

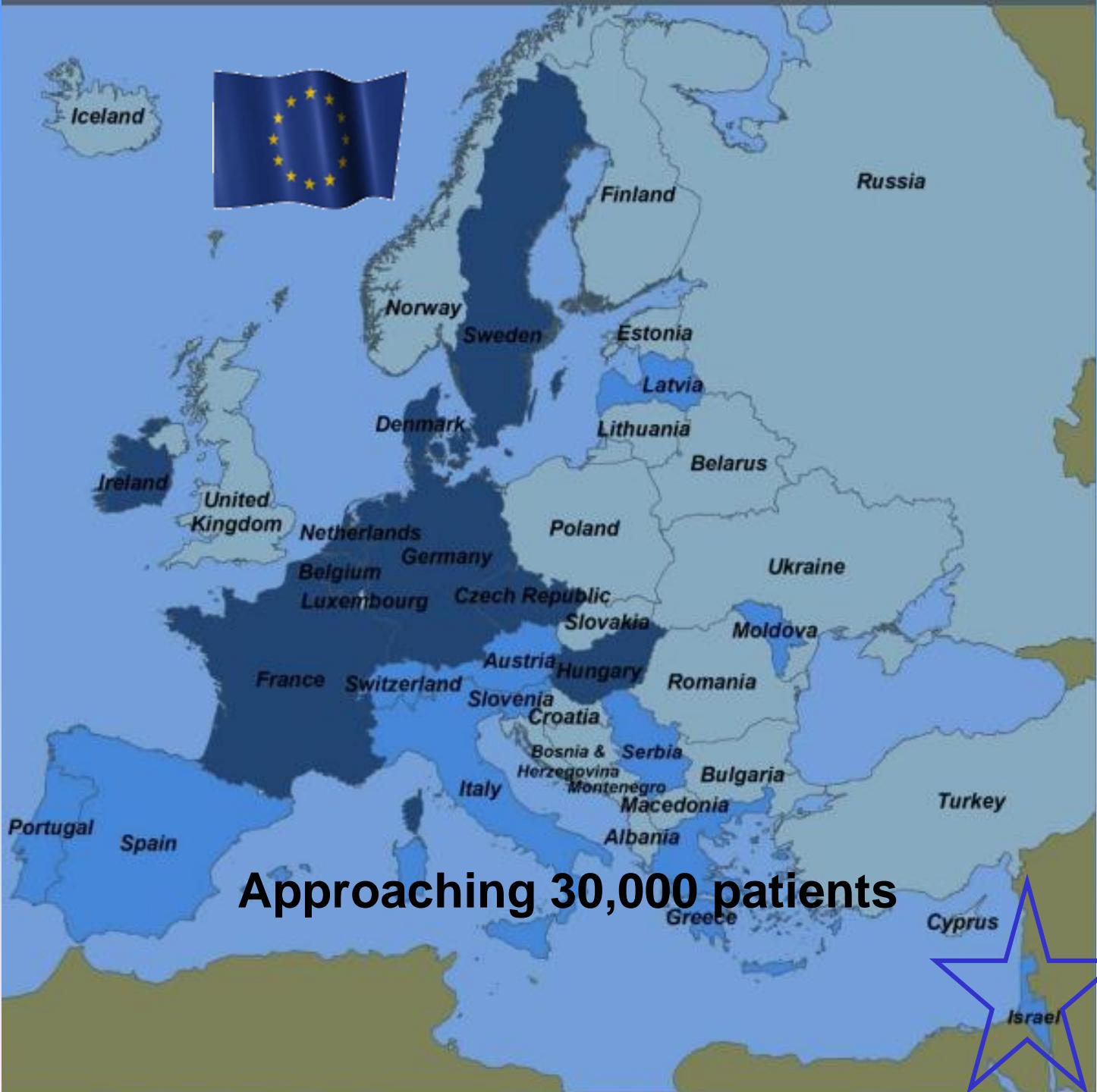
The Israeli CF Database Update



Dr. Meir Mei-Zahav
Graub Cystic Fibrosis Center
Schneider Children's Medical Center of Israel

Issues to discuss....

- What is the ECFR?
- What happened in Europe since the registry was started?
- Some data   - where we are?
- Some data on Israel 2004-2010



Approaching 30,000 patients



DATA IN THE ECFS PATIENT REGISTRY

	2003	2004	2005	2006	2007	2008	2009
Austria				107	116	347	352
Belgium	860	888	912	1036	1052	1086	1129
Bulgaria					95		
Czech Republic				457	502	490	507
Denmark	412	425	441	438	447	452	451
France	4104	4533		4994	5147	5366	5640
Germany		4581	4758	4894	5039	5533	5048
Greece					121	94	92
Hungary					372	547	555
Ireland	325			832		1021	
Israel			498	447	507	516	533
Italy	4142			771	774	529	539
Latvia						30	29
Moldova							41
Netherlands					1113	1168	1249
Portugal					116	61	117
Serbia						118	122
Slovenia					50	57	66
Spain						582	740
Sweden	362			120		367	578
Switzerland						173	190
United Kingdom		4912			4408	6082	7375

Summary of data report

Outcome		Females	Males	Total
Patients registered this year	n (%)	9,084 (47.8)	9,915 (52.2)	18,999
Age (years)	mean	18.3	18.8	18.6
	median	17.0	17.0	17.0
Patients over 18 years	%	46.8	49.1	48.0
Age at diagnosis	mean (years)	3.8	3.4	3.6
	median (months)	6.0	4.8	6.0

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Patients with at least one F508del allele	%	82.8	83.2	83.0
Patients deceased during this year	n (%)	94 (1.0)	76 (0.8)	170 (0.9)

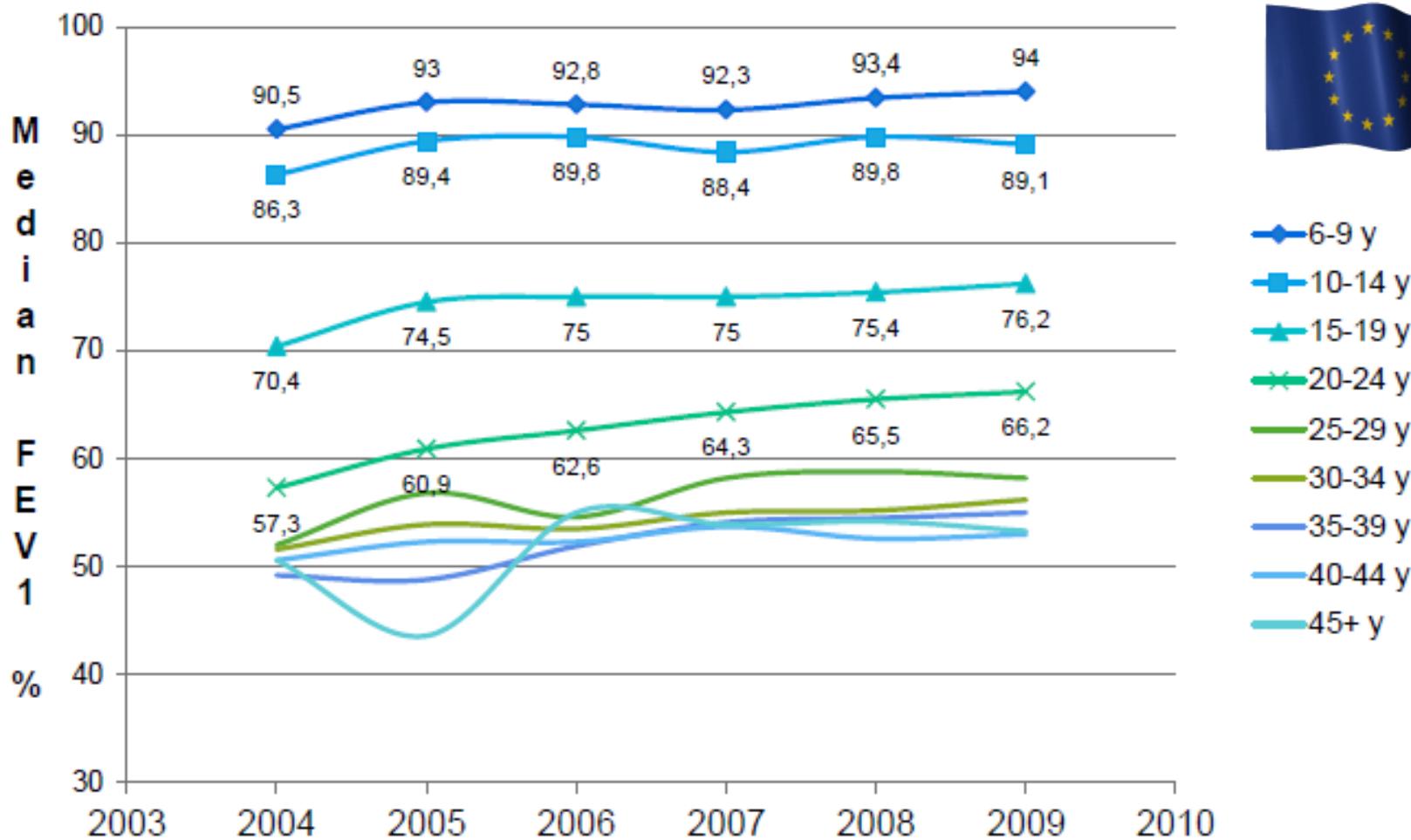
Summary of data report

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Age at death (years)	mean	24.2	29.9	26.7
	median	23.0	29.0	25.0
Patient living with lung transplant	n (%)	409 (4.6)	395 (4.0)	804 (4.3)
Patient living with liver transplant	n (%)	41 (0.5)	52 (0.5)	93 (0.5)

Issues to discuss....

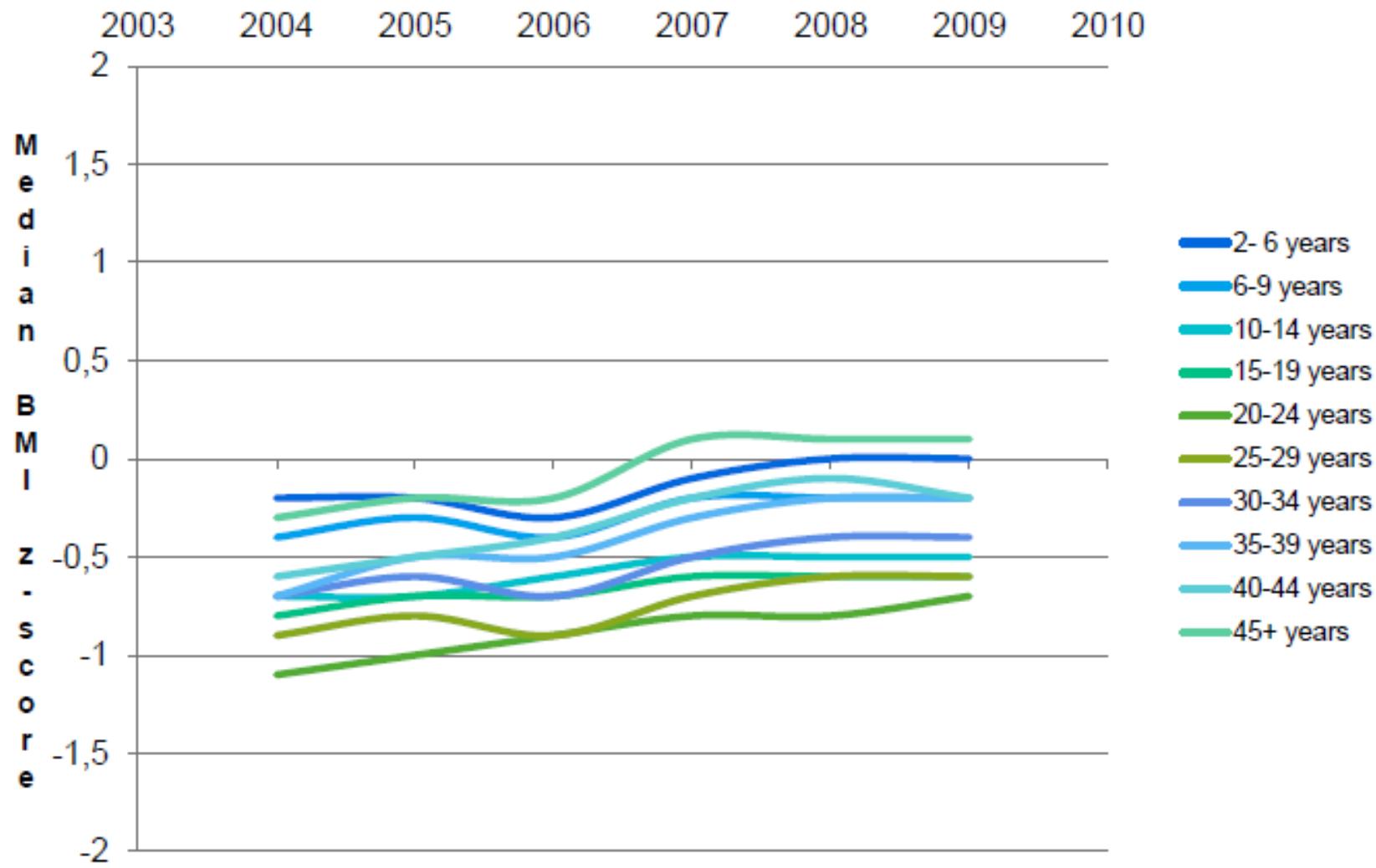
- What is the ECFR?
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- Some data – Europe-Israel- where we are?
- Some data on Israel 2003-2010

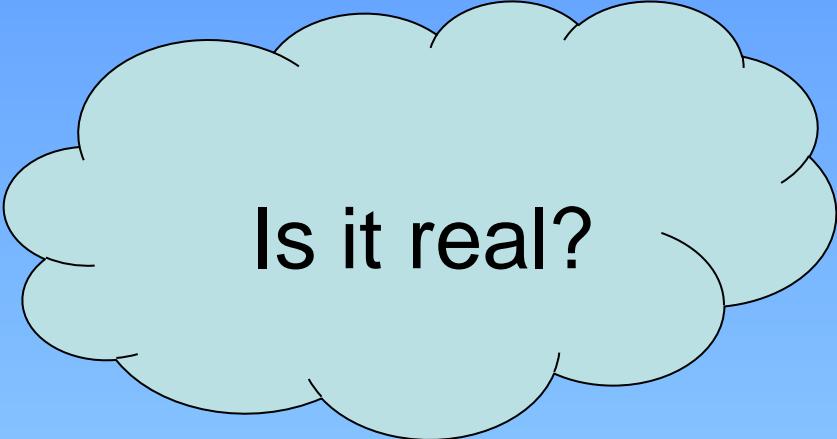
FEV1 MEDIAN BY AGE GROUP FOR TOTAL*



*Only non-transplanted patients

BMI Z-SCORE BY AGE GROUP FOR TOTAL





Is it real?

