



# The ESID Online Registry

## Current figures and perspectives

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- **Introduction**
- **Overview**
- **Statistics for everyone!**
- **Perspectives**

## Introduction (1) - Basics

- Database hosted in Freiburg (Germany)
- Went online in August 2004
- Database for more than 180 PIDs
- Disease-specific subregistries
- Accessible via standard internet browsers

## Introduction (2) - Aims

### The aims of the Registry:

- to compile clinical and laboratory data of patients with primary immunodeficiencies (PID)
- to improve diagnosis
- to improve classification
- to improve prognosis
- to improve therapy
- to provide continuous long-term compilation of data on PID
- to enable genetic and therapeutic trials within ESID
- to enable research across centres

## Introduction (3)

- **Bonus system:**
  - **Compensation** for secretarial assistance (**EUR 10** per patient per year – also for follow-up documentation)
  - **Travel grants** for the top documenting centers (totalling **EUR 3.000**)
  - **Publication award** of max. **EUR 15.000** per year

# Contents

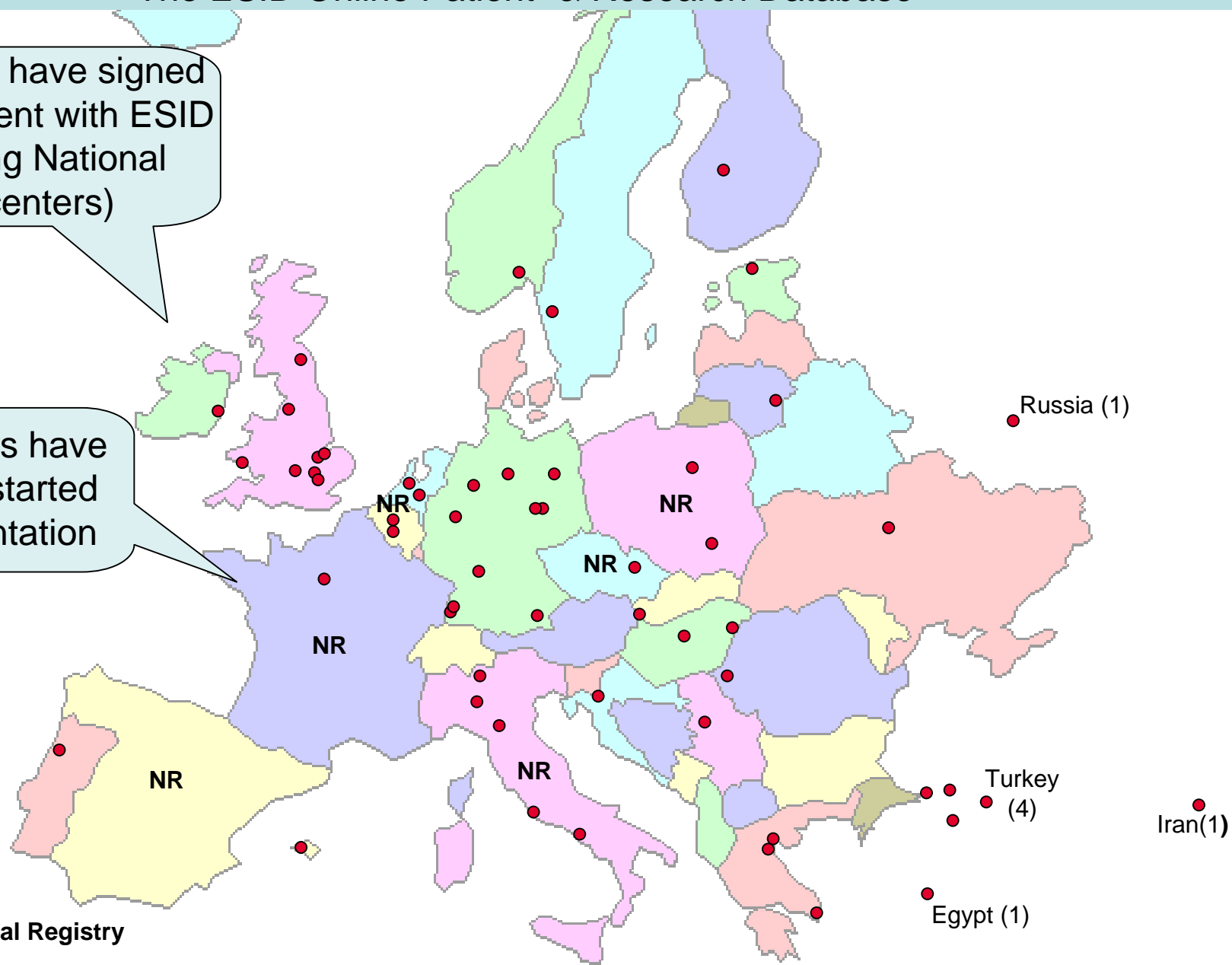
- Introduction
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# EUROPEAN SOCIETY FOR IMMUNODEFICIENCIES

