic microangiopathy is a syndrome that comprises several disorders characterised by localised or diffuse microvascular thrombosis. These include the catastrophic antiphospholipid syndrome (CAPS), thrombotic thrombocytopenic purpura (TTP), hemolytic uremic syndrome (HUS), disseminated intravascular coagulation (DIC) in the context of systemic infections or malignancies, hypertension-related, pregnancy-related and drug-related microangiopathic syndromes, and heparin-induced thrombocytopenia. Some of these clinical scenarios such as severe preeclampsia and HELLP syndrome may be suspected given the appropriate clinical context such as pregnancy. In other cases, the most important point is to perform a systematic and complete clinical history and physical examination looking for previous thrombosis or pregnancy morbidity, uncontrolled hypertension, bloody diarrhoea, and exposure to heparin or some concrete drugs such as ticlopidine, clopidogrel, chemotherapy agents, and alendronate that have been identified as probable cause of thrombotic microangiopathy. In addition, unexplained weight loss, gradual onset of symptoms, hepatomegaly or splenomegaly, or palpable lymphadenopathies may be indicate the existence of malignancy triggering the thrombotic microangiopathy. High-grade fever accompanying chills may be the signs of systemic infection in the form of severe sepsis. A history of acute gastroenteritis with bloody diarrhea caused by verocytotoxin (Shiga-like toxin)-producing Escherichia coli but also Shigella dysenteriae type I and Citrobacter freundii might indicate HUS. In cases of severe hypertension, fundoscopic exam is mandatory to rule out the evidence of exudates and papilla oedema pointing to a malignant hypertension.

However, differential diagnosis is not so easy in the real-world setting. On the one hand, the clinical picture may be very similar between different diseases. Kidney and neurologic involvement may be present in CAPS but also in malignant hypertension, severe sepsis, and TTP/HUS. Conversely, physicians should remember that in most causes of thrombotic microangiopathy, other than CAPS, antiphospholipid antibodies (aPL) may also be present. The aPL have been reported in some patients with TTP/HUS, but usually at low titers. In this context, a decrease in ADAMTS13 activity of fewer than 5% points to TTP as the most probable diagnosis. Infections are capable of inducing aPL but normally at low titer and they are not persistent over time. With all these data, in front of a patient with thrombotic microangiopathy, a comprehensive diagnostic workup should be carried out.

Learning Objectives
- Explain the main challenges in the differential diagnosis of microangiopathy in SLE
- Describe the treatment options for microangiopathy in SLE
- Discuss new trends in research on new markers for microangiopathy in SLE

REFERENCES