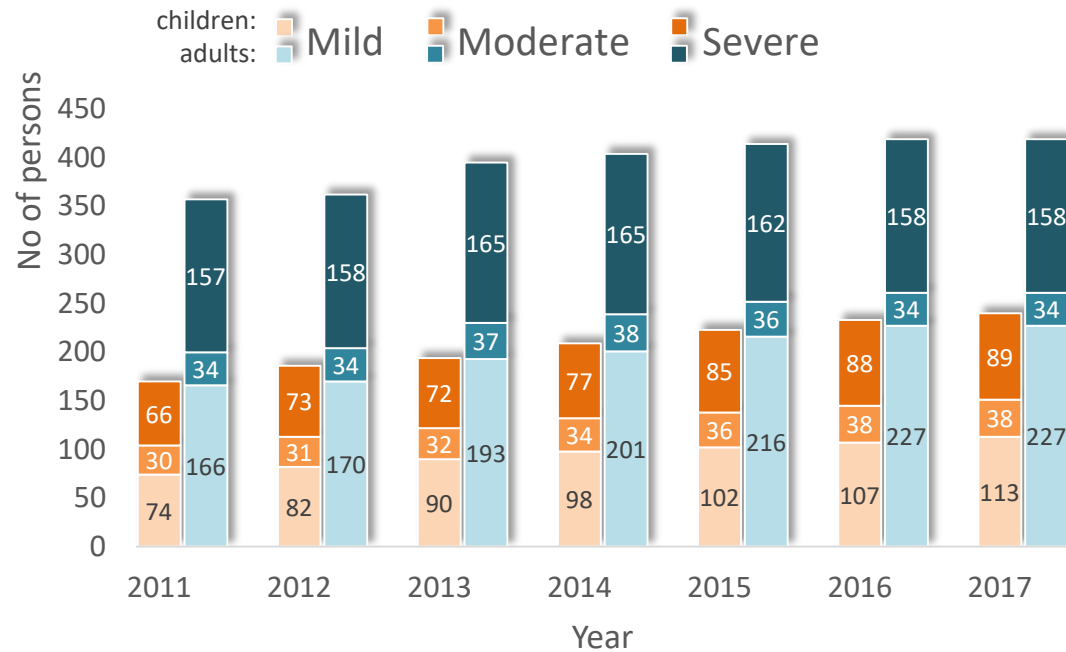


Trends in the haemophilia treatment 2011-2017

ČNHP data, January 2019

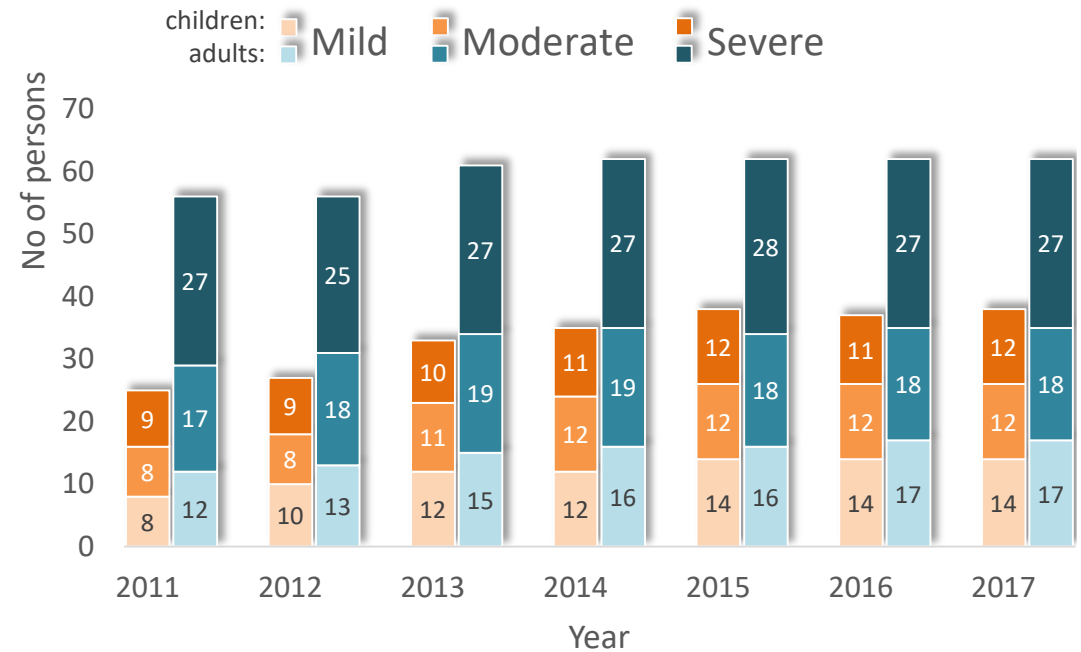
Number of persons registered in the CNHP in time According to the type, severity and age

