



**SPNP**

Sociedade Portuguesa  
de Nefrologia Pediátrica

# Registo da DRC5 em TSFR -2020-

**Conceição Mota  
Clara Gomes  
Margarida Abranches  
Rosário Stone**

# Movimento -ano de 2020-

<b><i>INCIDENTES</i></b>	
Novos doentes DRC5 em TSFR Diálise e TR <i>preemptive</i>	9

<b><i>SAÍDOS</i></b>	
Mortos	1
Recuperação da FR	1
Saída de registo → transferência “Adultos”	15

<b><i>Mudança de TSFR</i></b>	
TR → HD ou DP	-
HD → DP	1
DP → HD	2

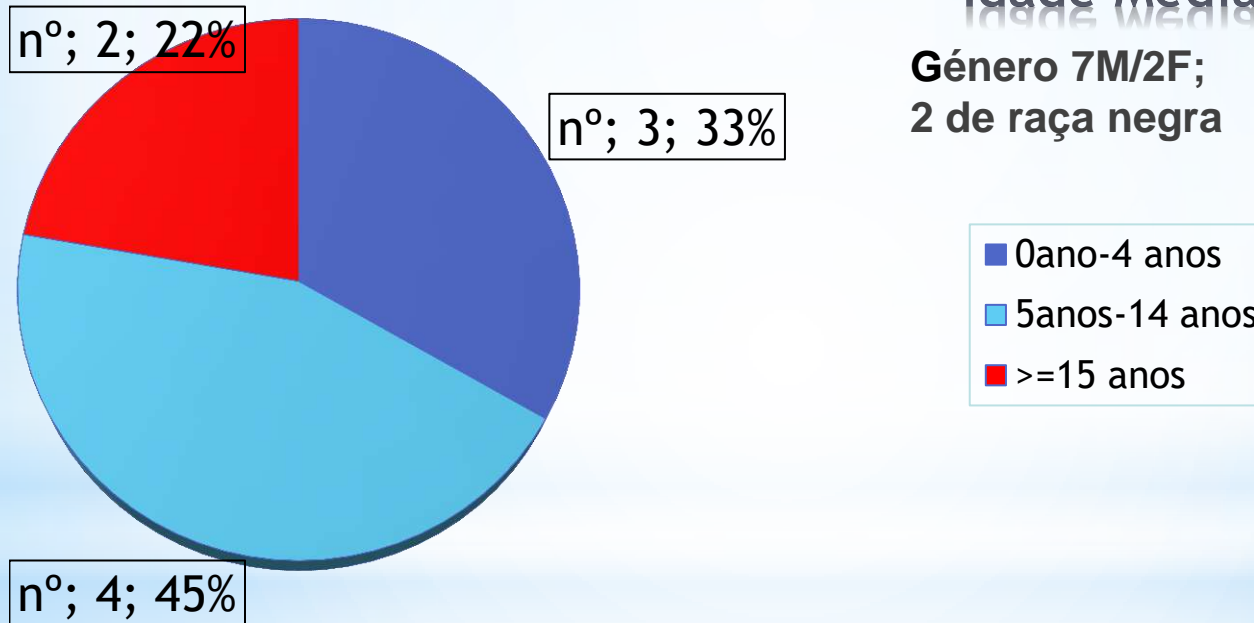
<b><i>Transplantes</i></b>	5
----------------------------	---

# Doentes Incidentes em 2020 (1º TSFR)

9 doentes

Idade Média=9,8± 6,9anos

Género 7M/2F;  
2 de raça negra



# Evolução do registo

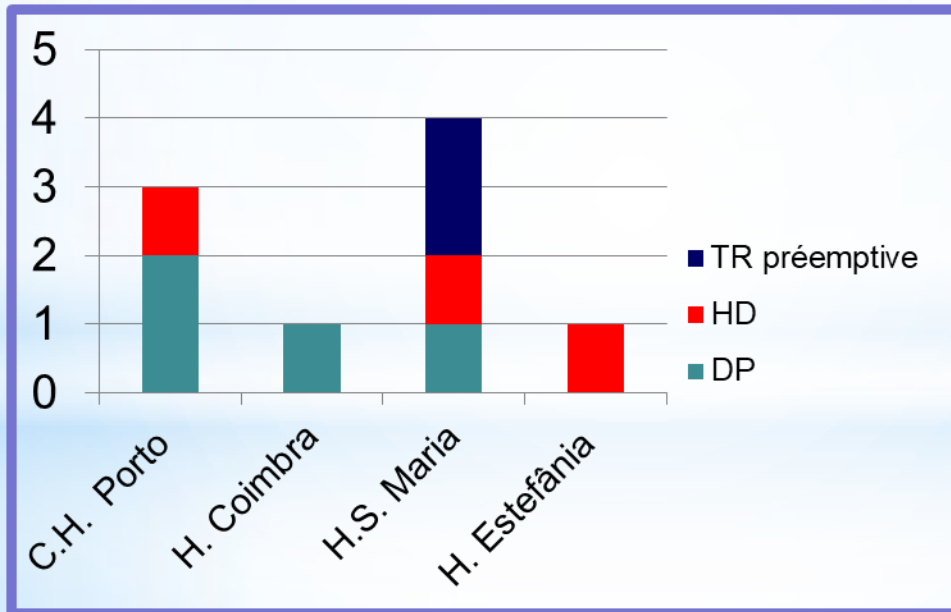
**Novos doentes/TR**

<b>Ano</b>	<b>Novos doentes</b>	<b>Transplante/ano</b>
2007	19	17
2008	16	18
2009	18	16
2010	17	17
2011	18	16
2012	24	17
2013	10	17
2014	17	7
2015	16	20
2016	11	15
2017	13	13
2018	14	6
2019	15	10
2020	9	5

