Registo da DRC5 em TSFR 2017

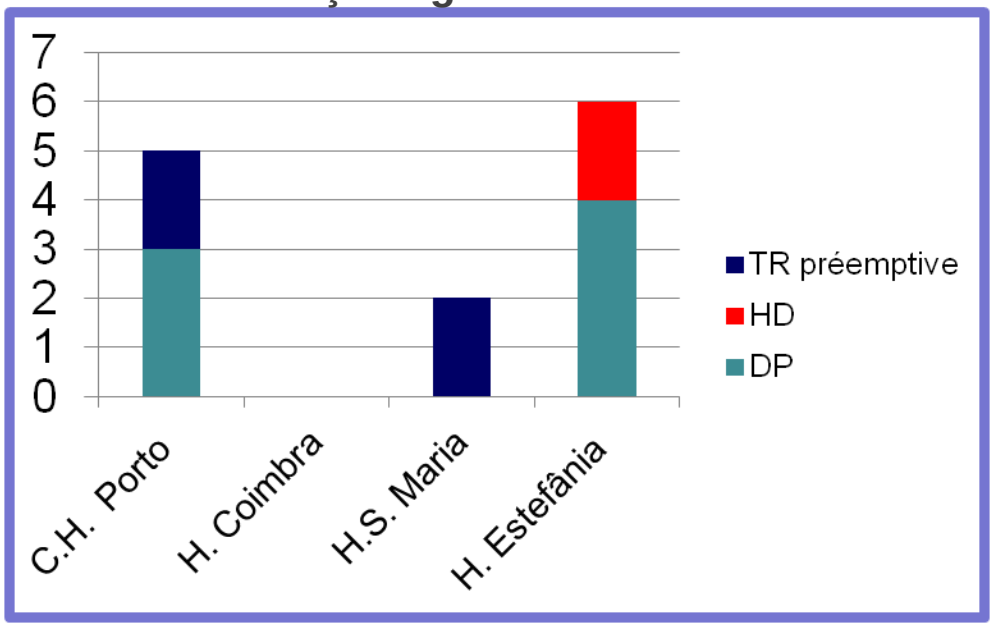
- **Fonte: Registo enviado anualmente para *European Society of Pediatric Nephrology (*ESPN), com a contribuição das Unidades de Nefrologia Pediátrica Portuguesa.**

- Conceição Mota (C.H.Porto)
- Rosário Stone (H.S. Maria, Lisboa)
- Margarida Abranches (H. D. Estefânia, Lisboa)
- Clara Gomes (H. Pediátrico de Coimbra)

Doentes que iniciaram TSFR no ano de 2017

N=13

8 género masculino;
2 de raça negra

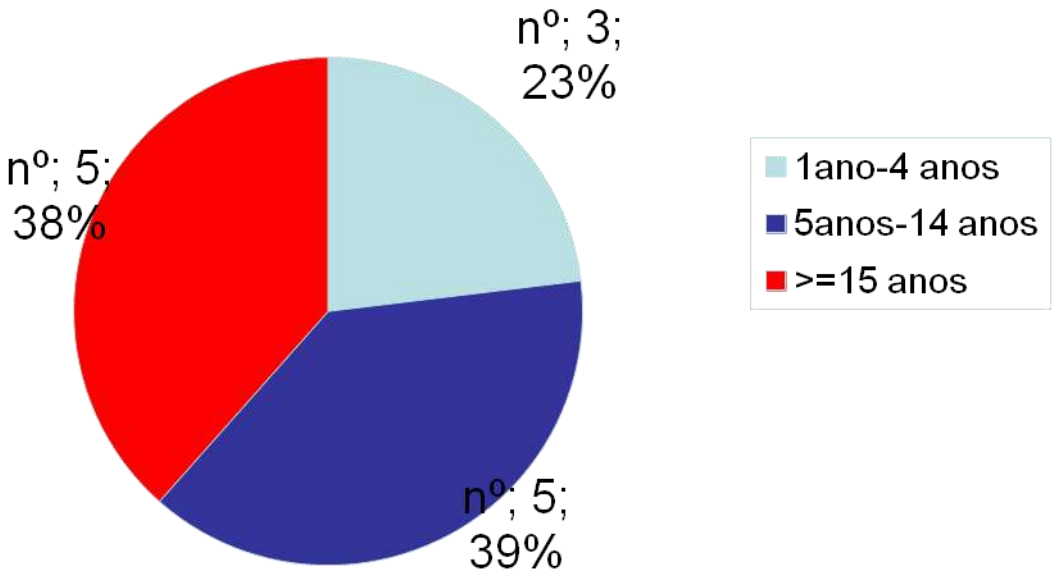


Tipo de TSFR (dia1)
7 em DP
2 em HD
4 TR préemptive.

