

# **The Israeli CF Database**

## **2015**

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Schneider Children's Medical Center of Israel

# European CF Patient Registry (ECFSPR)

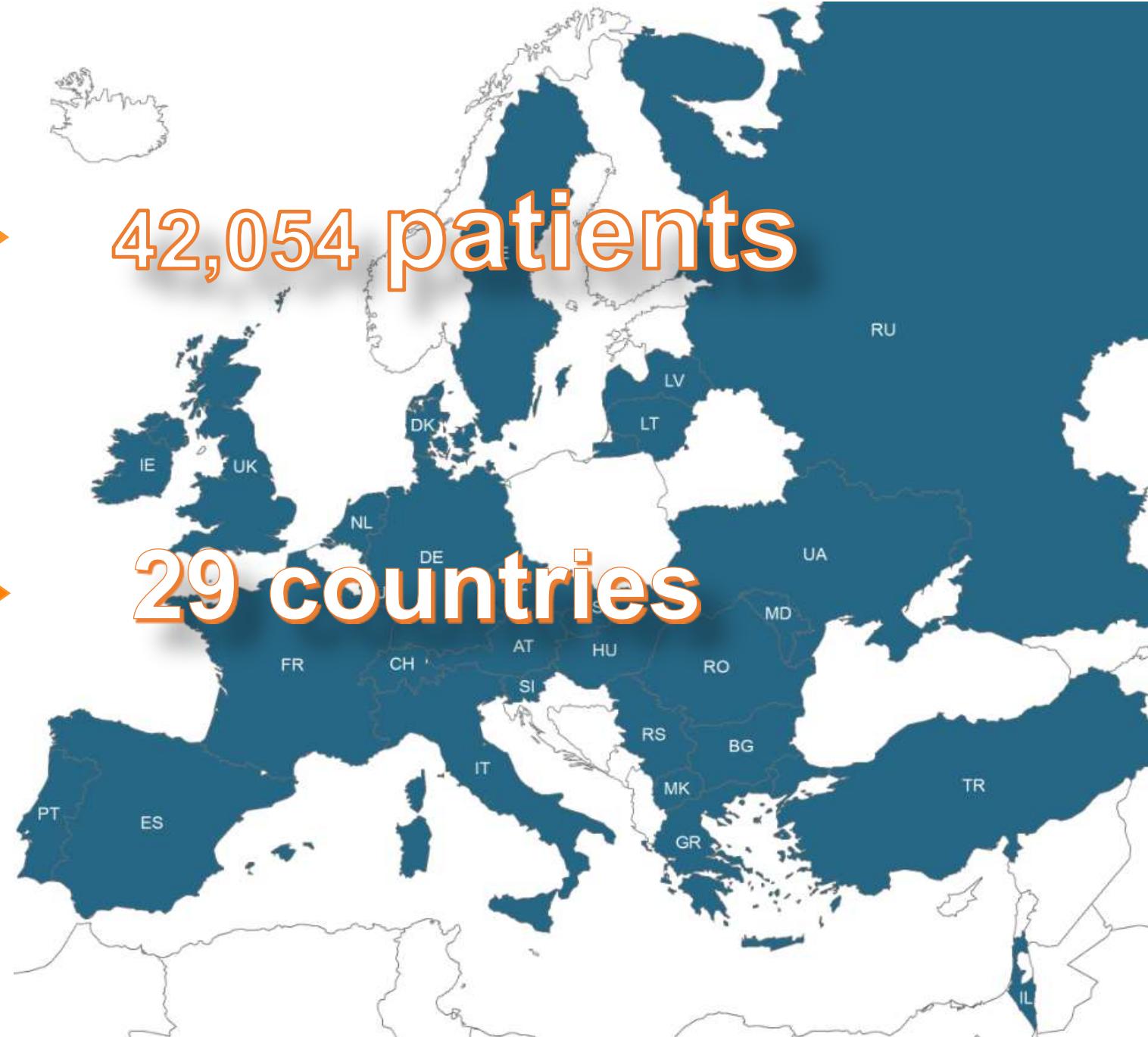




42,054 patients



29 countries





EUR



Austria



Belgium



Bulgaria



Czech  
Republic



Denmark



France



Germany



Greece



Hungary



Ireland



Israel



Italy



Latvia



Lithuania



Macedonia



Moldova



Netherlands



Portugal



Romania



Russia



Slovakia



Slovenia



Serbia



Spain



Sweden



Switzerland



Ukraine

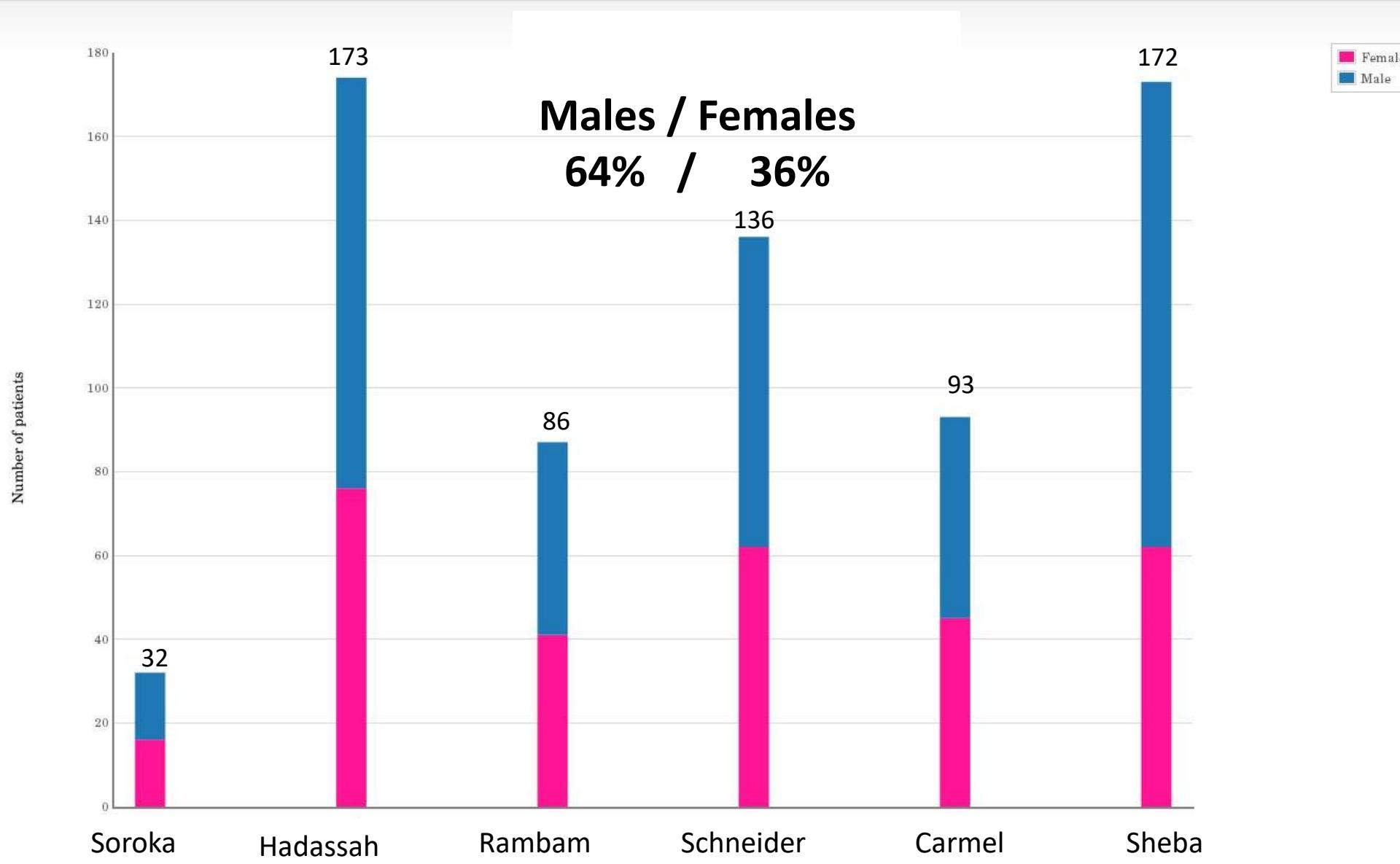


UK

# The Israeli CF Database Update 2015

# Demographics

# Number of Patients in the Registry 2015



# Number of Patients

## In Israel

- Patients registered – 665
- Seen in 2015- 548
- Missing – 17.5%

## In Europe

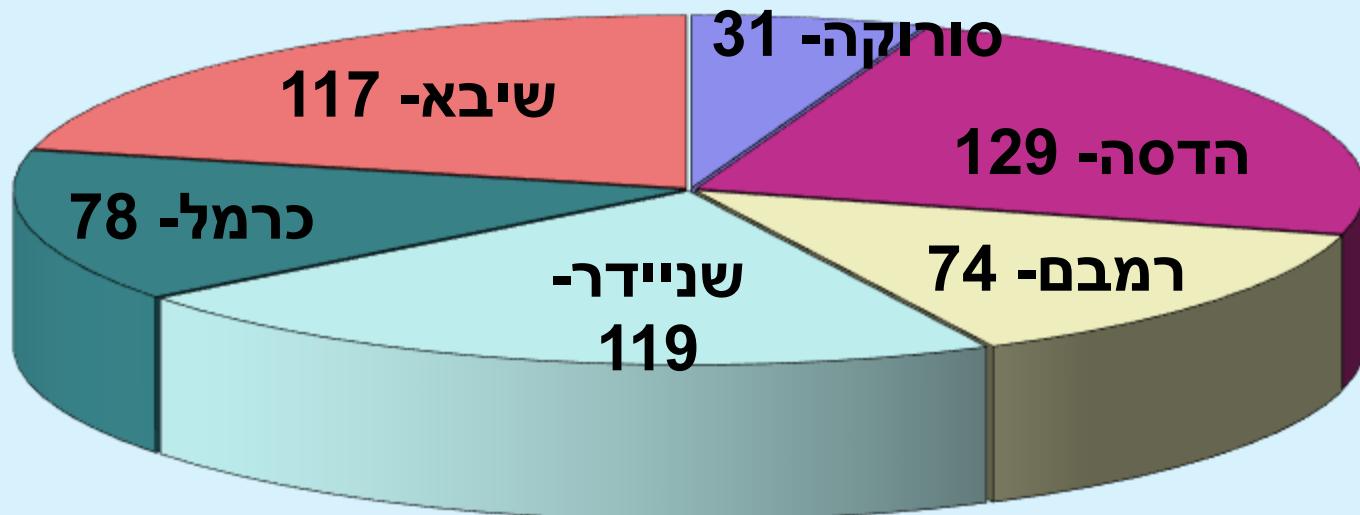
- Patients registered – 42,054
- Seen in 2015- 40,098
- Missing – 4%

# Number of Patients

So, where are they?

- Doubles (patients moving/ visiting 2 centers)
- Tourists / Palestinian
- Patients who come infrequently  
(atypical/mild disease)

