NYU Langone Health

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2020 HIGHLIGHTS

COVID-19 Risk for Immunosuppressed Patients

Research Determines Whether Patients Prescribed Immunosuppressants Face Increased Risk

See page 2.

Reducing Risks for Lupus Patients Across the Age Spectrum

Studies Seek to Identify and Reduce Adverse Outcomes During Pregnancy and for Seniors

See page 4.

Lupus Patient Facing rombocytopenia and Anemia

Treatment Regimen Prevents Relapse and Provides E ective Solution

See page 7.

To help clarify the in ammatory process,
Dr. Abramson and collaborators, including

As an extension of the PATCH study, the researchers investigated the potential for cardiac toxicity in the neonates of anti-SSA/Ro antibody–positive mothers taking HCQ. Past research had reassuringly suggested that such toxicity manifest as a dangerously lengthened QTc interval on electrocardiograms (ECGs) did not occur. But Dr. Buyon notes that most such studies relied on women's self-reported adherence to HCQ despite well-documented adherence shortfalls.

e PATCH study overcame such bias by measuring HCQ levels during each trimester in maternal blood and cord blood and then comparing these levels with the newborn's QTc intervals. Study collaborator Deborah Friedman, MD, a pediatric cardiologist at New York Medical College in Valhalla, evaluated the neonate ECGs and measured their QT intervals to detect any abnormalities.

Excluding those neonates diagnosed with heart block, the resulting analysis, published in the journal *Circulation: Arrh thmia and Electro-ph siolog* , showed no relationship between HCQ levels and the QTc interval. Of the 45 neonates included in the analysis, ECG measurements suggested that ve did have a prolonged QT interval, with three deemed only marginally abnormal and two clearly abnormal. "We saw no negative clinical consequences in any of these

J N , MD

e bound peptides can then be presented on the cell surface—a key step in T cell recognition and activation. Although ERAP1 normally cuts to a 9-mer target, the Hap10 variant's low enzymatic activity results in insu cient trimming and overly long peptides.

Dr. Nowatzky hypothesizes that packaging of longer peptides in HLA molecules, including HLA-B51, alters the immune response in a way that can lead to Behçet's syndrome and its characteristic uveitis. To study the potential contribution of ERAP1 and HLA-B*51, his lab is using CRISPR-Cas9 genome editing to knock out or modify the ERAP1 gene and study the resulting activity of HLA-bearing cells in immunofunctional assays. "When we change this gene, we're looking at how the peptidome changes, meaning all of the proteins that are bound to HLA-B51," Dr. Nowatzky says. " en we look at how these genetically modi ed cells behave with other cells they are supposed to stimulate or to inhibit, especially CD8 T cells."

e researchers are also comparing the immune phenotypes of patients with severe Behçet's syndrome from Turkey and the United States who carry the ERAP1 Has o2 (hrr.6 (-0.01G(a)-2)7)-7.7(m)1.10.6 en we

r m12.4 (i)1.7 (ck (e)-5.3 (n w)-1(e)-8.2 (l)-360P.7 (l o)0.8)-8.2(i)-6

H. Michael Belmont, MD, professor in the Department of Medicine and medical director at NYU Langone Orthopedic Hospital, says clinicians should be aware of the broad di erential diagnoses of thrombocytopenia in SLE. Most commonly, these include autoimmune thrombocytopenia (AITP) with antibodies to platelet membrane antigens (such as glycoproteins IIb/IIIa, Ib/IX, or Ia/IIa); AITP plus autoimmune hemolytic anemia (AIHA), also known as Evans syndrome; and a non-criteria manifestation of antiphospholipid syndrome.

Clinicians, though, also need to consider less common etiologies, including those in which the thrombocytopenia is accompanied by evidence of a thrombotic microangiopathic hemolytic anemia (TMHA) process such as thrombotic thrombocytopenic purpura (TTP) with an immuno-globulin inh

By mid-July 2019, the patient reported feeling well at a follow-up appointment, with no rash, alopecia, or arthritis, previous hallmarks of her SLE. Further, she had no fever, oral ulcers, shortness of breath, pleuritic chest pain, nausea, vomiting, abdominal pain, edema, or headaches. Her platelet count had rebounded to 170,000, and her ADAMTS13 activity remained high, at 81 percent.

Since her hospitalization, the patient has remained on a 5-mg dose of prednisone, 200 mg of hydroxychloroquine three times a week, and 1,000 mg of mycophenolate mofetil twice a day. She has not had a relapse of her thrombocytopenia since the initial event.

Dr. Belmont is providing maintenance courses of rituximab at six-month intervals to prevent a recurrence of TTP and reappearance of the anti-ADAMTS13 antibody. is case, he says, illustrates the potential for severe lupus exacerbation despite progression to end-stage kidney disease and initiation of hemodialysis. In addition, he says, it underscores the importance of considering TMHA as the mechanism of thrombocytopenia in patients with lupus with a di erential diagnosis that includes secondary TTP on the basis of an acquired immunoglobulin inhibitor of ADAMTS13. ■

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