

Report of the activities of the ESPN WG CAKUT /UTI/bladder dysfunction AUG 2018

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. Amira Peco-Antic has left the board and was replaced by Julia Hofele. We thank Amira for her dedication and contribution to the WG.

Coordinator: Ann Raes

Board members: Giovanni Montini, Silvio Maringhini, Fatos Yalcinkaya, Julia Höfele

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, Per Brandström, Nica Temizkan Dincel

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. Additional ESPN WG Meeting was held in Brussels (2015) and Glasgow (2017) . A new ESPN WG meeting is planned in Antalya, 05/10/2018
- b) 32 members have joined the WG CAKUT/UTI/Bladder Dysfunction since foundation

2) CME and Educational activities

- a) During the 50th 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 was 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 7th 2017 a symposium on 'The past, present and future of paediatric CAKUT, UTI and bladder disorders' was held. S. Weber presented 'Advances and new horizons in genetics of CAKUT' and A. Raes presented 'Bladder dysfunction in the paediatric recipient' followed by oral presentations on different research topics.
Link to the program '<http://espn2017.org/scientificprogramme.php>'
- c) During the 51th Congress of the European Society for Pediatric nephrology ESPN in Antalya 4-6/10/ 2018 a symposium on CAKUT: From early diagnosis to treatment and outcome: toward human reality ' will be held with following topics: 1)Exploring the genetic landscape of CAKUT: presenter : presented by Velibor Tasic 2) A practical algorithmic approach to assessment and

management of AHN: presented by Fatos Yalcinka 3) CAKUT and prevention of CKD: transition from paediatric to adult nephrourological care: presented by Luisa Murer

d) A clinical quiz with CAKUT as design has recently (dec 2017) been published on the ESPN website.

3) Registry Activities

a) Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) account for approx. 40 % of cases of end stage renal failure until the age of 30 and therefore pose a major disease and economic burden. CAKUT comprises a vast spectrum of both clinically and genetically complex disorders. More than 40 genes have been implicated in monogenic CAKUT forms (isolated and syndromic), yet explaining only up to about 20 % of cases. There is strong evidence that more monogenic CAKUT forms exist, judging from the observation of familial occurrence and monogenic mouse models.

The **EURECA Registry** (European Registry for Familial CAKUT Cases) has been implemented as a webbased registry open to all members of the society (<https://eureca-registry.eu/>). The aim of the EURECA Registry is to set up a standardized registry for familial CAKUT throughout Europe. This registry will feature both genetic and clinical data, with the possibility of an ongoing follow-up of the course of the disease. Information material and informed consent in English language are available on the website after registration. Approval of the local ethics committee for the EURECA Registry is applied. The **official start** of the registry is planned for **January 2019**. An update will be presented by J Höfele at the CAKUT WG meeting at the ESPN meeting in Antalya (Oct 05th ,2018).

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* "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. Rönnholm, S. Sørensen, E. Verrina, S. Weber, J. Harambat, F. Schaefer, K. J Jager, J. W Groothoff. *Pediatr Nephrol.* 2018 Jan;33(1):117-124. doi: 10.1007/s00467-017-3770-9. Epub 2017 Aug 4

c) An update on the international PKD registry ARegPKD (by M Liebau) and on ADPedKD: a global initiative (by D. Mekahli) will be presented at the CAKUT WG meeting at the ESPN meeting in Antalya (Oct 05th ,2018).

3. Clinical studies

1) 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. 2018 '. 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. J Nephrol. 2018 Feb;31(1):95-102. doi: 10.1007/s40620-017-0417-7. Epub 2017 Jun 24

2) G. Montini will present an update on the PREDICT-Trial at the CAKUT WG meeting at the ESPN meeting in Antalya (Oct 05th, 2018).

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 uses induced pluripotent stem cell technology to model human renal malformations.

b) Publication of paper on pharmacokinetic study of ciprofloxacin in children in 'Antimicrob Agents Chemother 2018 '. Results of a Multicenter Population Pharmacokinetic Study of Ciprofloxacin in Children with Complicated Urinary Tract Infection'. Meesters K, Michelet R, Mauel R, Raes A, Van Bocxlaer J, Vande Walle J, Vermeulen A. Antimicrob Agents Chemother 2018 Aug 27;62(9). pii: e00517-18. doi: 10.1128/AAC.00517-18.

b) **2017-2018**: A survey regarding the off label use of quinolones in children was adapted to a broader topic 'uroprophylaxis'. The questionnaire will be discussed at the CAKUT WG meeting at ESPN 2018 and after comments and approval it will be distributed to members of different societies (international group of pediatricians, pediatric nephrologists and urologists). Data analysis can then be performed.

5. Guidelines

1) 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 14-15th. Results will be presented in Glasgow Sept 8th at the business meeting. 11/2017: A draft of the proposed ERBP consensus paper has been finalized. February 2018: proposed to our ERBP stakeholder on how to adapt the concept paper. June 2018: Recommendations received and now in stage of adapting the manuscript according to 'systematic review with statements'. The updated manuscript will be discussed with the corer group for comments and finalization at the ESPN 2018.

2) Initiation of collaboration with the ERKNet WG on CAKUT and Ciliopathies to start a project of a consensus paper on „Renal dysplasia“, based on the recently published work of the ESPN questionnaire by G. Montini.

6. Financial report

The two years budget will be spent to cover the expenses of the survey on the use of quinolones . 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 2018.

9. ESPN WG Meeting

A next WG Meeting with all members is planned in October 2018 at the ESPN Glasgow (3-6 th September). Members are invited on Friday oct 5th in Room Romano 1

For the ESPN WG CAKUT/UTI/Bladder Dysfunction

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