# Doentes que iniciaram TSFR 2020

N=9

**ANO 2020**  
Tipo de TSFR (dia1)  
4 em DP  
3 em HD  
2 TR *préemptive*.



# Transplantação- ano de 2020

N=5 transplantes

-CHUPorto; n=1

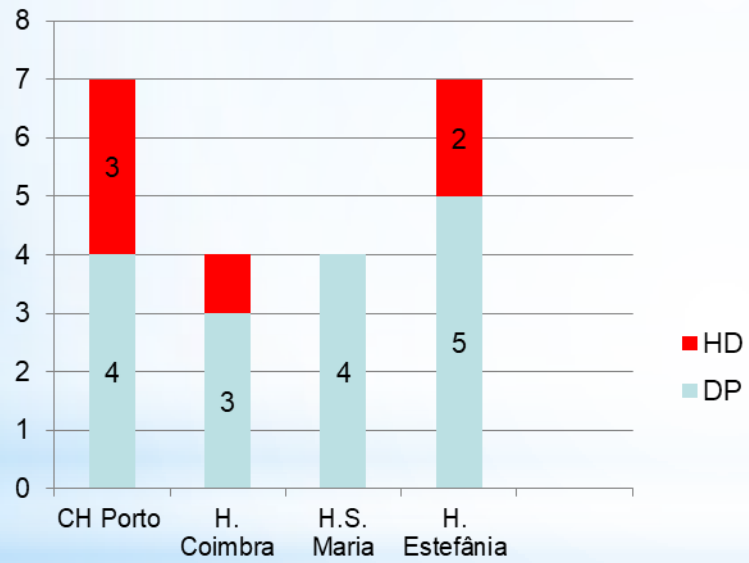
-H. S. Maria, CH Lisboa Norte; n=4

-Dador: Rim de dador cadáver n=3; dador vivo n=2

-TR *préemptive*: n= 2

# Prevalência pontual de doentes em diálise

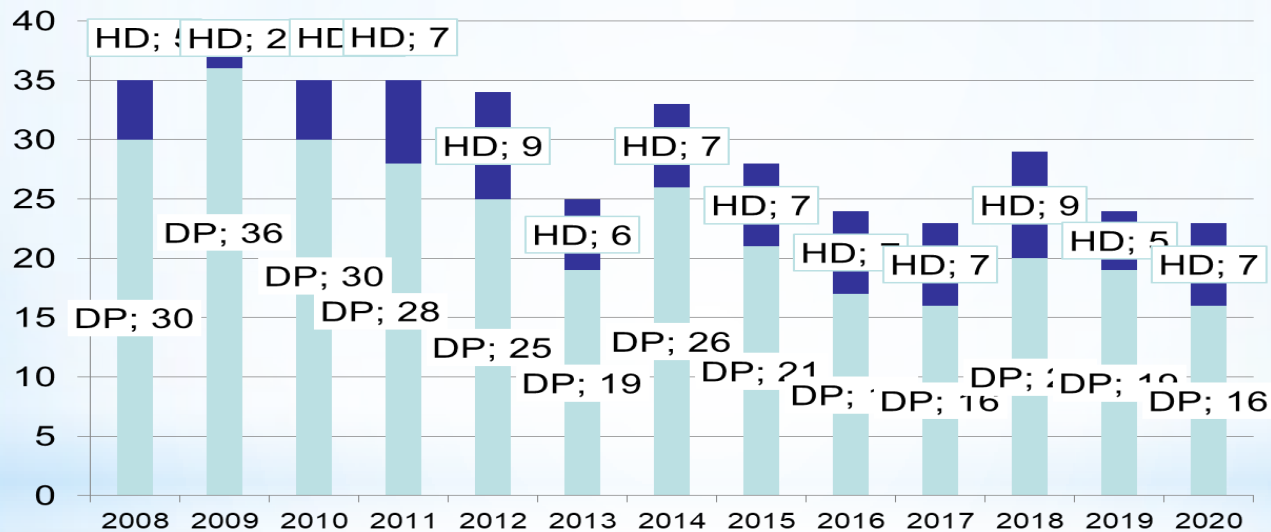
## 31/12/2020



**Diálise=23**  
**DP=16 (69,5%)**  
**HD=7 (30,5%)**

# Evolução do registo

Prevalência pontual de doentes em diálise  
a 31 de Dezembro





# Prevalência pontual de doentes com TR funcionando 31/12/2020

TR=80

Unidade	n
H S Maria-Lisboa	48
C Hospitalar Porto	24
H Pediátrico Coimbra	8

## ANNUAL REPORT

Table 5: Prevalent Patients

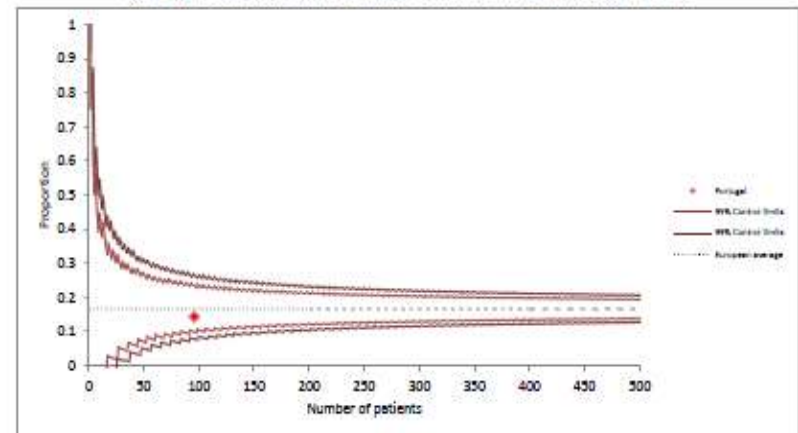
Prevalent paediatric patients on renal replacement therapy on the 31<sup>st</sup> of December 2012. Prevalent counts and prevalence per million age related population, by age groups.

Country	Total RRT patients 0-14 years		Age Groups		Adolescents 10-14 years
	N	pmarp	Infants 0-4 years	Children 5-9 years	
Albania	5	8.8	0.0	5.6	17.6
Austria	47	38.4	17.8	39.4	56.6
Belarus	30	20.5	9.1	17.4	37.7
Bosnia and Herzegovina	10	16.5	37.3	0.0	17.8
Bulgaria	12	12.2	0.0	0.0	38.2
Croatia	26	40.6	37.6	24.4	58.3
Czech Republic	35	22.6	1.7	43.1	26.4
Denmark	53	53.3	28.1	36.0	93.8
Estonia	2	9.6	0.0	14.3	16.5
Finland	75	84.3	56.0	91.5	106.3
France	414	34.1	13.2	34.3	54.6
FYR of Macedonia	5	14.2	0.0	26.5	16.2
Germany*	185	23.6	7.8	27.6	28.3
Greece	57	35.1	19.9	28.0	57.8
Hungary	43	30.0	10.7	33.0	47.5
Iceland	8	120.5	85.2	92.6	187.8
Italy*	262	31.4	16.8	30.7	46.5
Lithuania	9	20.4	6.7	22.2	32.2
Malta	4	65.1	0.0	51.5	138.2
Moldova	1	1.7	0.0	0.0	5.1
Norway	44	47.5	19.2	56.0	67.7
Poland	237	40.8	19.9	39.6	64.7
Portugal	84	53.8	20.6	57.8	78.9
Romania	46	14.4	7.7	10.4	24.8
Russia	322	15.0	6.7	13.6	26.4
Serbia	31	30.0	9.0	28.5	51.4
Slovakia	19	22.8	3.4	15.0	51.1
Slovenia	12	40.5	9.1	21.3	97.8
Spain	279	39.4	16.7	42.6	61.0
Sweden	90	56.3	31.6	57.7	83.7
Switzerland	58	48.3	9.9	53.8	81.3
the Netherlands	133	46.1	24.0	43.8	68.1
Turkey*	328	17.4	7.7	11.8	31.8
United Kingdom	585	52.2	24.5	51.4	83.9
Total*	3038	32.6	14.7	32.0	52.2

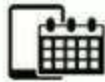
\* Data from Germany are only based on transplantation patients from 26 out of 20 transplantation centers, dialysis patients are not included and are therefore an underestimation of the true prevalence. Each year, around 120 patients are transplanted, of which 10% pre-emptively. The prevalence for Turkey is an underestimation of the true prevalence. Therefore, Germany and Turkey were excluded from the overall prevalence. In Italy, (pre-emptive) transplantation patients are not included; these numbers are an underestimation of true prevalence.

### Preliminary Benchmarking Report

Age-adjusted proportion of dialysis patients with overweight (BMI\*)



## ESPN/ERA-EDTA Registry: a successful collaboration



**12** consecutive years of data collection about **100** variables



Data on **>24,000** individual patients from **40** countries



**22** research fellows from **11** countries



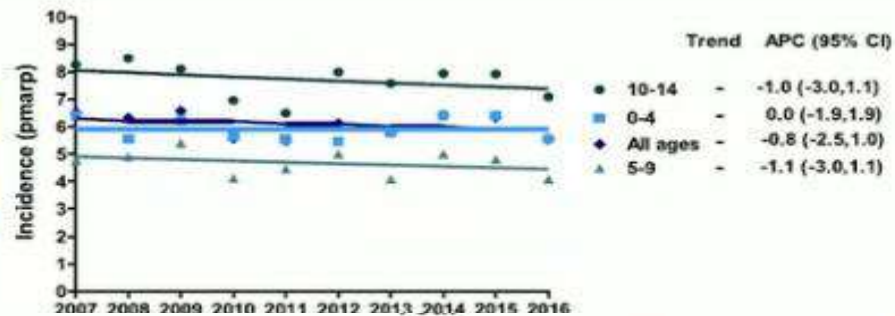
**48** publications in **10** medical journals