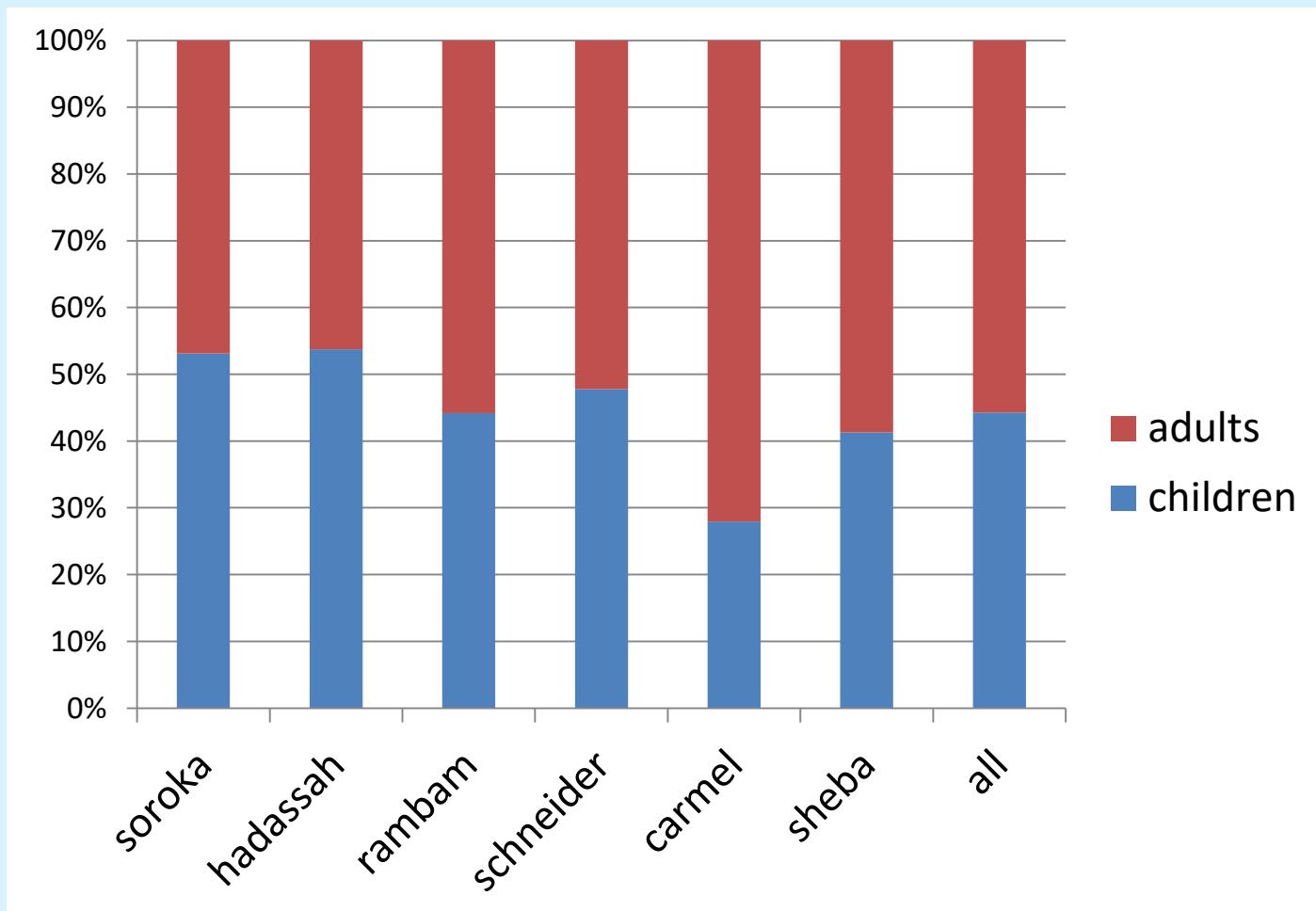
# Israeli CF Centers seen in 2015



Seen in 2015- 548

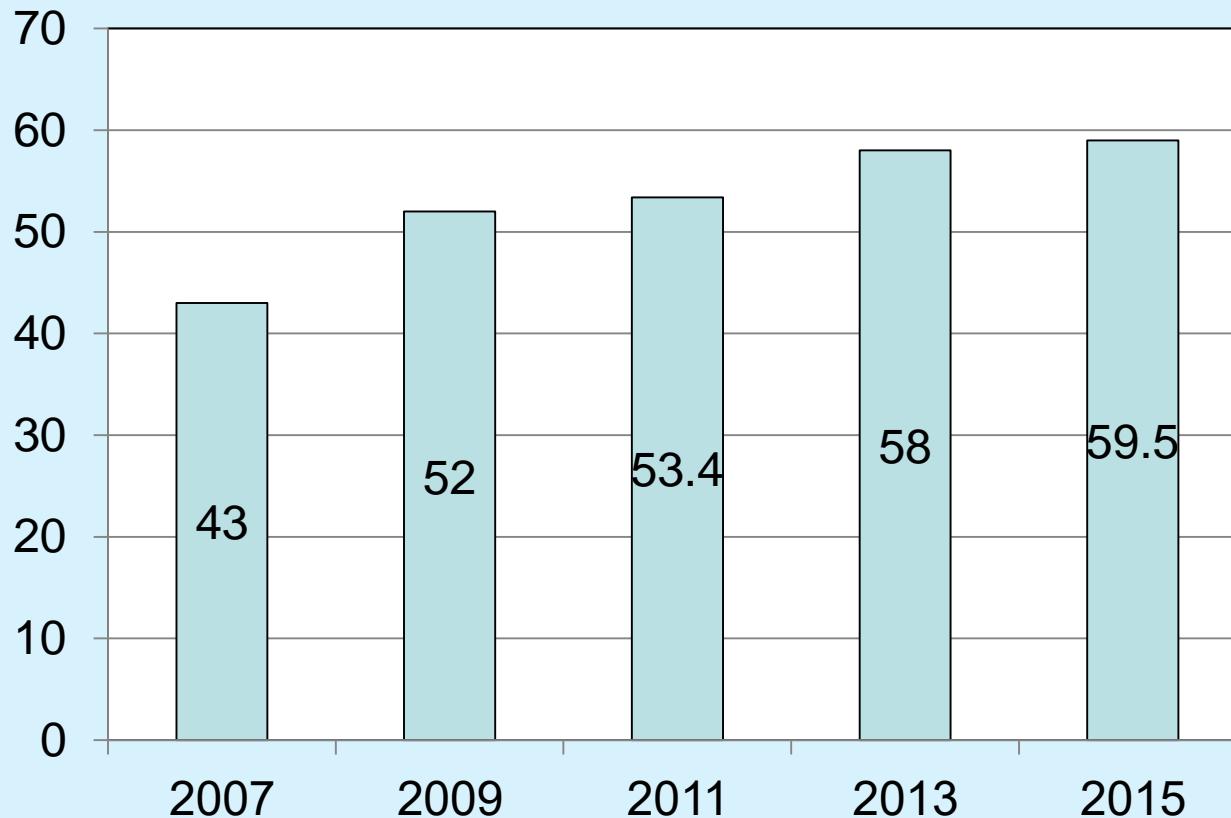
# Children/Adults per Center

Total children-280/692- 40.5%



# % Adults

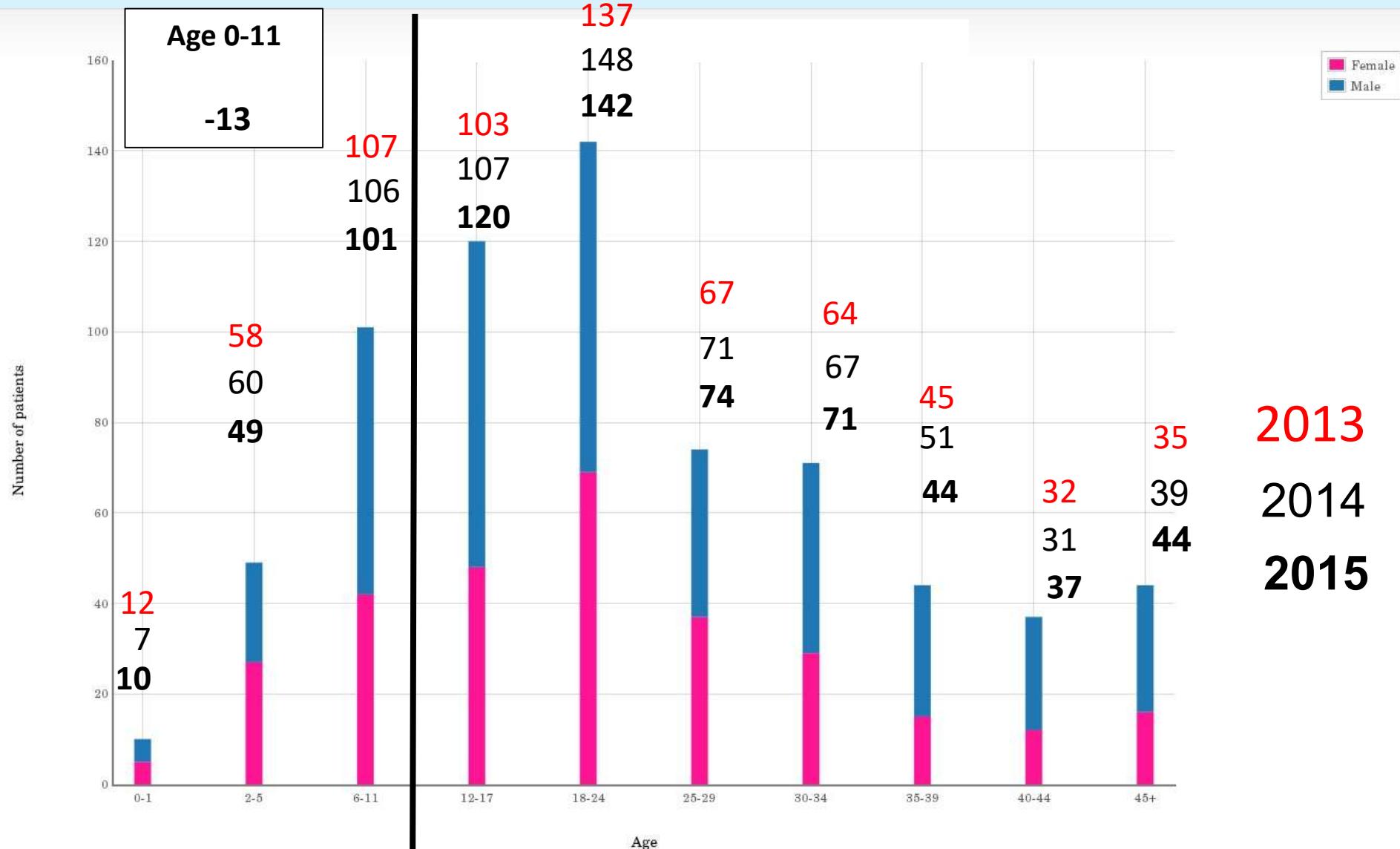
2007-15



EU  
52%

P<0.05

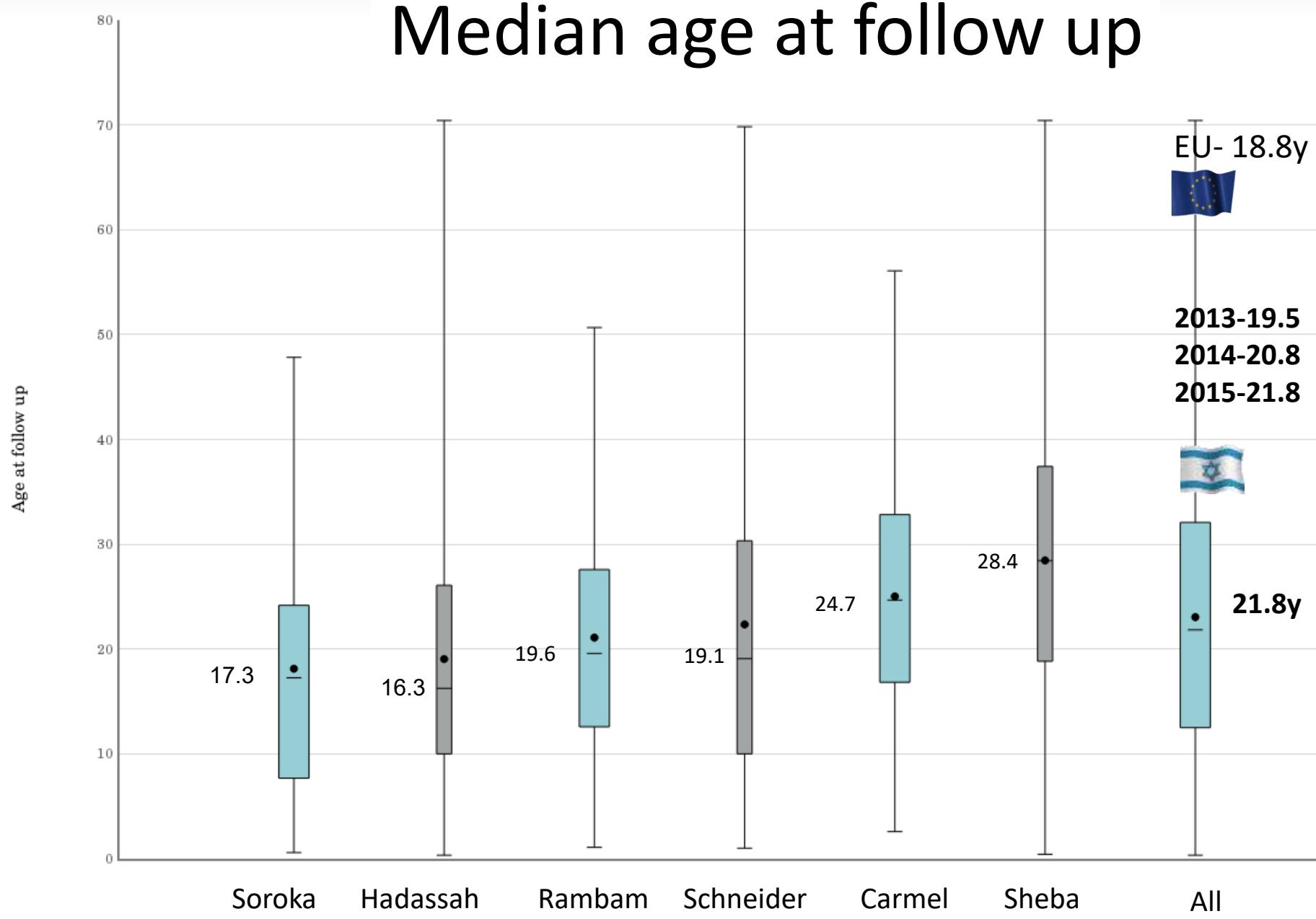
# Age & Gender



# Age at follow up



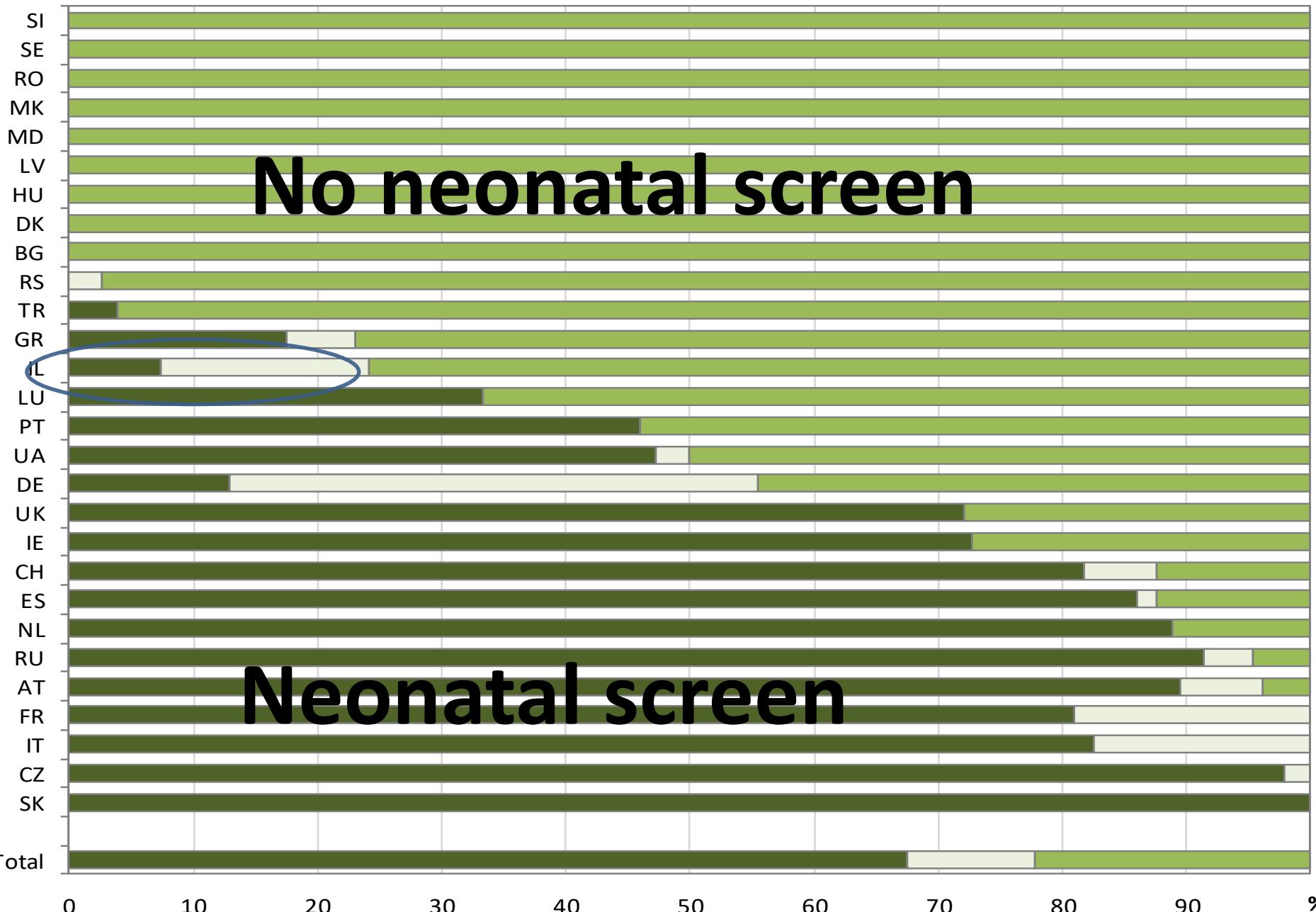
# Median age at follow up



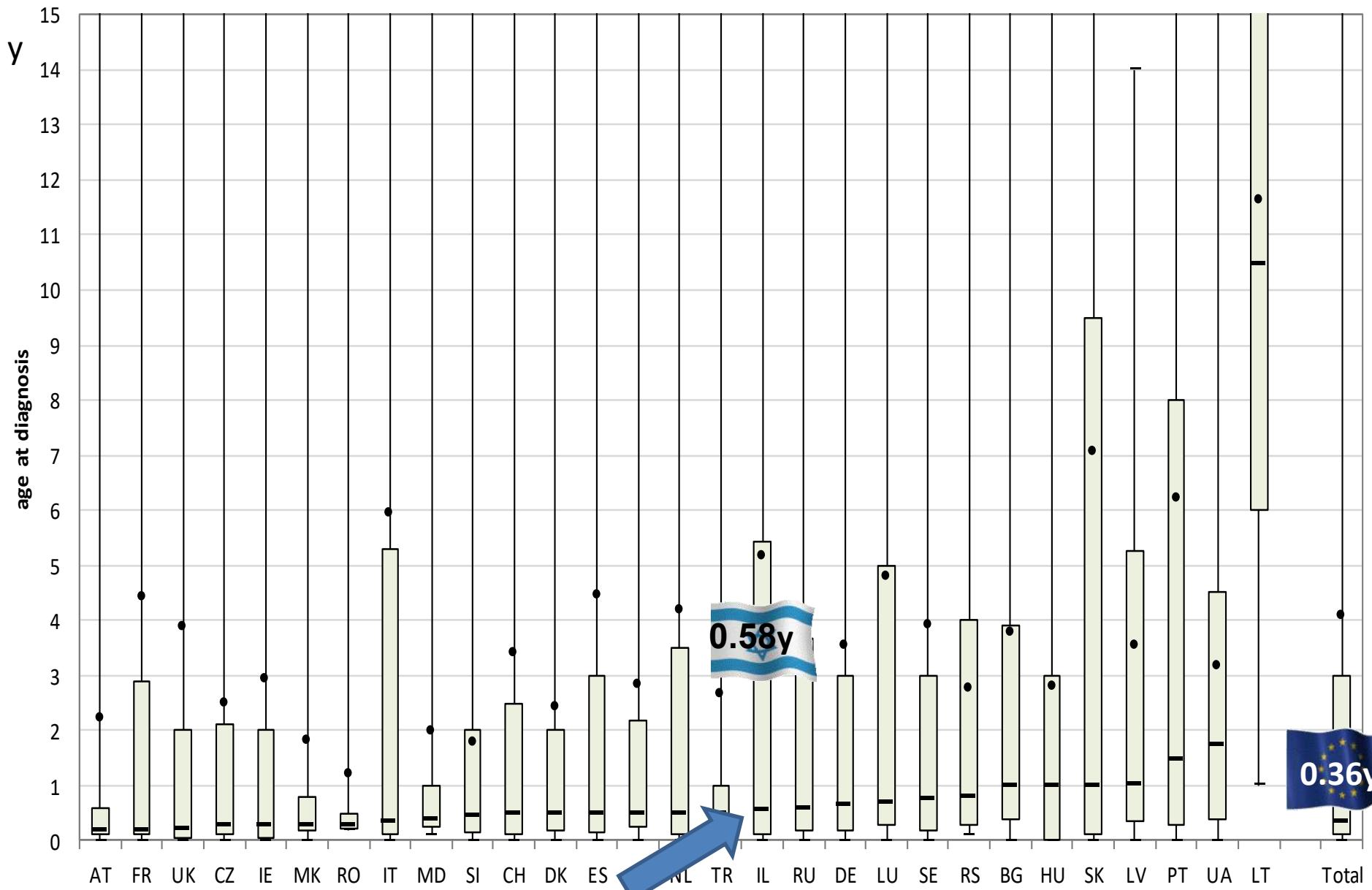
# **Diagnosis**

# Neonatal Screen

■ Performed    □ Missing/Unknown    ▨ Not performed



# Median Age at Diagnosis



# Genetics

- Genetic analysis performed- 547/548!!!
- 2 mutations found- 80.3% of pts
- 74 mutations in the Israeli registry

# Common Mutations in Israel

mutation	Frequency	Percent
F508del	334	24.1
W1282X	297	21.5
unknown	213	15.3
D1152H	68	4.9
G542X	70	5.1
N1303K	46	3.3
3849+10kbC->T	40	2.9
G85E	26	1.9
405+1G->A	19	1.4
Q359K/T360K	19	1.4
I1234V	13	0.9
S549R(A->C)	10	0.7

