



An update on the Registry- December 2016



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As members of the ESPN/ERA-EDTA Registry committee we would like to thank you again for your participation and efforts to the Registry. Currently, 38 countries are participating in the Registry, providing information on more than 10,000 patients who started RRT before the age of 20, between 1997 and 2014.

In 2016, nine papers based on Registry data have been accepted for publication by different journals and many others have been submitted. We are very happy to announce that one of our papers has even been accepted by The Lancet. The full publication list can be found below.

Evgenia Preka from Brussels, Belgium visited the AMC in June. She worked on a collaborative project between the Registry and the ESPN Dialysis WG on eGFR at dialysis initiation and its association with outcomes. Additionally, the relationships with other ESPN working groups are well maintained, resulting in several proposals for collaborative future

studies. Recently, Bram Smit, a medical student from Amsterdam, finished his 16-week scientific internship on comorbidity data.

If you are interested in performing a research project on the Registry or would like to know more about participating in the ESPN/ERA-EDTA registry, please contact Marjolein Bonthuis:

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We would like to thank you again for your collaboration and hope to work with you in the future to improve paediatric nephrology care and research in Europe.

Data analyses and publications

The ESPN/ERA-EDTA Registry collects data on RRT on an annual basis via the national renal registries in Europe. So far, data have been included from eight subsequent years.

In 2014, the overall incidence was 5.7 per million age-related population (pmarp) and ranged from 0.0, as no patients started RRT in that year, to 19.8 pmarp. The prevalence also shows a wide range from 7.3 to 89.3 pmarp. Five-year survival was 93.6% after start of RRT. Most patients died because of infections or other identified death causes.

Again, the country reports provided insights on the performance in each country compared to the average European performance by showing benchmarking figures.

Nine papers have been accepted and published in the previous months. Recently, The Lancet accepted our paper on mortality risk disparities showing considerable variation in mortality rates across Europe. Three papers have been accepted by AJKD. One on racial disparities and access to transplantation, one on transplantation strategies in ARPKD, and one on infant dialysis. The first showed less favorable outcomes for black and Asian patients. ARPKD patients receiving a combined liver-kidney transplant were found to have a higher mortality compared with ARPKD patients receiving a kidney transplant alone.

AJT has published a paper on gender disparities and access to

transplantation, showing that girls had a lower probability of receiving a preemptive kidney transplant.

Kidney International has published a paper on mortality on dialysis, showing a higher mortality risk for children starting on HD compared to those on PD.

Furthermore, Pediatric Nephrology published a summary paper of the ESPN/ERA-EDTA Registry, and accepted a paper on Finnish type nephropathy, and NDT accepted our paper on racial variation in cardiovascular risk factors.

These papers would not have been possible without your help and efforts, for which we are very grateful.

Thank you all for making this possible.

Table 1: Incident patients

Incident paediatric patients accepted for renal replacement therapy in 2014 and general population characteristics of countries contributing 2014 data to the ESPN/ERA-EDTA Registry.