**>400** authorships with **230** unique authors from the ESPN

## Incidence of KRT

Stable incidence of KRT  
over 10 years: ~ 6 pmc in  
children < 15 years



N=4459  
Children 0-14 years old  
22 countries



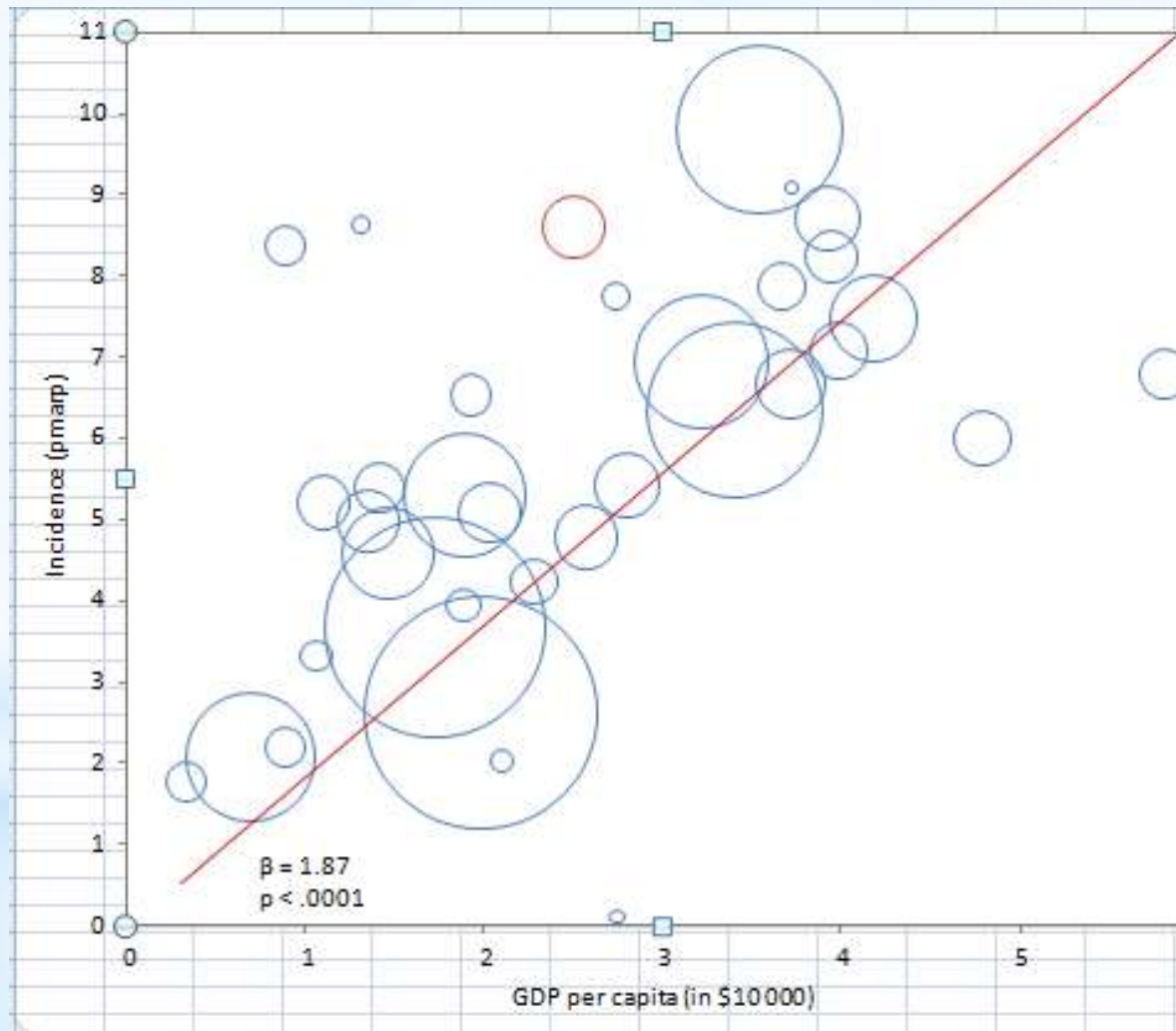
Bonthuis, Ped Nephrol (2021)





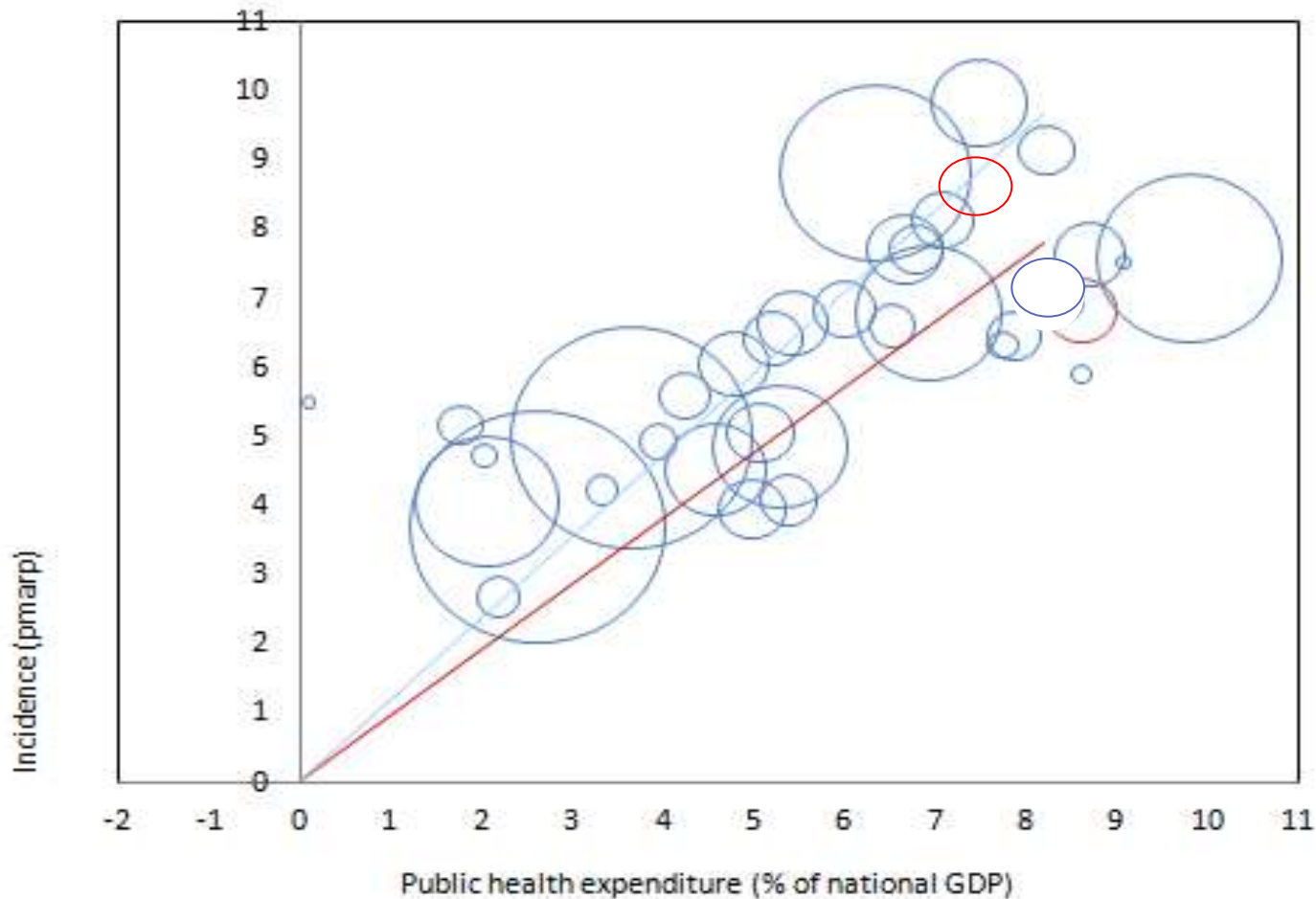
# Disparities in treatment rates of paediatric end-stage renal disease across Europe insights from the ESPN/ERA-EDTA Registry

Incidence of KRT correlated with GDP



# Disparities in treatment rates of paediatric end-stage renal disease across Europe insights from the ESPN/ERA-EDTA Registry

Incidence of KRT associated with public health expenditure (% of national GDP)



# Incidência de doentes com idade pediátrica em TSFR

(<sup>1</sup>)*pmart*=per million of the age -related population) 1451624

Ano	Incidência (0-14 anos) <i>pmart</i> Europa	Incidência (0-14 anos) <i>pmart</i> <sup>(1)</sup> Portugal
2007	6,5	9,1
2008	6,3	7,3
2009	6,6	10,5
2010	5,6	5,0
2011	5,5	9,5
2012	6,1	12,2
2013	5,8	4,6
2014	6,4	8,6
2015	6,3	6,8
2016	5,5	4,8
2017	6,1	7,9
2018	ND	6,4
2019	ND	6,4
2020	ND	4,8
	Média =6,0	<b>Média=7,4</b>

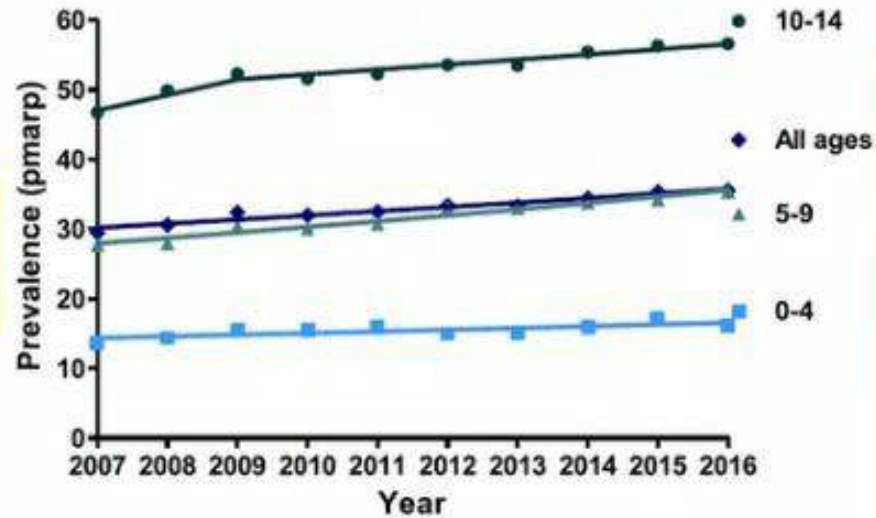
8,3

6,5



# Prevalence of KRT

Increasing prevalence of KRT over 10 years (2%/yr): ~ 35 pmc in children < 15 years