The ESID Online Patient- & Research Database

70 centers have signed an agreement with ESID (including National Subcenters)

30 centers have already started documentation



NR = National Registry

Main Category	Category	Subregistry	data model submitted	under developm.	Demo System	productive System
<b>current status of Subregistries</b>	<b>Agammaglobulinemias</b>	<b>XLA (Btk)</b>				
		<b>all other Agammas</b>				
	<b>Hypogammaglobulinemias</b>	<b>CVID</b>				
		<b>Second. Hypogammagl.</b>				
		<b>Selective IgM def.</b>				
		<b>Selective IgA def.</b>				
		<b>Second. selective IgA def.</b>				
		<b>IgG subclass def.</b>				
		<b>Deficiency of specific IgG</b>				
		<b>Transient Hypogammagl. of infancy</b>				
<b>Predominantly Antibody Disorders</b>		<b>ICOS def.</b>				
		<b>CD19 def.</b>				
		<b>TACI</b>				
		<b>BaffR</b>				
	<b>CSR/HIGM</b>	<b>CD40L</b>				
		<b>AID/UNG</b>				
<b>Predominantly T-cell deficiencies</b>	<b>T-B-Scid</b>	<b>RAG1</b>				
	<b>ADA</b>	<b>ADA</b>				
	<b>DGS</b>	<b>DiGeorge syndrome</b>				
<b>Other well defined PIDs</b>	<b>Hyper IgE syndromes</b>	<b>HIES</b>				
	<b>DNA breakage disorder</b>	<b>AT</b>				
		<b>NBS</b>				
	<b>Osteopetrosis</b>	<b>TCIRG1</b>				
	<b>ICF syndrome</b>	<b>DNMT3B</b>				
	<b>Comel Netherton Syndrome</b>	<b>CNS (SPINK5)</b>				
<b>Autoimm./immunodysreg.</b>	<b>IPEX</b>	<b>FOXP3</b>				



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## Statistics for everyone!

- New ESID Website ([www.esid.org](http://www.esid.org)) went online in February 2006
- Contains a new log in site for the database
- Contains a new section called  
**“Database Statistics”**

# EUROPEAN SOCIETY FOR IMMUNODEFICIENCIES

## The ESID Online Patient- & Research Database

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### ESID Working Parties

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### ESID Members

[ESID Members & Membership](#)

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### ESID Website Statistics

Visitors Online: 3

[View Website Statistics](#)

## Registry

[Link to ESID Online Registry](#) | [ESID Database Statistics](#) | [Link to ESID Test Registry](#)



Every effort has been made to ensure that the information contained in this website has been obtained from reliable sources. However ESID is not responsible for any errors or omissions, or for the consequences of this information. ESID does not guarantee completeness, accuracy or reliability of the information obtained from the use of this database.

Click to register patients

Click to see current figures

### Login Quick Guide

This is a short guide to the ESID registry

[More information](#)

### Frequently Asked Questions (FAQ)

[More information](#)

### Test registry

If you would like to learn more about the ESID registry we have set up a TEST version for you to look at and enter test patients.