Country	Total		General Population Characteristics		
	RRT patients		Children	Total Population	Children
	0-14 years		0-14 years	0-99 years	0-14 years
	N	pmarp	N	N	percent
Albania	0	0.0	546,473	2,894,476	18.9
Austria	8	6.6	1,218,844	8,507,786	14.3
Belarus	8	5.3	1,518,669	9,480,868	16.0
Belgium	5	2.6	1,910,700	11,231,212	17.0
Bosnia and Herzegovina	3	4.9	607,160	3,507,928	17.3
Bulgaria	0	0.0	997,171	7,223,941	13.8
Croatia	4	6.4	624,344	4,236,063	14.7
Cyprus	1	7.2	139,401	852,504	16.4
Czech Republic	9	5.7	1,589,249	10,525,346	15.1
Denmark	5	5.2	965,658	5,643,476	17.1
Estonia	0	0.0	208,812	1,314,542	15.9
Finland	11	12.3	895,815	5,461,514	16.4
France	64	5.2	12,340,667	66,262,304	18.6
FYR of Macedonia	0	0.0	348,030	2,067,139	16.8
Georgia	9	11.8	760,455	4,483,230	17.0
Germany-KFH*	27	2.5	10,664,577	80,982,502	13.2
Germany-CERTAIN*	6	0.6	10,664,577	80,982,502	13.2
Greece	8	5.0	1,589,011	10,892,416	14.6
Hungary	8	5.6	1,426,500	9,866,469	14.5
Iceland	0	0.0	66,906	327,384	20.4
Italy*	35	4.2	8,415,627	60,789,138	13.8
Latvia	1	3.4	296,052	1,993,783	14.8
Lithuania	0	0.0	427,775	2,932,366	14.6
Malta	0	0.0	61,331	427,362	14.4
Norway	9	9.7	931,668	5,137,231	18.1
Poland	40	7.0	5,716,758	38,011,737	15.0
Portugal	13	8.6	1,506,048	10,401,062	14.5
Republic of Serbia	6	5.9	1,024,133	7,130,576	14.4
Romania	9	2.9	3,087,847	19,908,982	15.5
Russia	96	4.5	21,534,456	142,368,368	15.1
Slovakia	4	4.8	830,053	5,418,649	15.3
Slovenia	6	19.8	302,682	2,061,982	14.7
Spain	36	5.1	7,058,015	46,480,882	15.2
Sweden	17	10.2	1,664,067	9,696,107	17.2
Switzerland	6	4.9	1,224,981	8,237,666	14.9
the Netherlands	17	6.0	2,838,571	16,865,008	17.0
Turkey*	61	3.2	18,856,122	77,181,882	24.4
Ukraine	26	3.9	6,710,689	45,309,294	14.8
United Kingdom	108	9.5	11,407,724	64,596,752	17.7
Total*	537	5.7	94,376,715	591,756,405	15.9

* Data from the German transplantation registry are based on 18 transplantation centers. In 2014, 117 patients under the age of 21 years were transplanted. The incidence in Turkey is an underestimation of the true incidence. In Italy, (pre-emptive) transplantation patients are not included; these numbers are an underestimation of true incidence. Therefore, Germany, Turkey, Italy, and Turkey, were excluded from the overall incidence.

Table 2: Treatment modality at start of RRT in 2014

Treatment modality at day 1, among patients < 15 years of age starting RRT in 2014.
Patients from Germany, Italy, and Switzerland are excluded.

	N	Percent	Pmarp
HD at start	221	41.2	2.34
PD at start	212	39.5	2.25
Pre-emptive transplantation	99	18.4	1.05
Unknown	5	0.9	0.05

Table 3: PRD distribution at start of RRT in 2014

Cause of renal failure, among patients < 15 years of age, starting RRT in 2014 according to new and old PRD coding.

	N		Percent		Pmarp	
	New	Old	New	Old	New	Old
CAKUT	193	163	35.9	30.4	2.07	1.75
Glomerulonephritis	86	69	16.0	12.8	0.92	0.74
Cystic kidney disease	68	71	12.7	13.2	0.73	0.76
Hereditary nephropathy	-	31	-	5.8	-	0.33
Metabolic and tubulointerstitial disorders	25	12	4.7	2.2	0.27	0.13
Toxic/ischemic renal failure	10	8	1.9	1.5	0.11	0.09
HUS	36	36	6.7	6.7	0.39	0.39
Vascular	12	10	2.2	1.9	0.13	0.11
Miscellaneous	87	61	16.2	11.4	0.93	0.66
Unknown	20	76	3.7	14.2	0.21	0.82

Table 4: eGFR at start of RRT

Estimated GFR based on age, height and serum creatinine levels, calculated according to the new bedside Schwartz formula, among incident patients, age <15 years in 2014.

	N	Percent
eGFR<8 ml min ⁻¹ per 1.73 m ²	122	39.2
eGFR 8- 15 ml min ⁻¹ per 1.73 m ²	147	47.3
eGFR>15 ml min ⁻¹ per 1.73 m ²	42	13.5

Table 5: Prevalent Patients

Prevalent paediatric patients on renal replacement therapy on the 31st of December 2014. Prevalent counts and prevalence per million age related population, by age groups.