Idade à data da 1º TSFR

Doentes incidentes em 2017

Média=10,3± 6,1anos



Doentes pediátricos que foram transplantados no ano de 2017

N=13 transplantes

- Centro Hospitalar do Porto; n=8
- H. S. Maria, Lisboa; n=5

- Dador. Rim de dador cadáver/vivo=10/3
- TR *préemptive*. n= 4

Movimento
-ano de 2017-

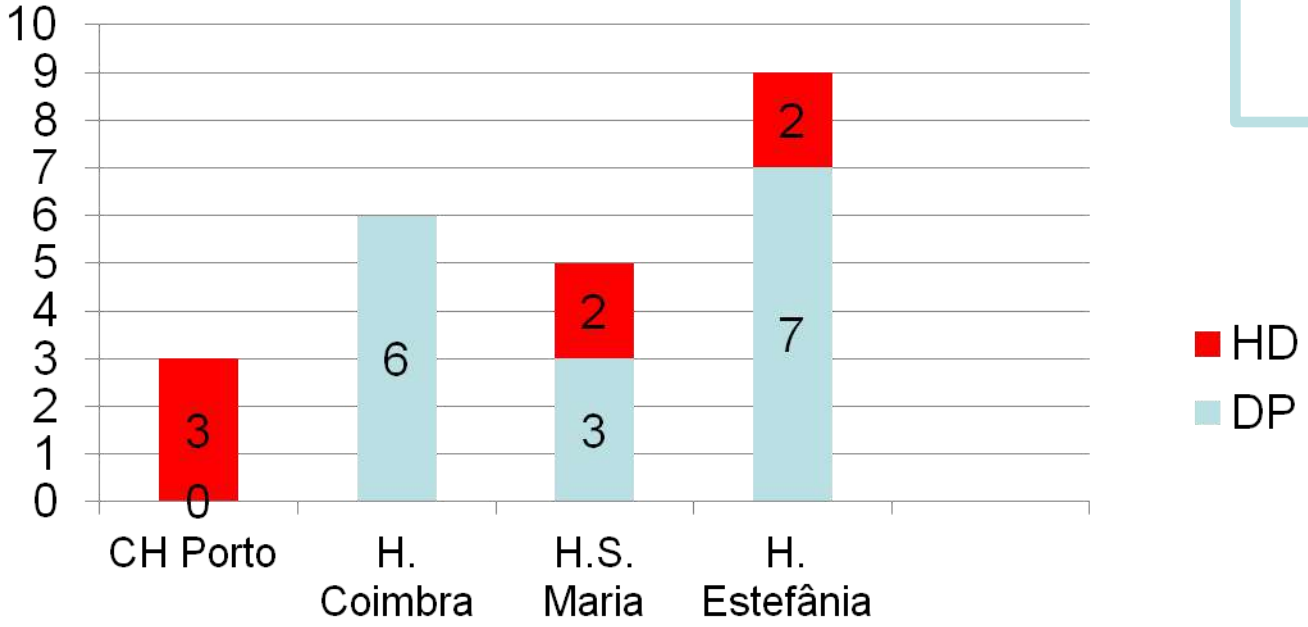
IN	
Novos doentes	13

OUT	
Número de TR	13
Mortos	0
Recuperação da FR	0
Saída de registo→ transferência	10(+1)

Mudança de TSFR	
TR→HD	1 (rejeiçao celular)
TR→DP	1(não função imediata)

Prevalência pontual de doentes em diálise em ambiente pediátrico
(31/12/2017)

N=23

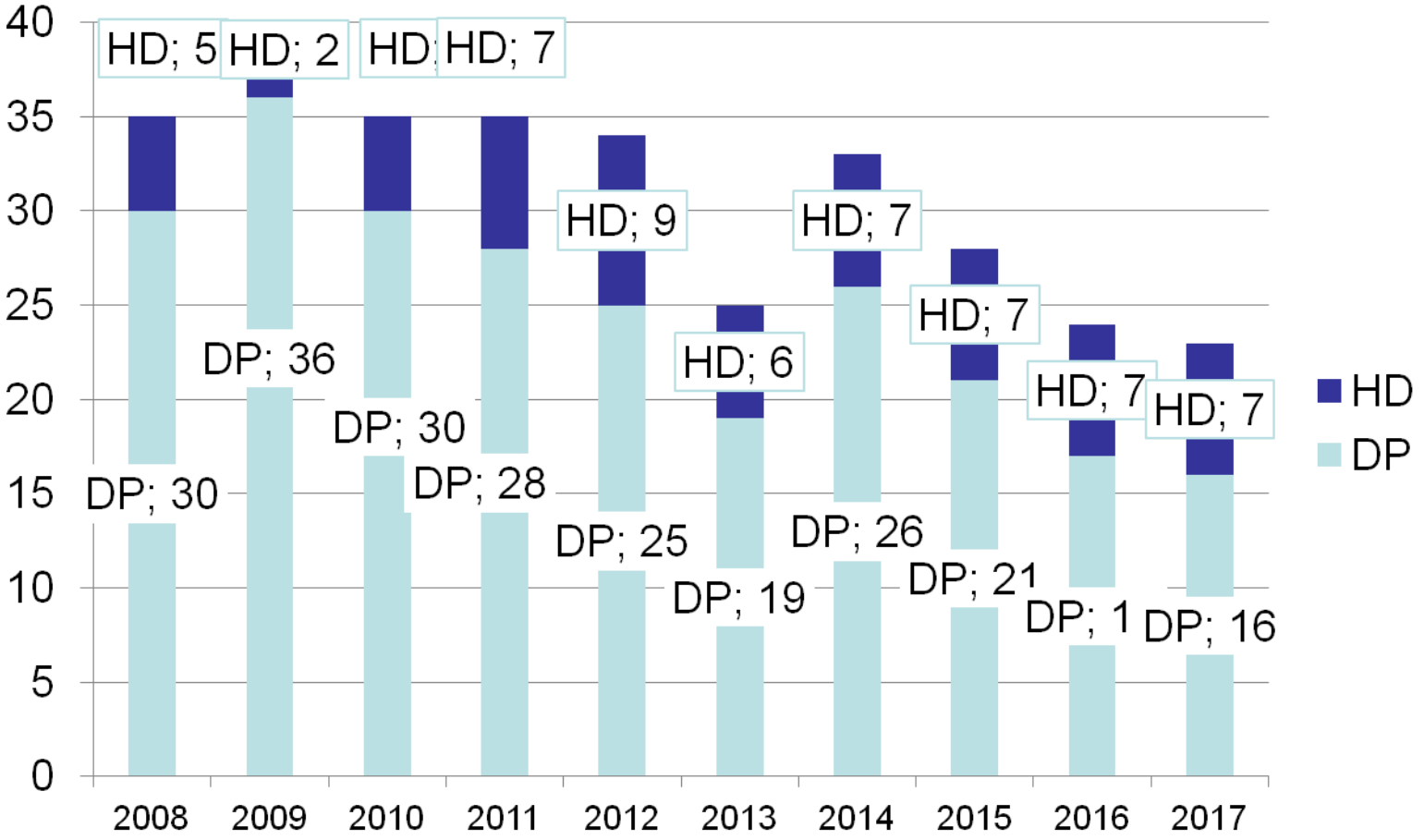


DP=16(69,5%)
HD=7(30,5%)

Evolução do registo

SPP
SOCIEDADE PORTUGUESA DE PEDIATRIA
Sociedade de Nefrologia

Prevalência pontual de doentes em diálise a 31 de Dezembro



Prevalência pontual de doentes com transplante renal (enxerto funcionante) com seguimento em Unidades de Nefrologia Pediátrica a 31/12/2017

N=95

H S Maria-Lisboa	50
C Hospitalar Porto	35
H Pediátrico Coimbra	10

Evolução do registo

SPP
SOCIEDADE PORTUGUESA DE PEDIATRIA
Sociedade de Nefrologia

Ano	Novos doentes	Transplante / ano
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

