Report of ESPN WGIKD Board 2022 (February)

WG composition

Chair: E. Levtchenko

Board: G. Ariceta, F. Emma, D. Haffner, R. Lennon, M. Liebau

Members: 60 members (2021: 19 new members, 2022: 1 new member)

Published papers 2021 & 2022

- Boyer O, Schaefer F, Haffner D, Bockenhauer D, Hölttä T, Bérody S, Webb H, Heselden M, Lipska-Zie Tkiewicz BS, Ozaltin F, Levtchenko E, Vivarelli M. Nat Rev Nephrol. Management of congenital nephrotic syndrome: consensus recommendations of the ERKNet-ESPN Working Group. 2021 Apr;17(4):277-289.
- Trepiccione F, Walsh SB, Ariceta G, Boyer O, Emma F, Camilla R, Ferraro PM, Haffner D, Konrad M, Levtchenko E, Lopez-Garcia SC, Santos F, Stabouli S, Szczepanska M, Tasic V, Topaloglu R, Vargas-Poussou R, Wlodkowski T, Bockenhauer D. Distal renal tubular acidosis: EKRNet/ESPN clinical practice points. Nephrol Dial Transplant. 2021 Aug 27;36(9):1585-1596.
- Emma F, Hoff WV, Hohenfellner K, Topaloglu R, Greco M, Ariceta G, Bettini C, Bockenhauer D, Veys K, Pape L, Hulton S, Collin S, Ozaltin F, Servais A, Deschênes G, Novo R, Bertholet-Thomas A, Oh J, Cornelissen E, Jansse M, Haffner D, Ravà L, Antignac C, Devuyst O, Niaudet P, Levtchenko E.An international cohort study spanning five decades assessed outcome of nephropathic cystinosis. Kidney Int. 2021 Jul 6:S0085-2538(21)00648-7.
- Burgmaier K, Brinker L, Erger F, Beck B, Benz M, Bergmann C, Boyer O, Collard L, Dafinger C, Fila M, Kowalewska C, Lange-Sperandio B, Massella L, Mastrangelo A, Mekahli D, Miklaszewska M, Ortiz-Bruechle N, Patzer L, Prikhodina L, Ranchin B, Ranguelov N, Schild R, Seeman T, Sever L, Sikora P, Szczepanska M, Teixeira A, Thumfart J, Uetz B, Weber LT, Wühl E, Zerres K, ESCAPE Study group, GPN study group, Dötsch J, Schaefer F, Liebau MC for the ARegPKD consortium. Refining genotype-phenotype correlations in 304 patients with autosomal recessive polycystic kidney disease (ARPKD) and PKHD1 variants. Kidney Int. 2021 Sep;100(3):650-659.
- Burgmaier K, Kilian S, Arbeiter K, Atmis B, Büscher A, Derichs U, Dursun I, Duzova A, Eid LA, Galiano M, Gessner M, Gokce I, Haeffner K, Hooman N, Jankauskiene A, Körber F, Longo G, Massella L, Mekahli D, Miloševski-Lomić G, Nalcacioglu H, Rus R, Shroff R, Stabouli S, Weber LT, Wygoda S, Yilmaz A, Zachwieja K, Zagozdzon I, Dötsch J, Schaefer F, Liebau MC on behalf of the ARegPKD consortium. Early childhood height-adjusted total kidney volume as a risk marker of kidney survival in ARPKD. Scientific Reports 2021 Nov 4;11(1):21677.
- Verploegen MFA, Vargas-Poussou R, Walsh SB, Alpay H, Amouzegar A, Ariceta G, Atmis B, Bacchetta J, Bárány P, Baron S, Bayrakci US, Belge H, Besouw M, Blanchard A, Bökenkamp A, Boyer O, Burgmaier K, Calò LA, Decramer S, Devuyst O, van Dyck M, Ferraro PM, Fila M, Francisco T, Ghiggeri GM, Gondra L, Guarino S, Hooman N, Hoorn EJ, Houillier P, Kamperis K, Kari JA, Konrad M, Levtchenko E, Lucchetti L, Lugani F, Marzuillo P, Mohidin B, Neuhaus TJ, Osman A, Papizh S, Perelló M, Rookmaaker MB, Conti VS, Santos F, Sawaf G, Serdaroglu E, Szczepanska M, Taroni F, Topaloglu R, Trepiccione F, Vidal E, Wan ER, Weber L, Yildirim ZY, Yüksel S, Zlatanova G, Bockenhauer D, Emma F, Nijenhuis T.Parathyroid hormone and phosphate homeostasis in patients with Bartter and Gitelman syndrome: an international cross-sectional study. Nephrol Dial Transplant. 2022 Feb 7:gfac029.

Online CME Course 2021, November 25

Rachel Lennon	Collagen IV nephropathies + 2 case presentations
Dieter Haffner	X-linked hypophosphatemic rickets + 1 case presentation
Detlef Bockenhauer	distal RTA + 2 case presentations

294 registrations, 135 participants from 43 countries

Ongoing registries

dRTA Registry	D. Bokkenhauer
ADPKD Registry	D. Mekahli
ARPKD Registry	M. Liebau
Cystinosis Registry	A. Servais & E. Levtchenko

Ongoing projects

- Glucose-metabolism in early ADPKD: a multi-center study (supported by ESPN research grant). A. Dachy, D. Mekahli
- Mutations in the sodium-phosphate cotransporter genes – survey
  L. Brunkhorst, D. Haffner: > 180 patients, data analysis ongoing
- Collecting duct water reabsorption in cystinosis
  A. Ferrulli, G. Valenti, E. Levtchenko
- Use of NSAID in patients with tubulopathies
  F. Emma, D. Bockenhauer: > 400 patients, data analysis ongoing

Planned New Guidelines & Clinical Practice Recommendations

<u>For 2022:</u>

- XLH update
- NDI (together with ERKNet)
- Dent (together with ERKNet)

<u>For 2023</u>:

- Disorders of Na-Pi transporters
- update ARPKD

WGIKD CME course 2022

November 24, 2022

3-6 pm CET

How to diagnose and how to treat hereditary kidney diseases?

Digital, 3 hours, 3 topics followed by case presentations and breakout sessions

Draft Program

Hereditary FSGS

ARPKD

Congenital anomalies of kidney and urinary tract

ESPN Board Examination questions on inherited kidney disorders

WGIKD board will participate in writing questions for the next ESPN Board on Inherited Kidney Disorders