Country	Total RRT		Age Groups		
	N	0-14 years	Infants	Children	Adolescents
		pmarp	0-4 years	5-9 years	10-14 years
		pmarp	pmarp	pmarp	pmarp
Albania	4	7.3	0.0	6.0	14.4
Austria	46	37.7	15.0	49.4	48.2
Belarus	35	23.0	7.0	21.9	45.1
Belgium	70	36.6	23.1	37.0	50.4
Bosnia and Herzegovina	11	18.1	43.5	0.0	17.8
Bulgaria	8	8.0	0.0	0.0	25.2
Croatia	23	36.8	28.9	33.4	48.2
Cyprus	7	50.2	41.4	65.1	44.4
Czech Republic	42	26.4	10.7	28.4	43.1
Denmark	40	41.4	13.3	36.2	72.0
Estonia	2	9.6	0.0	0.0	32.2
Finland	80	89.3	69.6	82.6	116.5
France	460	37.3	14.7	33.7	62.8
FYR of Macedonia	4	11.5	0.0	35.5	0.0
Georgia	13	17.1	14.2	8.7	28.1
Germany-KFH*	115	10.8	10.4	11.2	10.7
Germany-CERTAIN*	205	19.2	5.5	22.9	28.4
Greece	52	32.7	13.7	29.3	54.5
Hungary	48	33.6	8.8	30.4	60.4
Iceland	4	59.8	0.0	88.0	94.1
Italy*	275	32.7	18.2	31.7	47.2
Lithuania	10	23.4	13.3	14.6	42.8
Malta	4	65.2	0.0	50.3	146.0
Norway	45	48.3	25.8	41.5	77.9
Poland	229	40.1	17.9	39.3	64.9
Portugal	79	52.5	21.9	35.7	93.4
Republic of Serbia	28	27.3	6.1	41.2	33.9
Romania	42	13.6	4.2	15.9	19.8
Russia	382	17.7	7.3	17.3	30.8
Slovakia	17	20.5	10.4	14.4	37.9
Slovenia	11	36.3	36.4	19.7	54.7
Spain	286	40.5	17.5	41.9	62.1
Sweden	100	60.1	34.4	56.7	92.7
Switzerland*	52	42.4	16.7	32.1	79.6
the Netherlands	123	43.3	16.8	51.5	59.2
Turkey*	251	13.3	5.4	10.5	23.9
Ukraine	84	12.5	3.2	11.7	25.9
United Kingdom	646	56.6	26.6	57.8	89.7
Total*	3035	32.2	14.4	32.1	53.1

* Data from the German transplantation registry are based on 18 transplantation centers. In 2014, 117 patients under the age of 21 years were transplanted. The prevalence for Turkey is an underestimation of the true prevalence. In Switzerland, not all patients provided informed consent resulting in an underestimation of the true prevalence. In Italy, (pre-emptive) transplantation patients are not included; these numbers are an underestimation of true prevalence. Therefore, Germany, Switzerland, Italy, and Turkey were excluded from the overall prevalence.

Table 5: Prevalent Patients (continued)

Prevalent paediatric patients on renal replacement therapy on the 31st of December 2014. Prevalent counts and prevalence per million age related population, by gender and treatment modality.