Haemophilia A



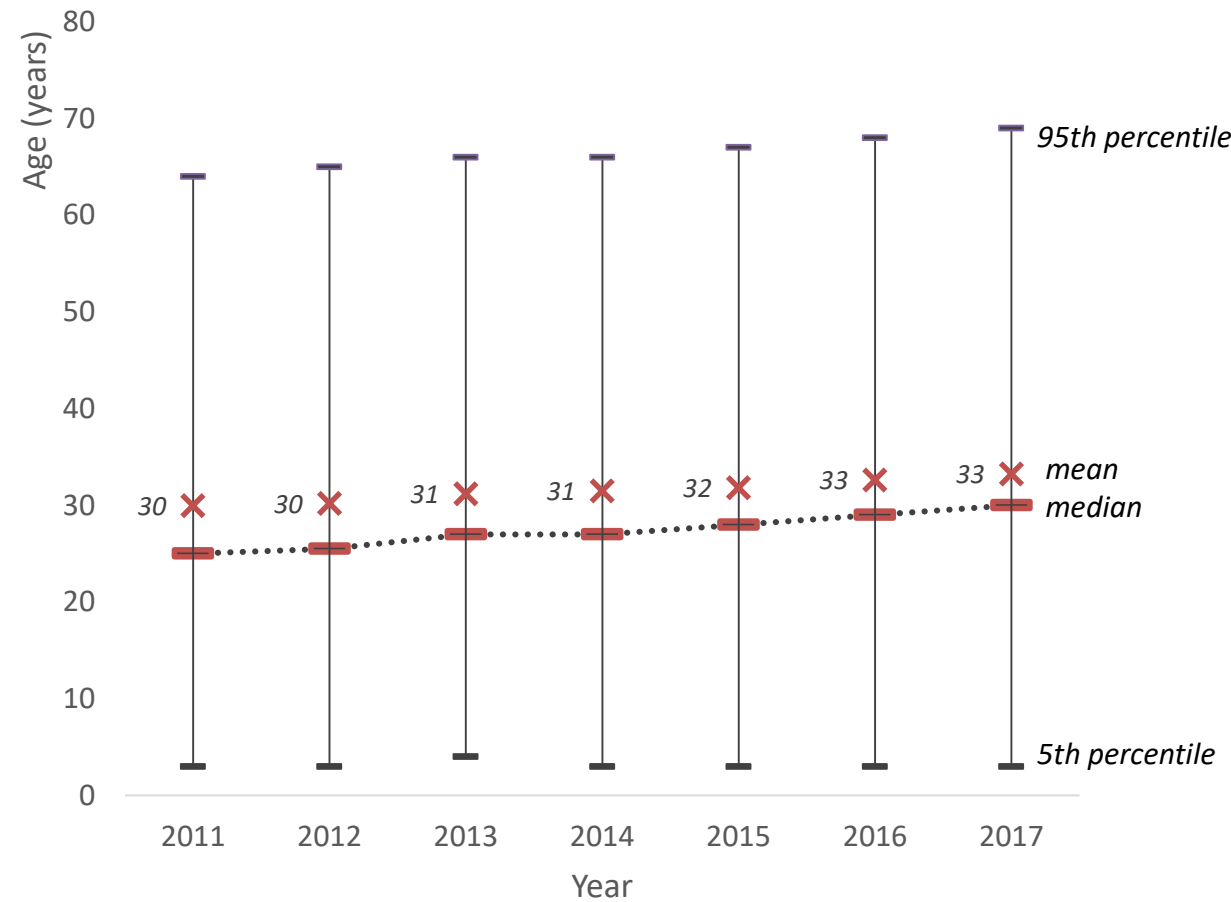
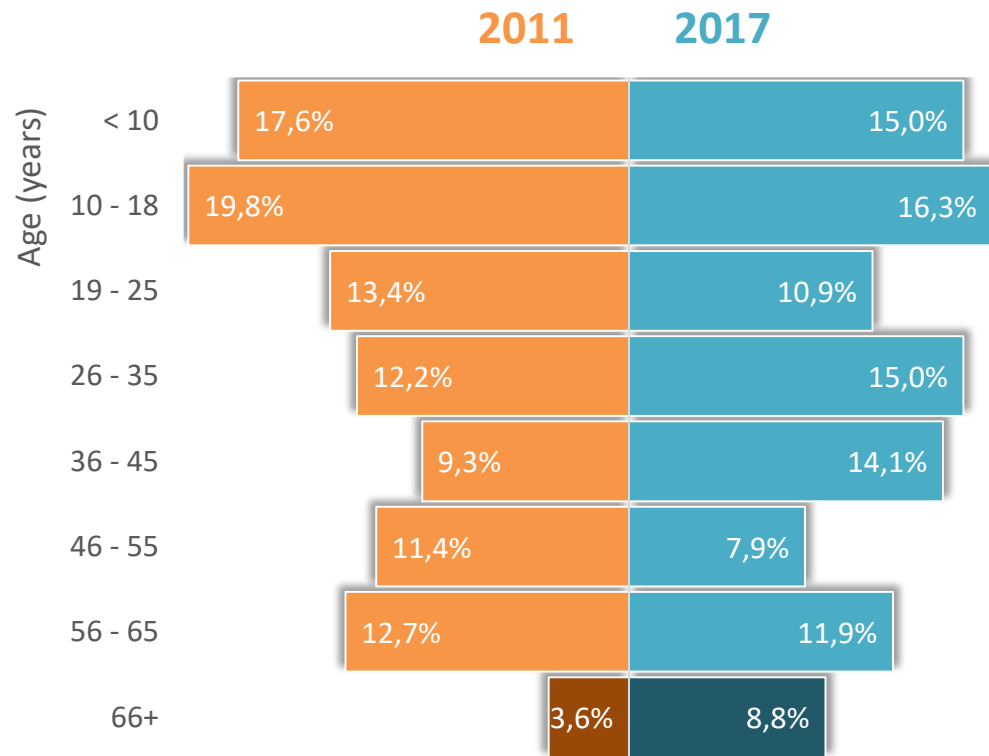
936 males suffered from haemophilia A at the end of 2017, i.e. about 1.79 patients per 10,000 men in the Czech Republic. 659 (70%) of them are registered within the CNHP registry.

Haemophilia B



141 males suffered from haemophilia B at the end of 2017, i.e. about 0.27 patients per 10,000 men in the Czech Republic. 100 (71%) of them are registered within the CNHP registry.

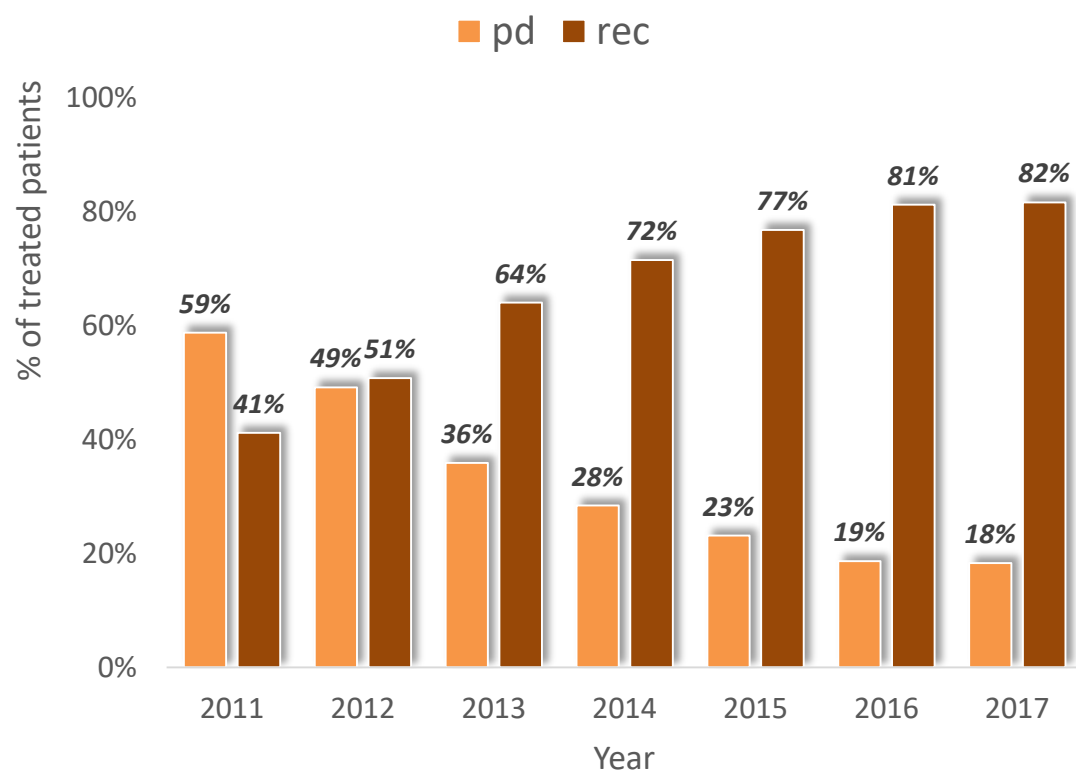
Age of persons with haemophilia



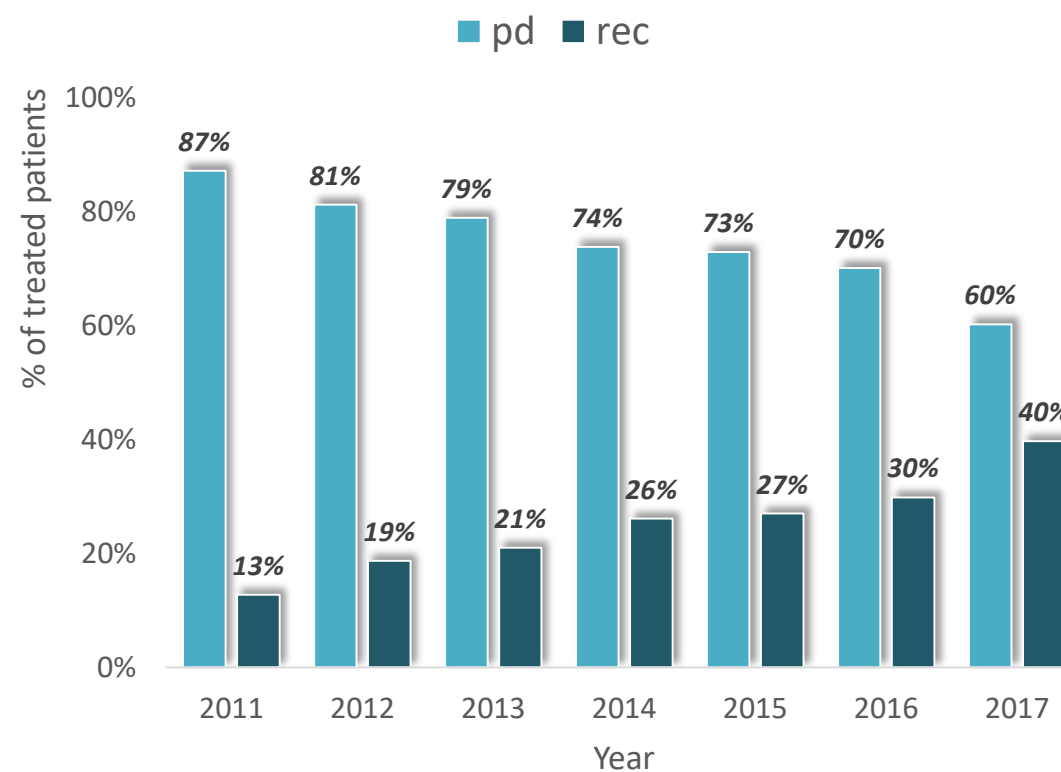
Percentage of persons older than 65 years was 3.6% in 2011, whereas 8.8% in 2017. Mean changed from 30 to 33 years of age within this period.

