

TAKING CPSS TO THE NEXT LEVEL

2ND MEETING OF EXPERTS

July 1-2, 2022

Onsite & online congress

Centre Médical Universitaire (CMU), Geneva



Taking CPSS to the next level

2nd meeting of experts

Context

The management of congenital portosystemic shunts (CPSS) is challenging.

Aims

To improve patient care by addressing controversies in CPSS management focusing on liver nodules, portopulmonary hypertension (POPH), endocrine abnormalities, and the challenges of recommending shunt closure.

Public

Professionals (physicians, scientists, patient organisations, allied health professionals)

Language

All presentations will be in English.

Type of congress

Onsite and online (hybrid meeting)

Organizing committee

Dr. Anna Baiges, Prof. Maurice Beghetti, Dr. Ana Maria Calinescu-Tuleasca, Prof. Andrea De Gottardi, Prof. Stéphanie Franchi-Abella, Prof. Florent Guérin, Dr. Solène Le Cam, Dr. Josefina Martinelli, Prof. Valérie McLin, Dr. Aurélie Plessier, Prof. Tudor Pop, Prof. Laurent Savale, Prof. Hubert Van der Doef, Dr. Julie Wacker, Prof. Barbara Wildhaber

Scientific committee

Prof. Maurice Beghetti, Dr. Lorenzo D'Antiga, Prof. Dominique Debray, Prof. Stéphanie Franchi-Abella, Prof. Emmanuel Gonzales, Prof. Florent Guérin, Dr. Virginie Lambert, Dr. José Lipsich, Prof. Periklis Makrythanasis, Prof. Valérie McLin, Prof. Laurent Savale, Prof. Marie-Victoire Senat, Dr. Frank Van Steenbeek, Prof. Barbara Wildhaber

Program

Friday July 1, 2022

8:30 Welcome & Introduction

[Prof. Stéphanie Franchi-Abella](#), Pediatric radiology, Paris-Saclay University, Hôpital Bicêtre, Hôpitaux Paris-Saclay APHP, France; [Prof. Valérie McLin](#), Swiss Pediatric Liver Center, Gastroenterology, hepatology and pediatric nutrition unit, University Hospitals of Geneva, Geneva, Switzerland

I. Refresher

8:35 Present day understanding of presentation, diagnosis and management

[Prof. Emmanuel Gonzales](#), Pediatric Hepatology and Liver Transplantation Unit, Hôpital Bicêtre, Reference Centre for Rare Pediatric Liver Diseases, FILFOIE, ERN RARE LIVER, FHU Hepatinov, INSERM UMR-S 1193, AP-HP. Université Paris-Saclay, Le Kremlin-Bicêtre, France

8:55 Survey on current practices

[Mehdi Gadiri](#), Faculty of Medicine, University of Geneva, Geneva, Switzerland; [Dr. Josefina Martinelli](#), Swiss Pediatric Liver Center, Gastroenterology, hepatology and pediatric nutrition, University Hospitals of Geneva, Geneva, Switzerland

9:05 Update on the International Registry of Congenital Porto-Systemic Shunts (IRCPSS)

[Simona Korff PhD](#), Swiss Pediatric Liver Center, Gastroenterology, hepatology and pediatric nutrition unit, University Hospitals of Geneva, Geneva, Switzerland

II. Personalizing Care

9:15 Prenatal diagnosis and neonatal management: "wait and see" versus invasive management

[Dr. Grégoire Dumery](#), Gynécologie-Obstétrique, Hôpital Bicêtre, Paris, France; [Prof. Stéphanie Franchi-Abella](#), Pediatric Radiology, Paris-Saclay University, Hôpital Bicêtre, Hôpitaux Paris-Saclay APHP, Paris, France

Friday July 1, 2022

9:35 Late diagnosis of a congenital extrahepatic portosystemic shunt with multiple liver nodules

[Prof. Tudor Lucian Pop](#), University of Medicine and Pharmacy "Iuliu Hatieganu" Cluj-Napoca, Romania

9:45 Coffee break

III. Liver Nodules

Moderators: [Dr. Anne-Laure Rougemont](#), Swiss Pediatric Liver Center, Pediatric pathology, University Hospitals of Geneva, Geneva, Switzerland; [Prof. Barbara Wildhaber](#), Swiss Pediatric Liver Center, Pediatric surgery, University Hospitals of Geneva, Geneva, Switzerland

10:15 Imaging characteristics

[Prof. Valérie Vilgrain](#), Radiology, Hôpital Beaujon, Paris, France

10:35 Histopathology and molecular biology/role of biopsy

[Prof. Alberto Quaglia](#), Department of Cellular Pathology, Royal Free London NHS Foundation Trust/UCL Cancer Centre, London, England

Whom to resect:

11:05 French pediatric experience

[Prof. Florent Guérin](#), Pediatric Surgery Unit, Université Paris-Saclay, Assistance Publique Hôpitaux de Paris, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