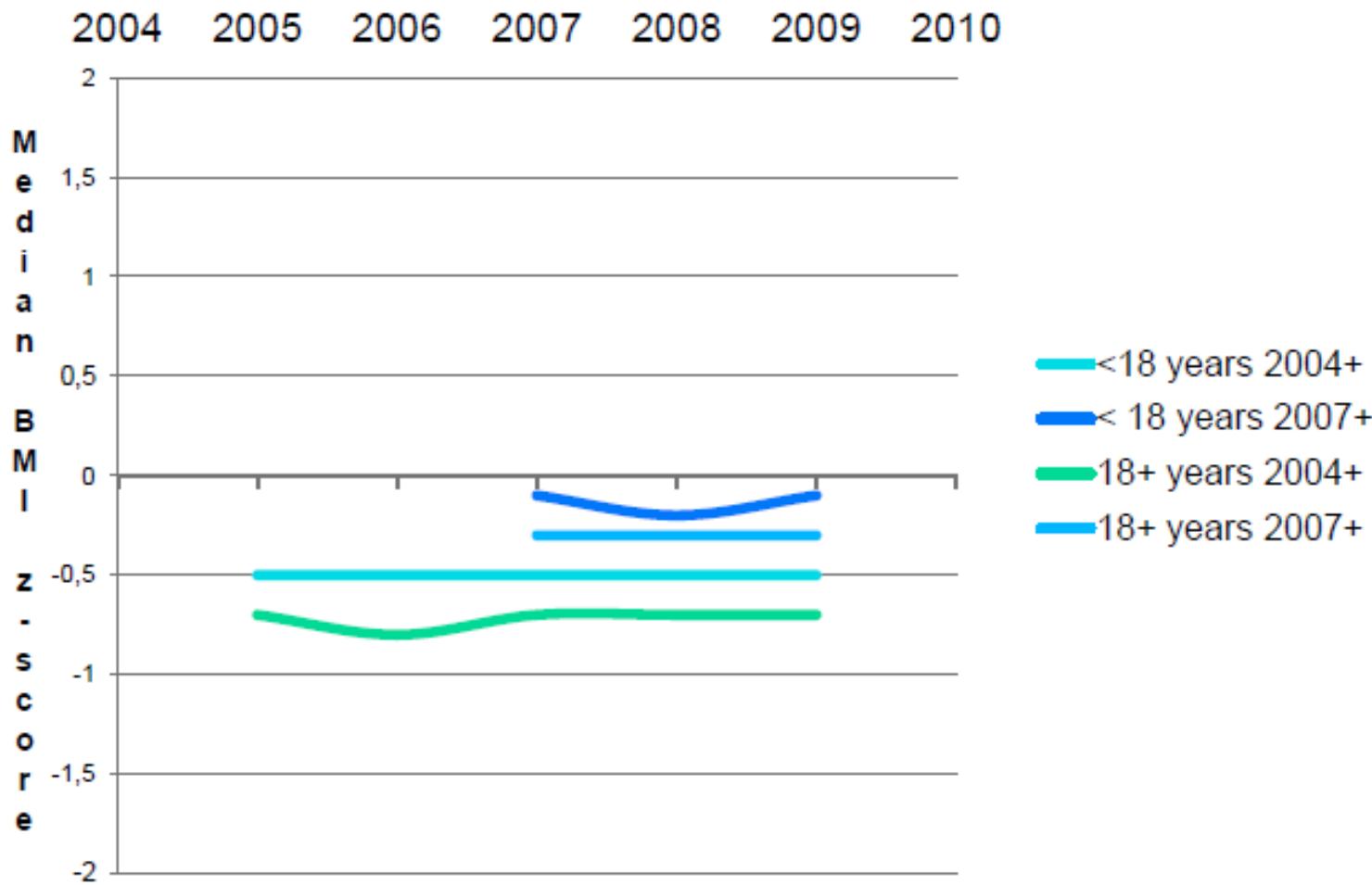


FEV-1 BY CHILDREN AND ADULTS BY CONTRIBUTOR-GROUP*



*Only non-transplanted patients

BMI Z-SCORE CHILDREN AND ADULTS BY CONTRIBUTOR GROUP



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-

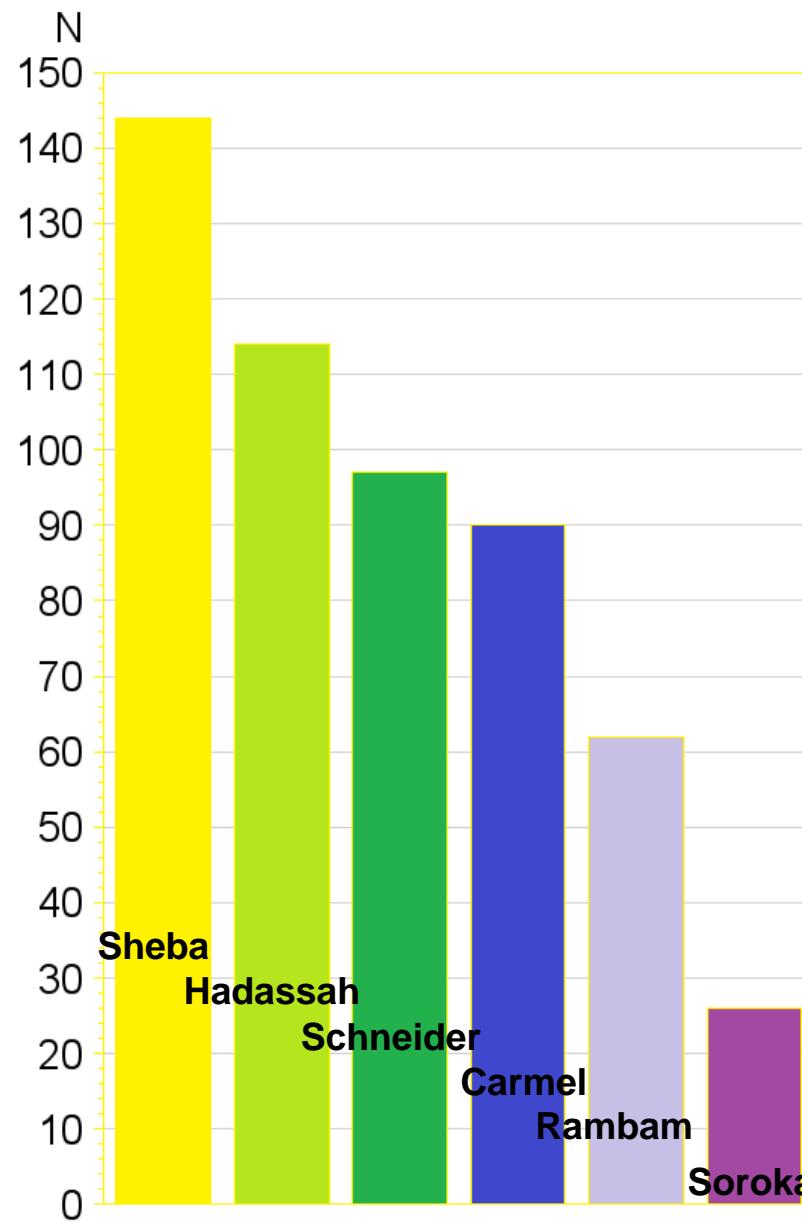
Israel CF Registry Data

- Years
 - 2009 – all centers- 533 patients
 - 2010- without Sheba MC- 436 patients

- Demographics
- Clinical Characteristics
- Treatment

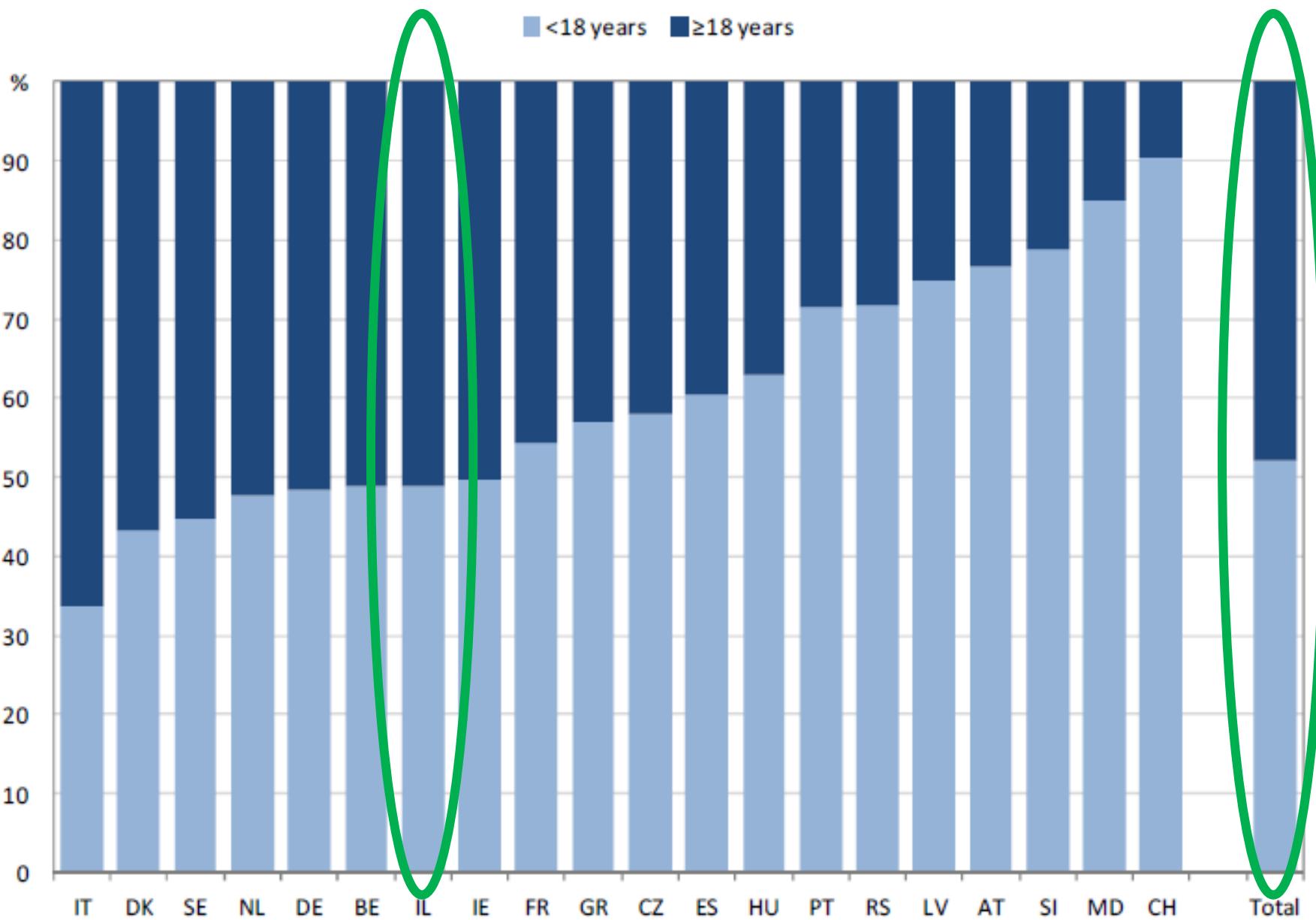
- Demographics
- Clinical Characteristics
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Number of patients -533
Year 2009





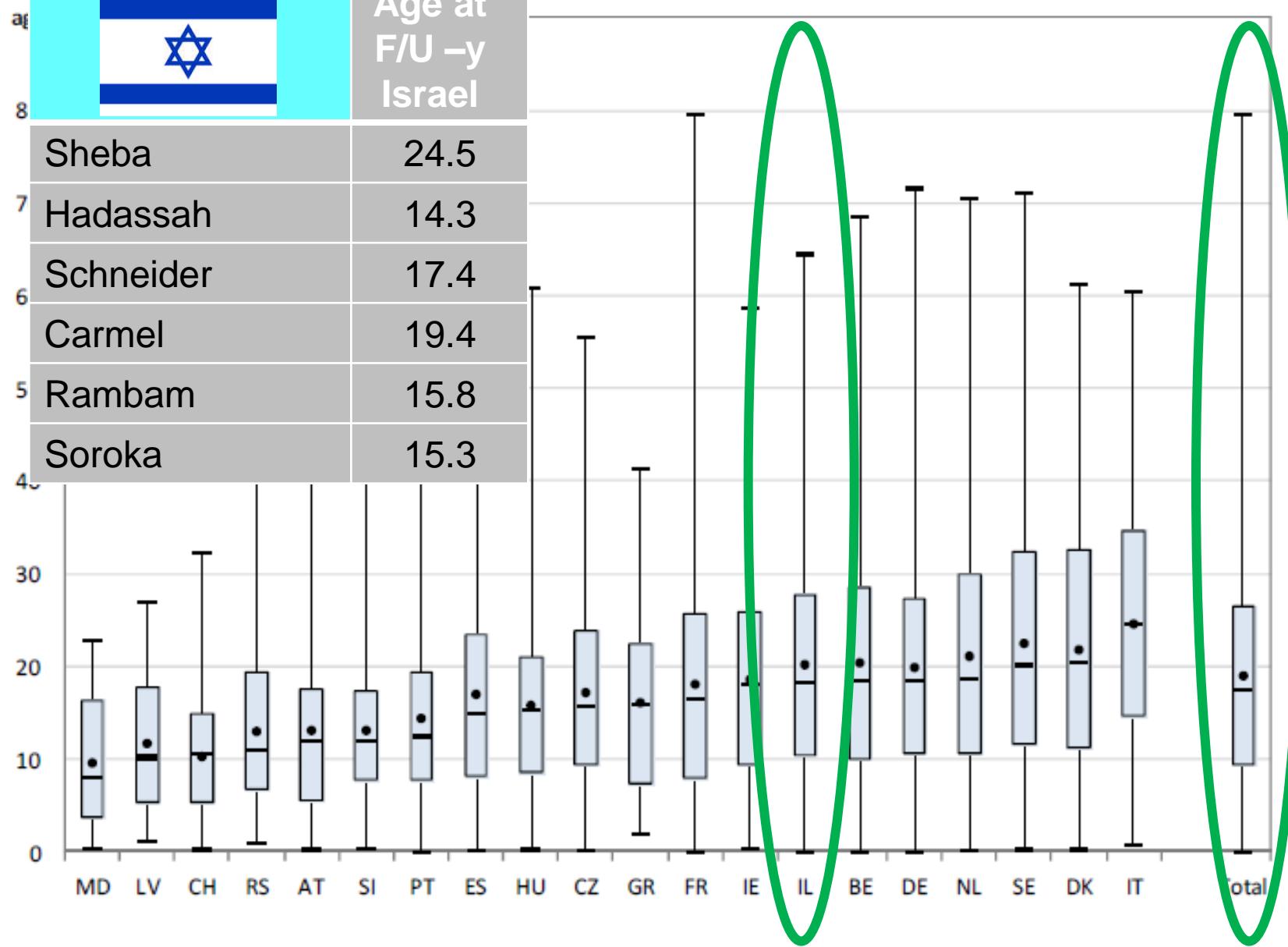
CF Kid and adults



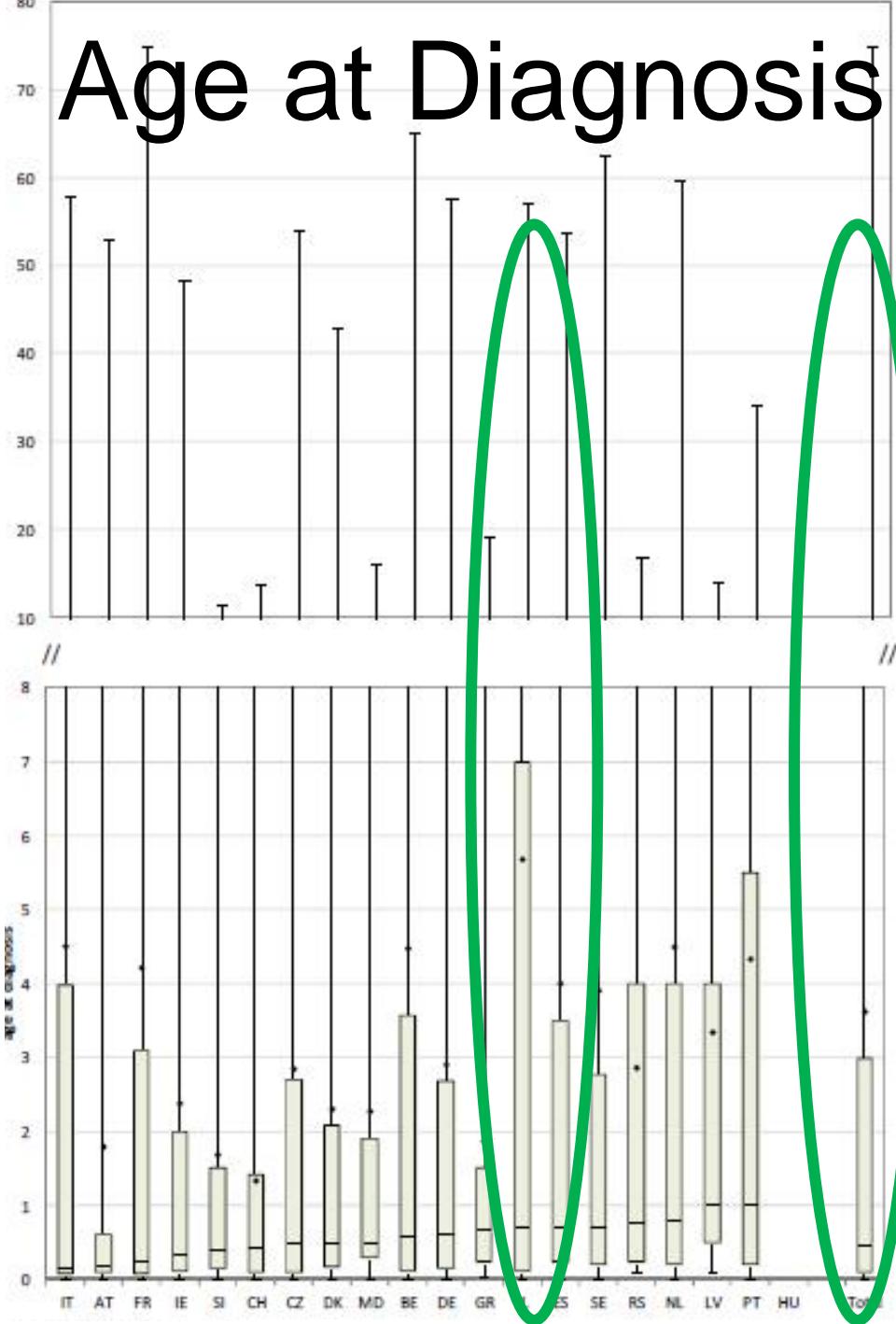


Age at F/U

Ap	Age at F/U -y Israel
Sheba	24.5
Hadassah	14.3
Schneider	17.4
Carmel	19.4
Rambam	15.8
Soroka	15.3