Incidência de doentes com idade pediátrica em TSFR

	Incidência (0-14 anos) Nº Portugal	Incidência (0-14 anos) pmart[1] Portugal	Incidência (0-14 anos) pmart Europa	Incidência (0-18 anos) Nº Portugal
2008	13	7,9	5,2	16
2009	15	9,3	5,8	18
2010	11	6,7	4,5	17
2011	15	9,5	4,4	18
2012	16	10,2	3,9	24
2013	7	4,6	4,7	11
2014	13	8,6	5,4	17
2015	10	6,8	6,0	16
2016	7	4,7	.5,6	11
2017	8	5,5	ND	13

[1]*pmart=per million of the age –related population) 1451624*

Prevalência pontual de Doentes Pediátricos em TSFR

Populacao 0'-14 a-1 451 624
Populaçao 0-18ªa -3 084 001

Ano	Prevalência (0-14 anos) Nº Portugal	Prevalencia (0-14 anos) Pmart Portugal	Prevalencia) (0-14 anos) pmart Europa	Prevalencia 0-18 anos (pmart Portugal
31/12/2007	64	39,2	ND	-
31/12/2008	56	34,4	ND	-
31/12/2009	70	43,2	ND	-
31/12/2010	77	47,1	ND	-
31/12/2011	77	49,1	26,1	-
31/12/2012	84	53,8	32,6	91,1
31/12/2013	78	50,8	29,7	80,2
31/12/2014	79	52,5	38,8	84,9
31/12/2015	80	54,2	35,5	92,8
31/12/2016	73	50,3	34,9	102.1
31/12/2017	75	52,8	ND	105,5

european society for paediatric nephrology

ESPN/ERA-EDTA Registry

ESPN http://espn-online.org/index.php ESPN/ERA-EDTA Registry http://www.espn-reg.org

An update on the Registry- November 2018

Jaap Groothoff and Jérôme Harambat

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, 36 countries are participating in the Registry, providing information on nearly 22,000 patients who started RRT before the age of 20.

Since its start in 2007, the Registry published 40 scientific papers in high-ranking medical journals. So far, in 2018, four papers based on Registry data have been accepted for publication by different journals and some others have been submitted. The full publication list can be found below.

An important part of the Registry's research activities arise from the successful internship programme, which has so far led to 18 fellows from 11 European countries visiting the Registry. In 2018, two fellows joined the Registry. Michael Böhm from Vienna, Austria, started a project on the minimum weight at which infants can be transplanted.

Enrico Vidal from Padova, Italy, obtained an ERA-EDTA educational grant to perform a fellowship on antihypertensive medications in October and November 2018.

If you are also 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: m.bonthuis@amc.uva.nl.

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

european society for paediatric nephrology

ESPN/ERA-EDTA Registry

era edta
LEADING EUROPEAN NEPHROLOGY

ESPN http://espn-online.org/index.php ESPN/ERA-EDTA Registry http://www.espn-reg.org

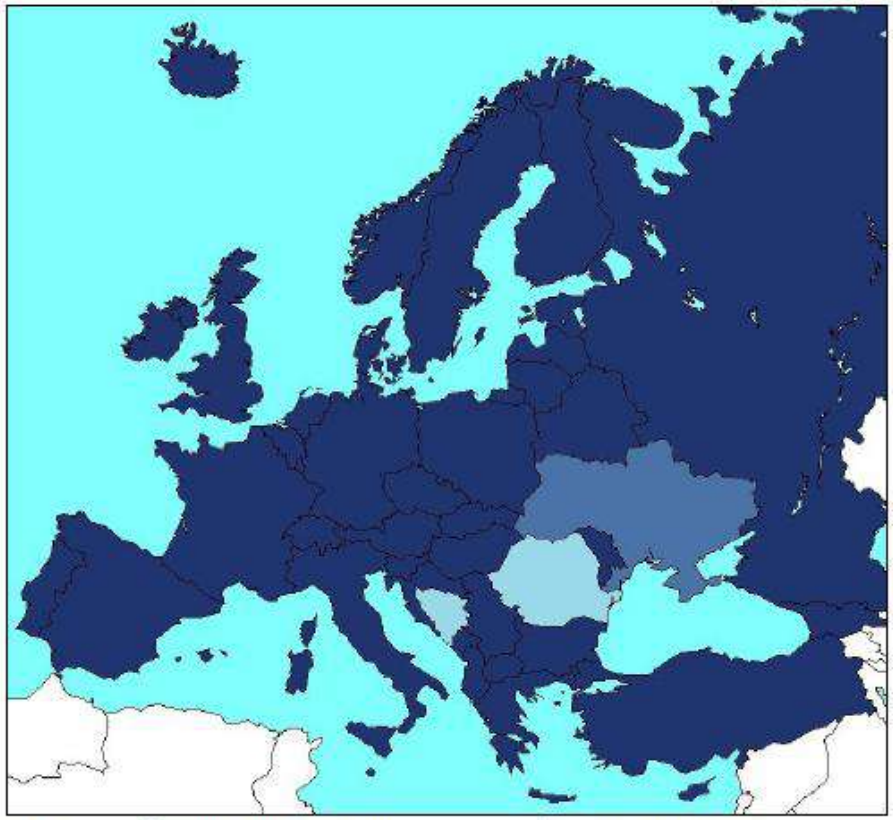
An update on the Registry- November 2018

- Registo da ESPN de 9 anos consecutivos.
- 36 países
- População: 22000 doentes DRC5 em TSFR com idade < 20 anos.

- Apresentação do Registo/Análise anual.
- Publicações em revistas médicas indexadas e com ↑ fator de impato.

An update on the Registry- November 2018

National registries that contributed data as of November 2018



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

Incident Patients (2016)

Incident paediatric patients accepted for renal replacement therapy in 2016 and general population characteristics of countries contributing 2016 data to the ESPN/ERA-EDTA Registry.