Country	Gender		Treatment Modality		
	Males 0-14 years pmarp	Female 0-14 pmarp	HD 0-14 years pmarp	PD 0-14 years pmarp	Transplantation 0-14 years pmarp
Albania	10.5	3.8	3.7	0.0	3.7
Austria	48.0	27.0	0.8	4.1	32.8
Belarus	26.9	19.0	2.0	6.6	14.5
Belgium	42.0	31.1	8.9	3.7	21.5
Bosnia and Herzegovina	25.8	10.1	11.5	3.3	3.3
Bulgaria	9.8	6.2	2.0	2.0	3.0
Croatia	43.6	29.7	1.6	24.0	11.2
Cyprus	28.0	73.7	0.0	14.3	35.9
Czech Republic	34.3	18.1	2.5	5.7	18.2
Denmark	54.6	27.6	1.0	3.1	37.3
Estonia	9.3	9.9	0.0	0.0	9.6
Finland	100.4	77.7	1.1	5.6	82.6
France	44.2	30.0	5.9	3.4	28.0
FYR of Macedonia	11.1	11.9	5.7	2.9	2.9
Georgia	22.5	11.1	5.3	9.2	2.6
Germany-KFH*	12.8	8.7	3.8	6.7	-
Germany-CERTAIN*	23.0	15.2	-	-	18.7
Greece	44.2	20.6	8.8	9.4	14.5
Hungary	41.0	25.9	3.5	5.6	24.5
Iceland	88.0	30.5	0.0	0.0	59.8
Italy*	39.5	25.5	5.2	10.1	-
Lithuania	18.2	28.8	0.0	9.4	11.7
Malta	63.4	67.2	0.0	0.0	65.2
Norway	58.7	37.4	1.1	1.1	46.2
Poland	45.0	34.1	4.5	8.6	26.9
Portugal	58.3	46.3	2.7	14.6	35.2
Republic of Serbia	19.0	36.2	4.9	2.0	20.5
Romania	14.5	12.7	8.1	4.9	0.6
Russia	21.8	13.5	3.5	7.0	7.2
Slovakia	21.1	19.8	1.2	10.8	8.4
Slovenia	51.4	20.4	6.6	9.9	19.8
Spain	46.8	33.9	2.4	3.3	34.9
Sweden	70.2	49.4	3.0	6.0	50.5
Switzerland*	42.3	35.3	4.9	3.3	33.5
the Netherlands	54.4	31.7	3.2	2.1	38.0
Turkey*	14.6	12.0	2.3	8.4	2.4
Ukraine	12.8	12.1	5.1	3.0	4.3
United Kingdom	70.0	42.6	7.4	6.1	42.5
Total*	38.3	26.1	4.6	6.9	22.1

* Data from the German transplantation registry are based on 18 transplantation centers. In 2014, 117 patients under the age of 21 years were transplanted. The prevalence for Turkey is an underestimation of the true prevalence. In Switzerland, not all patients provided informed consent resulting in an underestimation of the true prevalence. In Italy, (pre-emptive) transplantation patients are not included; these numbers are an underestimation of true prevalence. Therefore, Germany, Switzerland, Italy, and Turkey were excluded from the overall prevalence.

Table 6: Hypertension and height in children on RRT

Height z-score based on recent national reference charts, or, if unavailable, on newly developed reference charts for Northern and Southern Europe (Bonthuis et al, PLoS ONE 7(8): e42506. doi:10.1371/journal.pone.0042506).

	Dialysis	Transplantation
Blood pressure		
% of patients with hypertension	47.0 (45.2-48.8)	26.6 (25.3-27.9)
Mean z-score systolic blood pressure	1.28 (1.20-1.35)	0.81 (0.77-0.85)
Mean z-score diastolic blood pressure	1.11 (1.05-1.17)	0.63 (0.60-0.67)
Height		
% of patients with height z-score < -2	50.2 (48.8-51.6)	40.9 (39.6-42.3)
Mean height z-score	-2.10 (-2.16; -2.04)	-1.81 (-1.86; -1.77)

Figure 1: Five-year survival

Incident RRT patients under the age of 15 starting RRT from 2007 onwards. Follow-up till 31st of December 2014.

