Neonatal lupus erythematosus (NLE) is an uncommon disease that occurs in the neonate from transplacental passage of maternal autoantibodies in utero. The most common autoantibodies passed from the mother are anti-Ro/SSA and anti-La/SSB, though other autoantibodies have been implicated. It commonly manifests as skin lesions or congenital heart block but can involve the liver. Rarely, hepatobiliary disease is the sole clinical manifestation of the disease and can present similarly to gestational alloimmune liver disease (GALD). We present a case of severe liver disease in an infant born to a mother with systemic lupus erythematosus (SLE) with histopathological features of perinatal autoimmune hepatitis.

CASE SUMMARY
An infant was delivered at 37 weeks gestational age for growth restriction. The mother was a 34-year-old G1P1 who was diagnosed at age 16 years with SLE with pericarditis, severe joint pain, fever, alopecia, and malar rash, without liver involvement. The mother’s SLE had been stable for the past nine years, including during pregnancy during which she was treated with azathioprine and hydroxychloroquine. At age one month, the infant presented with jaundice and thrombocytopenia. Liver biopsy showed cirrhosis with a micronodular pattern, increased iron deposition, and minimal inflammation (Figure 1). Rapid progression to liver failure at age five months required liver transplant. Grossly, the explant was diffusely nodular with fibrotic septa and areas of dense fibrosis (Figure 2). Microscopically, there was mixed coarse and micronodular cirrhosis with prominent periportal inflammatory infiltrate. The overall features suggested GALD progressing to autoimmune hepatitis with cirrhosis, a diagnosis at odds with the young age of the patient. We propose that the liver disease in this infant is related to transfer of maternal antibodies associated with SLE to the fetus. Serologic testing of the infant at the time of presentation revealed positive anti-ANA (1:320) and anti-ribonucleoprotein (anti-RNP) antibodies; post-transplantation testing was negative for both markers. The mother was positive for anti-ANA (>1:1280) and anti-RNP antibodies, along with low-positives for anti-dsDNA and anti-LC1 antibodies. Both mother and infant were negative for anti-SSA and anti-SSB antibodies.

DISCUSSION
The overall features suggested GALD progressing to autoimmune hepatitis with cirrhosis, a diagnosis at odds with the young age of the patient. We propose that the liver disease in this infant is related to transfer of maternal antibodies associated with SLE to the fetus. Serologic testing of the infant at the time of presentation revealed positive anti-ANA (1:320) and anti-ribonucleoprotein (anti-RNP) antibodies; post-transplantation testing was negative for both markers. The mother was positive for anti-ANA (>1:1280) and anti-RNP antibodies, along with low-positives for anti-dsDNA and anti-LC1 antibodies. Both mother and infant were negative for anti-SSA and anti-SSB antibodies. While rare cases of NLE associated with anti-RNP have been reported, liver disease has not been previously described. This case represents a unique hepatobiliary manifestation of presumed neonatal lupus involving anti-RNP antibodies, resulting in a liver disorder that initially with histological features of GALD that rapidly progressed to a phenotype of end-stage autoimmune hepatitis.