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Kawasaki's Disease with Cholangitis in an Operated Biliary Atresia

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Kawasaki disease has not been reported in children with cholangitis and biliary atresia.

Case : A 1 years old girl, operated case of biliary atresia (Kasai surgery) presented with fever, clay colored stools and upper respiratory symptoms in April 2012. She had been treated for enterobacter septicemia and pneumonia in March 2012. Currently in April 2012, she had jaundice, hepatosplenomegaly with dilated veins over abdomen and bilateral wheeze. Ultrasound of abdomen showed hepatosplenomegaly with portal collaterals suggestive of portal hypertension. Her repeat blood culture did not grow any organism. She was treated with antibiotics in view of suspected cholangitis. She responded to the same and became afebrile. However after 10 days, she again developed fever, leukocytosis and strawberry tongue with cervical nodes. Her blood culture was still sterile. In view of increasing platelets, high CRP (232) and high ESR (120mm at end of 1 hour) along with clinical features, she was suspected to have Kawasaki's disease. An echocardiography showed left coronary ectasia. She was subsequently treated with intravenous immunoglobulin and fever subsided within 24 hours, leucocyte count and platelets normalized, CRP, ESR started decreasing. Aspirin was not started in view of portal hypertension and underlying liver disease. On follow-up, echocardiography, in June 2012, there was persistence of the coronary artery ectasia and intermittent fever. She was subsequently treated with prednisolone which was tapered in next 21 days. Her echocardiography in July 2012 showed decrease in coronary dimensions and her fever disappeared.