

Problems and reality of Russian Registry of PID

**Paschenko O., Kondratenko I., Sidorenko I.,
Ilyina N., Kousnetsova M., Pashanov E.,
Bologov A.**

**Russian State Children`s Hospital,
Research Institute for Pediatric Hematology,
Institute of Immunology,
Medical Academy
Moscow, Russia**

- 1991 - USSR registry of PID
372 patients with 18 forms of PID
for 1982 – 1991 years
- 1996 - Russian Registry of PID
154 patients with PID
- 2002 - renewed Russian Registry
307 patients - 22 PID forms
 - 2004 – Russian Registry
377 patients – 30 PID forms

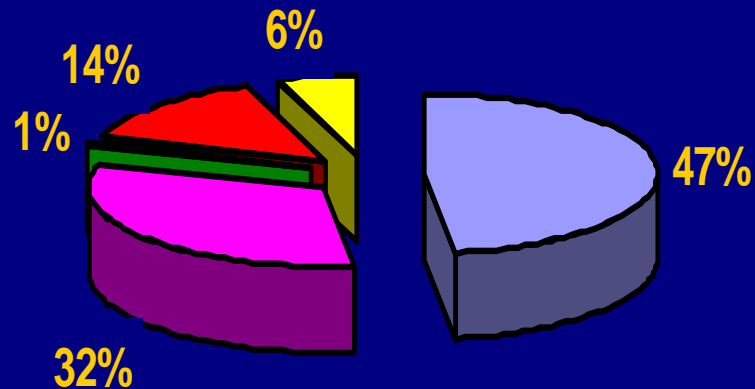
The form of Russian registry includes the data about

- the age of the first manifestation**
- common clinical features – infectious and noninfectious complications**
- gene and molecular defects**
- immune defects**
- therapy and it`s effects**

Russian database of PID

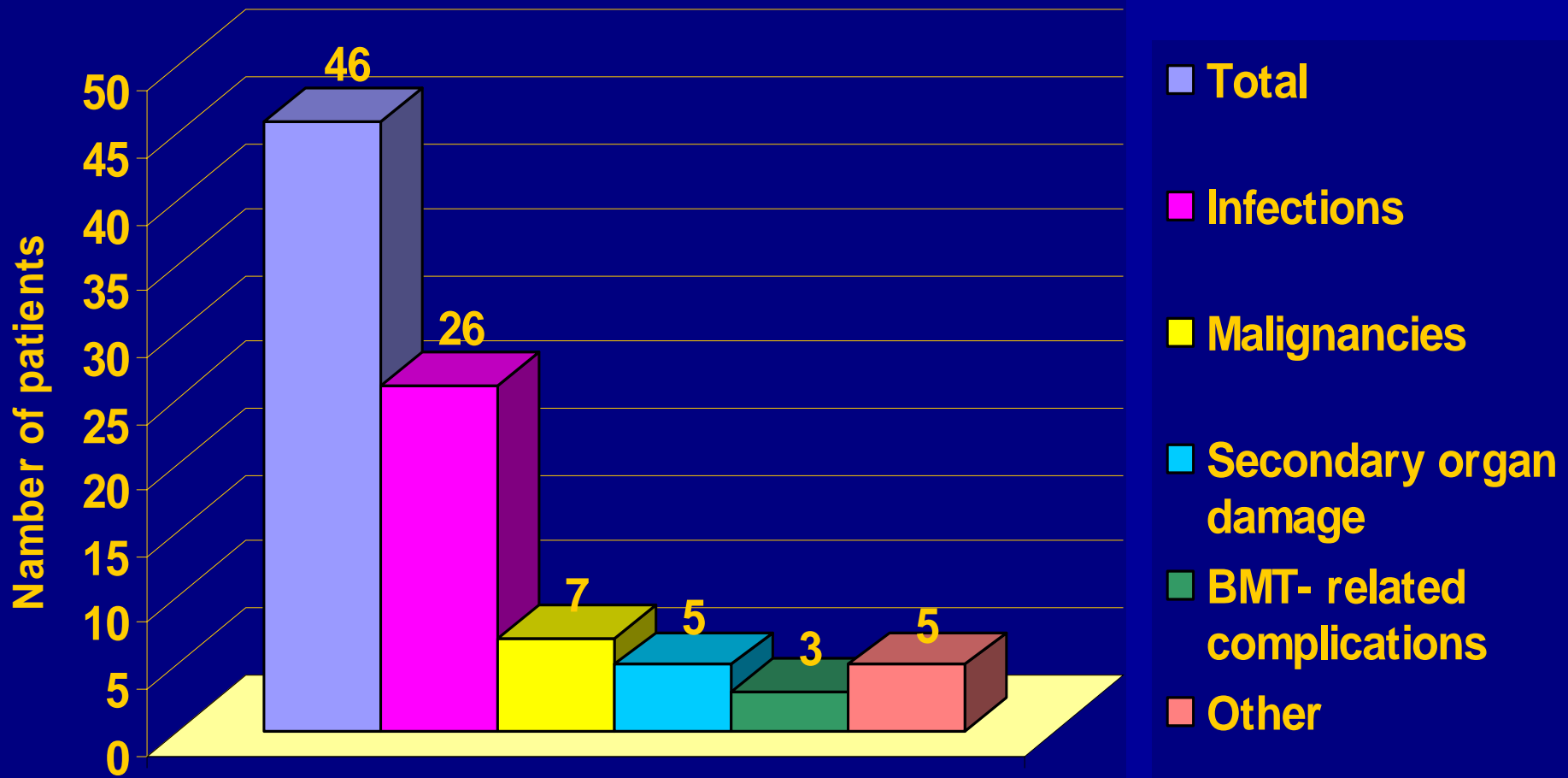
- **377 patients with 30 different forms of PID**
- **Children – 325 patients (86%)**
- **Adults – 52 patients (14%)**
- **50 regions of Russia**
- **Mortality - 12,2%**