[More information](#)

### ESID Information Leaflet

Information on the ESID Database including patient consents in English

[More information](#)

### Data protection and security tutorial

[More information](#)

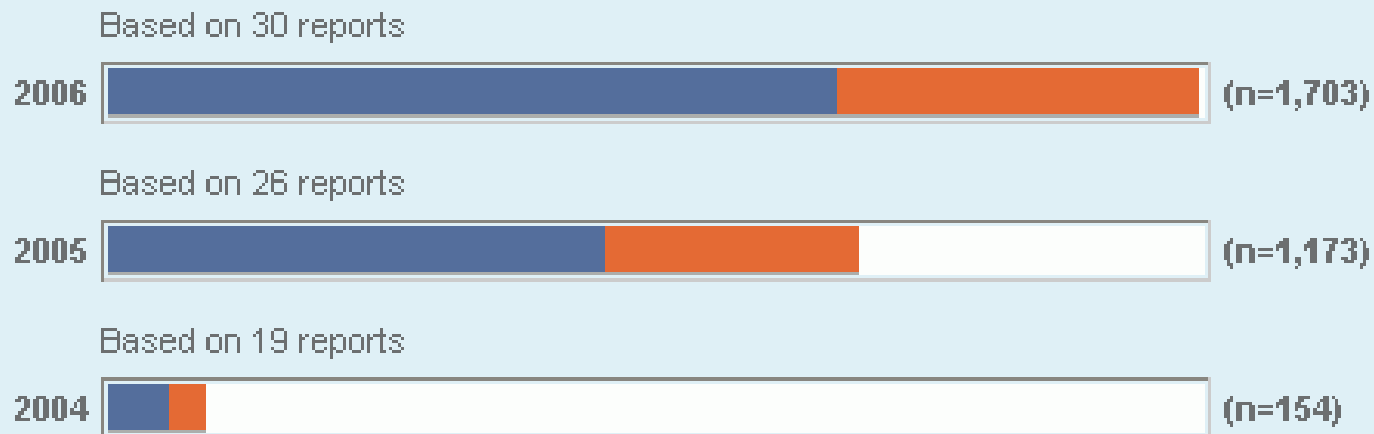
### Patient consent forms

Before entering patient data into the ESID Database you have to obtain an *informed* patient consent. Patient consent forms are available here in many languages. If you have any questions please contact us.

# Statistics

## » Total number of patients

Based on reports received from ESID documenting centers.

