

Outcome of Extrahepatic Portal Vein Obstruction in Children

Abstract

Background: Extrahepatic portal vein obstruction (EHPVO) developing due to thrombotic occlusion of the portal vein in children is generally a benign disease. The aim of this retrospective study was to describe clinical presentation, risk factors, complications and treatment in children with EHPVO presented to the Pediatric Hepatology Unit, Cairo University Children's Hospital, Egypt.

Materials and methods: The medical records of 145 children (89 male, median age 10 years, range 7 months- 18 years) presenting with EHPVO between 1988 and 2008 were reviewed retrospectively.

Results: Clinical presentation included bleeding in 58% and splenomegaly in 37%. Prolonged prothrombin time was seen in 26% and elevated liver enzymes in 6%. On first endoscopy, oesophageal varices were present in 131 (90%) patients; of those subjects, 101 (77%) received sclerotherapy, 7 (5.3%) had band ligation and 14 (10.6%) received both. Thrombophilia investigation was performed in 40 patients revealed: factor V Leiden mutation in 12, protein C deficiency in 11, factor II mutation in 6 and antithrombin III deficiency in 1 patients. Liver biopsies were performed in 68 patients revealed mixed portal cirrhosis in one and minimal fibrosis in four patients.

Conclusion: the aetiology of EHPVO in the majority of patients remains unknown. The overall prognosis is good. Sclerotherapy and banding are effective treatments for bleeding varices with good long term outcome.