Bonthuis, Pediatr Nephrol 2021



N=4459  
Children 0-14 years old  
22 countries



Bonthuis, Ped Nephrol (2021)

# Prevalência pontual de Doentes Pediátricos em TSFR

População 0'-14 a-1 451 624  
População 0-18 a -3 084 001

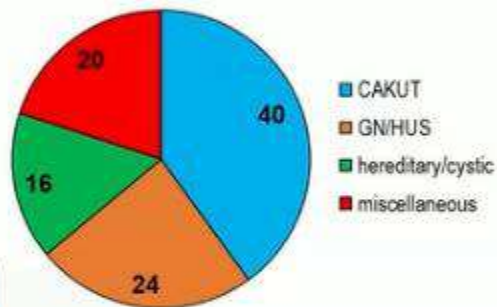
Ano	Prevalencia) (0-14 anos) p mart Europa	Prevalencia (0-14 anos) p mart Portugal	
31/12/2007	29,4	38,2	48,7
31/12/2008	30,0	40,9	
31/12/2009	32,4	46,9	
31/12/2010	32,0	45,4	
31/12/2011	32,5	50,5	
31/12/2012	33,4	59,6	
31/12/2013	33,5	54,7	
31/12/2014	34,4	57,1	51,7
31/12/2015	35,3	56,3	
31/12/2016	35,6	50,3	
31/12/2017	37,1	52,8	
31/12/2018	ND	51,6	
31/12/2019	ND	46,4	
21/12/2020	ND	47,5	

# Causes of of kidney failure

ESPN/ERA-EDTA Registry  
(Children < 15 years)

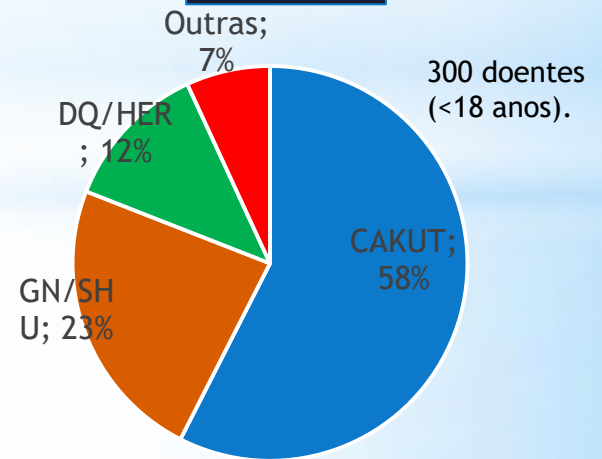


ESPN/ERA-EDTA Registry



European Society for Paediatric Nephrology

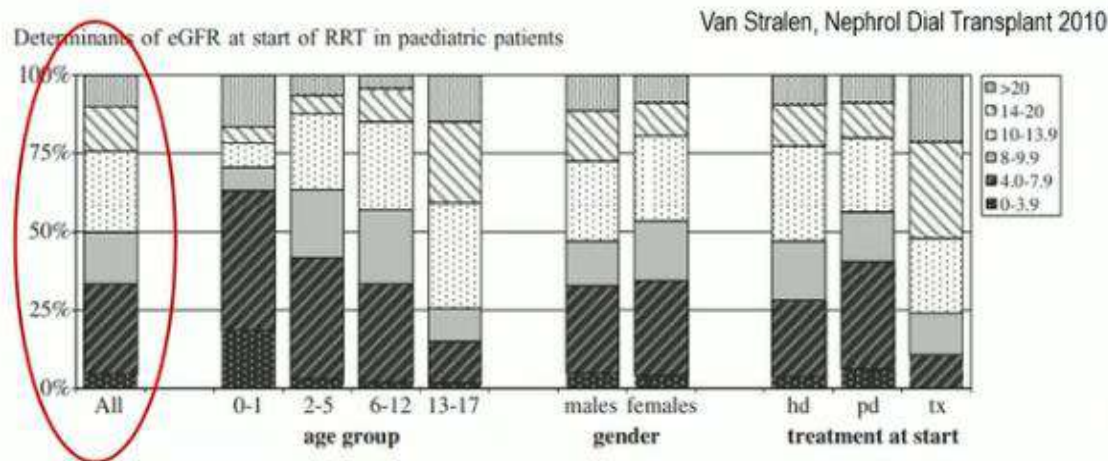
Portugal



■ CAKUT ■ GN/SHU ■ DQ/HER ■ Outras

# When to start kidney replacement therapy?

## eGFR at start of KRT



Median eGFR-10,4 ml/min/1,73 m<sup>2</sup>.

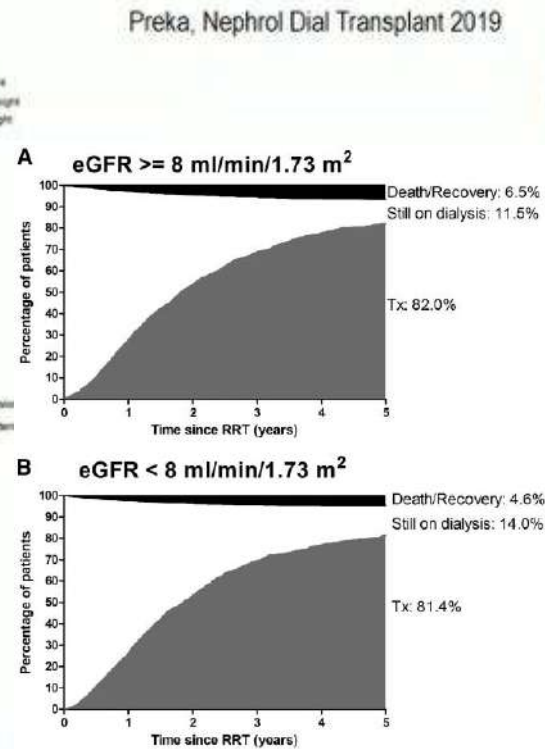
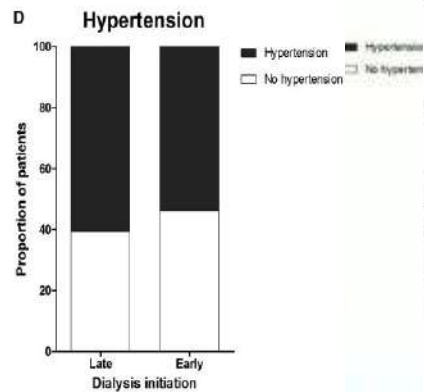
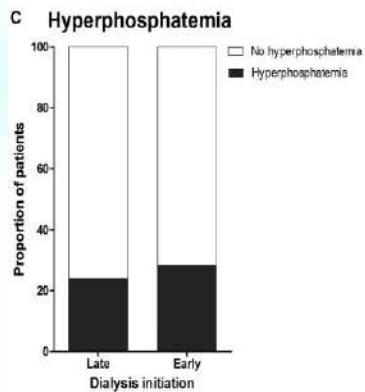
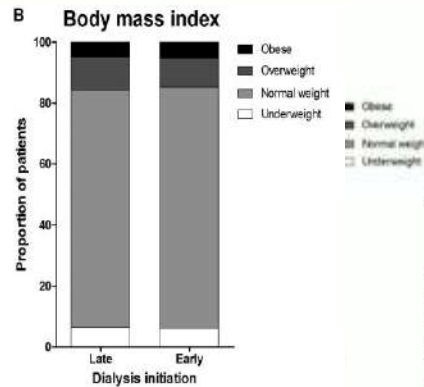
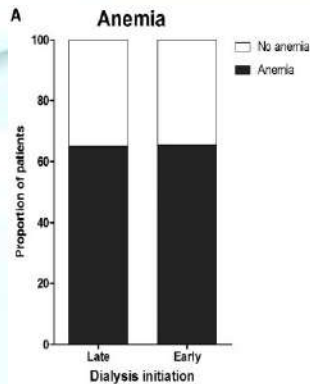
Lower eGFR at start:

- younger children,
- dialysis vs preemptive TR,
- late referral.



Research is needed to determine the consequences of these differences.

