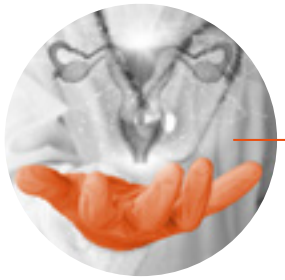

3.7.2 Molecular Hepatology Group

Publications: 8
Q1: 7

COMPOSITION

Paloma Jara Vega. Emérita Asistencial. Hospital Universitario La Paz

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María Ángeles Pajares Tarancón. Investigadora Científica. CSIC

Luiz Stark Aroeira. Investigador Postdoctoral. Hospital Universitario La Paz

David Vicent López. Investigador Senior (Contrato Miguel Servet - I2). Hospital Universitario La Paz

STRATEGIC OBJECTIVE

Our research interest is focused on the study of the molecular mechanisms underlying the most severe paediatric liver disorders, namely cholestasis, which results from the impaired secretion of bile from the liver to the intestine.

As such, it represents a clinical and biochemical syndrome that is produced by a wide variety of disease processes that affect the liver. Individuals with cholestasis manifest jaundice, severe itching, malabsorption of fats and lipidsoluble vitamins and, in many cases, progressive liver damage. These clinical manifestations are due to the accumulation in blood and tissues of substances normally secreted in the bile, such as bilirubin, bile acids, and cholesterol and to the absence of bile from the intestine.

When manifested in early infancy, cholestasis is often life threatening and usually requires liver transplantation. Extrahepatic biliary atresia (EHBA), Alagille syndrome and progressive familial intrahepatic cholestasis (PFIC) constitute the main paediatric cholestatic disorders. EHBA is an enigmatic disease of unknown aetiology, characterised by a precocious and accelerated obstruction of the biliary tree. Alagille syndrome is associated with mutations in the Jag1 gene and is characterised by a paucity or absence of intrahepatic bile ducts. PFIC encompasses a heterogeneous group of autosomal recessive diseases that exhibit similar clinical features. These diseases are caused by mutations in proteins located in the canalicular membrane of the hepatocyte and in proteins involved in bile secretion, such as the bile salt export pump (BSEP; ABCB11), the phospholipid transport protein MDR3 (ABCB4) and the aminophospholipid translocase FIC1 (ATP8B1). These cholestatic disorders constitute the most common indication for liver transplantation in childhood.

RESEARCH LINES

- Molecular basis of paediatric liver diseases.
- Liver Pathobiology.
- Biomarker identification.

RESEARCH ACTIVITY

Publications

- Baumann U, Karam V, Adam R, Fondevila C, Dhawan A, Sokal E, Jacquemin E, Kelly DA, Grabhorn E, Pawlowska J, D'Antiga L, Vega PJ, Debray D, Polak WG, de Goyet JD, Verkade HJ. Prognosis of children undergoing liver transplantation: a 30-year european study. Pediatrics. 2022; 150(4): e2022057424. Article. IF: 8; D1
- de Kleine RH, Lexmond WS, Buescher G, Sturm E, Kelly D, Lohse AW, Lenz D, Jorgensen MH, (Muñoz Bartolo G, Hierro LLanillo L). Severe acute hepatitis and acute liver failure of unknown origin in children: a questionnaire-based study within 34 paediatric liver centres in 22 European countries and Israel, April 2022. Euro Surveill. 2022; 27(19): 6-12. Article. IF: 19; D1
- Molera C, Sarishvili T, Nascimento A, Rtskhiladze I, Bartolo GM, Cebrian SF, Fernández JV, Cabello BM, Graham RJ, Miller W, Sepulveda B, Kamath BM, Meng H, Lawlor MW. Intrahepatic cholestasis is a clinically significant feature associated with natural history of X-linked myotubular myopathy (XLMTM): A case series and biopsy report. J Neuromuscul Dis. 2022; 9(1): 73-82. Article. IF: 3.3; Q2
- Montalva E, Rodríguez-Peralvarez M, Blasi A, Bonanad S, Gavin O, Hierro L, Llado L, Llop E, Pozo-Laderas JC, Colmenero J. Consensus statement on hemostatic management, anticoagulation, and antiplatelet therapy in liver transplantation. Transplantation. 2022; 106(6): 1123-31. Review. IF: 6.2; D1
- Platero L, García-Sánchez P, Sainz T, Calvo C, Iglesias I, Esperon F, de la Fuente R, Frauca E, Pérez-Martínez A, Méndez-Echevarría A. Pets for pediatric transplant recipients: To have or not to have. Front Vet Sci . 2022; 9: 974665. Article. IF: 3.2; D1
- Sánchez-Zapardiel E, Alos M, Nozal P, González-Muñoz M, Frauca-Remacha E, Gavilán LB, Quiles MJ, Hierro L, López-Granados E. Humoral and cellular immune responses to Pfizer-BioNTech BNT162b2 SARS-CoV-2 vaccine in adolescents with liver transplantation: Single center experience. Front Immunol. 2022; 13: 1049188. Article. IF: 7.3; Q1
- Semova I, Levenson AE, Krawczyk J, Bullock K, Gearing ME, Ling AV, Williams KA, Miao J, Adamson SS, Shin DJ, Chahar S, Graham MJ, Crooke RM, Hagey LR, Vicent D, de Ferranti SD, Kidambi S, Clish CB, Biddinger SB. Insulin prevents hypercholesterolemia by suppressing 12 alpha-hydroxylated bile acids. Circulation. 2022; 145(13): 969-82. Article. IF: 37.8; D1
- van Beek J, La Fraaij P, Giaquinto C, Shingadia D, Horby P, Indolfi G, Koopmans M, (Muñoz Bartolo G, Hierro LLanillo L). Case numbers of acute hepatitis of unknown aetiology among children in 24 countries up to 18 April 2022 compared to the previous 5 years. Euro Surveill. 2022; 27(19): 2-5. Article. IF: 19; D1