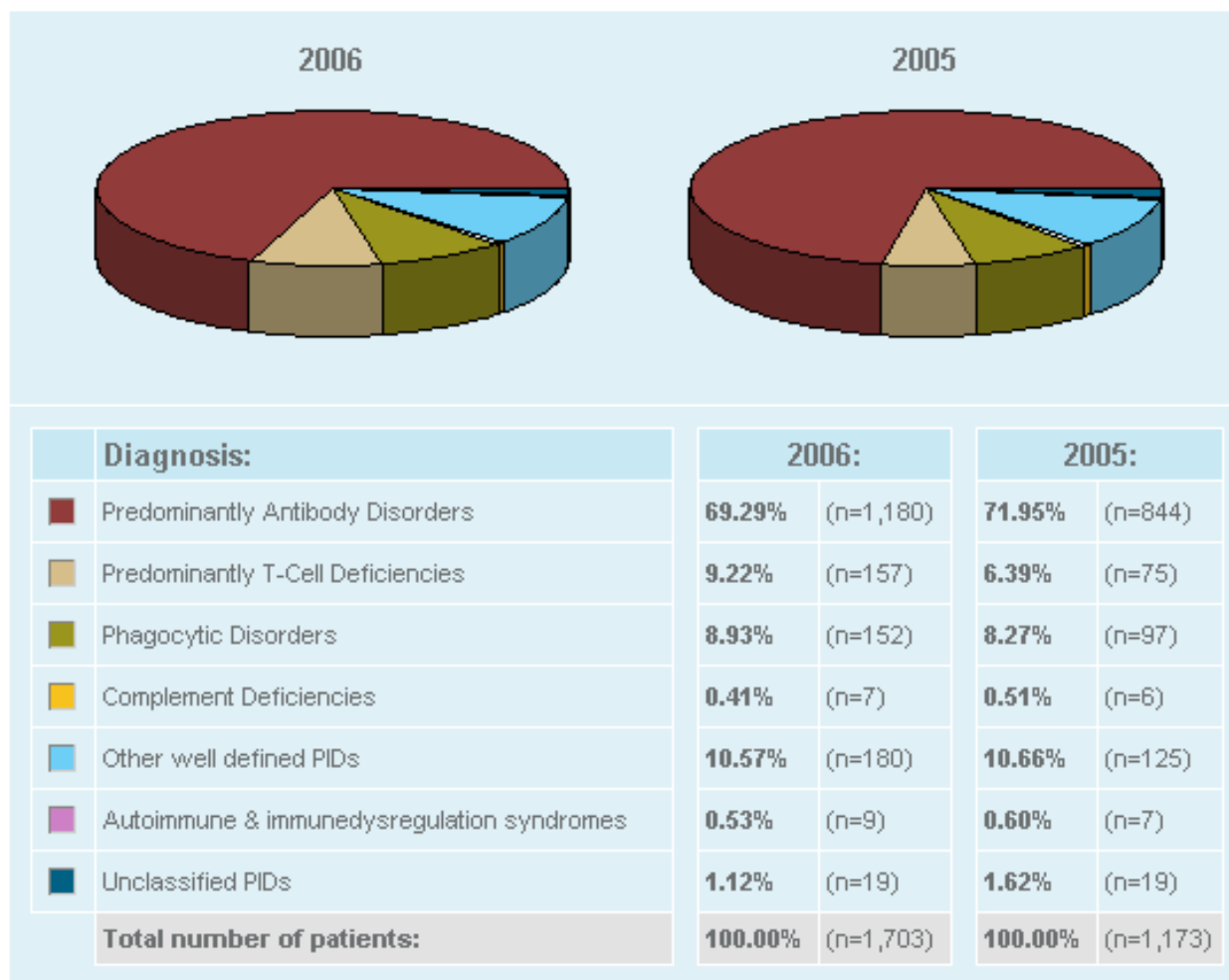


### Distribution between the sexes:

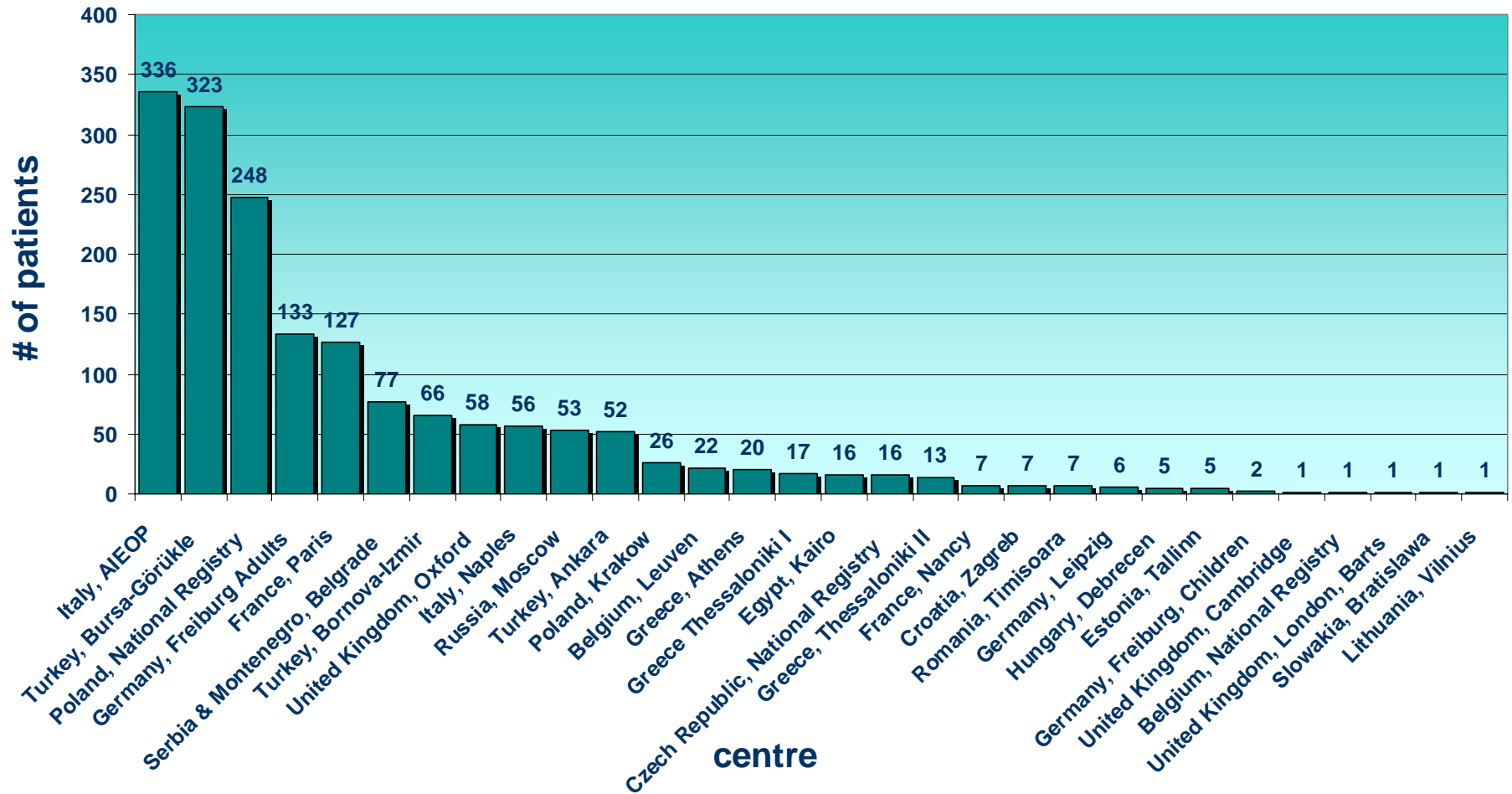
Male
  Female
  Unknown

