

## **Report of the activities of the ESPN WG CAKUT /UTI/bladder dysfunction AUG 2017**

### **Board**

The duration of board membership is 3 years. This year one membership had to be replaced. This was done by voting, according to the ESPN council. Khalid Ismaili left the board and was replaced by Silvio Maringhini . We thank Khalid for his dedication and contribution to the WG.

Coordinator: Ann Raes

Board members: Giovanni Montini, , Amira Peco-Antic, Fatos Yalcinkaya

Members: S. Weber, H. Jardim , A. Yilmaz , A. Bael ,P. Parvex , L. Masella , M. Zaniew , H. Alpay , S. Paunova ,S. Maringhini , K. Gulleroglu , S. Caliskan , F. Lale Sever , S. Yuksel ,L. Murer, B. Ozcakar , A. Van Eerde, A. Woolf , K. Tullus , V. Tasic,I. Rtskhalidaze , A. Yilmaz, D. Hacıhamdioglu, E. Gotsiridze

Liaison ESPN Registry: Marjolein Bonthuis

Liaison Council: Gema Ariceta

CME Education Officer: Paloma Parvex

### **1) General issues**

- a) A first ESPN WG Meeting had been held in Porto, Feb 3rd, 2014. Participants were both members of the WG and interested attendees of the ESPN Annual Meeting. A second ESPN WG Meeting was held in Brussels, Sept 3rd, 2015 . A new ESPN WG meeting is planned in Glasgow, sept 8<sup>th</sup>, 2017
- b) 27 members have joined the WG CAKUT/UTI/Bladder Dysfunction since foundation

### **2) CME and Educational activities**

- a) During the 50<sup>th</sup> Congress of the European Society for Pediatric nephrology in Glasgow 6-9/09/2017 a nephro-urological educational ICCS course on different topics on enuresis and LUT dysfunction will be covered.  
Link to program '<http://espn2017.org/scientificprogramme.php>'
- b) During the 50th Congress of the European Society for Pediatric nephrology in Glasgow sept 7<sup>th</sup> 2017 a symposium on 'The past, present and future of paediatric CAKUT, UTI and bladder disorders' will be held. S. Weber will present 'Advances and new horizons in genetics of CAKUT' and A. Raes will present 'Bladder dysfunction in the paediatric recipient' followed by oral presentations on different research topics.  
Link to the program '<http://espn2017.org/scientificprogramme.php>'

### **3) Registry Activities**

- a)The **EURECA Registry** (European Registry for Familial CAKUT Cases) has been implemented as a webbased registry open to all members of the society (<http://www.nefrokid.it/EURECA/v0/>). This

registry involves the most recent web technologies (AngularJS framework, responsive design, REST API pattern) and will be accessible by mobile devices. We expect to generate a comprehensive collection of familial CAKUT cases throughout Europe by the means of this Registry initiative. As CAKUT (in its overall presentation) is rather frequent we believe that the rate of success of identification of new causative genes will be much higher when restricting the study patients to familial cases. A multinational approach of experts in the field will further strengthen the outcome of the study. A web application will integrate pedigree draw in order to better understand the genetic implications of the disease. The registry will be supported by DNA collections and biobanking. An ESPN Research Grant to financially support this initiative has been approved and donated by the ESPN Council in September 2015. Informed consent and Information material has been created in English language. A translation in different important languages is planned after approval of the local ethics committee. The Registry is still under construction since approval by the ethics committee is awaited. As soon as approval is received we will motivate members of society to register. **Action plan during business meeting Glasgow 2017**

b) **ESPN/ERA-EDTA Registry** data analysis on longterm outcomes in patients with Prune-Belly Syndrome(PBS) on renal replacement therapy has been performed by F. Yalcinkaya (University Children's Hospital, Ankara, Turkey) and Beyza Doğanay Erdoğan (Department of Biostatistics at Ankara University, Turkey) in collaboration with the ESPN/ERA-EDTA Registry team (Marjolein Bonthuis). 86 patients with PBS were compared to 1035 patients with congenital obstructive uropathy and 1905 patients with congenital renal hypodysplasia.

The paper has recently been **published** in 'Pediatr Nephrol. 2017 Aug 4' "Outcomes of renal replacement therapy in boys with prune belly syndrome: findings from the ESPN/ERA-EDTA registry" F. Yalcinkaya, M. Bonthuis, B.Erdogan, K. J van Stralen, S. Baiko, H. Chehade, H. Maxwell, G. Montini, K. Rönholm, S. Sørensen, E. Verrina, S. Weber, J. Harambat, F. Schaefer, K. J Jager, J. W Groothoff.

### 3. Clinical studies

A questionnaire assessing the management of kidney dysplasia through-out Europe has been developed by Giovanni Montini for the WG. After being presented and discussed at the ESPN WG Meeting in Porto on Sept 18th, 2014 and approval of the WG and the ESPN Council, the questionnaire was distributed to all ESPN members and via the ESPN mailing list of national societies. Two requests using MonkeySurvey were sent out in Nov 2014 and Jan 2015. 248 physicians from 54 countries worldwide completed the survey.

The paper has been **published** in 'J Nephrol. 2017 Jun 24'. A questionnaire survey of radiological diagnosis and management of renal dysplasia in children. G. Montini, M. Busutti, F. Yalcinkaya, S. Weber on behalf of the CAKUT ESPN working group.

### 4. Scientific studies

a) Adrian Woolf (Manchester, UK) plans a project within the WG focusing on "Renal tract malformations: from human genomics to novel therapies". At this moment the University of

Manchester has started to use induced pluripotent stem cell technology to model human renal malformations.

b) **2017:** A survey regarding the off label use of quinolones in children will be developed and it is the intention to send it to an international group of pediatricians, pediatric nephrologists and urologists, on their use of quinolone, an off label drug in pediatrics. The questionnaire will be distributed by email to the members of the CAKUT WG for discussion and comments. After approval the questionnaire can then be distributed to members of different societies. We expect that the questionnaires will be returned within a dead-line of 3 months. Data analysis can then be performed. To be started

### **5. Guidelines**

In the second half of 2016 we initiated a standardization paper/ consensus guidelines on urinary tract infections in children. The core group exists of G. Montini, K. Tullus, T. Mattoo, A. Raes, J. Vande Walle, P. Brandström, K. Meesters. This is in collaboration with the European Society of Pediatric Urology (P. Hoebeke, S. Tekgül, G. Manzoni) and the ESPN work group CAKUT. April 2017: Inventarisation on missing items in existing guidelines (ESPU, AAP) is ongoing. A core group meeting took place on June 14-15<sup>th</sup>. Results will be presented in Glasgow Sept 8<sup>th</sup> at the business meeting.

### **6. Financial report**

The two years budget will be spent to cover the expenses of the survey on the use of quinolones and the standardization meeting planned in late spring 2017. To be defined.

### **7. Website Educational Material**

Educational material has been posted on the ESPN website covering the following topics: UTI Bladder Dysfunction, CAKUT Genetics, Antenatal Management of CAKUT

### **8. ESPN regulations**

Replacement of one member of the board of the CAKUT WG took place in January-February 2017.

### **9. ESPN WG Meeting**

A next WG Meeting with all members is planned in September 2017 at the ESPN Glasgow (6-9<sup>th</sup> September). Members are invited on Friday Sept 8<sup>th</sup> in Room Fyne.

For the ESPN WG CAKUT/UTI/Bladder Dysfunction

Ann Raes

Department of Pediatric Nephrology

University Ghent, Belgium

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