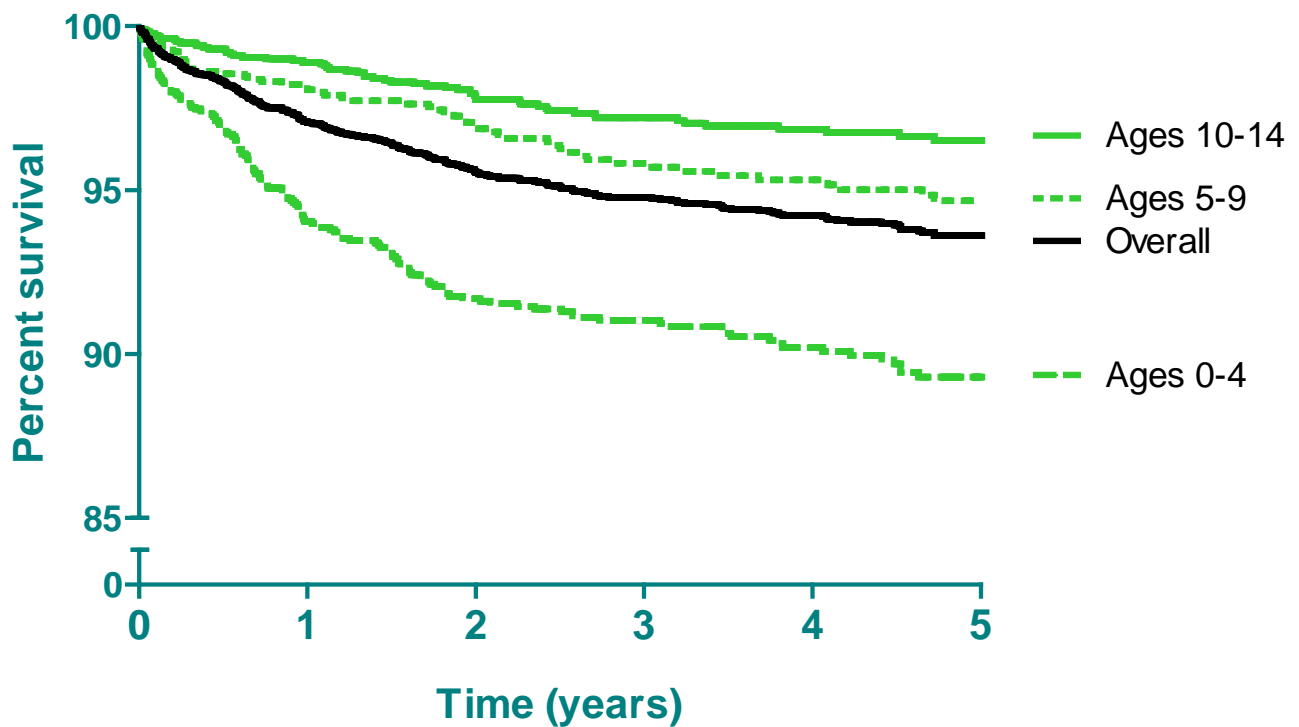


Table 7: Causes of Death

Causes of death according to the ERA-EDTA coding lists. Incident RRT patients under the age of 15 starting RRT from 2007 onwards. Follow-up till 31st of December 2014.

Cause of death	Number of deaths	Percent
Myocardial ischemia and infarction	1	0.3
Cardiac failure	23	7.8
Cardiac arrest/sudden death other cause	30	10.2
Cerebro-vascular accident	24	8.1
Infection	52	17.6
Suicide /refusal or cessation of treatment	4	1.3
Treatment withdrawn	2	0.7
Malignant disease	9	3.1
Other identified cause of death	51	17.3
Cause of death uncertain/not determined	99	33.6

ESPN/ERA-EDTA Registry Scientific Committee

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Publication list 2016

1. Racial disparities in access to and outcomes of kidney transplantation in children, adolescents and young adults: Results from the ESPN/ERA-EDTA Registry. Tjaden LA, Noordzij M, van Stralen KJ, Kuehni CE, Raes A, Cornelissen EA, O'Brien C, Papatristou F, Schaefer F, Groothoff JW, Jager KJ. *Am J Kidney Dis.* 2016 Feb;67(2):293-301.

