

Agenda

All Times UK (BST)

Talks with a final have been pre-recorded

14:00-14:10 Welcome

David Hartley, the XLP Research Trust, UK

XLP1

14:10 – 14:30 Aplastic anaemia as an initial manifestation in a patient with X-linked lymphoproliferative disease type 1

Asst. Prof. Akihiro Hoshino Tokyo Medical and Dental University, Japan

14:30 – 14:50 Cheating Death: Mechanisms of Apoptosis Resistance in SAP-deficient T cells, and Consequences for XLP-1

Dr. Andrew Snow Uniformed Service University of the Health Sciences, Bethesda, USA

14:50 – 15:10 Diacylglycerol kinase activity regulation by SAP and WASp

Dr. Suresh Velnati Institute for Research and Cure of Autoimmune Diseases, University of Piemonte Orientale, Italy



15:20-15:40 SAP and Diacylglycerol kinase alpha (DGK α) control TCR expression at cell surface by regulating its trafficking.

Prof. Andrea Graziani Molecular Biotechnology Center, University of Torino, Italy

15:40 – 16:00 Allosteric SHP2 inhibition in XLP patient T cells rescues humoral and cytotoxicity defects, *in vitro*.

Dr. Neelam Panchal UCL Great Ormond Street Institute of Child Health, London, UK

16:00 – 16:20 Safety and pharmacokinetics of an EBNA1 inhibitor for the treatment of XLP- associated disease

Dr. Troy Messick The Wistar Institute, Philadelphia, USA

16:20 – 16:40 Update on progress towards a clinical trial for T cell gene therapy in XLP

Dr. Claire Booth & Dr. Amairelys Barroeta
UCL Great Ormond Street Institute of Child Health, London, UK

16:40 – 17:00 Excellent outcomes for adults following allogeneic stem cell transplantation for primary immunodeficiencies for XLP and XIAP

Thomas Fox

UCLH and UCL Institute of Immunity and Transplantation, London, UK

17:00 to 17:10 *Comfort Break*



XIAP

17:10 – 17:30 Immune phenotyping of XIAP deficient patients

Dr. Lynn Wong Institute of Experimental Immunology, University of Zurich, Switzerland

17:30 – 17:50 XIAP and inflammatory cell death in innate immunity



Dr. Monica Yabal Institute of Molecular Immunology & Experimental Oncology, Technical University München, Germany

17:50 – 18:10 Long term outcomes for adults with X-linked inhibitor of apoptosis (XIAP) Deficiency: What do we need to know?

Prof. Siobhan Burns Institute of Immunity and Transplantation, London, UK

18:10 – 18:30 Inflammatory bowel disease associated with XIAP deficiency can be cured by allogenic hematopoietic cell transplantation

Dr. Shintaro Ono Tokyo Medical and Dental University, Japan

18:30 – 18:40 Wrap up, thanks and future events

David Hartley, The XLP Research Trust, UK

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Thank you