Type of the treatment (treated patients only)

Children



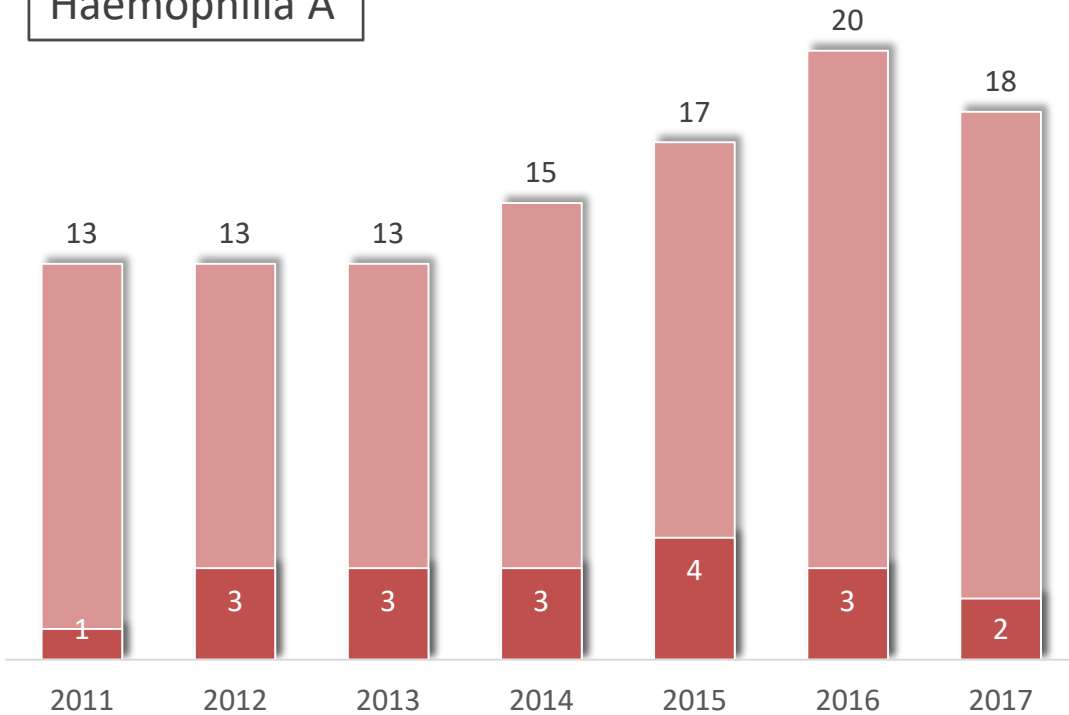
Adults



Inhibitors in patients with haemophilia

■ Number of inhibitors ■ Newly developed

Haemophilia A



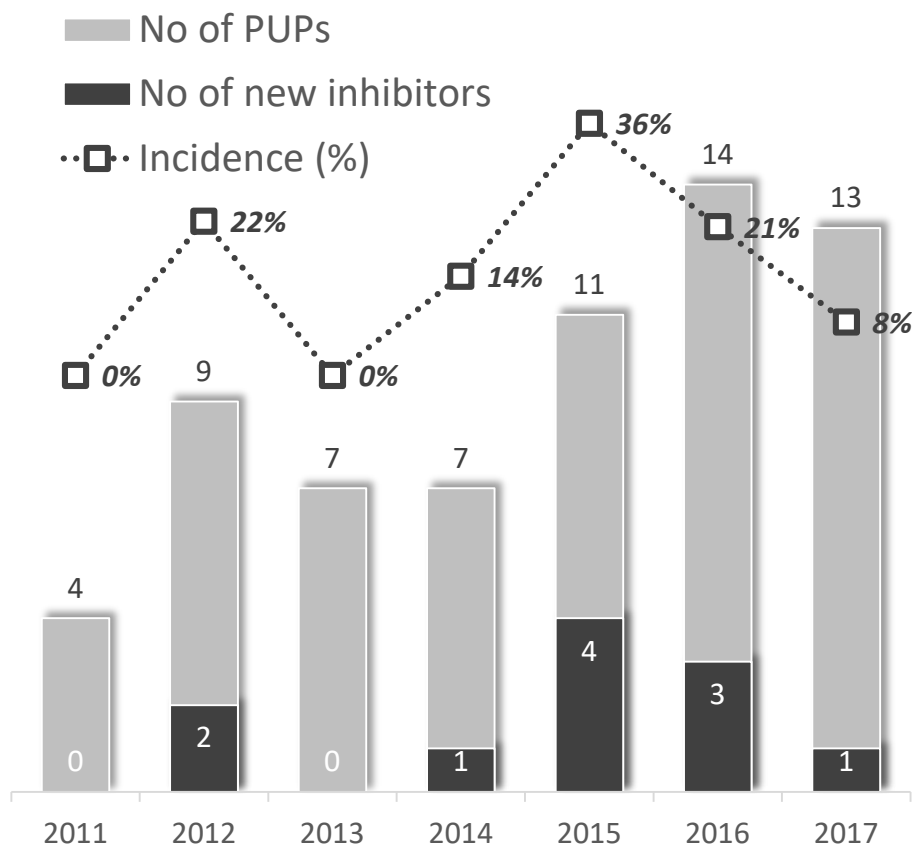
Inhibitors were present in 31 PWHA and in 1 PWHB between years 2011 and 2017

- 29 in severe, 1 in moderate and 2 in mild haemophilia
- 21 high-titre ($\geq 5\text{BU}$) and 10 low-titre (N/A in one patient)
- **18 inhibitors historically developed in patients treated with plasma-derived factors and 14 inhibitors developed in patients treated with recombinant factors**

Out of 31 PWHA, inhibitors newly developed in 19 of them during this period

Inhibitors in PUPs with severe haemophilia A

Number of inhibitors developed in PUPs with severe haemophilia A (PUP – up to 50 ED)



Inhibitors developed in 11 PUPs with severe haemophilia A between 2011 and 2017

- 8 high-titre (≥5BU) and 3 low-titre
- **1 inhibitor developed in patient treated with plasma-derived factors and 10 inhibitors developed in patients treated with recombinant factors**

*The incidence rate of inhibitors in PUPs with severe haemophilia A was **0.169** (11 new inhibitors per 65 patient-years), i.e. **16.9 new inhibitors per 100 patient-years** (compared to incidence rate **5.6 new inhibitors per 100 patient-years between years 2003-2013***).*

