

centers in Europe. Patients were followed up at 1, 3, 6, 9 and 12 months, or until death or LT. The primary endpoint was 42-day mortality (LT as a competing event). We also aimed to develop a futility score for TIPS placement, and to validate it in a previously published retrospective multicenter European cohort.

Results: Overall, 177 pts were included (73.4% men, age 54 years, alcohol-related cirrhosis 76%, Child-Pugh score 9, MELD score 16, ACLF 23%, salvage/rescue TIPS 78.5%/21.5%). Forty-two-day mortality was 24.3%, with no significant difference between salvage and rescue TIPS ($p=0.14$). In multivariate analysis, factors independently associated with 42-day mortality were higher age, ascites, higher bilirubin, INR and creatinine and vasopressors at TIPS placement. Only one patient had a Child-Pugh score ≥ 14 , precluding validation of this criterion. Among the 27 patients with a MELD score ≥ 30 and/or lactate ≥ 12 mM, the 42-day transplant-free survival rate was 37%. A futility score was developed using the previously listed factors associated with 42-day mortality: the AUROC for the prediction of 42-day mortality was 0.808 and 42-day mortality was 100% in patients with a score ≥ 90 in our derivation cohort. In the validation cohort of 164 patients, 29 patients (17.7%) had a score ≥ 90 , with a 42-day mortality rate of 93.1%.

Conclusion: In this prospective series, rescue/salvage TIPS was associated with a better prognosis than previously reported, possibly due to patient's selection. Previously described futility criteria were not validated. A new score should be used to discuss rescue/salvage TIPS on a case-by-case basis in patients who are not candidate for LT.

OS-085-YI

The natural history of porto-sinusoidal vascular disorder without portal hypertension: a multicenter retrospective VALDIG study

Edeline Kaze¹, Sarah Raevens², Géraldine Dahlqvist³, Margherita Saracco⁴, Lucile Moga⁵, Nicola Pugliese⁶, Elisa Farina⁷, Emma Murcia⁸, Lorenz Balcar⁹, Carina Boros¹⁰, Martina Luca¹¹, Lorenzo Argiento¹¹, Heloise Giudicelli-Lett¹², Thomas Vanwolleghem¹³, Ann Driessen¹³, Sarah Shalaby¹⁴, Wagner Enrique Ramirez¹⁴, Djamal Tazibt¹⁵, Odile Gorla¹⁶, Laure Elkrief¹⁷, Jean Delwaide¹⁸, Astrid Marot¹⁹, Quentin Schoevaerdts¹⁹, Christophe Moreno²⁰, Jean Paul Cervoni¹⁵, Virginia Hernández-Gea¹⁴, Juan Carlos García-Pagán¹⁴, Sven Francque¹³, Dominique Thabut¹², Carla Cafasso²¹, Marianne Zioli¹⁰, Pierre Nahon¹⁰, Georg Semmler⁹, Bernhard Scheiner⁹, Christophe Bureau⁸, Mauro Viganò⁷, Alessandro Loglio⁷, Alessio Aghemo⁶, Aurélie Plessier⁵, Pierre-Emmanuel Rautou⁵, Pamela Baldin³, Laurine Verset²⁰, Anne Hoorens², Pierre Deltenre²². ¹Hôpital Erasme-Cliniques Universitaires de Bruxelles, cliniques de l'Europe, Bruxelles, Belgium; ²Ghent University Hospital, Gent, Belgium; ³Cliniques Universitaires Saint-Luc, Bruxelles, Belgium; ⁴Azienda Ospedaliera Città della Salute e della Scienza di Torino, Torino, Italy; ⁵Hôpital Beaujon, Université Paris-Cité, Clichy, France; ⁶Humanitas University, Rozzano, Italy; ⁷ASST Papa Giovanni XXIII, Bergamo, Italy; ⁸Hôpital Rangueil, Toulouse, France; ⁹Medizinische Universität Wien, Wien, Austria; ¹⁰Hôpital Avicenne, Bobigny, France; ¹¹Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Milano, Italy; ¹²Hôpital Universitaire de la Pitié-Salpêtrière, Paris, France; ¹³Antwerp University Hospital, Edegem, Belgium; ¹⁴Hospital Clínic De Barcelona, Barcelona, Spain; ¹⁵Chu Besançon, Besançon, France; ¹⁶Hôpital Charles Nicolle, Rouen, France; ¹⁷Hôpital Trousseau CHU de Tours, Tours, France; ¹⁸Centre Hospitalier Universitaire Sart Tilman, Liege, Belgium; ¹⁹CHU UCL Namur-Site de Godinne, Yvoir, Belgium; ²⁰Hôpital Erasme-Cliniques Universitaires de Bruxelles, Bruxelles, Belgium; ²¹University of Milan, Milano, Italy; ²²Hôpital Erasme-Cliniques Universitaires de Bruxelles, Clinique Saint-Luc Bouge, CHU UCL Namur-Site Godinne, Bruxelles, Belgium
Email: kazeedeline@gmail.com

Background and aims: Data on the natural history of patients with histological lesions of porto-sinusoidal vascular disorder (PSVD) who do not show signs of portal hypertension (PH) are scarce. We aimed to

describe the natural history of these patients and identify risk factors for progression.