# Outcome of KRT by eGFR at start: < 8 vs. ≥ 8 ml/min/1.73 m<sup>2</sup>



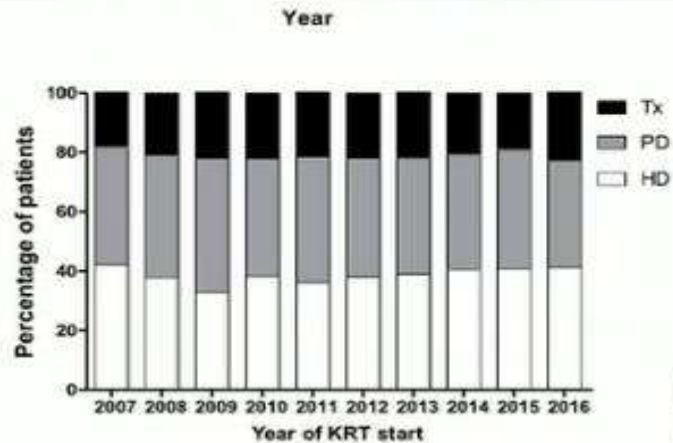
Higher eGFR at dialysis initiation is not associated with clinical benefit in children - mortality and access to transplant (except slightly higher BP in late starters)



## Modality of the first KRT

N=4459  
Children 0-14 years old  
22 countries

Modality at start of KRT:  
40% HD, 40% PD, 20% Tx



Bonthuis, Pediatr  
Nephrol 2021

# Recovery after start of chronic dialysis

Bonthuis, CJASN 2018

## What is the likelihood that children starting maintenance dialysis therapy will recover kidney function?

**CJASN**  
Clinical Journal of American Society of Nephrology

### Methods and Cohort



ESPN/ERA-EDTA Registry  
36 European countries



N= 6574  
Age <15 years  
Maintenance dialysis initiation 2000-2014



Outcome:  
Dialysis recovery = discontinuing  
dialysis for 30 days or more

### Recovery (entire cohort)

Recovery at  
2 years  
**2%**

Median time  
to recovery  
**5 months**  
(IQR 2-9.6)

### Recovery (by cause of kidney failure)



CAKUT

**0.8%**

Adjusted HR  
(95% CI)

ref



Vasculitis

**11%**

20.4  
(9.7-42.8)



Ischemia

**12%**

11.4  
(5.6-23.1)



HUS

**13%**

15.6  
(8.9-27.3)

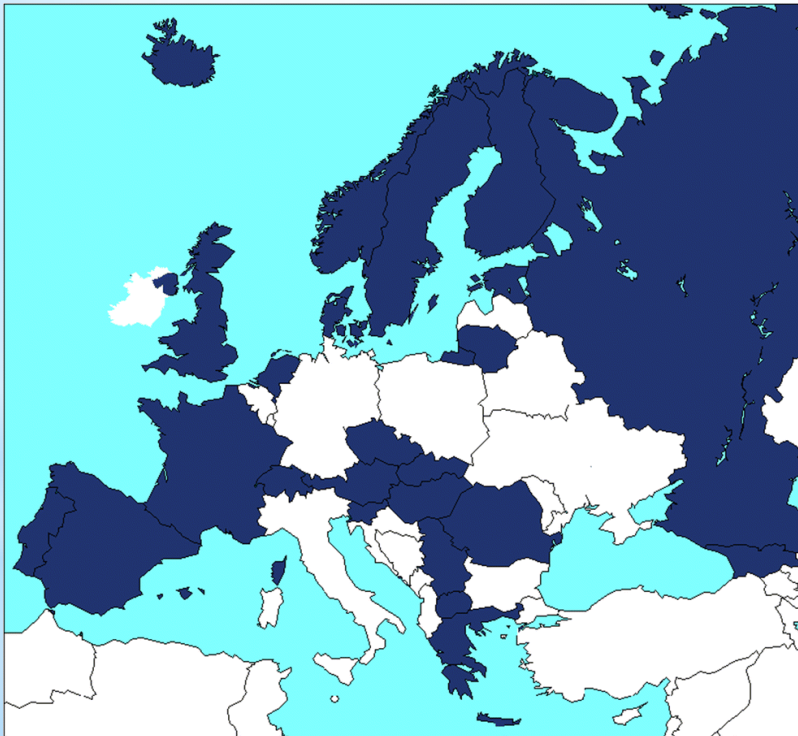
**Conclusions** There was a recovery rate of 2% within 2 years after initiation of maintenance dialysis in children. There is a clinically important chance of recovery in children with vasculitis, ischemic kidney failure and HUS.

Marjolein Bonthuis, Jérôme Harambat, Edienne Bérand, Karlen Cramberg, Ali Ozsova, Liliana Garateza, Maria Herbelius, Adrian C. Lungu, Timo Jahnukainen, Lukas Kalmegger, Genia Aricorta, Elisabeth Maurer, Runolfur Palsson, Manish D. Sethi, Sara Testa, Jaap W. Groothoff, and Kitty J. Jager. **Recovery of Kidney Function in Children Treated with Maintenance Dialysis.**  
doi: 10.2215/CJN.01500218

# Mortality

Ten year trends in epidemiology and outcomes of pediatric kidney replacement therapy in Europe: data from the ESPN/ERA-EDTA Registry .

Methods: All children aged <15 years starting KRT 2007-2016 in 22 European countries, 4459 patients aged 0-14 years participating.  
ESPN/ERA-EDTA Registry



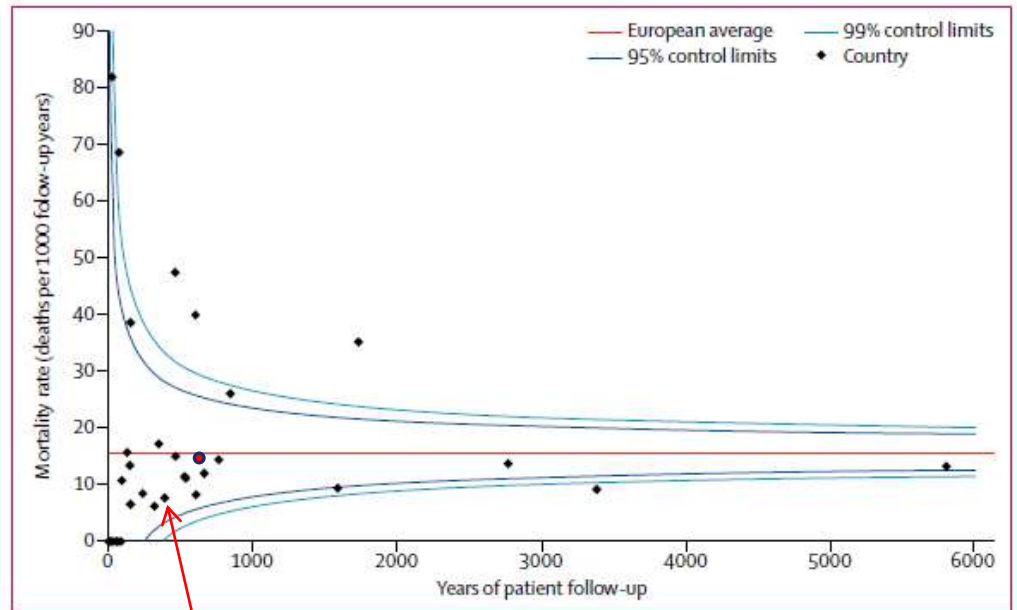
## Conclusions

Five-year patient survival was good, around 94%. It was unchanged over time

Bonthuis M et al. *Pediatr Nephrol* (2021)



# Country-specific KRT mortality rate



Portugal

Chesnaye et al, Lancet 2017

5 year RRT mortality rate was 15,8 deaths per 1000 patients years

## Country-specific KRT mortality rate

### Mortality rate significantly associated:

-Macroeconomics: Public health expenditure (per 1 SD increase);