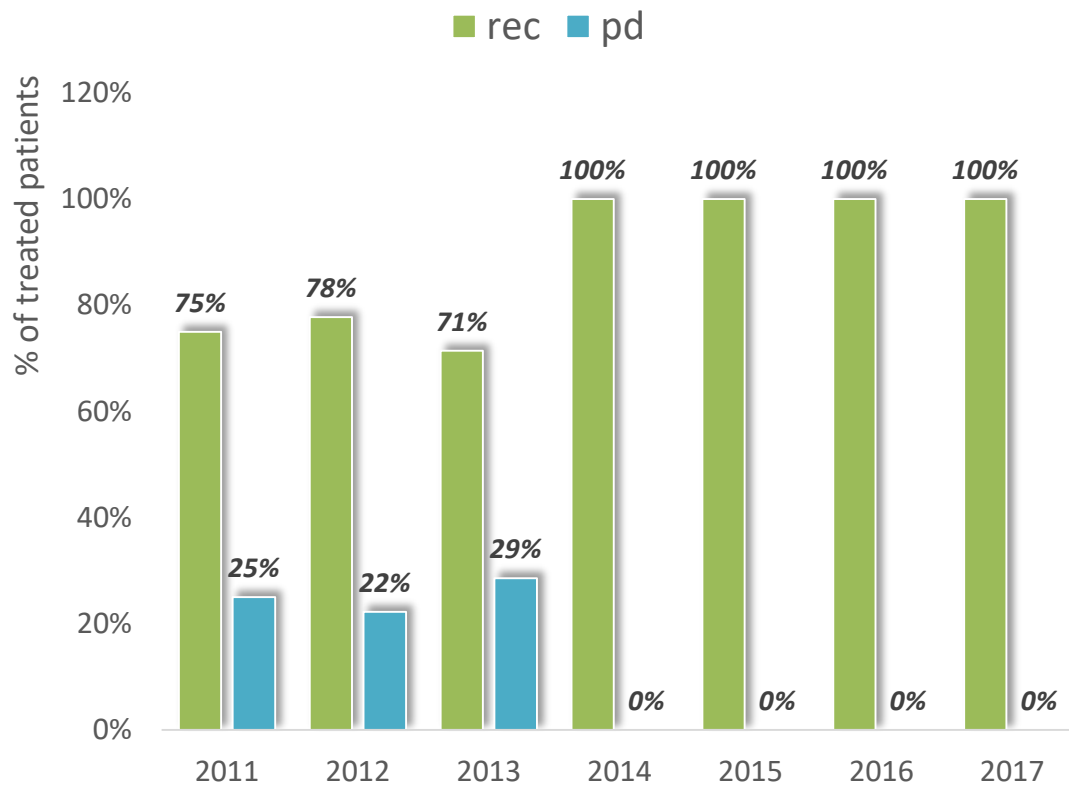
*The absolute incidence of inhibitors in PUPs with severe haemophilia A was **33%** (11 new inhibitors in 33 PUPs), compared to **26% between years 2003-2013***.*

* Blatny J et al. Inhibitors incidence rate in Czech previously untreated patients with haemophilia A has not increased since introduction of recombinant factor VIII treatment in 2003. Blood Coagul Fibrinolysis. 2015 Sep;26(6):673-8.

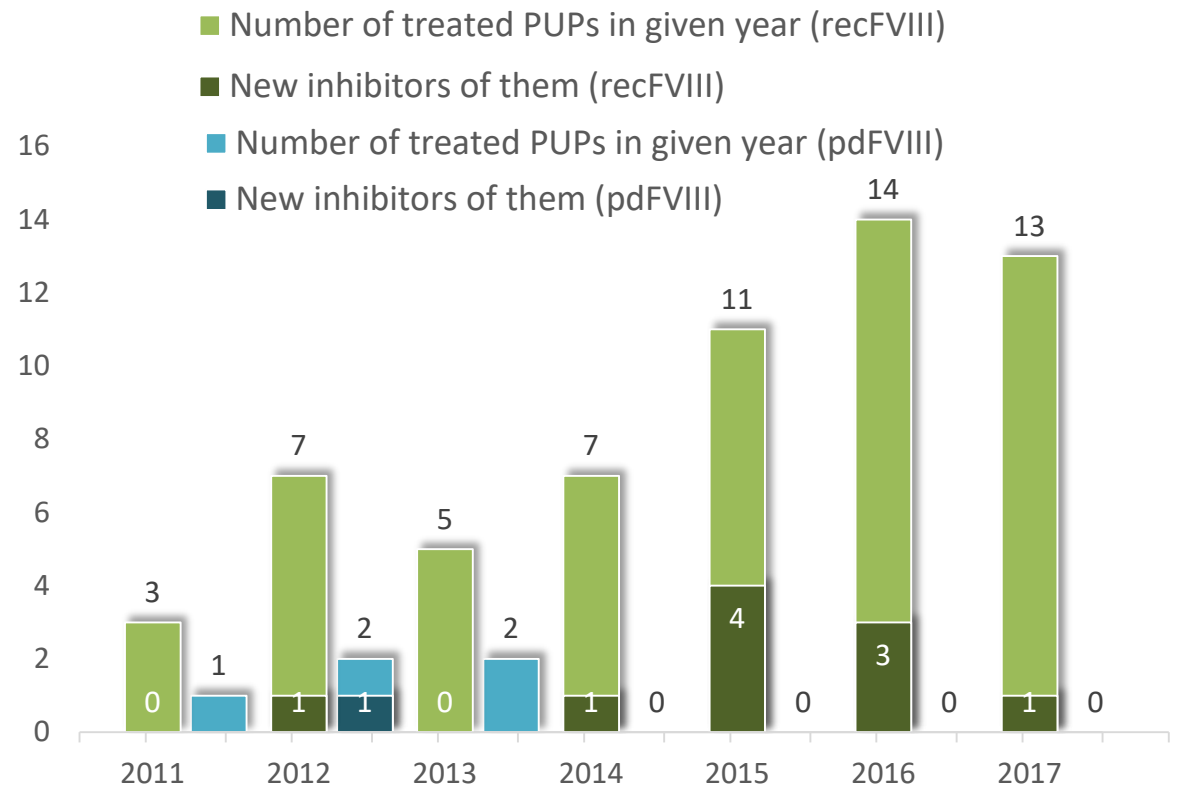
Type of treatment in PUPs with severe haemophilia A (only treated patients)

