

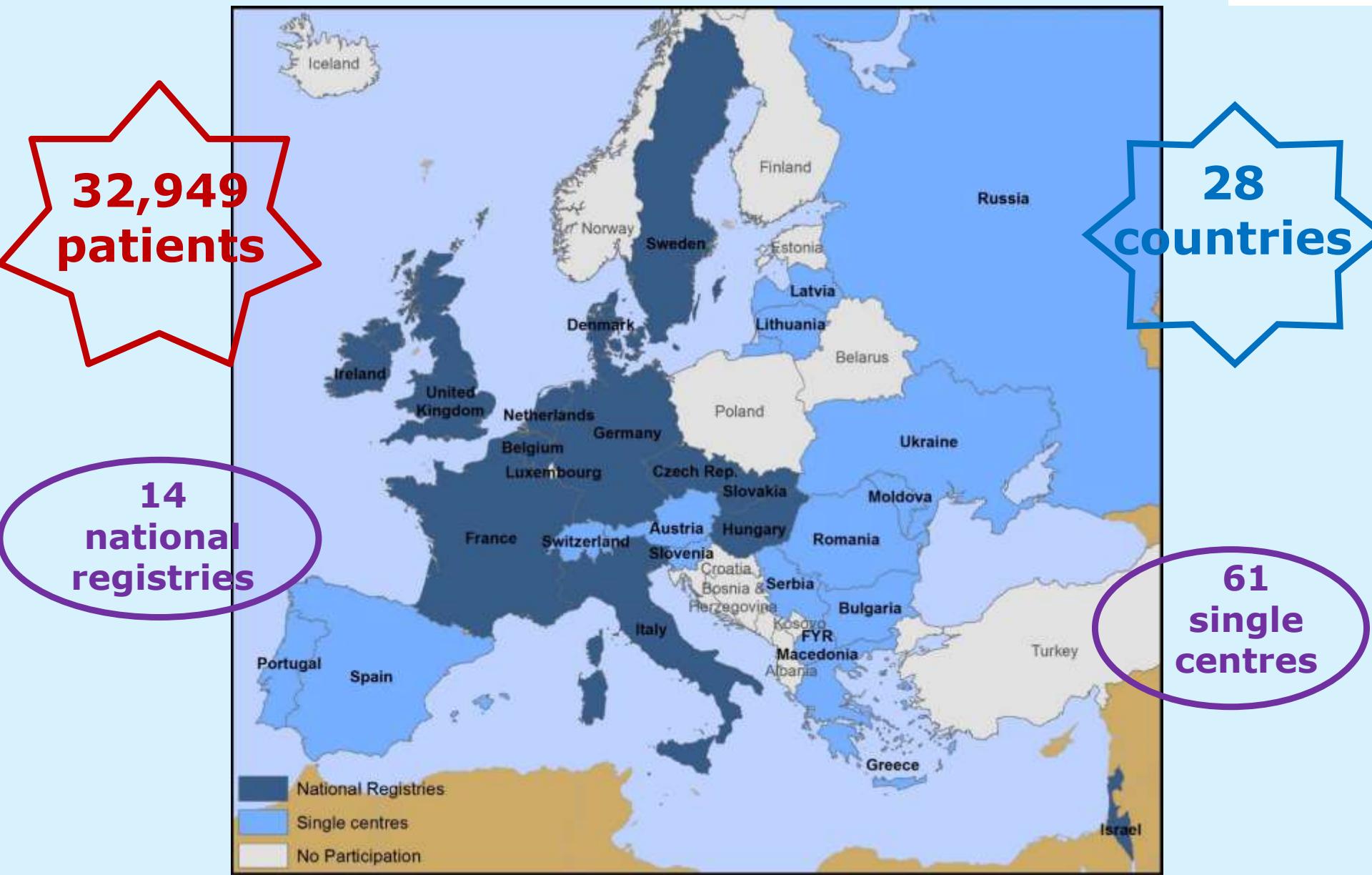
European CF Patient Registry (ECFSPR)

2013

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



Participating 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



- Web-based software
- User friendly
- Remote management
- European Data Protection compliant
- Future add-on modules

Data Collection - Variables

Demographic

age, gender, status of patient, etc.

Diagnosis

age at diagnosis, sweat test, meconium Ileus, neonatal screening

Genotype

Growth/lung function

Microbiology

Nontuberculous mycobacteria, Pseudomonas aeruginosa, Staphylococcus aureus

Complications

Diabetes, liver disease, pancreatic status, malignancy

Therapy

Antibiotic, bronchodilators, Oxygen therapy, pancreatic enzymes

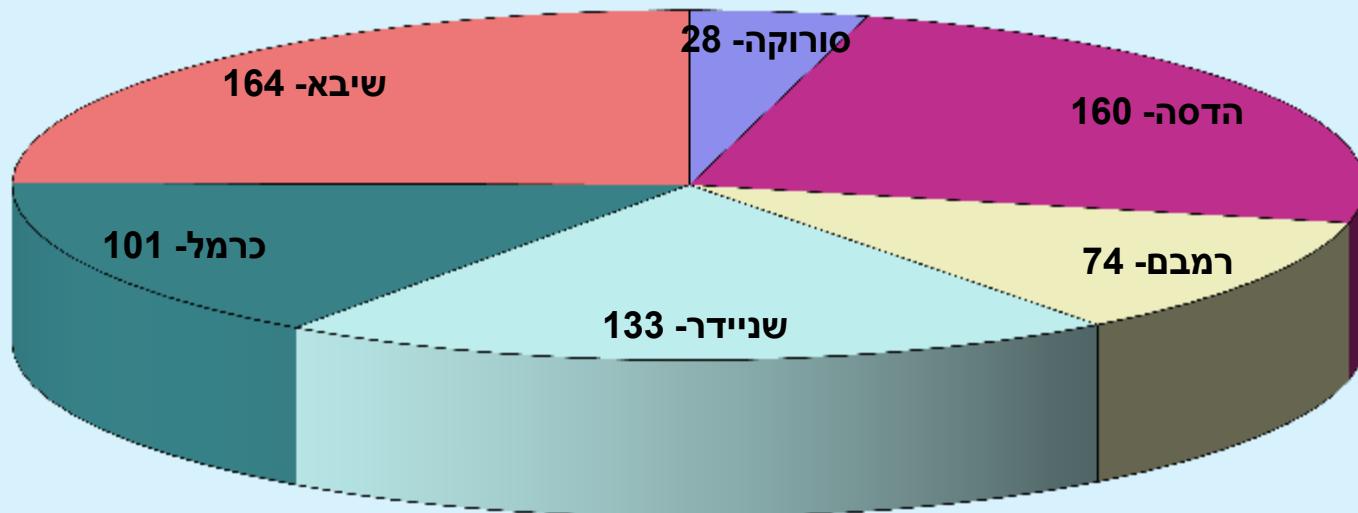
Lung/liver transplant

The Israeli CF Database Update 2013



Demographics

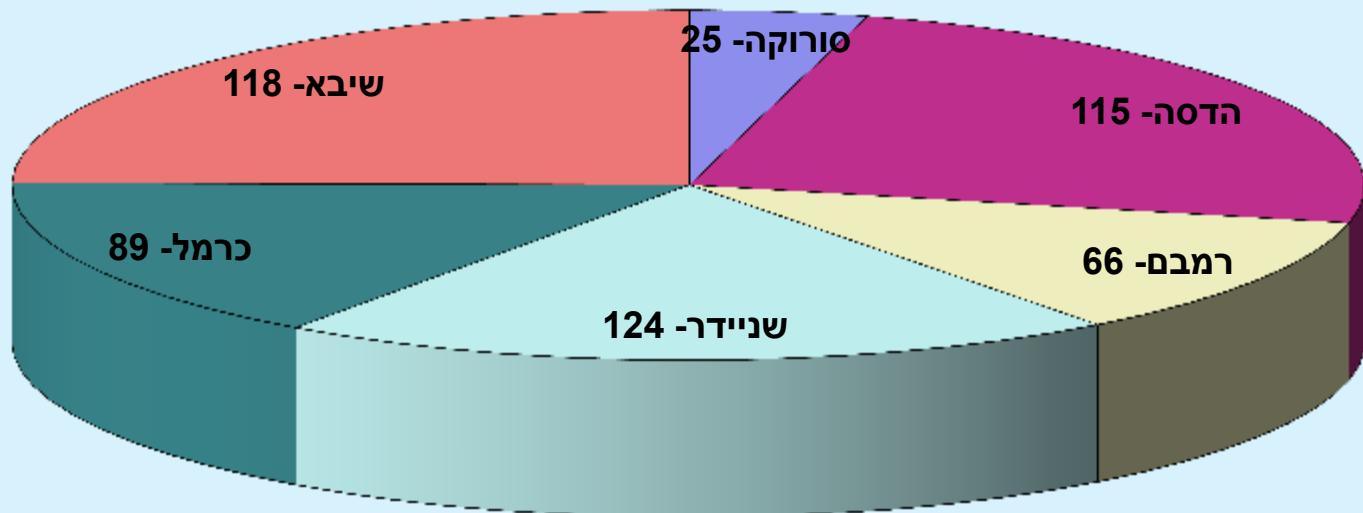
Israeli CF Centers



N=660

Israel 2009

Israeli CF Centers 2013

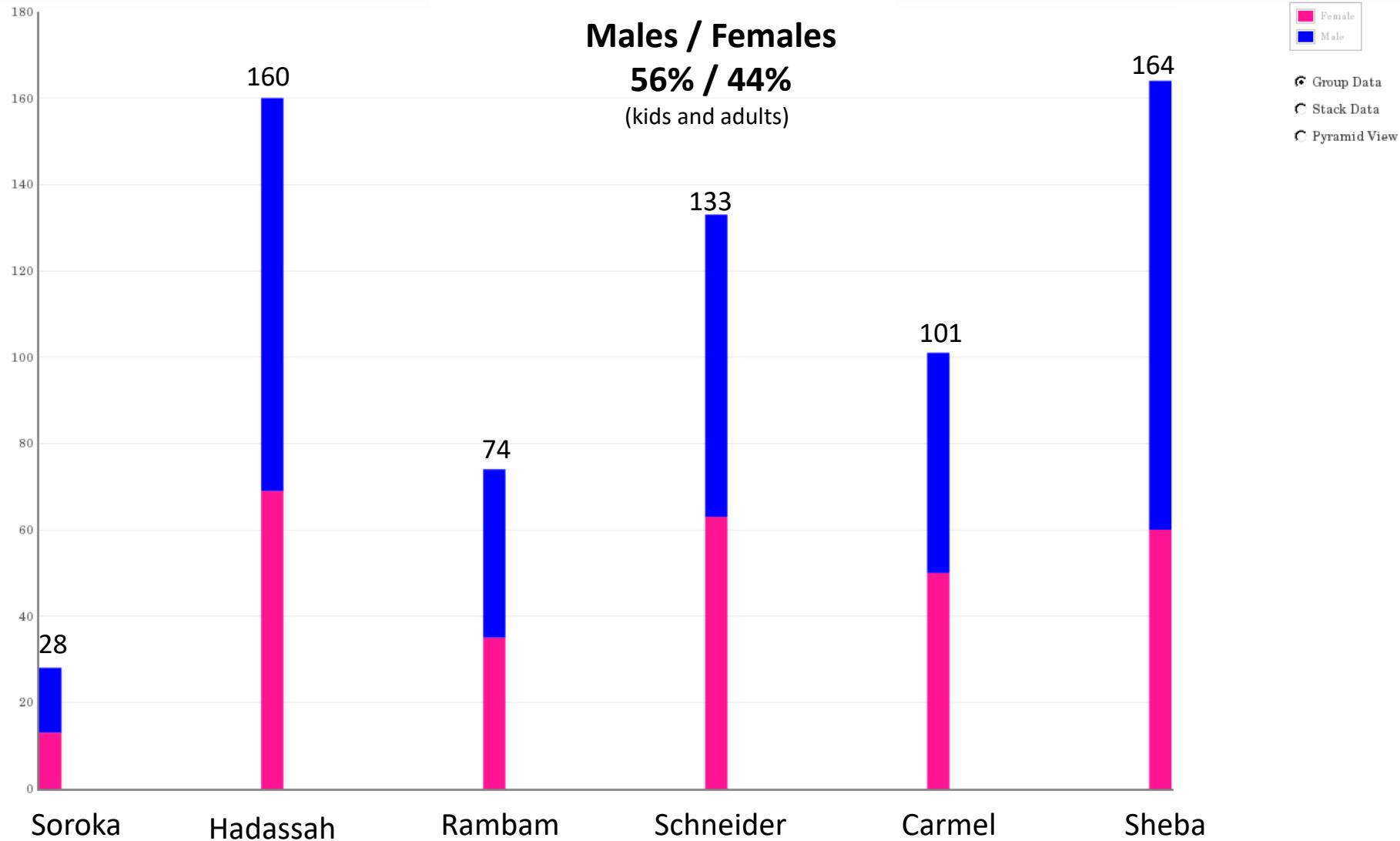


Seen in 2013- 529

Demographics

Gender

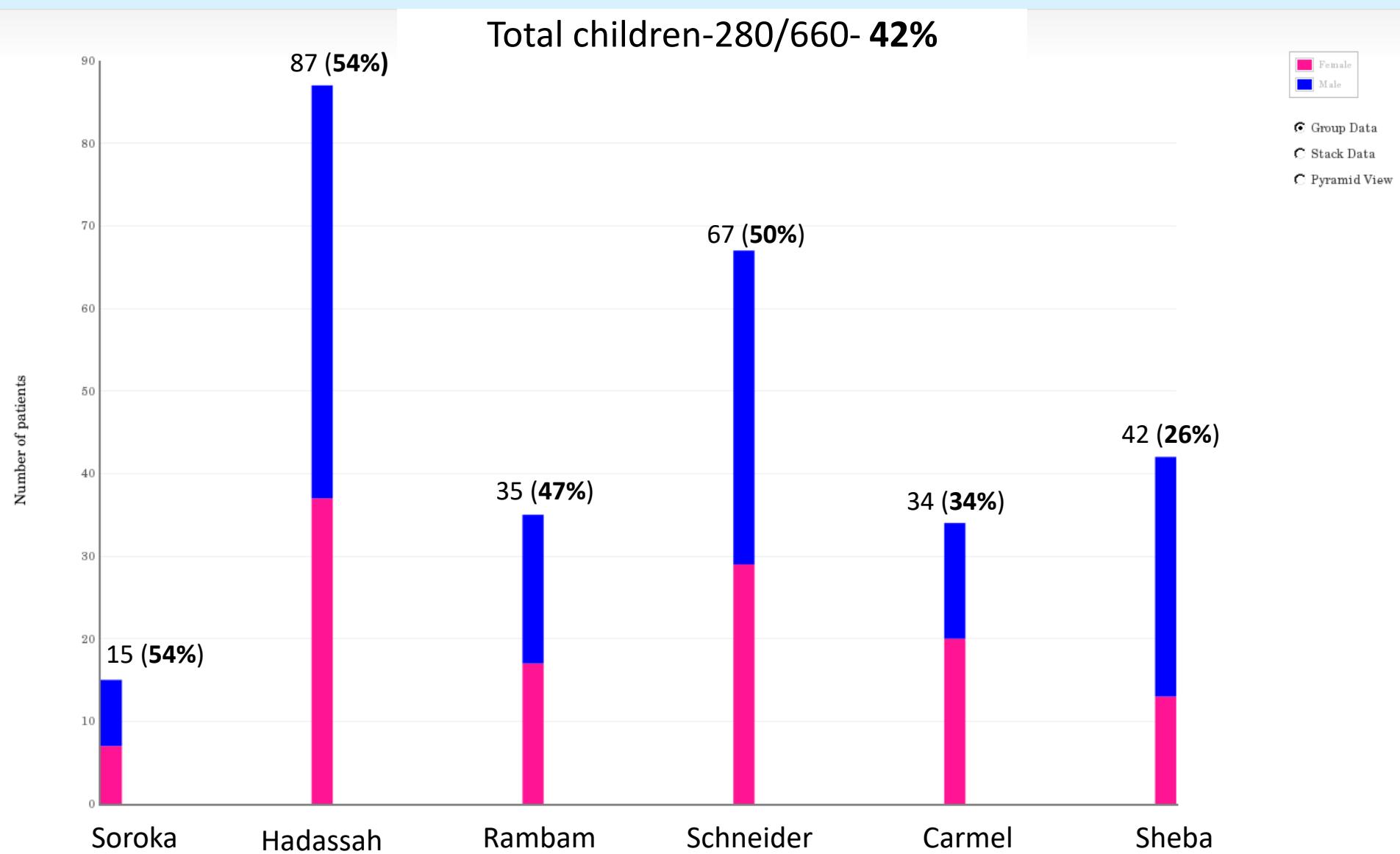
Gender



Demographics

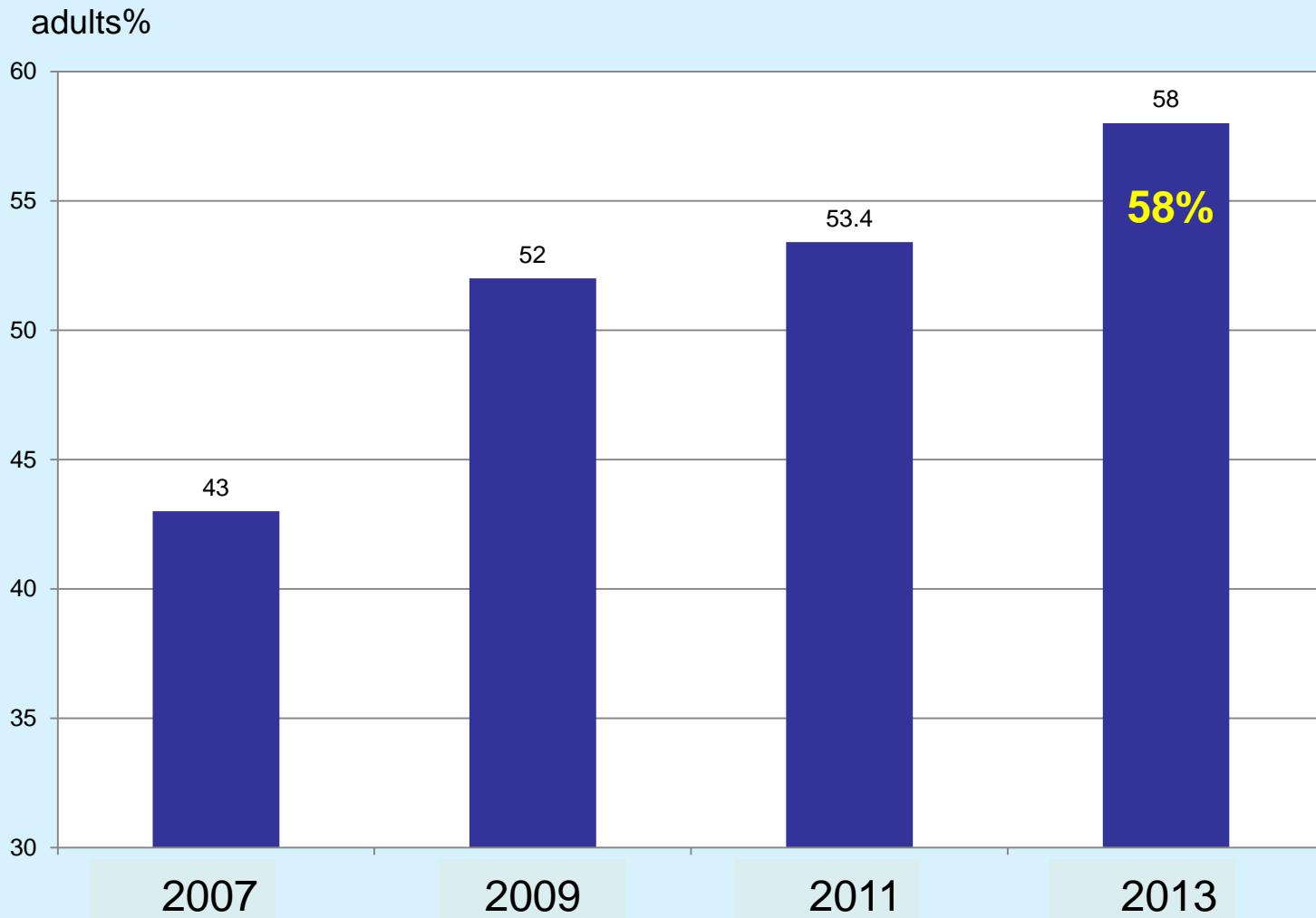
Children and Adults

Number of Children per Center (%)