Country	Total RRT patients 0-14 years N	pmarp	General Population Characteristics Children 0-14 years N	Total Population 0-99 years N	Children 0-14 years percent
Albania	5	9.7	515,961	2,881,308	17.9
Austria	8	6.4	1,246,847	8,700,471	14.3
Belarus	6	3.8	1,579,277	9,504,704	16.6
Bosnia and Herzegovina	1	1.8	543,719	3,531,159	15.4
Bulgaria	2	2.0	999,613	7,127,823	14.0
Croatia	7	11.5	607,461	4,172,439	14.6
Cyprus	2	14.3	139,423	851,561	16.4
Czech Republic	7	4.3	1,635,496	10,566,334	15.5
Denmark	5	5.2	960,922	5,728,010	16.8
Estonia	0	0.0	212,526	1,315,788	16.2
Finland	3	3.4	895,101	5,495,301	16.3
France	82	6.7	12,293,404	66,859,768	18.4
FYR of Macedonia	0	0.0	344,114	2,0722,40	16.6
Georgia	4	5.3	760,455	4,483,230	17.0
Germany-KFH*	43	3.9	10,964,847	82,348,668	13.3
Germany-CERTAIN*	10	0.9	10,964,847	82,348,668	13.3
Greece	4	2.6	1,555,716	10,775,971	14.4
Hungary	9	6.3	1,423,656	9,814,023	14.5
Iceland	0	0.0	66,678	335,436	19.9
Ireland	12	11.9	1,006,479	4,755,336	21.2
Italy*	25	3.0	8,232,222	60,627,497	13.6
Lithuania	1	2.4	422,934	2,868,229	14.7
Malta	1	15.5	64,455	455,356	14.2
Norway	5	5.3	935,655	5,234,518	17.9
Portugal	7	4.8	1,451,624	10,325,451	14.1
Republic of Serbia	5	4.9	1,016,587	7,058,325	14.4
Romania	12	3.9	3,061,009	19,702,332	15.5
Russia	77	3.6	21,534,456	142,368,368	15.1
Slovakia	2	2.4	836,136	5,430,798	15.4
Slovenia	3	9.8	307,492	2,065,042	14.9
Spain	50	7.1	7,015,292	46,484,065	15.1
Sweden	15	8.6	1,739,070	9,923,085	17.5
Switzerland	9	7.2	1,254,991	8,419,950	14.9
the Netherlands	18	6.4	2,790,770	16,939,923	16.6
Turkey*	62	3.3	18,906,001	79,277,963	23.8
Ukraine	26	4.0	6,535,536	42,414,905	15.4
United Kingdom	104	8.9	11,67,832	65,648,054	17.8
Total*	492	5.6	87,429,687	544,399,695	16.1

* In 2016, 110 patients under the age of 21 years were transplanted in 18 transplant centres in Germany. In Italy, (pre-emptive) transplantation patients are not included; these numbers are an underestimation of true incidence. The incidence in Turkey is an underestimation of the true incidence. Therefore, Germany, Italy, and

Treatment modality at start of RRT in 2016

Treatment modality at day 1, among patients < 15 years of age starting RRT in 2016.
Patients from Germany, Italy, and Turkey are excluded.

	N	Percent	Pmarp
HD at start	204	41.4	2.35
PD at start	187	38.0	2.15
Pre-emptive transplantation	100	20.3	1.15
Unknown	1	0.2	0.01

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 2016.

	N	Percent
eGFR<8 ml min^{-1} per 1.73 m^2	81	36.3
eGFR 8- 15 ml min^{-1} per 1.73 m^2	111	49.8
eGFR>15 ml min^{-1} per 1.73 m^2	31	13.9

Table 3: PRD distribution at start of RRT in 2016

Cause of renal failure, among patients < 15 years of age, starting RRT in 2016 according to new and old PRD coding.

	N		Percent		Pmarp	
	New	Old	New	Old	New	Old
CAKUT	179	146	36.4	29.7	2.05	1.67
Glomerulonephritis	98	83	19.9	16.9	1.12	0.95
Cystic kidney disease	50	55	10.1	11.2	0.57	0.63
Hereditary nephropathy	-	37	-	7.5	-	0.42
Metabolic and tubulointerstitial disorders	23	7	4.7	1.4	0.26	0.08
Toxic/ischemic renal failure	11	6	2.2	1.2	0.13	0.07
HUS	24	24	4.9	4.9	0.27	0.27
Vascular	9	7	1.8	1.4	0.10	0.08
Miscellaneous	89	83	18.1	16.9	1.02	0.95
Unknown	9	44	1.0	8.9	0.10	0.50

Prevalent Patients (2016)

Table 5: Prevalent Patients (continued)

Prevalent paediatric patients on renal replacement therapy on the 31st of December 2016. 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	3.7	16.2	5.8	0.0	3.9
Austria	51.5	31.4	4.8	3.2	33.7
Belarus	28.3	15.7	2.5	3.8	17.7
Bosnia and Herzegovina	25.1	11.3	12.9	0.0	5.5
Bulgaria	13.6	6.2	3.0	2.0	5.0
Croatia	54.4	47.4	1.6	31.3	18.1
Cyprus	55.9	88.5	7.2	35.9	28.7
Czech Republic	33.4	21.3	3.1	4.9	19.6
Denmark	50.7	29.9	2.1	4.2	33.3
Estonia	18.3	19.3	0.0	0.0	18.8
Finland	96.4	61.7	1.1	4.5	74.9
France	44.9	31.3	6.9	3.7	27.7
FYR of Macedonia	16.9	12.0	5.8	5.8	2.9
Georgia	22.5	16.7	5.3	10.5	3.9
Germany-KFH*	14.4	10.1	4.6	9.6	-
Germany-CERTAIN*	32.1	12.8	-	-	22.5
Greece	42.6	23.8	9.0	11.6	13.5
Hungary	47.9	30.3	2.6	4.9	31.6
Iceland	88.1	0.0	0.0	0.0	45.0
Ireland	83.6	67.1	7.9	10.9	56.6
Italy*	35.9	24.5	4.6	7.5	-
Lithuania	18.5	24.3	0.0	9.5	11.6
Malta	30.0	32.1	0.0	0.0	31.0
Norway	64.7	46.2	0.0	2.1	54.5
Portugal	57.9	42.4	1.4	9.6	39.3
Republic of Serbia	28.7	30.4	7.9	3.0	18.7
Romania	16.5	15.5	10.1	4.6	1.3
Russia	24.3	16.4	5.0	9.0	6.5
Slovakia	18.7	19.7	7.2	7.2	4.8
Slovenia	37.9	26.8	9.8	6.5	16.3
Spain	58.1	36.2	5.0	4.0	38.2
Sweden	67.1	47.4	2.3	8.1	47.2
Switzerland*	41.9	34.4	2.4	7.2	28.7
the Netherlands	55.3	36.0	3.2	2.1	40.1
Turkey*	16.3	15.1	2.4	6.9	6.3
Ukraine	13.7	12.6	3.8	2.8	6.6
United Kingdom	70.1	41.1	7.5	7.2	40.9
Total*	41.1	27.5	5.4	6.1	22.5

* In 2016, 110 patients under the age of 21 years were transplanted in 18 transplant centres in Germany. In Italy, (pre-emptive) transplantation patients are not included; these numbers are an underestimation of true prevalence. The prevalence in Turkey is an underestimation of the true prevalence. In Switzerland, not all patients provided informed consent resulting in an underestimation of the true prevalence. Therefore, Germany, Italy, Turkey, and Switzerland were excluded from the overall prevalence.

