

European Cystic Fibrosis Registry Report 2004/2005 data

This report contains data collected from existing national registries in Europe. All known national registries were contacted and asked for their co-operation. In case of a positive response, data were collected by a common Microsoft Office Excel[®] spreadsheet with core demographic and clinical data, and definitions for some variables. The same spread sheet was sent to countries all over Europe via the project EuroCareCF WP 2 (Registry) to collect primarily demographic data. The demographic data from these countries will be presented elsewhere, whereas this report only presents data from the existing registries.

The data were extracted from the national registries into the spreadsheet by the national registry organizations according to the fields in the national registries. If the fields were not compatible some variables were omitted whereas others were redefined for the transfer after communication with the ECFR.

All data were transferred anonymously (gender, month/year of birth and country) according to the regulations of the Danish Data Protection agency, where the ECFR is registered. Data analysis was performed by the Institute of Biostatistics at the University of Milan, Italy (Istituto di Statistica Medica e Biometria G.A. Maccacaro).

These data were collected in 2007, and were first presented at the 30th European Cystic Fibrosis Conference in Belek, Turkey. The analyses have, however, been re-done after the registry consensus meeting in Århus, Denmark October 2007. This re-analysis was done for two major reasons:

1. The discussion about common reference values for anthropometric measures and FEV-1 as well as the convenience of using z-scores instead of percentiles. In the data collected, for BMI international references were used, whereas for FEV-1 national references were used.
2. Some of the clinical data, e.g. complications were not valid because of very different definitions in the national registries. For that reason we have omitted these data entirely and will be working on common ECFR definitions, so these variables can be included in future reports.

We would like to thank the participating countries for their patience and cooperation. Special thanks to the people involved in the extraction and evaluation of national data, as well as the final report:

Linda Foley, Ireland
Elaine Gunn, UK
Herwig Jansen, Belgium
Meir Mei-zahav, Israel
Sophie Ravilly, France
Paul Wenzlaff, Germany

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Hanne Vebert Olesen
European CF Registry Steering Committee, Denmark

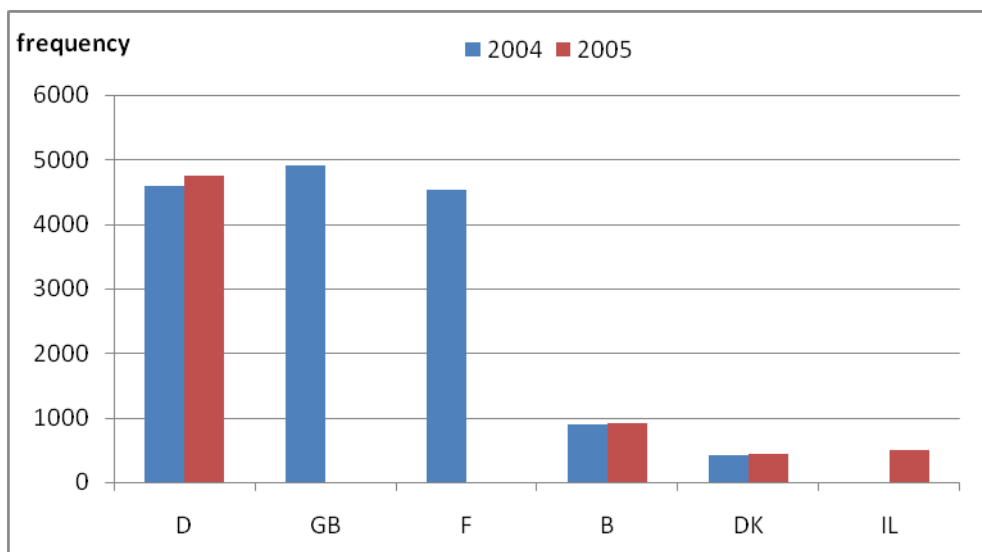
Laura Viviani
Istituto di Statistica Medica e Biometria, Italy

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Table 1 Number of patients reported by each country, by year

country	2004	2005
B	888	912
D	4581	4758
DK	425	441
F	4533	---
GB	4912	---
IL	---	498
<i>Total</i>	<i>15339</i>	<i>6609</i>

