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Topic: *AS02 HEPATOLOGY / AS02a General Hepatology*

DEVELOPMENT AND INITIAL RESULTS OF THE INTERNATIONAL MULTI-CENTRE PAEDIATRIC PORTAL HYPERTENSION REGISTRY (IMPPHR)

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Objectives and Study: Due to limited data, the management of variceal hemorrhage (VH) is controversial in paediatrics. IMPPHR aims to derive large-scale international data informing management of VH.

Methods: The major foci of IMPPHR's retrospective data collection are, 1) morbidity and mortality of first VH, 2) feasibility of primary prophylaxis of VH , 3) approaches to secondary prophylaxis . Each centre provides characteristics and clinical activity accrued between 1/1/18 and 31/12/19. Potential subjects are children with first VH or primary endoscopic prophylaxis between 1/7/12 and 30/6/17 with follow up until 30/6/20.

Results: As of 30/11/22, of the > 50 centres expressing interest, 7 centres (median population 6M) located in 5 countries on 4 continents have entered centre-specific and patient level data (n=136). Centre characteristics and 2-year activity include (median:25%-75%ile): hospital beds (289:76-354), ICU beds (32:16-42), interventional radiologists (4:2-5), OLT (34:7-80), LRD OLT (0:0-10), mesorex bypass (1:0-6), variceal ligation sessions (22:18-25), sclerotherapy sessions (4:2-45). 71 patients entered for 1st VH (biliary atresia-19, EHPVO-30, eleven miscellaneous diagnoses–22) undergoing ligation-41, sclerotherapy-17, no endoscopic intervention-6 (7 unknown). 65 entered for primary prophylaxis (EHPVO-16, biliary atresia-14, autoimmune hepatitis-7, CHF/ARPKD-5, CF-5, PSC-4, eight miscellaneous diagnoses-14) undergoing ligation-52, sclerotherapy-4 or both-2 (7 unknown). Variceal eradication was accomplished in 52% after 1st VH (4.0:2-6 sessions) and 62% undergoing primary prophylaxis (4.0:3.0-5.5 sessions). 6 underwent mesorex bypass and 6 portosystemic shunting (5 EHPVO) during follow-up. Status at final follow-up for 1stVH and primary prophylaxis was alive with native liver (57 and 65%), OLT (30 and 33%), died (11 and 2%, including 2 (2.8%) within 6 weeks of 1st VH).

Conclusions: IMMPHR is feasible and generated novel preliminary data – the target of > 1000 cases will provide critical information guiding clinicians caring for children with portal hypertension. Supported by the Spain Family and an ESPGHAN Networking Grant.

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