



The ESID Online Database - Introduction -

www.esid.org

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Overview

- An online database for more than **200 primary immunodeficiencies (PID)**
- went online in August 2004
- accessible world-wide via **web browsers**
- Administration and programming at University Hospital Freiburg
- Funding currently by the **PPTA** (Plasma Protein Therapeutics Association), **BMBF** (German ministry for education and research) and **EU** (EURO_PAD_NET)

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Diagnosis, classification, prognosis & therapy

PID prevalence numbers

improve

**Long-term compilation of
PID patient data to**

enable

combined evaluation of genetic and clinical data

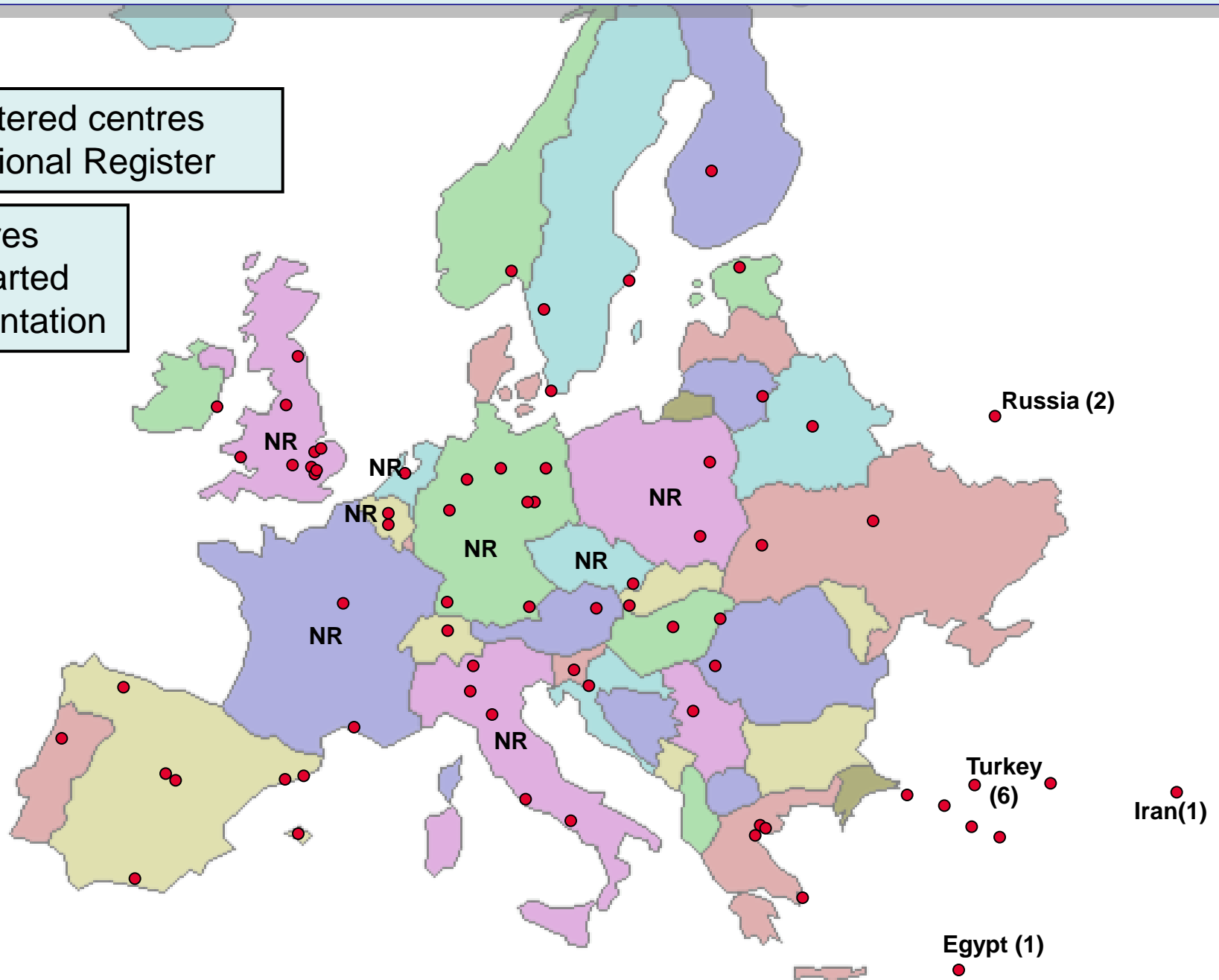
genetic and therapeutic trials within ESID