Figure 1 Number of patients reported by each country, by year

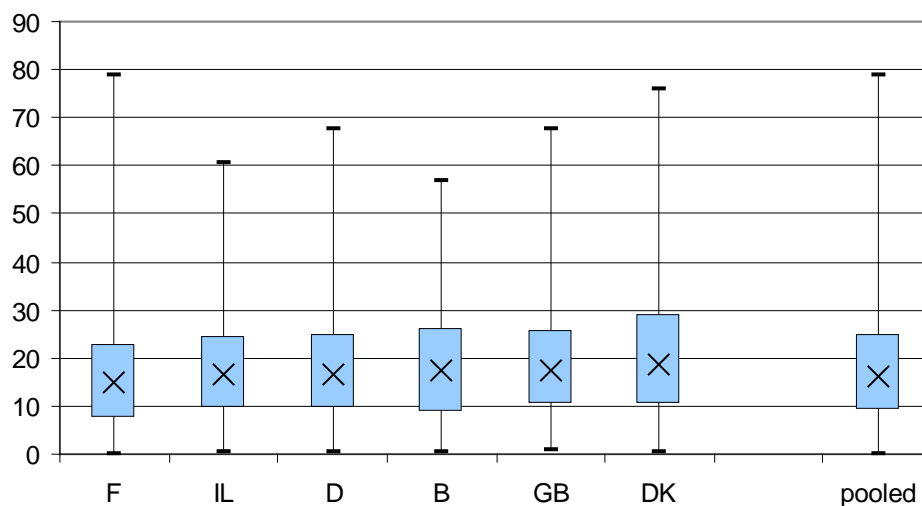


Not all countries could supply data from both years (2004 and 2005) for various reasons. Since we would not expect huge changes in these core values over a 1 year period, for the countries with two years of data we have chosen the year with most valid data for the comparison with the other countries.

Table 2 Age of patients at the end of each year (in years)

Country	Mean	Min	25 th Pctl	50 th Pctl	75 th Pctl	Max	% adults (≥18 years)
B	18.56	0.46	8.87	17.38	26.05	56.87	47.80
D	18.17	0.29	9.39	16.75	24.90	67.47	45.24
DK	20.24	0.21	10.46	18.55	28.88	75.87	52.02
F	16.38	0.04	7.46	14.87	22.80	78.87	38.12
GB	19.13	0.96	10.21	17.46	25.71	67.71	48.37
IL	18.31	0.21	9.74	16.50	24.62	60.42	46.34
<i>total</i>	<i>18.04</i>	<i>0.04</i>	<i>9.21</i>	<i>16.38</i>	<i>24.80</i>	<i>78.87</i>	<i>44.55</i>

Figure 2 Boxplot of age of patients at the end of each year (in years)



Boxplots: cross represents median, box represents 25 to 75% percentiles, whiskers represent minimum and maximum.

Figure 3 Gender distribution (pink=females, light blue=males)