Severe Haemophilia A
PUPs (up to 50 ED) born ≥ 2010

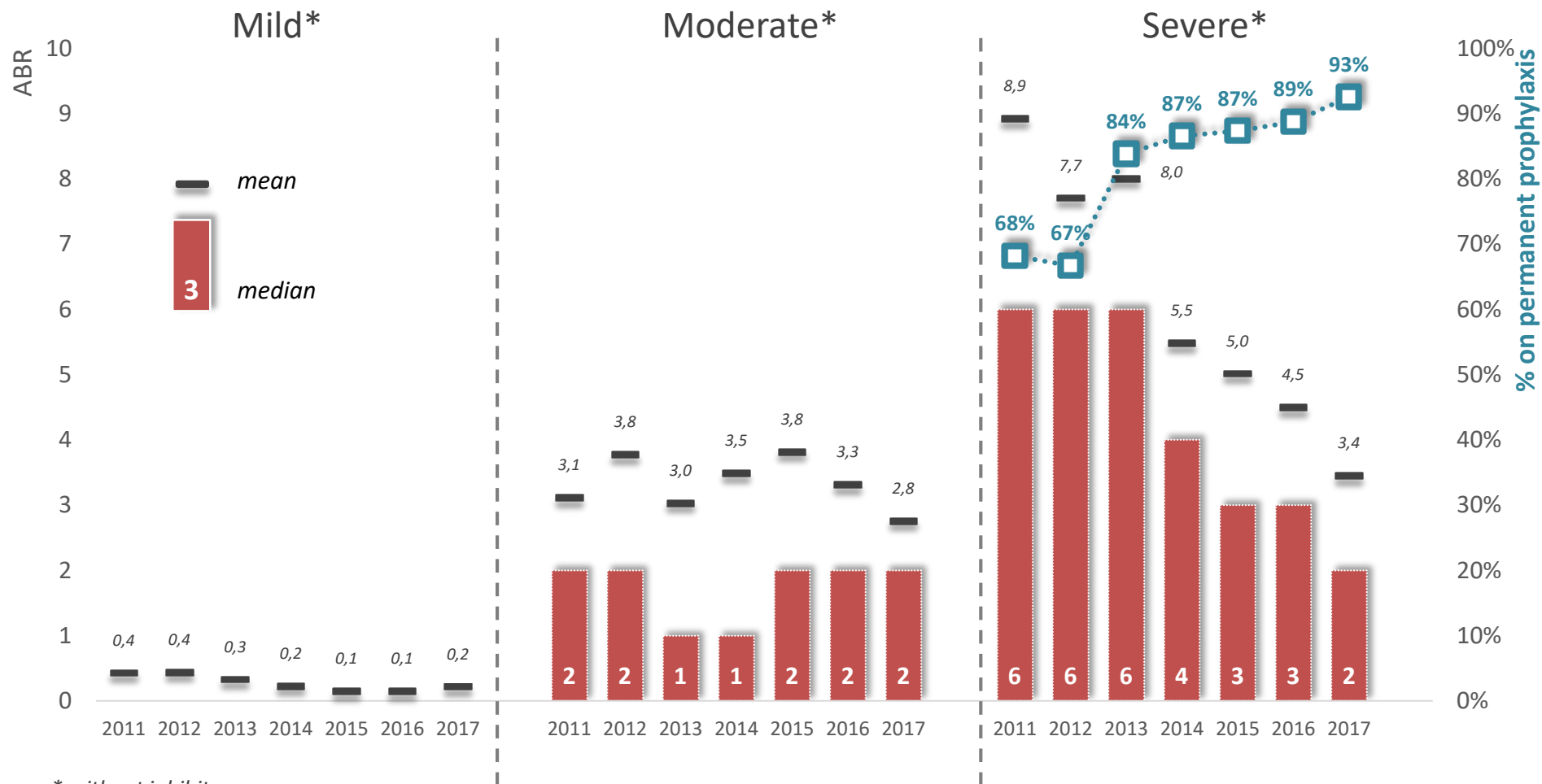
Children



New inhibitors in PUPs with severe Haemophilia A

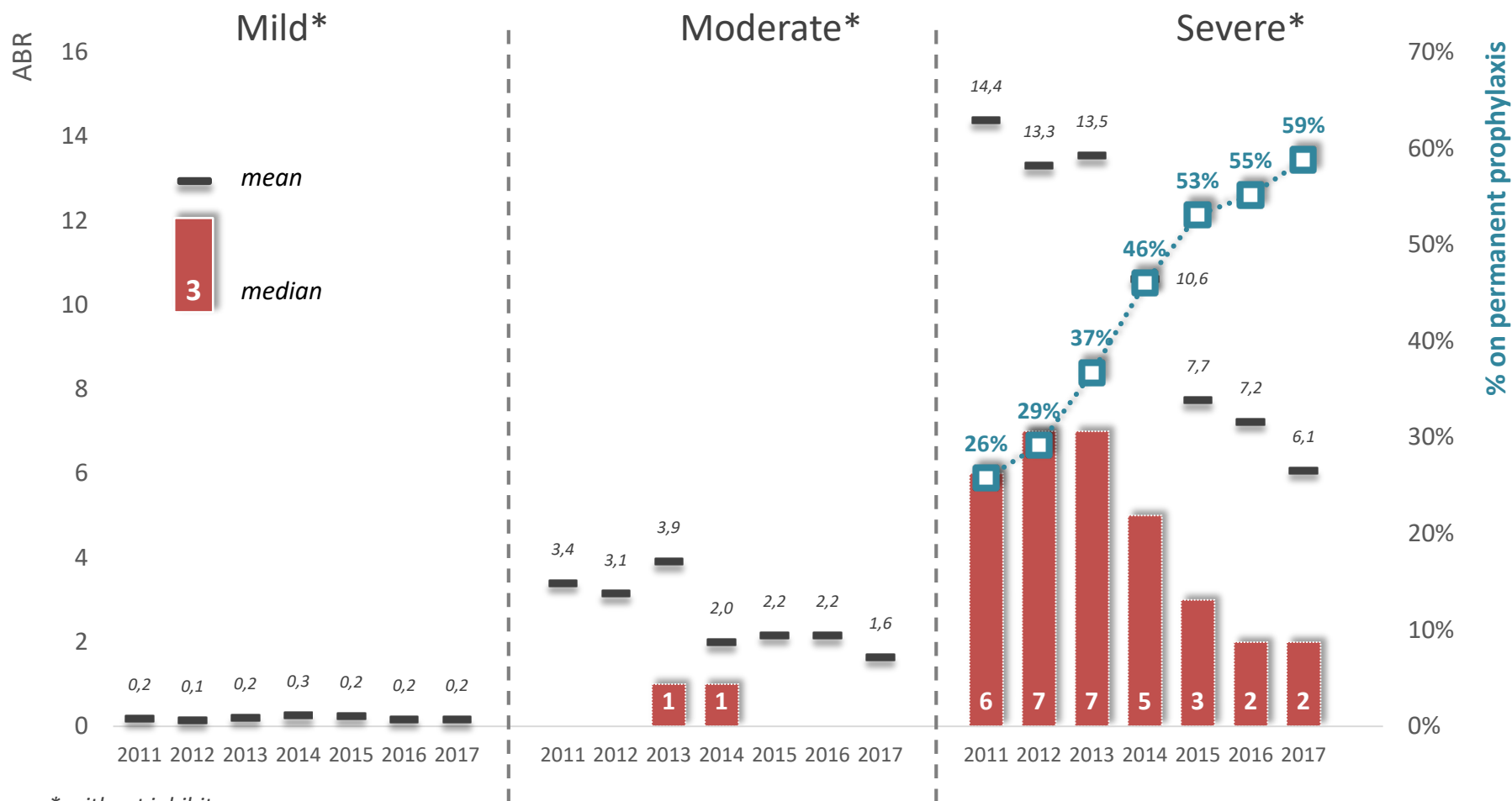


Children: change of ABR in time According to the severity of haemophilia



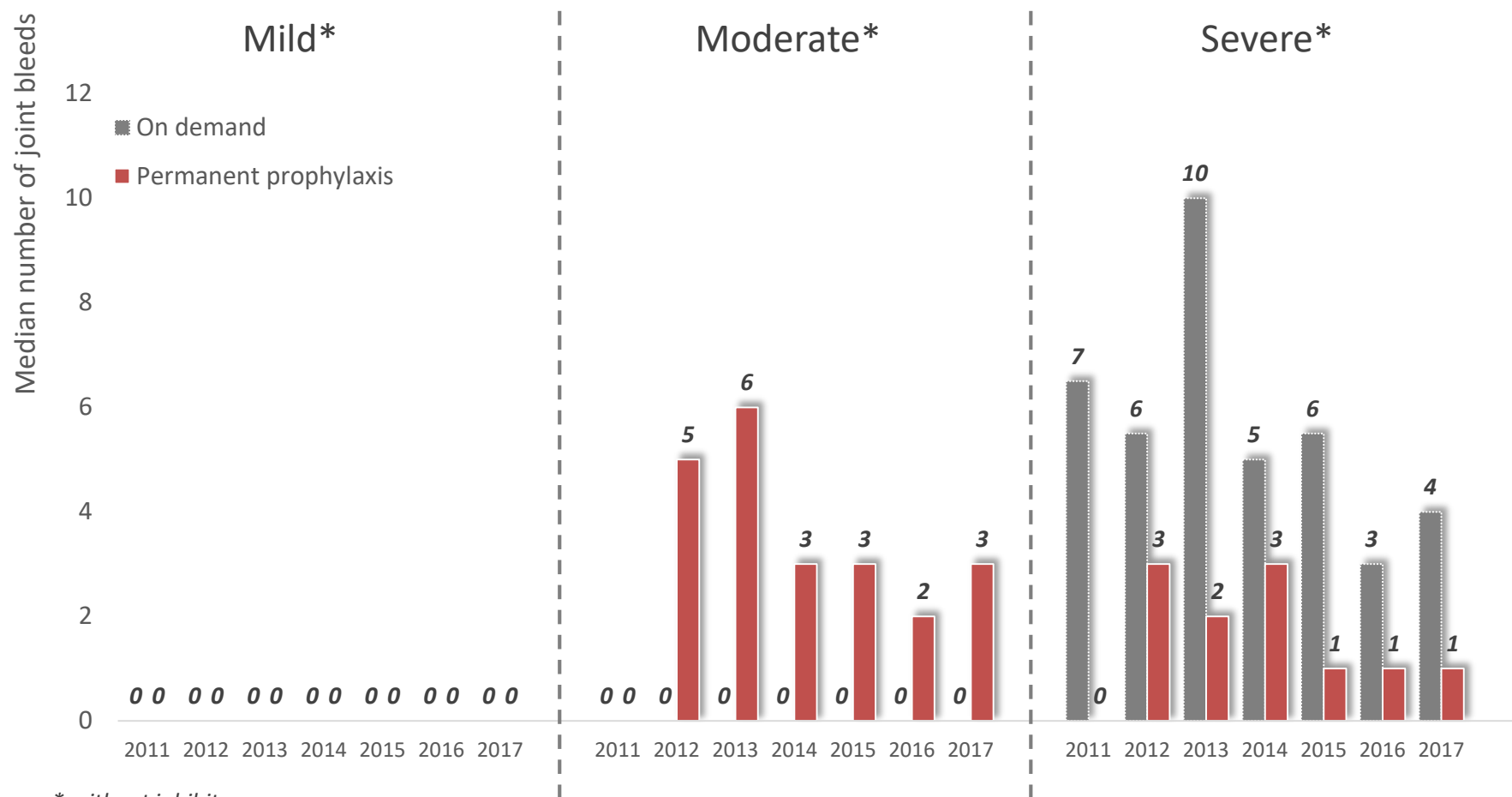
