

PROGRAMME

Friday 12 May 2023

Introductory Remarks Plenary

08:00 – 08:10
Chairs:
Massimo PINZANI, *United Kingdom*
Mario STRAZZABOSCO, *United States*

General concepts Plenary

Chairs:
Chantal HOUSSET, *France*
Detlef SCHUPPAN, *Germany*

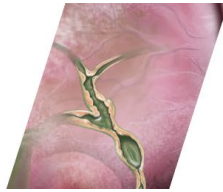
08:10 - 08:35 **A brief history of bile secretion and cholestasis**
James L. BOYER, *United States*

08:35 - 09:00 **The cholangiocyte in health and disease**
Mario STRAZZABOSCO, *United States*

09:00 - 09:25 **Biliary immunobiology in health and biliary disease**
Niklas BJÖRKSTRÖM, *Sweden*

09:25 - 09:50 **Cholangiocyte and hepatocyte adaption in cholestasis**
Michael TRAUNER, *Austria*

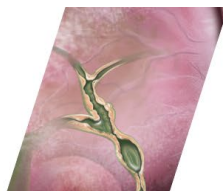
09:50 - 10:15 **Molecular pathways to fibrogenesis in cholangiopathies**
Massimo PINZANI, *United Kingdom*



ePoster session 1 and break

ePoster area

- | | |
|---------------|---|
| 10:15 – 10:45 | Chairs:
Chantal HOUSSET, <i>France</i> |
| 10:15 - 10:15 | Acute hepatocytic loss of endosomal sorting complex Retromer increases proliferation of biliary epithelial cells (BECs)
Markus BARBOSA, <i>Netherlands</i> |
| 10:15 - 10:15 | CRISPR-engineered chemically-derived hepatic progenitor cancer organoid recapitulates intrahepatic cholangiocarcinoma carcinogenesis and pathophysiology
Dongho CHOI, <i>Korea, Rep. of South</i> |
| 10:15 - 10:15 | Quantitative MRCP differentiates between malignant and benign biliary obstructions
Sarah FINNEGAN, <i>United Kingdom</i> |
| 10:15 - 10:15 | Characterizing and manipulating GPR35-dependent mast cell activity in PSC
Georg SCHNEDITZ, <i>Norway</i> |
| 10:15 - 10:15 | Necroptosis contributes to liver inflammation and fibrosis in experimental primary sclerosing cholangitis
Pierre-Antoine SORET, <i>France</i> |
| 10:15 - 10:15 | Multimodal decoding of the mesenchymal landscape of human biliary fibrosis
David WILSON, <i>United Kingdom</i> |
| 10:15 - 10:15 | Silencing CNNM4 in cholangiocarcinoma inhibits tumoral progression by means of non-canonical ferroptosis
Maria MERCADO-GÓMEZ, <i>Spain</i> |
| 10:15 - 10:15 | Harnessing immune dysregulation in the search for serum biomarkers for diagnosis and prognosis of primary sclerosing cholangitis
Lander HEYERICK, <i>Belgium</i> |



- 10:15 - 10:15 **Beta 7 integrin and L-selectin contribute to cholestatic liver disease in mice**
 Agnes SEEGER, *Germany*
- 10:15 - 10:15 **Dynamics of spleen stiffness measurement in patients with primary biliary cholangitis predict disease progression: a pilot study**
 Micol CITTONE, *Italy*

Fibrosing congenital cholangiopathies

Plenary

Chairs:
 Sophie LOTERSZTAJN, *France*
 Christian TRAUTWEIN, *Germany*

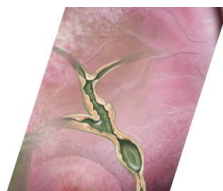
- 10:45 - 11:05 **Biliary development**
 Frédéric LEMAIGRE, *Belgium*
- 11:05 - 11:25 **Biliary atresia: Update on pathogenesis**
 Alexander MIETHKE, *United States*
- 11:25 - 11:45 **Unmet clinical needs and therapeutic strategies in biliary atresia: beyond Kasai and Liver Transplantation**
 Etienne SOKAL, *Belgium*
- 11:45 - 12:05 **Innate immunity and the gut liver axis in cholangiopathies: The case for cystic fibrosis related liver disease**
 Romina FIOROTTO, *United States*
- 12:05 - 12:25 **Targeting bile composition as a strategy for modifying cholestatic liver disease progression**
 Ulrich BEUERS, *Netherlands*

LUNCH FORUM: The non-invasive assessment of fibrosis in biliary diseases

Plenary

Chairs:
 Mette VESTERHUS, *Norway*

- 12:25 - 12:35 **Serum Markers**
 Diana LEEMING, *Denmark*



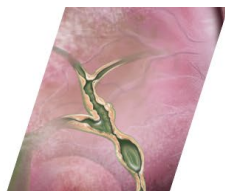
- 12:35 - 12:45 **Elastography**
Mette VESTERHUS, *Norway*
- 12:45 - 12:55 **Imaging**
Christoph SCHRAMM, *Germany*
- 12:55 - 13:25 **Panel Discussion**
Mette Vesterhus, Diana Leeming, Christoph Schramm

