SCIENTIFIC SEMINAR

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Understanding liver biology to combat liver disease. Familial Cholestasis

Progressive familial intrahepatic cholestasis type 3 (PFIC3) is a severe and rare liver disease, affecting between 1 in 50,000 to 1 in 100,000 children. It is caused by mutations in the phosphatidylcholine transporter ABCB4 (MDR3), leading to intrahepatic accumulation of free bile acids and subsequent liver damage. PFIC3 typically manifests early in life, progresses rapidly, and carries a poor prognosis.

To better understand the pathogenesis of PFIC3, we conducted an integrated proteomics and phosphoproteomics study using human liver samples, followed by validation of key functional hypotheses in a PFIC3 mouse model and liver organoids. Given that liver transplantation remains the only therapeutic option, we also investigated the liver regeneration process to identify prognostic factors that could enhance patient monitoring after liver resection.

Our findings highlight the modulation of one-carbon metabolism as a critical factor and a potential indicator of hepatocyte differentiation and functional status. Collectively, these results provide insights into the mechanisms and protein mediators of chronic liver injury, which may aid in improving the follow-up and management of affected patients.

CIC bioGUNE
MEMBER OF BASQUE RESEARCH & TECHNOLOGY ALLIANCE
EXCELENCIA SEVERO OCHOA

Tuesday
September 16
Atrio 800
12.00H

BRTA
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