### Statistics (3)

#### » Major Immunodeficiency Groups



## Statistics (4)

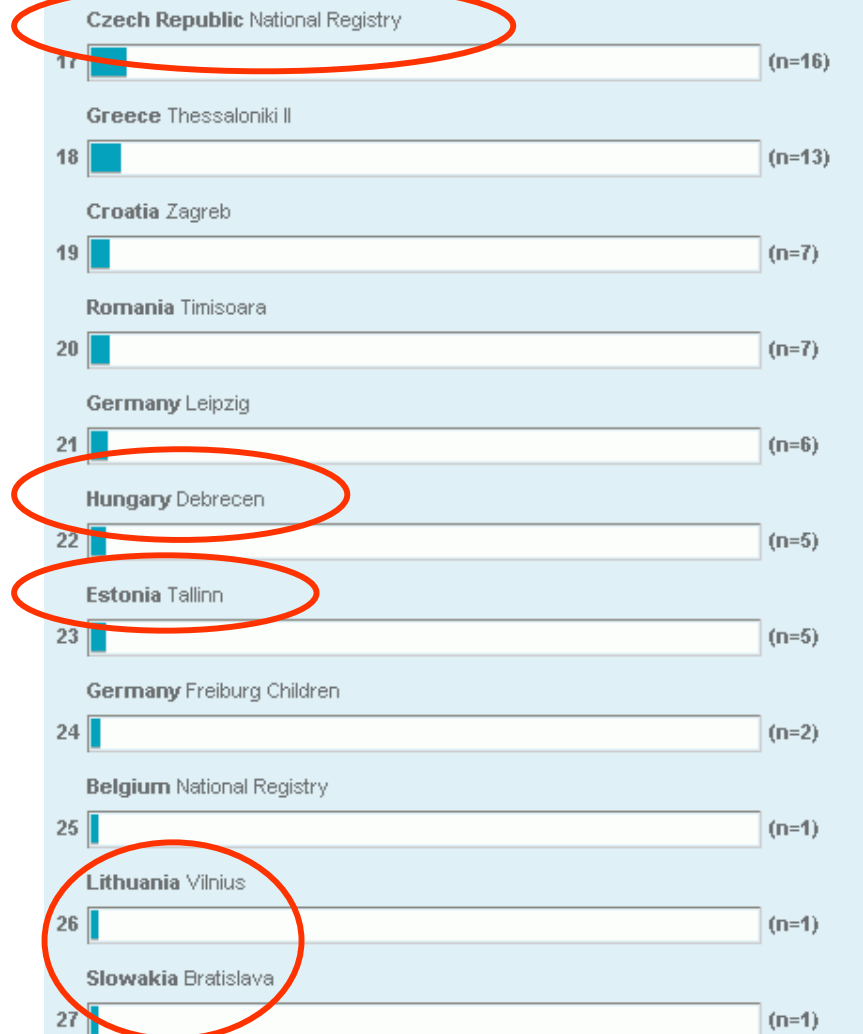
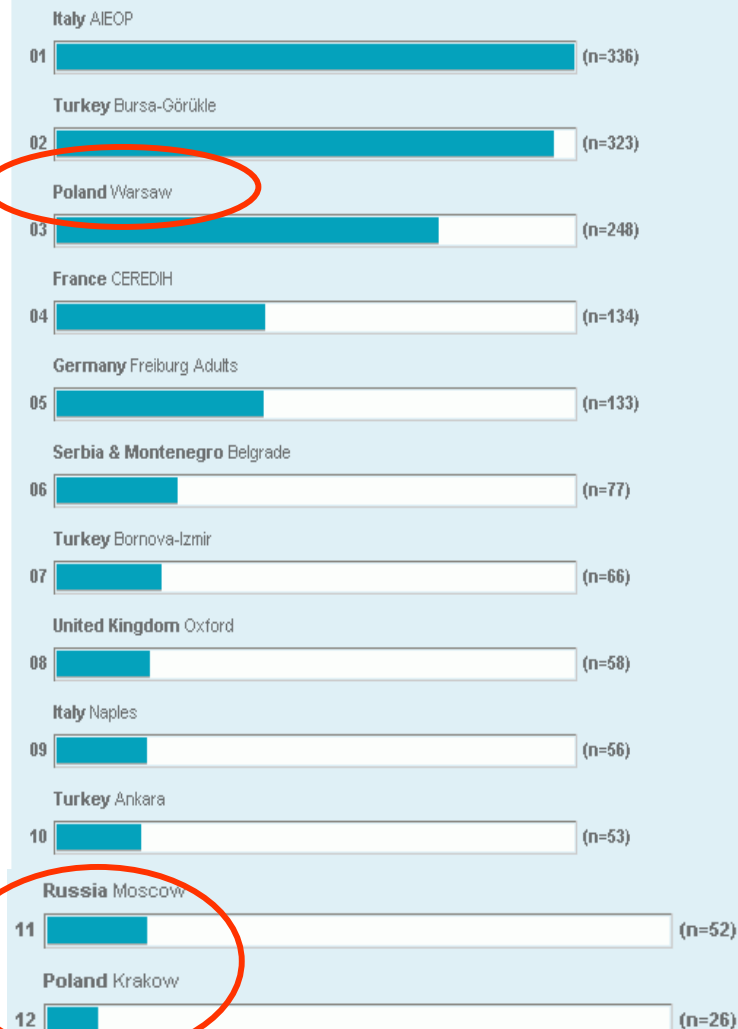


### » Number of patients per documenting center

Ranking of all documenting centers regarding their absolute number of patients.