11:15 Japanese pediatric experience

[Dr. Hajime Uchida](#), Organ Transplantation Center, National Center for Child Health and Development, Tokyo, Japan

11:25 Experience in adults

[Prof. Nigel Heaton](#), Professor of Transplantation, Consultant in adult and pediatric liver transplantation and HPB surgery, Kings College Hospital, London, England

11:35 Discussion

12:15 Lunch

IV. Portopulmonary Hypertension

Moderators: [Prof. Maurice Beghetti](#), Pediatric cardiology, University Hospitals of Geneva, Geneva, Switzerland; [Dr Frédéric Lador](#), Service de Pneumologie, University Hospitals of Geneva, Geneva, Switzerland; [Prof. Laurent Savale](#), Department of Respiratory and Intensive Care Medicine, Pulmonary Hypertension National Referral Centre, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

13:30 Pathophysiology of pulmonary vascular diseases induced by porto-systemic shunts

[Prof. Laurent Savale](#), Department of Respiratory and Intensive Care Medicine, Pulmonary Hypertension National Referral Centre, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

13:50 Role of BMP9 in the pathophysiology of hepatopulmonary syndrome

[Fabien Robert](#), INSERM UMR_S 999, Pulmonary Hypertension: Pathophysiology and Novel Therapies, Hôpital Marie Lannelongue, Le Plessis Robinson, France

14:00 Cardiovascular disorders and CPSS

[Dr. Virginie Lambert](#), Institut mutualiste Montsouris, Paris, France; APHP, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

14:20 How to screen for and confirm the diagnosis of POPH?

[Dr. Sebastien Hascoet](#), Pediatric and Adult Congenital Heart Diseases, Hôpital Marie Lannelongue, Le Plessis-Robinson, France

14:40 Who, when and how to treat POPH

[Dr. Julie Wacker](#), Pediatric cardiology unit, University Hospitals of Geneva, Geneva, Switzerland; Centre Universitaire Romand de Cardiologie et Chirurgie Cardiaque Pédiatrique, University of Geneva and Lausanne, Switzerland

15:00 Liver or lung transplantation. Which indications for which patients?

[Prof. Olivier Sitbon](#), Department of Respiratory and Intensive Care Medicine, Pulmonary Hypertension National Referral Centre, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

15:20 Two clinical cases:

A case of POPH worsening after CPSS closure

[Dr. Audrey Baron](#), Department of Respiratory and Intensive Care Medicine, Pulmonary Hypertension National Referral Centre, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

A case of POPH improvement after CPSS closure

[Dr. Julie Wacker](#), Pediatric cardiology unit, University Hospitals of Geneva, Geneva, Switzerland; Centre Universitaire Romand de Cardiologie et Chirurgie Cardiaque Pédiatrique, University of Geneva and Lausanne, Switzerland

15:30 Discussion

16:00 Coffee break

V. Controversies Prior to Closure

Moderators: [Dr. Valeria Casotti](#), Ospedale Pediatric Hepatology, Gastroenterology and Transplant Centre, ASST Ospedale Papa Giovanni XXIII, Bergamo, Italy; [Prof. Valérie McLin](#), Swiss Pediatric Liver Center, Gastroenterology, hepatology and pediatric nutrition unit, University Hospitals of Geneva, Geneva, Switzerland

16:30 Role of liver biopsy: debate

[Prof. Dominique Debray](#), Unité d'Hépatologie pédiatrique et transplantation, Centre de référence de l'AVB et cholestases génétiques, Centre de compétences des hépatites auto-immunes, cholangites sclérosantes, maladies vasculaires du foie et maladie de Wilson, Hôpital Necker, Paris, France

16:40 Discussion

16:50 Genetics: toward the development of a shunt-ome

[Prof. Periklis Makrythanasis](#), Assistant Professor, Medical School, National and Kapodistrian University of Athens, Athens, Greece; [Dr. Frank G. Van Steenbeek PhD](#), Assistant Professor, Utrecht University, faculty of veterinary medicine, The Netherlands

17:10 Discussion

17:20 Ethical aspects of preemptive closure and patient/proxies perspectives

[Dr. Nicolas Foureur](#), médecin responsable du Centre d'éthique clinique de l'AP-HP, Hôpital Cochin, Paris, France

18:10 Discussion

20:00 Dinner at Restaurant Café Papon

Rue Henri-Fazy 1, 1204 Geneva



Program

Saturday July 2, 2022

VI. Endocrinology of the Liver and Consequences of Portosystemic Bypass in Children and Adults

Moderators: [Dr. Hubert van der Doef](#), MD PhD, Pediatric hepatologist and gastroenterologist, University Medical Center Groningen, Groningen, the Netherlands; [Dr. Béatrice Kuhlmann](#), Pediatric Endocrinology, Cantonal Hospital Aarau KSA, Aarau, Switzerland