Selected oral presentations

Plenary

Chairs:
 Jesús M. BAÑALES, *Spain*
 Fabio MARRA, *Italy*

- 13:30 - 13:45 **Targeting epithelia-matrix interactions halts cyst growth in polycystic liver disease**
Scott WADDELL, *United Kingdom*
- 13:45 - 14:00 **Hepatic single-cell transcriptome in primary sclerosing cholangitis**
Oleksandr PETRENKO, *Austria*
- 14:00 - 14:15 **Dual alpha-v/beta-6 and alpha-v/beta-1 integrin inhibitors reduce TGF-beta signaling and fibrosis in multiple preclinical rodent models of biliary fibrosis**
Johanna SCHAUB, *United States*
- 14:15 - 14:30 **Immuno-oncology biomarkers for improved diagnosis of cholangiocarcinoma in primary sclerosing cholangitis: insights from the inflammatory and fibrotic tumour microenvironment**
Andres GARCIA SAMPEDRO, *United Kingdom*
- 14:30 - 14:45 **Osteopontin prevents excessive macrophage-mediated bile duct injury in experimental sclerosing cholangitis and correlates with liver injury in human patients**
Kevin DE MUYNCK, *Belgium*
- 14:45 - 15:00 **The planar cell polarity pathway as a master regulator of terminal biliary morphogenesis during liver development**
Michaela RAAB, *United Kingdom*

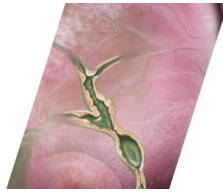


- 15:00 - 15:15 **Liver transcriptome analysis reveals PSC-specific gene signature in biliary fibrosis**
Alena LASCHTOWITZ, *Germany*
- 15:15 - 15:30 **Quantitative MRCP metric to distinguish IgG4-sclerosing cholangitis from primary sclerosing cholangitis**
Emmanuel SELVARAJ, *United Kingdom*

ePoster session 2 and break

ePoster area

- 15:30 – 16:00 **Chairs:**
Marco CARBONE, *Italy*
- 15:30 - 15:30 **Liquid biopsy protein biomarkers of cholangiocarcinoma risk, early diagnosis and survival mirroring tumor cells**
Ainhoa LAPITZ, *Spain*
- 15:30 - 15:30 **Functional dichotomy of biliary primary cilia in liver injury and cholangiocarcinoma development**
Geoff MCCAUGHAN, *Australia*
- 15:30 - 15:30 **Hematopoietic-biliary organoids as a model for liver development and injury**
Susanna QUACH, *Germany*
- 15:30 - 15:30 **Identification of the role of osteopontin in biliary atresia**
Natalia NIETO, *United States*
- 15:30 - 15:30 **Transcriptomic risk score predicts response to obeticholic acid and identifies genes linked with metabolic remodelling, autophagy, senescence and fibrosis in patients with PBC**
Hussain SYED, *Canada*
- 15:30 - 15:30 **CD8+ T cells from patient with primary biliary cholangitis express e-cadherin upon T cell receptor activation and invade biliary epithelial cells through e-cadherin-beta-catenin interaction**
Vincenzo RONCA, *United Kingdom*



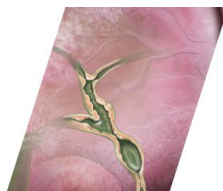
- 15:30 - 15:30 **Mitochondrial dynamics and autophagy are disturbed in experimental models of primary biliary cholangitis, contributing to disease pathogenesis**
 Irune LASA, *Spain*
- 15:30 - 15:30 **Identification of a functional relationship between MEK5-ERK5 pathway and hypoxia inducible factor 1-alfa**
 Alessio MENCONI, *Italy*
- 15:30 - 15:30 **Primary cilia as a targetable node between biliary injury, senescence and regeneration in liver transplantation**
 Sofia FERREIRA-GONZALEZ, *United Kingdom*
- 15:30 - 15:30 **Neutrophils in the pathogenesis of primary sclerosing cholangitis**
 Nidhi JALAN SAKRIKAR, *United States*

