



Activity report of ESPN Working Group on Inherited Kidney Disorders (IKDWG)

September 2021

IKDWG COMPOSITION

Chair: E. Levtchenko

New Board: D. Bockenbauer (leaving 2021), D. Haffner, R. Lennon, M. Liebau

Vacant position: will be announced in October 2021

Liaison ERA-EDTA WG: F. Emma

Members: 59 (2021: 19 new members)

IKDWG MEETINGS

- Board meetings: February 5, 2021

Fall 2021 (TBC)

- Plenary meetings: April 22, 2021

September 16, 2021

PUBLISHED PAPERS

2019

Treatment and long-term outcome in primary distal renal tubular acidosis. Lopez-Garcia SC et al. Nephrol Dial Transplant. 2019; 34(6): 981-991. ADPKD: a global platform on the management of children with ADPKD. De Rechter S et al. Kidney Int Rep. 2019; 4(9): 1271-1284.

Clinical practice recommendations for the diagnosis and management of X-linked hypophosphatemia. Haffner D et al. Nat Rev Nephrol. 2019 Nov;15(11):713-726.

2020

Severe neurological outcomes after very early bilateral nephrectomies in patients with autosomal recessive polycystic kidney disease (ARPKD). Burgmaier K, Ariceta G, Bald M, Buescher AK, Burgmaier M, Erger F, Gessner M, Gokce I, König J, Kowalewska C, Massella L, Mastrangelo A, Mekahli D, Pape

L, Patzer L, Potemkina A, Schalk G, Schild R, Shroff R, Szczepanska M, Taranta-Janusz K, Tkaczyk M, Weber LT, Wühl E, Wurm D, Wygoda S, Zagodzón I, Dötsch J, Oh J, Schaefer F, Liebau MC; ARegPKD consortium. Sci Rep. 2020 Sep 29;10(1):16025.

2021

Management of congenital nephrotic syndrome: consensus recommendations of the ERKNet-ESPN Working Group. 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. 2021 Apr;17(4):277-289.

Distal renal tubular acidosis: EKRNet/ESPN clinical practice points. 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. Nephrol Dial Transplant. 2021 Aug 27;36(9):1585-1596.

An international cohort study spanning five decades assessed outcome of nephropathic cystinosis. 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, Devuyt O, Niaudet P, Levtchenko E. Kidney Int. 2021 Jul 6:S0085-2538(21)00648-7.

ONGOING PROJECTS

Registries

dRTA Registry	D. Bockenhauer
ADPKD Registry	M. Liebau
ARPKD Registry	D. Mekahli
Cystinosis Registry	A. Servais

Surveys

Mutations in the sodium-phosphate cotransporter genes – survey D. Haffner, L. Brunkhorst

FINALIZED PROJECTS

PTH levels in Bartter and Gitelman syndromes F. Emma et al, in association with ERKNet

Cystinosis sibling study. Koenraad Veys & E. Levtchenko

NEW PROJECTS

Suggestions presented in April 2021 at IKDWG meeting

Glucose-metabolism in early ADPKD: a multi-center study A. Dachy, D. Mekahli

Collecting duct water reabsorption in cystinosis A. Ferrulli, G. Valenti, E. Levtchenko