Age at Diagnosis



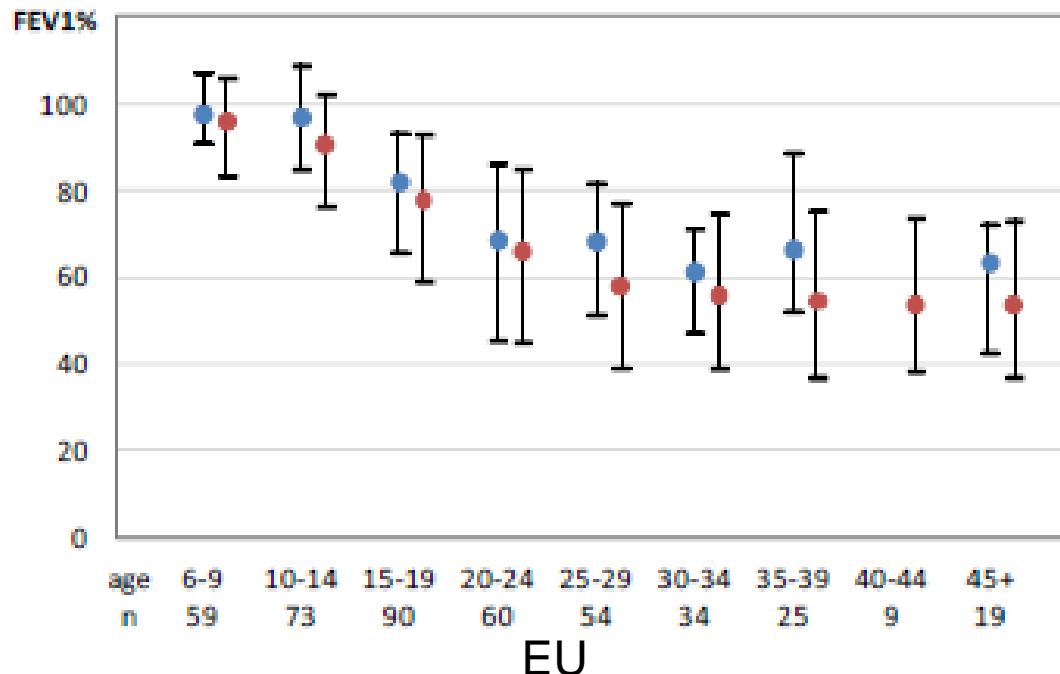
Neonatal screen??

Israel 2010-
9 months

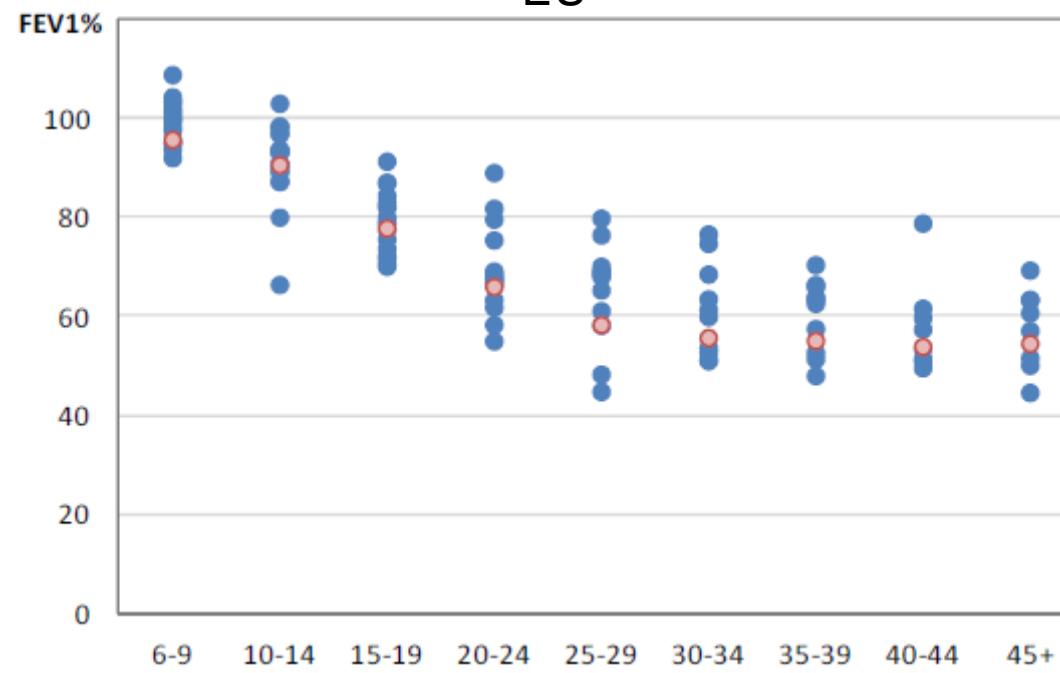
- Demographics
- Clinical Characteristics
- Treatment

Pulmonary

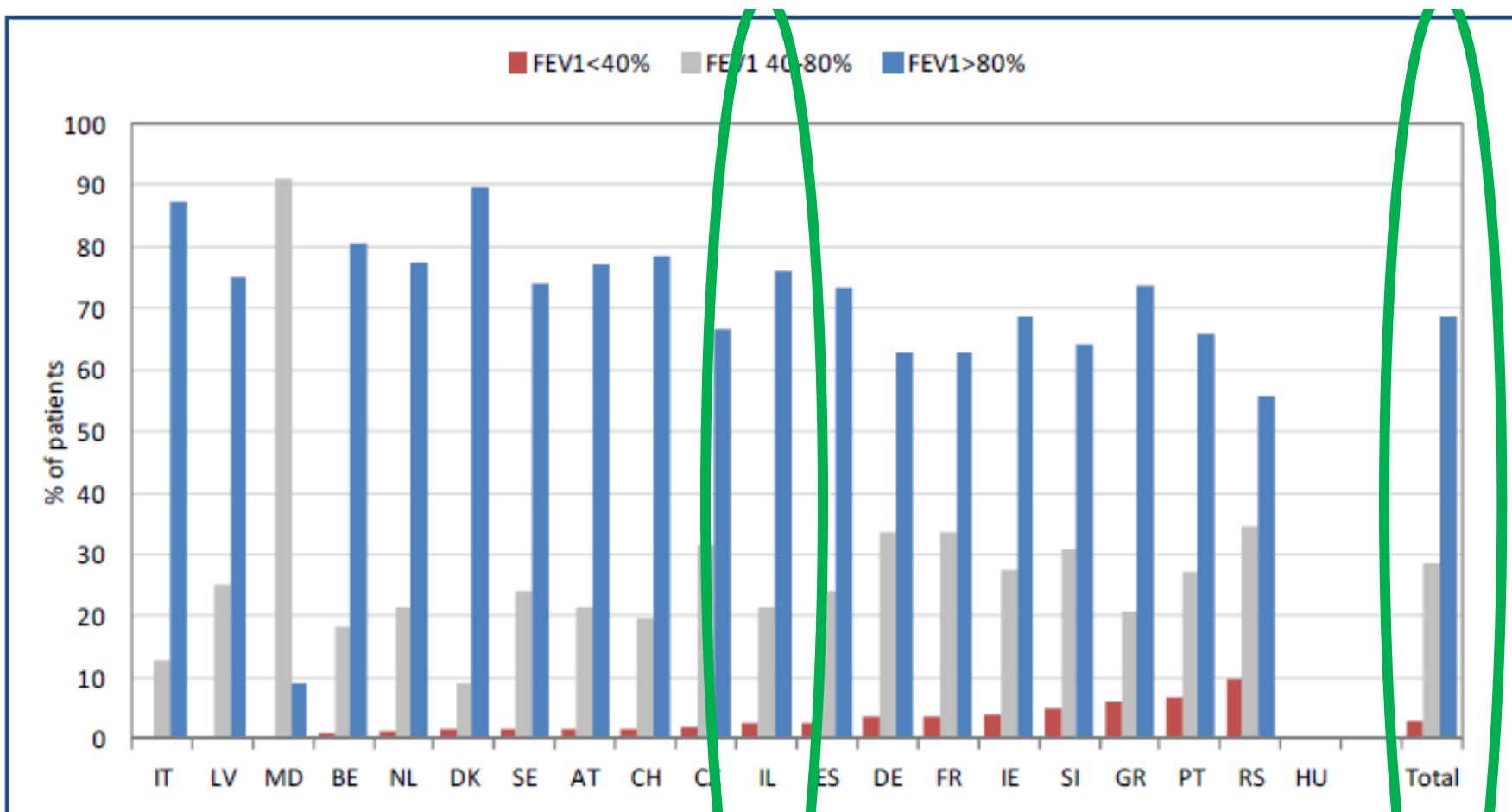
Quartiles of FEV1%: Israel



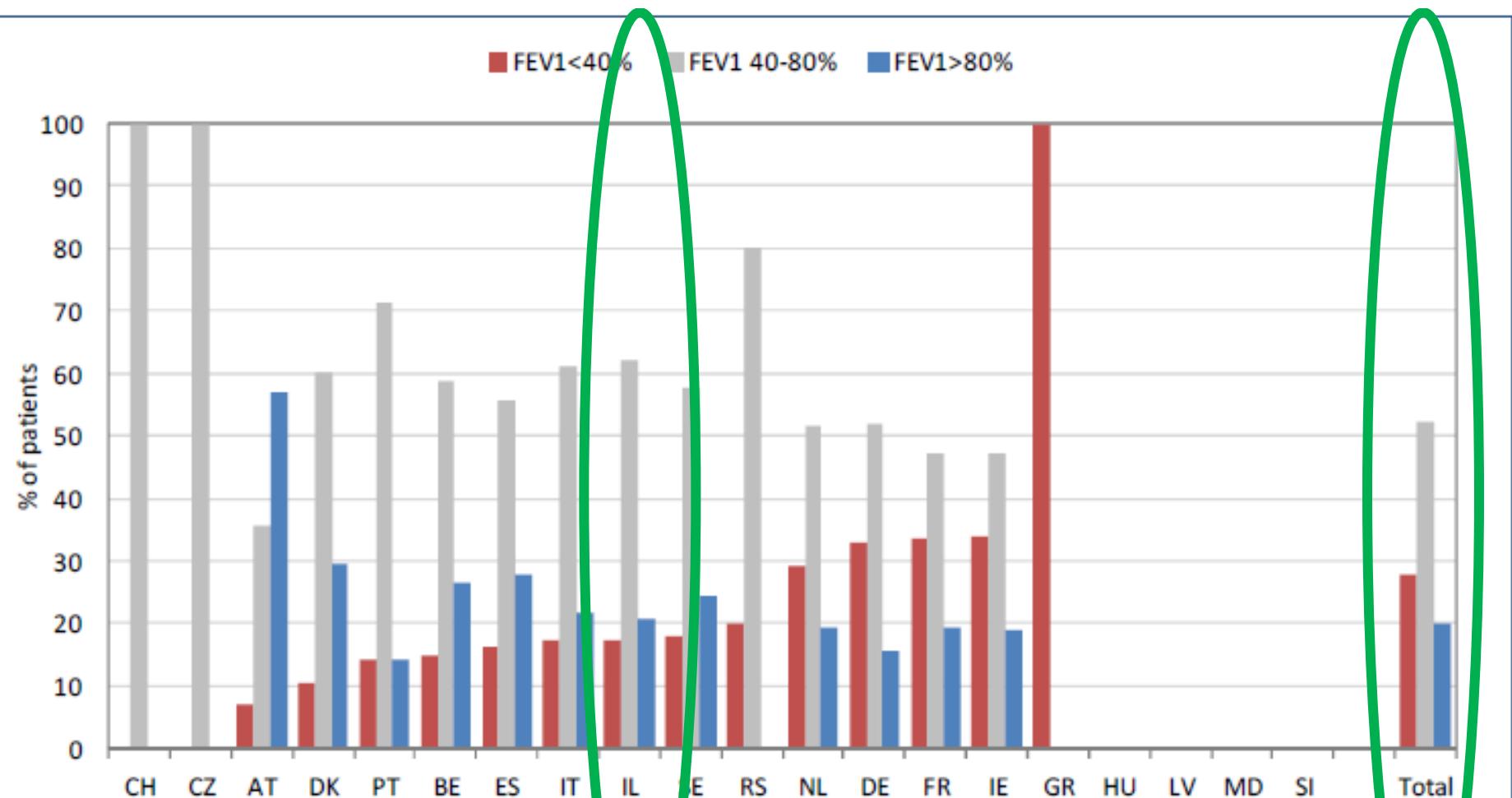
EU



FEV1-Divided by Severity - Children



FEV1-Divided by Severity – Adults>30y



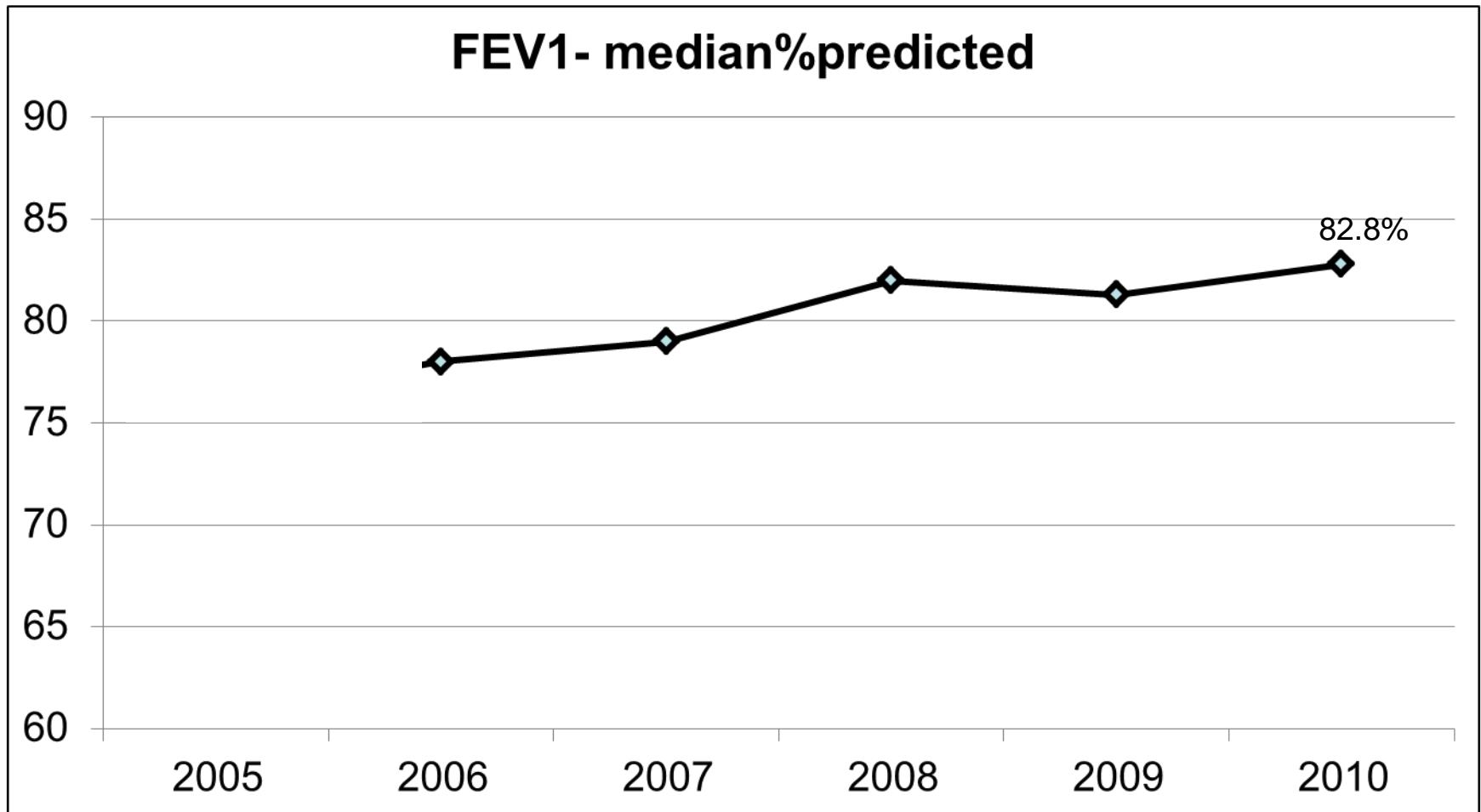
Note: not all the countries reported the best FEV1 value of the year:

France reported the last FEV1 of the year,

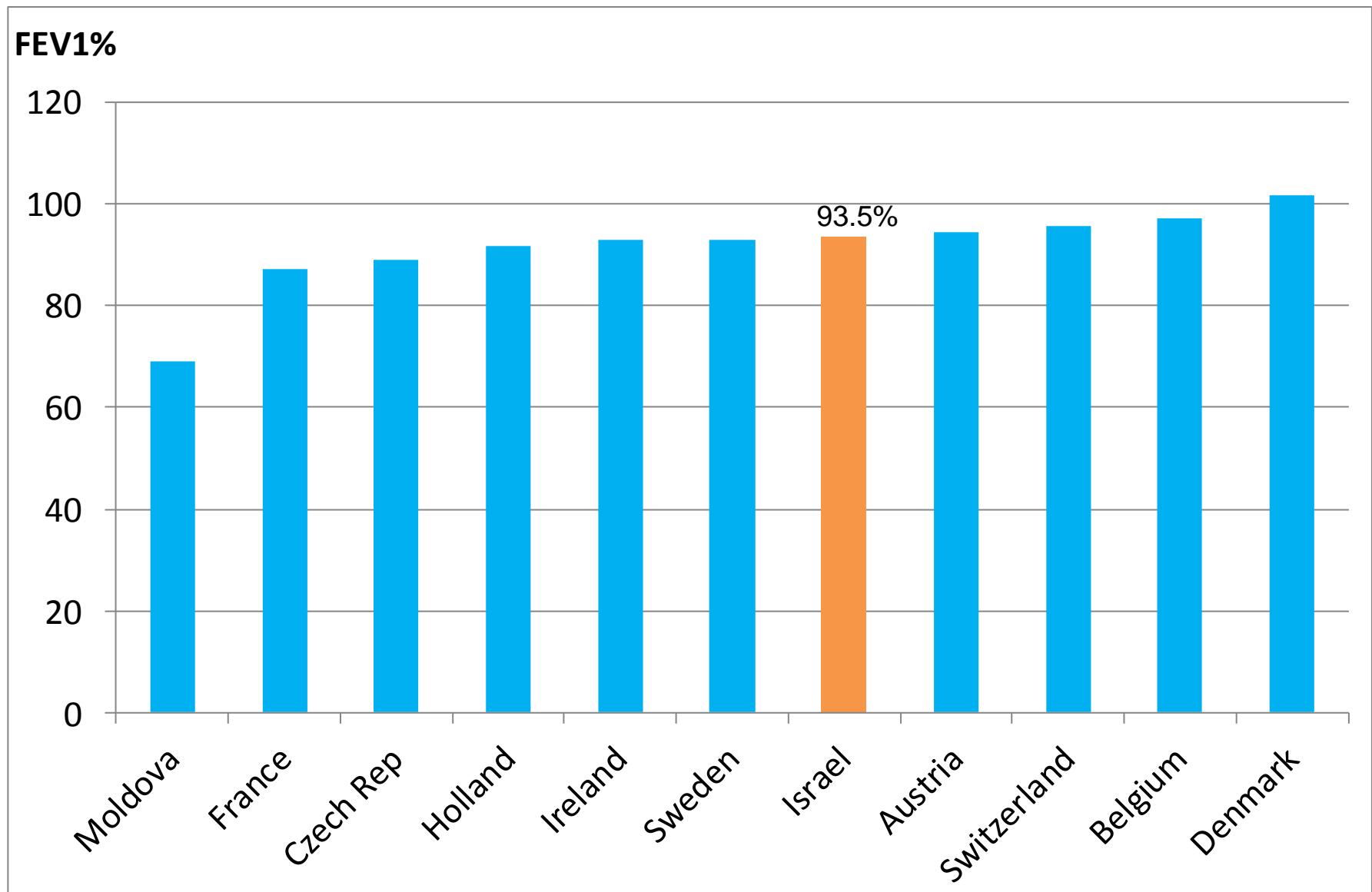
Germany reported the FEV1 value closest to the patient's birthday (without exacerbation),

Sweden reported FEV1 collected at the time of the annual review.