Figure 1: Five-year survival

Incident RRT patients under the age of 15 starting RRT from 2007 onwards. Follow-up till 31st of December 2016.

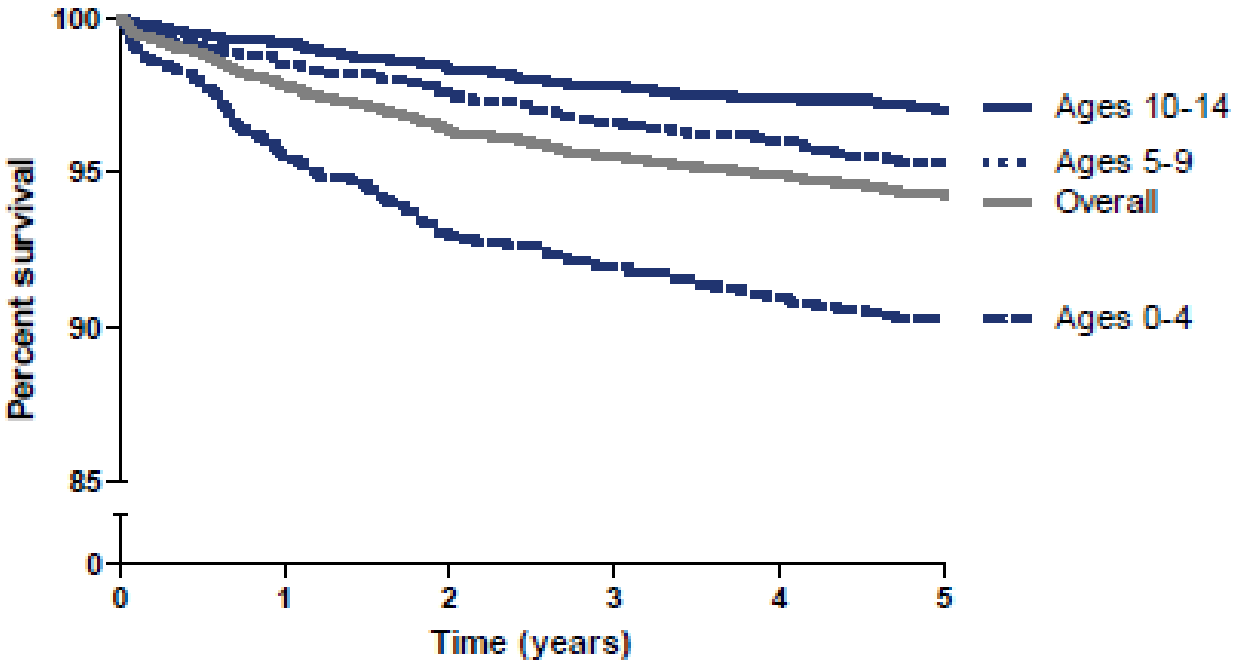


Table 7: Causes of Death

Causes of death according to the ERA-EDTA coding list. Incident RRT patients under the age of 15 starting RRT from 2007 onwards are included. Follow-up till 31st of December 2016

Cause of death	Number of deaths	Percent
Myocardial ischemia and infarction	2	0.6
Cardiac failure	31	8.9
Cardiac arrest/sudden death other cause	36	10.3
Cerebro-vascular accident	23	6.6
Infection	71	20.3
Suicide/refusal or cessation of treatment	5	1.4
Treatment withdrawn	6	1.7
Malignant disease	11	3.1
Other identified cause of death	60	17.1
Cause of death uncertain/not determined	105	30.0