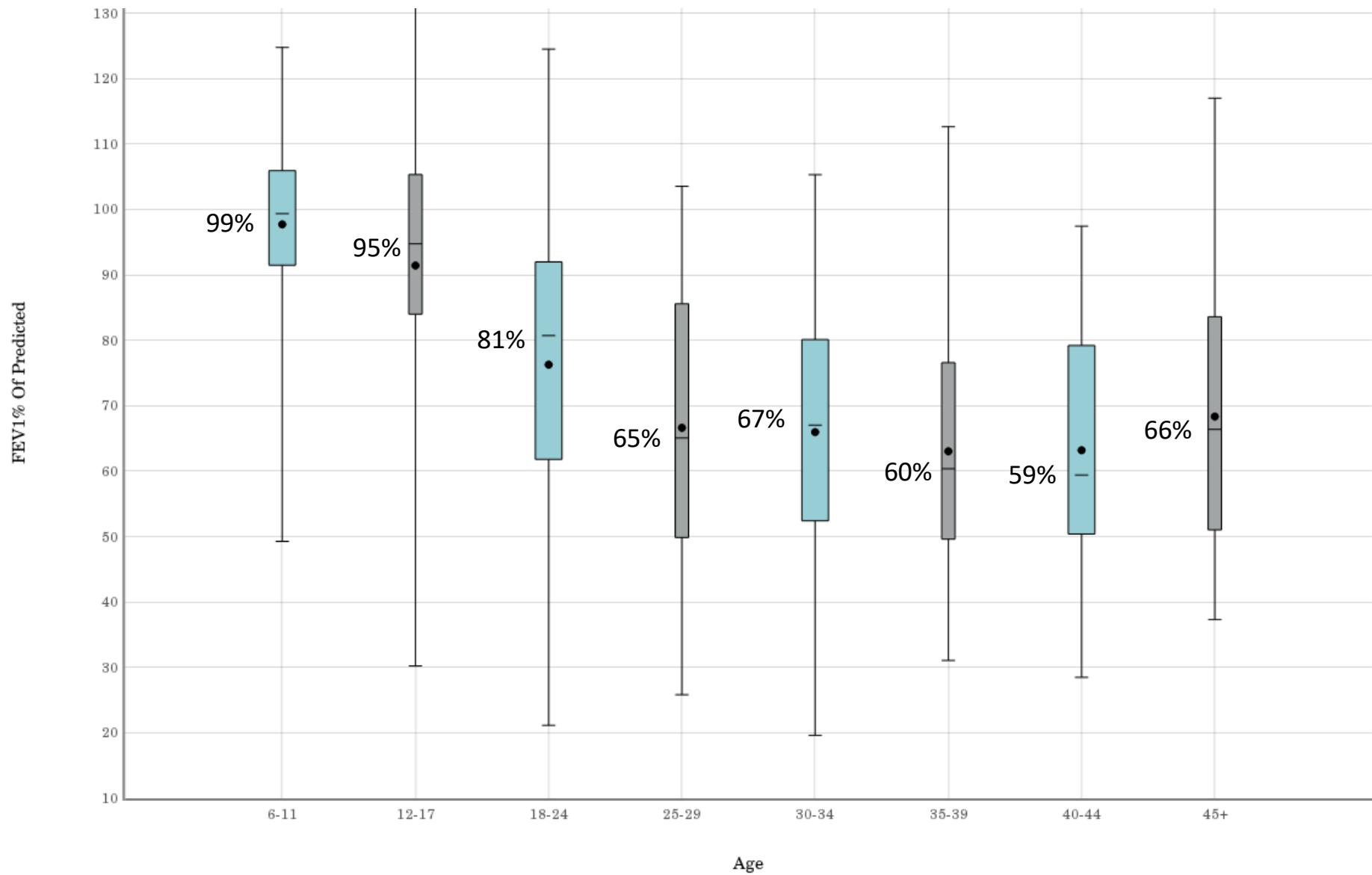
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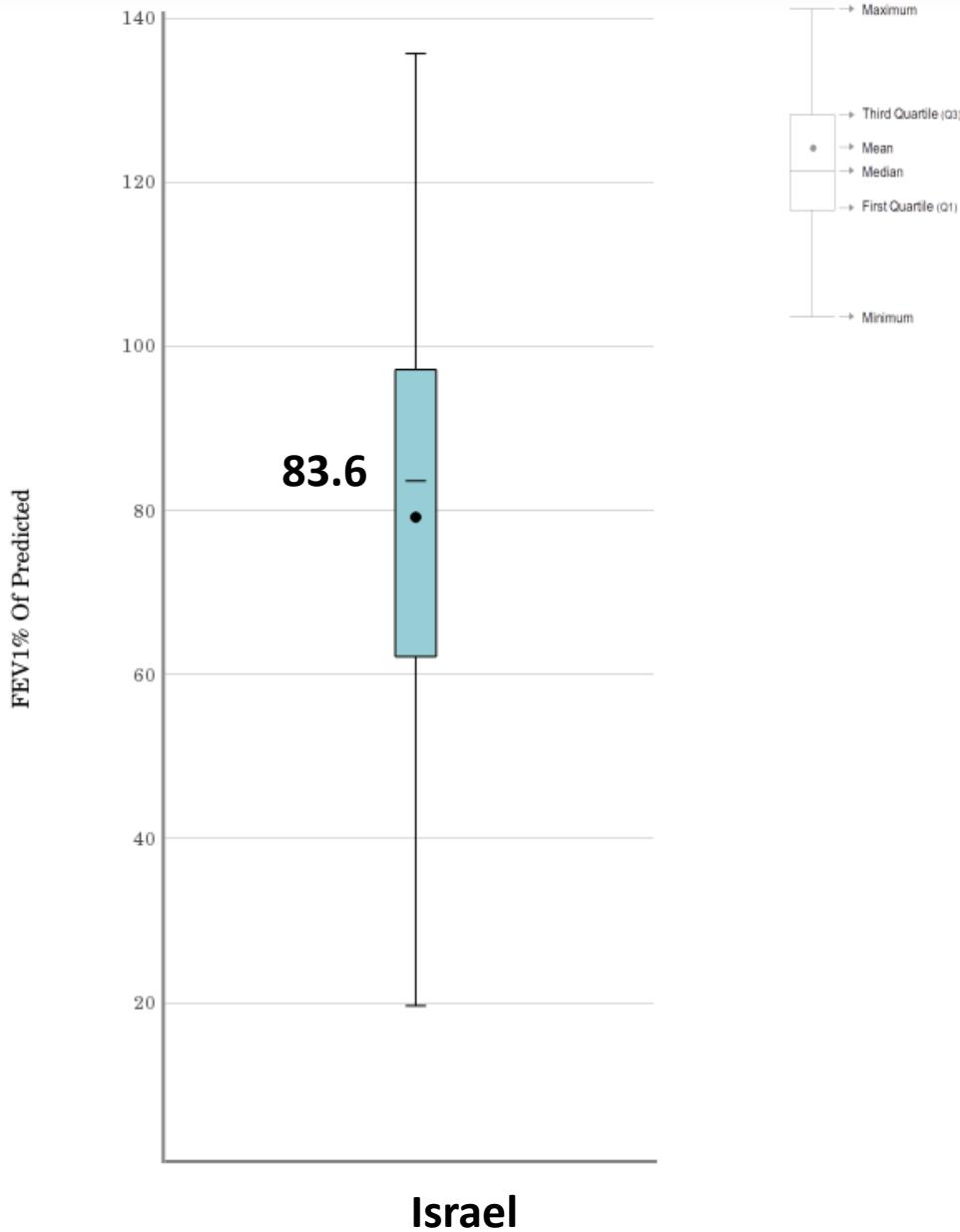
Mutation name	Number of alleles	Percentage among tested	Country with highest allele frequency
F508del	48801	61.51	Denmark (82.01%)
G542X	2098	2.64	Spain (5.96%)
N1303K	1652	2.08	Turkey (7.14%)
G551D	1124	1.42	Ireland (8.77%)
W1282X	891	1.12	Israel (22.95%)
R117H	805	1.01	Ireland (2.64%)
2789+5G->A	752	0.95	Turkey (6.59%)
1717-1G->A	691	0.87	Switzerland (3.02%)
R553X	678	0.85	Lithuania (7.14%)
3849+10kbC->T	677	0.85	Lithuania (17.86%)
CFTRdel2,3	631	0.8	Czech Republic (6.40%)
621+1G->T	526	0.66	Greece (5.79%)
2183AA->G	458	0.58	Turkey (4.40%)
R1162X	426	0.54	Slovenia (4.35%)
D1152H	424	0.53	Israel (5.10%)
R347P	403	0.51	Luxembourg (3.85%)

# **Clinical - Pulmonary**

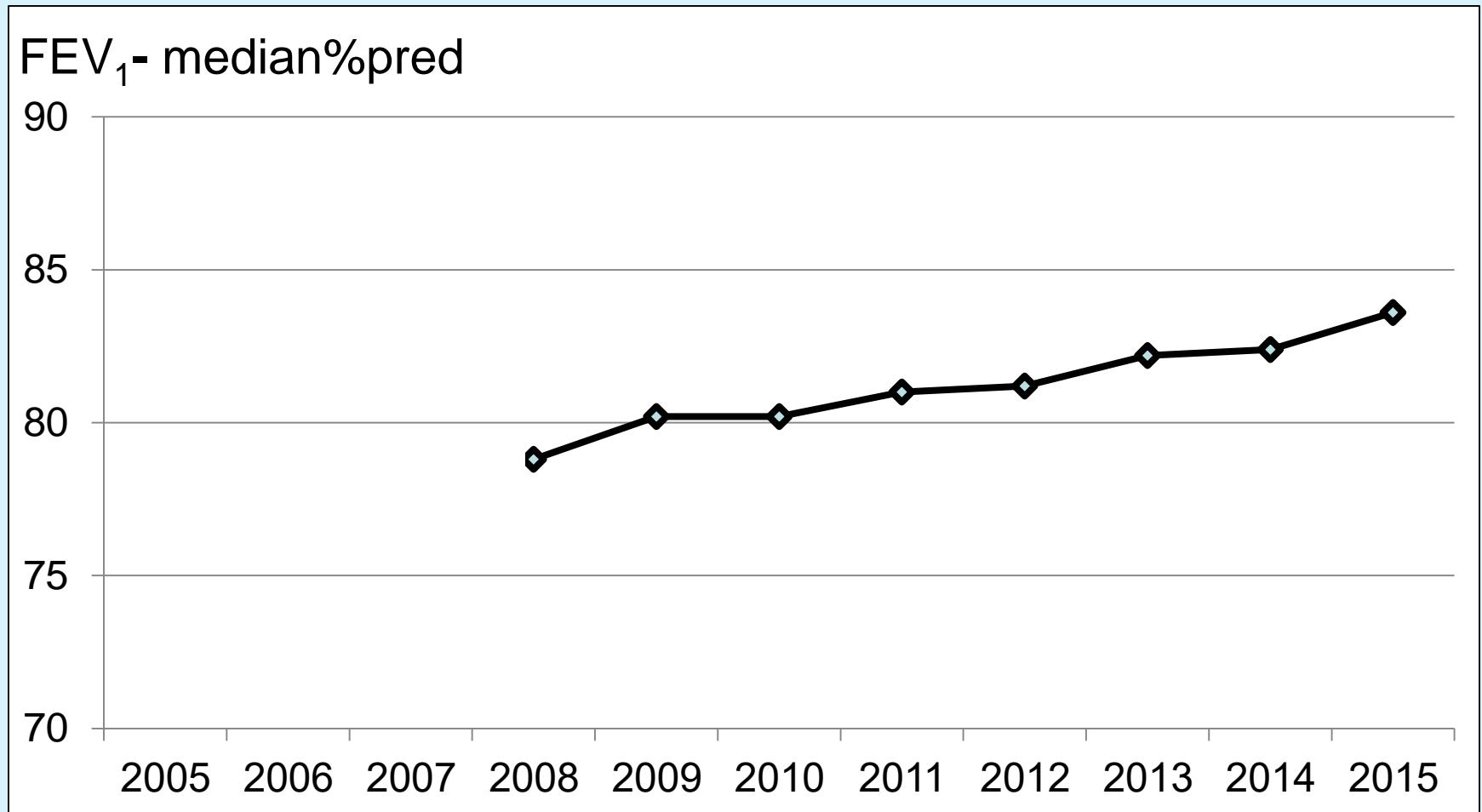
# FEV<sub>1</sub> with Age- Israel

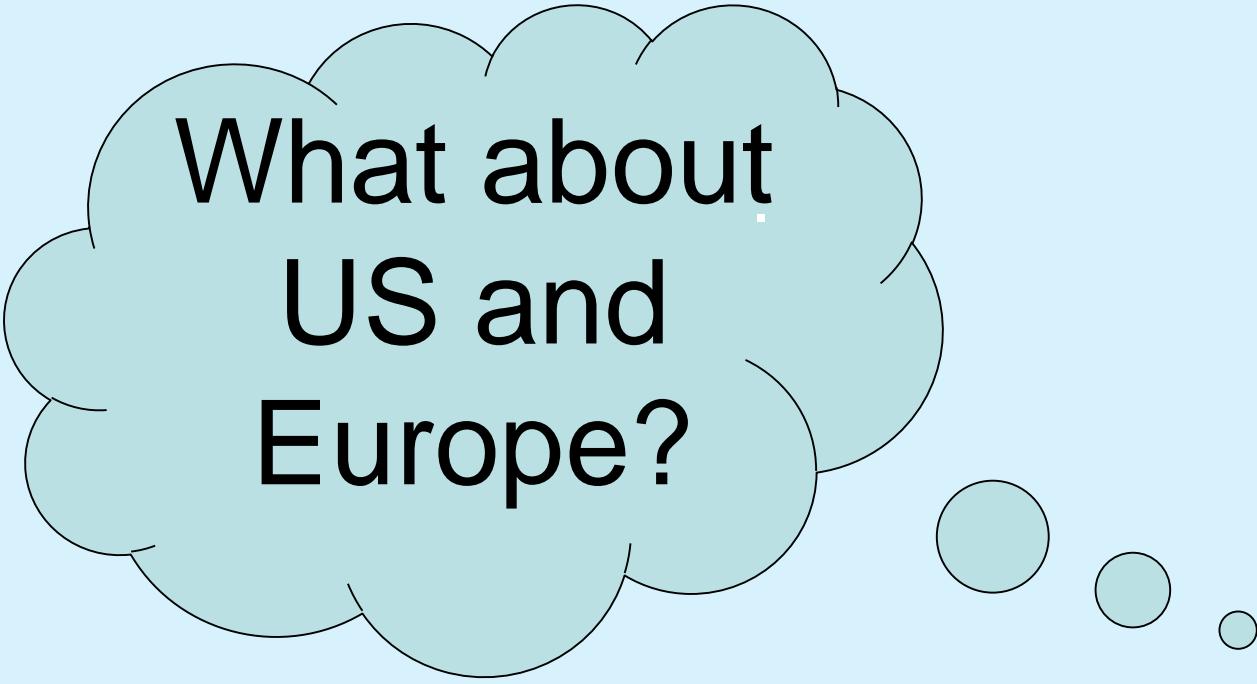


# FEV<sub>1</sub> median- Israel



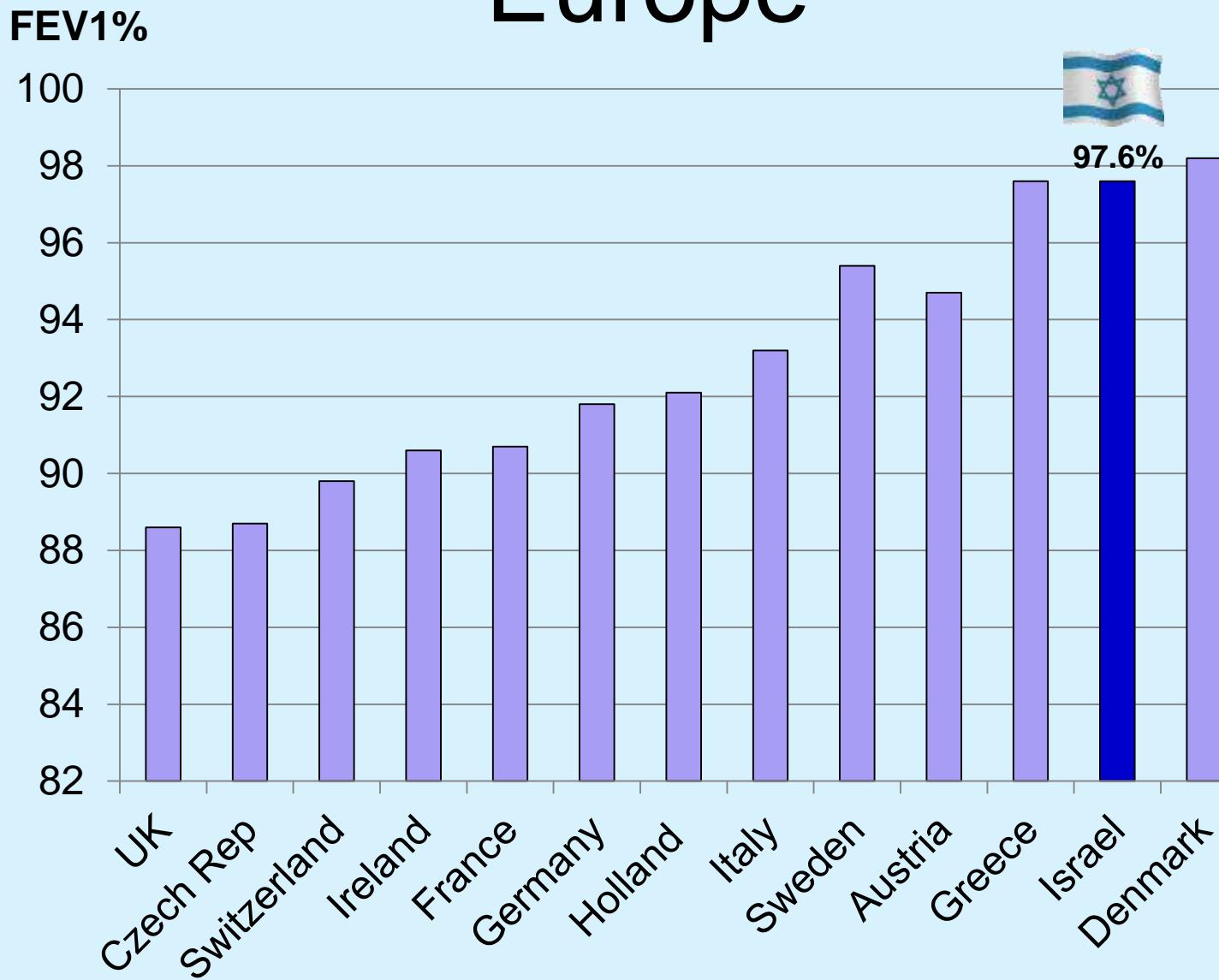
# $\text{FEV}_1(\text{median})$ - Trend – Israel