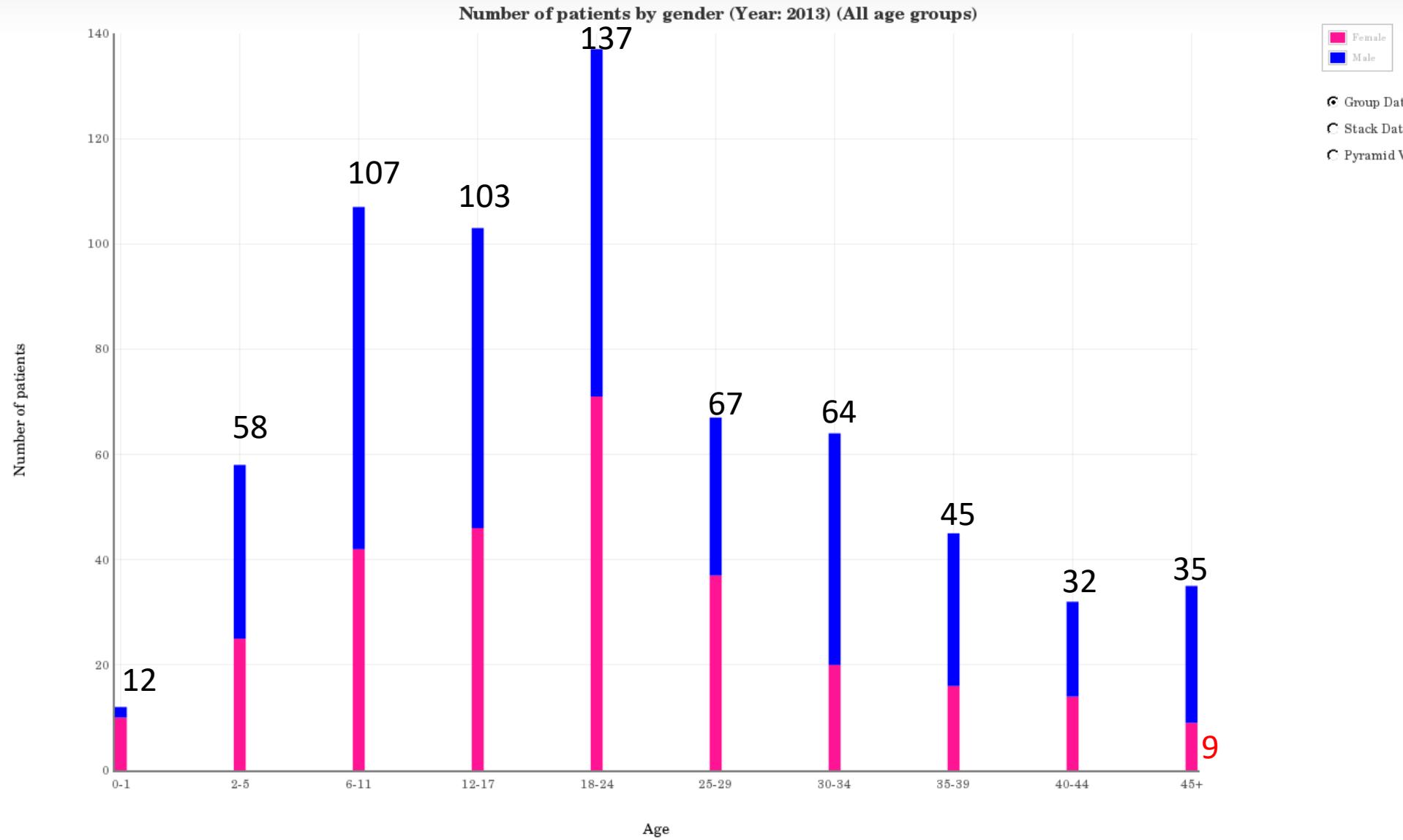


%Adults

2007-13



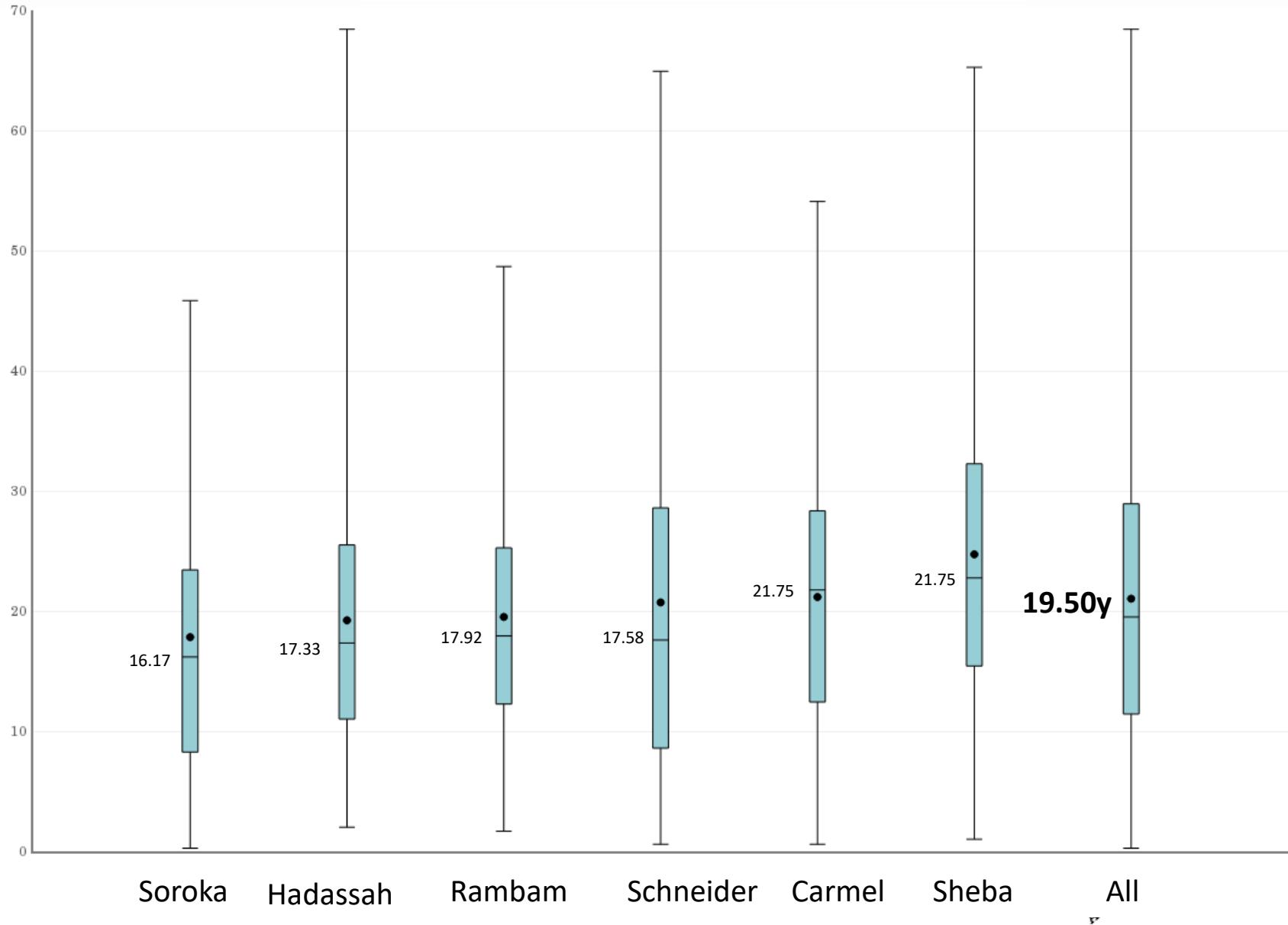
Age & Gender



Age at follow up



Age at follow up

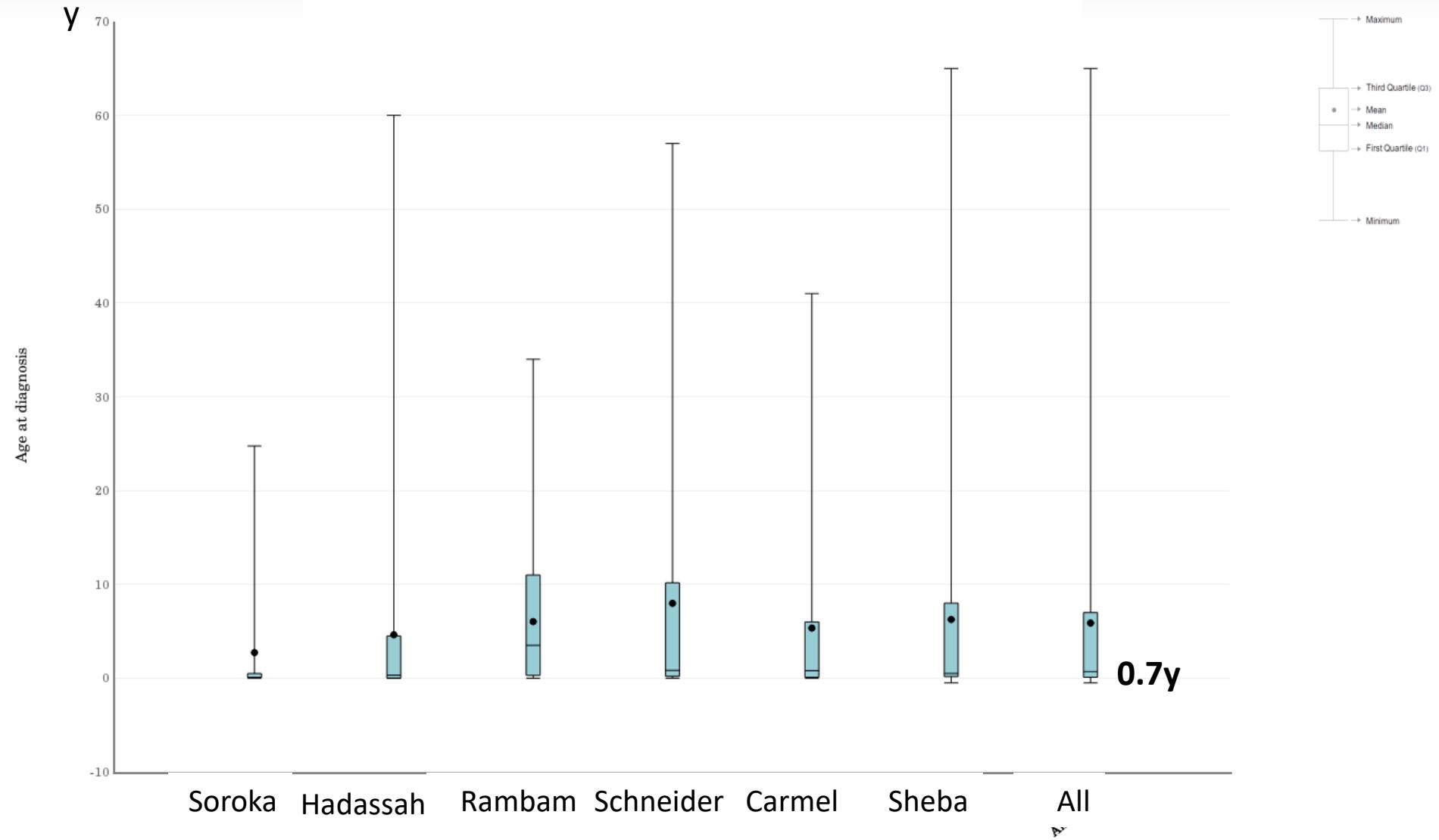


Diagnosis

Neonatal Diagnosis

- Neonatal screen- 1.13% (6/517)
- Meconium ileus- 13.6% (73/529)

Age at Diagnosis



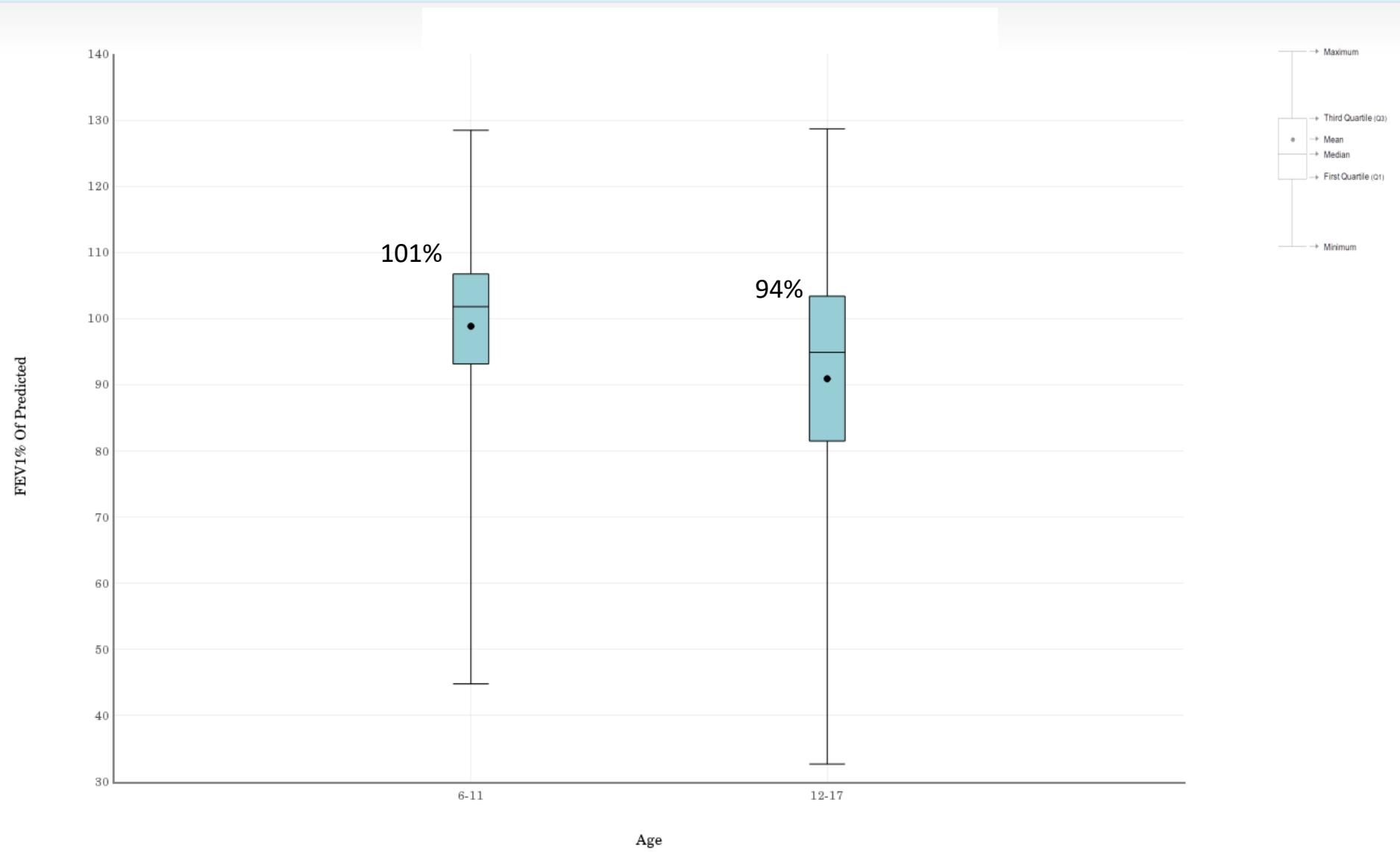
Genetics

- Genetic analysis performed- 528/529!!!
- 2 mutations found- 80.1% of pts
- Mutations found- 84.7% of alleles
- 74 mutations in the Israeli registry