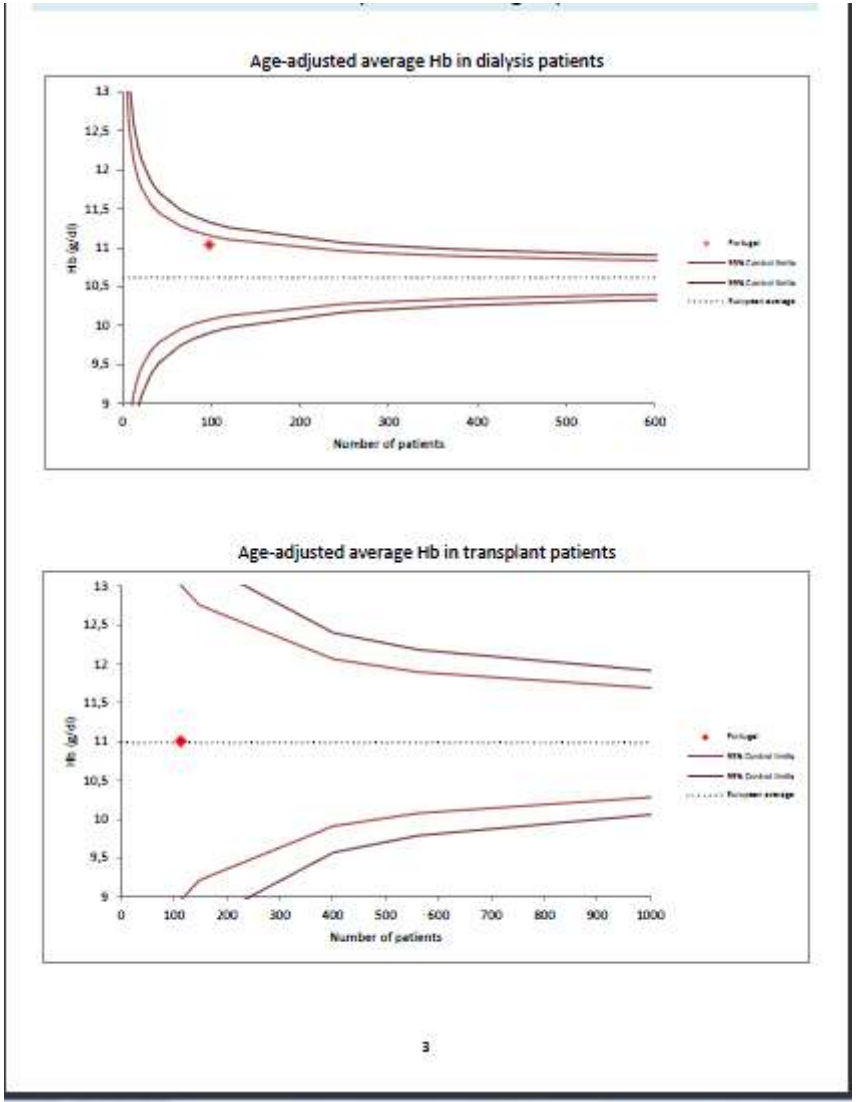
Preliminary Benchmarking Report

Funnel-plots allow us to objectively compare the performace of individual countries. For each clinical indicator, country estimates (y-axis) are plotted against the number of patients (x-axis). The control limits form a 'funnel' around the European averagen and reflect the precision of the estimate based on the number of patients in each country. Countries that fall outside these limits are doing either better or worse compared to the European average. Nevertheless, in countries with a small number of patients (<10), these limits may be imprecise. The plots are based on patient measurements collected since 2007 for patients aged 0-14 years. All country estimates are adjusted for the effect of age. Details on the methods used can be found in the appendix.

Dados fornecidos em Novembro de 2018 pelo
ESPN registry
Comparação de Portugal com a média de todos os 36 países europeus que contribuem para o registo da ESPN

Benchmarking Report

Age-adjusted average Hb



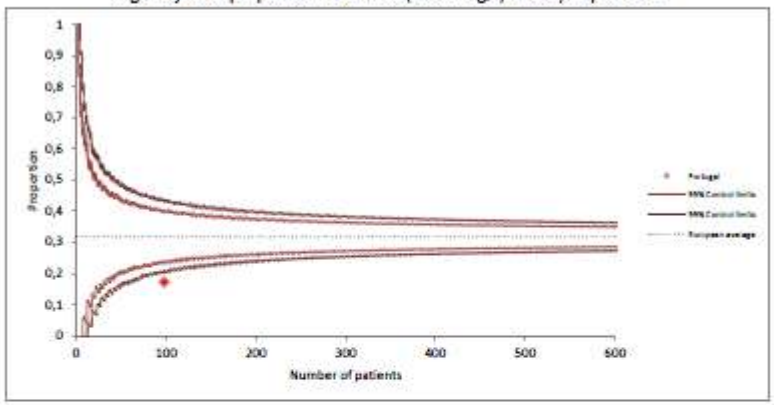
Ponto vermelho Portugal

Benchmarking Report

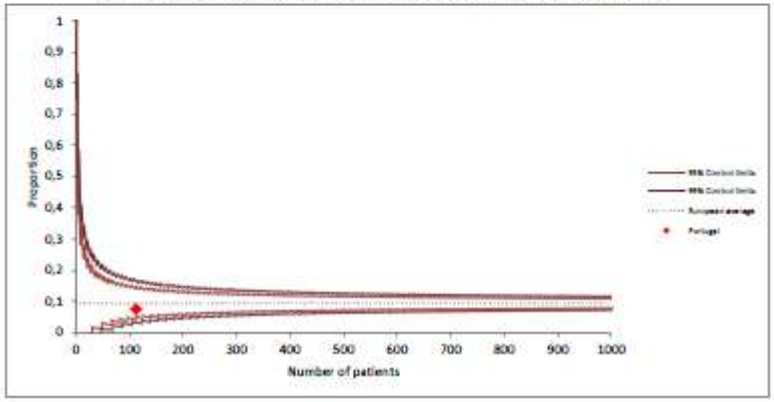
Age-adjusted proportion of anemia

Preliminary Benchmarking Report

Age-adjusted proportion of anemia (Hb < 10 g/l) in dialysis patients



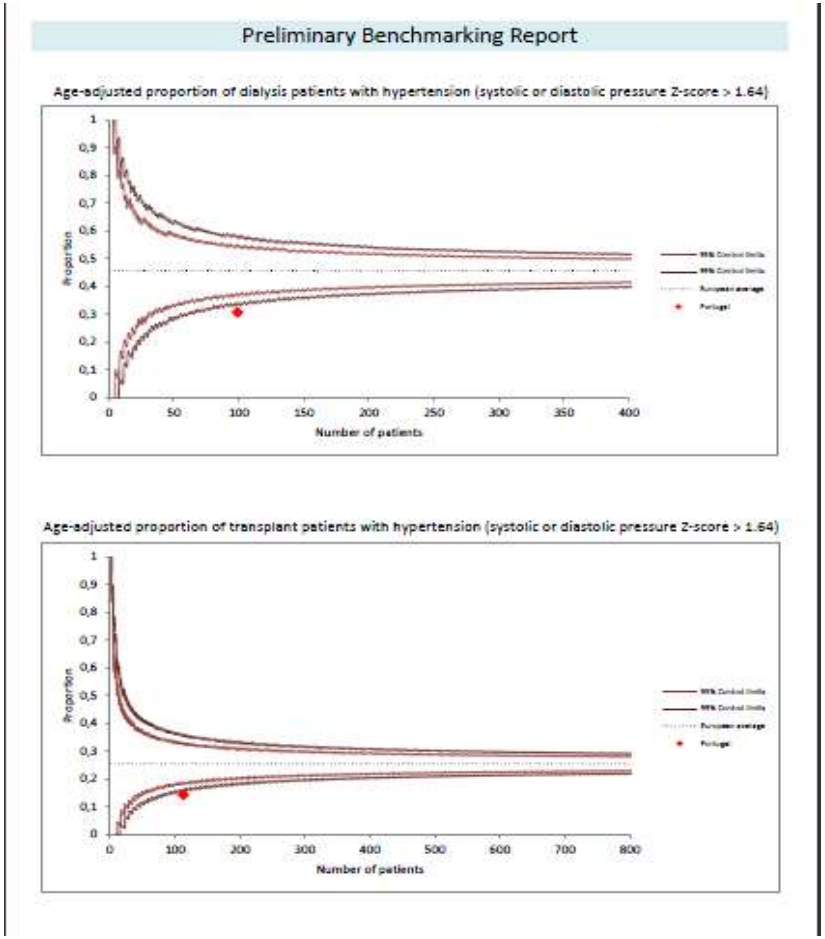
Age-adjusted proportion of anemia (Hb < 10 g/l) in transplant patients