research across centres

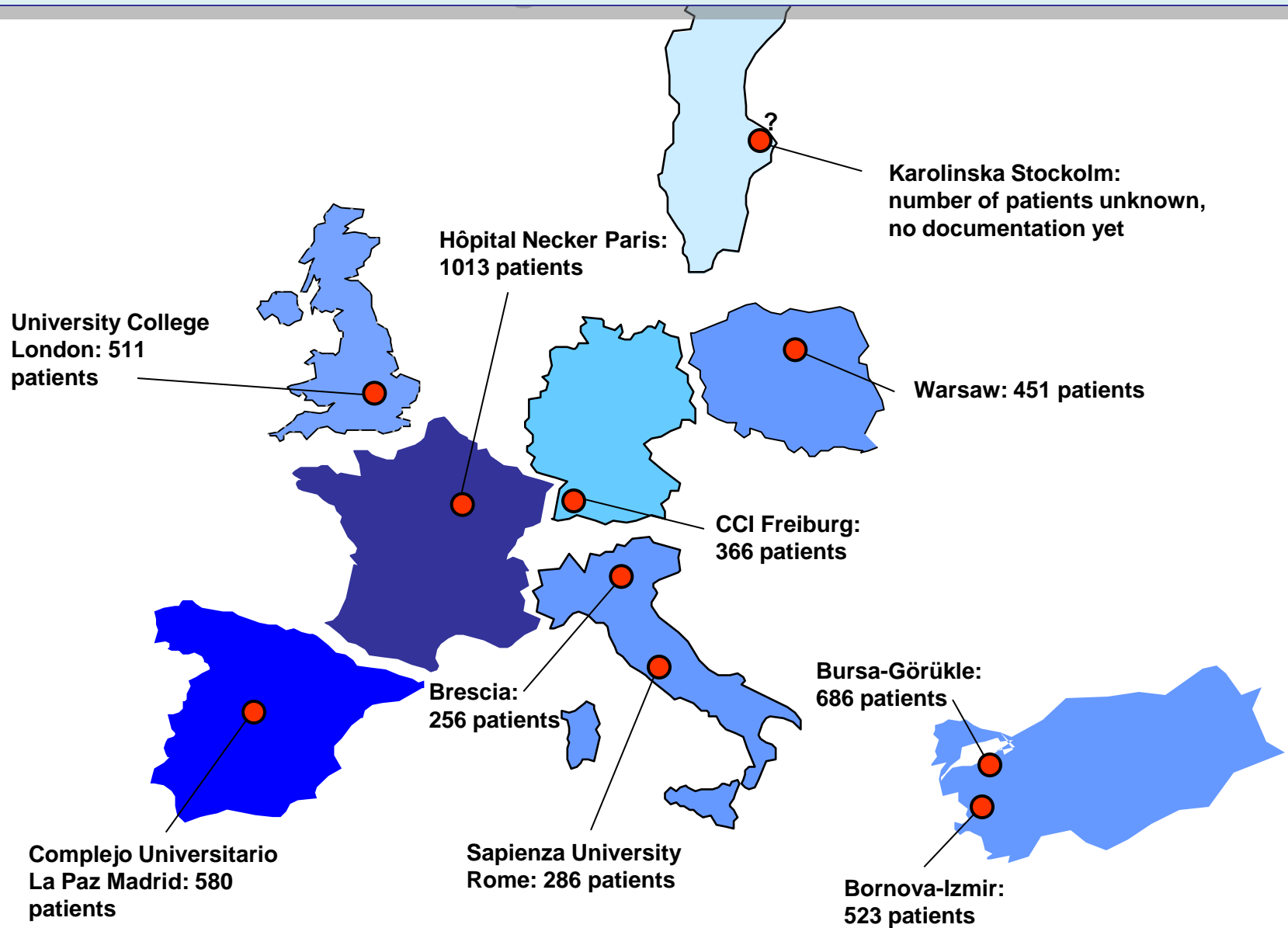
Current status of documenting centres

79 registered centres
NR=National Register

59 centres
have started
documentation



Largest centres*



*based on the number of registered patients seen at this centre over the past five years

The screenshot shows the ESID.org website interface. At the top left is the ESID logo with a map of Europe. Below it is the date and time: May 20, 2008, 2:45 PM. A navigation menu on the left includes 'Home', 'Contact ESID Webmaster', 'ESID Society' (with sub-links for Board, President's Corner, News, Meetings, etc.), and 'ESID Working Parties' (with sub-links for Bone Marrow Transplantation, Clinical, Education, Genetics, Registry, Juniors, and PID Care). The main content area features a 'Registry' section with a red arrow icon. Below this is a banner for 'ESID ONLINE REGISTRY' with a 'Test Version' label and 'ESID DATABASE STATISTICS'. A callout bubble points to the 'Registry' link, stating 'Productive system can be accessed here'. Another callout bubble points to a link in the 'New:' section, stating 'Click HERE to USE the DEMO-System'. The page also contains an 'Important notice' and 'General information' sections.

ESID.org

May 20, 2008, 2:45 PM

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ESID Society

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ESID Working Parties

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> Registry

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

ESID ONLINE REGISTRY **Test Version** **ESID ONLINE REGISTRY** **ESID DATABASE STATISTICS**

New: Reporting Tool for registered database users. [Click here](#)

Important notice: It is the responsibility of the attending physicians and their documents to ensure that data is entered accurately and that their user ID and passwords are kept confidential. Data security is ensured by CwebRD. ESID is not responsible and ESID does not guarantee completeness or accuracy of the results obtained from the use of this database. In particular, ESID makes no representations as to the accuracy and safety of the link to the IMT Mutation Database in Tampere, Finland.

General information

Information for anyone interested in the ESID database.

More information

Information for users

Information on **latest updates** to the database.
Documents required by registered users of the ESID Database.

More information

Productive system
can be accessed
here

Click HERE to
USE the
DEMO-System

» ESID online patient and research database network

Search for diagnosis: (min. 2 chars)

[Search](#) [Reset](#)

- [+ Predominantly antibody deficiencies](#)
- [+ Predominantly T-cell deficiencies](#)
- [+ Phagocytic disorders](#)
- [+ Complement deficiencies](#)
- [+ Other well defined PID's](#)
- [+ Autoimmune and immunedysregulation syndromes](#)
- [+ Autoinflammatory syndromes](#)
- [+ Unclassified immunodeficiencies](#)
- [+ **Main registry \(Live Statistics, Center administration\)**](#)
- [+ Reporting Tool \(Additional password required\)](#)

Subregistries can be accessed via search tree

Live-statistics and Centre-Administration

Reports via a separate query-tool

Common Core dataset, layout depends on login/role

New Patient **Select Patient** **Show all Patients** **Show patient report** **Info & Color Coding** **Logout** **Change Password** **Sponsors**

ESID XLA-subregistry
USER ID: 104
Login: test.gerhard
PATIENT ID: 2243
Menu:

Personal information
Core Dataset:
General, Diagnosis, QoL
Ig-replacement & Antibiotics
Other medication & Adverse Events
Laboratory values
CEREDIH modules:
CEREDIH CODES
Microbiology
Questionnaire
Alternating Antibiotic
Additional modules:
Patient & Diagnosis
Concomitant diseases
Infections
Family history
Quality of Life & Clinical Investigator
Additional Investigation
Immunisations
Core & Extended Laboratory
Sample & Biopsy & Surgery
SF36 Survey
Percentiles

Core data: General Visit Date: 2007-05-09

Patient Info

Patient ID 2243 Patient consent Full consent Familial case —
Date of birth (YYYY-MM-DD) 2000 - 2 - 2 Sex Female if yes: Family Number
Date of death (YYYY-MM-DD) Country of residence Serbia-Montenegro Consanguinity
Current status — Twin
Submitted to REDIP? —Please select—

Submit **Clear**

Diagnosis

PID Diagnosis X-linked agammaglobulinemia
Date of diagnosis 2001 - 1 - 3
Onset of symptoms 20
Submit **Clear**

Quality of life & Clinical Investigation

Date 2007-05-09
Days missed at school/work since 2006-11-28 12 Entry made in old format:
not answered because: not applicable unemployed retired
Days in hospital since 2006-11-28 8 Entry made in old format:
Infectious episodes since 2006-11-28 5 Of these, serious bacterial infections: 1
Weight (kg) 45.0
Submit **Clear**

Status of therapy on selected visit date

Therapy on 2007-05-09 changed not changed no (current) therapy
Submit **Clear**

Additional Questions

Core Dataset

Center specific modules

Extended disease specific dataset

Additional features

Baxter **Biotest** **CSL Behring** **GRIFOLS** **octapharma** **KEDRION** **Talecris** **Sum**
BioScience microsystems

Lab values

[New Patient](#)
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 - Core Dataset:**
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 - SF36 Survey
 - Percentiles

Core Laboratory

Visit Date: 2007-05-09

Date of blood sample 2007-05-09

	Status	Value	Unit
Leukocytes	done	100.0	10 ⁹ /L
Thrombocytes	—		10 ⁹ /L
Erythrocytes	—		10 ⁶ /μL
Hemoglobin	done	2.0	g/dL
(total) Lymphocytes (%)	done	3.0	%
Neutrophils (%)	—		%
Eosinophils (%)	—		%
Basophils (%)	—		%
Monocytes (%)	—		%
IgG	done	= 4.0	g/L
IgA	done	= 5.0	g/L
IgM	—	=	g/L
IgE	—	=	IU/mL
CD3 (%)	—		%
CD4 (%)	—		%
CD8 (%)	—		%
CD19 (%)	—		%
or CD20 (%)	—		%
CD56/CD16 (%)	—		%
CD19+/CD27+ (%)	—		%
CD20+/CD27+ (%)	—		%
NK-T-Cells (%)	—		%
IgG1	—	=	mg/dL
IgG2	—	=	mg/dL
IgG3	—	=	mg/dL
IgG4	—	=	mg/dL

	Status	Value	Unit
(total) Lymphocytes (abs.)*	done	3.0	10 ⁹ /L
Neutrophils (abs.)*	—		10 ⁹ /L
Eosinophils (abs.)*	—		10 ⁹ /L
Basophils (abs.)*	—		10 ⁹ /L
Monocytes (abs.)*	—		10 ⁹ /L

IgG measured as trough level before initiation of Ig-repl.
 other unknown

Days passed since last IVIG-substitution:

	Status	Value	Unit
CD3 (abs.)*	—		10 ⁹ /L
CD4 (abs.)*	—		10 ⁹ /L
CD8 (abs.)*	—		10 ⁹ /L
CD19 (abs.)*	—		10 ⁹ /L
or CD20 (abs.)*	—		10 ⁹ /L
CD56/CD16 (abs.)*	—		10 ⁹ /L
CD19+/CD27+ (abs.)*	—		10 ⁹ /L
CD20+/CD27+ (abs.)*	—		10 ⁹ /L
NK-T-Cells (abs.)*	—		10 ⁹ /L

* abs. = absolute



Additional features: Patient Report

Show patient report



ESID Online Database

Patient Report sheet

Date of printout: 26.09.2007

ESID Database ID: 5880

Mr. Testpatient, Manfred

Date of birth (Year-Month-Day): 1960-7-4

Diagnosed with: Deficiency of specific IgG
 Country of residency: Netherlands
 Followed at ESID Center: DEVELOPMENT
 Type of patient consent: Research and genetics

Date of diagnosis (Year-Month-Day):	2001	-	-	-
Onset of symptoms (Year-Month-Day):	1999	-	-	-

Clinical data for visit date: 2007-07-30

Body weight (kg):	78.0
Body height (cm):	185.0
Head circumference (cm):	

Quality of life data

From	until	Days missed at school or work	Days in hospital	All infections	Serious infections
2007-03-23	2007-07-30	0	10	1	0
2001-05-07	2007-03-23	10	30	2	1
one year before	2001-05-07	10	9	5	2

Current medication

Since (year)	Drug group	Generic name	Brand	route	Dose	Unit	Times	per
2004	Ig-Replacement	Human Gammaglobulin	Subcuvia	s.c.	20.0	g	1.0	week
1999	Corticosteroids	Cortison		oral	20.0	mg	1.0	day

Blood count

Date	Leukocytes	Unit	Thrombocytes	Unit	Erythrocytes	Unit	Hemoglobin	Unit
2007-07-30	5.9	10 ⁹ /L	206.0	10 ⁹ /L	4.8	10 ⁶ /μL	12.0	g/dL
2007-03-23	9.7	10 ⁹ /L	196.0	10 ⁹ /L	4.8	10 ⁶ /μL	16.0	g/dL
2001-05-07	6.8	10 ⁹ /L	198.0	10 ⁹ /L		10 ⁶ /μL		g/dL

Immunology lab values

Date	IgG	Unit	IgA	Unit	IgM	Unit	Relation to Ig replacement
2007-07-30	7.9	g/L	2.0	g/L	<0.23	g/L	trough level
2007-03-23	9.0	g/L	<0.5	g/L	<0.23	g/L	other
2001-05-07	2.1	g/L	0.5	g/L	<0.23	g/L	before any Ig repl.