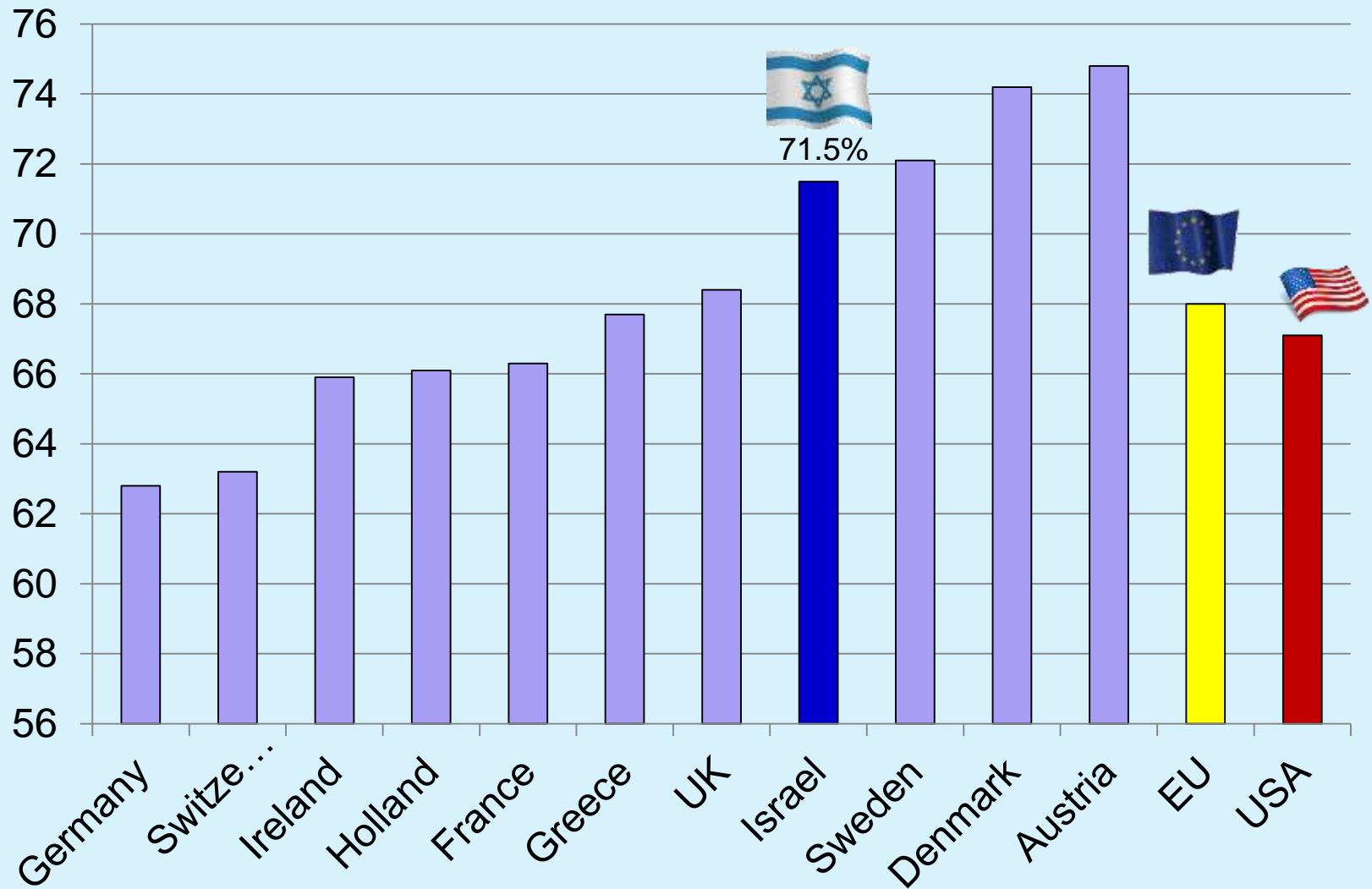


**What about  
US and  
Europe?**

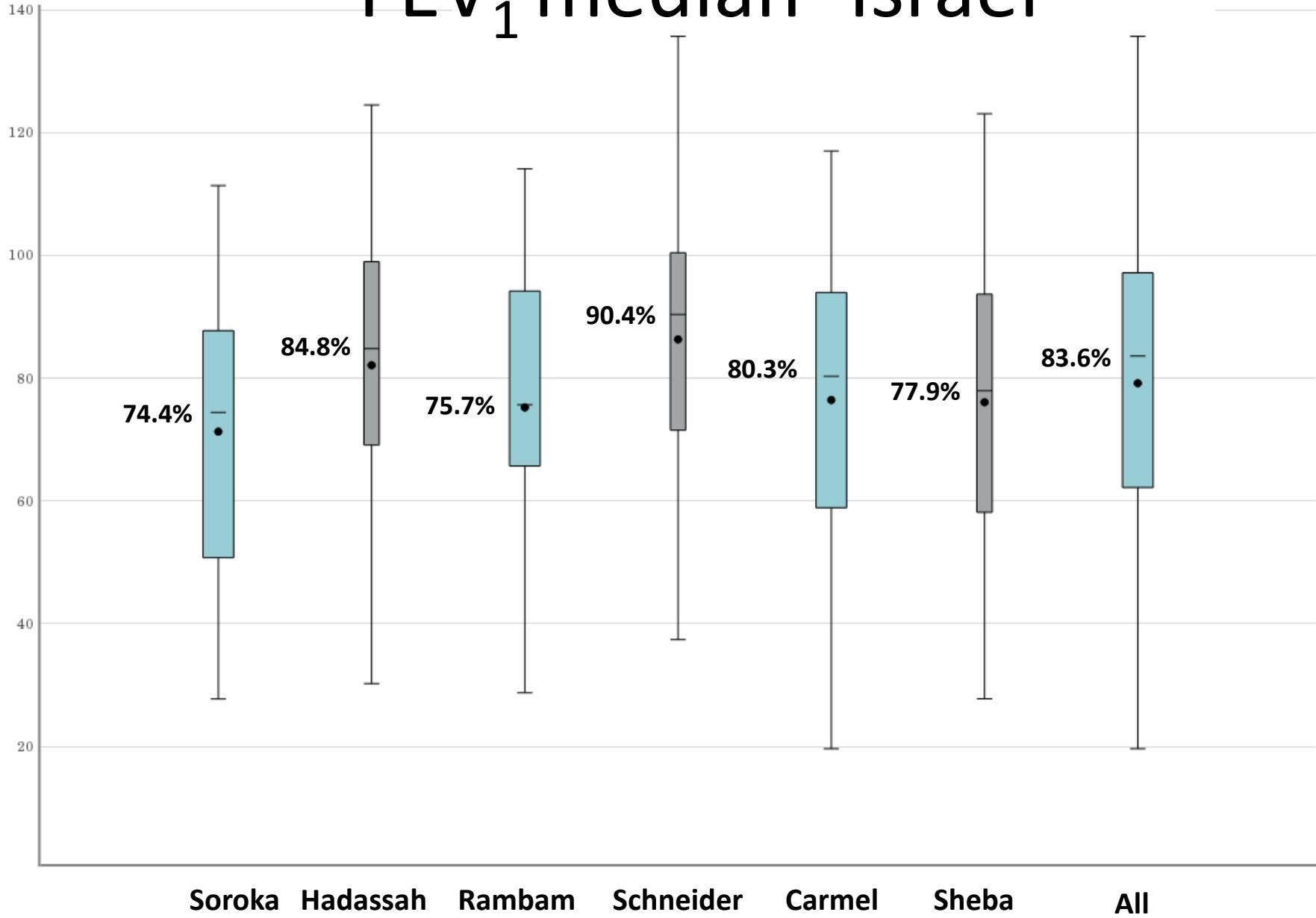
# FEV<sub>1</sub>(median) Children Europe