# Statistics (4)

2006



# Statistics (5)

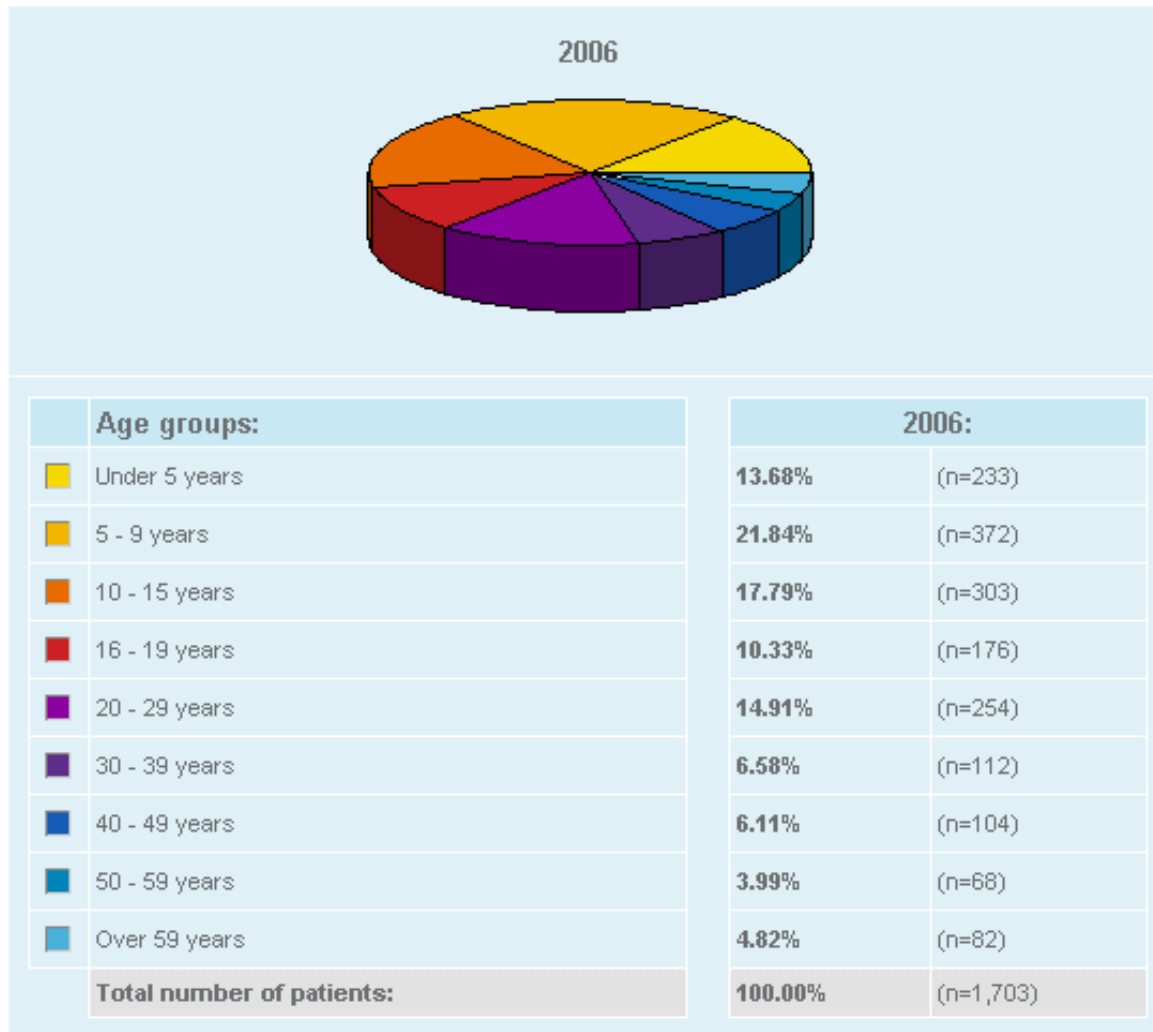
## » Additional statistics

Additional statistics:		2006:	
How many patients underwent BMT?		(n=76)	4.46%
How many patients are on Ig replacement?		(n=773)	45.39%
How many of these are on IVIG?		(n=694)	40.75%
How many of these are on SCIG?		(n=75)	4.40%
How many of these are on IMIG?		(n=0)	0.00%
<b>Total number of patients:</b>		<b>(n=1,703)</b>	<b>100.00%</b>



## Statistics (6)

» Distribution between age groups



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# Perspectives

- **Studies**
- **New functionalities**
  - Predefined queries
- **Personalised version**

## Studies



CVID

Bodo Grimbacher, Freiburg



Agammaglobulinemias/HIGM,  
CEREDIH (in cooperation with ESID programmers, adding fields, etc.)

Alain Fischer,



X-linked Agammaglobulinemia,

Edvard Smith, Stockholm



DiGeorge Syndrome,

Anna Sediva, Prague



Nijmegen Breakage Syndrome,

Ewa Bernatowska, Warsaw



Studies on CD3 deficiency, FoxN1 and Wiskott Aldrich syndrome (WAS) have already been submitted and will be launched in October 2006 at the Budapest meeting.