Additional features: Patient List

Patient List (download)



ESID Online Database

Patient list

Freiburg Development Centre 2

Date of printout: 25.09.2007

List of patients by diagnosis

Total number of patients: 10

ID	Disease	Last name	First name	Year of birth	Year of death	Sex
4391	Common variable immunodeficiency (CVID)	Bellabella	Isabella	1983		female
4361	Common variable immunodeficiency (CVID)	Bo	Stakka	1983		male
4923	Common variable immunodeficiency (CVID)	Bond	James	1983		male
4298	Common variable immunodeficiency (CVID)	Leone	Sergio	1983		male
4384	Common variable immunodeficiency (CVID)	Müller	Hans	1983		male
4379	Common variable immunodeficiency (CVID)	Parker-Bowles	Camilla	1983		female
3929	Common variable immunodeficiency (CVID)	Valerien	Harry	2000		male
5458	Deficiency of specific IgG	Skipielsko	Igor	1975		
5459	Isolated IgG subclass deficiency	Megawatti	Ohma	1900		female
3936	Nijmegen breakage syndrome (NBS1)	Nein	Ja	1985		male

Additional features: Mutation Browser

New Patient

Select Patient

Show all Patients

Logout

Change Password

Sponsors

ESID SCID-subregistry

USER ID: 133

Login: beavis

PATIENT ID:

432

Menu:

Core Dataset

Core Laboratory

Mutation Browser

Info & Color Coding

Patient & Diagnosis & History

Donor Information

Bone Marrow Transplantation

Results

Add. Diagnosis

Quality of Life & Clinical Inv

Additional Investigation

Immunisations & Serology & Lymphoproliferation

Core & Extended Laboratory

Sample & Biopsy & Surgery

Therapy & Adverse Events



Mutation Browser

Press the "Mutation submission" button to submit a new mutation event. Note that you will be redirected to the UTA-IDbases website in order to validate your data. Once the mutation is successfully validated and submitted please press the "Mutation Browser" webmenu option to display it.

Please check previously that your browser allows pop-up windows from the ESID website.

Gene name: **RAG1**

Reported cDNA mutations in **RAG1**:

Allele 1

Allele 2

```
GAGAGCAGAGAACACACTTTGCCTTCTCTTTGGTATGAGTAATATCAACCAAAATTGCAG GAGAGCAGAGAACACACTTTGCCTTCTCTTTGTTTGGTAAATATCAACCAAAATTGCAG
ACATCTCAACACTTTGGCCAGCCAGCCTGCTGGAGCAAGGTACCTCAGCCAGCATGGCAGC ACATCTCAACACTTTGGCCAGCCAGCCTGCTGGAGCAAGGTACCTCAGCCAGCATGGCAGC
CTCTTTCCCAACCCACTTTGGGACTCAGTTCCTGCCCATGCAAAATTCAGCAGCCACATAT CTCTTTCCCAACCCACTTTGGGACTCAGTTCCTGCCCATGCAAAATTCAGCAGCCACATAT
TAAATTTTTCAGAAATGGAATTTAAAGCTGTTCCCGGGTATGCAAAATTCAGCAGCCACATAT CTCTTTCCCAACCCACTTTGGGACTCAGTTCCTGCCCATGCAAAATTCAGCAGCCACATAT
AGAAGCTCAAAAGGAAAAGGAAAGGATTCTTTGAGGGC http://www.esid.org:8080 - M... ATTTAAGCTGTTCCCGGGTGAATCTCTTTGAAAAGACACCTGA
GAAGGATTCTTTGAGGGGAAACCCCTCTCTGGAGCAATCTCC
TGATGGTCAGAAAGCCAGTCCCACTCAGCCATTGTAAAAGC
GAAATTTTCAGCAACCGAGAAAGCAAGAGGCAAGCGATCCA
TCTCTGCCCATCTGTGGGAATCTTTTAGAGCTGATGAGCA
CCATGGTCTGTGGATGGTAAACCCCTAGCCCTTTTACGAAA
TTCTGGCCGGACCTCATTGCCAAGGTTTTCCGGATCGATGT
GATCCACCCCACTGAGTTCGCCATAACTGTGGAGCATCAT
TGCCCATGTGAGGTTTTACTTCCCGAGGAAAGCTGACCATGGA
ATCCTGTGACATCTGCAACACTGCCCGTCCGGGACTCAAAGG
CTTGACGCTCAGCAAAAACTCAAAACTGTGCTTGACCAAGC
CAAGAGAAAGAGCTCAGGCCAAGGATCAGCAGCAAGGATGTCAT
CAGTAAGATACATCTTAGTACCAAGCTCCTGCAGTGGACTT
CCCAGAGCACTTTGTGAAATCCATCTCTGCCAGATCTGTGAACACATTCTGGCTGACCC
TGTGGAGACCAACTGTAAGCATGTCTTTTGGCCGGTCTGCATTCTCAGATGCCCTCAAAGT
CATGGGCAGCTATTGTCCCTCTTGGCCGATATCCATGTCTCCCTACTGACCTGGAGAGTCC
AGTGAAGTCTTTCTGAGCGTCTTGAATTCCTGATGGTGAATGTCCAGCAAAAAGAGTG
CAATGAGGAGGTGAGTTTGGAAAAATAAATCAACCAATCTCAAAGTCAAGGAATCAA
AGAGATTTTGTGCACATTAATAAAGGGGGCCGGCCGCCAACATCTTCTGTGCTGTGAC
TCGGAGAGCTCAGAAAGCCAGCGGACTCAAAGTCAAAAGCCTTTGCTGA TCGGAGAGCTCAGAAAGCCAGCGGCTGAGGGACTCAAAGTCAAAAGCCTTTGCTGA
CAAAAGAAAGGTGGAGATGTGAAGTCCGTTGTCATGACCTTGTCTGTGCTGTGAG
CAAAAGAAAGGTGGAGATGTGAAGTCCGTTGTCATGACCTTGTCTGTGCTGTGAG
GGCGAGGAATGAGCACAGGCAAGCTGATGAGCTGGAGGCCATCATGCAGGAAAGGGCTC
TGGCCTGCAGCCAGCTGTTGCTTGGCCATCCGTTGTCAAACCTTCTCAGCTGCAGTCA
TGACCAAAAGATGTACAGGACTGTGAAAGCCATCAGAGGACTGTGAAAGCCATCAGGGGACAGATTTTTACGCTTT
GCATGCCCTTCGGAATGCTGAGAAGGTAATCTTCTGCCAGGCTACCACCCTTTGAGTGCCA
GCCACCTCTGAAGAATGTGCTTCCAGCACTGATGTTGGCATTATGATGGGCTGTCTGG
ACTATCATCTCTGTGGAATGATTACCCAGTGGACACCATTTGCAAAAGGTTCCGCTATGA
TTCAGCTTTGGTGTCTGCTTTGATGGACATGGAAGAAGACATCTTGAAGGCCATGAGATC
CCAAAGCCTTGATGATTAACCTGAAATGGCCCTTCACTGTGGTGGTGAAGGAGTCTTGTGA
TGGAAATGGGAGACGTTGAGTGAGAAGCATGGGAGTGGGCTGTAGTTCOCAGAAAAGCGAAT
CCGTTTTTTCATTCACAAATCAAGAAAATTAATTTGCCCCAGCTCTCAGAAATGTGAAAGT
ATTTGAAGAAAGCCAAACCTAACTCTGAAGTGTGTTGCAAGCCATTTGTGCCTTATGTCTGCC
AGATGAGTCTGACCAAGCAGTACTGCACTGCACTCCTGACTCCTCCTGCTGAGAGGGA
GGCCATGAAGAGCAGTGAATTAATGCTTGGCTGGGAGGCCATTCTCCGAGTTTCAAAGTT
CATCTTCAGGGCCACCCGCTATGATGAAAACCTTGTGCCGGAAAGTGGAAAGCCCTCAGAGC
TCTTGGCTCAGTCTACATTTGACTCTTTGTGATGCCACCCGCTGGAAGCCCTCTCAAAA
TCTTGTCTTCCACTCTATAACCGAAGCCATGCTGAGAAAGCTGGAAGCTTATGAGGCTCTG
```

Gene = RAG1
Molecule Type = CDNA
Start position = 35

Reference Sequence:
T

Mutated Sequence:
C

Fertig

Additional features: Percentile curves

subregistry

USER ID: 105
Login: test.benjamin

PATIENT ID:
273

Menu:

Patient Data

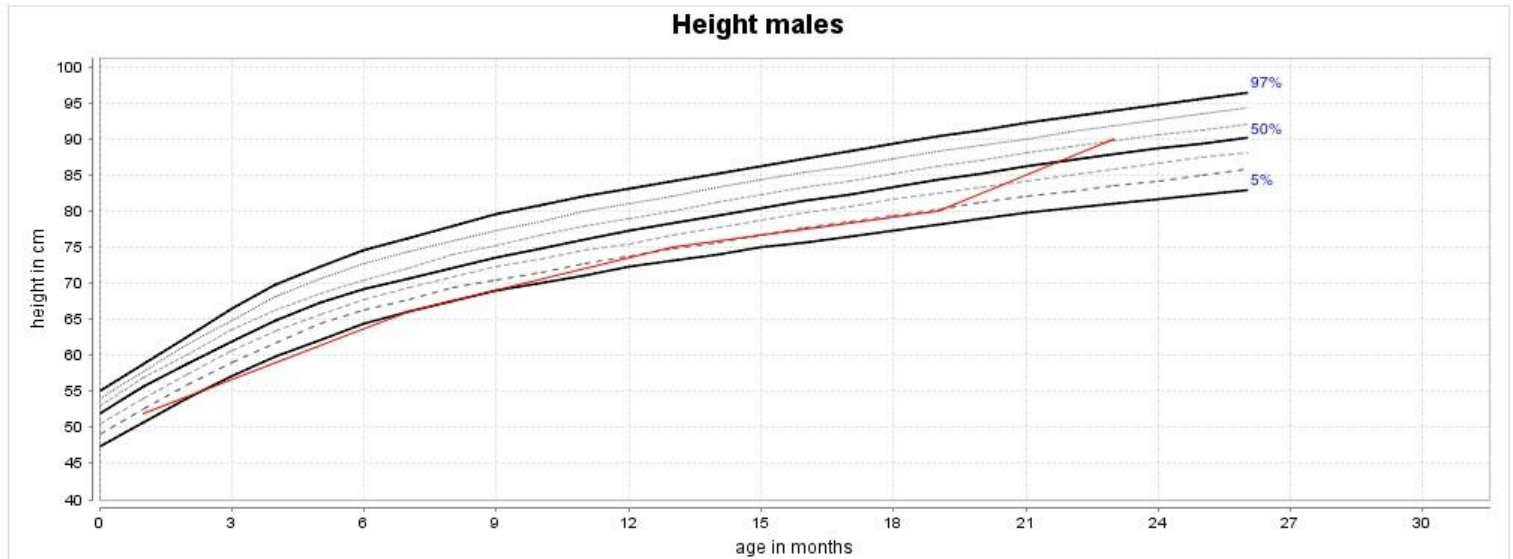
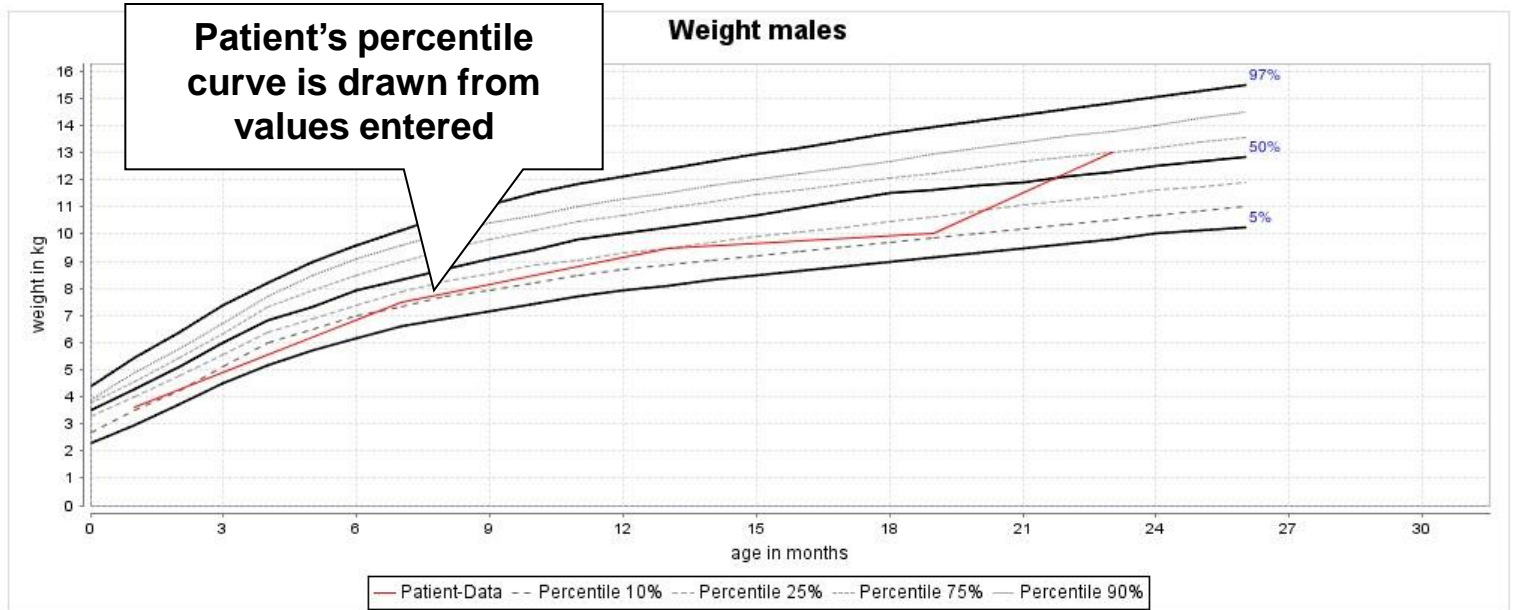
Core

Dataset

Core

Laboratory

Percentiles



Additional features: SF36 Health Questionnaire

ESID CVID-subregistry

USER ID: 98
Login: viviane.knerr
PATIENT ID:
422

Menu:

- Core Dataset
- Core Laboratory
- Info and Color Coding
- Diagnosis & Etiology
- History & Add Diagnosis
- Quality of Life & Clinical Inv.
- Additional Investigation
- Immunisations & Serology
- Core & Extended Laboratory
- Sample & Biopsy & Surgery
- Therapy & Adverse Events
- SF36 Survey
- Normal Ranges
- User Administration
- Role Administration
- Center Administration

SF36 Survey

Visit Date: 2003-11-17

This survey asks for your views about your health. This information will help keep track of how you feel and how well you are able to do your usual activities.

Please answer these questions by "check-marking" your choice.
Please select only one choice for each item

1. In general, would you say your health is:

- Excellent Very good Good Fair Poor

2. Compared to ONE YEAR AGO, how would you rate your health in general NOW?

- MUCH BETTER than one year ago. Somewhat BETTER now than one year ago.
 About the SAME as one year ago. Somewhat WORSE now than one year ago.
 MUCH WORSE now than one year ago.

3. The following questions are about activities you might do during a typical day.

Does your health now limit you in these activities?

If so, how much?