8:30 Overgrowth and CPSS

[Dr. Atessa Bahadori](#), DFEA, University Hospitals of Geneva, Geneva, Switzerland

8:45 Clinical case of severe hypoglycemia revealing syndromic CPSS

[Dr. Solène Le Cam](#), Pediatric radiology, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

8:50 Hypoglycemia, hyperinsulinism and CPSS

[Dr. Mirjam van Albada](#), endocrinologist UMCG, Groningen, the Netherlands

9:05 Adrenal insufficiency and CPSS

[Dr. Moinak Sen Sarma](#), Pediatric gastroenterology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India

9:20 Hypothyroidism and CPSS

[Dr. Nitash Zwaveling](#), endocrinologist Amsterdam UMC, Amsterdam, the Netherlands

9:35 Clinical case of amenorrhea and hyperandrogenism

[Dr. Jai Patel](#), interventional radiologist, Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom

9:40 Discussion

10:30 Coffee break

Saturday July 2, 2022

VII. Controversies in the Management of CPSS

Moderators: Prof. Nigel Heaton, Professor of Transplantation, Consultant in adult and pediatric liver transplantation and HPB surgery, Kings College Hospital, London, England; Dr Alexis Ricoeur, Unité de radiologie abdominale et interventionnelle, Département diagnostic, University Hospitals of Geneva, Geneva, Switzerland

11:00 Preoperative imaging workup

Dr. Hajime Uchida, Organ Transplantation Center, National Center for Child Health and Development, Tokyo, Japan

11:20 Contraindications to closure

Dr. Riccardo A. Superina, Transplant and Hepatobiliary Surgery, Robert E. Schneider Chair in Transplantation Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, United States

11:40 Timing and preemptive management

Prof. Jean De Ville de Goyet, Pediatric abdominal surgery and liver transplantation, ISMETT, Palermo, Italy

12:00 Complications of closure and their management

Dr. Paolo Marra, Department of Radiology, Papa Giovanni XXIII Hospital, Bergamo, Italy - School of Medicine and Surgery, University of Milano-Bicocca, Milano, Italy

Outcomes following closure:

12:20 Predictors of post-closure portal hypertension

Prof. Stéphanie Franchi-Abella, Pediatric radiology, Paris-Saclay University, Hôpital Bicêtre, Hôpitaux Paris-Saclay APHP, Paris, France

12:35 Long term follow up after symptomatic/asymptomatic shunt closure (children/adults)

Dr. Anna Baiges, Hepatology, Hospital Clínic de Barcelona, Barcelona, Spain

12:55 Lunch or lunch box sessions

VIII. Challenging Case Presentations and Discussion

13:45 Pre-selected illustrative abstracts

Moderators:

Dr. Anna Baiges, Hepatology, Hospital Clínic de Barcelona, Barcelona, Spain

Dr. Ana Maria Calinescu-Tuleasca, Swiss Pediatric Liver Center, Pediatric Surgery, University Hospitals of Geneva, Geneva, Switzerland

Dr. Solène Le Cam, Pediatric radiology, Hôpital Bicêtre, Le Kremlin Bicêtre, Paris, France

Dr. Josefina Martinelli, Swiss Pediatric Liver Center, Gastroenterology, hepatology and pediatric nutrition unit, University Hospitals of Geneva, Geneva, Switzerland

Dr. Julie Wacker, Pediatric cardiology unit, University Hospitals of Geneva, Geneva, Switzerland; Centre Universitaire Romand de Cardiologie et Chirurgie Cardiaque Pédiatrique, University of Geneva and Lausanne, Switzerland

Speakers:

Prof. Ron Bardin, Helen Schneider's Hospital for Women, Rabin Medical Center, Petach Tikva, Israel

Dr. Debalina Chakrabati, Avadh Hospital and Heart Center, Lucknow, India

Dr. Caroline Lemoine, Ann and Robert Lurie Children's Hospital of Chicago, Chicago, USA

Dr. Valeria Casotti, Hospital Papa Giovanni XXII, Bergamo, Italy

Dr. Vittoria Camelli, Regina Margherita Children's Hospital, Torino, Italy

Dr. Vanessa Steger, Swiss Pediatric Liver Center, Geneva, Switzerland

IX. Wrap Up

16:00 *Prof. Valérie McLin, Swiss Pediatric Liver Center, Gastroenterology, hepatology and pediatric nutrition unit, University Hospitals of Geneva, Geneva, Switzerland; Prof. Barbara Wildhaber, Swiss Pediatric Liver Center, Pediatric surgery, University Hospitals of Geneva, Geneva, Switzerland*

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Practical information



INFO

Post-graduate credits

SSPC: 12

SSP/SGP: 15

SGG/SSG: 16

SGR/SSR: 2

Venue

Centre Médical Universitaire (CMU),
Auditorium Alex-F. Müller (A250)
Avenue de Champel 7, 1205 Genève

Contact

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