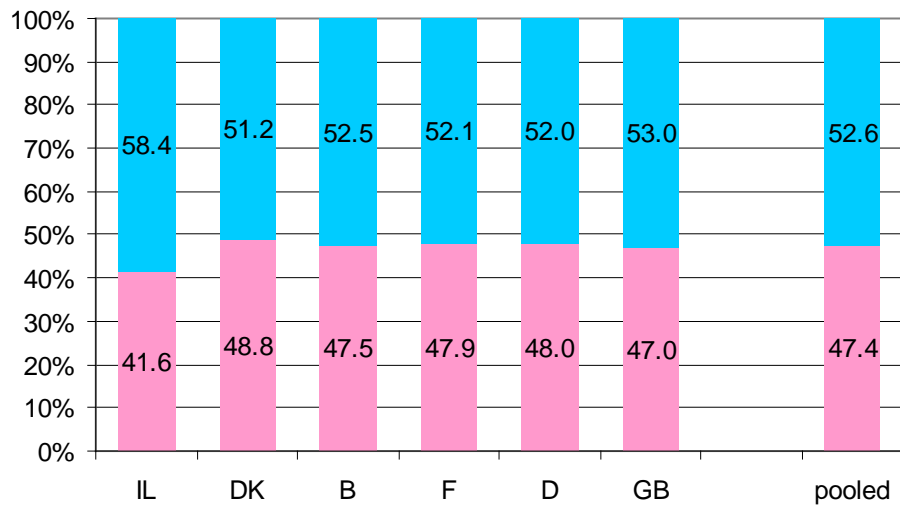
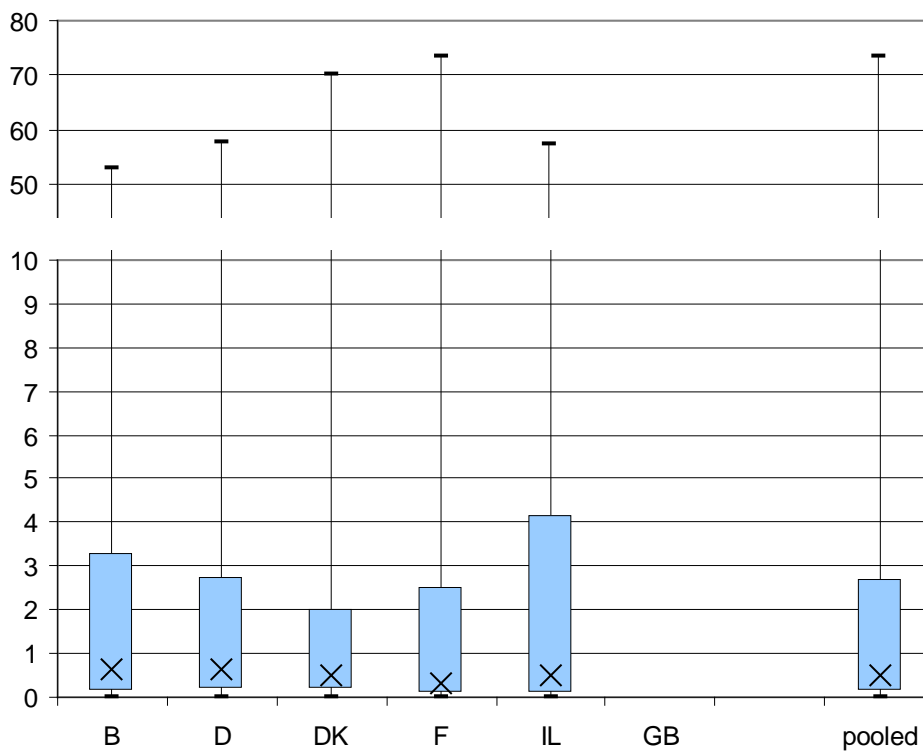


Table 3 Age at diagnosis (in years)*

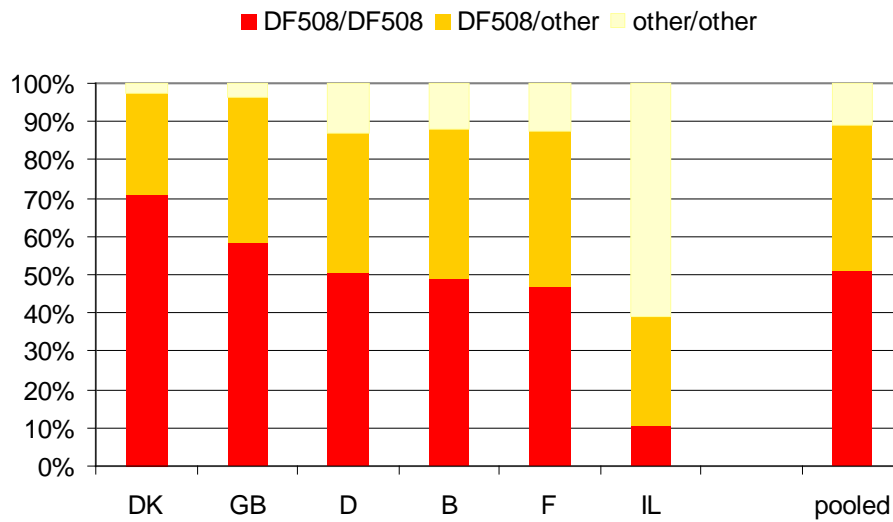
country	Mean	Min	25 th Pctl	50 th Pctl	75 th Pctl	Max	% adults (≥18 years)	% infants (<1 year)
B	3.87	at birth	0.15	0.65	3.30	52.69	6.56	58.90
D	2.92	at birth	0.18	0.63	2.75	57.55	3.33	57.46
DK	2.16	at birth	0.17	0.50	2.00	70.00	1.18	63.76
F	3.33	at birth	0.08	0.33	2.50	73.25	5.20	66.87
IL	4.72	at birth	0.10	0.50	4.17	57.17	8.58	62.01
GB	-	-	-	-	-	-	-	-
<i>total</i>	<i>3.20</i>	<i>at birth</i>	<i>0.12</i>	<i>0.50</i>	<i>2.70</i>	<i>73.25</i>	<i>4.46</i>	<i>61.70</i>

Figure 4 Age at diagnosis (in years)



*No data available on age at diagnosis from the UK registry

Figure 5 Genotype: frequencies of F508del mutation



Percentages were computed considering only patients undergone genetic analysis. In the category "other", unknown mutations are included.