Ponto vermelho Portugal

Benchmarking Report

Age-adjusted proportion of patients with hypertension (systolic or diastolic blood pressure Z-score>1,64)



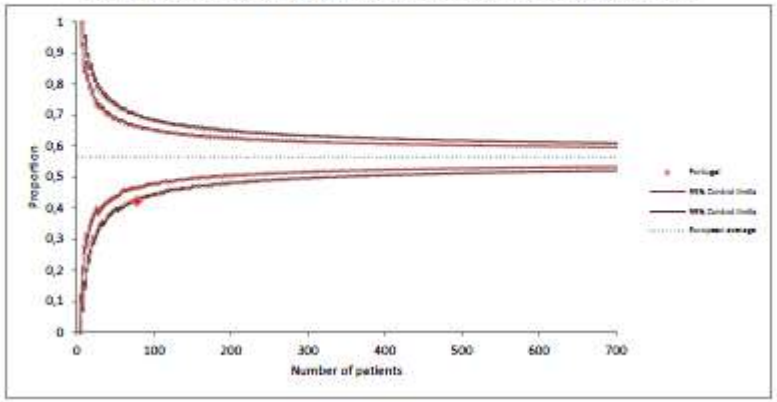
Ponto vermelho Portugal

Benchmarking Report

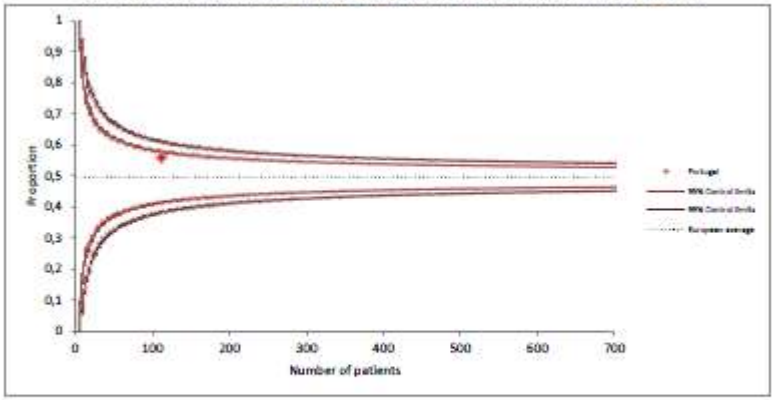
Age-adjusted patients treated with antihypertensives