2. Mortality risk in European children with end-stage renal disease on dialysis. Chesnaye NC, Schaefer F, Groothoff JW, Bonthuis M, Reusz G, Heaf JG, Lewis M, Maurer E, Paripović D, Zagozdzon I, van Stralen KJ, Jager KJ. *Kidney Int* 2016 Jun;89(6):1355-62.

3. Gender disparities in access to pediatric renal transplantation in Europe: data from the ESPN/ERA-EDTA Registry. Hogan J, Couchoud C, Bonthuis M, Groothoff JW, Jager KJ, Schaefer F, van Stralen KJ. *Am J Transplant.* 2016 Jul;16(7):2097-105.

4. Lessons learned from the ESPN/ERA-EDTA Registry. Harambat J, Bonthuis M, Groothoff JW, Schaefer F, Tizard EJ, Verrina E, van Stralen KJ, Jager KJ. *Pediatr Nephrol.* 2016 Nov;31(11):2055-64.

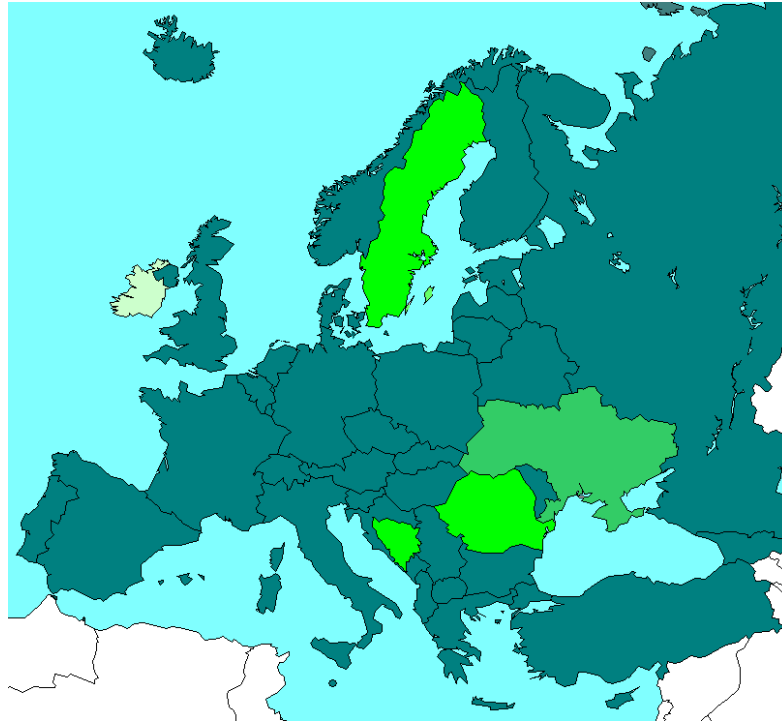
5. Kidney versus combined kidney and liver transplantation in children with autosomal recessive polycystic kidney disease: data from the ESPN/ERA-EDTA Registry. Mekahli D, van Stralen KJ, Bonthuis M, Jager KJ, Balat A, Benetti E, Godefroid N, Edvardsson VO, Heaf JG, Jankauskiene A, Kerecuk L, Marinova S, Puteo F, Seeman T, Zurowska A, Pirenne J, Schaefer F, Groothoff JW. *Am J Kidney Dis.* 2016 Nov;68(5):782-788.

6. Timing of renal replacement therapy does not influence survival and growth in children with congenital nephrotic syndrome caused by mutations in NPHS1-Data from the ESPN/ERA-EDTA Registry. Hölttä T, Bonthuis M, van Stralen KJ, Bjerre A, Topaloglu R, Ozaltin F, Holmberg C, Harambat J, Jager KJ, Schaefer F, Groothoff JW. *Pediatr Nephrol.* 2016 Dec;31(12):2317-2325.

7. Infants requiring maintenance dialysis: outcomes of hemodialysis and peritoneal dialysis. Vidal E, van Stralen KJ, Chesnaye NC, Bonthuis M, Holmberg C, Zurowska A, Trivelli A, Eduardo Esteves Da Silva J, Herthelius M, Adams B, Bjerre A, Jankauskiene A, Miteva P, Emirova K, Bayazit AK, Mache JC, Sánchez-Moreno A, Harambat J, Groothoff JW, Jager KJ, Schaefer F, Verrina E. *Am J Kidney Dis.* 2016 [Epub ahead of print].