The graph shows the different distribution of F508del mutation among countries, which reflects genetic heterogeneity.

Figure 6 Allelic frequencies of unknown mutations

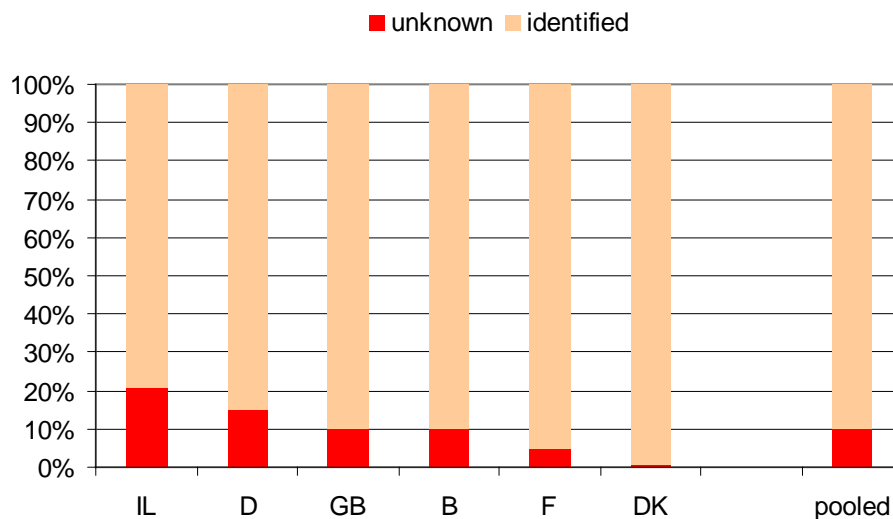
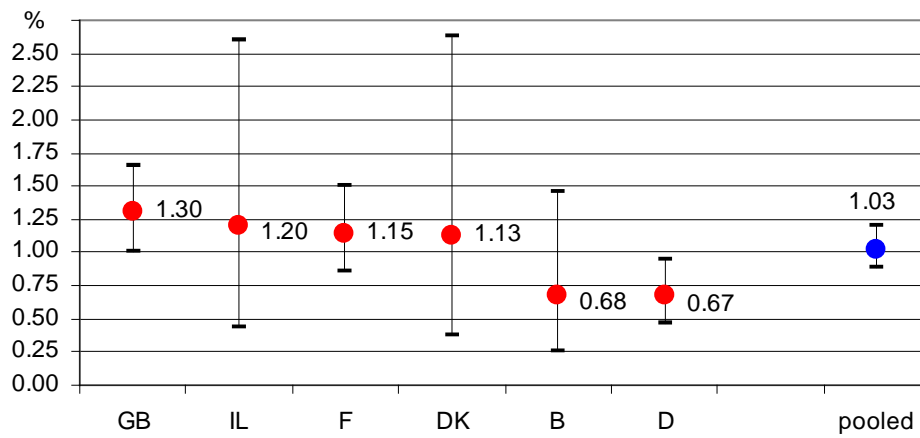
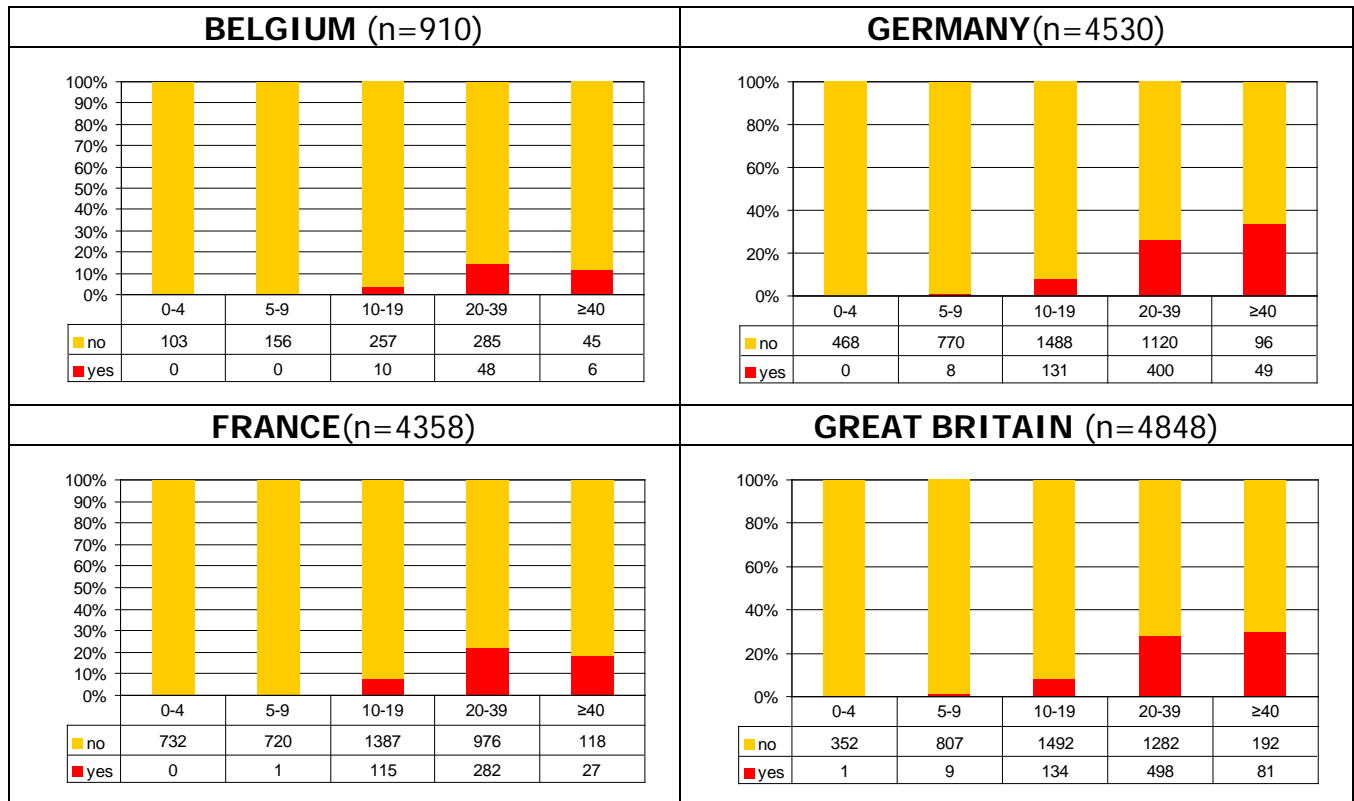