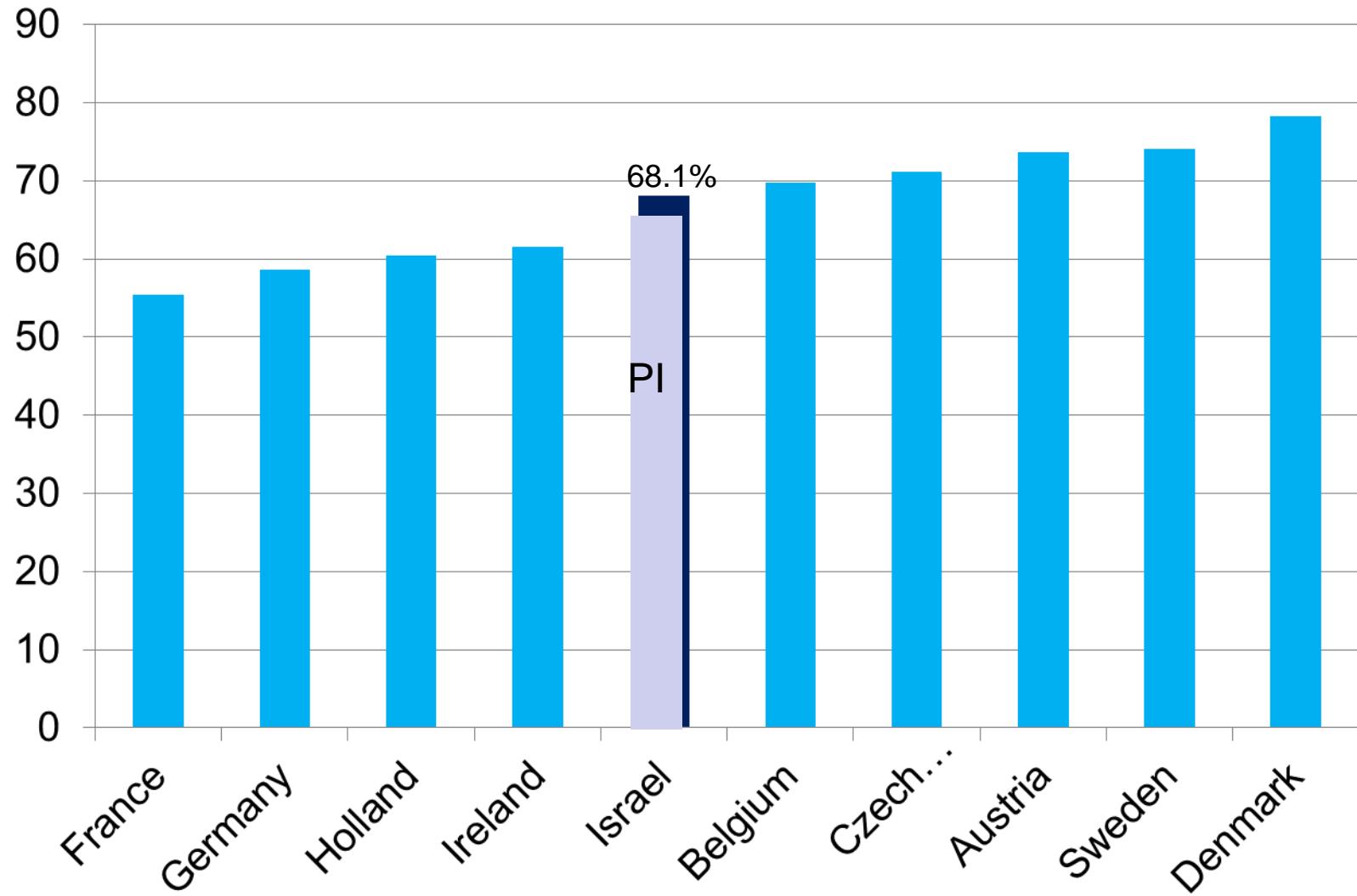
FEV1- Trend - Israel



FEV1 (med) in Children 6-17y

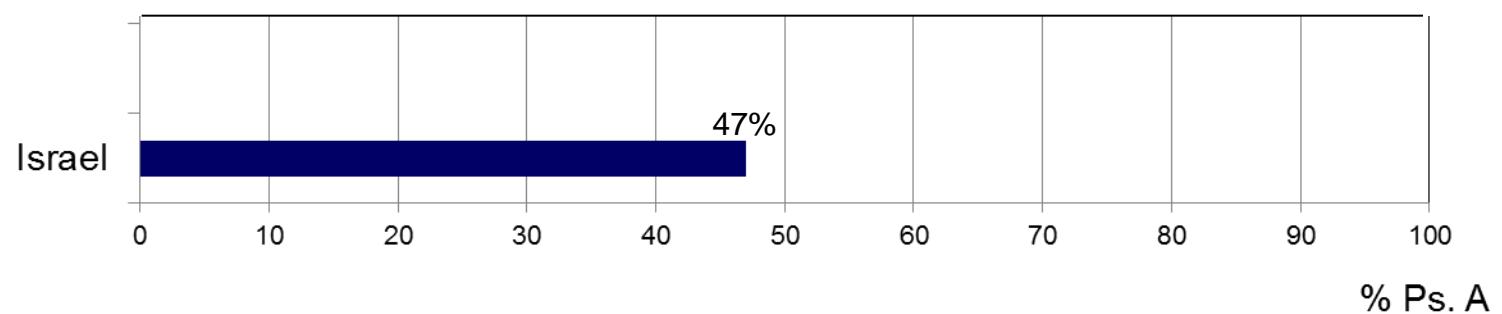


FEV1 (med) in Adults 18+y

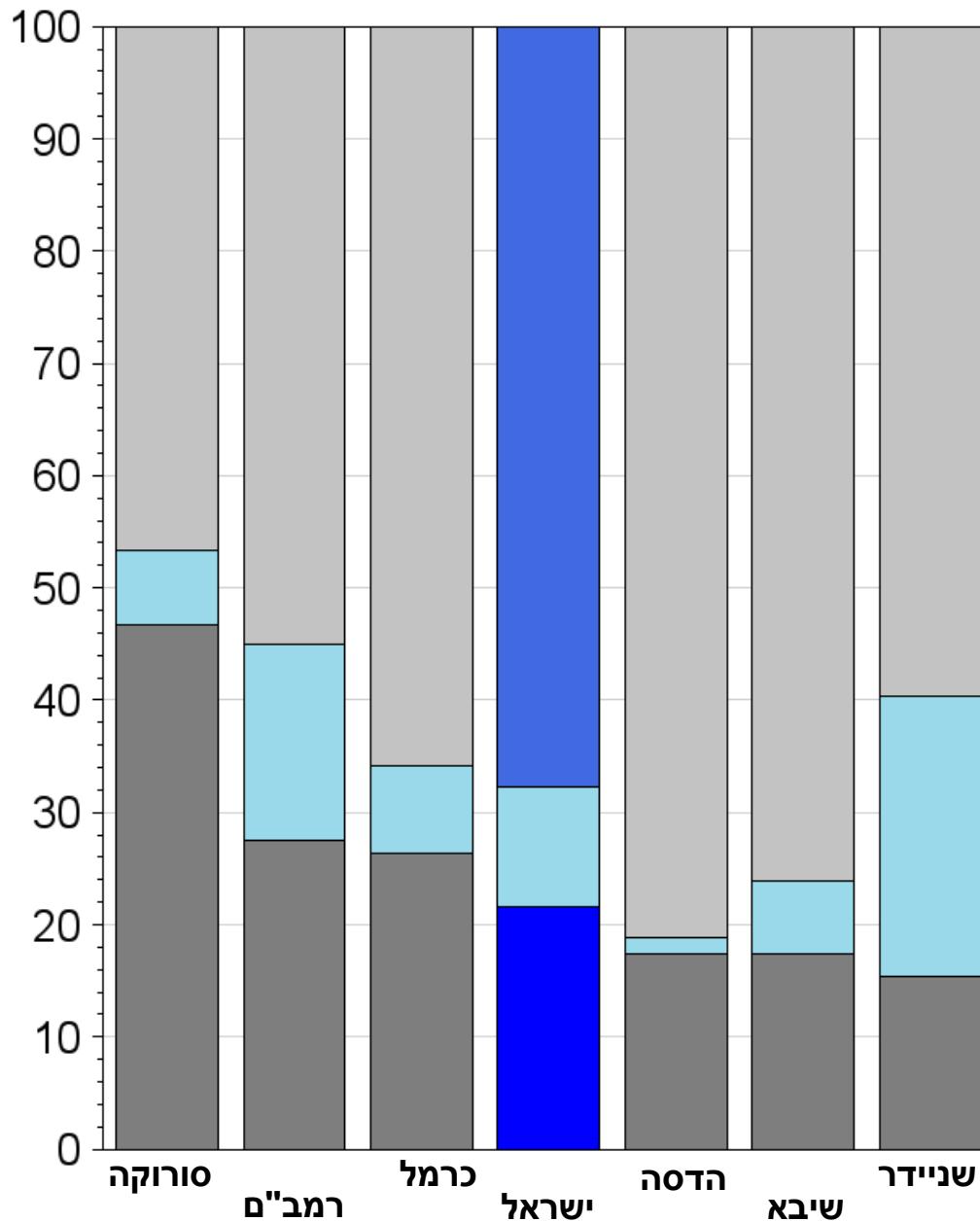




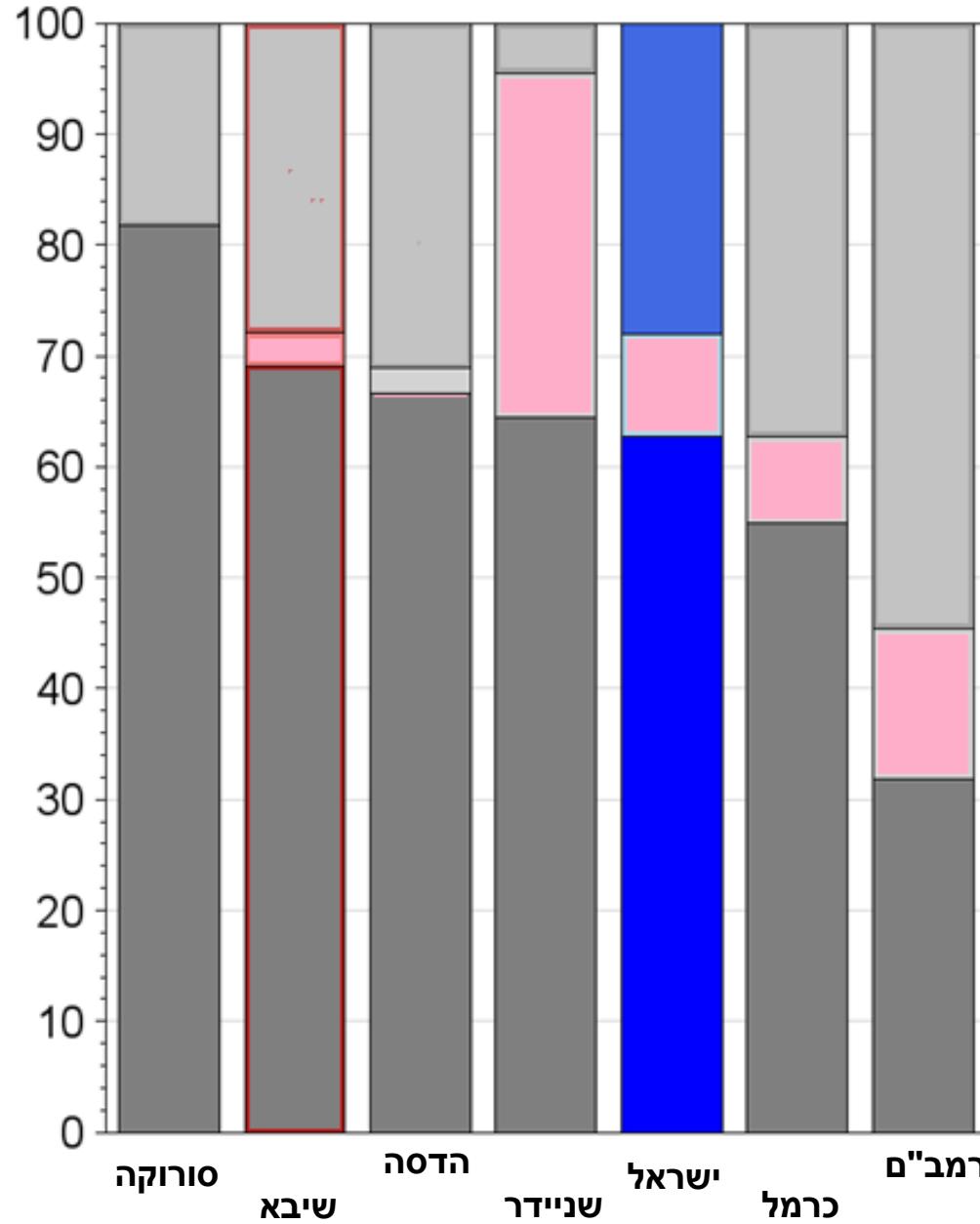
% Chronic Ps. A



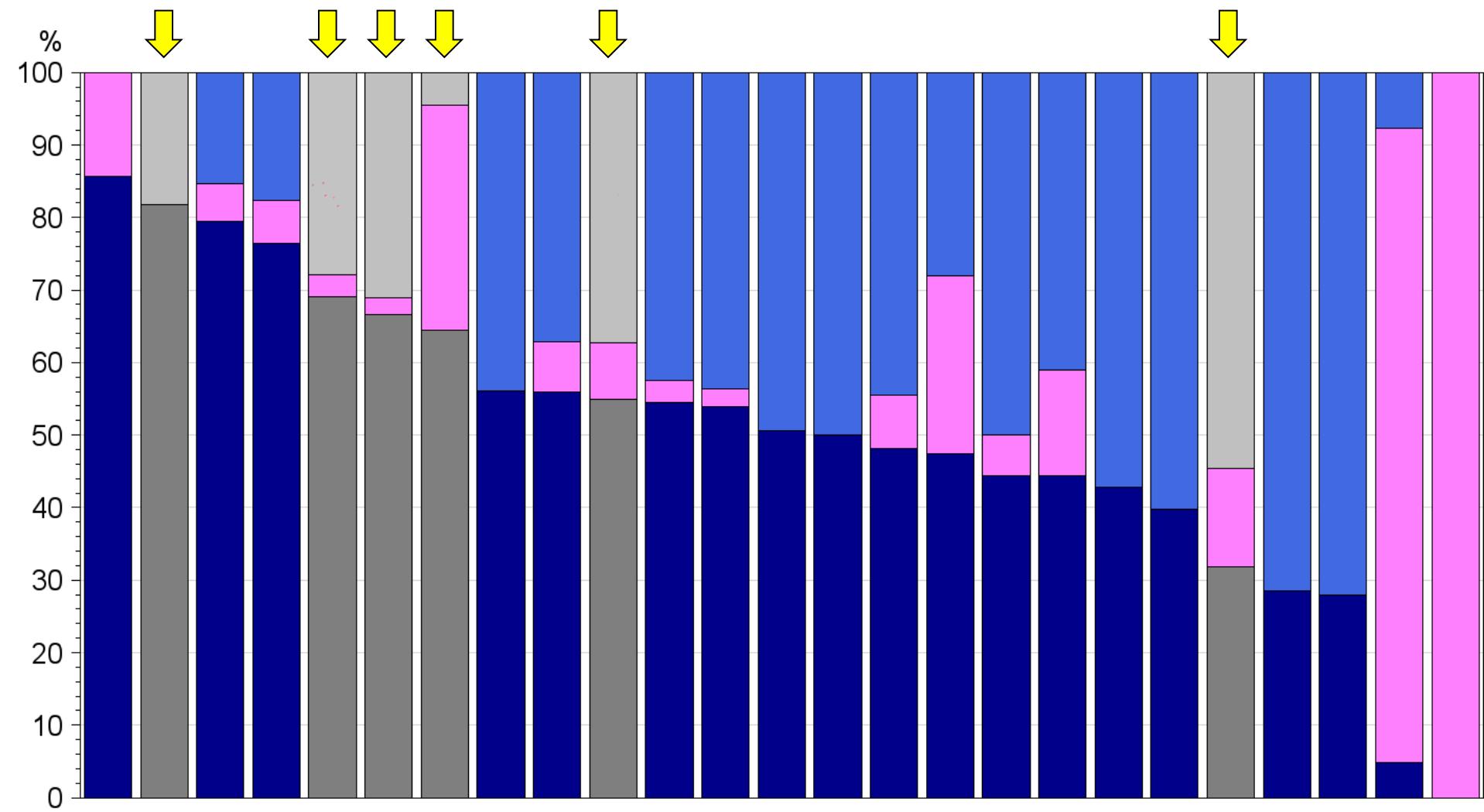
Presence of chronic *Pseudomonas aeruginosa* infection in patients aged less than 18 years



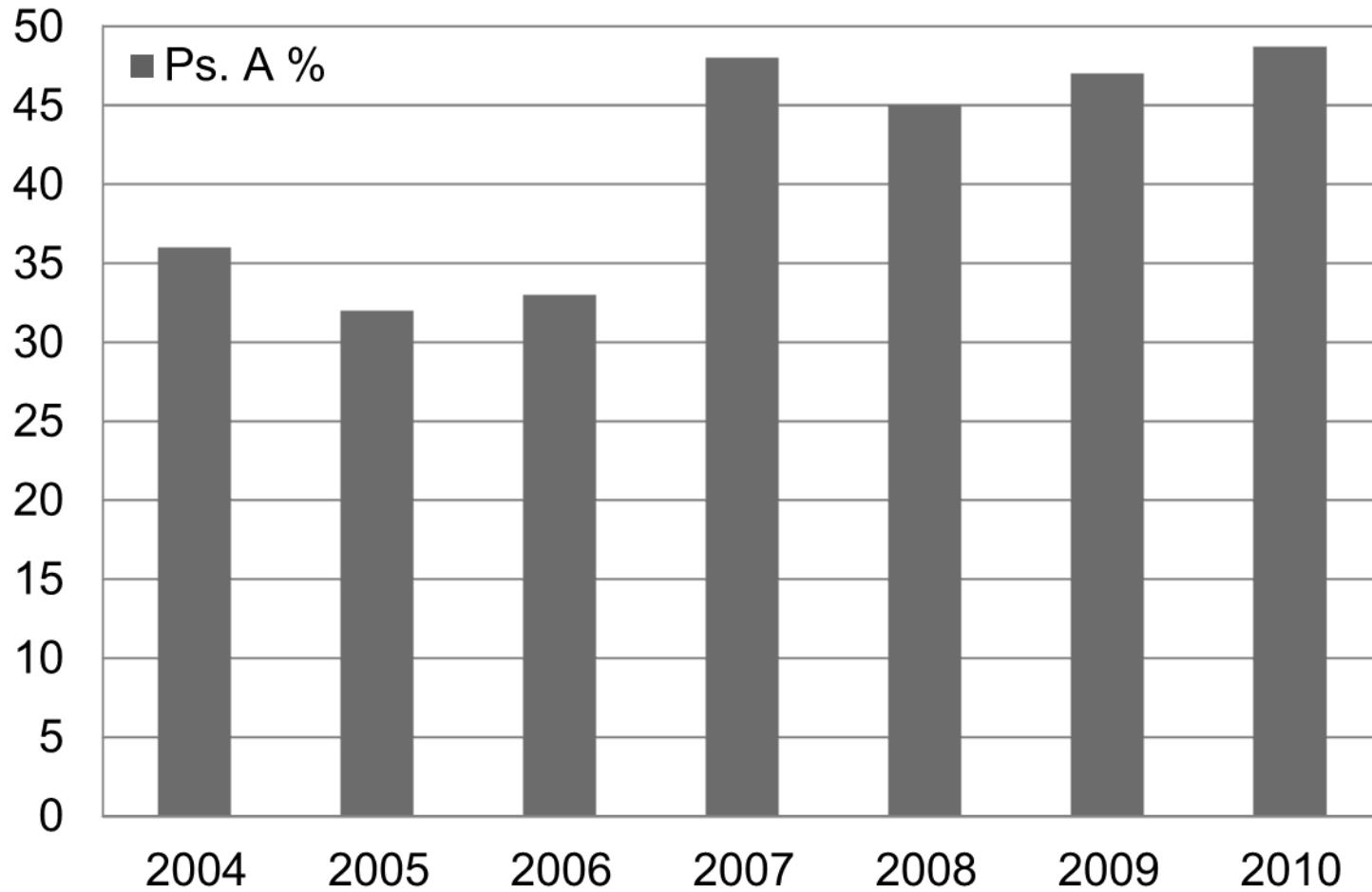
Presence of chronic Pseudomonas aeruginosa infection in patients aged 18 years or more



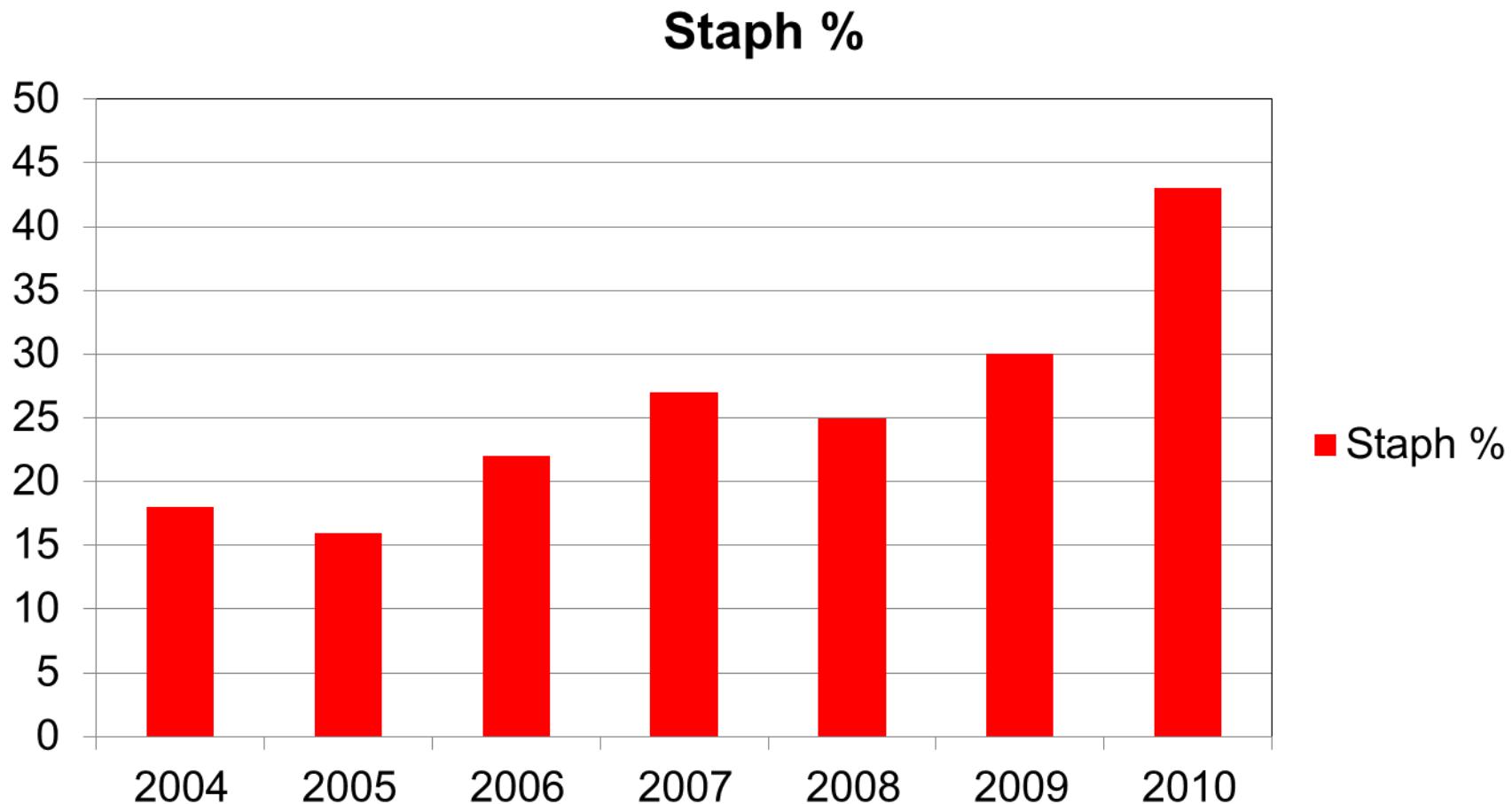
Presence of chronic Pseudomonas aeruginosa in patients aged 18 years or more



Ps. A in Israel- Trend



Staph. in Israel- Trend



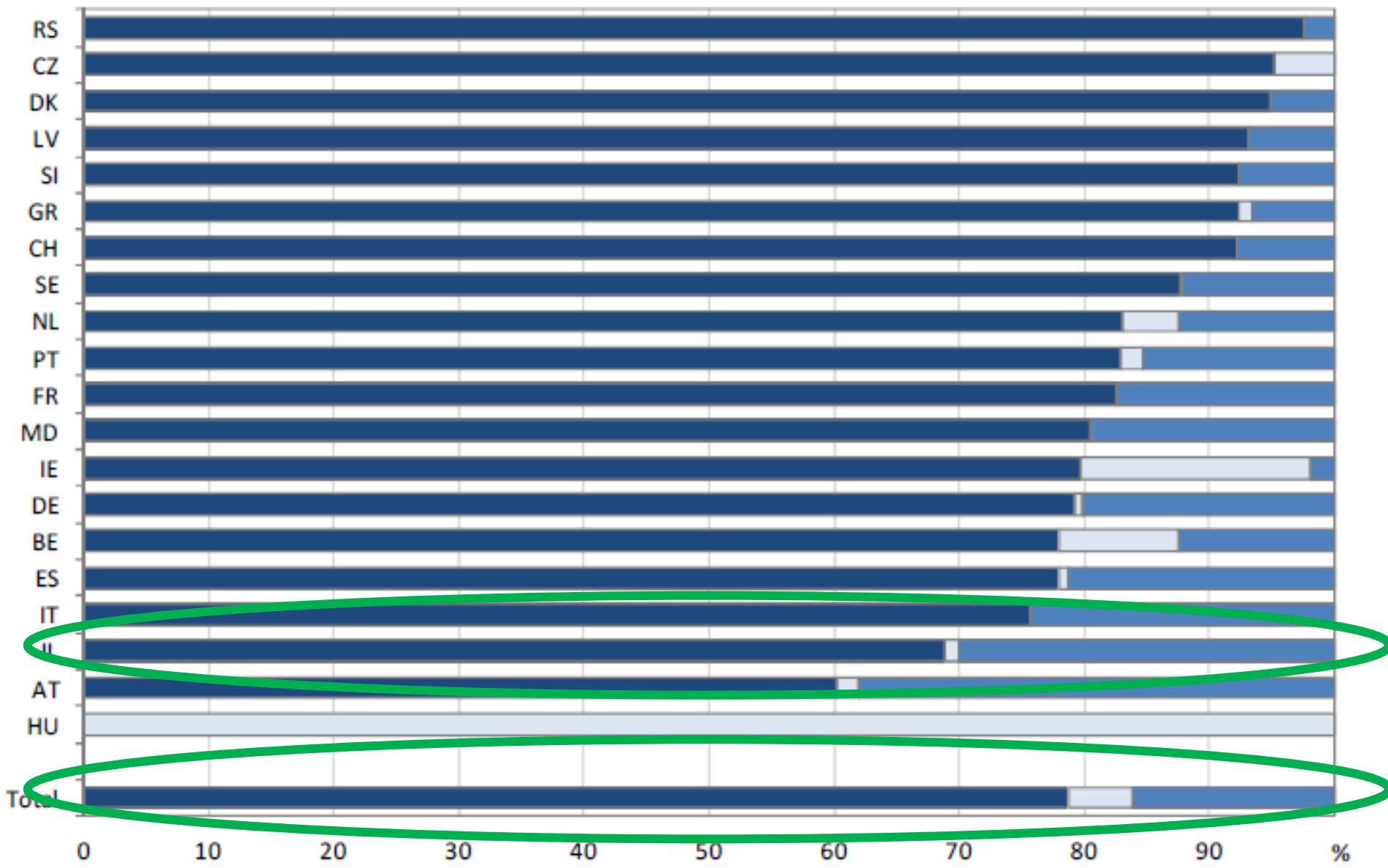
Other Infections – Israel 2010

- B Cepacia 3/378- **0.8%**
- Non Tub. MB 19/386- **19%**
- Stenotrophom. 15/380- **15%**