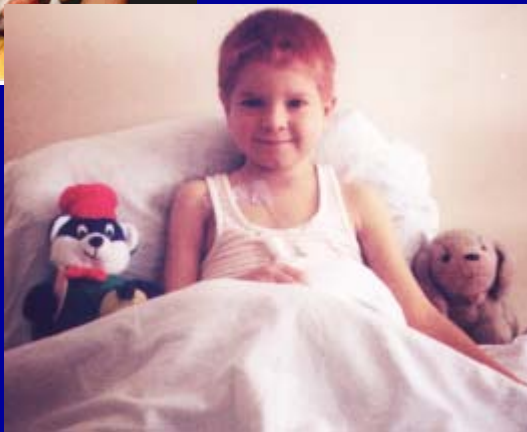
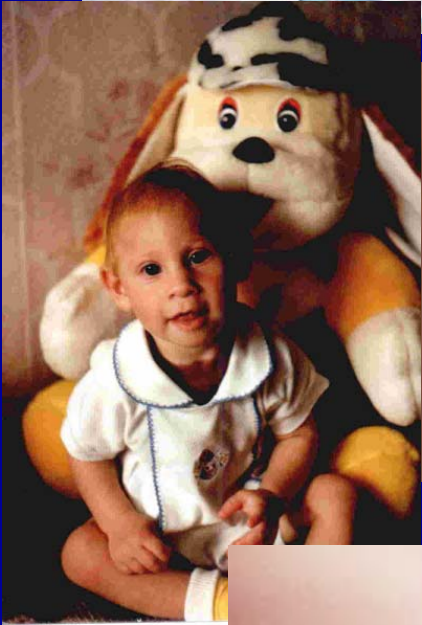
The forms of PID in Russian registry



- Antibody deficiencies - 180
- Combined deficiencies - 120
- Complement deficiencies - 2
- Fagocytic deficiencies - 54
- Other PIDs - 21

The causes of death of the patients with PID





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