Figure 7 Proportion of deceased patients in one year (95% C.I.)



Some countries reported loss to follow up as well as delays in deaths registration, which might affect mortality estimates.

Figure 8 Prevalence of CF-related diabetes during year of observation



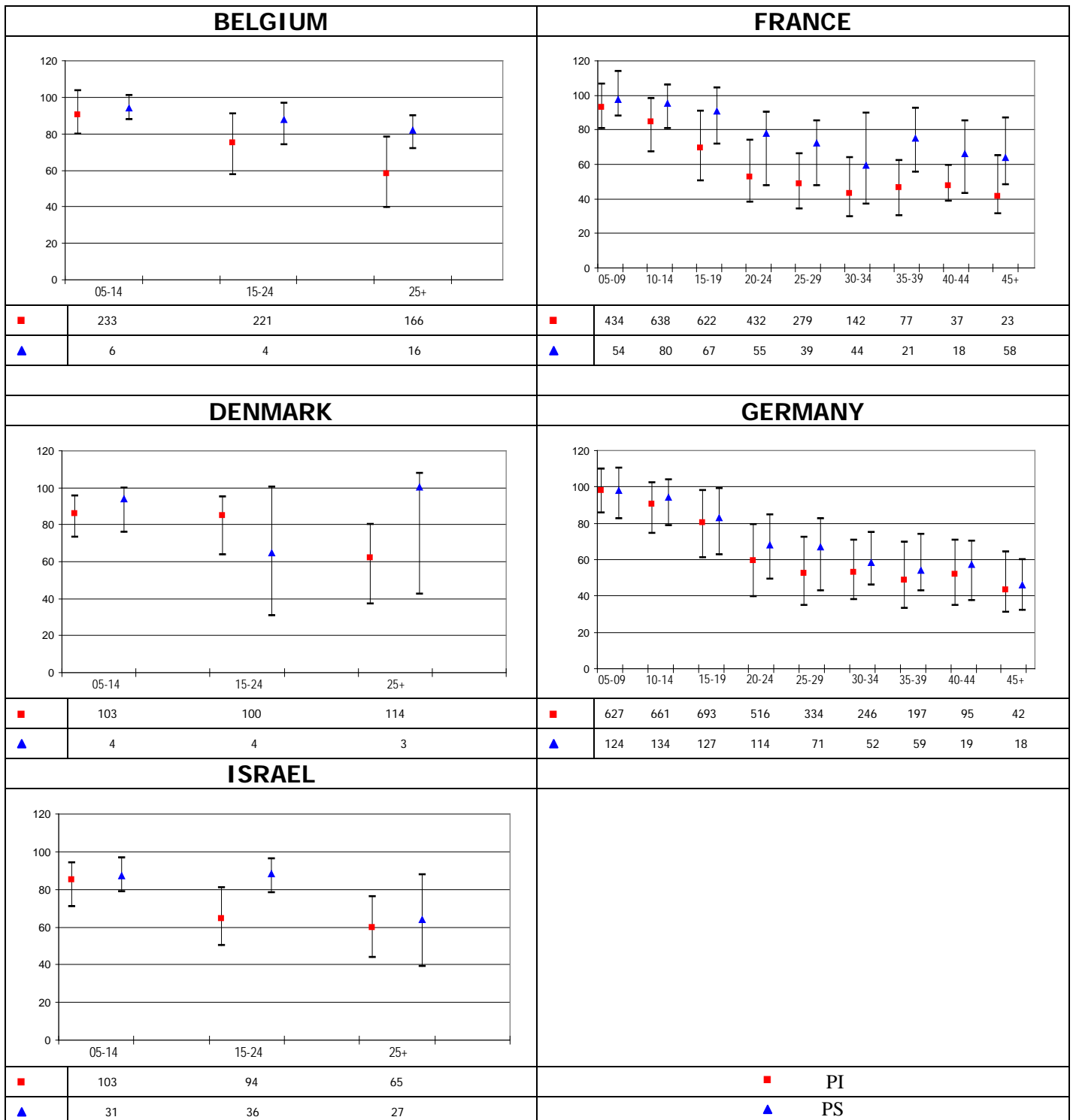
Only four countries shared a similar definition of CFRD. This figure is therefore also to illustrate that common definitions are very important and this issue will be addressed in future data collection.

Table 4 Frequencies of lung transplantation

Country	Transplantation performed			during this year before this year year unknown
	don't know	never	yes	
B	21 (2.30)	823 (90.24)	68 (7.46)	{ 9 (0.99) 59 (6.47) 0 (0.00)
D	0 (0.00)	4702 (98.82)	56 (1.18)	{ 10 (0.21) 30 (0.63) 16 (0.34)
DK	0 (0.00)	410 (92.97)	31 (7.03)	{ 6 (1.36) 25 (5.67) 0 (0.00)
F	0 (0.00)	4404 (97.15)	129 (2.85)	{ 37 (0.82) 92 (2.03) 0 (0.00)
GB	0 (0.00)	4882 (99.39)	30 (0.61)	{ 0 (0.00) 30 (0.61) 0 (0.00)
IL	1 (0.20)	493 (99.00)	4 (0.80)	{ 0 (0.00) 4 (0.80) 0 (0.00)
<i>total</i>	<i>22</i> <i>(0.14)</i>	<i>15714</i> <i>(97.88)</i>	<i>318</i> <i>(1.98)</i>	{ <i>62 (0.39)</i> <i>240 (1.49)</i> <i>16 (0.10)</i>

The proportion of transplanted patients is variable across countries. This may reflect different attitudes towards transplantation as well as different reporting.