# FEV<sub>1</sub>(median) Adults Europe



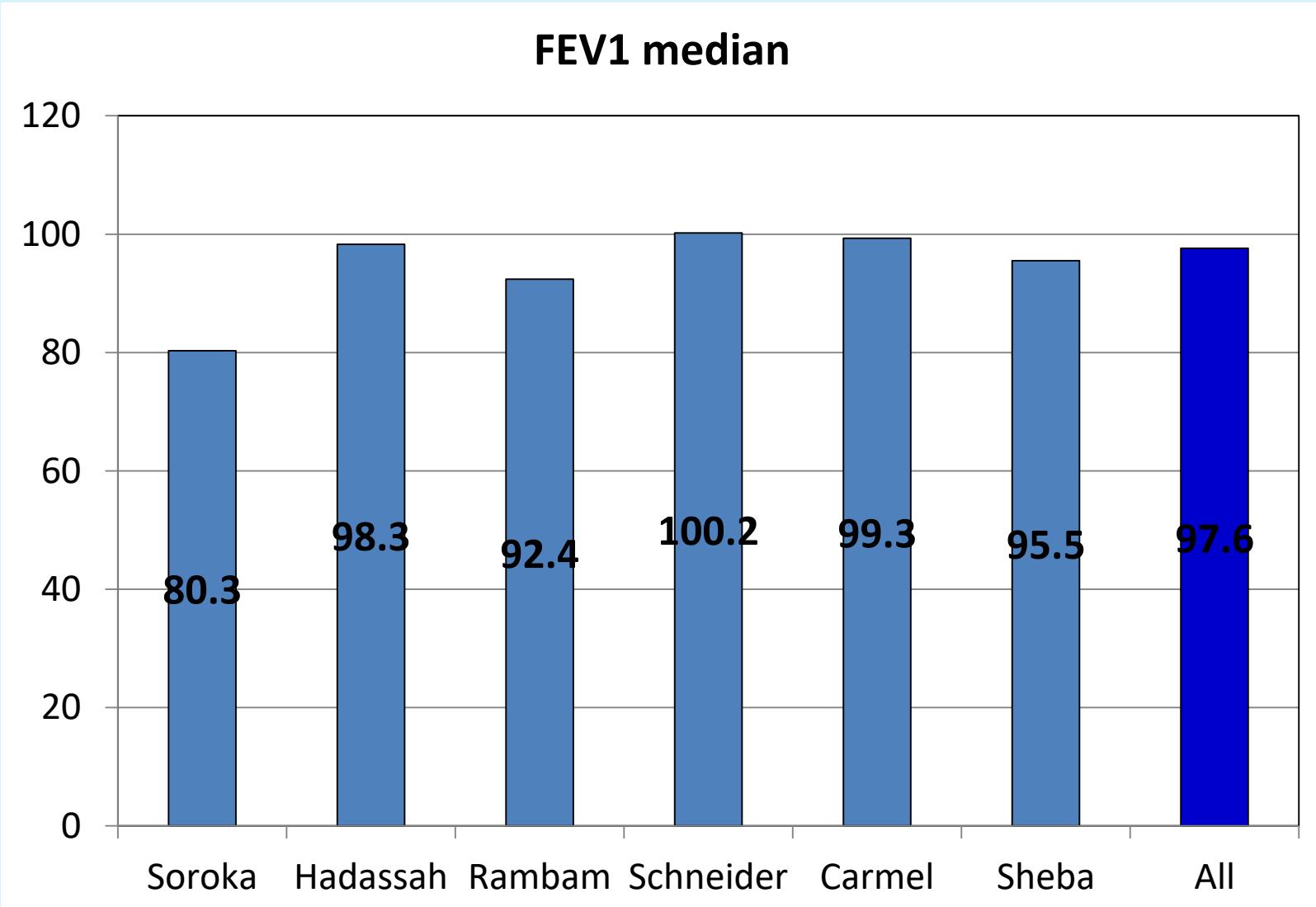
# FEV<sub>1</sub> median- Israel



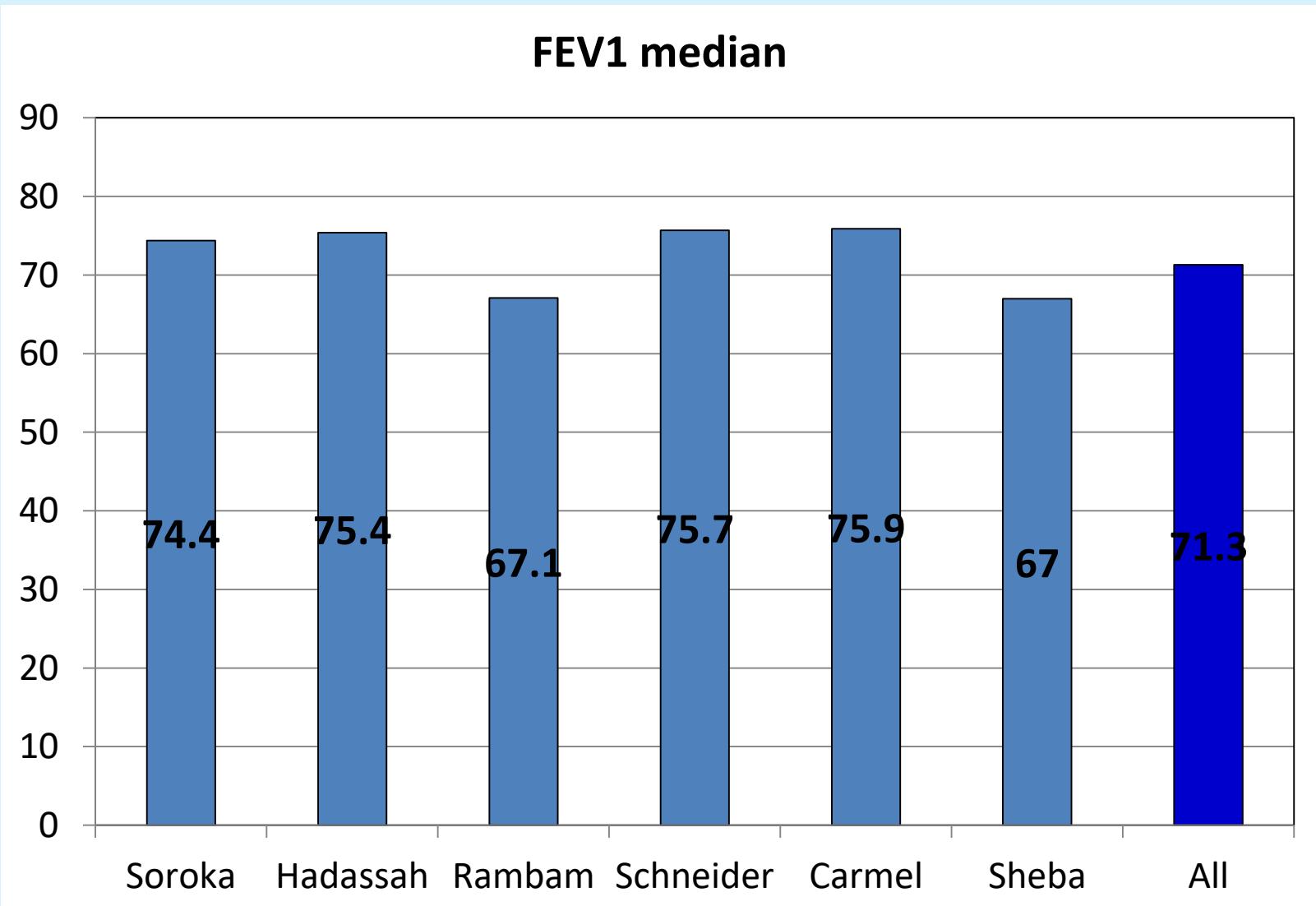
# FEV<sub>1</sub> median- Israel

Children / Adults

# $\text{FEV}_1$ - median- Children

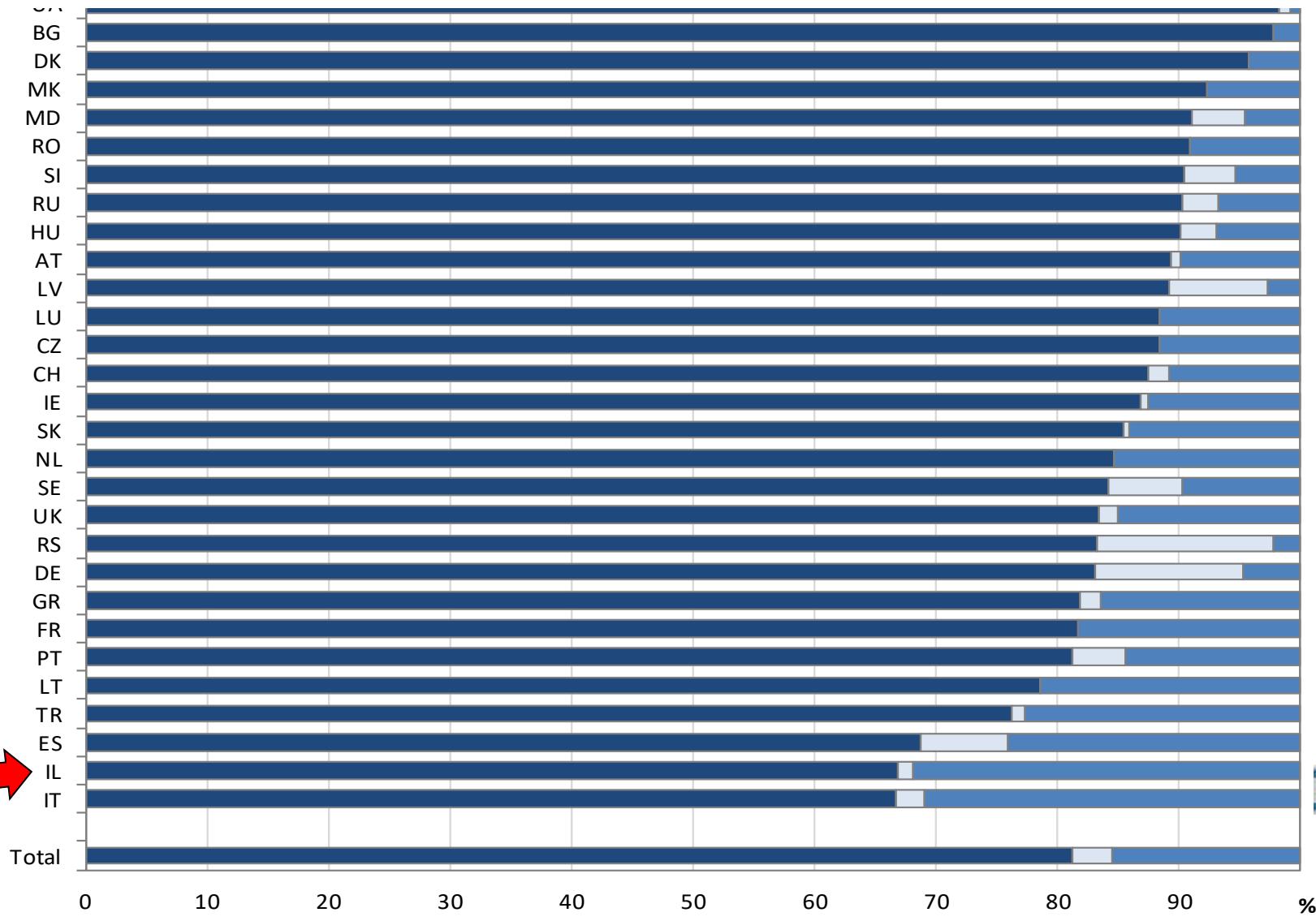


# FEV<sub>1</sub>- median- Adults



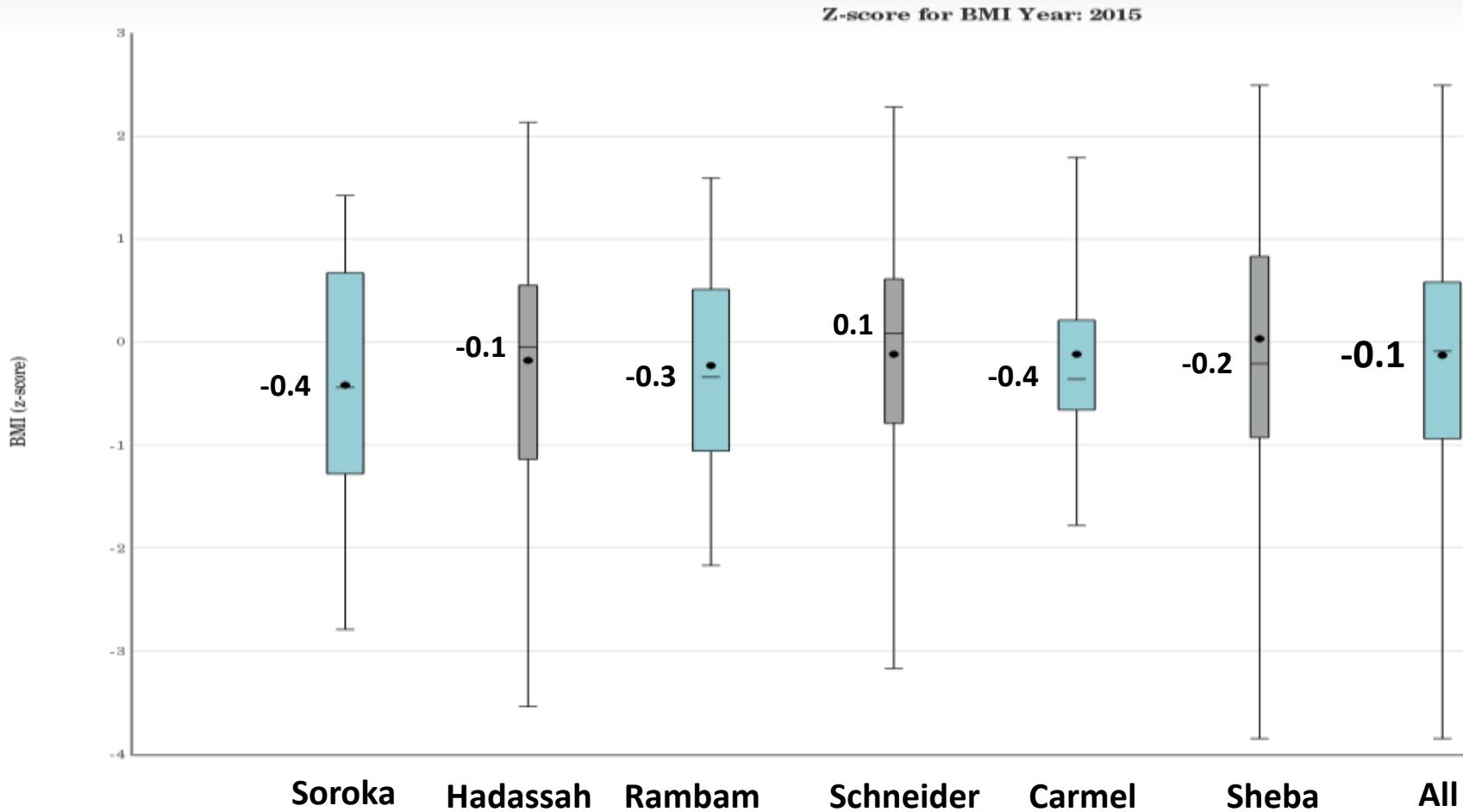
# **Clinical - Nutrition**

# Use of Pancreatic Enzymes





# BMI- Z score – children

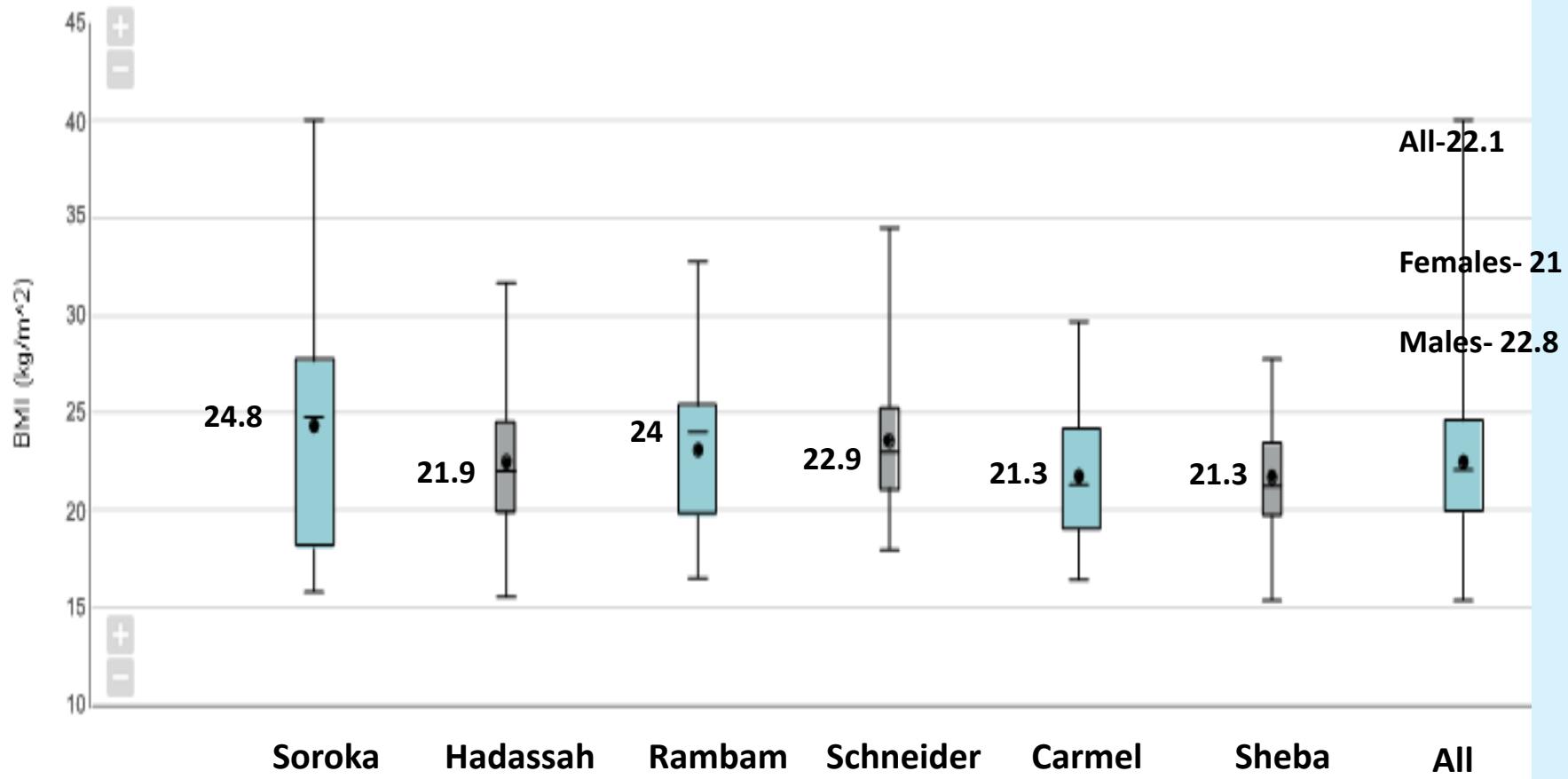




# BMI- (M+F) Adults (2014)

Graph