GI

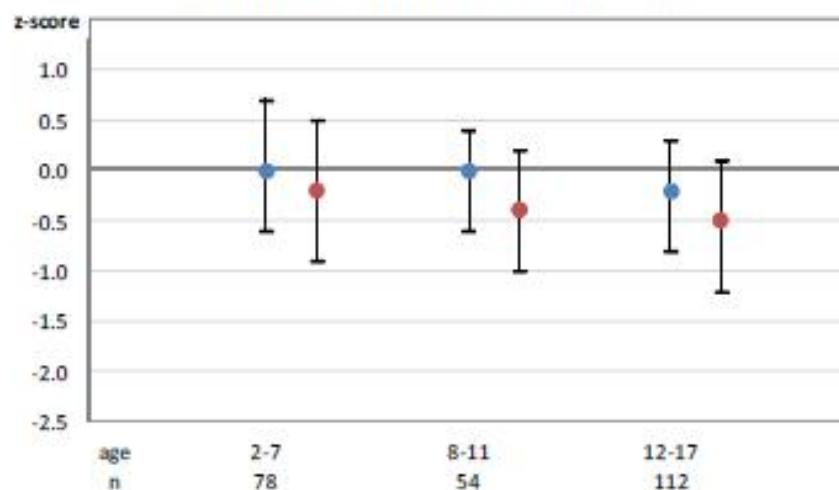
Use of Pancreatic Enzymes

■ Yes □ Missing/Unknown ■ No



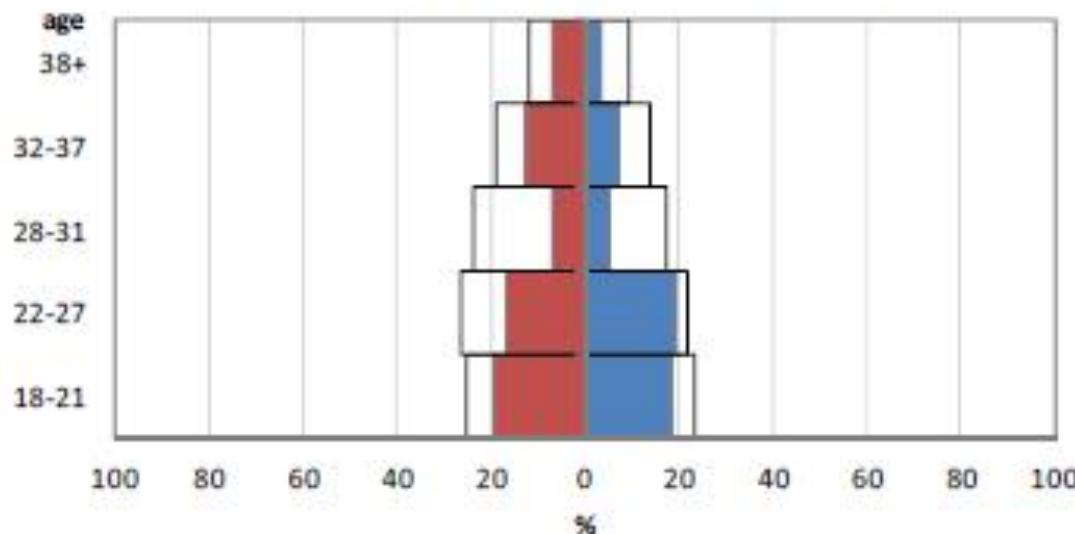


Quartiles of z-scores for BMI: Israel

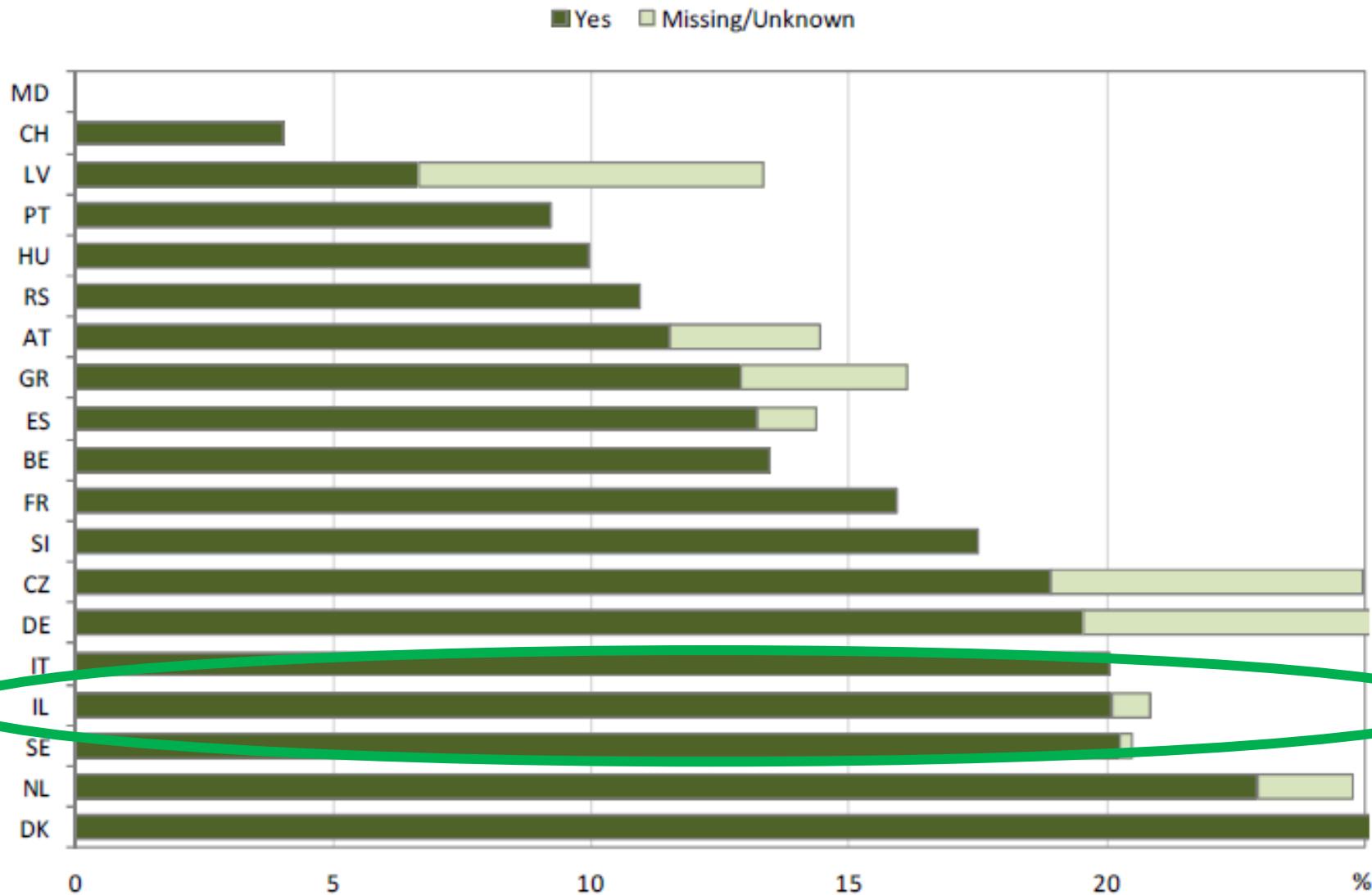


BMI<18.5 kg/m²: Israel

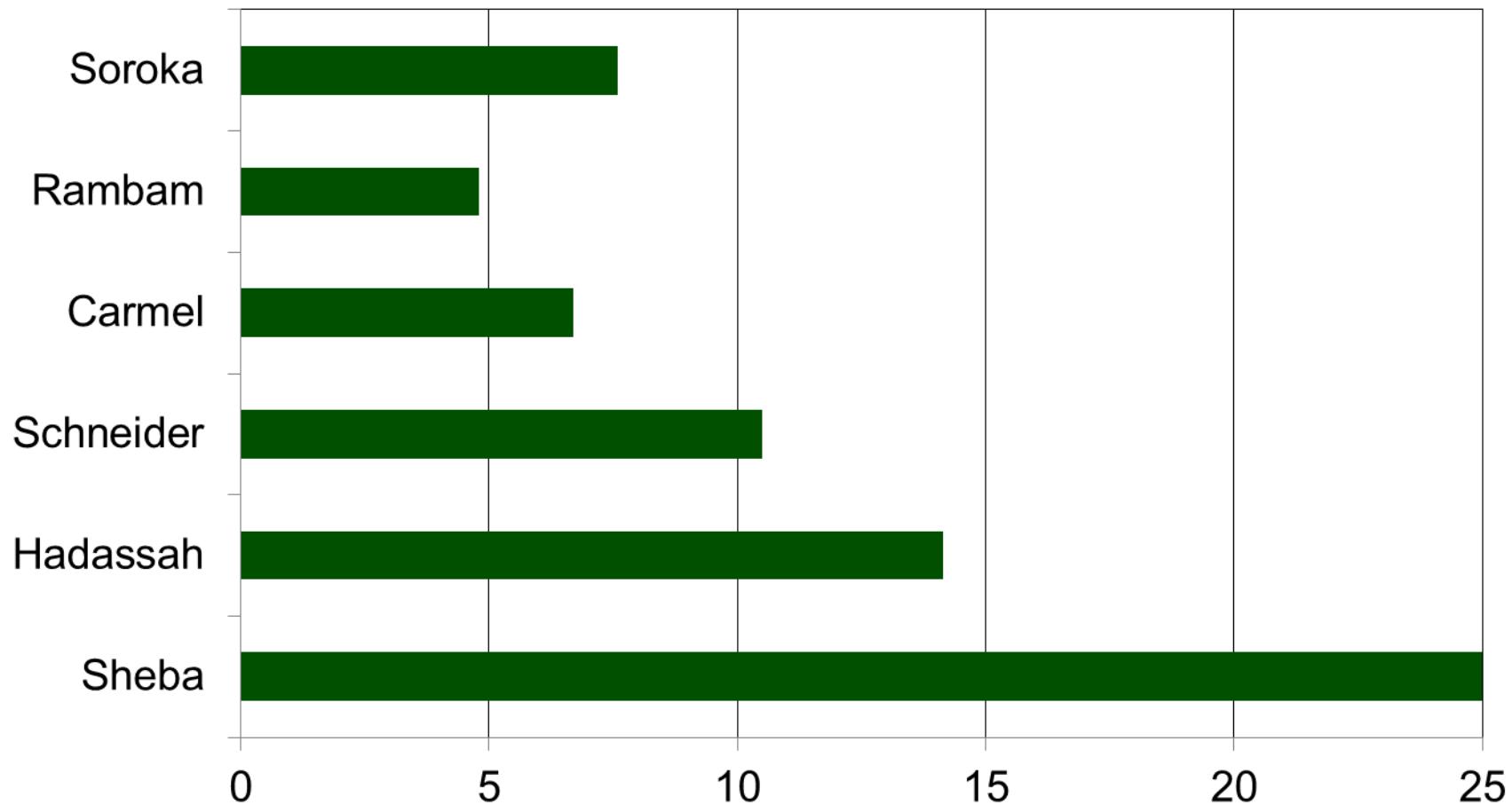
□ Other countries ■ Israel (F) ■ Israel (M)



CFRD



CFRD



Other Complications – Israel 2010

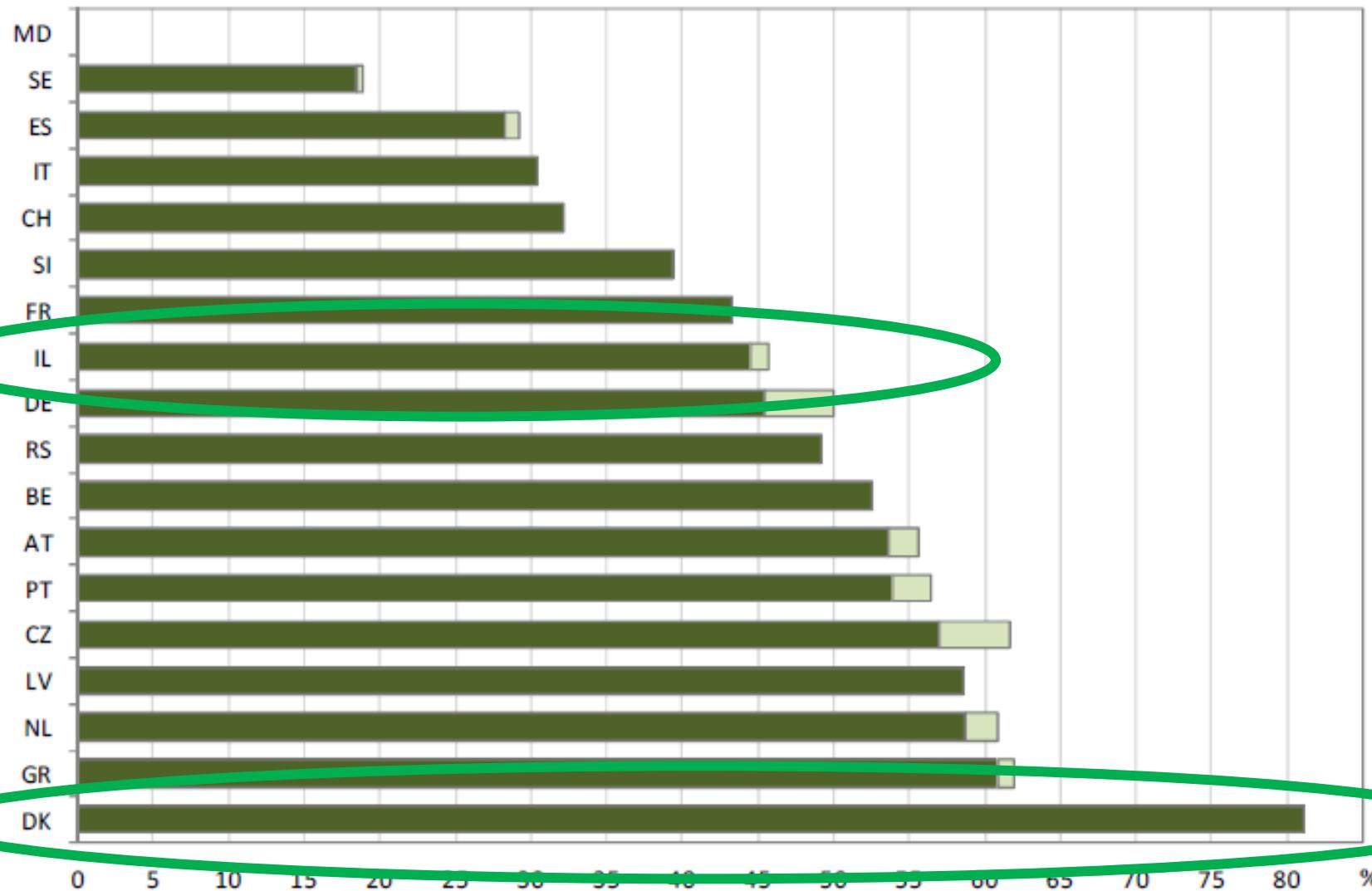
- ABPA- 5.3%
- Liver Dis- 17.8%
- Major hemoptysis- 1.4%