Method: A retrospective study was conducted to collect health-related data on patients with a histopathological diagnosis of PSVD and no sign of PH. Patients with any sign of PH were excluded and a liver biopsy of at least 15 mm was required. Associated conditions were defined as the use of any medication or the presence of any disease that could be associated with PSVD. They were considered severe if they were potentially associated with reduced life expectancy. Main endpoints were the development of signs or complications of PH and death.

Results: 227 patients from 18 centers were included (92 men [41%], median age 46 years). Indication for liver biopsy was abnormal liver tests in 186 patients (82%). Histological findings were nodular regenerative hyperplasia, obliterative portal venopathy, incomplete septal cirrhosis, a combination of these histological lesions, or sinusoidal dilatation with or without other signs indicative of PSVD, in 126 (56%), 41 (18%), 2 (1%), 14 (6%) and 44 (19%) patients, respectively. Associated conditions were observed in 100 patients (46%), of which 55 (24%) were considered as severe. During a median follow-up of 37 months (95% CI: 27–46), 3 patients (1.3%) developed PH-related manifestations (ascites in 2 patients, esophageal varices in 1 patient). The cumulative incidence of PH-related manifestations at 10 years was 6% (95% CI: 2–20%). Nineteen patients (8%) died, 17 from non-hepatic causes and 2 from complications related to preexisting cholangiocarcinoma. No patient died from PH-related complications. Independent predictors of death were age (relative risk [RR]: 1.07, 95% CI: 1.03, 1.11, $p<0.001$) and presence of a severe associated condition (RR: 4.76, 95% CI: 1.55–14.65, $p=0.006$). The 10-year survival rate was 84% (95% CI: 77–92%); 95% (95% CI: 89–100) in patients without an associated condition, 100% in patients with a non-severe associated condition, and 58% (95% CI: 41–76) in patients with a severe associated condition ($p<0.001$).

Conclusion: Most patients with PSVD without PH will not develop it. Sixteen percent of patients die within 10 years, mostly from non-hepatic causes, and the risk of death is independently associated with the severity of the associated condition.

Immune mediated II

OS-086

Event-free survival of maralixibat-treated patients with progressive familial intrahepatic cholestasis compared with aligned real-world data of untreated patients from NAPPED

Pedro Miranda Afonso¹, Alise D.E. de Groot², Mark Nomden², Paola Mian², Benjamin Shneider³, Emmanuel Gonzalès⁴, Irena Jankowska⁵, Etienne Sokal⁶, Richard J. Thompson⁷, Andrea Goldstein⁸, Douglas B. Mogul⁸, Shannon Vandriel⁸, Seema Alam⁹, Cigdem Ankan¹⁰, Henrik Arnell¹¹, Madeleine Aumar¹², Amer Azaz¹³, Emna Barkaoui¹⁴, Maria Belen Pallitto¹⁵, Marisa Beretta¹⁶, Jernej Breclj¹⁷, Pier Luigi Calvo¹⁸, Mara Cananzi¹⁹, Huey-Ling Chen^{20,21,22}, Guillermo Costaguta^{23,24}, Elisa de Carvalho²⁵, Dominique Debray²⁶, Antal Dezsöfi-Gottl²⁷, Angelo Di Giorgio²⁸, Ozlem Durmaz²⁹, Alexandre Fabre³⁰, Sandra Ferreira³¹, Ryan T. Fischer³², Björn Fischler¹¹, Cristina Campos Gonçalves^{33,34}, Enke Grabhorn³⁵, Tassos Grammatikopoulos⁷, Girish Gupta³⁶, Winita Hardikar³⁷, Loreto Hierro Llanillo³⁸, Giuseppe Indolfi³⁹, Emmanuel Jacquemin⁴, Carolina Jimenez-Rivera⁴⁰, Marianne Hørby Jørgensen⁴¹, Agustina Kadaristiana⁷, Wikrom Karnsakul⁴², Nanda Kerkar^{43,44}, Kyung Mo Kim⁴⁵, Jae Sung Ko⁴⁶, Denitza Kofinova⁴⁷, Nolwenn Laborde⁴⁸, Patryk Lipiński⁴⁹, Rohan Malik⁵⁰, Saeed Mohammad⁵¹, Yael Mozer-Glassberg⁵², Gabriella Nebbia⁵³, Emanuele Nicastro⁵⁴, Felipe Ordoñez⁵⁵, Anna Orłowska-Wójcicka⁵, Arti Pawaria⁵⁶,