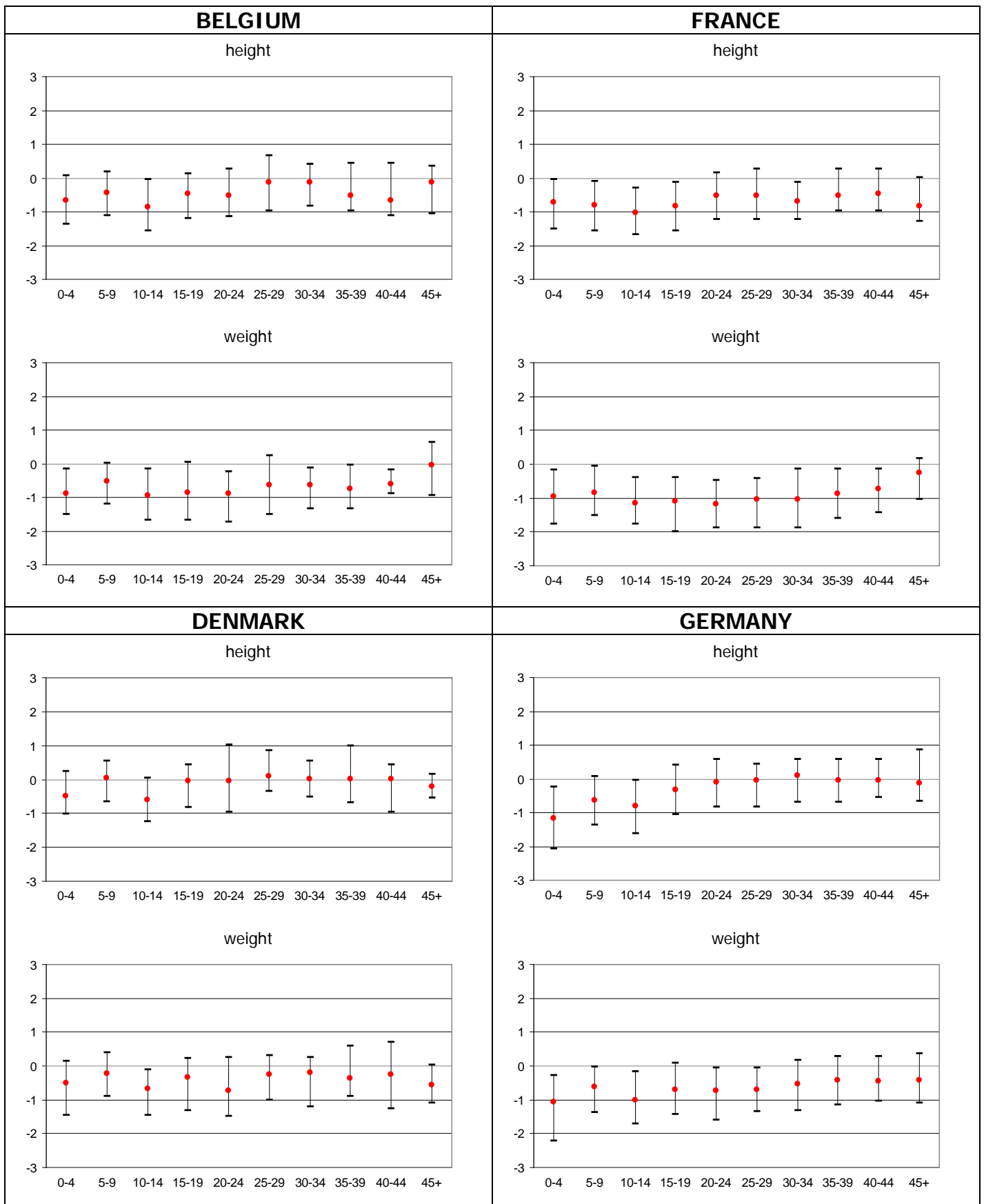
Figure 9 FEV1(%pred) inter-quartile range, by age and pancreatic status*

