Vanishing bile duct syndrome in HIV infected patients: a case series

E Mabonga1, K Childs2, R Brum1, S Jebakumar3, S Ariyanayagam4, M Nelson3, K Agarwal5, M Tenant-Favours1, C Taylor1
1Department of Sexual Health and HIV, King’s College Hospital NHS Foundation Trust, 2Institute of Liver Studies, King’s College Hospital NHS Foundation Trust, 3Department of GUM/HIV, Peterborough City Hospital, 4Chelsea & Westminster NHS Foundation Trust

Background
Vanishing bile duct syndrome (VBDS) is a rare acquired disorder associated with progressive destruction and disappearance of intrahepatic bile ducts and ultimately cholestasis. The diagnosis is made on histology. “Ductopenia” refers to the absence of interlobular bile ducts from within the portal and exists when there is loss of interlobular bile ducts in more than 50% of portal tracts.

Multiple aetiologies have been identified including infections, neoplastic disorders, autoimmune conditions and drugs. In HIV negative patients the commonest causes of VBDS in published case reports are drugs and Hodgkin lymphoma.

We report the first case series of VBDS in HIV infected patients.

Methods
Case notes and electronic patient records were reviewed of patients known to have a histological diagnosis of VBDS

Results
Five patients were identified, all male. All presented with symptoms of cholestatic jaundice. There was no evidence of cholangiopathy on magnetic resonance imaging. Biopsy of their livers demonstrated cholestasis with severe ductopenia (figures 1 & 2).

• The initial liver biopsy in patient A demonstrated features of VBDS; a second biopsy was done 15 months later as his symptoms did not improve which revealed Hodgkin lymphoma.
• Patient B presented with jaundice, weight loss and cervical lymphadenopathy. A lymph node biopsy revealed Hodgkin lymphoma. He was initially managed with chemotherapy (ABVD). He was referred to King’s as he had persistently raised bilirubin. He went on to have a liver transplant with resolution of symptoms.
• Patient C had poor adherence with uncontrolled HIV viraemia. Histology revealed features of early VBDS (figure 2). He has been lost to follow up.
• In patient D the initial aetiology for VBDS was Darunavir toxicity. He has subsequently developed features suggestive of lymphoma and is awaiting a lymph node biopsy.
• Patient E the aetiology for VBDS is unknown.
• All patients with the exception of patient E had chronic HIV infection with features of immune suppression with or without detectable HIV viraemia.
• Compared to the literature in HIV more of our cases were unrelated to drug exposure.

Conclusion
In HIV-1 infected patients who present with cholestasis and normal intrahepatic bile ducts on imaging, a diagnosis of VBDS should be considered. Early biopsy needs to be performed to establish a diagnosis. Lymphoproliferative malignancy and drugs are the commonest associations in our series.

References