A EUROPEAN REGISTRY OF CHILDREN WITH HENOCH SCHOENLEIN NEPHRITIS TO DETECT CLINICAL, GENETIC AND IMMUNOLOGICAL RISK FACTORS

RATIONALE

Henoch-Schönlein purpura (HSP) is the most common vasculitis in children, with variable extrarenal signs and variable renal involvement, conditioning long term prognosis. The ESPN-ERA-EDTA Registry has recently reported a stable incidence of children undergoing renal replacement treatment because of HSP nephritis.

A new study investigating outcome of recent cases of HSP in a pan-European cohort to detect risk factors for progression has been launched and granted by ESPN, the ideal setting for a large scale multifaceted approach taking also advantage from the collaboration with Columbia University (NY, USA) for GWAS studies addressed to explore the genetic variants predisposing to primary IgAN and HSP IgAN.

This project is articulated in 2 parts:

a) HSP NEPHRITIS REGISTRY: a retrospective data-base collecting clinical data in European children with HSP presented in the last 20 years.

aims: identification of a European retrospective cohort for the search of

- clinical risk factors
- pathology risk factor
- genetic conditioning in a subgroup of subjects

b) PROSPECTIVE STUDY FOR VALIDATION OF IMMUNOLOGIC RISK FACTORS: on a selected cohort of 50 incident children with HSP nephritis a immunological panel of studies will be performed at time of renal biopsy together with the genetic study

aims: collection of a new incident cohort of children with HSP nephrits to study the immunological asset before treatment evaluating

- immunoproteasome switch
- TLR expression
- altered glycosylation of IgA1 and antibodies against deGAl IgA1
- complement activation
- oxidative stress markers (AOPP)

OUTCOME AND RELEVANCE

We expect a great impact of the outcomes of this proposal.

The scientific power and the cooperation capacity of the European Society for Paediatric Nephrology will face with the challenging aim of organizing a complete data-base of children with HSP nephritis, with full clinical and histological data at onset, at renal biopsy and during follow-up, finalized to evaluate the long term outcome of these children and enrolling patients with HSP for immunologic studies. This cooperative effort will be a strong European contribution for the study of genetic conditioning of onset and progression of the disease.

If you are interested in joining this challenging project and receiving the whole protocol with detailed enrolment instructions you can contact:

Licia Peruzzi

Regina Margherita Children's Hospital
Torino, Italy

e-mail: licia.peruzzi@unito.it