Projects launched in 2014

1) Joint activity ESPN and ERA-EDTA Working Groups on immune mediated renal diseases

At the end of March 2014 a renal biopsy survey was sent to Members of the two Societies in order to gather a broad overview of clinical indications and technical details for renal biopsy in children (and in adults) with glomerular diseases on native or on transplanted kidneys.

The questionnaire was divided into 5 parts

Part 1: Information on the corresponding Nephrologist Part 2: Questions for Centers performing renal biopsies in adults Part 3: Questions for Centers performing renal biopsies in children Part 4: Questions for Centers performing renal biopsies in grafted kidneys (adults and children) Part 5: Post biopsy survey

At September 1th , 2014, a total of 597 responses were gathered for all over Europe and 228 from Pediatric Nephrology Centers. The expected results of a European survey and also from the non EU Mediterranean were reported in Porto 2014 at the parallel meeting of the WG. The material gathered is highly suitable for a publication in 2015.

2) Study proposal in collaboration with Columbia University: GWAS in children with Henoch-Schoenlein Purpura (HSP) and primary IgA nephropathy (IgAN) . Recent studies from A.Gharavi and K.Kyryluck defined IgAN as an autoimmune trait of complex architecture with a strong genetic determination. Several common genetic variants were found to predispose to IgAN in adults through a large-scale genome-wide association study (GWAS). More recently, a strong relationship between the number of GWAS risk alleles and the age of disease onset was discovered, suggesting that pediatric patients with IgAN may carry a significantly higher burden of genetic risk factors. Accordingly, we are now extending our genetic investigations to children affected by IgAN and Henoch-Schönlein purpura without or with nephritis. The recruitment of patients will consist of a single blood draw and provision of basic clinical and biopsy data.

3) The WG has successfully submitted an application to the first Research call of ESPN presenter Licia Peruzzi on behalf of the ESPN WG of Immune mediated renal disorders.

The Board of ESPN WG J. C. Davin, R. Topaloglu, E. Baskin, C. Pecoraro and the WG coordinator A. Amore, will take active part in following the development of this project.

The project represents an expansion, with completely new initiatives, of the ongoing project of the Immune mediated renal disorders WG 2014 programme (point n2), which was based on a cooperation between ESPN and the Genetics at Columbia University, NY, USA. The project was developed among the Board and collaborators of the ESPN WG in strict collaboration with the ERA-EDTA Immunonephrology WG and open to collaboration with EULAR (European League Against Rheumatism)

## TITLE: A EUROPEAN REGISTRY OF CHILDREN WITH HENOCH SCHONELEIN NEPHRITIS TO DETECT CLINICAL, GENETIC AND IMMUNOLOGICAL RISK FACTORS

There is a need of a new study investigating outcome of recent cases of HSP in a pan-European cohort and there is a need to detect risk factors for progression.

This proposal is articulated into 2 parts:

a) A retrospective data-base to build up a Disease Registry correlated with search for genetic risk factors: data from children with HSP nephritis (done on the appropriate forms which were used for the recent VALIGA: validation study of the Oxford classification) is aimed at investigating, in European children with HSP presented in the last 20 years

Clinical presentation Pathology features classified according to ISKD and Oxford classification Updated outcome Pathology risk factors Clinical risk factors Genetic conditioning (in connection with the Columbia University GWAS)

b) A prospective study for validation of recently detected immunologic risk factors On a selected cohort of 50 children with severe HSP nephritis at onset, the immunoproteasome switch, altered glycosylation of IgA1, antibodies against deGAI IgA1, complement activation and oxidative stress markers (AOPP) to validate these tests as biomarkers for HSP nephritis at risk of progression.

The project will proceed in 2015 with a call to all ESPN members for patients' enrolment.