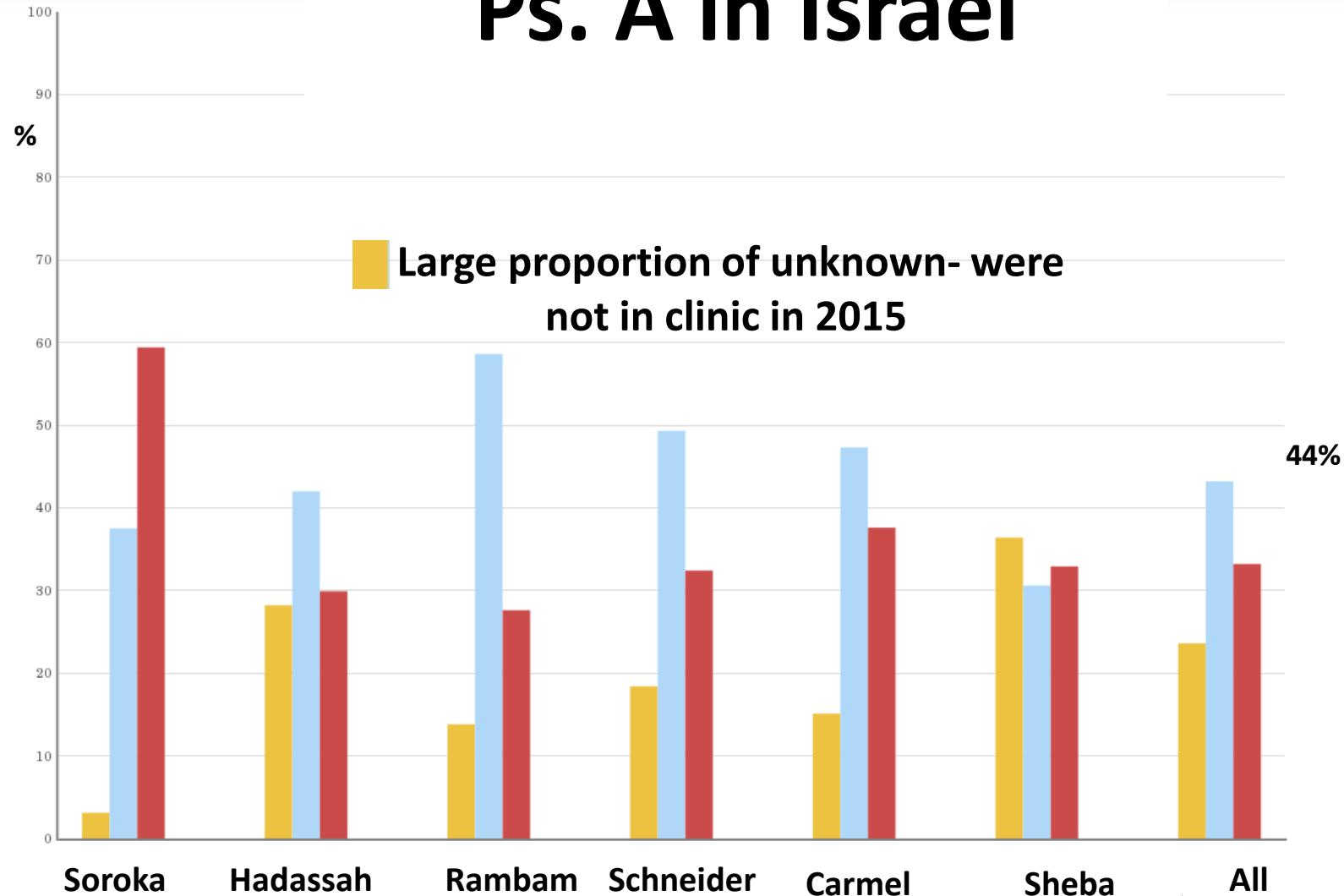
Table



# Bacteriology

# Ps. A in Israel

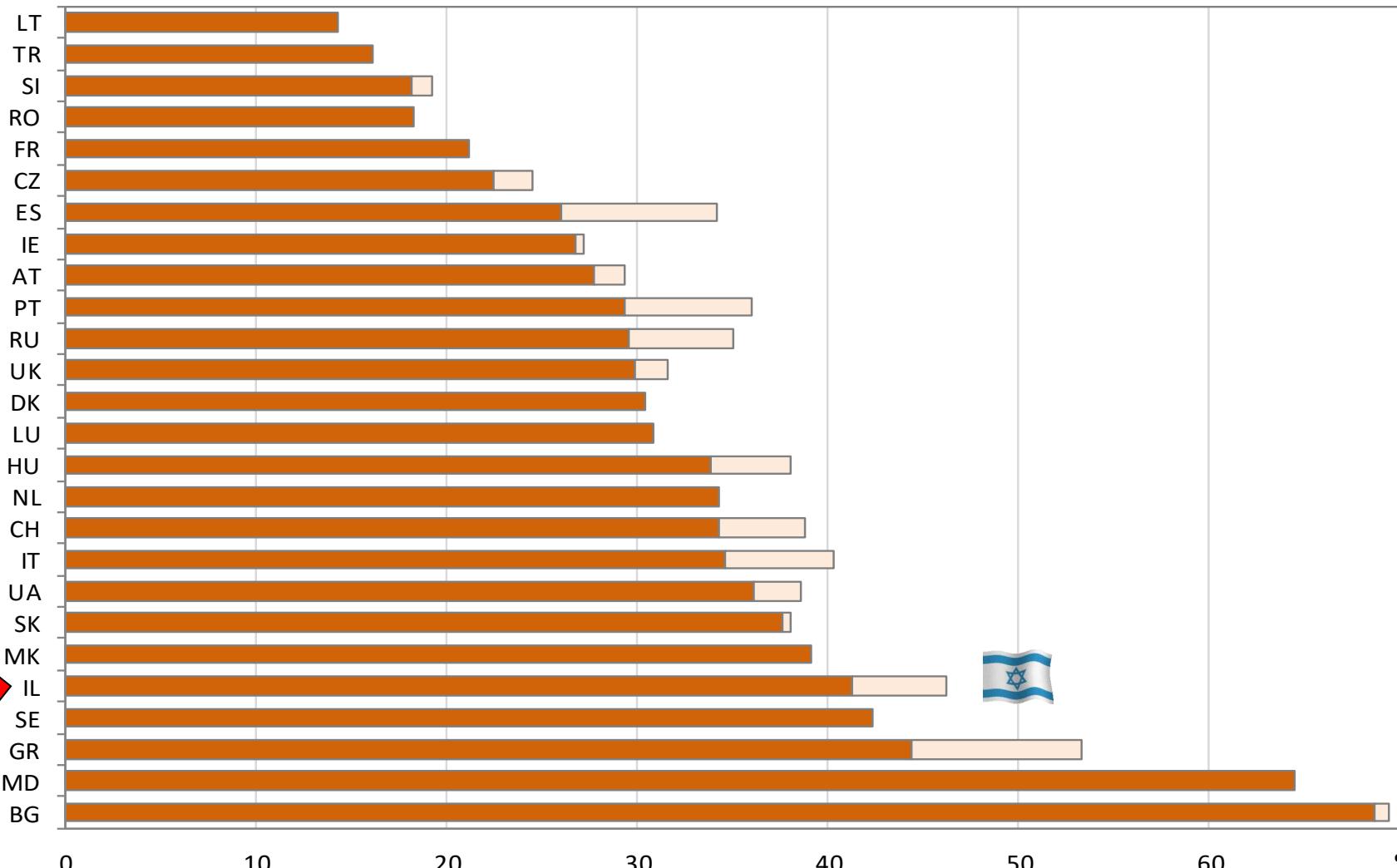
Unknown  
No  
Yes



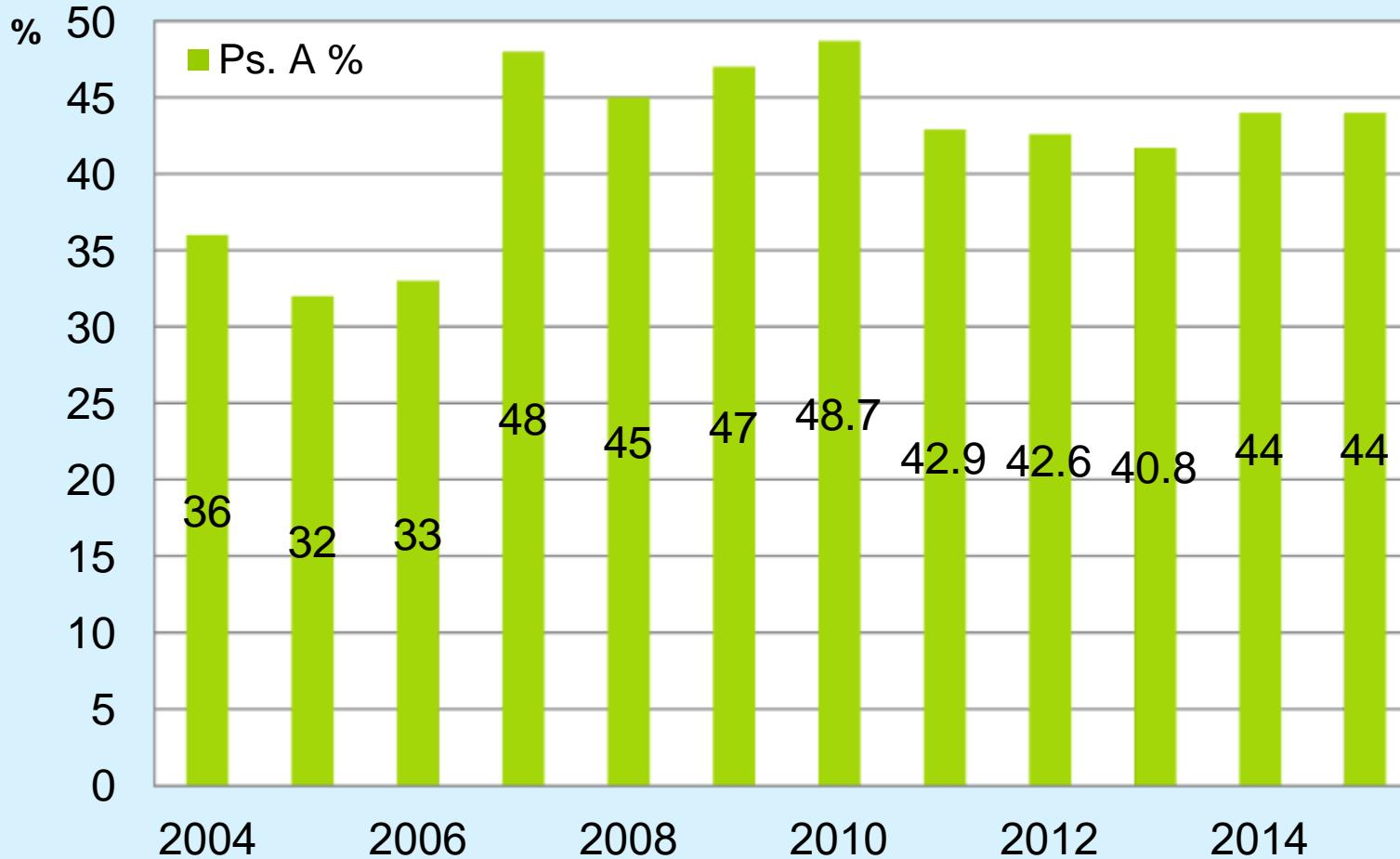


# Ps. A in Europe

■ Yes □ Missing/Unknown

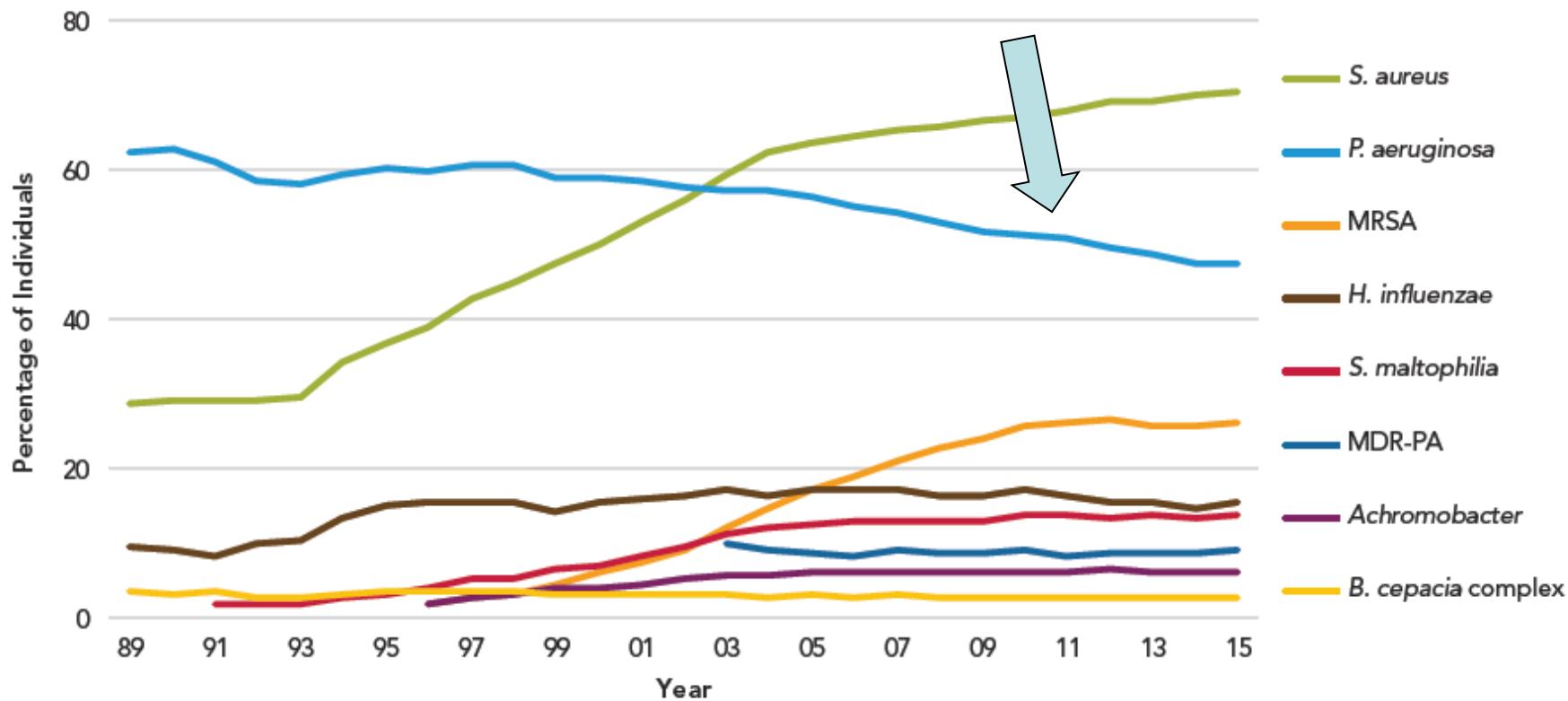


# Ps. A in Israel- Trend

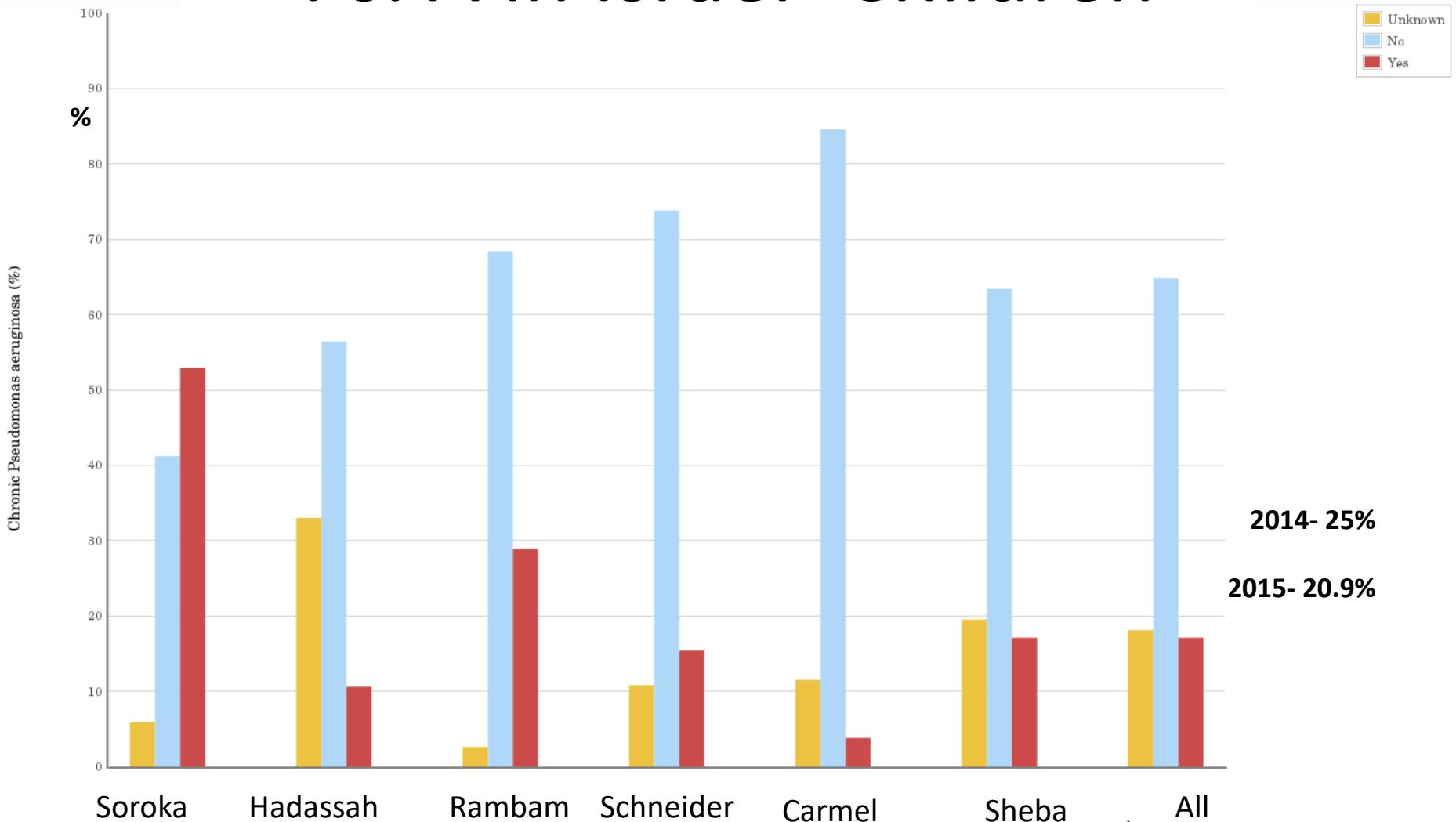


# Ps. A in the US- Trend

Prevalence of Respiratory Microorganisms, 1989–2015



# Ps. A in Israel- Children



Report generated by:

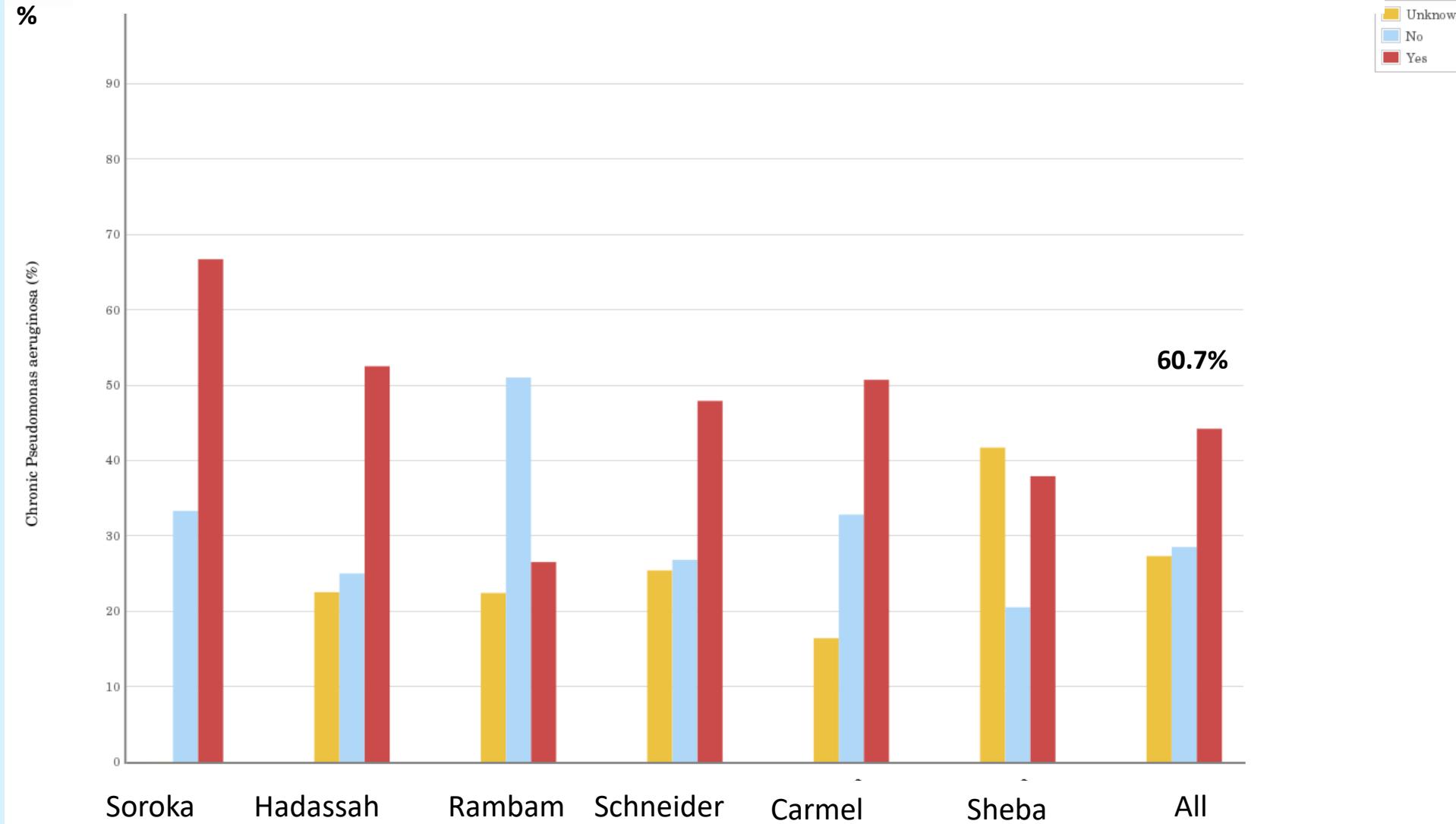
**Meir Mei-Zahav**

Report date:

**Thu, 19 Oct 2017 21:27:12 GMT**

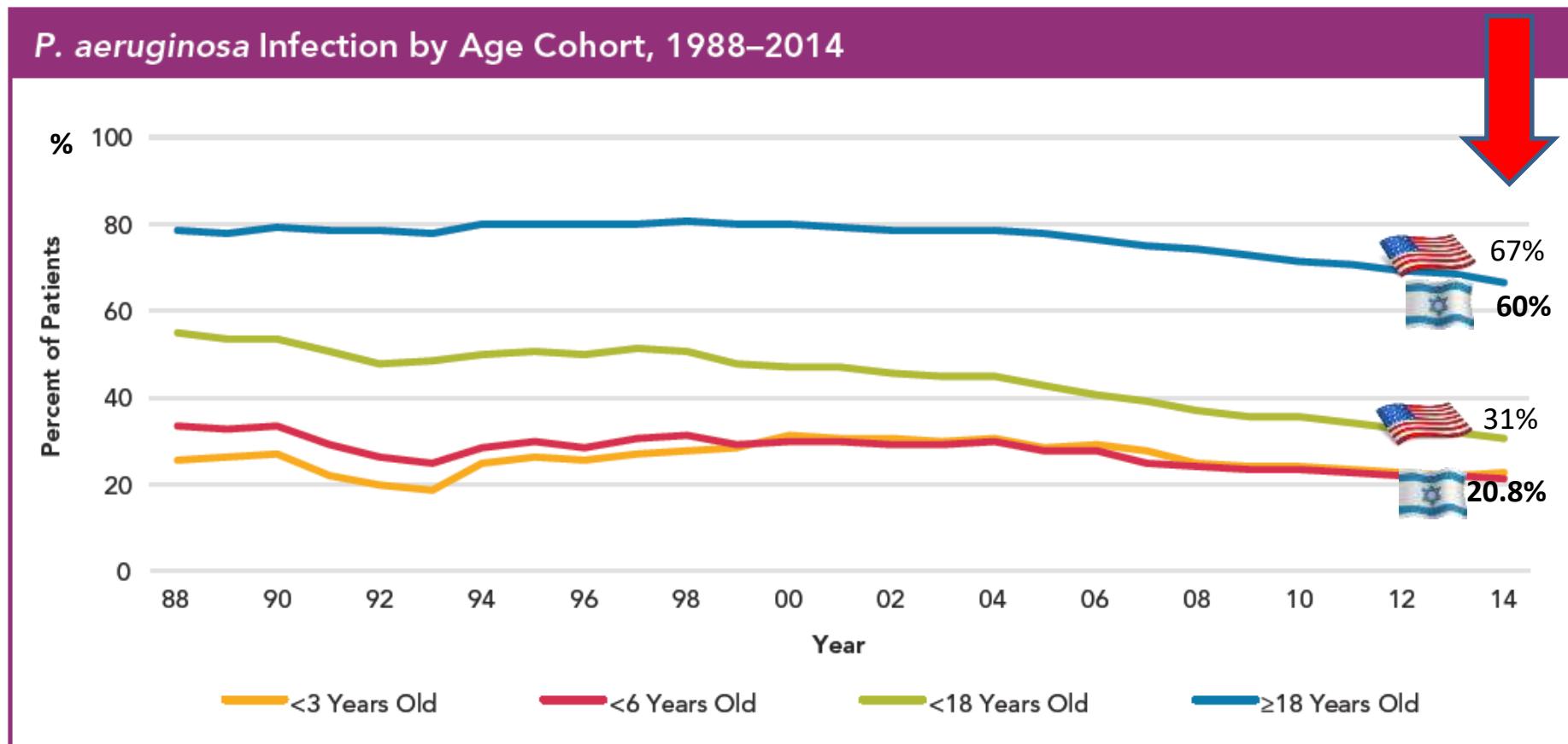
ECFSPR validation status: **No**

# Ps. A in Israel- Adults

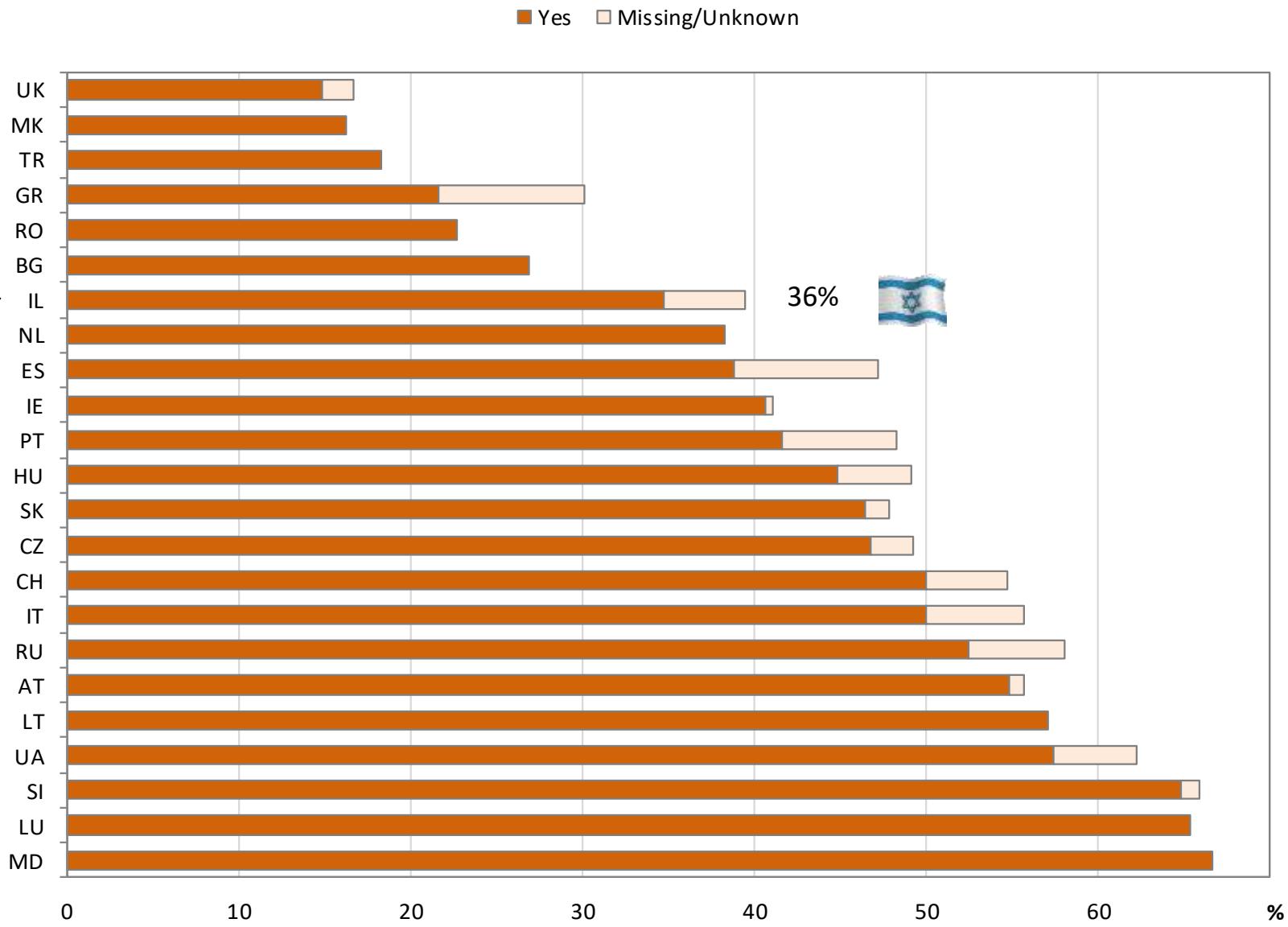


# Ps. A in the US

2014

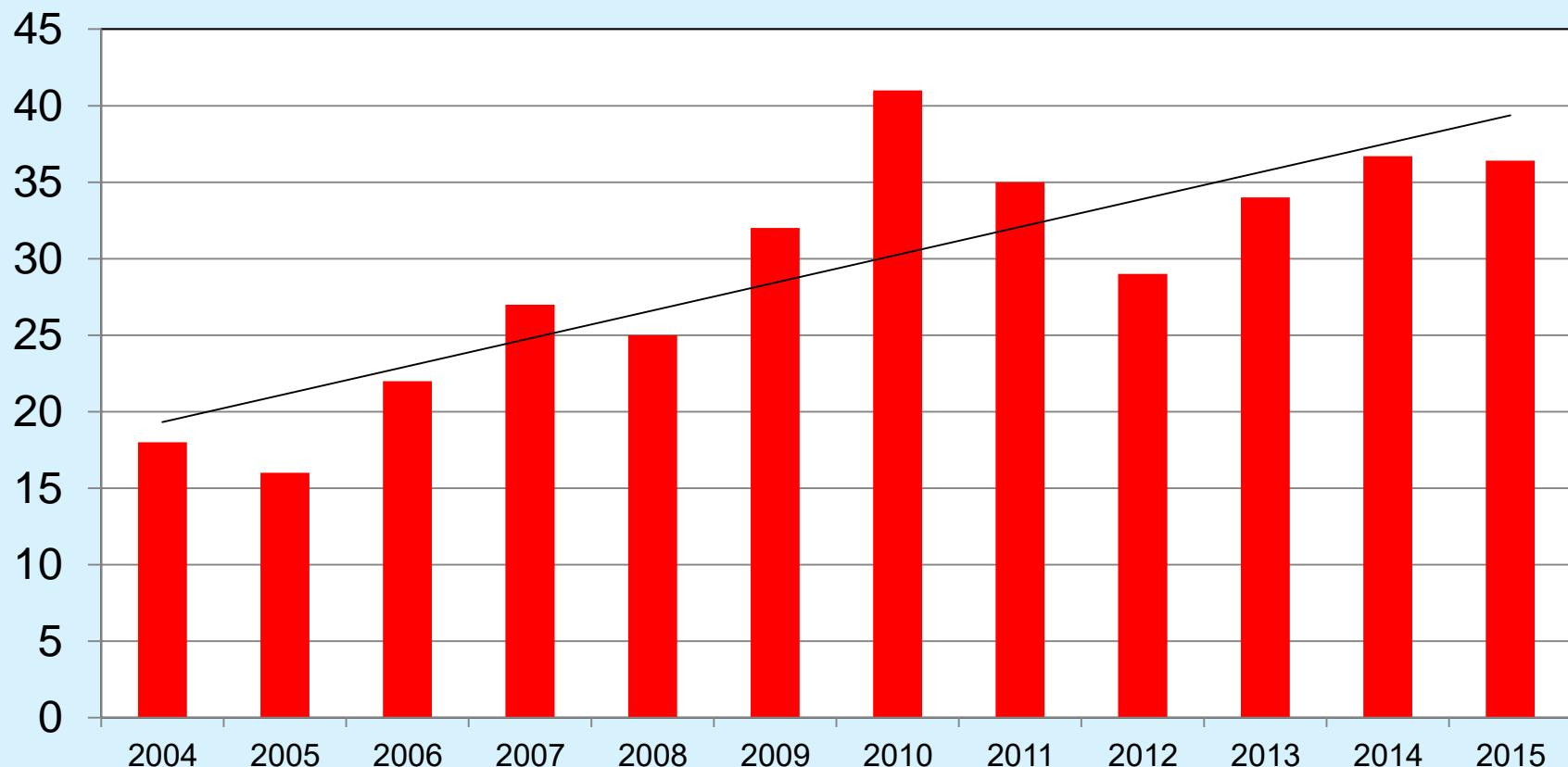


# Staph A. in Europe

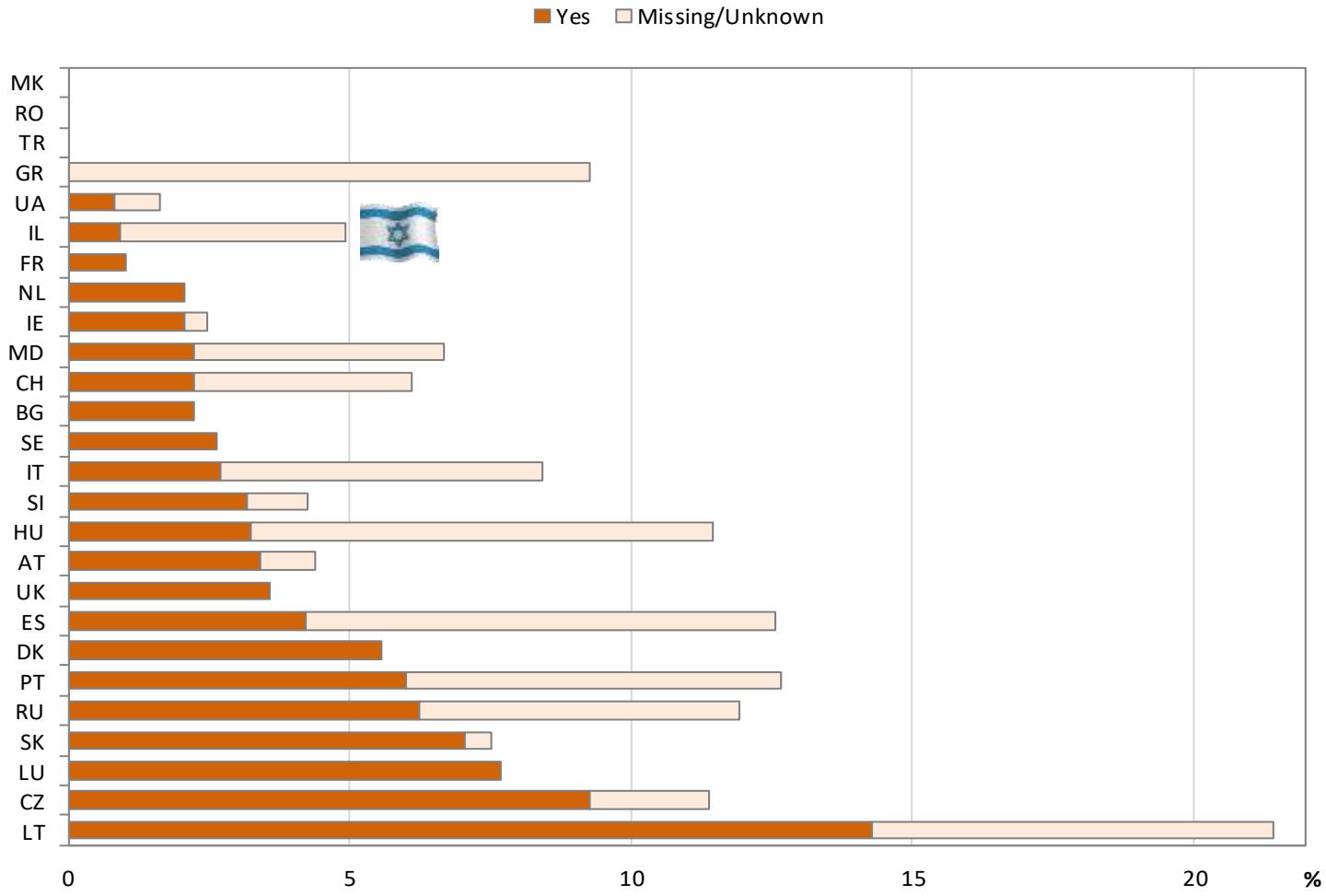


# Staph. in Israel- Trend

Staph %



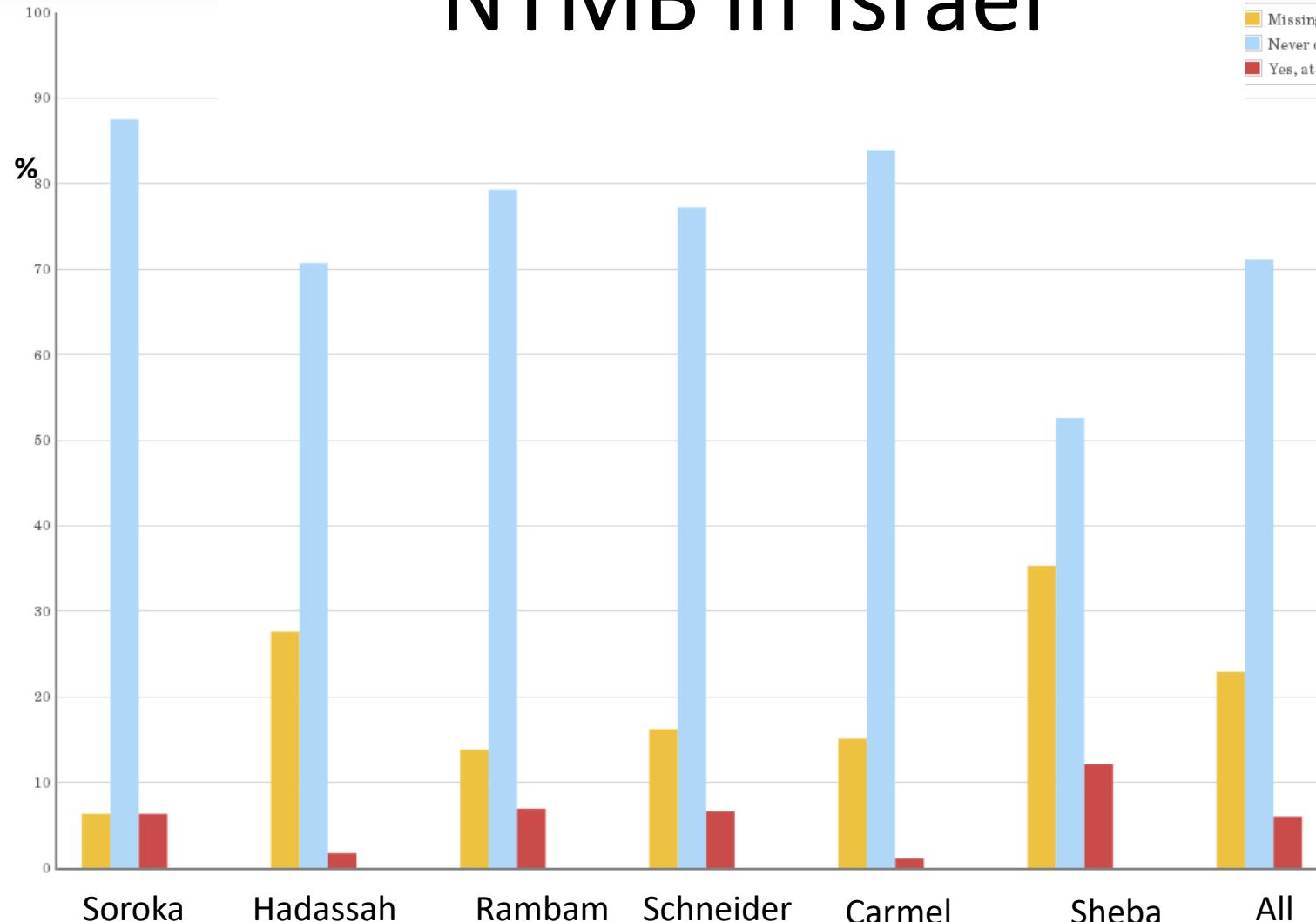
# B. Cepacia in Europe



# NTMB in Israel

Missing/Unknown  
Never during the year of follow-up  
Yes, at least once during the year of follow-up

Nontuberculous mycobacteria (%)



Report generated by: Meir Mei-Zahav

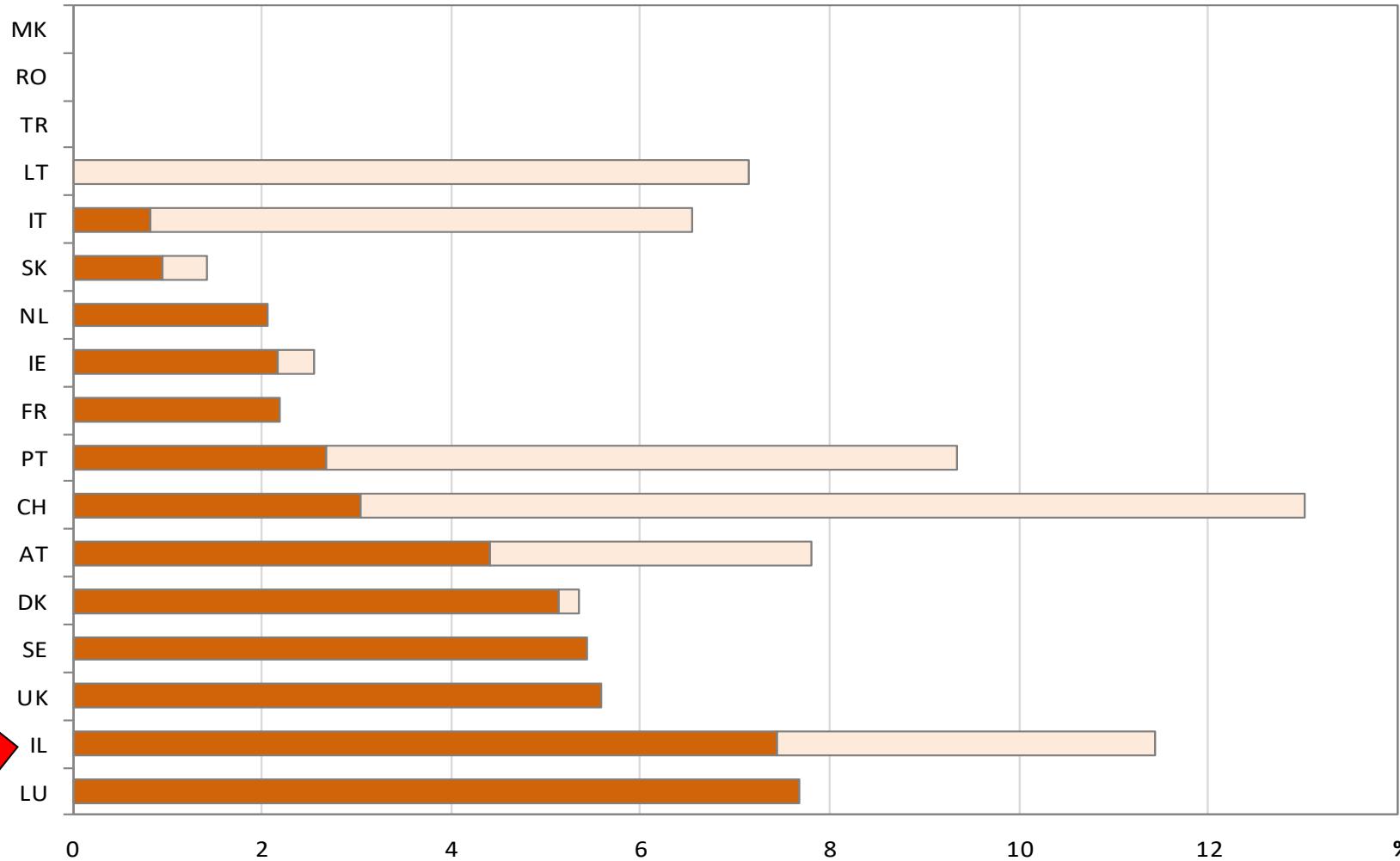
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ECFSPR validation status: No



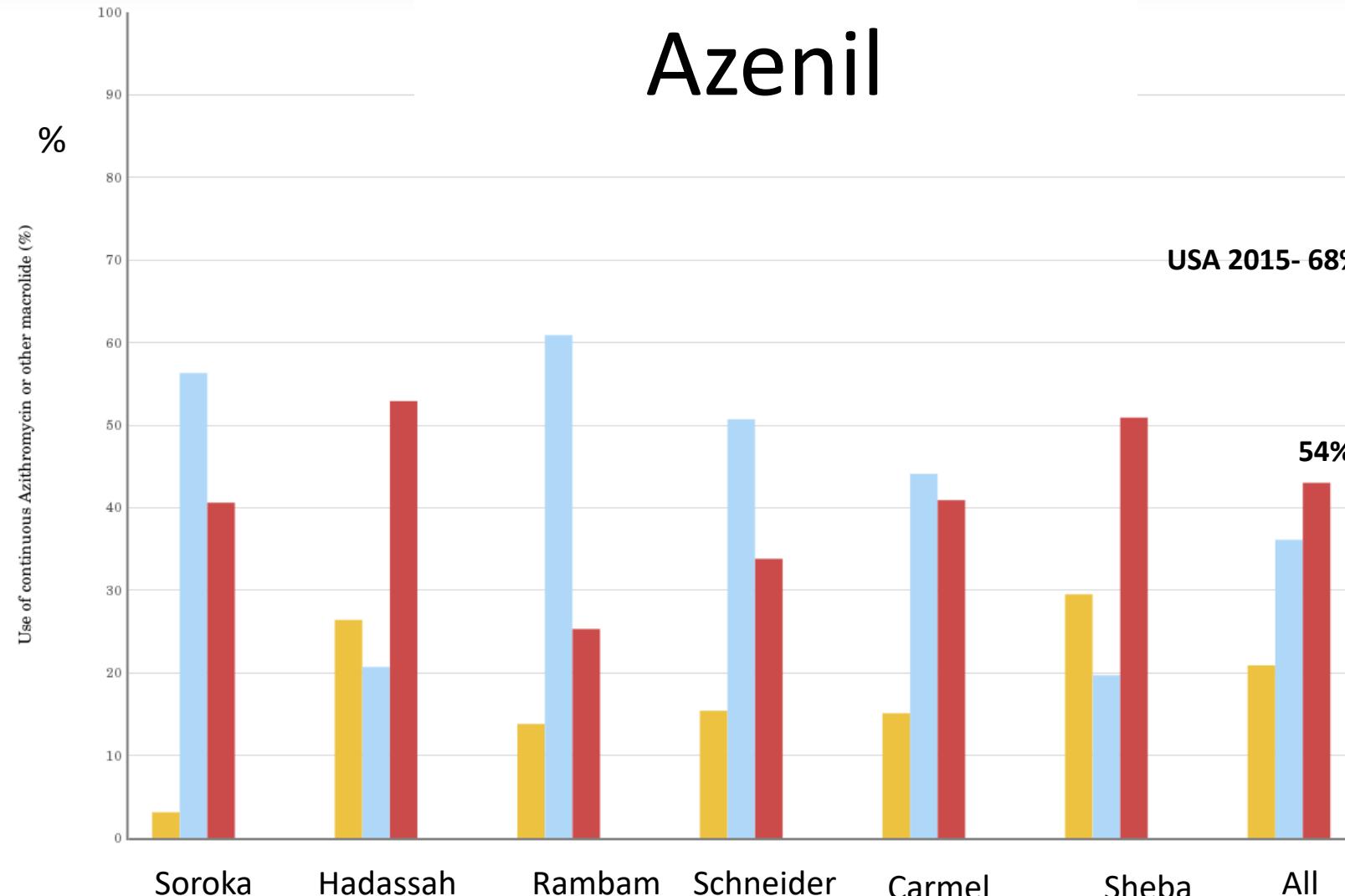
# NTM in Europe

■ Yes ■ Missing/Unknown



# Treatment

# Azenil



USA 2015- 68%



54%

Report generated by:

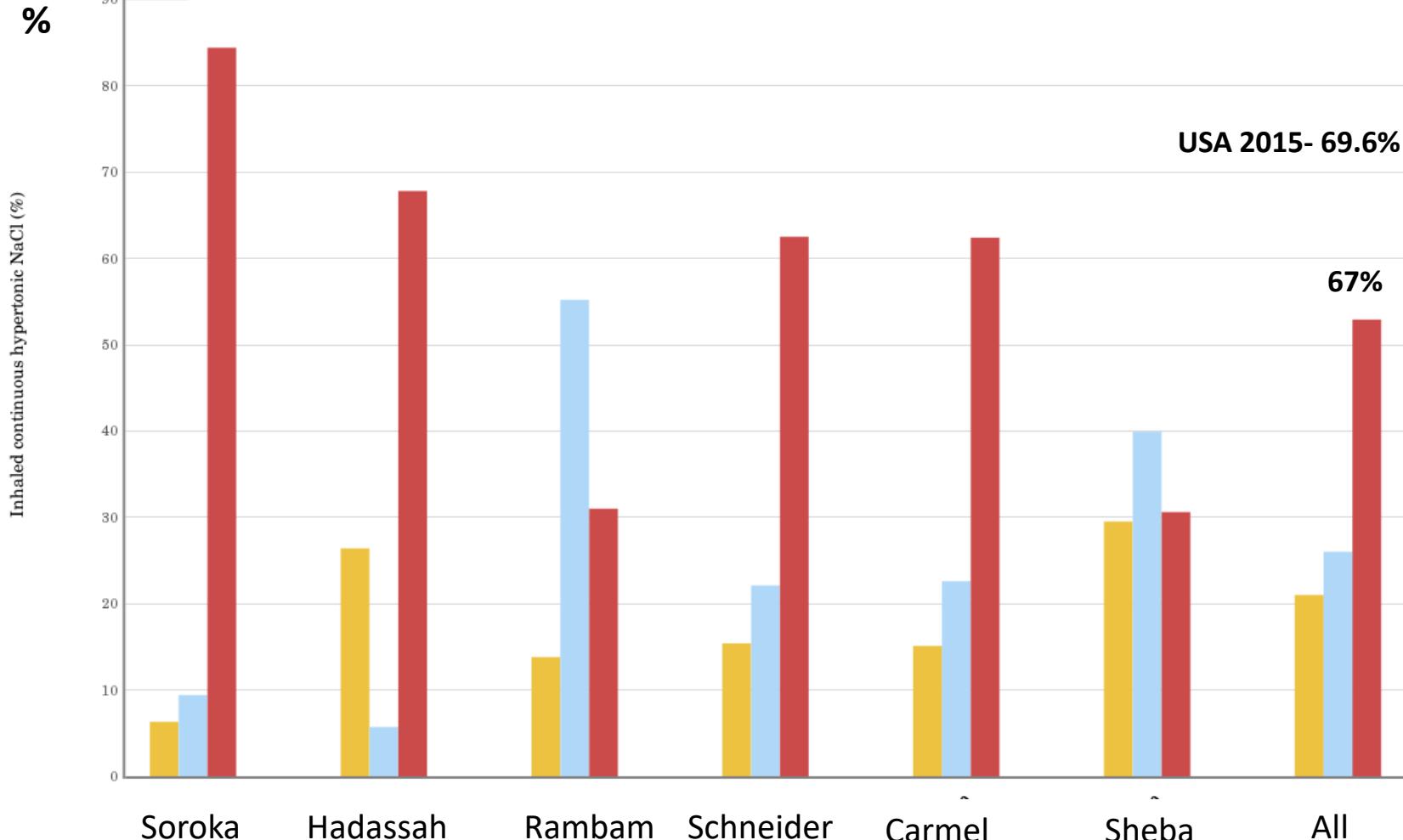
**Meir Mei-Zahav**

Report date:

**Thu, 19 Oct 2017 21:53:30 GMT**

ECFSRP validation status: **No**

# Hypertonic Saline



Report generated by:

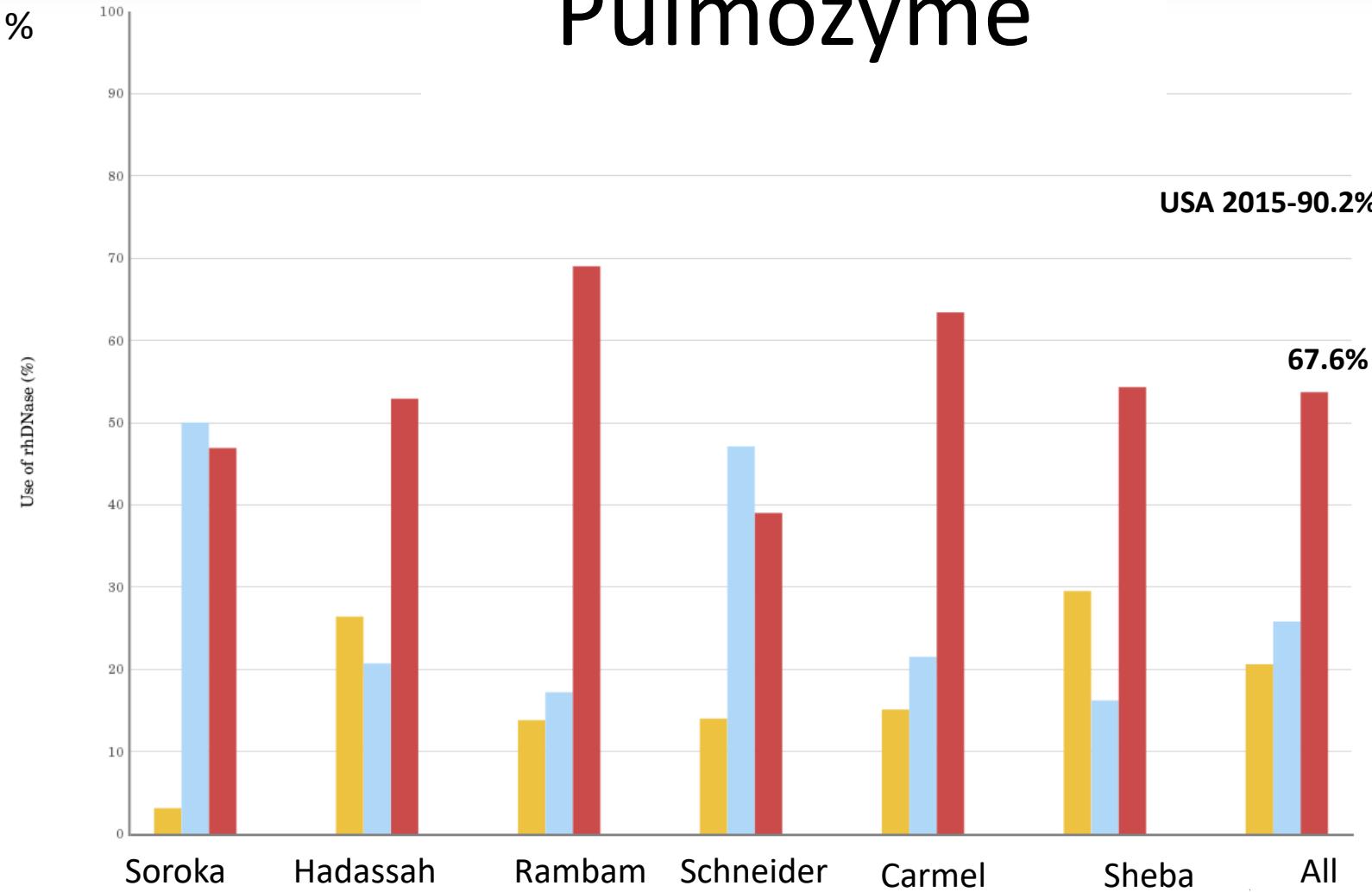
**Meir Mei-Zahav**

Report date:

**Thu, 19 Oct 2017 21:58:42 GMT**

ECFSPR validation status: **No**

# Pulmozyme



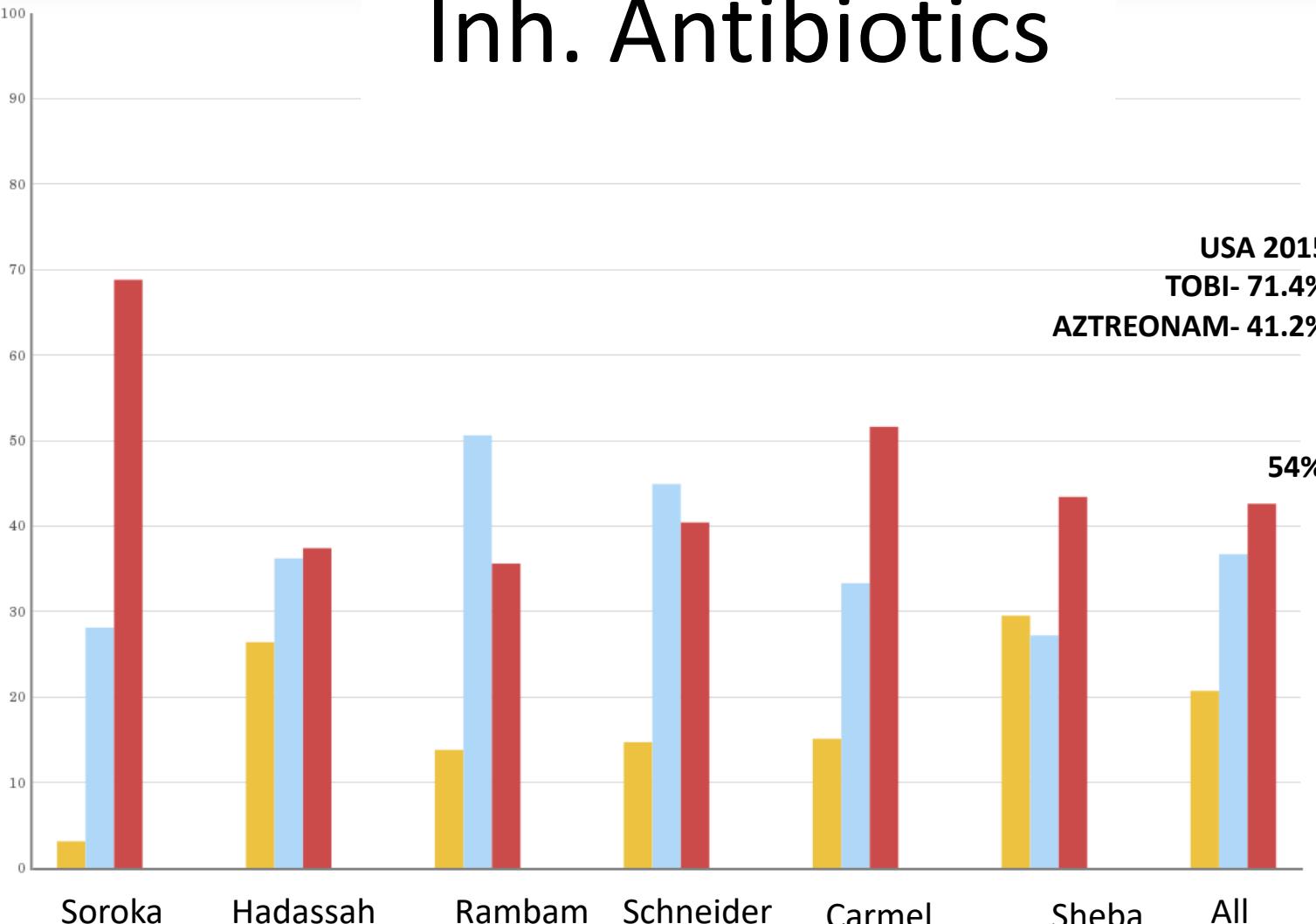
# Inh. Antibiotics

Unknown  
No  
Yes

USA 2015  
TOBI- 71.4%  
AZTREONAM- 41.2%



Inhaled continuous antibiotic (%)



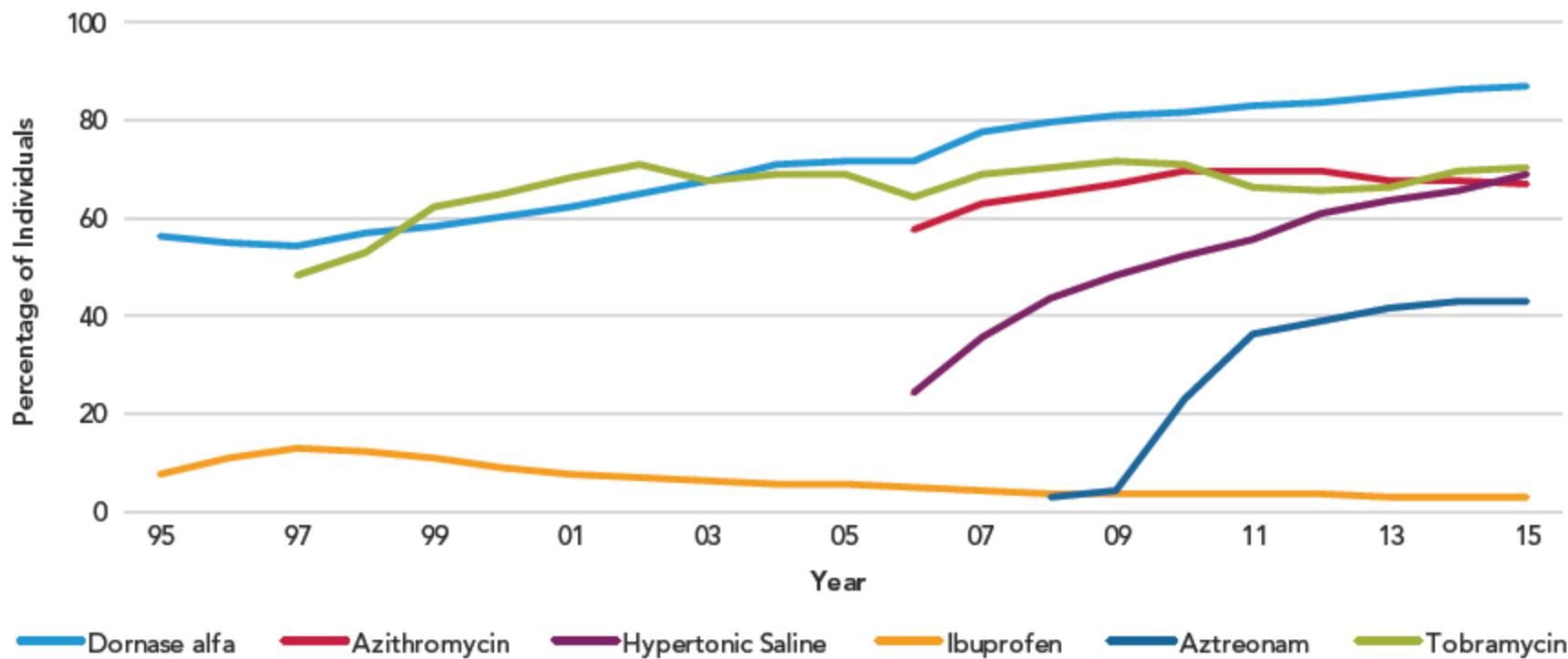
Report generated by: Meir Mei-Zahav

Report date: Thu, 19 Oct 2017 22:04:44 GMT

ECFSRP validation status: No

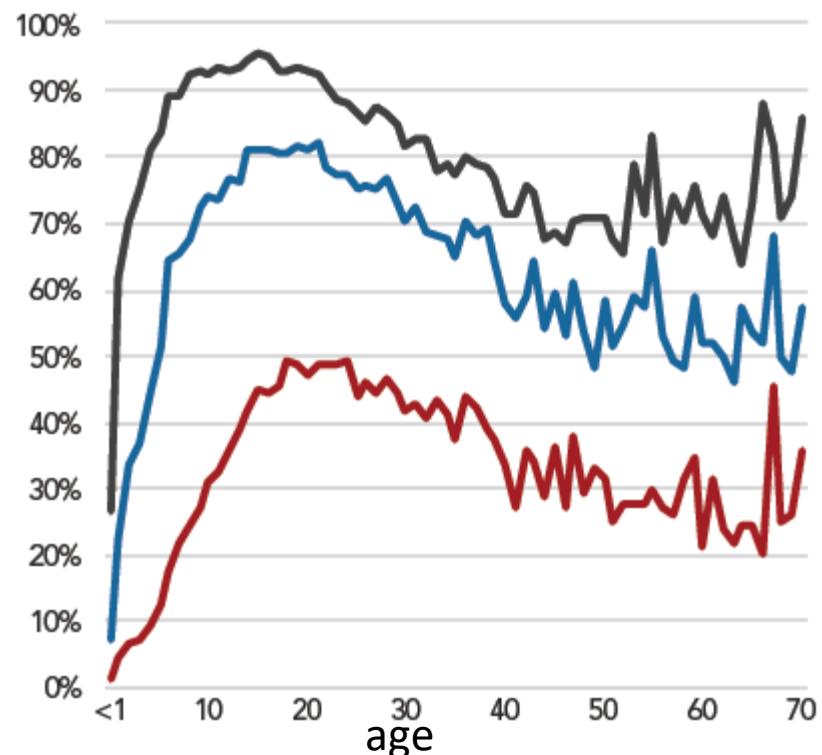
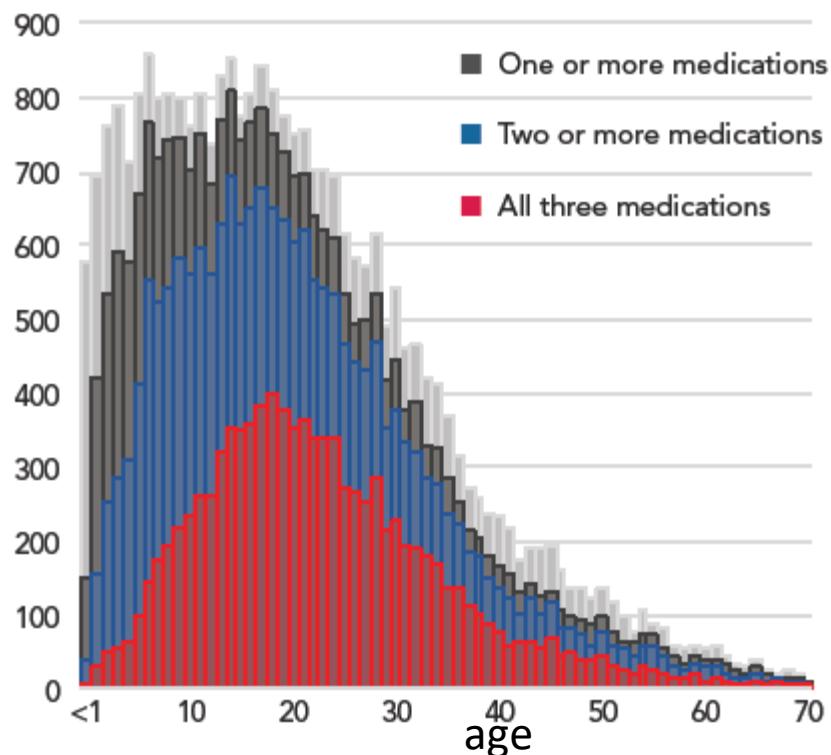
# Use of Meds in the US

Chronic Medication Prescription in Eligible Patients, 1995-2015



# Use of Meds in the US

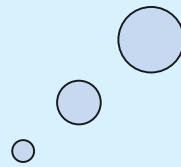
Inhaled Medication (Dornase Alfa, Hypertonic Saline, Inhaled Antibiotic) Prescription by Age in Years, 2015



# Complications

- ABPA- 36 (6.4%)
- Hemoptysis- 19 (3.7%)
- Diabetes (insulin treated)- 104 (19%)
  - Children- 8 (3%)
  - Adults- 96 (31%)
- Liver dis-
  - Chirrosis with portal HTN- 12 (4%)
  - Chirrosis w/o portal HTN- 5 (0.9%)
  - Liver dis, no chirrosis- 72 (13%)

# Summary



# Summary

- CF in Israel is becoming an adult disease
- Pulmonary function are comparable to best European countries
- PsA is a major pathogen starting in childhood, with no change in the last years ( in children?)
- Staph prevalence is rising
- BMI is within the normal range but wide variability
- Medication use is high but lower than in the US

# Summary

- This is only the beginning....

- Thanks to:
  - The patients that gave consent to be in the database
  - The staff at all centers in charge of the data entry
  - The ECFStracker