- Demographics
- Clinical Characteristics
- Treatment



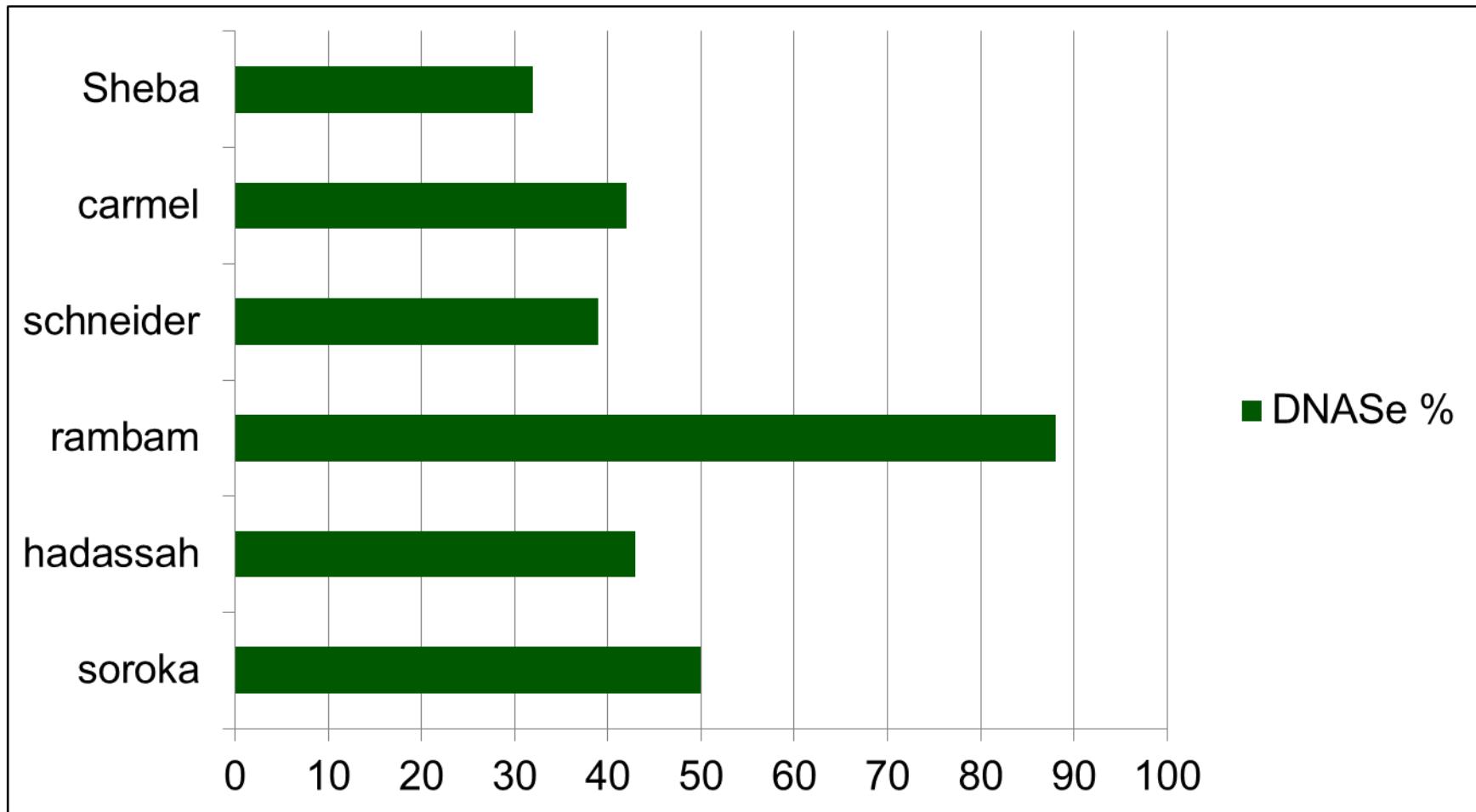
DNASE- Europe

■ Yes ■ Missing/Unknown

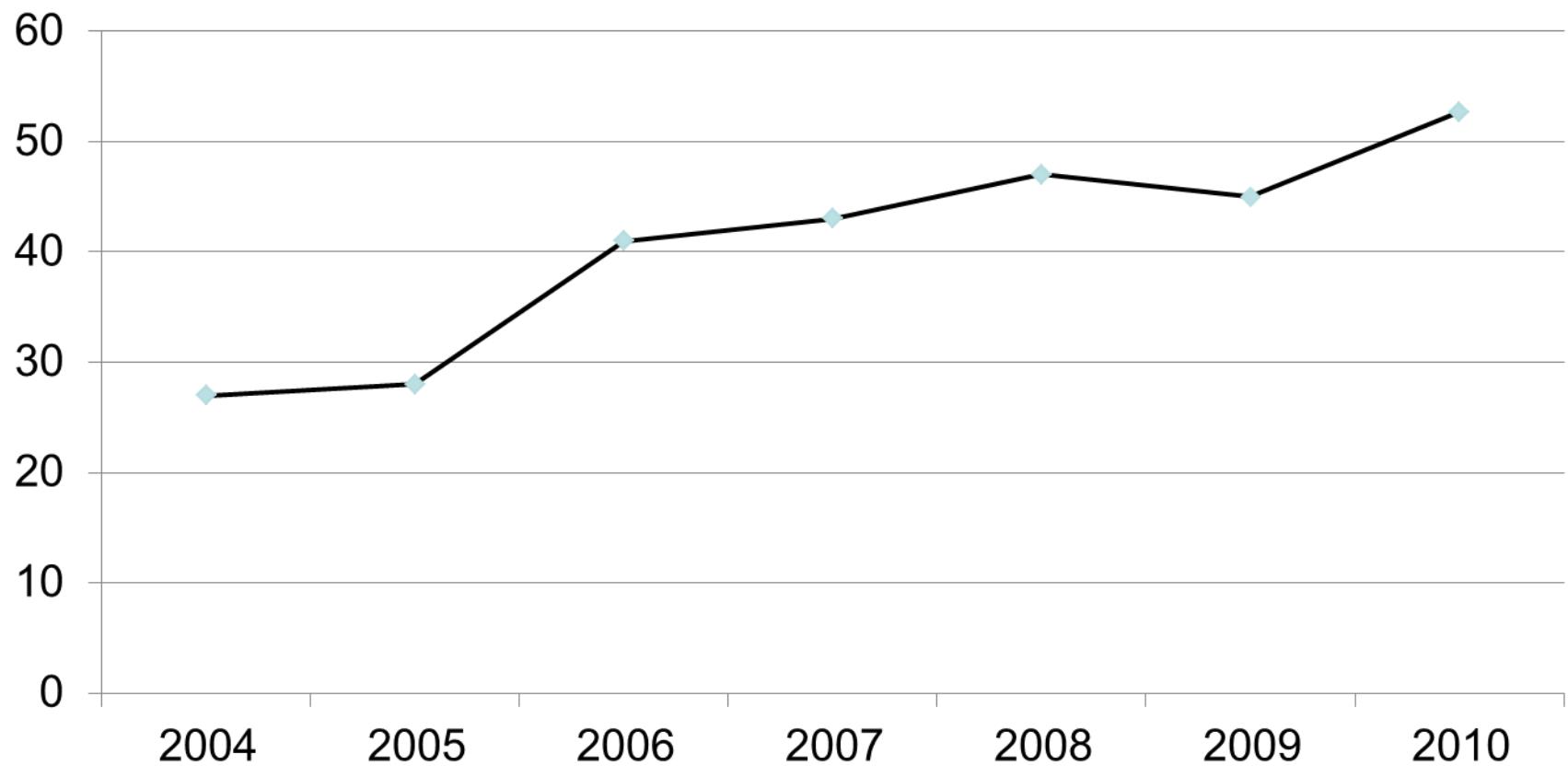




DNASE

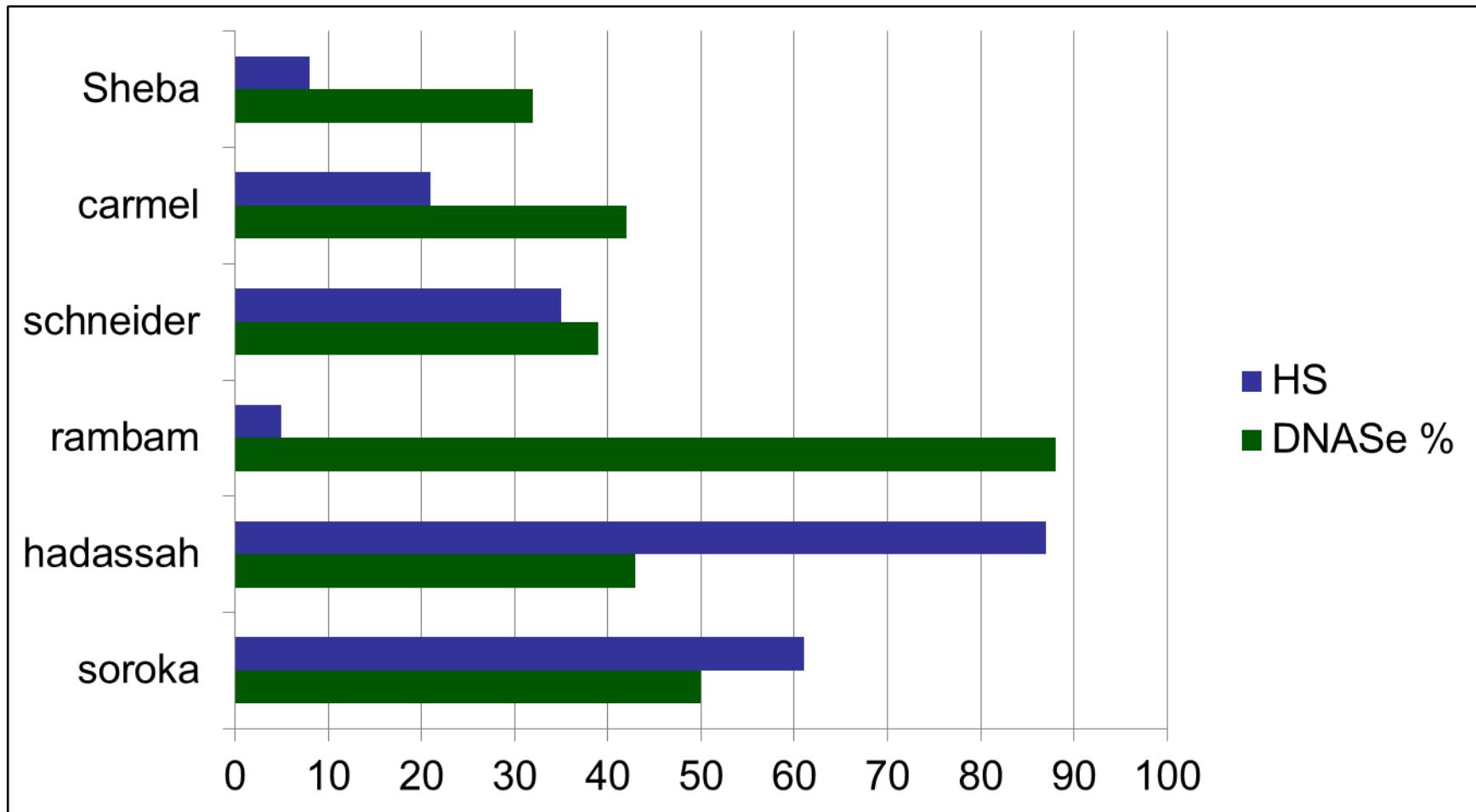


DNASE %



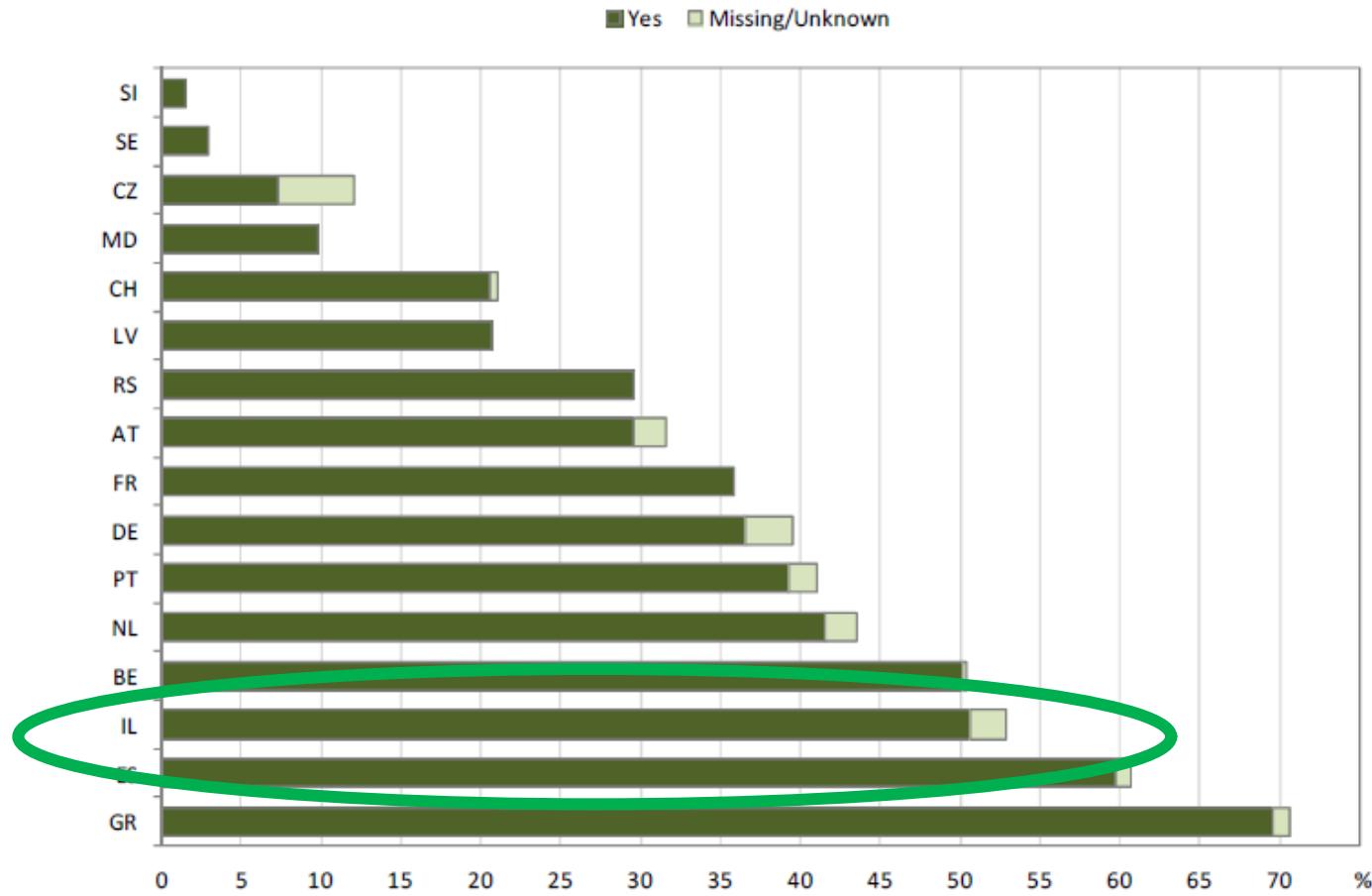


DNASE & HS



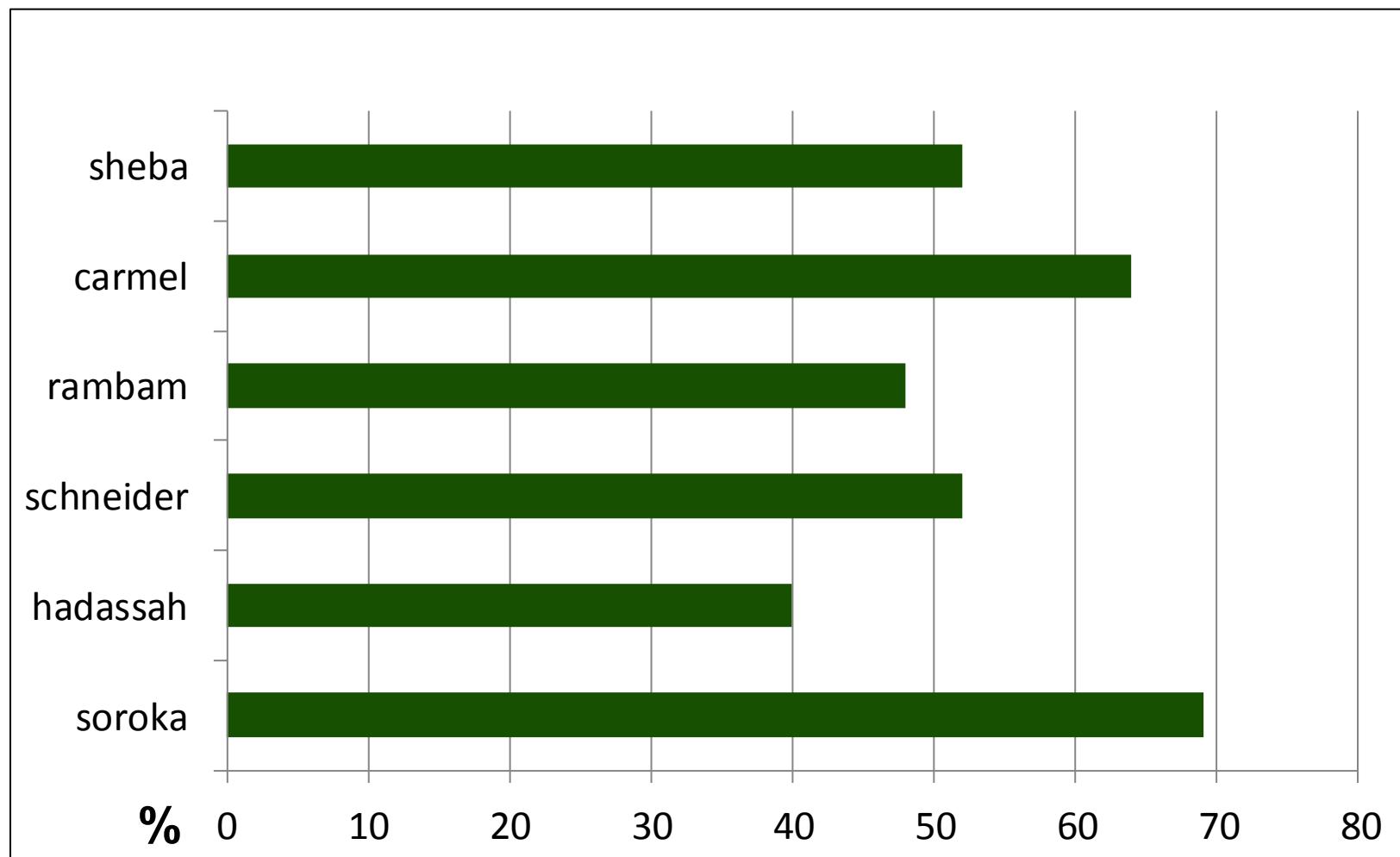


INH Antibiotics- Europe

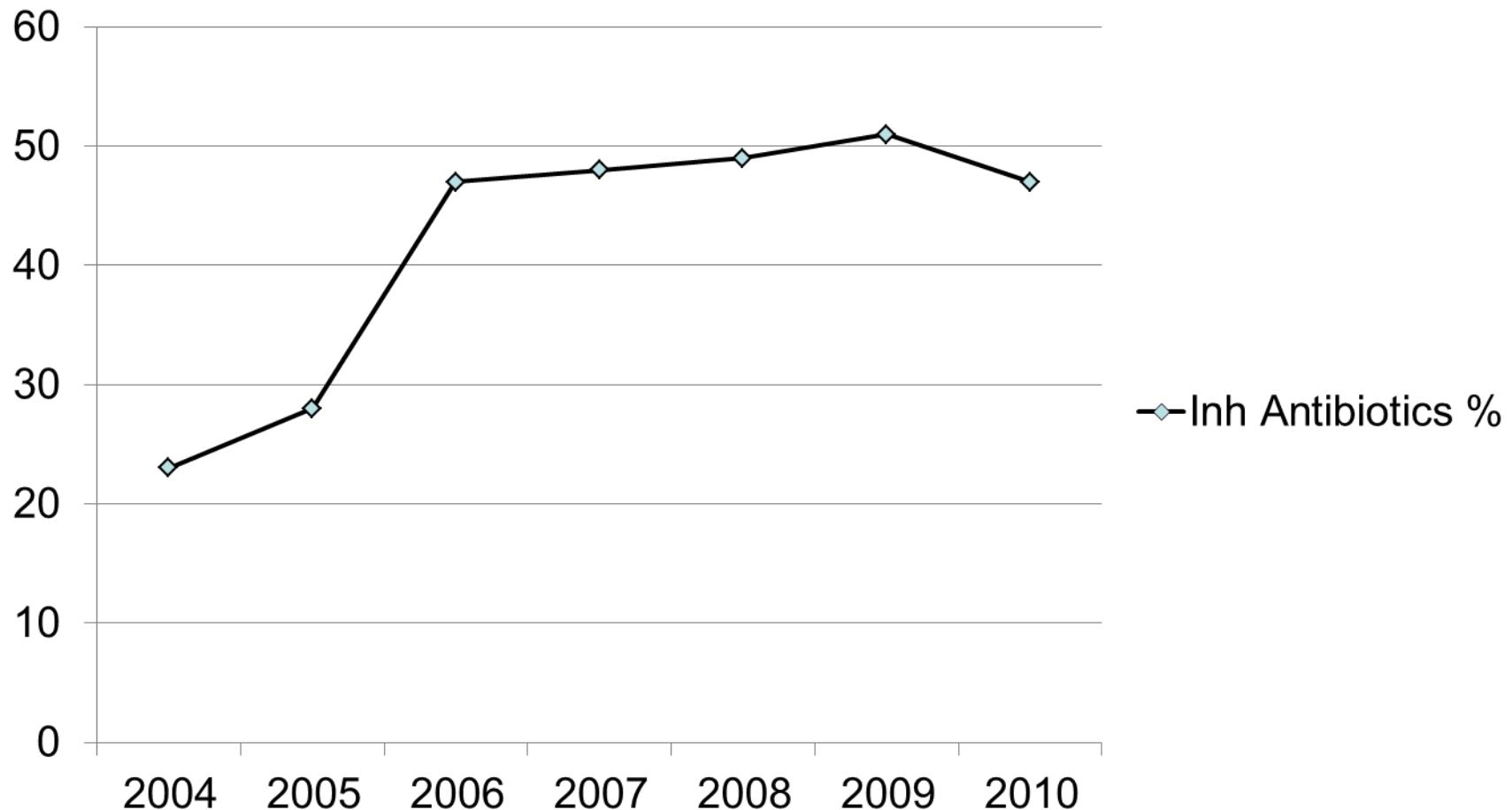




INH Antibiotics



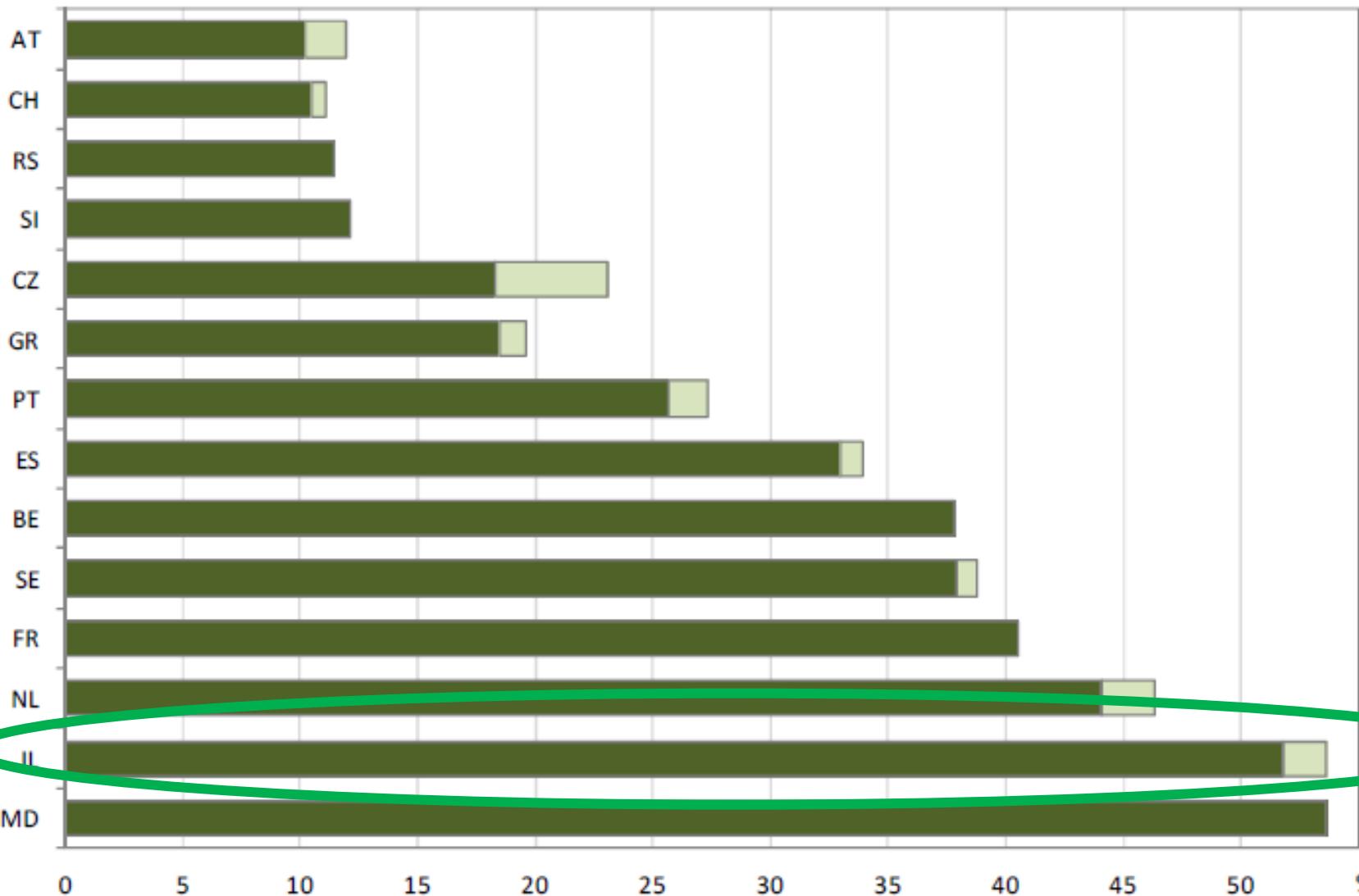
Inhaled Antibiotics- %





AZENIL

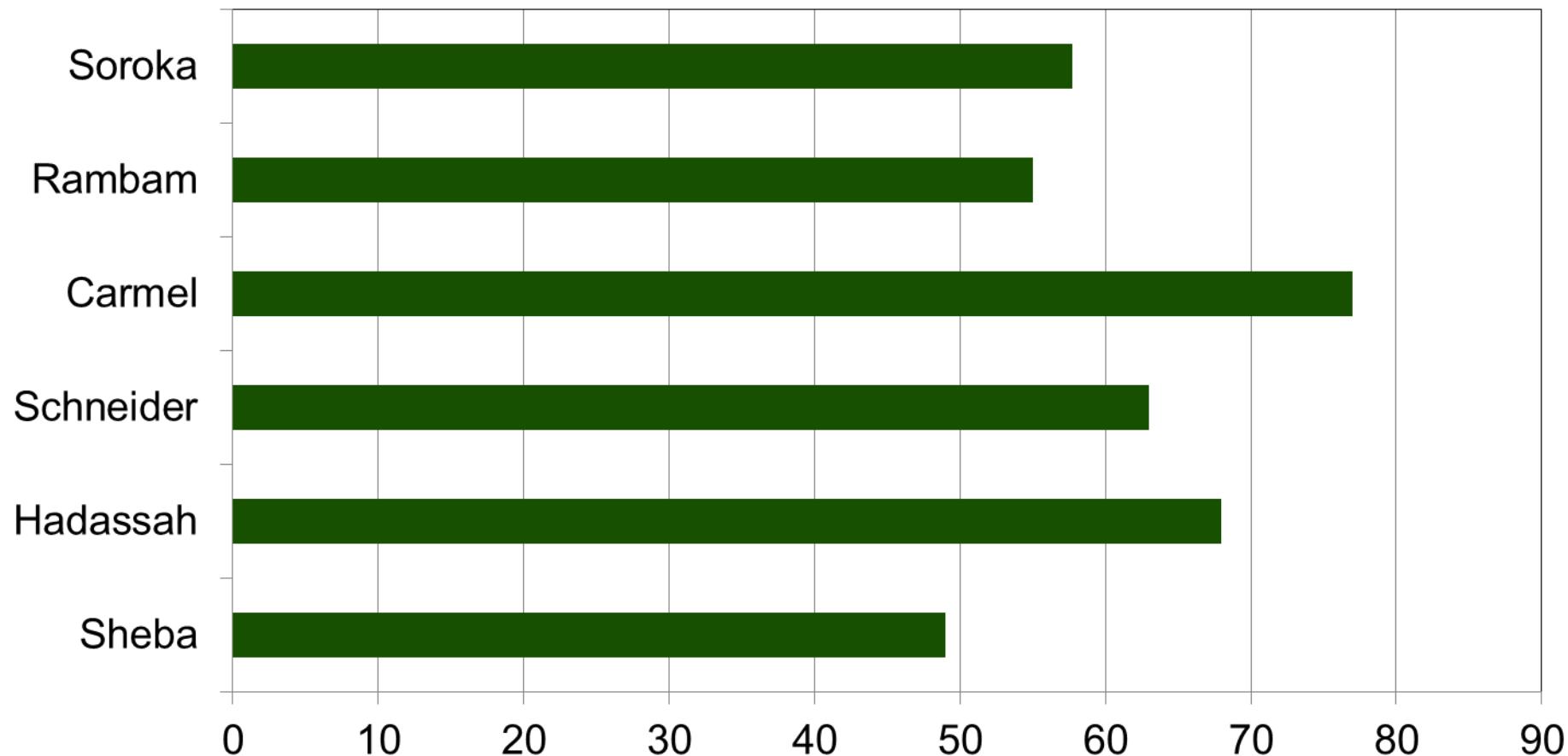
■ Yes ■ Missing/Unknown