Morphogenetic signaling and biliary fibrosis

Plenary

Chairs:
 Christoph SCHRAMM, *Germany*
 Ana LLEO, *Italy*

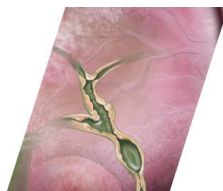
- 16:00 - 16:20 **Biliary stem cells in health and biliary disease**
 Vincenzo CARDINALE, *Italy*
- 16:20 - 16:40 **Morphogenetic signaling in biliary repair and fibrosis**
 Luca FABRIS, *Italy*
- 16:40 - 17:00 **Alagille Syndrome and the morphogenetic role of Notch signaling in development and liver disease: Translational opportunities?**
 Emma ANDERSSON, *Sweden*
- 17:00 - 17:20 **May the targeting of cholangiocyte signaling be useful in the clinical setting? The case of Polycystic and Fibropolycystic liver diseases**
 Jesús M. BAÑALES, *Spain*



ePoster session 3 and break

ePoster area

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|---------------|---|
| 17:20 – 17:50 | Chairs:
Emma ANDERSSON, <i>Sweden</i> |
| 17:20 - 17:20 | An organotypic model of ductular reaction reveals a role of lipid biosynthesis in biliary cell activation
Giovanni SORRENTINO, <i>Italy</i> |
| 17:20 - 17:20 | Metabolic rewiring of cystic cholangiocytes improves mitochondrial dynamics and bioenergetics ameliorating the pathogenesis of polycystic liver disease in experimental models
Enara MARKAIDE, <i>Spain</i> |
| 17:20 - 17:20 | Large CD103+ CD69+ CD8+ T cells consistently invade biliary epithelial cells in patients with primary biliary cholangitis
Scott DAVIES, <i>United Kingdom</i> |
| 17:20 - 17:20 | Targeting OX40 signalling to modulate regulatory T cells and reduce biliary fibrosis
Wei-Yu LU, <i>United Kingdom</i> |
| 17:20 - 17:20 | Combined inhibition of bile salt synthesis and intestinal bile salt uptake ameliorates cholestatic liver damage and reduces colonic bile salt levels in mice
Stan VAN DE GRAAF, <i>Netherlands</i> |
| 17:20 - 17:20 | Beta-caryophyllene a CB2 specific agonist prevents the development of hepatopulmonary syndrome in experimental model of biliary cirrhosis
AKASH KUMAR MOURYA, <i>India</i> |
| 17:20 - 17:20 | Patient proteomic data and mouse model reinforce the proinflammatory role of CCL24 in cholestatic disease
Matthew FRANKEL, <i>Israel</i> |
| 17:20 - 17:20 | Non-invasive evaluation of progression to primary biliary cholangitis in patients with positive antimitochondrial antibodies
Marta ALONSO-PEÑA, <i>Spain</i> |



- 17:20 - 17:20 **Primary sclerosing cholangitis with features of autoimmune hepatitis: phenotypic characterization and prognosis**
Mirjam KOLEV, *Switzerland*
- 17:20 - 17:20 **A3907, a systemic ASBT inhibitor, improves cholestasis and biliary fibrosis in mice by multi-organ activity and shows translational relevance to human**
Francisco J. CABALLERO, *Spain*

PBC and PSC and cholangiocarcinoma

Plenary

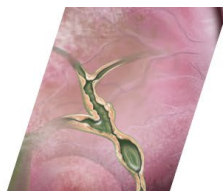
Chairs:
Tom Hemming KARLSEN, *Norway*
Chiara RAGGI, *Italy*

- 17:50 - 18:15 **The past and the present of PBC and PSC treatment: where are we going?**
David N. ASSIS, *United States*
- 18:15 - 18:40 **Cellular senescence in biliary diseases**
Stuart J. FORBES, *United Kingdom*
- 18:40 - 19:05 **Targeting ductular reactive cells as an anti-fibrotic strategy in biliary fibrosis**
Gregory GORES, *United States*
- 19:05 - 19:25 **Immune cell infiltration and fibrosis in PSC**
Christoph SCHRAMM, *Germany*

ePoster session 4 and networking reception

ePoster area

- 19:25 – 19:55
Chairs:
Romina FIOROTTO, *United States*
- 19:25 - 19:25 **Chronic ketamine-induced sclerosing cholangitis associated with early inflammatory bowel disease after long term topical use**
Gerond LAKE-BAKAAR, *United States*
- 19:25 - 19:25 **Mice lacking the NO receptor develop biliary fibrosis and become worsen upon feeding Western diet**
Muhammad ASHFAQ-KHAN, *Germany*