Activities

a. Vigorous activities,

such as running, lifting heavy objects, participating in strenuous sports

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

b. Moderate activities,

such as moving tables, pushing a vacuum cleaner, bowling or playing golf

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

c. Lifting or carrying groceries

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

d. Climbing **several flights** of stairs

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

e. Climbing **one** flight of stairs

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

f. Bending, kneeling or stooping

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

g. Walking **more than a mile**

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

h. Walking **several hundred yards**

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

i. Walking **one hundred yards**

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

j. Bathing or dressing yourself

1. Yes, limited a lot 2. Yes, limited a little 3. No, not limited at all

4. During the **past 4 weeks**,

how much of the time have you had any of the following problems with your work or other regular activities as a result of your physical health?

a. Cut down on the **amount of time** you spent on work or other activities All of the time Most of the time Some of the time A little of the time None of the time

b. **Accomplished less** than you would like All of the time Most of the time Some of the time A little of the time None of the time

c. Were limited in the **kind** of work or other activities All of the time Most of the time Some of the time A little of the time None of the time

d. Had **difficulty** performing the work or other activities (for example it took extra effort) All of the time Most of the time Some of the time A little of the time None of the time

Predefined Queries (Live statistics, database users)

→ General numbers
for all registered
patients



Live statistics

Number of registered patients

Total patient count in database: 9136

Gender distribution in database

Gender:	female	Count:	3716
Gender:	male	Count:	5417
Gender:	not given	Count:	1
Gender:	unknown	Count:	2

Patient distribution in centres

The centre	Paris, CEREDIH	has registered	3149	patients.
The centre	REDIP (Spain) (without single centers)	has registered	871	patients.
The centre	Bursa-Gorukle, University Medical Faculty	has registered	630	patients.
The centre	Italy, ipinet (AIEOP)	has registered	584	patients.
The centre	Bornova-Izmir, Ege University	has registered	490	patients.
The centre	Warsaw, Children's Memorial Health Institute	has registered	406	patients.
The centre	London, UCL, Royal Free Hospital	has registered	243	patients.
The centre	Freiburg 1, Immunology	has registered	227	patients.
The centre	London, UCL, Institute of Child Health/GOSH	has registered	213	patients.
The centre	Newcastle, General Hospital	has registered	143	patients.
The centre	Manchester	has registered	138	patients.
The centre	Munich, Childrens Hospital Dr. von Hauner	has registered	111	patients.
The centre	Freiburg 2, Children's Hospital	has registered	102	patients.
The centre	Hannover 2, Pneumology	has registered	100	patients.
The centre	Mallorca, Hospital Son Dureta	has registered	97	patients.
The centre	Dublin, St. James's Hospital	has registered	93	patients.

→ Individual numbers for every centre

Number of registered patients in my centre

I have patients registered in my centre.

Distribution of PID-diagnoses in my centre

I have patients registered from my centre with PID-Diagnosis:

<input type="checkbox"/>	I have	<input type="text" value="94"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="CVID"/>
<input type="checkbox"/>	I have	<input type="text" value="8"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="IgG subclass deficiency"/>
<input type="checkbox"/>	I have	<input type="text" value="5"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="ICOS deficiency"/>
<input type="checkbox"/>	I have	<input type="text" value="4"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="Other Hypogammaglobulinemias"/>
<input type="checkbox"/>	I have	<input type="text" value="3"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="CSR defects and HIGM syndromes with unknown gene"/>
<input type="checkbox"/>	I have	<input type="text" value="3"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="Secondary hypogammaglobulinemia"/>
<input type="checkbox"/>	I have	<input type="text" value="3"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="TACI deficiency"/>
<input type="checkbox"/>	I have	<input type="text" value="2"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="Agammaglobulinemias with unknown genetic cause"/>
<input type="checkbox"/>	I have	<input type="text" value="2"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="Unclassified immunodeficiencies"/>
<input type="checkbox"/>	I have	<input type="text" value="2"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="selective IgA deficiency"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="ADA deficiency"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="AID deficiency (AICDA)"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="BAFFR deficiency"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="CD4-selective"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="CGD with unknown genetic cause"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="Good-syndrome (associated with thymoma)"/>
<input type="checkbox"/>	I have	<input type="text" value="1"/>	patients registered from my centre with PID-Diagnosis:	<input type="text" value="HIES"/>



May 26, 2006, 11:03 AM

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Every effort has been made to ensure that the information contained in this website has been obtained from reliable sources. However ESID does not accept any responsibility for errors or omissions, or for the results obtained from the use of this information. The accuracy, or timeliness of the results obtained

Click to see current figures

It is the responsibility of the attending physicians and their documenting team to ensure that data is entered accurately and that their user ID and passwords are kept undisclosed. Data security is ensured by CwebRPD. In particular, ESID makes no representations as to the accuracy and safety of the link to the IMT Mutation Database in Tampere, Finland.

Login Quick Guide

This is a short guide to the ESID registry

[More information](#)

Frequently Asked Questions (FAQ)

[More information](#)

[Test registry](#)

Statistics (Example, public)



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Number of patients

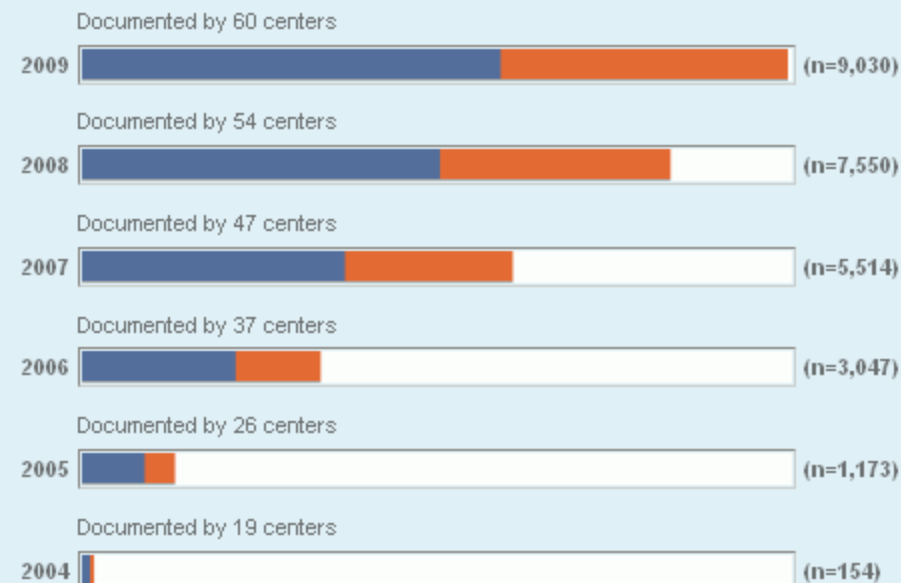
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>> Total number of patients (March 27th, 2009)

Registered in the ESID database by [ESID documenting centers](#).