8. Mortality risk disparities in children receiving chronic renal replacement therapy for the treatment of end-stage renal disease across Europe. An ESPN/ERA-EDTA Registry analysis. Chesnaye NC, Schaefer F, Bonthuis M, Holman R, Baiko S, Baskin E, Bjerre A, Cloarec S, Cornelissen EAM, Espinosa L, Heaf JG, Stone R, Shtiza D, Zagozdzon I, Harambat J, Jager KJ, Groothoff JW, van Stralen KJ. *Accepted by The Lancet.*

9. Racial variation in cardiovascular disease risk factors among European children on renal replacement therapy- Results from the ESPN/ERA-EDTA Registry. Tjaden LA, Jager KJ, Bonthuis M, Kuehni C, Lilien MR, Seeman T, Stefanidis CJ, Tse Y, Harambat J, Groothoff JW, Noordzij M. *Accepted by Nephrol Dial Transplant.*



Provided extended data to the ESPN/ERA-EDTA Registry

Provided limited data to the ESPN/ERA-EDTA Registry

Provided data via the ERA-EDTA Registry

Intend to contribute data in the near future

We sincerely thank the following countries and persons for their willingness to provide data to the Registry

Albania	D Shtiza	Italy	B Gianoglio, G Leozappa, B Minale
Austria	R Kramar		R Roperto, S Testa, E Vidal, E Verrina
Belarus	S Baiko, A Sukalo	Malta	V Said-Conti
Belgium	K van Hoeck and the Centre contributors to the Belgian Registry Committee	Moldova	S Gatcan, O Berbeca, N Zaikova, N Revenco
Bosnia Herzegovina	D Pokrajac	Montenegro	S Pavičević
Bulgaria	D Roussinov	Norway	T Leivestad, A Bjerre
Croatia	D Batinić, M Lemac, J Slavicek, D Milosevic	Poland	A Zurowska, I Zagozdzon
Cyprus	A Elia	Portugal	C Mota, R Stone, C Afonso
Czech Republic	T Seeman, K Vondrak	Romania	G Mircescu, L Garneata
Denmark	JG Heaf	Russia	EA Molchanova, NA Tomilina, BT Bikbov
Estonia	Ü Toots	Serbia	M Kostic, S Spasojevic- Dimitrijeva, G Milosevski-Lomic, D Paripovic, S Puric, D Kruscic
Finland	P Finne, C Grönhagen-Riska	Slovakia	L Podracka, G Kolvek
France	C Couchoud, M Lassalle, E Berard	Slovenia	N Battelino, G Novljan, J Buturovic-Ponikvar
FYR of Macedonia	E Sahpazova, N Abazi	Spain	A Alonso Melgar and the Spanish Paediatric Registry.
Georgia	T Davitaia	Sweden	S Schön, KG Prütz, L Bäckman, M Evans, M Stendahl, M Segelmark
Germany - CERTAIN	K Krupka, B Höcker, L Pape, B Tönshoff	Switzerland	CE Kuehni, E Maurer, GF Laube, S Tschumi, P Parvex
Germany - KfH	K Rascher, E Nüsken, L Weber, G von Gersdorff, Prof. Jörg Dötsch, F Schaefer	The Netherlands	L Heuveling and MH Hemmelder on behalf of the Nefrovisie foundation, JW Groothoff and all centres participating in the RICHQ-study
Greece	N Afentakis, A Kapogiannis, N Printza, C Stefanidis	Turkey	R Topaloglu, A Duzova
Hungary	G Reusz, Cs Berecki, A Szabó, T Szabó, Zs Györke, E Kis	Ukraine	DD Ivanov, SP Fomina
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Latvia	H Cerneviskis, V Kuzema		
Lithuania	S Rudaitis, A Jankauskiene		