* without inhibitor

Adults: change of ABR in time According to the severity of haemophilia



* without inhibitor

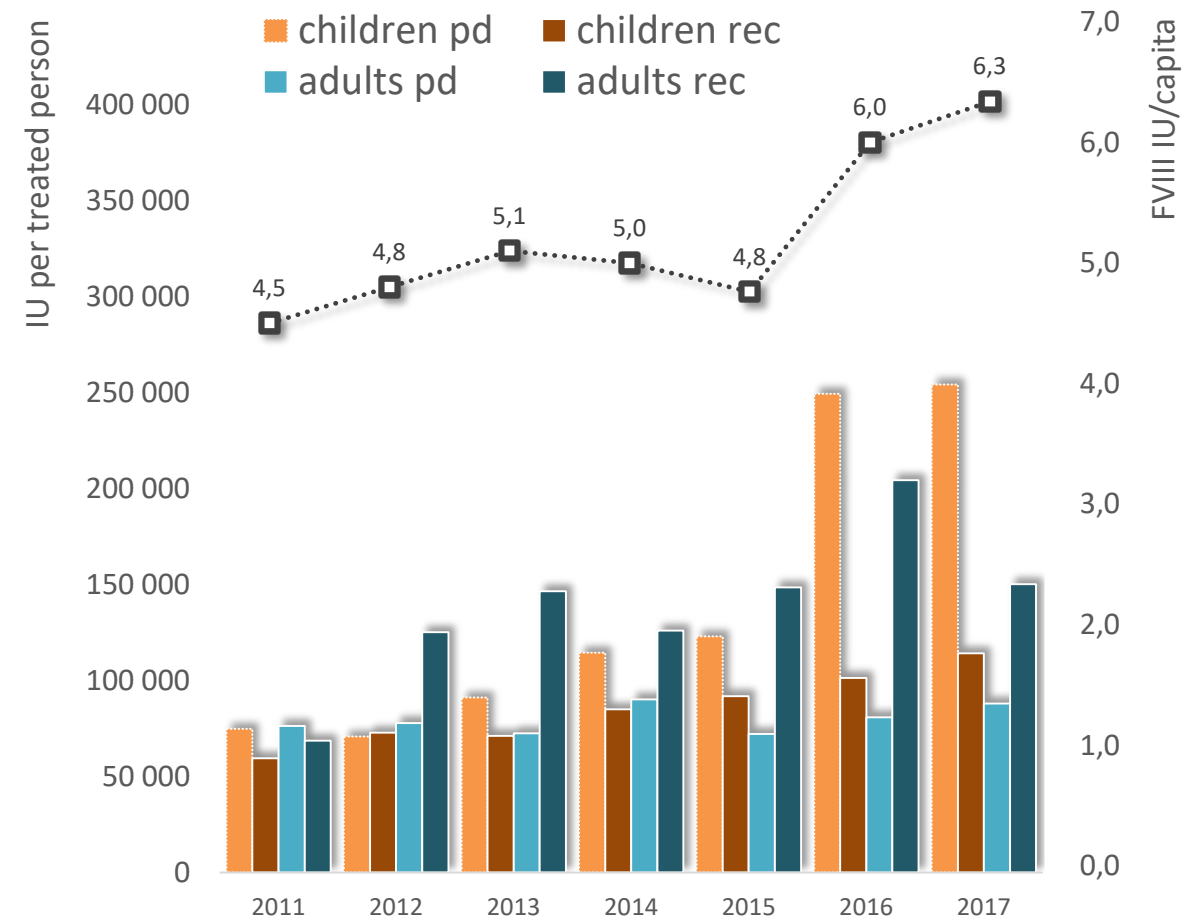
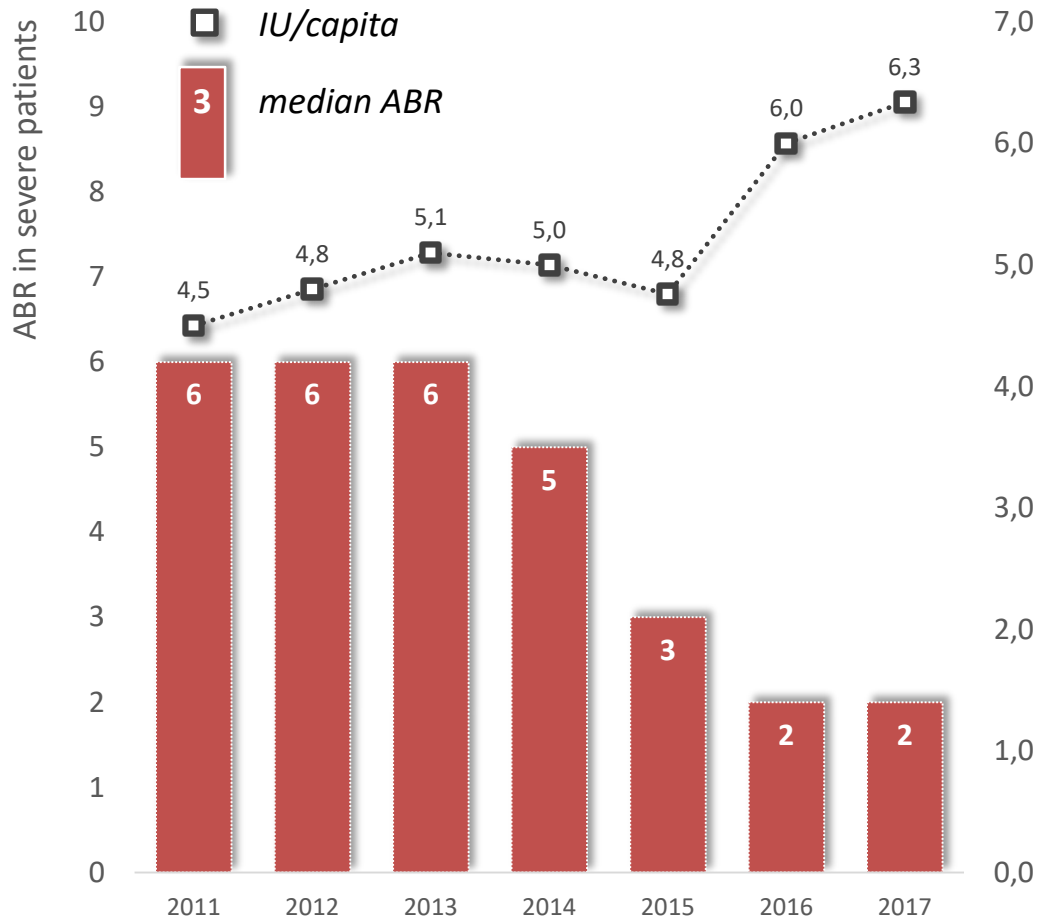
Adults: change of JOINT ABR according to the type of the treatment and severity