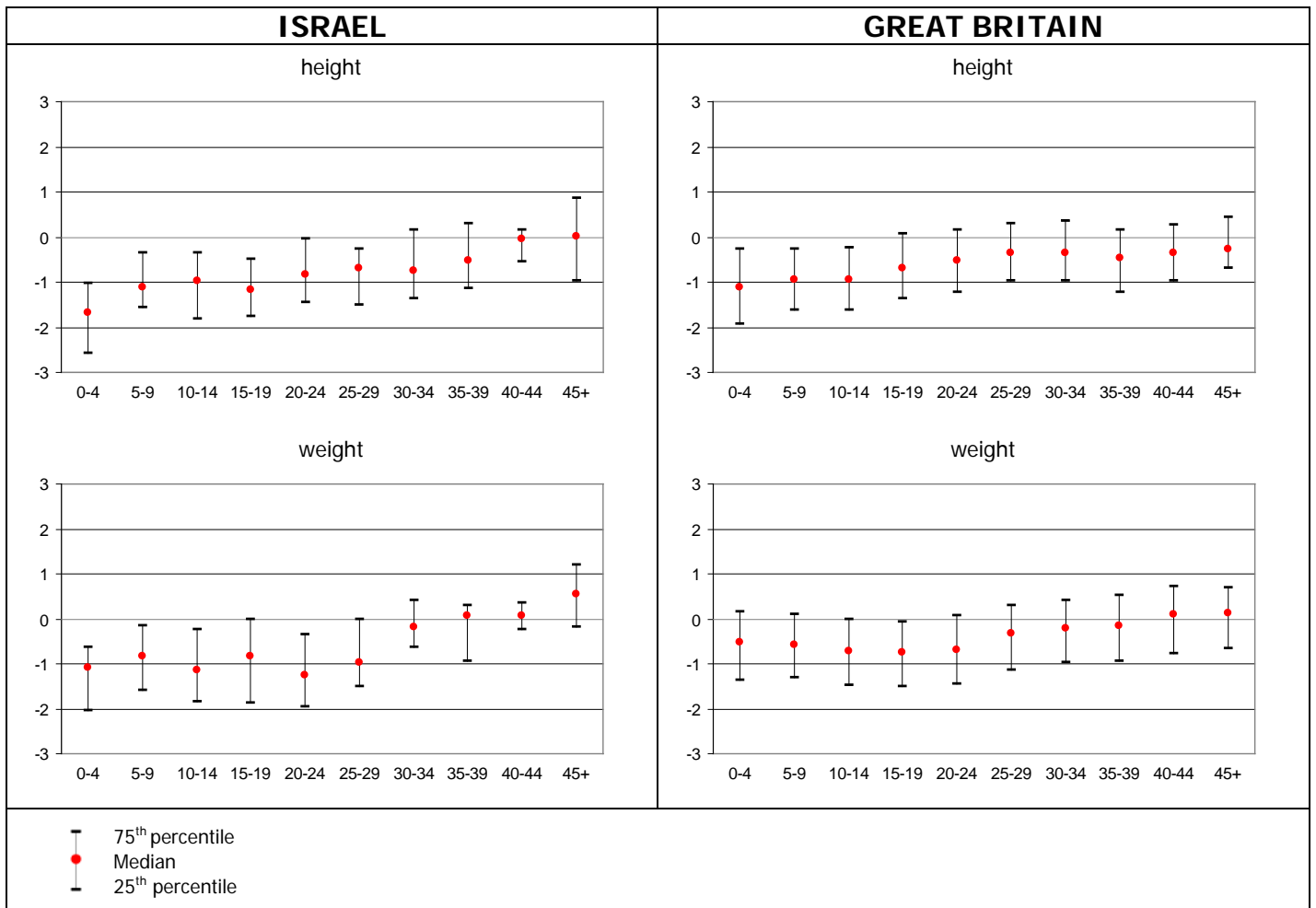


*transplanted patients were excluded from the analysis

FEV-1 % of predicted was calculated from the reference material used in the national registries because of lack of data for a common calculation. Due to low number of patients from Belgium, Denmark and Israel compared to France and Germany age was grouped differently. This makes comparison between countries more difficult, but makes comparison within countries (PI/PS) more informative.

Figure 10 Standard Deviation Scores for height and weight





International references were used¹. The use of common reference material allows comparison between CF populations across countries, but invalidates comparison between CF and NON-CF populations within countries. For future analyses, national references (where available) will be used.

¹ Kuczmarski RJ, Ogden CL, Guo SS et al. 2000 CDC growth charts for the United States: Methods and development. National Center for Health Statistics (2002). Vital Health Stat 11 (246): 1-190.

Figure 11 Standard Deviation Scores: height and weight, by gender