- 19:25 - 19:25 **Thy-1 expression in liver histology correlates with fibrosis grading in patients with primary biliary cholangitis**
Mirjam KOLEV, *Switzerland*
- 19:25 - 19:25 **Primary biliary cholangitis symptoms are reduced in M_cpip1f1/f1AlbCre mice after pharmacological treatment**
Katarzyna TRZOS, *Poland*
- 19:25 - 19:25 **Mertk-expressing macrophages promote the malignant features of cholangiocarcinoma cells**
Mirella PASTORE, *Italy*
- 19:25 - 19:25 **Biliary atresia human cholangiocyte organoids demonstrate increased oxidative stress response**
Yara HAMODY, *Israel*
- 19:25 - 19:25 **Oval cells as new players of the inflammatory response during cholestatic liver damage: role of the EGFR pathway**
Aránzazu SÁNCHEZ, *Spain*
- 19:25 - 19:25 **Radiomics features for risk stratification in primary sclerosing cholangitis: a proof-of-concept study**
Laura CRISTOFERI, *Italy*
- 19:25 - 19:25 **Bile duct injury in mdr2 knockout mice can be repaired with n-acetylcysteine treatment**
Adi HAR-ZAHAV, *Israel*
- 19:25 - 19:25 **Increased protein tyrosine phosphatase non-receptor type 2 activity in primary sclerosing cholangitis**
Madita DETERMANN, *Switzerland*

ePoster session 5 and networking reception

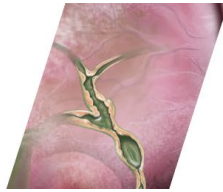
ePoster area

19:55 – 20:25

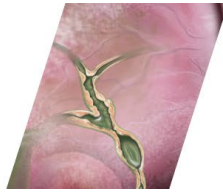
Chairs:
Marco CARBONE, *Italy*

19:55 - 19:55

Long-term treatment with fibrates and ursodeoxycholic acid in people with primary sclerosing cholangitis is safe and associated with persistent clinical and biochemical improvement
Matteo PEVIANI, *Italy*



- 19:55 - 19:55 **scRNA transcriptomics analysis of cellular cross talk in fibrocystin-defective mice reveals a central role for cholangiocytes, neutrophils and double-negative T cells**
Zehra SYEDA, *United States*
- 19:55 - 19:55 **Claudin-1 is a therapeutic target for primary sclerosing cholangitis**
Fabio DEL ZOMPO, *France*
- 19:55 - 19:55 **Dimethyl-fumarate attenuates cholestatic liver injury in mice**
Ester DOHNALKOVA, *Czech Republic*
- 19:55 - 19:55 **The tight junction protein jam-a is crucial for biliary pathophysiology in mice**
Grégory MERLEN, *France*
- 19:55 - 19:55 **Recurrence of primary sclerosing cholangitis after liver transplantation: a single center data**
Elisa CATANZARO, *Italy*
- 19:55 - 19:55 **Hepatic recompensation upon treatment with ursodeoxycholic acid in PBC patients with decompensated cirrhosis**
Lukas BURGHART, *Austria*
- 19:55 - 19:55 **Liver re-transplantation for recurrent primary sclerosing cholangitis: how far we can go?**
Chiara MAZZARELLI, *Italy*
- 19:55 - 19:55 **Biliary senescence affects the transcriptomic landscape of murine hepatocytes**
Marisa F. FERREIRA, *United Kingdom*
- 19:55 - 19:55 **Extracellular matrix remodeling in the progression of primary sclerosing cholangitis to cholangiocarcinoma: insights from an animal model**
Massimiliano CADAMURO, *Italy*



Saturday 13 May 2023

State of the Art Lecture: Gut-liver interactions in biliary diseases

Plenary

Chairs:
Michael TRAUNER, *Austria*

08:30 - 09:00

State of the Art Lecture: Gut-liver interactions in biliary diseases
Tom Hemming KARLSEN, *Norway*

New models and therapeutic approaches to biliary fibrosis

Plenary

Chairs:
Stuart J. FORBES, *United Kingdom*
Verena KEITEL-ANSELMINO, *Germany*

09:00 - 09:20

New cellular models/organoids
Fotios SAMPAZIOTIS, *United Kingdom*

09:20 - 09:40

The matrisome in biliary fibrosis
Giuseppe MAZZA, *United Kingdom*

09:40 - 10:00

Modulation of nuclear receptors
Saul J. KARPEN, *United States*

10:00 - 10:20

Clinical end-points in the treatment of cholangiopathies
Cyriel PONSIOEN, *Netherlands*

10:20 - 10:40

Use of Single cell transcriptomics to dissect the mesenchymal cell niche
Neil HENDERSON, *United Kingdom*

10:40 - 11:00

Role of Cholangiocyte derived Wnts during cholestatic injury
Vikas GUPTA, *United States*



Coffee break

ePoster area

11:00 – 11:25

Round table with industry: Future therapies in biliary fibrosis

Plenary

11:25 - 12:55

Chairs:
Detlef SCHUPPAN, *Germany*

Panel Discussion

Giuseppe Mazza, Scott Turner, Tariq Sethi, Bernard Allan, Thomas Wynn,
Matthew Frankel

Conclusions and adjourn

Plenary

12:55 – 13:00

Chairs:
Massimo PINZANI, *United Kingdom*
Mario STRAZZABOSCO, *United States*