* without inhibitor

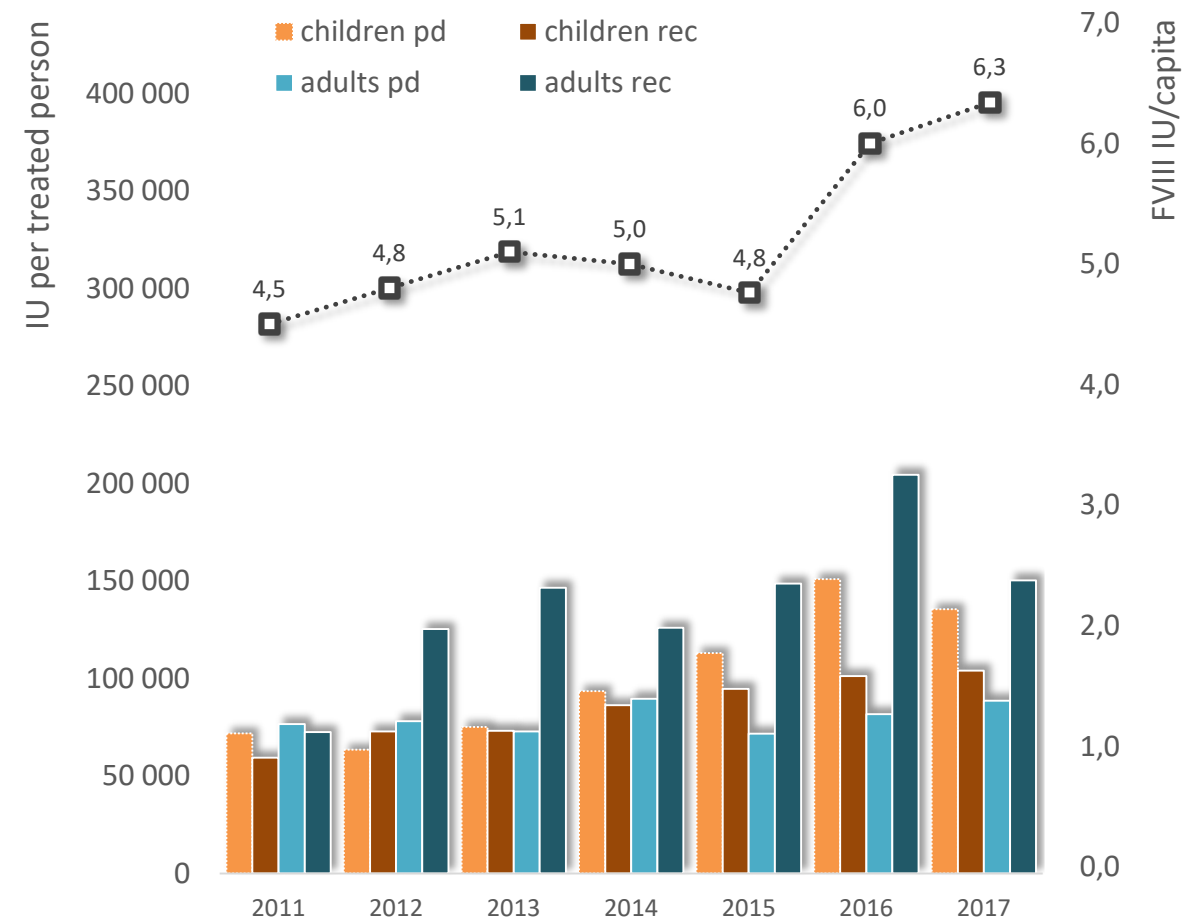
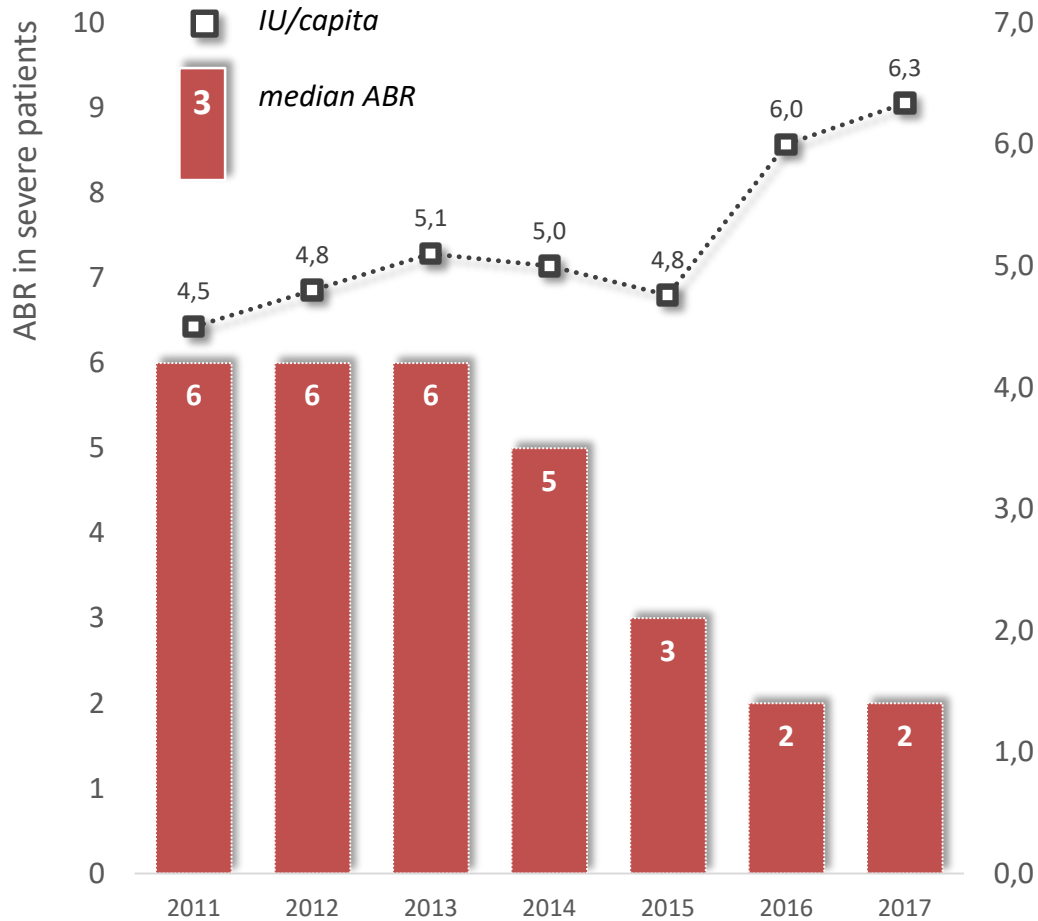
Consumption of FVIII in relation to ABR (including inhibitors and ITT)