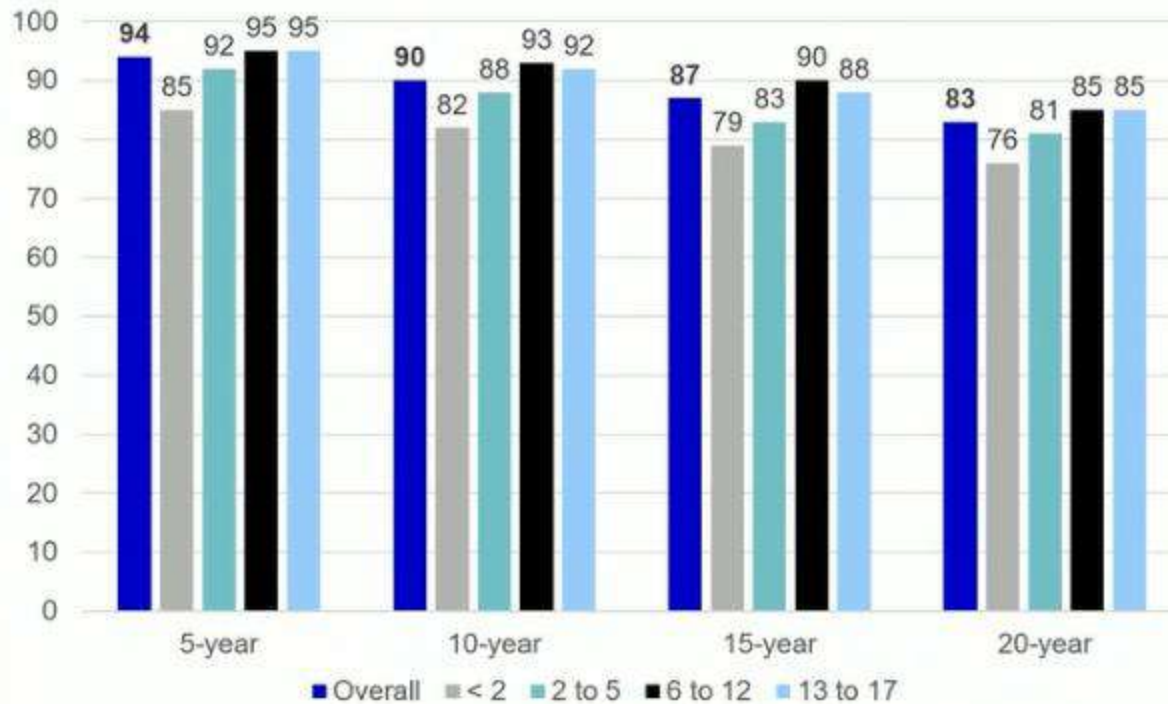
-Number pediatric nephrologists per million children ;

-Initial treatment modality . Dialysis (HD AND PD) compared a RT.

	aHR (95% CI)	P
<b>Macroeconomics</b>		
GDP per capita	-	-
Public health expenditure	0.69 (0.52-0.91)	0.008
Private health expenditure	0.87 (0.74-1.03)	0.11
<b>Child mortality</b>		
Neonatal mortality rate	1.21 (0.97-1.51)	0.10
Under-5 mortality rate	1.21 (0.96-1.53)	0.17
<b>Renal service indicators</b>		
Paediatric RRT incidence	1.02 (0.76-1.36)	0.92
Transplantation rate	0.86 (0.70-1.06)	0.16
Proportion pre-emptive Tx	1.00 (0.76-1.31)	0.98
No. PN centres pmc	0.92 (0.78-1.08)	0.30
No. paediatric nephrologists pmc	0.91 (0.85-0.98)	0.02
Reimbursement AHT	1.16 (0.72-1.87)	0.53
Reimbursement GH	1.49 (0.65-3.43)	0.35
<b>Initial treatment modality</b>		
HD	2.32 (1.53-3.52)	0.001
PD	1.76 (1.16-2.68)	0.008
Tx	Ref	Ref

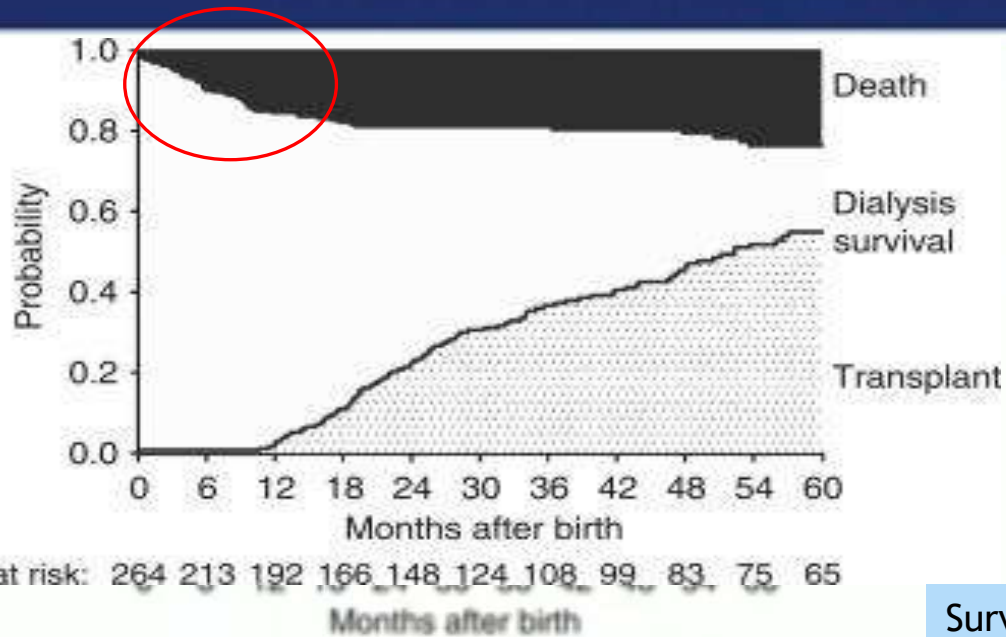
Chesnaye et al, Lancet 2017

## Long-term patient survival



Survival in pediatric patients in RRT - 94%, 90%, 87% and 83% at 5, 10, 15 and 20 years after 1st RRT treatment.

# Chronic KRT in neonates



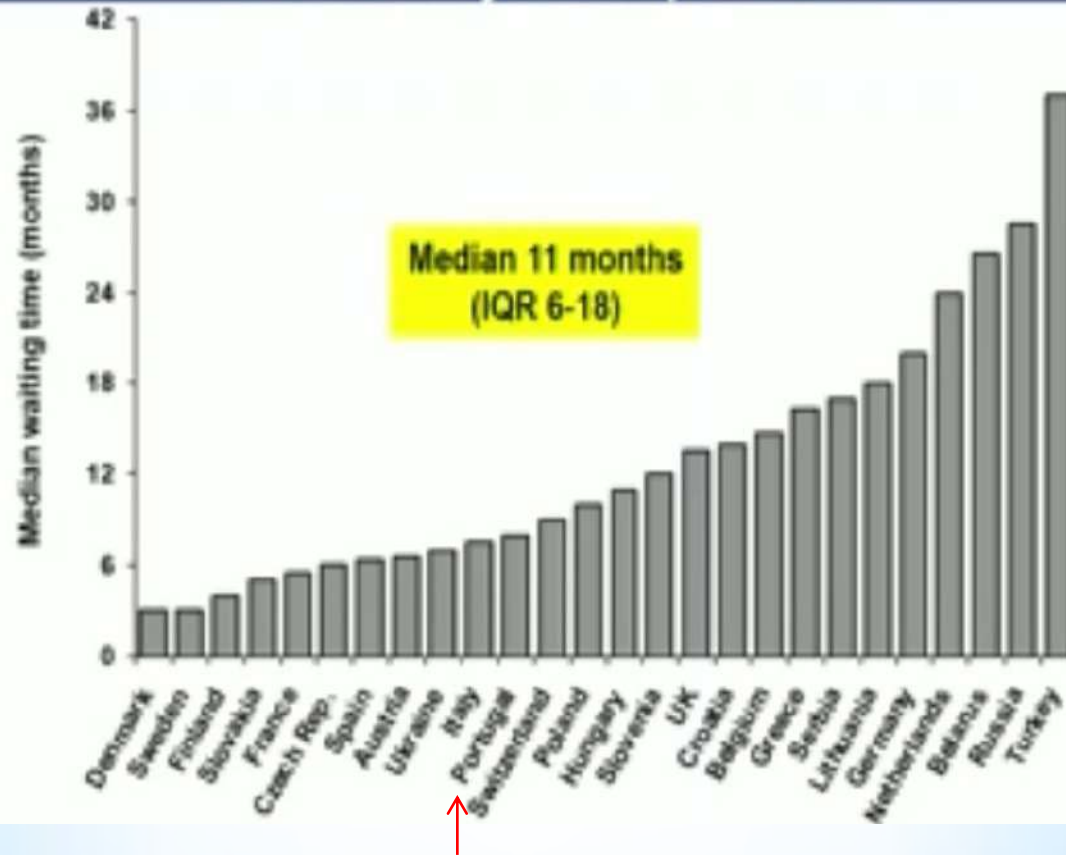
Survival after start RRT and the probability of getting a transplant

- . N=264
- . 45 (17%) died. 5-year survival rate of 76%
  - . Infection 35%
  - . Unknown 40%
- . 55% received a KT within 5 years