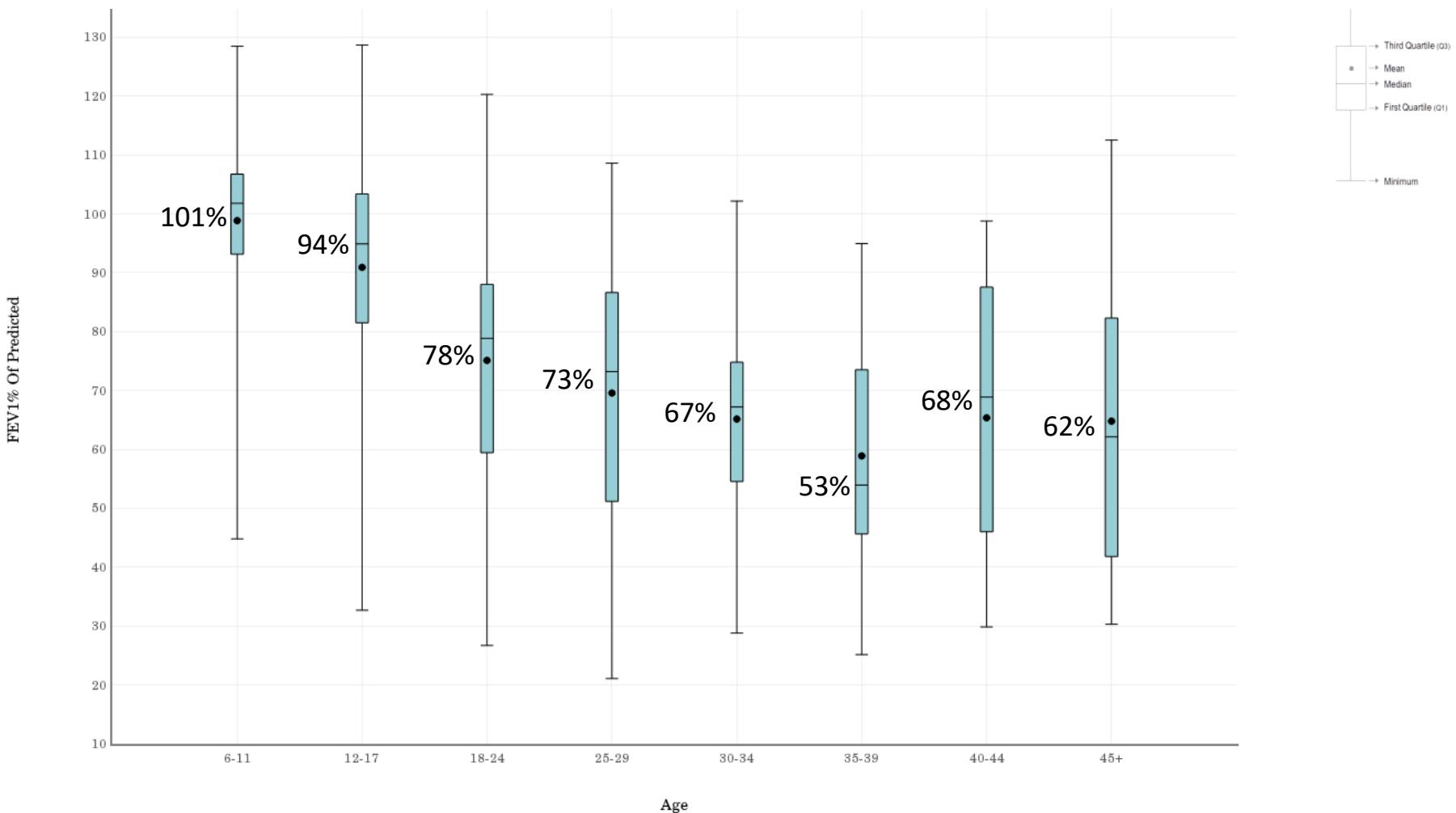
| mutation | Frequency | Percent |
|---------------|-----------|--------------|
| F508del | 251 | 23.77 |
| W1282X | 244 | 23.11 |
| unknown | 162 | 15.34 |
| D1152H | 61 | 5.78 |
| G542X | 57 | 5.40 |
| N1303K | 41 | 3.88 |
| 3849+10kbC->T | 34 | 3.22 |
| 5T | 22 | 2.08 |
| G85E | 21 | 1.99 |
| 405+1G->A | 17 | 1.61 |
| Q359K/T360K | 16 | 1.52 |
| I1234V | 10 | 0.95 |
| S549R(A->C) | 10 | 0.95 |

Clinical - Pulmonary

FEV₁- Kids - Israel



FEV₁ with Age- Israel



Report generated by:

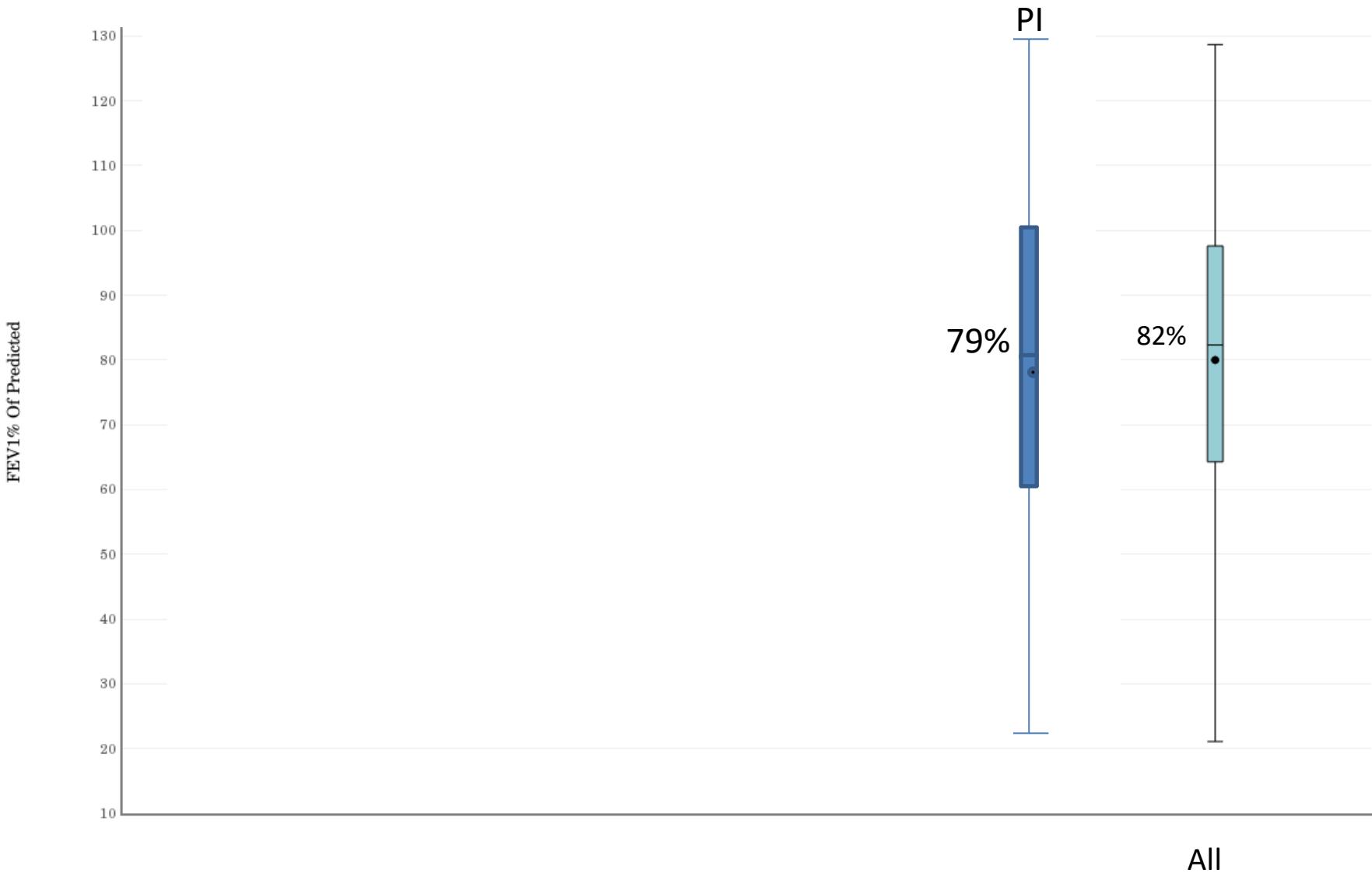
Meir Mei-Zahav

Report date:

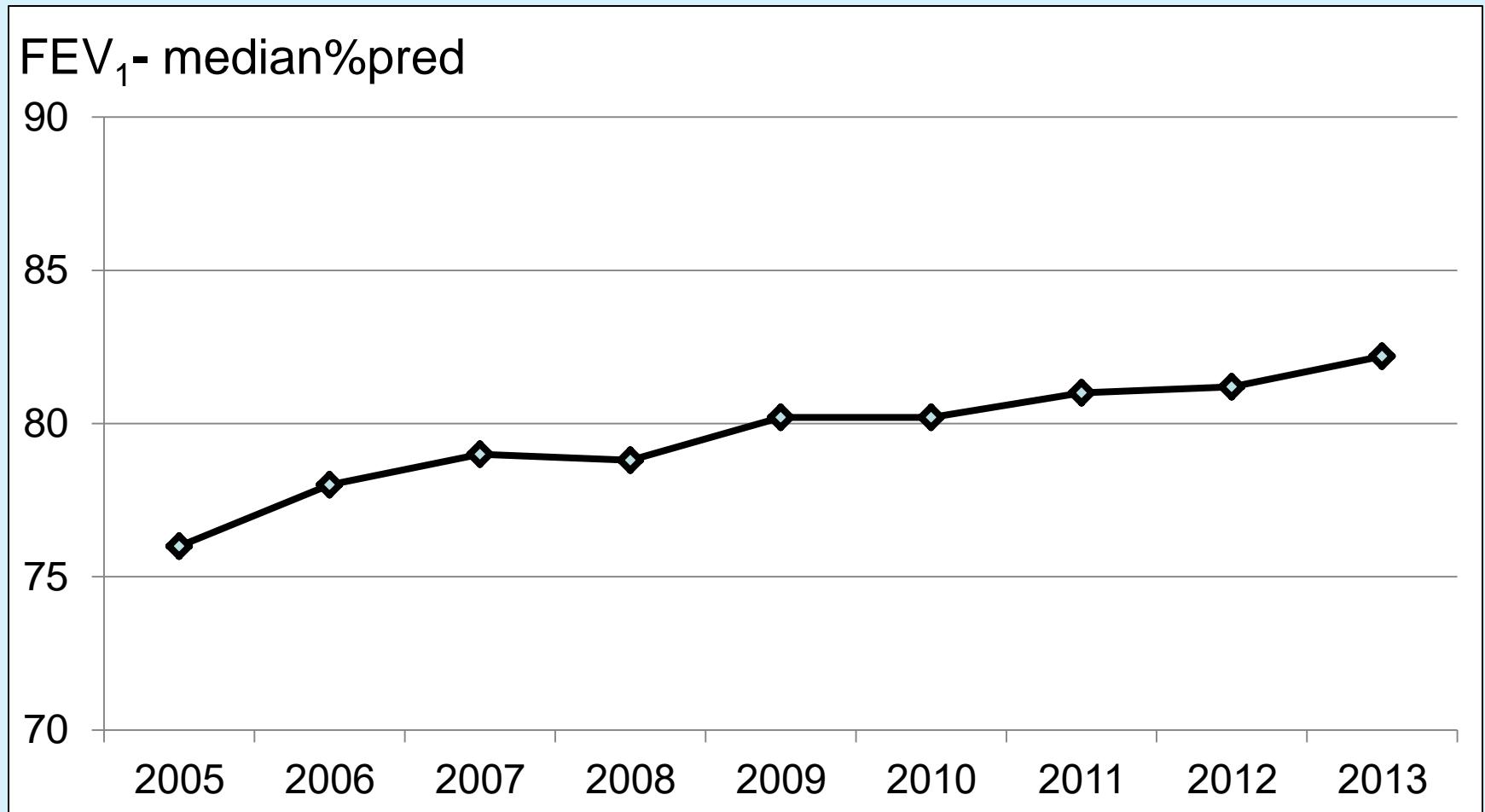
Sun, 25 Oct 2015 15:10:46 GMT

ECFSPR validation status: **No**

FEV₁ median - Israel



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



FEV₁- median- Children- Europe

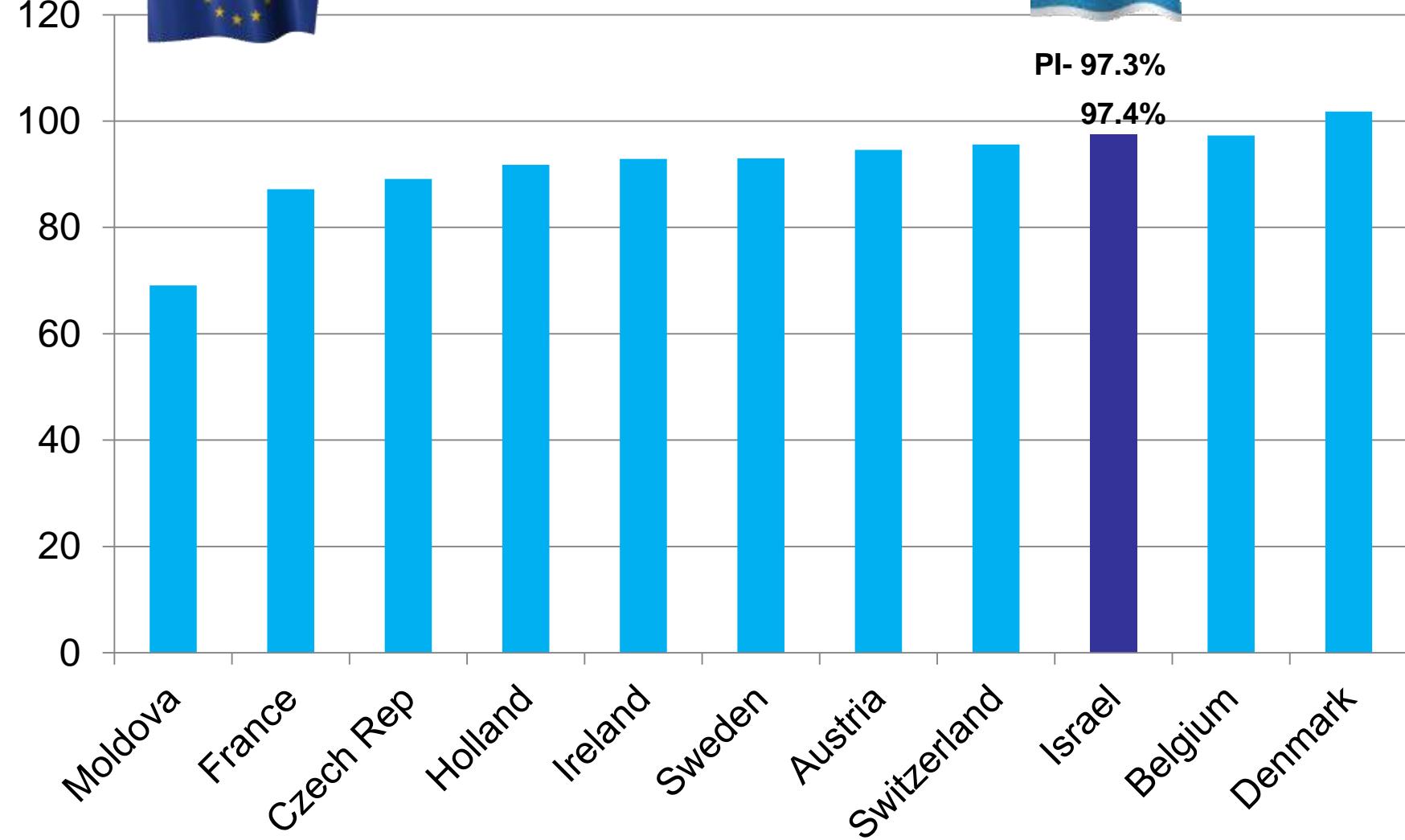


FEV1%

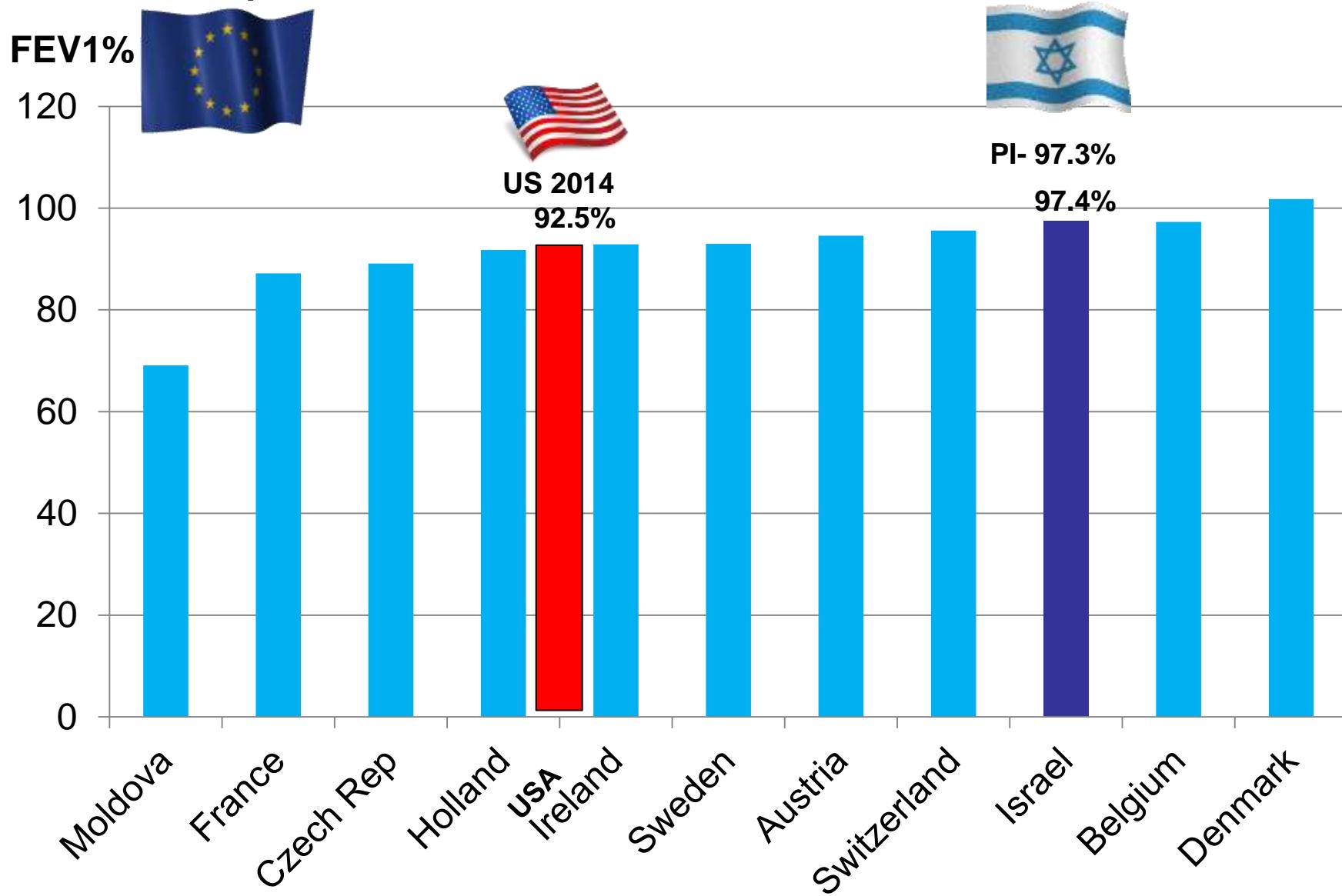


PI- 97.3%

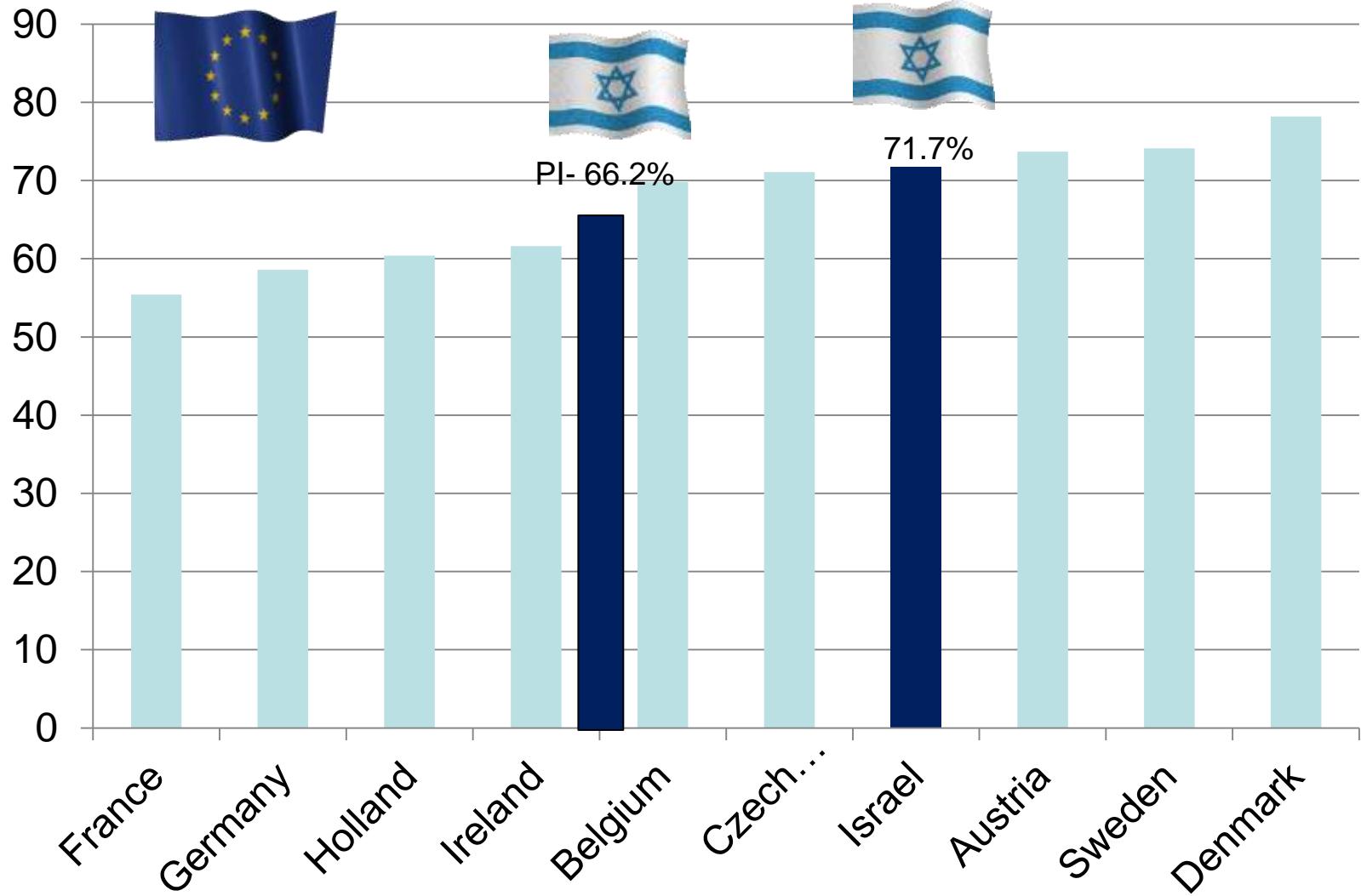
97.4%



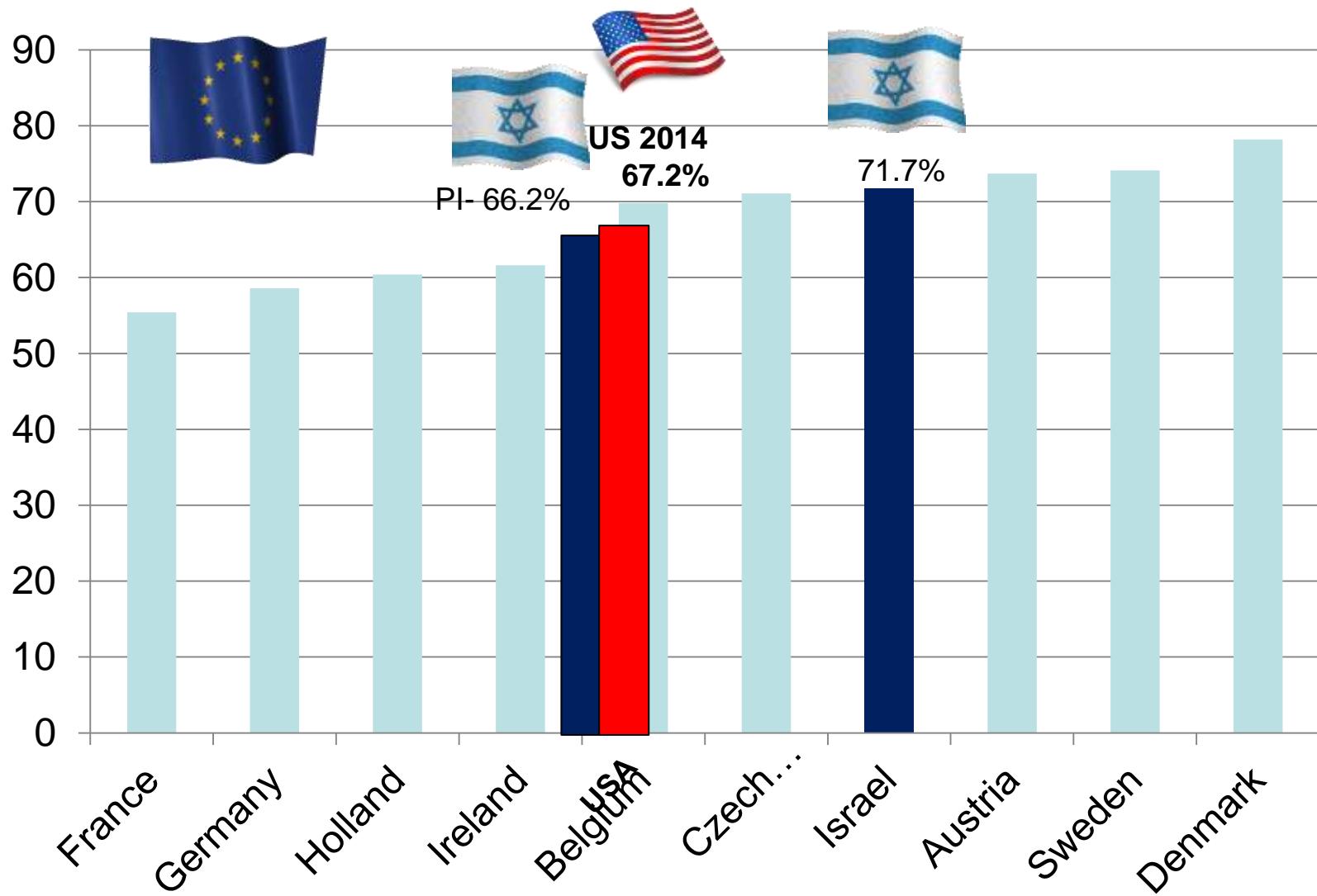
FEV₁- median- Children- Europe



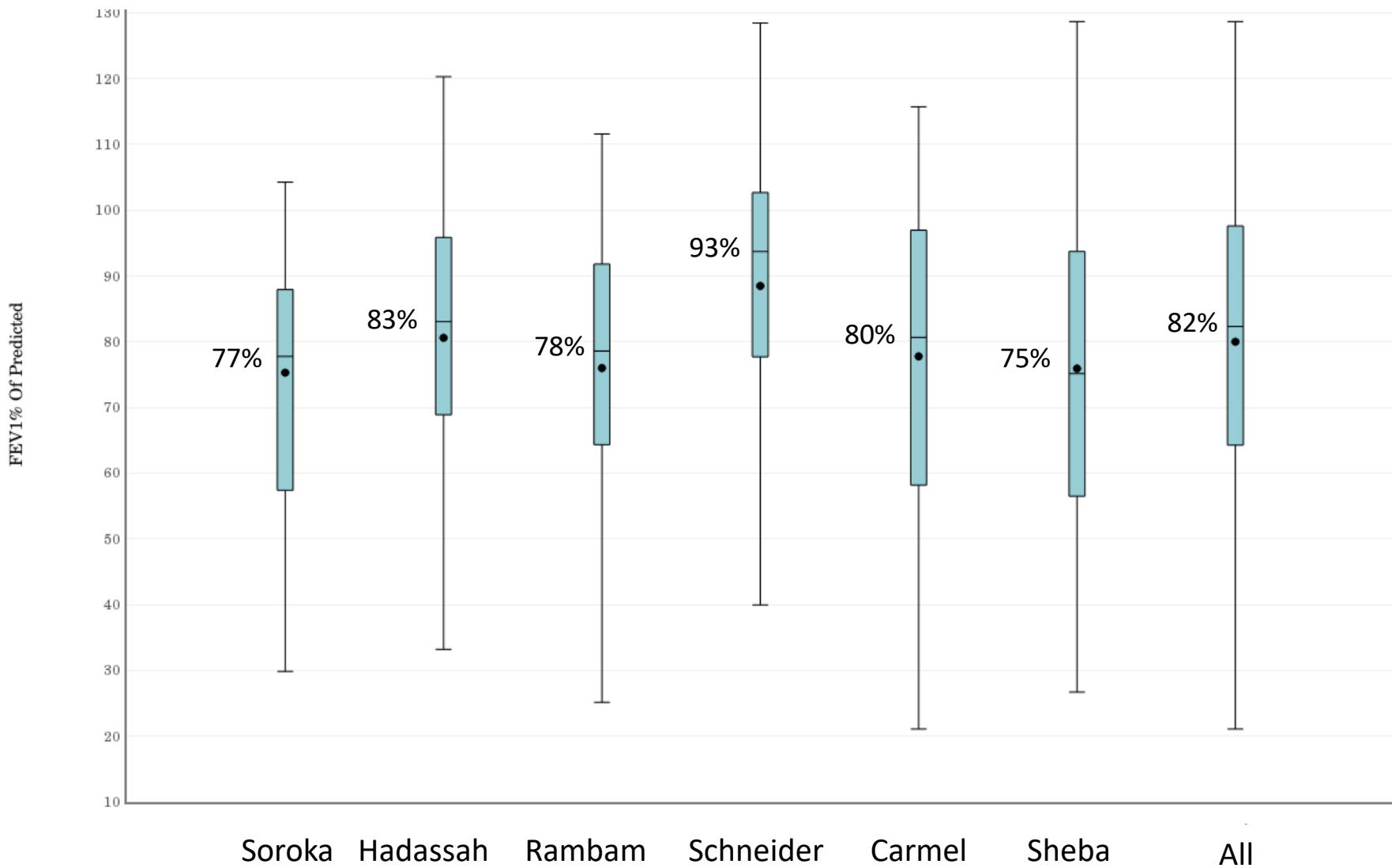
FEV₁- median- Adults - Europe