Preliminary Benchmarking Report

Age-adjusted proportion of dialysis patients treated with antihypertensives



Age-adjusted proportion of transplant patients treated with antihypertensives

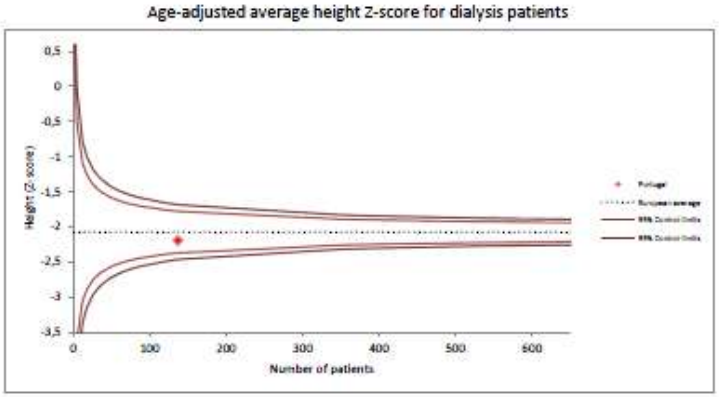


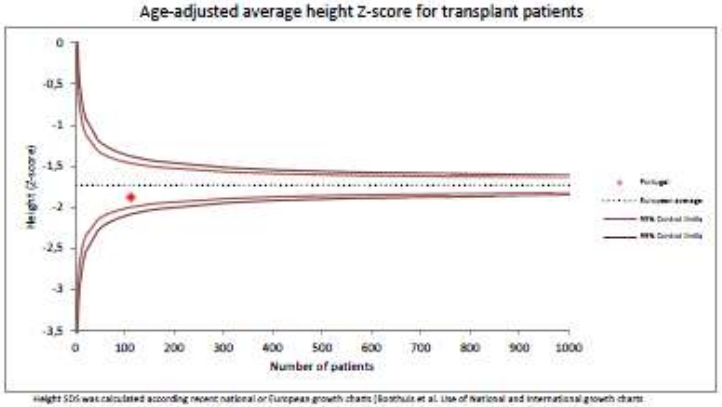
5

Ponto vermelho Portugal

Benchmarking Report

Age-adjusted average height Z-score



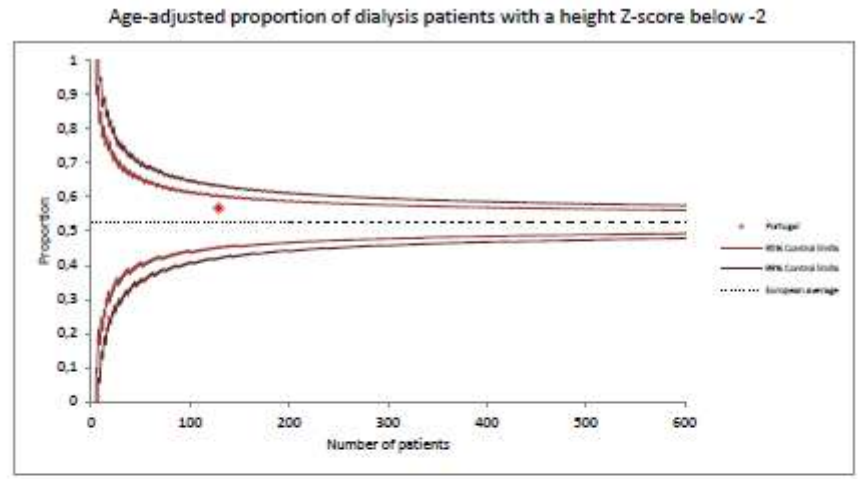


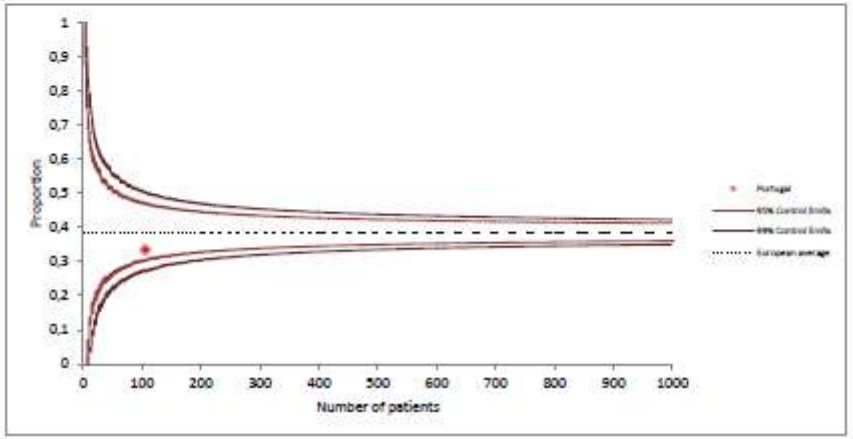
Height SDS was calculated according recent national or European growth charts [Bosthuis et al. Use of National and International growth charts for studying height in European children: development of up-to-date European height-for-age charts. PLoS ONE 2013; 7(8):e42506]

Ponto vermelho Portugal

Benchmarking Report

Age-adjusted proportion of patients wieh a height Z-score below -2



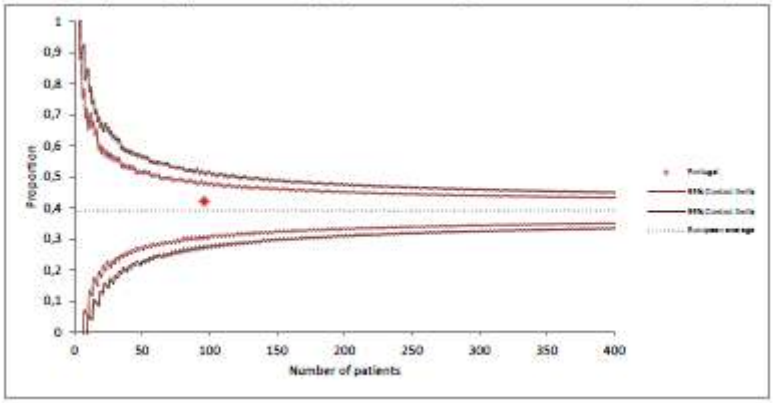


Ponto vermelho Portugal

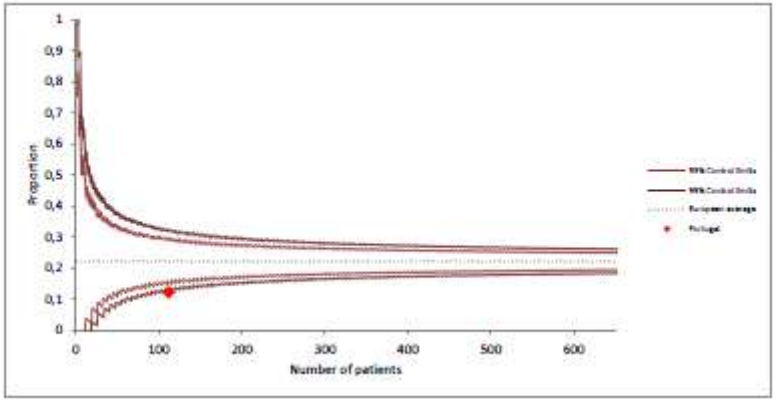
Benchmarking Report

Age-adjusted proportion of patients with hypercholesterolemia

Age-adjusted proportion of dialysis patients with hypercholesterolemia (>200 mg/dl)



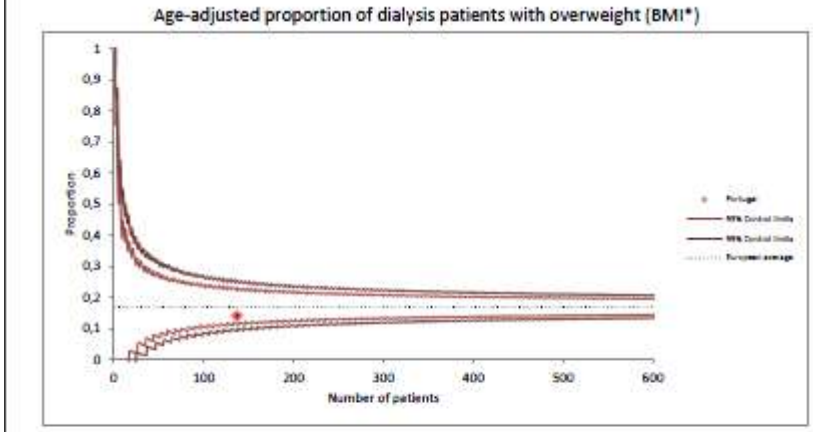
Age-adjusted proportion of transplant patients with hypercholesterolemia (>200 mg/dl)

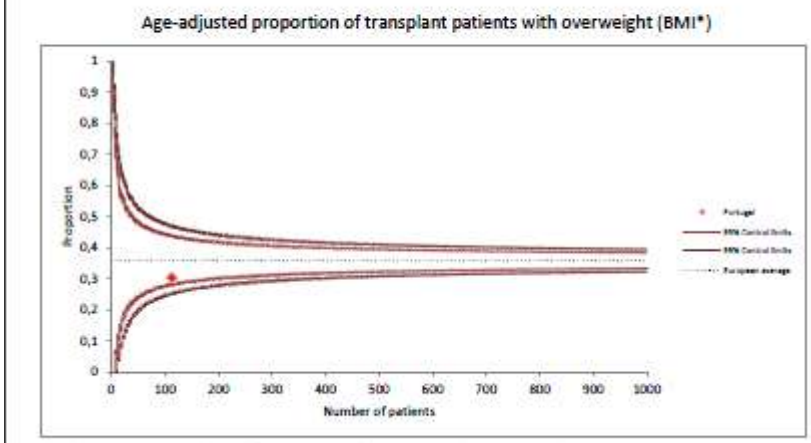


Ponto vermelho Portugal

Benchmarking Report

Age-adjusted proportion of patients with overweight





* For children <2 years, overweight was defined as Z-score > 2 based on WHO growth standards. For older patients, BMI was expressed according to height-age and categorized according to cut-offs defined by the International Obesity Taskforce

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