Van Stralen, Kidney Int 2014

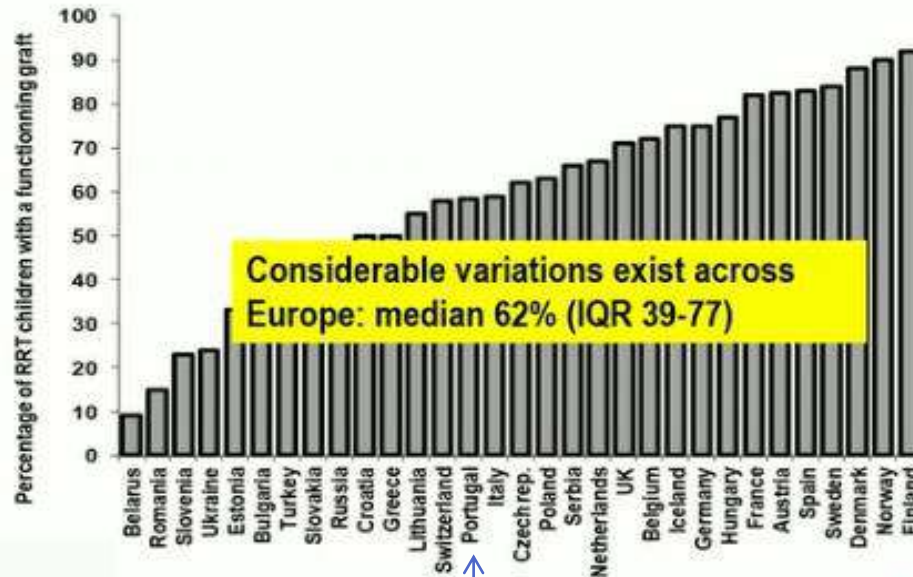
# Practice patterns and access to KT

## Disparities in waiting time on the list for DD kidney transplantation





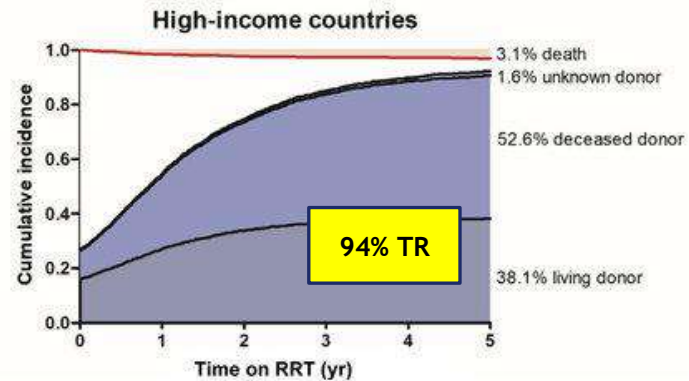
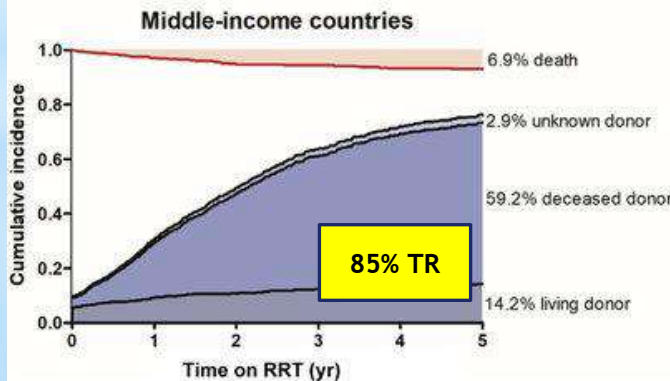
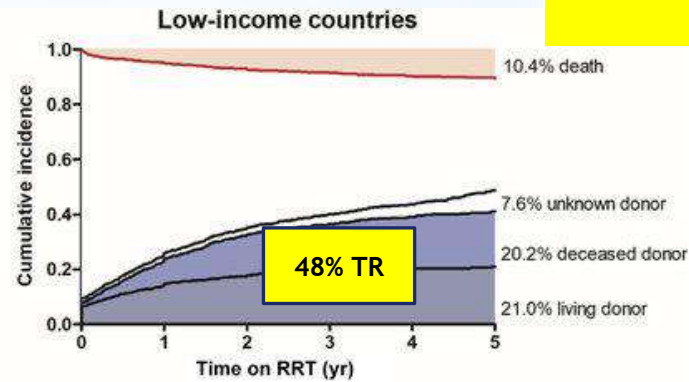
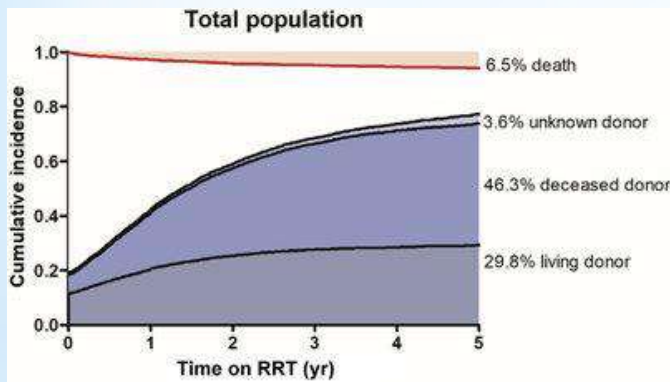
# Proportion of prevalent KRT children with a functioning graft by country



# Access to kidney transplantation (2007-2015)

Large disparities in access To KT, but no difference in graft survival between lower and higher income countries in Europe.

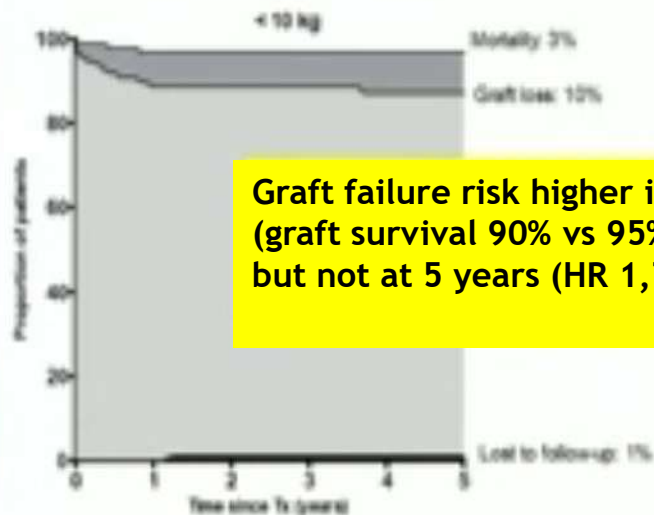
Bonthuis, Kidney Int 2020



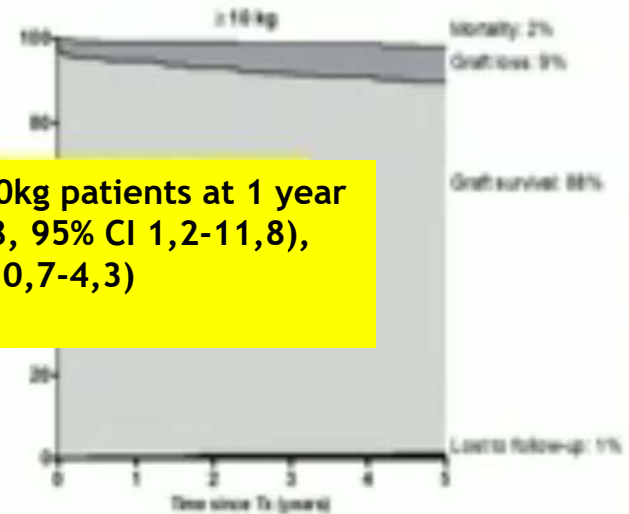
# Kidney transplantation in small children

601 children with start of KRT < 2.5 years and transplanted < 20 kg

KTx < 10 kg: N = 100



KTx ≥ 10 kg: N = 501



Graft failure risk higher in TR < 10kg patients at 1 year (graft survival 90% vs 95%; HR 3,8, 95% CI 1,2-11,8), but not at 5 years (HR 1,7 95% CI 0,7-4,3)