FEV₁- median- Adults - Europe

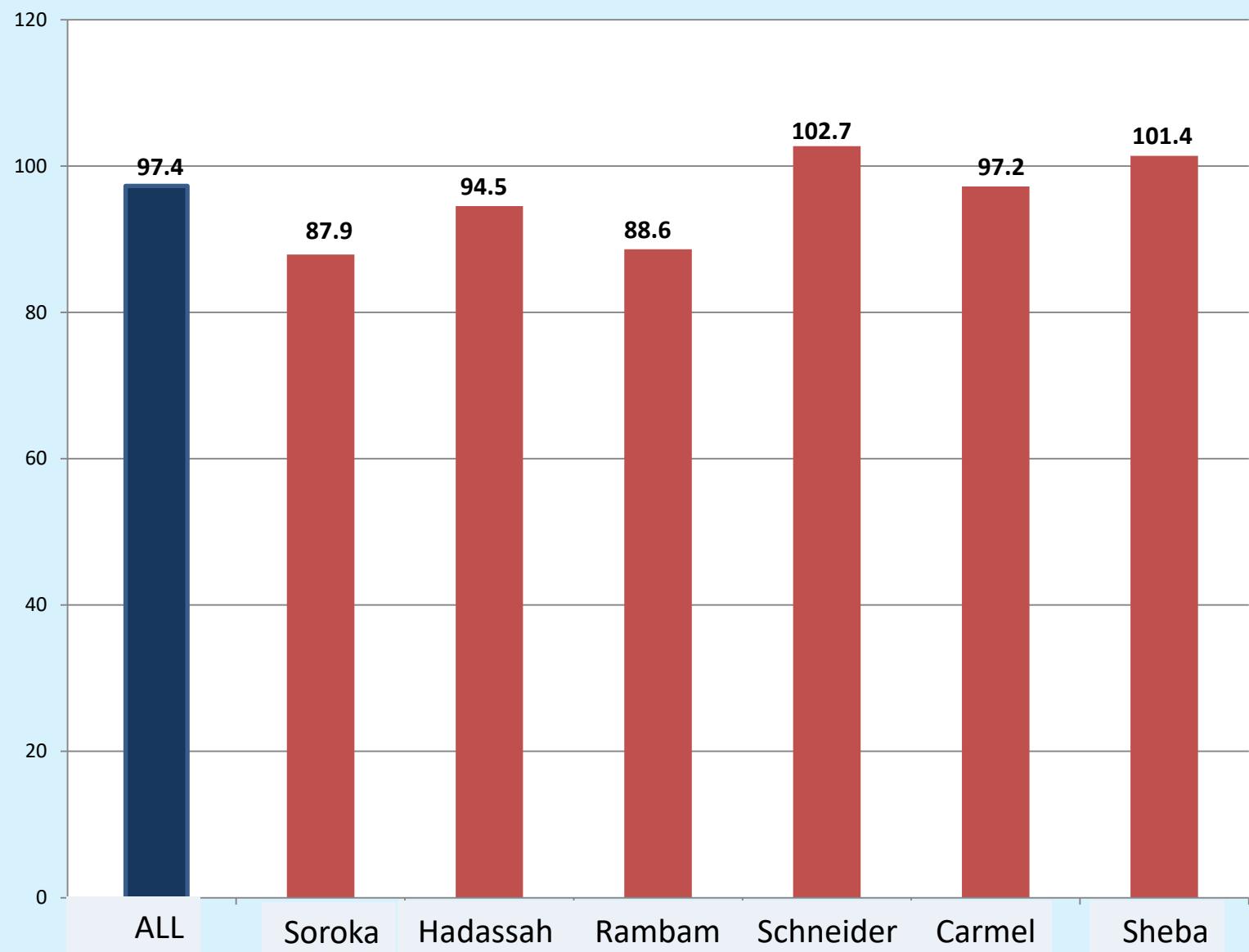


FEV₁- Centers - Israel



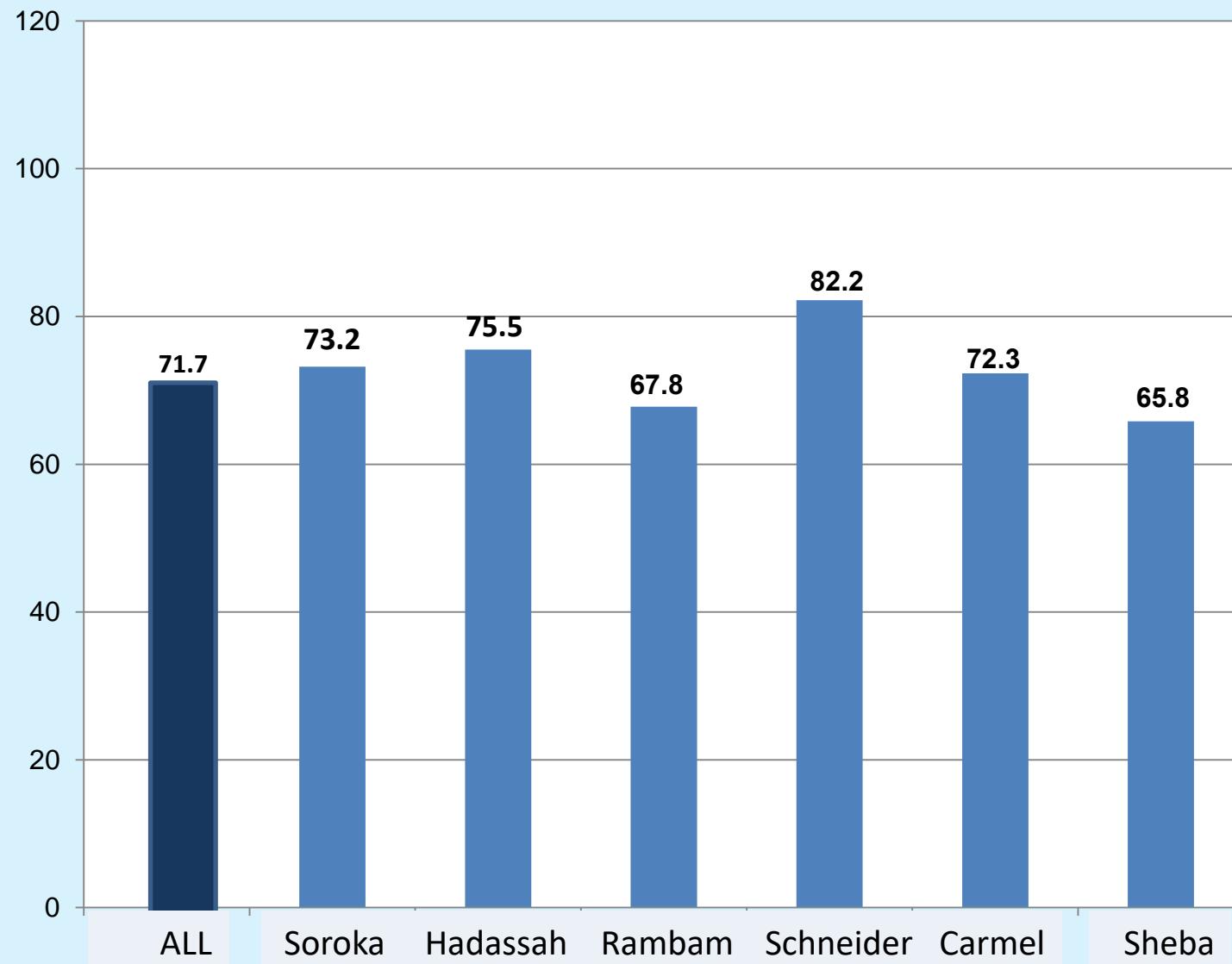
FEV_1 - median- Children

%



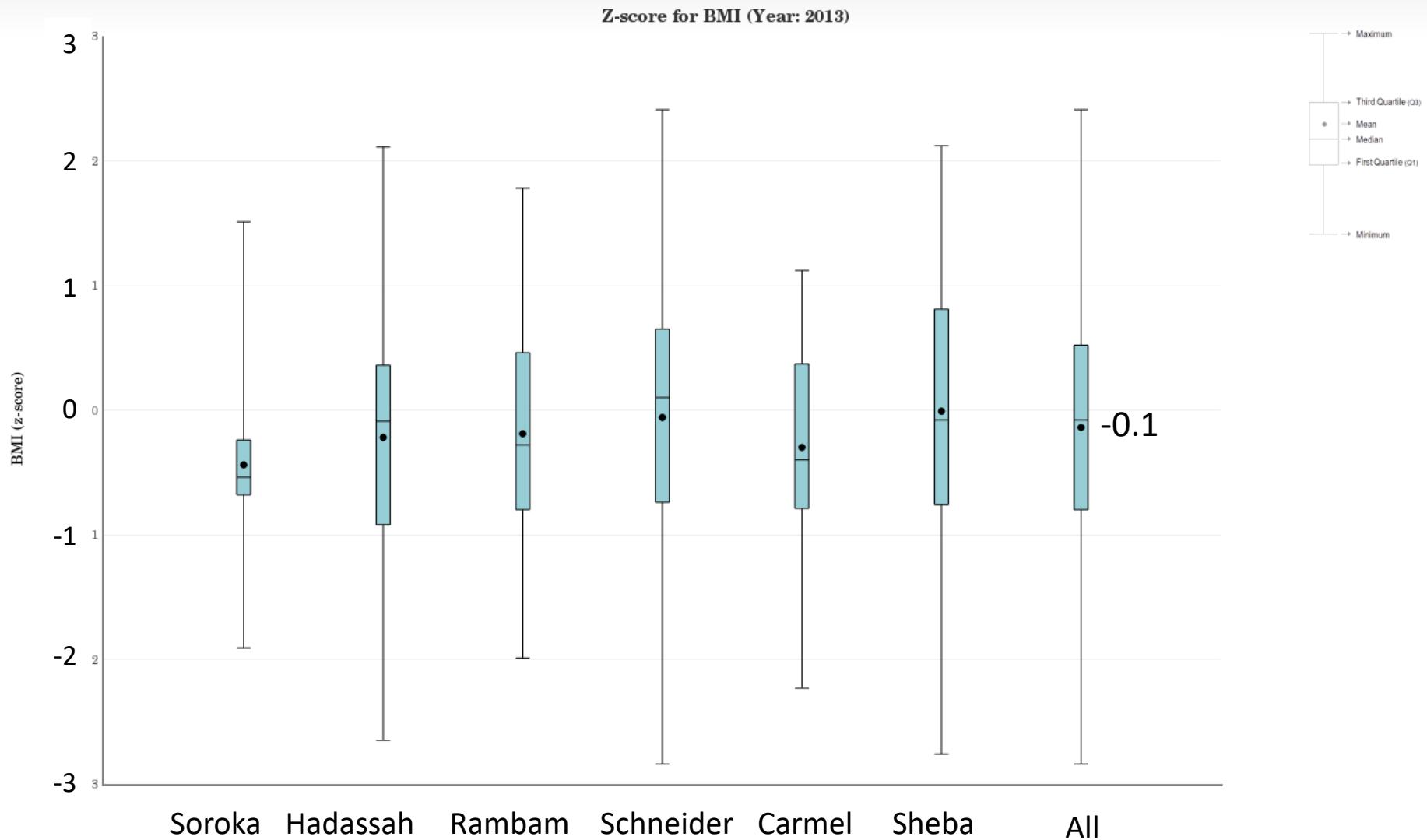
FEV_1 - median- Adults

%

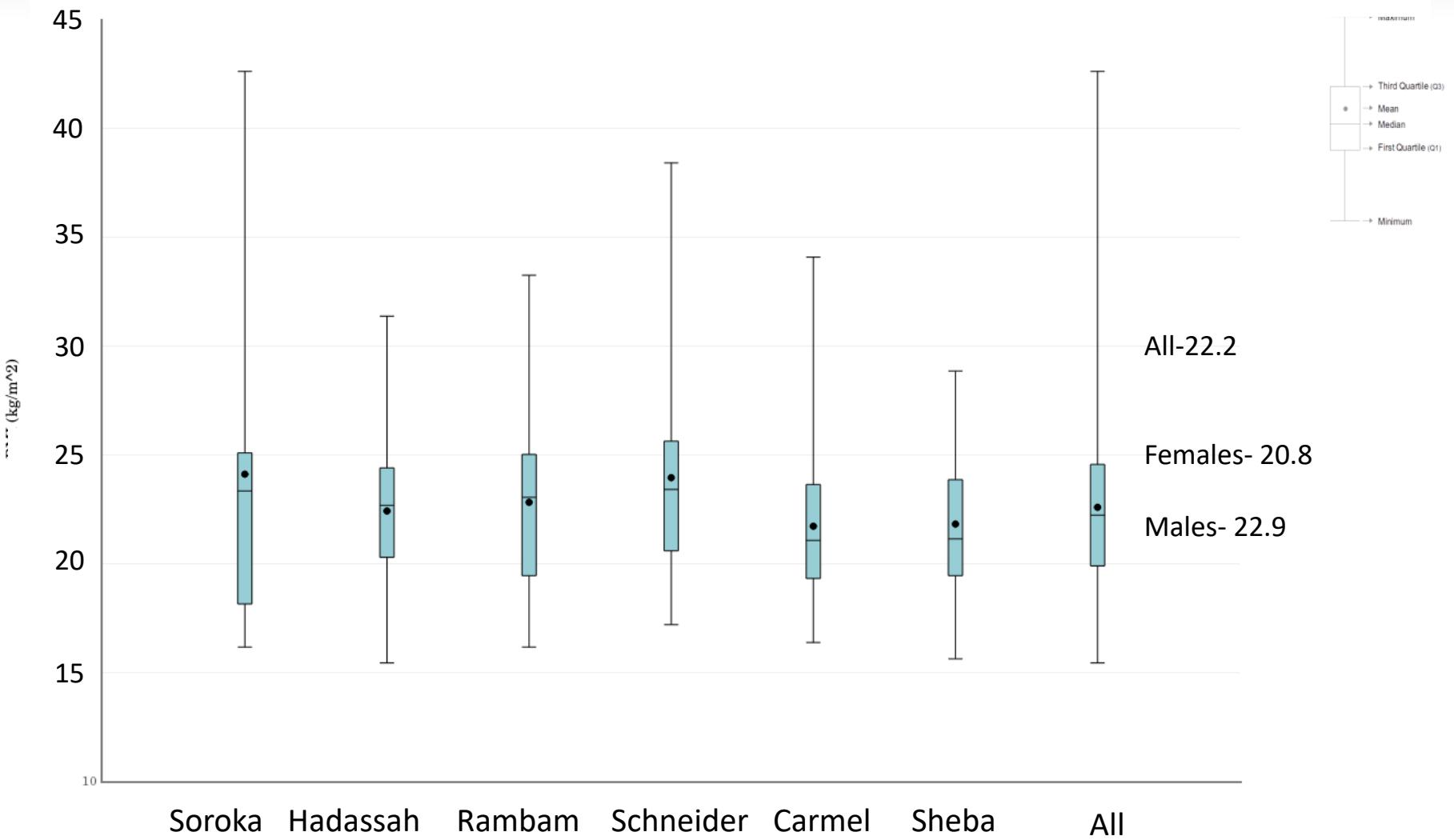


Clinical - Nutrition

BMI- Z score - children



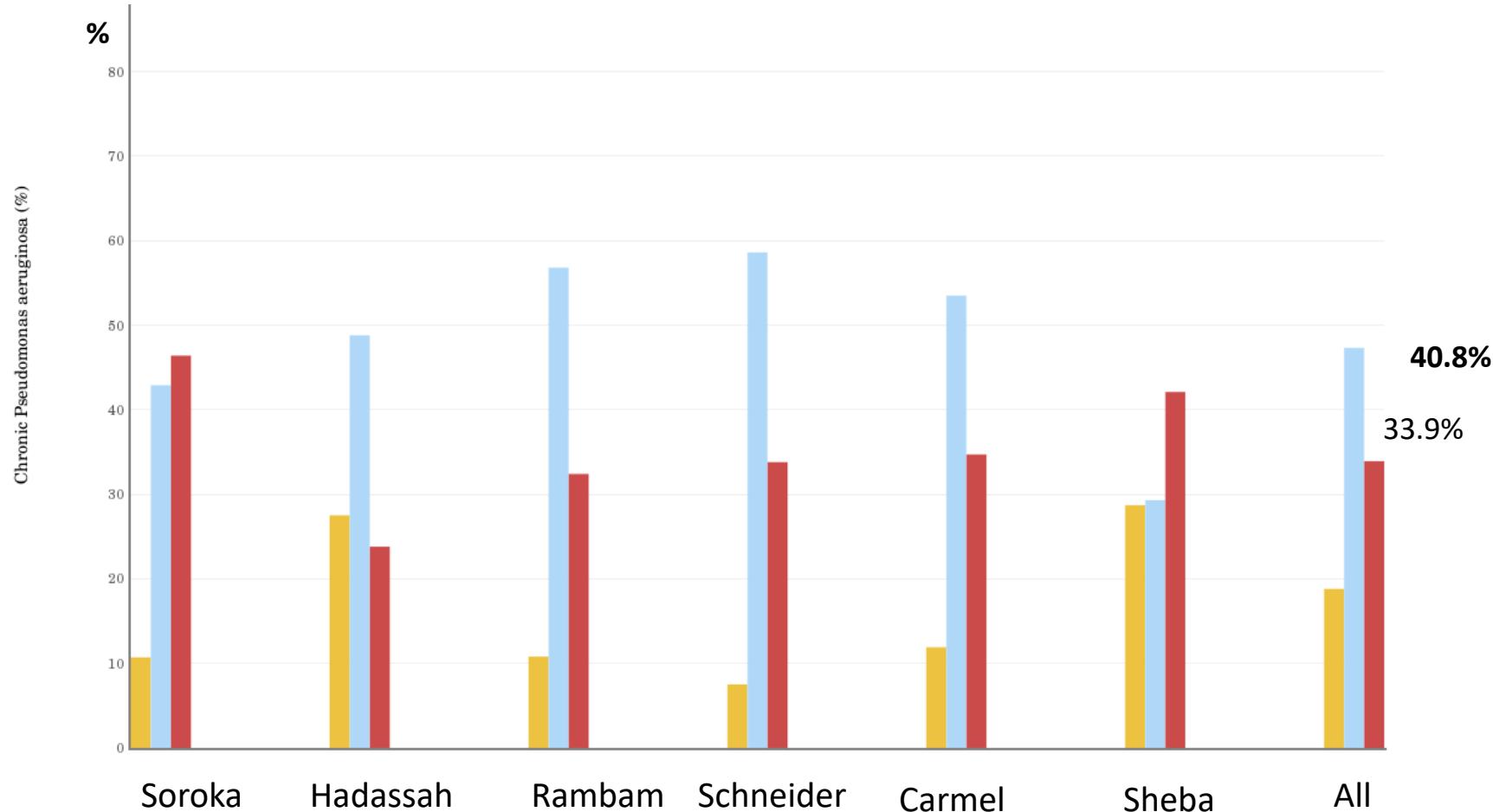
BMI- M+F Adults



Bacteriology

Ps. A in Israel

Unknown
No
Yes



Report generated by:

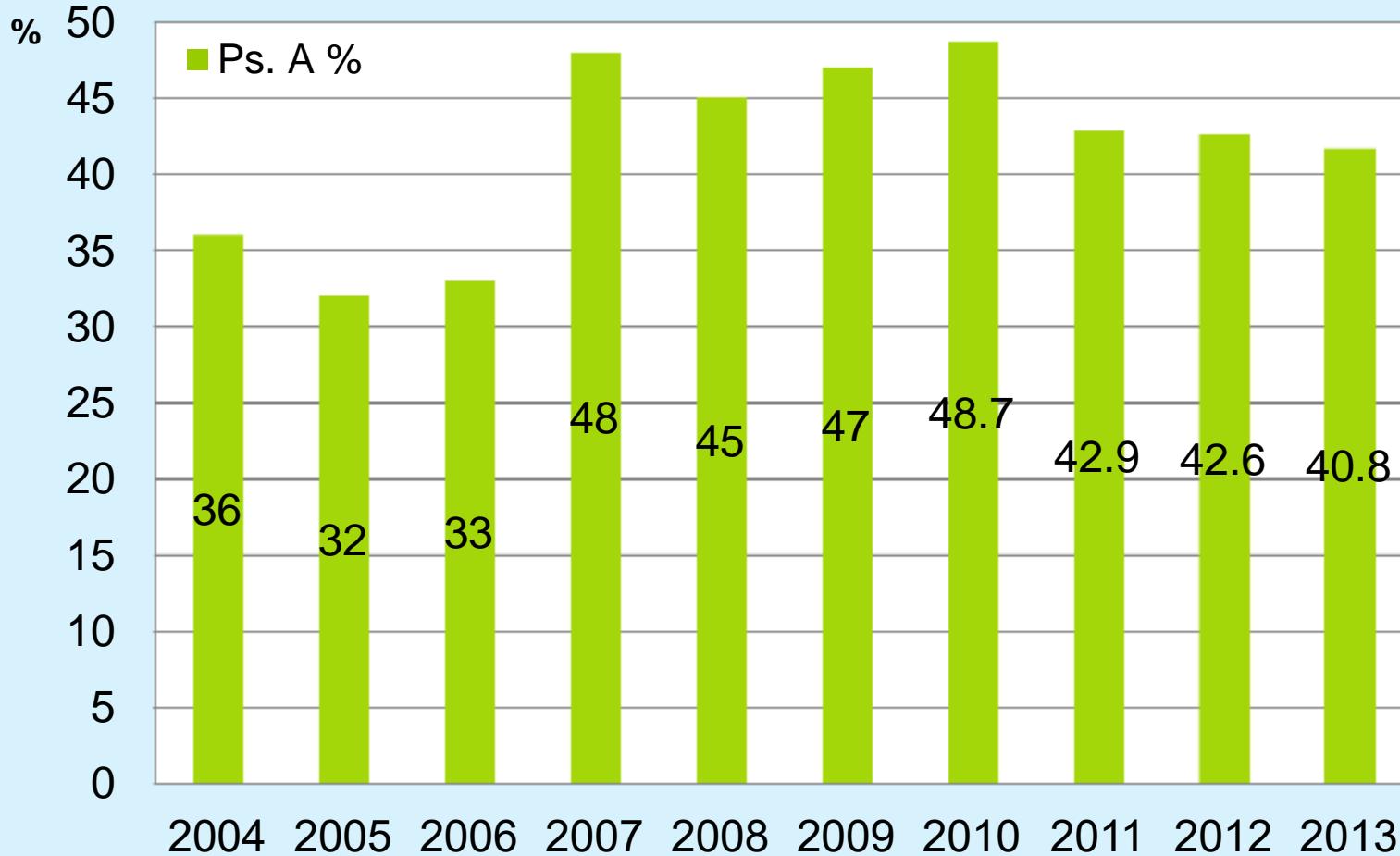
Meir Mei-Zahav

Report date:

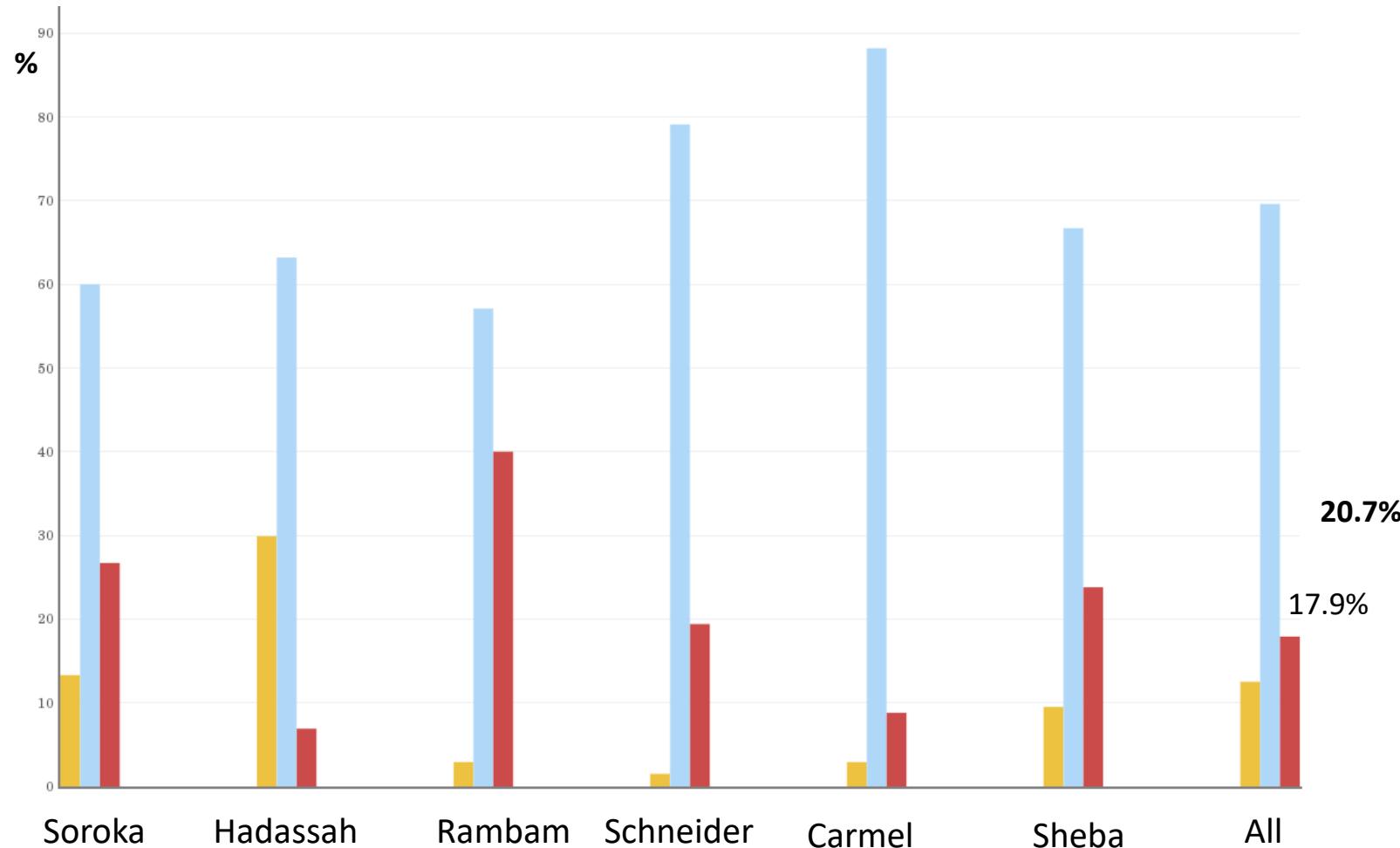
Mon, 26 Oct 2015 16:28:24 GMT

ECFSRP validation status: **No**

Ps. A in Israel- Trend



Ps. A in Israel- Children



Report generated by:

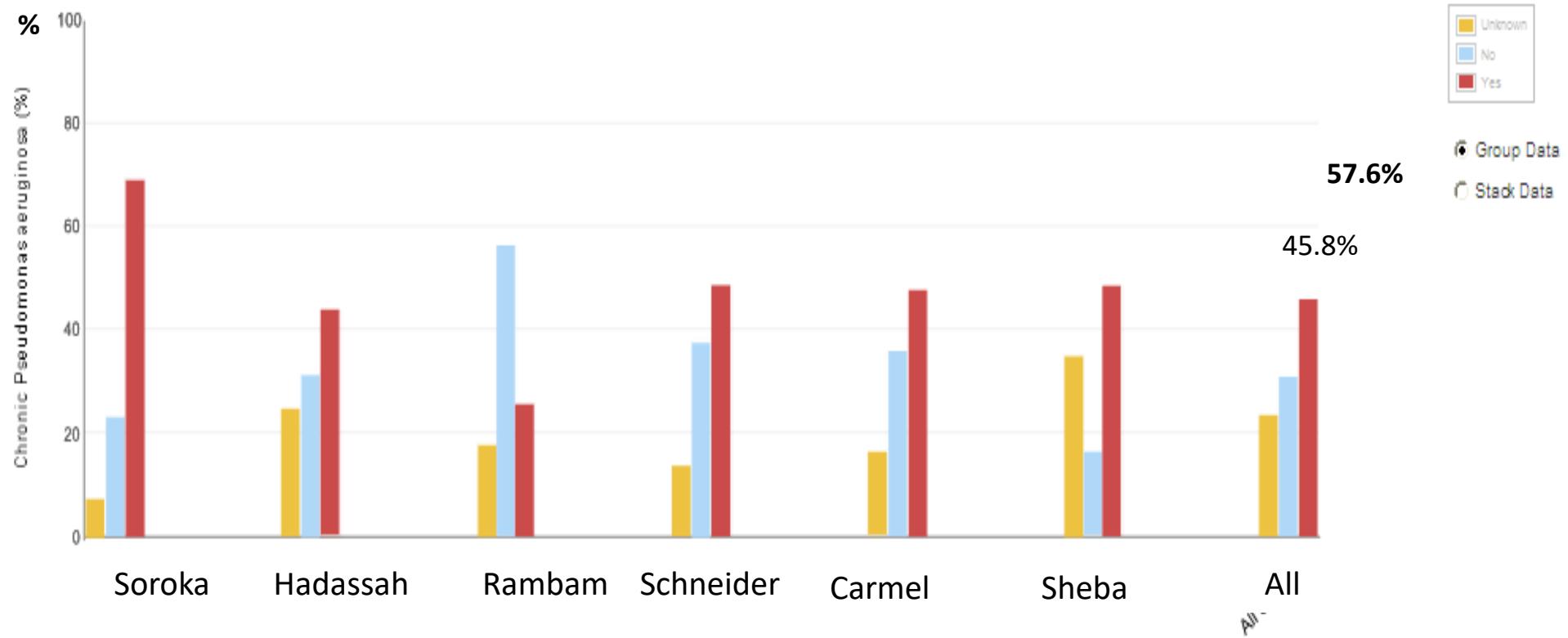
Meir Mei-Zahav

Report date:

Mon, 26 Oct 2015 16:38:03 GMT

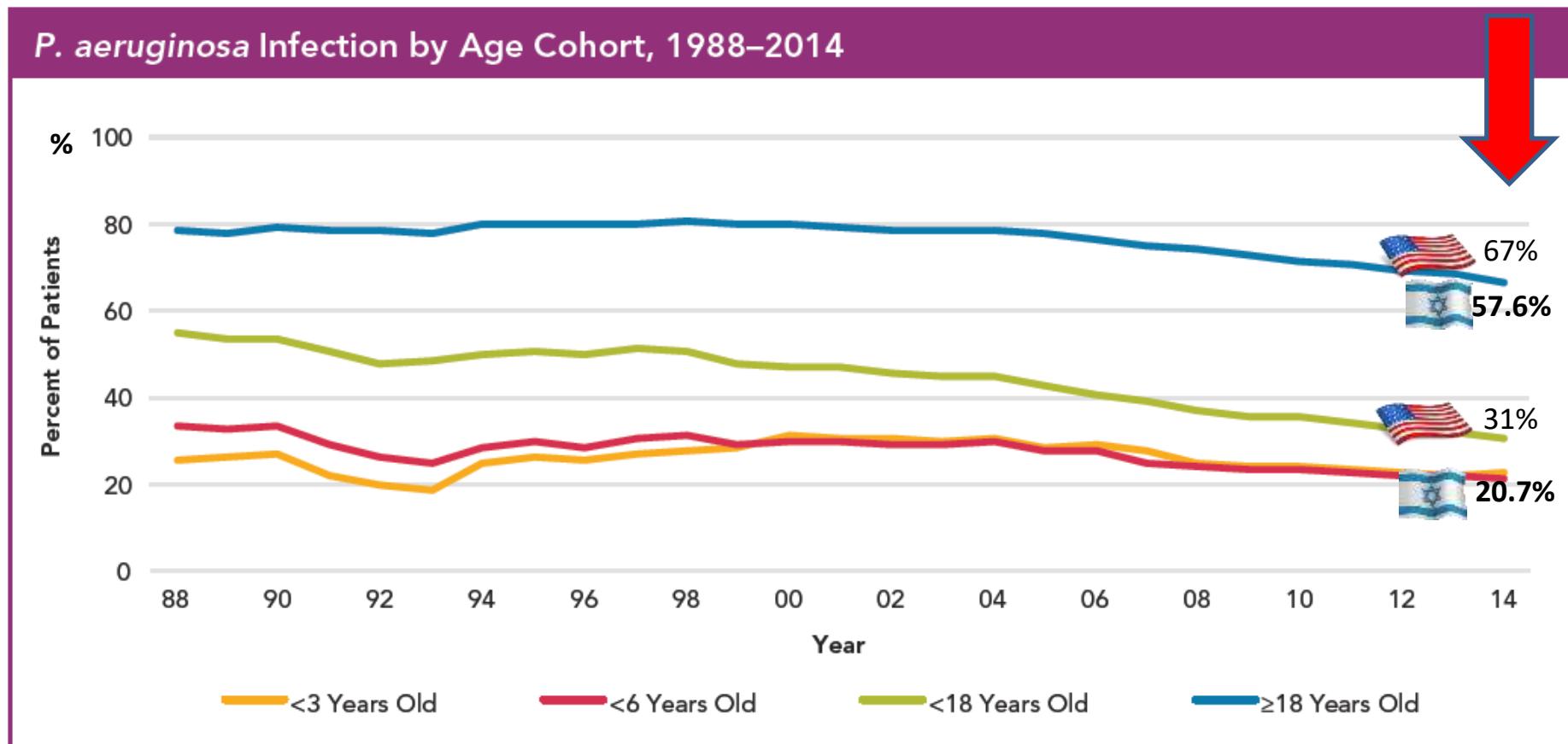
ECFSRP validation status: **No**

Ps. A in Israel- Adults

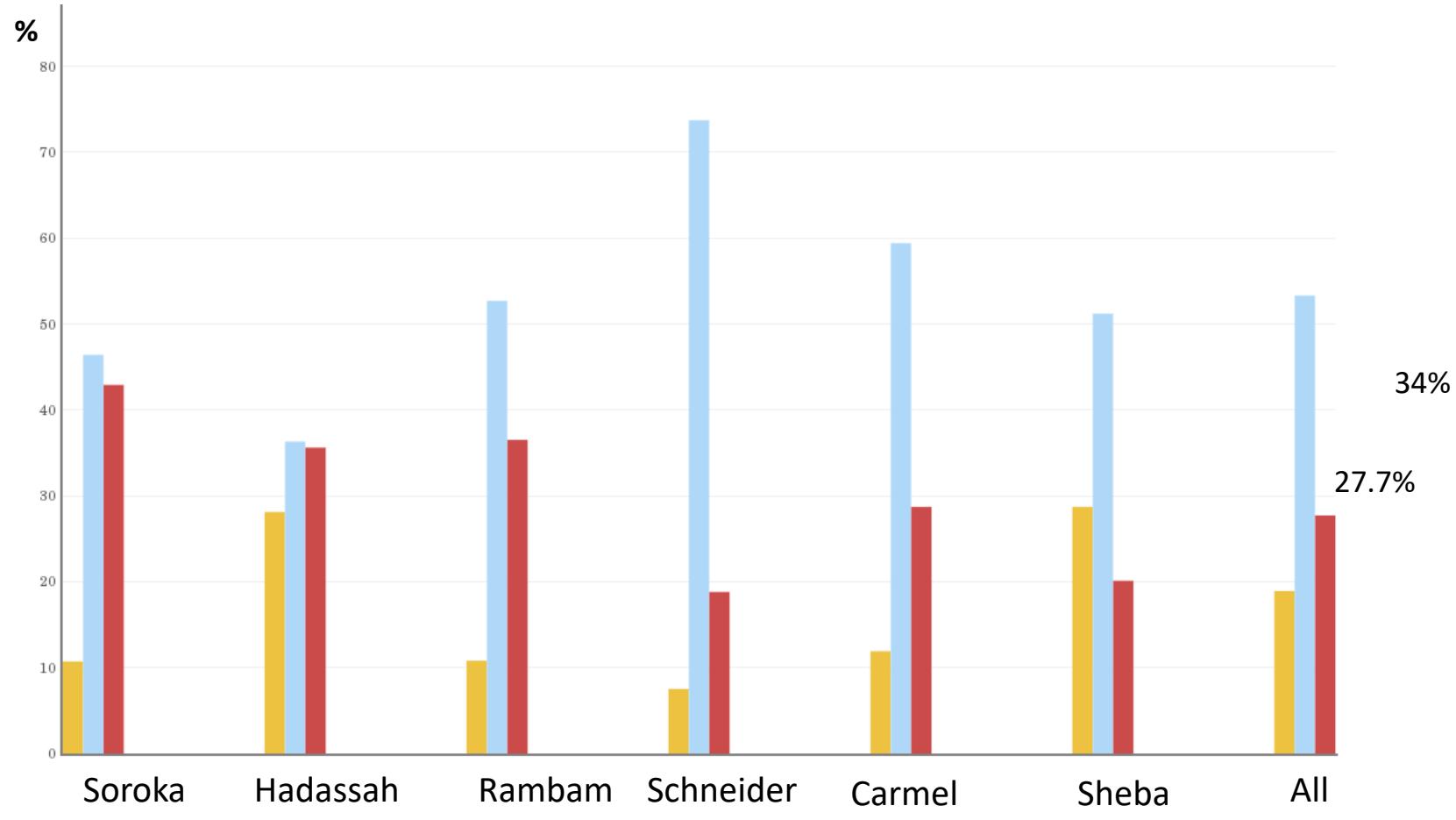


Ps. A in the US

2014



Staph. in Israel



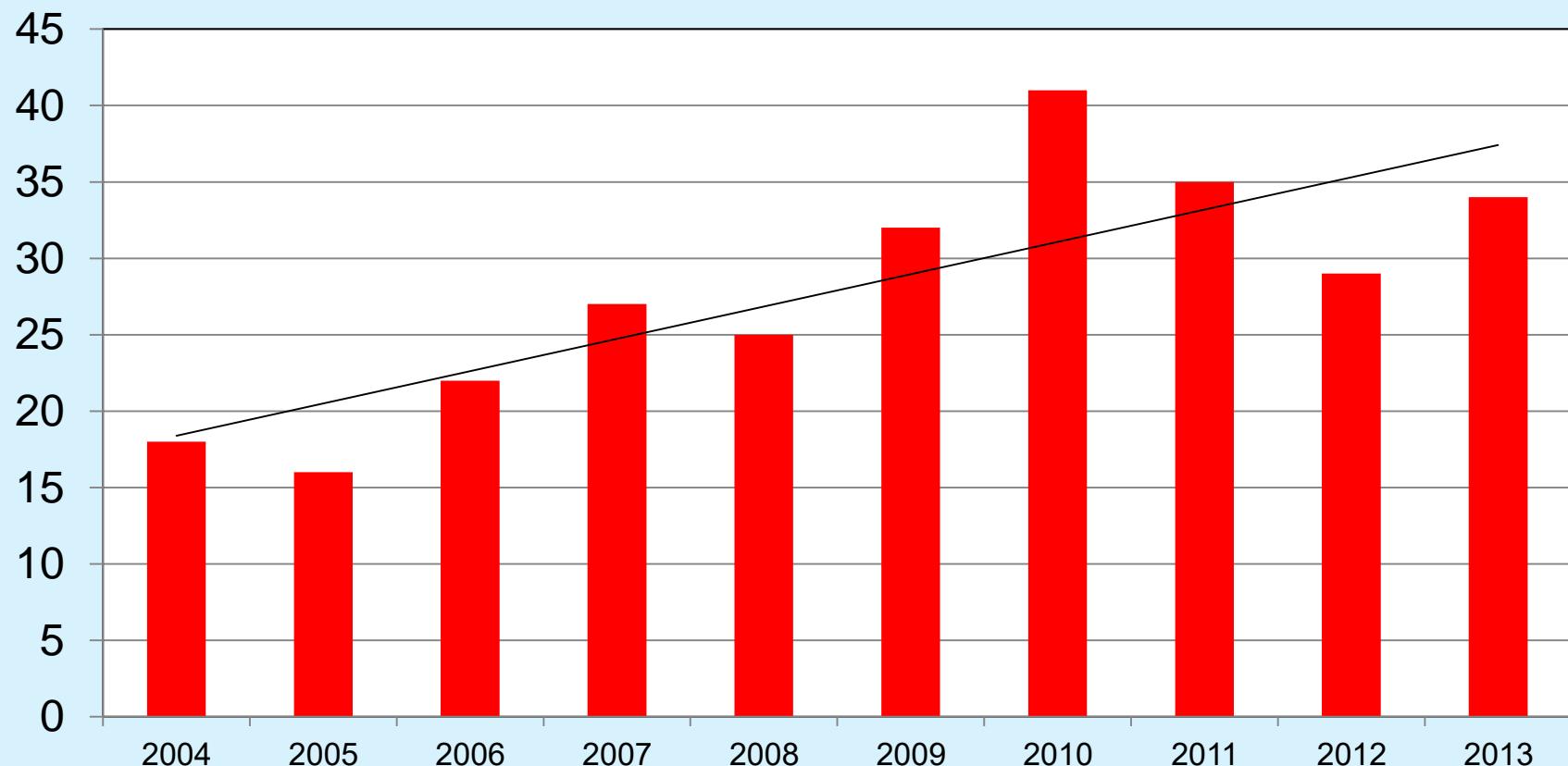
Report generated by: **Meir Mei-Zahav**

Report date: **Mon, 26 Oct 2015 16:35:02 GMT**

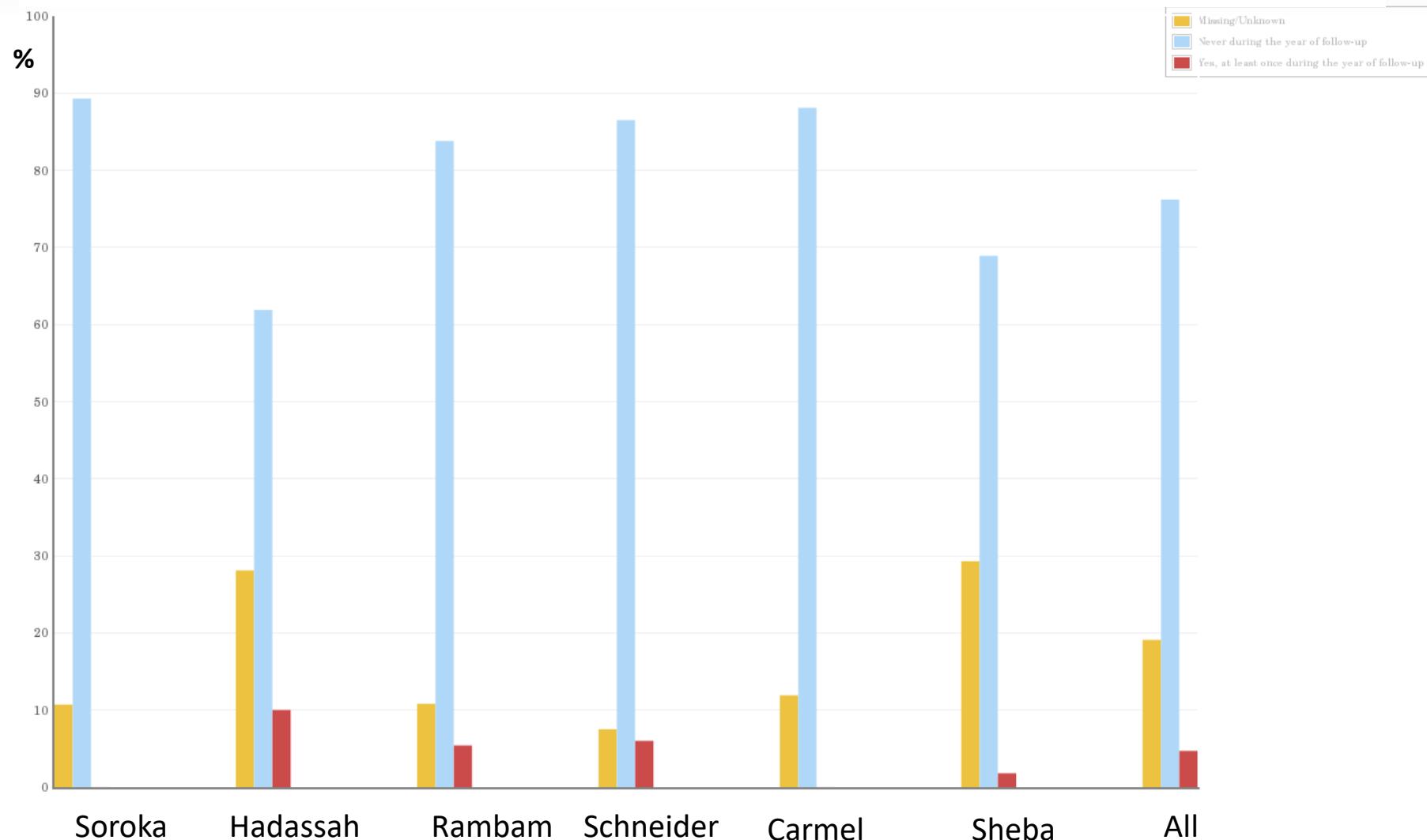
ECFSRP validation status: **No**

Staph. in Israel- Trend

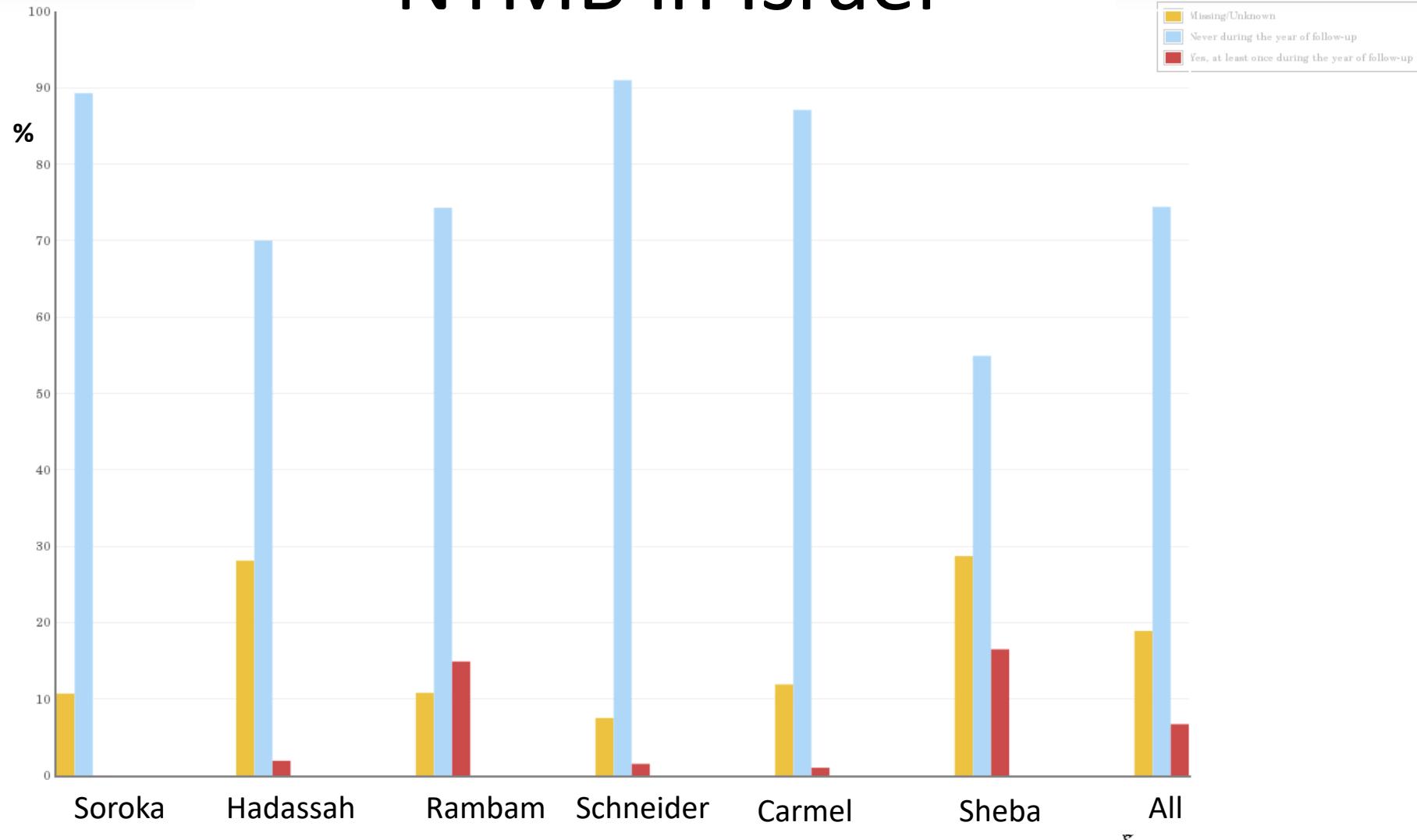
Staph %



Steno. in Israel



NTMB in Israel



Report generated by:

Meir Mei-Zahav

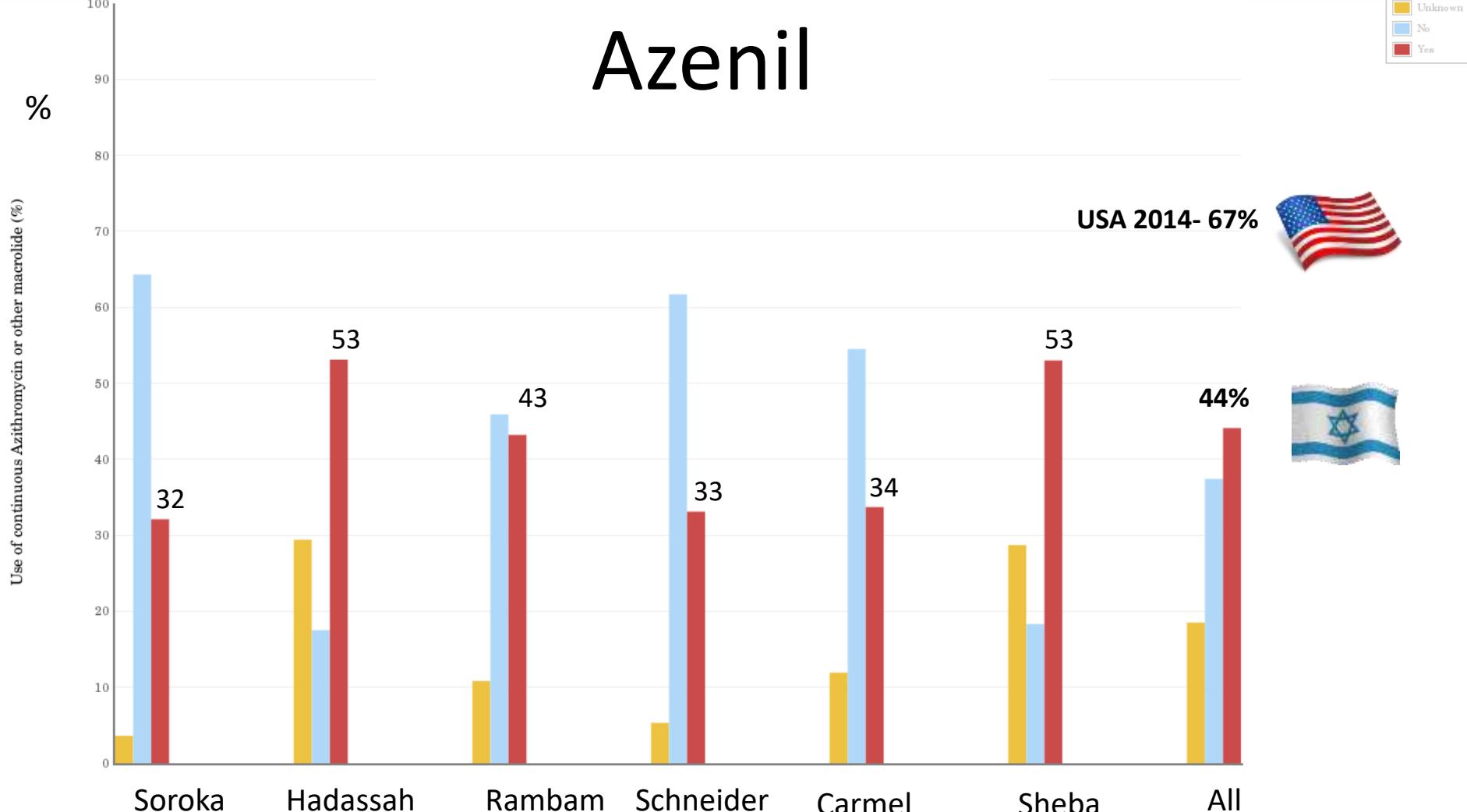
Report date:

Mon, 26 Oct 2015 16:33:40 GMT

ECFSPR validation status: **No**

Treatment

Azenil

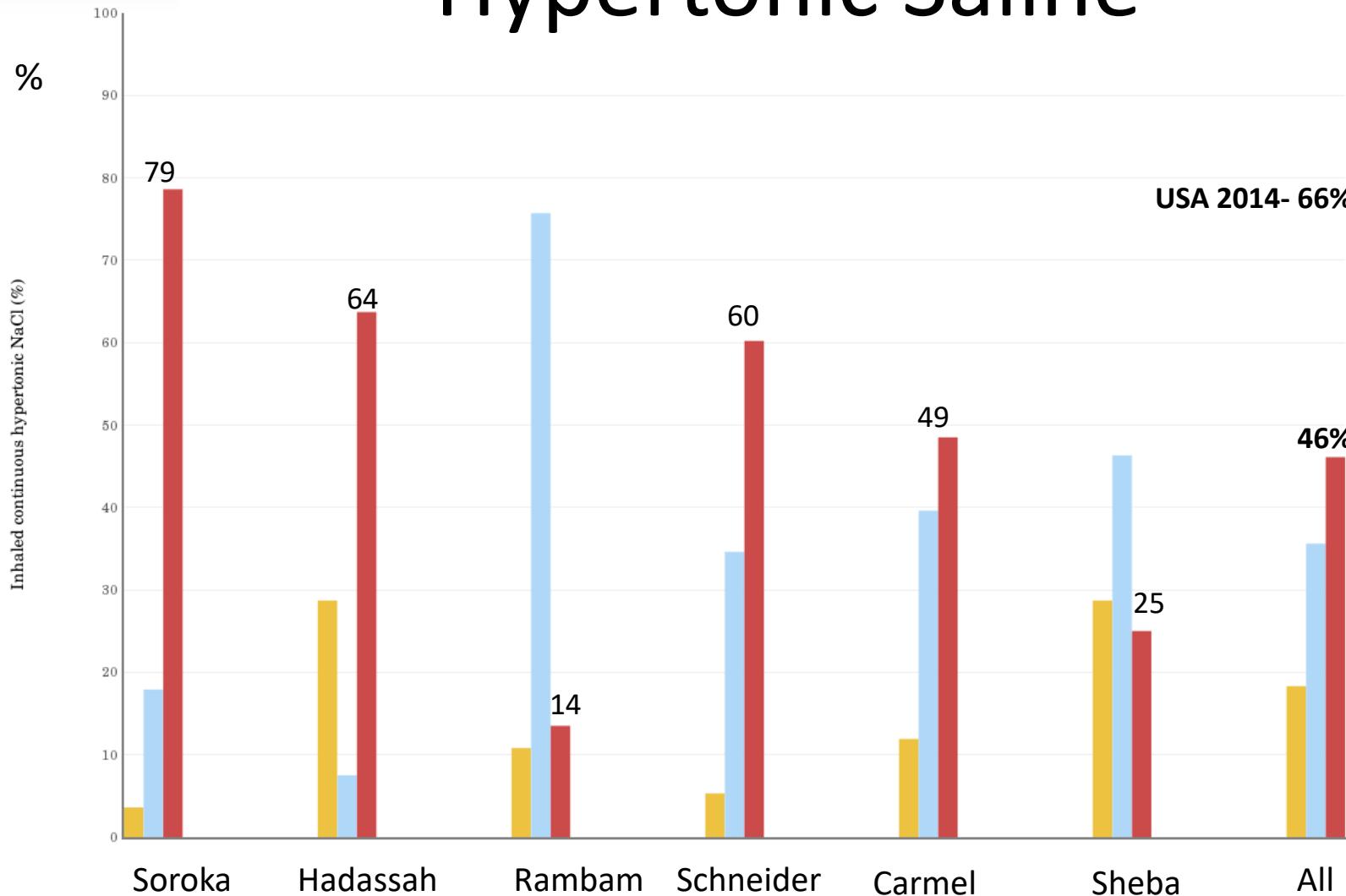


Report generated by: Meir Mei-Zahav

Report date: Mon, 26 Oct 2015 16:05:09 GMT

ECFSRP validation status: No

Hypertonic Saline



Report generated by:

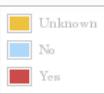
Meir Mei-Zahav

Report date:

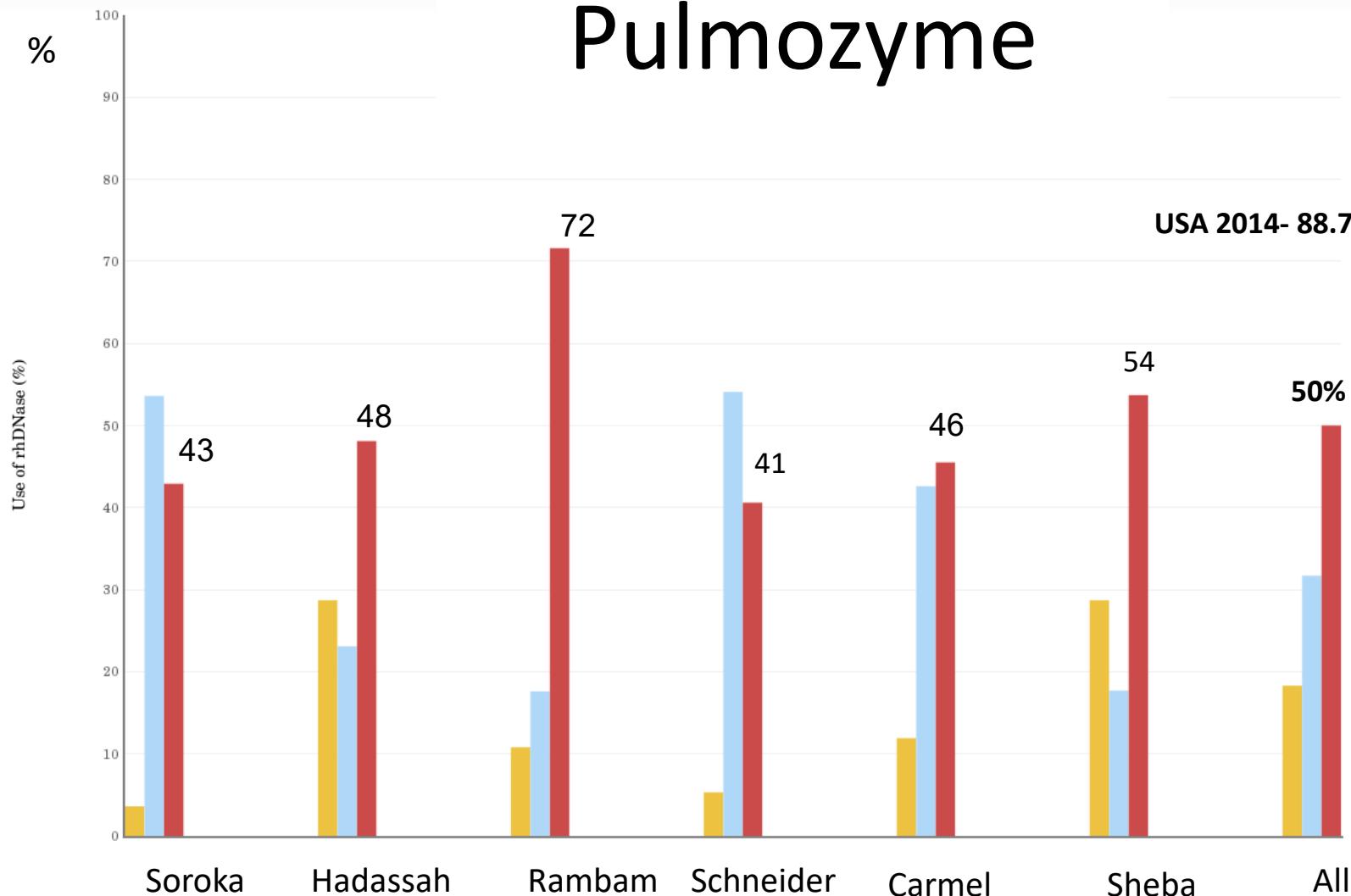
Mon, 26 Oct 2015 16:10:21 GMT

ECFSPR validation status: **No**

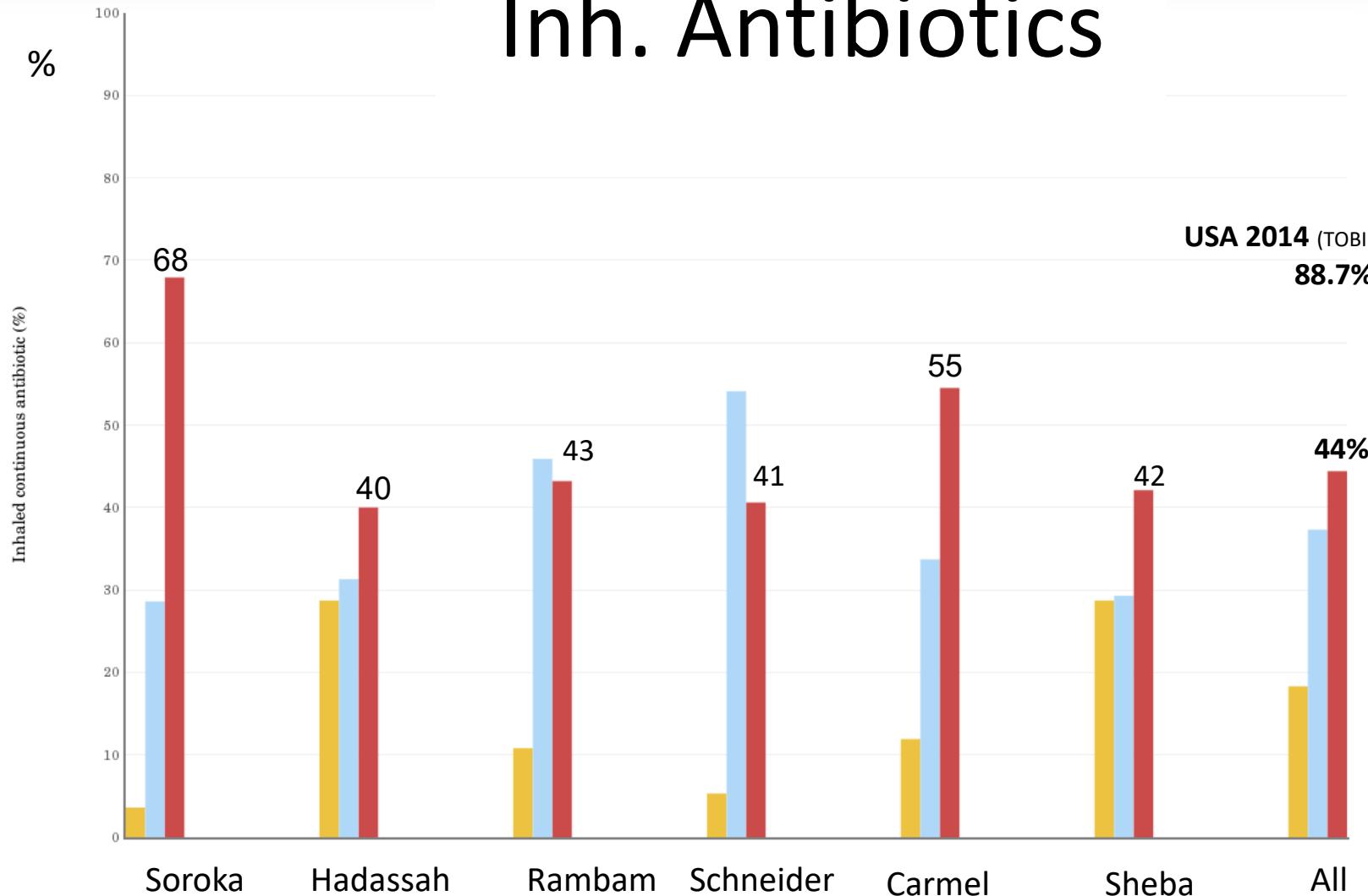
Pulmozyme



USA 2014- 88.7%



Inh. Antibiotics

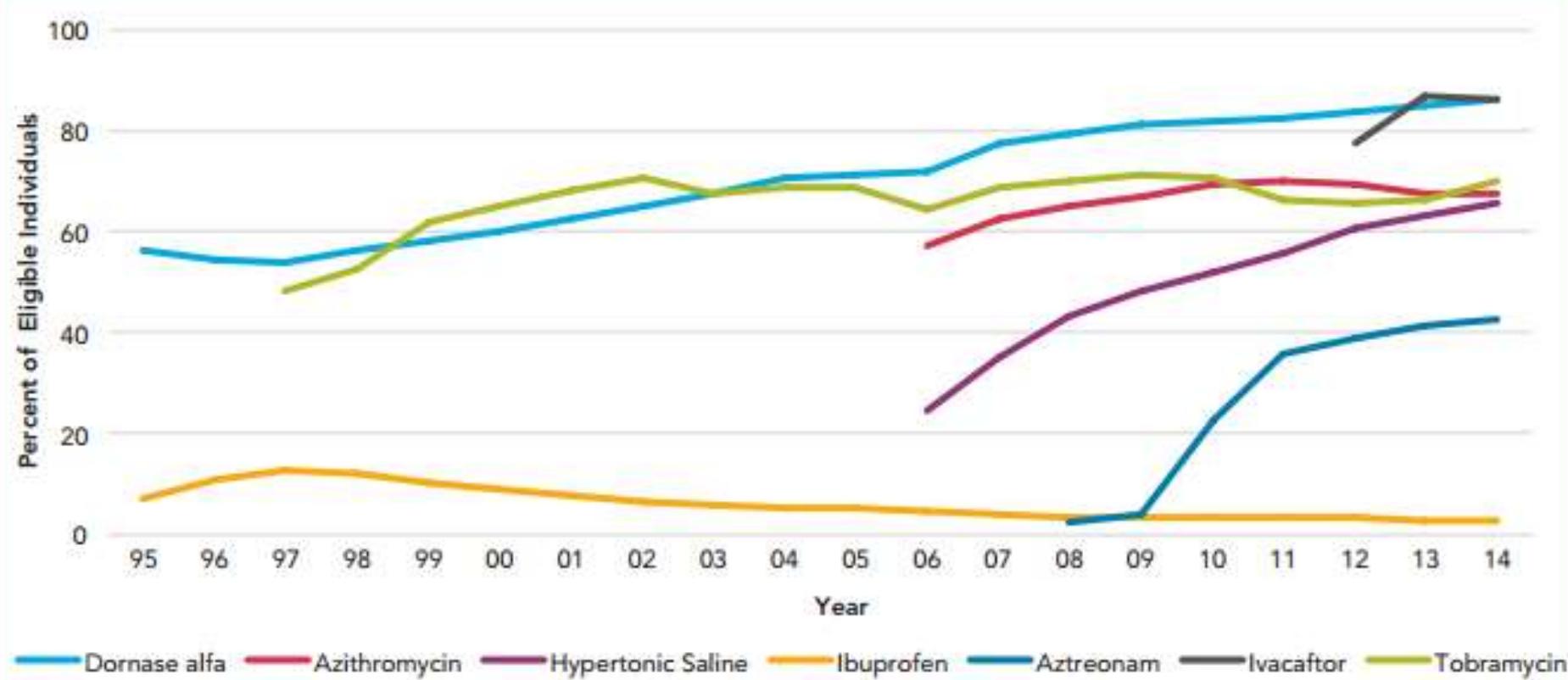


USA 2014 (TOBI)
88.7%



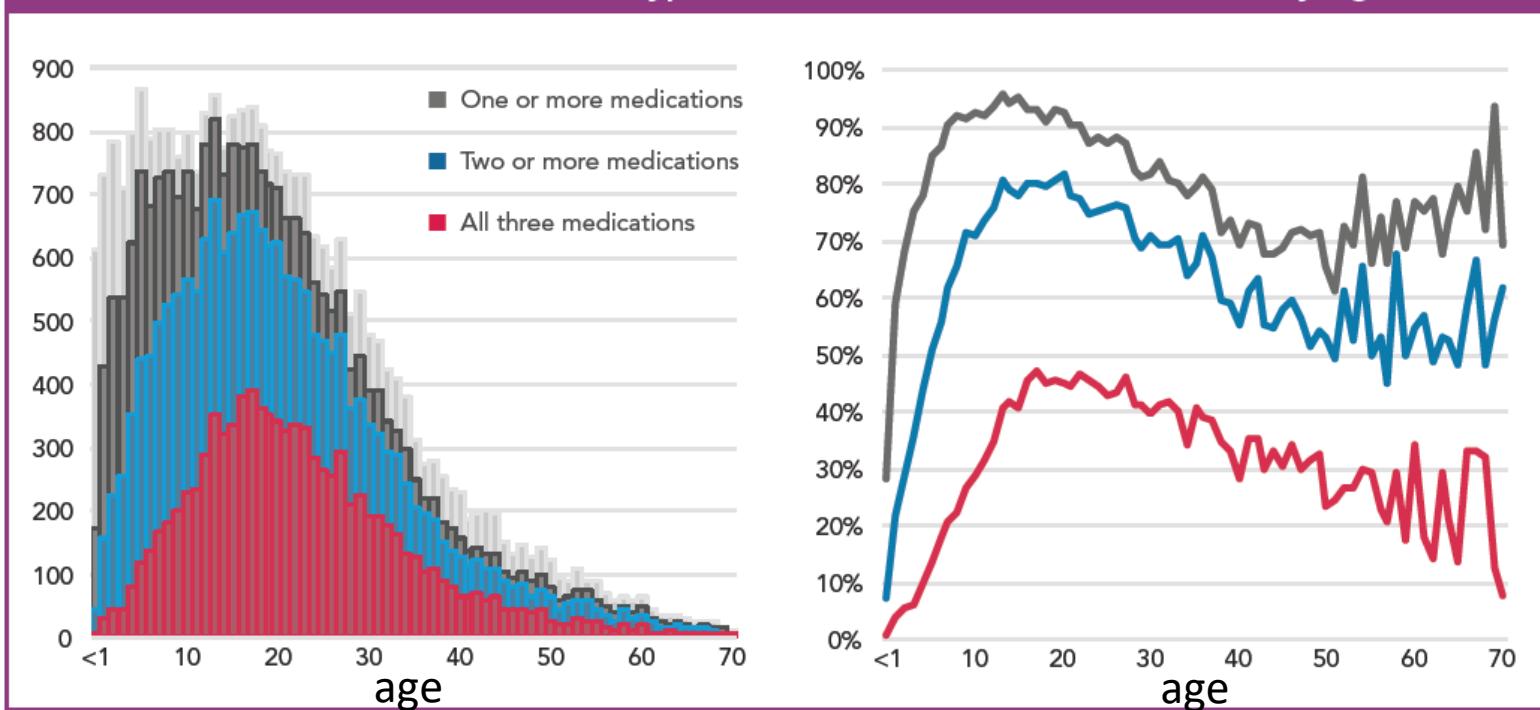
Use of Meds in the US

Chronic Medication Use in Eligible Individuals, 1995–2014



Use of Meds in the US

Inhaled Medication Use (Dornase Alfa, Hypertonic Saline, Inhaled Antibiotic) Use by Age, 2014



Complications

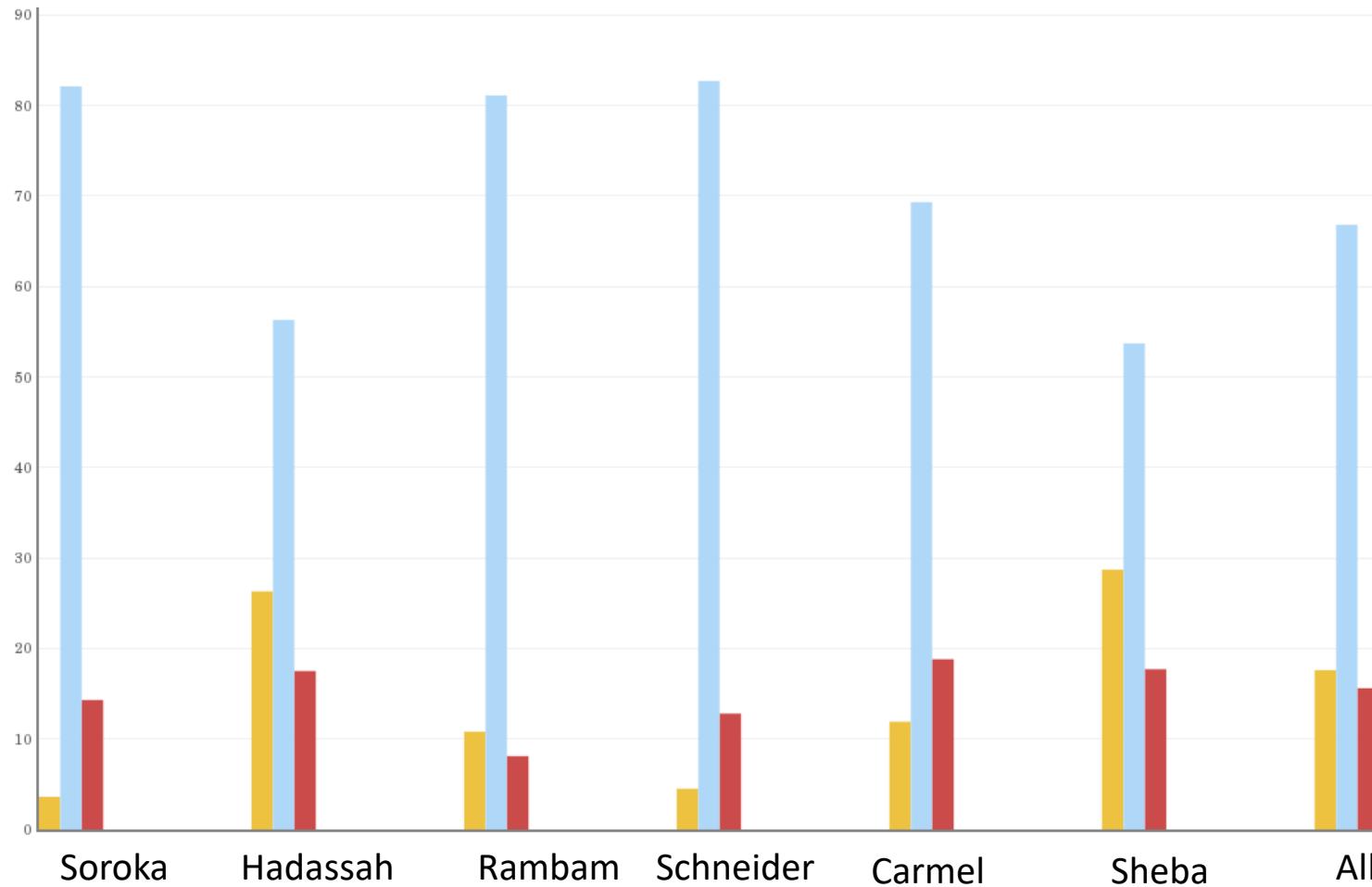
- ABPA- 28 (5%)
- Hemoptysis- 6 (1%)
- Diabetes (insulin treated)- 103 (19%)
 - Children- 15 (6%)
 - Adults- 88 (30%)
- Liver dis- 96 (18%)

All (except liver dis) are similar to the US 2014 data

CFRD

Unknown
No
Yes

Diabetes daily insulin treated (%)



Report generated by:

Meir Mei-Zahav

Report date:

Mon, 26 Oct 2015 16:46:45 GMT

ECFSPR validation status: **No**

Summary

- New friendly software
- CF in Israel is adults disease
- Pulmonary function are comparable to best European and US centers
- PsA is a major pathogen starting in childhood, but prevalence trend ↓ last years
- Staph prevalence is rising
- BMI is within the normal range but wide variability

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