Haemophilia A
(all severities incl. inh.)
Children+Adults



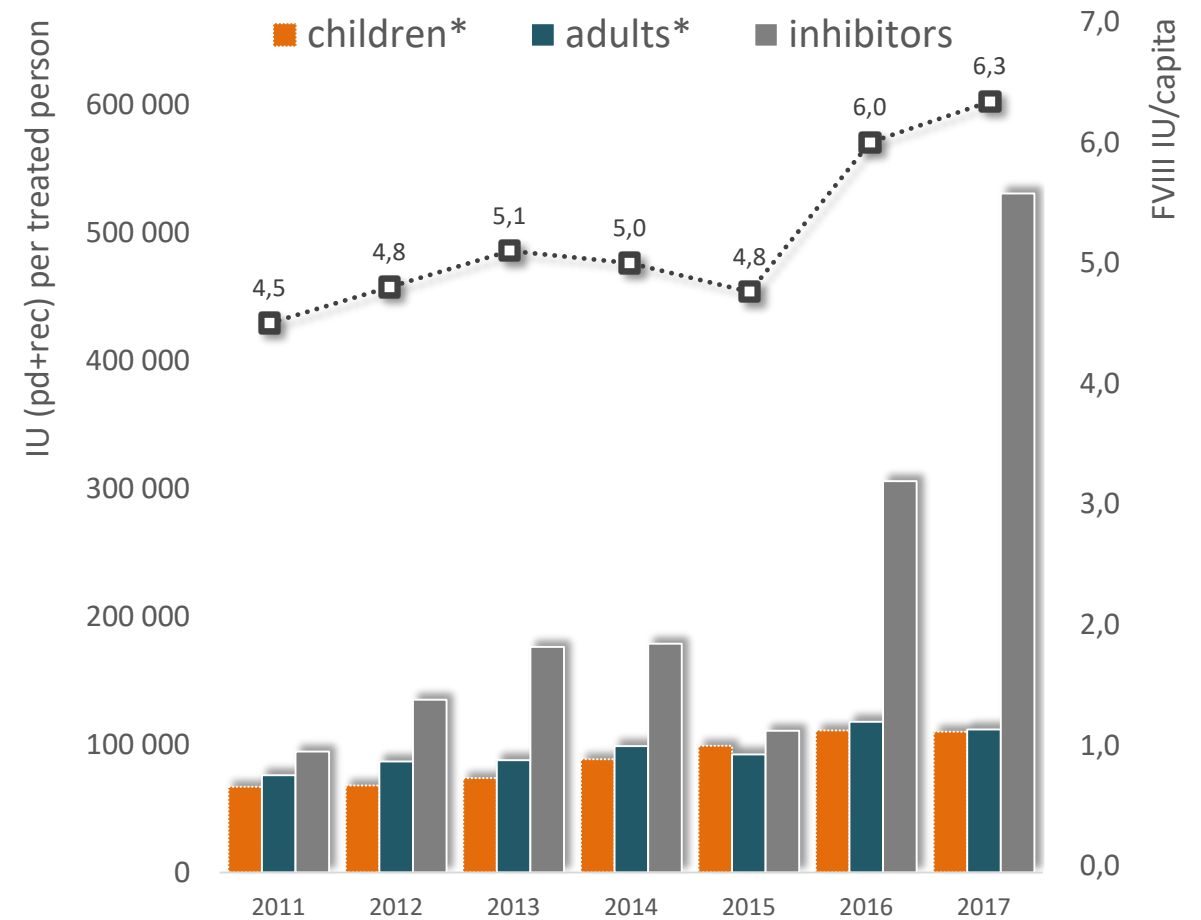
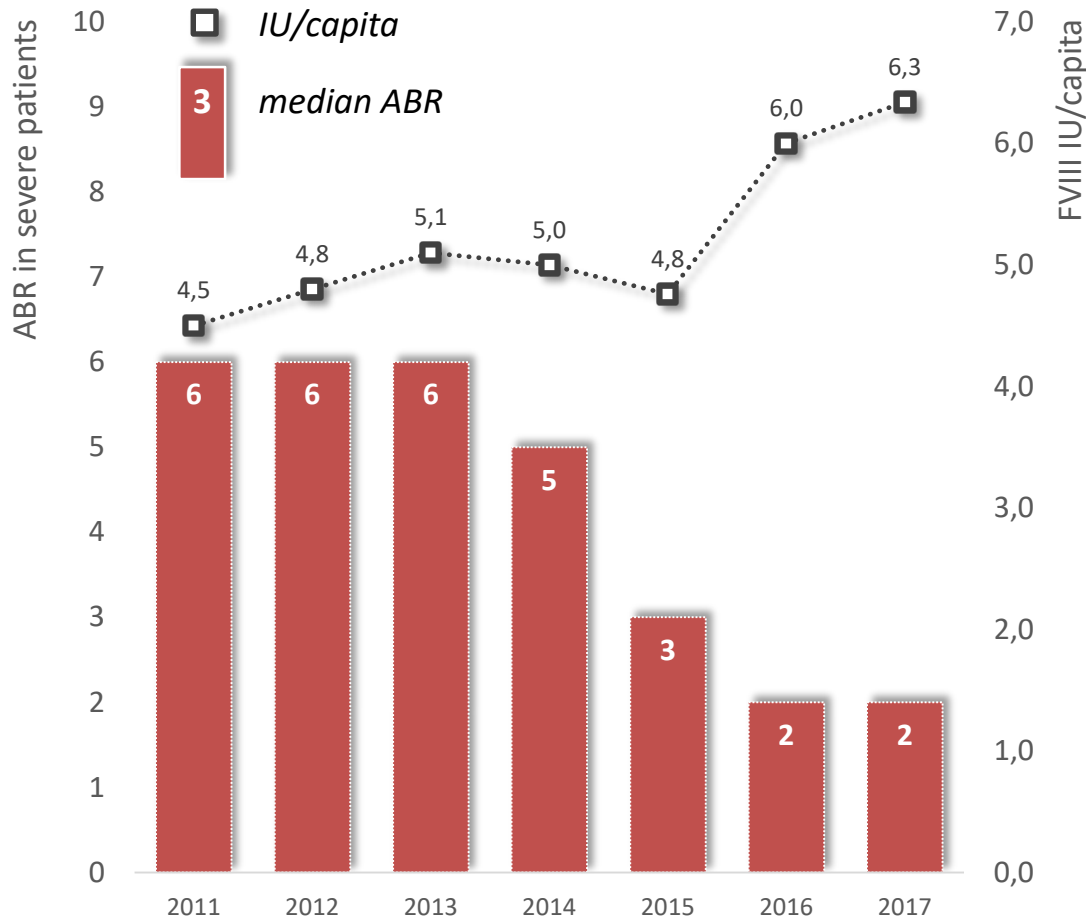
Consumption of FVIII in relation to ABR (excluding inhibitors and ITT)

Haemophilia A
(all severities excl. inh.)
Children+Adults



Consumption of FVIII in relation to ABR (including inhibitors and ITT)

Haemophilia A
(all severities incl. inh.)
Children+Adults



* without inhibitor