# Publicações da ESPN/ERA-EDTA (coautoria nefrologistas pediátricos portugueses)

1. [Determinants of eGFR at start of renal replacement therapy in paediatric patients.](#) Tizard EJ, Jager KJ, Schaefer F, Vondrak K, Groothoff JW, Podracká L, Holmberg C, Jankauskienė A, Lewis MA, van Damme-Lombaerts R, **Mota C**, Niaudet P, Novljan G, Peco-Antic A, Sahpazova E, Toots U, Verrina E. . *Nephrol Dial Transplant*. 2010 Oct;25(10):3325-32.
2. [Underweight, overweight and obesity in paediatric dialysis and renal transplant patients.](#) Bonthuis M, van Stralen KJ, Verrina E, Groothoff JW, Alonso Melgar A, Edefonti A, Fischbach M, **Mendes P**, Molchanova EA, Paripović D, Peco-Antic A, Printza N, Rees L, Rubik J, Stefanidis CJ, Sinha MD, Zagozdzon I, Jager KJ, Schaefer F; *NDT* 2013; 0:1-10
3. [Demographics of paediatric renal replacement therapy in Europe: a report of the ESPN/ERA-EDTA registry.](#) Chesnaye N, Bonthuis M, Schaefer F, Groothoff JW, Verrina E, Heaf JG, Jankauskiene A, Lukosiene V, Molchanova EA, **Mota C**, Peco-Antić A, Ratsch IM, Bjerre A, Roussinov DL, Sukalo A, Topaloglu R, Van Hoeck K, Zagozdzon I, Jager KJ, Van Stralen KJ; on behalf of the ESPN/ERA-EDTA registry. *Pediatr Nephrol*. 2014 Jul 21.
4. [Adult height in patients with advanced CKD requiring renal replacement therapy during childhood.](#) Harambat J, Bonthuis M, van Stralen KJ, Ariceta G, Battelino N, Bjerre A, Jahnukainen T, Leroy V, Reusz G, **Sandes AR**, Sinha MD, Groothoff JW, Combe C, Jager KJ, Verrina E, Schaefer F; ESPN/ERA-EDTA Registry. *Clin J Am Soc Nephrol*. 2014 Jan;9(1):92-9.

# Publicações da ESPN/ERA-EDTA (coautoria de nefrologistas portugueses)

5. **Mineral metabolism in European children living with a renal transplant: a European society for paediatric nephrology/european renal association-European dialysis and transplant association registry study.**  
[Bonthuis M](#), [Busutti M](#), [van Stralen KJ](#), [Jager KJ](#), [Baiko S](#), [Bakkaloğlu S](#), [Battelino N](#), [Gaydarova M](#), [Gianoglio B](#), [Parvex P](#), [Gomes C](#), [Heaf JG](#), [Podracka L](#), [Kuzmanovska D](#), [Molchanova MS](#), [Pankratenko TE](#), [Papachristou F](#), [Reusz G](#), [Sanahuja MJ](#), [Shroff R](#), [Groothoff JW](#), [Schaefer F](#), [Verrina E](#). *Clin J Am Soc Nephrol*. 2015 May 7;10(5):767-75. doi: 10.2215/CJN.06200614.
6. **Considerable variations in growth hormone policy and prescription in paediatric end-stage renal disease across European countries-a report from the ESPN/ERA-EDTA registry.**  
[van Huis M](#), [Bonthuis M](#), [Sahpazova E](#), [Mencarelli F](#), [Spasojević B](#), [Reusz G](#), [Caldas-Afonso A](#), [Bjerre A](#), [Baiko S](#), [Vondrak K](#), [Molchanova EA](#), [Kolvek G](#), [Zaikova N](#), [Böhm M](#), [Ariceta G](#), [Jager KJ](#), [Schaefer F](#)<sup>1</sup>, [van Stralen KJ](#), [Groothoff JW](#). *Nephrol Dial Transplant*. 2015 Apr 28. pii: gfv105.
7. **Infants requiring maintenance dialysis: outcomes hemodialysis and peritoneal dialysis.** [Vidal E](#), [van Stralen KJ](#), [Chesnaye NC](#), [Bonthuis M](#), [Holmberg C](#), [Zurowska A](#), [Trivelli A](#), [José 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](#), [van Stralen KJ](#), [Bonthuis M](#), [Groothoff JW](#), [Harambat J](#), [Schaefer F](#), [Verrina E](#). *Am J Kidney Dis*. 2017 May;69(5):617-625.
8. **Mortality risk disparities in children receiving 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](#). *Lancet*. 2017 May; 389(10084) :2128-2137.

# Publicações da ESPN/ERA-EDTA (coautoria de nefrologistas portugueses)

9. Association between timing of dialysis initiation and clinical outcomes in the paediatric population: an ESPN/ERA-EDTA registry study. [Evgenia Preka](#) , [Marjolein Bonthuis](#) , [Jerome Harambat](#) , [Kitty J Jager](#) , [Jaap W Groothoff](#) , [Sergey Baiko](#) , [Aysun K Bayazit](#) , [Michael Boehm](#) , [Mirjana Cvetkovic](#) , [Vidar O Edvardsson](#) , [Svitlana Fomina](#) , [James G Heaf](#) , [Tuula Holtta](#) , [Eva Kis](#) , [Gabriel Kolvek](#) , [Linda Koster-Kamphuis](#) , [Elena A Molchanova](#) , [Marina Muñoz](#) , [Gisela Neto](#) , [Gregor Novljan](#) , [Nikoleta Printza](#) , [Emilija Sahpazova](#) , [Lisa Sartz](#) , [Manish D Sinha](#) , [Enrico Vidal](#) , [Karel Vondrak](#) , [Isabelle Vrillon](#) , [Lutz T Weber](#) , [Marcus Weitz](#) , [Ilona Zagozdzon](#) , [Constantinos J Stefanidis](#) , [Sevcan A Bakkaloglu](#). *Nephrol Dial Transplant*. 2019 ; Nov 1;34(11):1932-1940.
10. Results in the ESPN/ERA-EDTA Registry suggest disparities in access to kidney transplantation but little variation in graft survival of children across Europe [Marjolein Bonthuis](#) , [Liz Cuperus](#) , [Nicholas C. Chesnaye](#) , [Sema Akman](#) , [Angel Alonso Melgar](#) , [Sergey Baiko](#) , [Antonia H. Bouts](#) , [Olivia Boyer](#) , [Kremena Dimitrova](#) , [Carmen do Carmo](#) , [Ryszard Grenda](#) , [James Heaf](#) , [Timo Jahnukainen](#) , [Augustina Jankauskiene](#) , [Lukas Kaltenecker](#) , [Mirjana Kostic](#) , [Stephen D. Marks](#) , [Andromachi Mitsioni](#) , [Gregor Novljan](#) , [Runolfur Palsson](#) , [Paloma Parvex](#) , [Ludmila Podracka](#) , [Anna Bjerre](#) , [Tomas Seeman](#) , [Jasna Slavicek](#) , [Tamas Szabo](#) , [Burkhard Tonshoff](#) , [Diletta D. Torres](#) , [Koen J. Van Hoeck](#) , [Susanne Westphal Ladfors](#) , [Jerome Harambat](#) , [Jaap W. Groothoff](#) and [Kitty J. Jager](#). *Kidney International* *Kidney International* 2020 ;98, 464-47
11. Growth patterns after kidney transplantation in European children over the past 25 years: An ESPN/ERA-EDTA Registry study. [Bonthuis M](#) , [Groothoff JW](#) , [Ariceta G](#) , [Baiko S](#) , [Battelino N](#) , [Bjerre A](#) , [Cransberg K](#) , [Kolvek G](#) , [Maxwell H](#) , [Miteva P](#) , [Molchanova MS](#) , [Neuhaus TJ](#) , [Pape L](#) , [Reusz G](#) , [Rousset-Rouviere C](#) , [Sandes AR](#) , [Topaloglu R](#) , [van Dyck M](#) , [Ylinen E](#) , [Zagozdzon I](#) , [Jager KJ](#) , [Harambat J](#). *Transplantation* 2020; 104(1):137-144.
12. Comorbidities. Disparities in treatment and outcome of kidney replacement therapy in children with comorbidities: An ESPN/ERA-EDTA Registry study. [Raphael Schild](#)\*1 , [Simeon Dupont](#)\*1 , [Jérôme Harambat](#)2 , [Enrico Vidal](#)3 , [Ayşe Balat](#)4 , [Csaba Berecki](#)5 , [Beata Bieniaś](#)6 , [Per Brandström](#)7 , [Francoise Broux](#)8 , [Silvia Consolo](#)9 , [Ivana Gojkovic](#)10 , [Jaap W. Groothoff](#)11 , [James G. Heaf](#)12 , [Holger Hubmann](#)13 , [Shaun Mannings](#)14 , [Tatiana E. Pankratenko](#)15 , [Fotios Papachristou](#)16 , [Lucy A. Plumb](#)17 , [Ludmila Podracka](#)18 , [Sylwester Prokurat](#)19 , [Anna V. Reisaeter](#)20 , [Carolina Cordinhã](#)21 , [Paula Seikku](#)22 , [Enkeledja Shkurti](#)23 , [Giuseppina Sparta](#)24 , [Karel Vondrak](#)25 , [Kitty J. Jager](#)26 , [Jun Oh](#)1 , [Marjolein Bonthuis](#)26. For submission to *Clinical Journal of the American Society of Nephrology*.