Distribution between the sexes:

■ Male ■ Female ■ Unknown

Statistics (Example, public)



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ESID Members Login

Username:

Password: *

*) Note! Not your ESID database password

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Selected year:

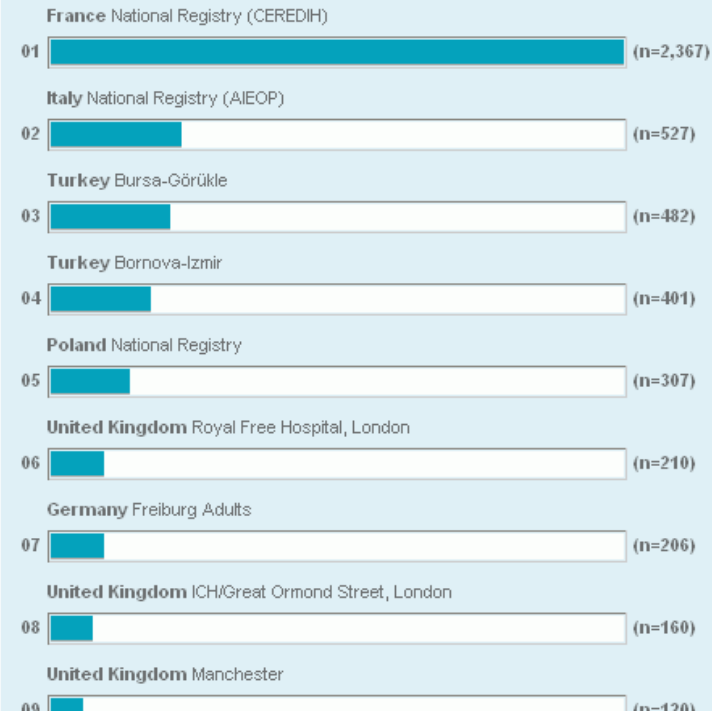
2008

[Select](#)

>> Number of patients per documenting center

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

2008



Issues

- **Documentation**

(,man-power‘, quality of data, frequency of documentation)

- **Data Protection** (legal / technical issues)

Different legislation, even within one country, e.g. Germany’s federal states

- **Ethics**

Ethics approval required for every centre

Approach

Documentation

- **Publication awards** to encourage the use of the system for studies (with data derived from the ESID-database) and publication of the results (5 awards with a total sum of 15.000 € per year)
- **Dedicated documenters** drastically increase the documentation rate (e.g. CEREDIH, France since 2006, Netherlands since summer 2009, starting in Germany in November 2009)

Approach

Data protection/Ethics: legal issues

- Assessments from the Freiburg **ethics committee** and the officials of the German federal states (e.g. Bavaria, Baden-Württemberg) can be used as a template to facilitate the process in other countries / states (but still much work! often causing significant delay in starting documentation)
- Every patient (or legal tutor) has to sign an **‘informed consent’** before documentation starts (translations of information and forms available on ESID website)

Approach

Data protection: Technical issues

- Servers are hosted within the ,secure server net‘ of the University Hospital’s computing centre, Freiburg
- Access to the system is password protected (with login-lock after 4 unsuccessful login-attempts)
- Communication with database is encrypted (https)
- User/role concept allows access to own’s centre data only

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The ESID Online Patient- & Research Database



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>> Publications

This section offers downloads of all publications on the ESID Online Database for Primary Immunodeficiencies.

Listed chronologically, starting with the latest publication.
(Last updated: January 16th, 2009)

 [Gathmann, Knerr eUpdate \(2008\)](#) (279k)

Articles on the ESID Database and the UK PID Registry published in the online journal of the International Patient Organisation for Primary Immunodeficiencies (IPOPI - www.ipopi.org)

 [Grimbacher, Knerr et al Med Klin \(2008, German\)](#) (526k)

An article focussing on data from Germany (includes editorial notes). (PMID: 18813884)

 [Knerr et al Curr Opin Allergy Clin Immunol \(2007\)](#) (195k)

This article reviews recent publications both on PID database systems themselves and the research performed using results from these databases. (PMID: 17989523)

 [Gathmann DSAI immun? \(2007, German\)](#) (430k)

An article published in the journal of the German PID patient organisation DSAI (www.dsai.de).

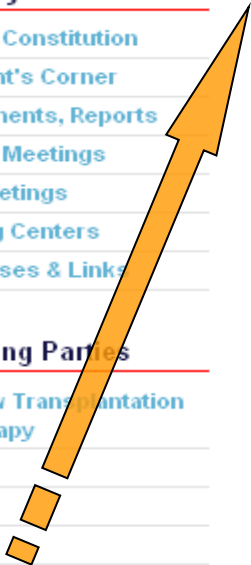
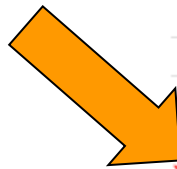
 [Guzman et al Bioinformatics \(2007\)](#) (455k)

A technical description of the database system (PMID: 17237056)

 [Eades-Perner et al Clinical Experimental Immunology \(2007\)](#) (237k)

First peer-reviewed publication of data in the ESID database (PMID: 17223972)

 [Knerr et al Central European J Immunology \(2006\)](#) (71k)



Thank you for your attention!



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