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Topic: AS02. HEPATOLOGY/AS02a. General Hepatology

FIRST VARICEAL HEMORRHAGE: INTERVENTIONS AND OUTCOMES - RESULTS FROM AN INTERIM ANALYSIS OF THE INTERNATIONAL MULTICENTER PAEDIATRIC PORTAL HYPERTENSION REGISTRY

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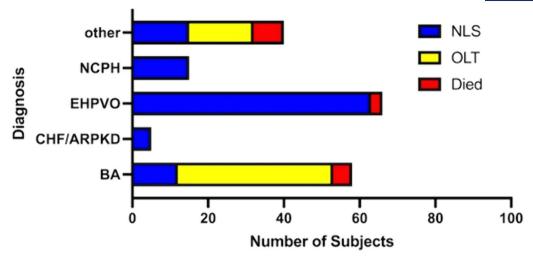
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Objectives and Study: There is a paucity of information about first variceal hemorrhage (1VH) in children, limiting evidence-based approaches to portal hypertensive variceal management. This multicenter study aims to provide important insights into this essential clinical question.

Methods: This international multicenter retrospective registry examines 1VH that occurred between 1/7/12 and 30/ 6/17 with follow up until 30/6/20. This interim analysis examines data accrued until 31/8/23.

Results: 190 subjects with 1VH were identified at 25 centres from 12 countries on 4 continents (mean + SD for age at 1VH 6.0 + 5.1 years). Biliary atresia (BA, 30.5%) and extrahepatic portal vein obstruction (EHPVO, 34.2%) were the most common diagnoses (Figure). Endoscopic secondary prophylaxis of VH included band ligation (n=110) and sclerotherapy (n=48). Band ligation resulted in a higher and faster eradication rate for varices (66%) compared to sclerotherapy (27%, p=.0002) with an average of 3.3 band ligation sessions vs 5.8 sclerotherapy sessions required for eradication. Re-bleeding occurred in 27 subjects <3 months of 1VH, and in 37 >3 months after 1VH. Other interventions after 1VH included TIPS (n=4, 3 OLT, 1 SNL), mesorex bypass for EHPVO (n=7, 4 no secondary prophylaxis, 7 SNL), portosystemic shunt for EHPVO (n=8, 3 no secondary prophylaxis, 8 SNL). Mortality from 1VH, defined per Baveno as mortality within 6 weeks of VH, occurred in 5 subjects (2.6%) none of whom had either BA or EHPVO (diagnoses – Fontan associated liver disease, cryptogenic cirrhosis, Budd-Chiari, Alagille syndrome, inborn error of metabolism). Seven subjects (3.6%) underwent OLT within 6 weeks of VH, 6 of whom had BA (4 poor or no bile flow). Primary etiology of liver disease determined the outcome (Figure).

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NCPH - noncirrhotic portal hypertension, EHPVO - extrahepatic portal vein obstruction, CHF/ARPKD - congenital hepatic fibrosis/autosomal recessive polycystic kidney disease, BA - biliary atresia, NLS - native liver survival, OLT - orthotopic liver transplant

Chi-square p < 0.0001

Conclusions: Early mortality from 1VH is low. Approaches and outcomes after 1VH are related to underlying disease. Nontransplant surgery is utilised for EHPVO. Supported by an ESPGHAN networking grant and the Spain Family.

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