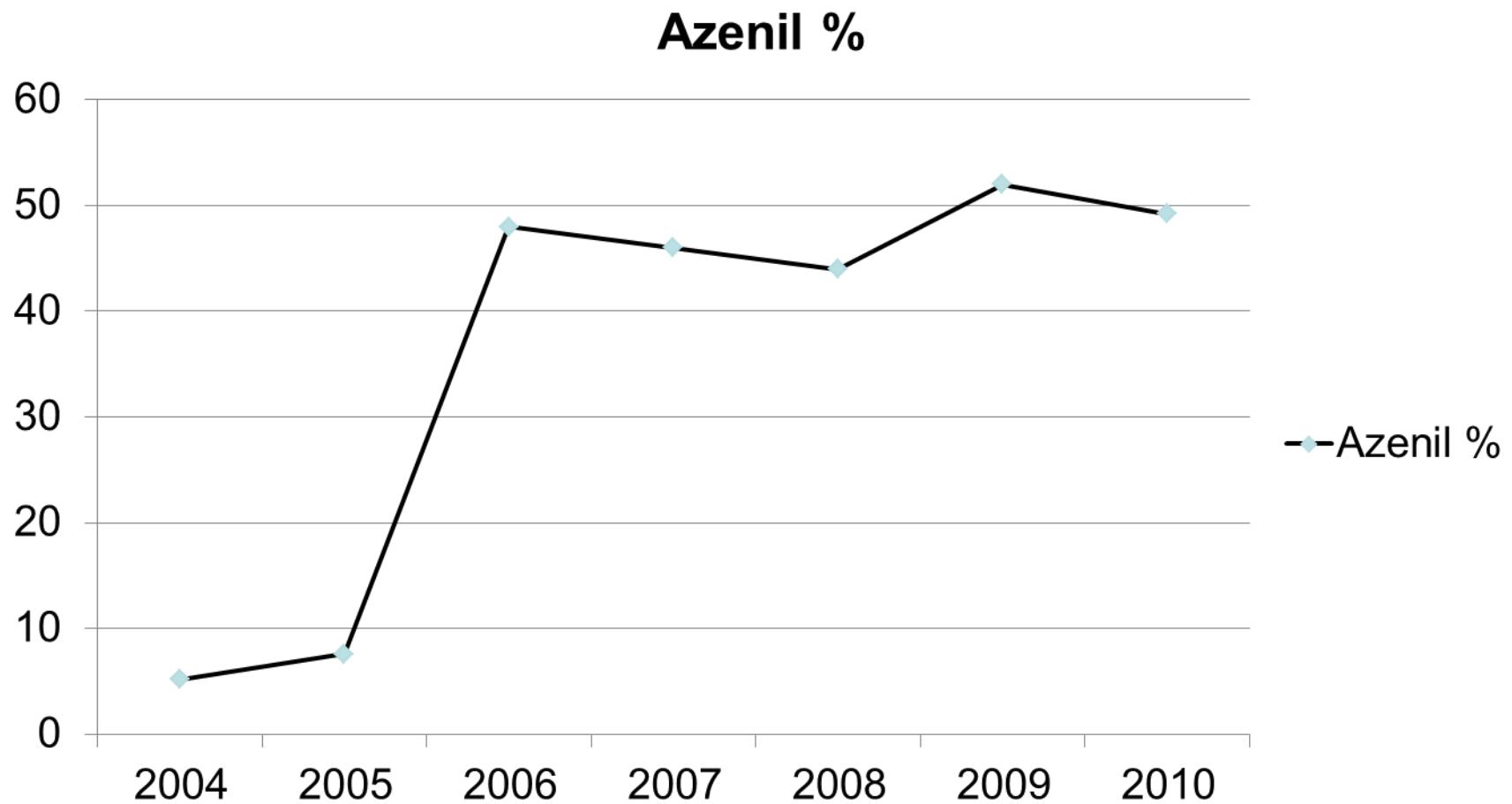


AZENIL

% on Azenil



Azenil Trend- Israel



Summary

- ECFR is growing, aiming to include all European countries
- Israel 2010- trend of delayed diagnosis compared to Europe
- Pulmonary outcome seems to be as good as in Europe
- Trend of improvement in FEV1 but higher rate of Ps. colonization

Summary

- Higher prevalence of *Pseudomonas* requires measures to reduce it
- Database collection is feasible but there is still a lot to improve....

Thanks

- ECFR
- Dr Hanne Olsen
- All team members at the Israeli center
- The patients and families