Research projects

Hierro Llanillo L. Desarrollo de métodos de diagnóstico molecular de enfermedades hepáticas infantiles de carácter hereditario. Fundación ACS. 2005-Ongoing.

Managment centre: FIBHULP

Hierro Llanillo L. Diagnóstico molecular de enfermedades hepáticas infantiles de carácter hereditario (PI-426). Fundación ACS. 2012-Ongoing.

Managment centre: FIBHULP

Hierro Llanillo L. Estableciendo una base de datos como parte de la red europea de referencia en enfermedades rara hepáticas. R-liver. UE. 2019-Ongoing.

Managment centre: FIBHULP

Hierro Llanillo L. Tomoxliver: estudio de la disfunción del hepatocito desde un abordaje multidisciplinar (S2017/BMD-3817). CM. 2018-2022.

Managment centre: FIBHULP

Hierro Llanillo L. Validación de marcadores genéticos y moleculares para el diagnostico de atresia biliar (PI20/01496). ISCIII. 2021-2023.

Managment centre: FIBHULP

Jara Vega P. Estudio epidemiológico, observacional sobre el riesgo de desarrollo de síndrome linfoproliferativo en pacientes pediátricos trasplantados hepáticos, en el HULP. Roche Farma S. A. 2008-Ongoing.

Managment centre: FIBHULP

Jara Vega P. PaEdiatric Transplantation European Registry (PETER PI-4389). UE. 2019-2023.

Managment centre: FIBHULP

Olveira A, Hierro Llanillo L. Impact of Wilson's disease on Spanish population: an observational study. Alexión Pharma Spain S.L. 2022-2022.

Managment centre: FIBHULP

Romero M, Hierro Llanillo L. Optimization of spleen VCTE examinations with FibroScan. Echosens. 2022-Ongoing.

Managment centre: FIBHULP

Ruiz de Valbuena R, Hierro Llanillo L. A prospective and retrospective cohort study to refine and expand the knowledge on patients with chronic forms of Acid Sphingomyelinase Deficiency (ASMD) (Estudio PIR16183). Sanofi . 2021-Ongoing.

Managment centre: FIBHULP

Vicent López D. Caracterización clínica del nuevo factor de riesgo cardiovascular trimetilamina-D-óxido en pacientes diabéticos obesos. Sección de Endocrinología y Nutrición Severo Ochoa. 2015-Ongoing.

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Vicent López D. Contrato Miguel Servet Categoria C (CES06/007). ISCIII. 2008-2025.

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Cibers and Retics

Hierro Llanillo L. ERN on Hepatological diseases (RARE-LIVER). EU. (31/12/2023). FIBHULP

Jara Vega P. ERN on Transplantation in children (TransplantChild). EU. (31/12/2023). FIBHULP