## Functionalities of the database

- Tool-Tips (exact definition of fields [?])
- Extensible drop-downs [+]
- ICD-10 code implementation
- Color coding



- Report-generation Tool  (in Freiburg)
- SQL query Tool  (in Freiburg)
- Mutation detection Tool  (x-linked & autosomal dominant)



- Predefined Queries (Query by Example) 05/2006
- Personalised version 05/2006



- Report-generation
- Pedigree-tool
- Recall-function
- Percentile Curves
- Search function for ESID-tree
- "Tree Jumper"
- Data warehouse



**Focus on OUTPUT in 2006!**

## Predefined Queries

- How many patients does my centre have?
- What is their distribution regarding diagnosis?
- How many patients do I have
  - who underwent BMT?
  - who are on Ig replacement?
  - who are on IVIG? SCIG? IMIG?
- How many female patients? How many male patients?
- How many < age 5, > < age 10, > < age 16, > < age 20, > < age 30, > < age 40, > < age 50, > < age 60, > age 60 (or other increments)
- Average number of sick days of my cohort this year and last years?
- Average number of hospital days of my cohort this year compared to last years
- My average trough IgG level on my patients on Ig replacement.

## Personalised version

- personalised data on separate servers
- easier and more comfortable working with the database
- database becomes more attractive, can be used as electronic patient chart
- coded version remains available
- data protection is guaranteed
- Schedule: Implementation at the beginning of May

## Personalised version (2)

current coded version, „Patient Info“

<a href="#">New Patient</a>	<a href="#">Select Patient</a>	<a href="#">Show all Patients</a>	<a href="#">User Administration</a>	<a href="#">Logout</a>	<a href="#">Change</a>
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<b>ESID CVID-subregistry</b> USER ID: 75 Login: test.veit PATIENT ID: 1187 Menu:		<b>Core dataset</b>	Visit Date: 2004-02-02 <input type="button" value="+"/>
<b>Patient Info</b>			
Patient ID 1187 <input style="font-size: 8px;" type="button" value="?"/>		Patient consent <input type="text" value="---"/> <input style="font-size: 8px;" type="button" value="?"/>	Familial case <input type="text" value="---"/> <input style="font-size: 8px;" type="button" value="?"/>
Date of birth (YYYY-MM-DD) 1979 - <input type="text" value=""/> - <input type="text" value=""/> <input style="font-size: 8px;" type="button" value="?"/>	Sex <input type="text" value="---"/> <input style="font-size: 8px;" type="button" value="?"/>	Consanguinity <input type="text" value="---"/> <input style="font-size: 8px;" type="button" value="?"/>	
Date of death (YYYY-MM-DD) <input type="text" value=""/> - <input type="text" value=""/> - <input type="text" value=""/> <input style="font-size: 8px;" type="button" value="?"/>	Country of residence <input type="text" value="Denmark"/> <input style="font-size: 8px;" type="button" value="*"/> <input style="font-size: 8px;" type="button" value="?"/>	Patient # in sibship <input type="text" value="---"/> <input style="font-size: 8px;" type="button" value="?"/>	
<input style="background-color: red; color: white;" type="button" value="Submit"/> <input type="button" value="Clear"/>			
<b>Diagnosis</b>			
PID Diagnosis <input type="text" value="CVID"/> <input style="font-size: 8px;" type="button" value="?"/>			

Core Dataset	Core Laboratory
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### personalised version „Patient Info“

**NEW!**

New Patient | Select Patient | Show all Patients | Report Preview | Logout | Change Pass

### ESID CVID-subregistry

USER ID: 32  
Login: demo

Menu

- Dataset
- Core Laboratory
- Info
- Report Documents
- Memo
- Diagnosis & Etiology
- History & Add.Diagnosis
- Quality of Life & Clinical Inv.
- Additional Investigation
- Immunisations & Serology
- Core & Extended Laboratory
- Sample & Biopsy & Surgery
- Therapy & Adverse Events

### Patient Data

Visit Date: 2005-09-20

Patient ID 191

Date of next app...  
Time of next appoin...

Date of birth (YYYY-MM-DD) 1940 2 18  
Sex Male  
Twin --  
Current status alive  
Date of death  
Country of birth United Kingdom  
Country of living United Kingdom  
ICD10-Code of cause of death  
ICD10-Text of cause of death

Send Patient Report

Submit Clear

### PHYSICIAN INFO

## Contact details

- Email: [registry@esid.org](mailto:registry@esid.org)
- Information, documents and demo version:  
[www.esid.org](http://www.esid.org) → „ESID Registry“

**Thank you  
for your attention!**

