

The Spectrum of Primary Immunodeficiency Diseases in Iran Iranian Primary Immunodeficiency Registry (IPIDR)



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CONTRIBUTING CENTERS

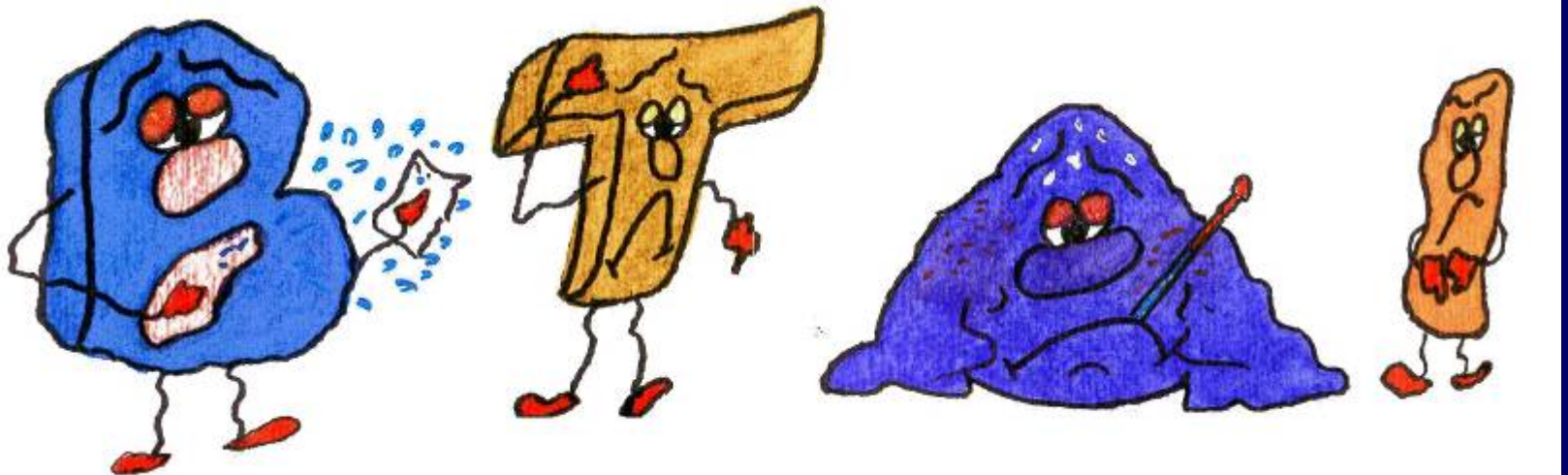
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BACKGROUND

Primary immunodeficiency disorders (PiD) are relatively rare disorders, characterized by an unusual susceptibility to infections.

There is wide geographical and racial variation in the prevalence and pattern of PiD.



OBJECTIVES



- To enhance the knowledge about these diseases among general practitioners and pediatricians
- To emphasize the importance of early diagnosis and treatment
- To determine the frequency of these diseases in Iran
- To stress the importance of teaching the clinical immunology in the medical curriculum
- To promote research about PiD in our country



IRANIAN PRIMARY IMMUNODEFICIENCY REGISTRY (IPIDR)

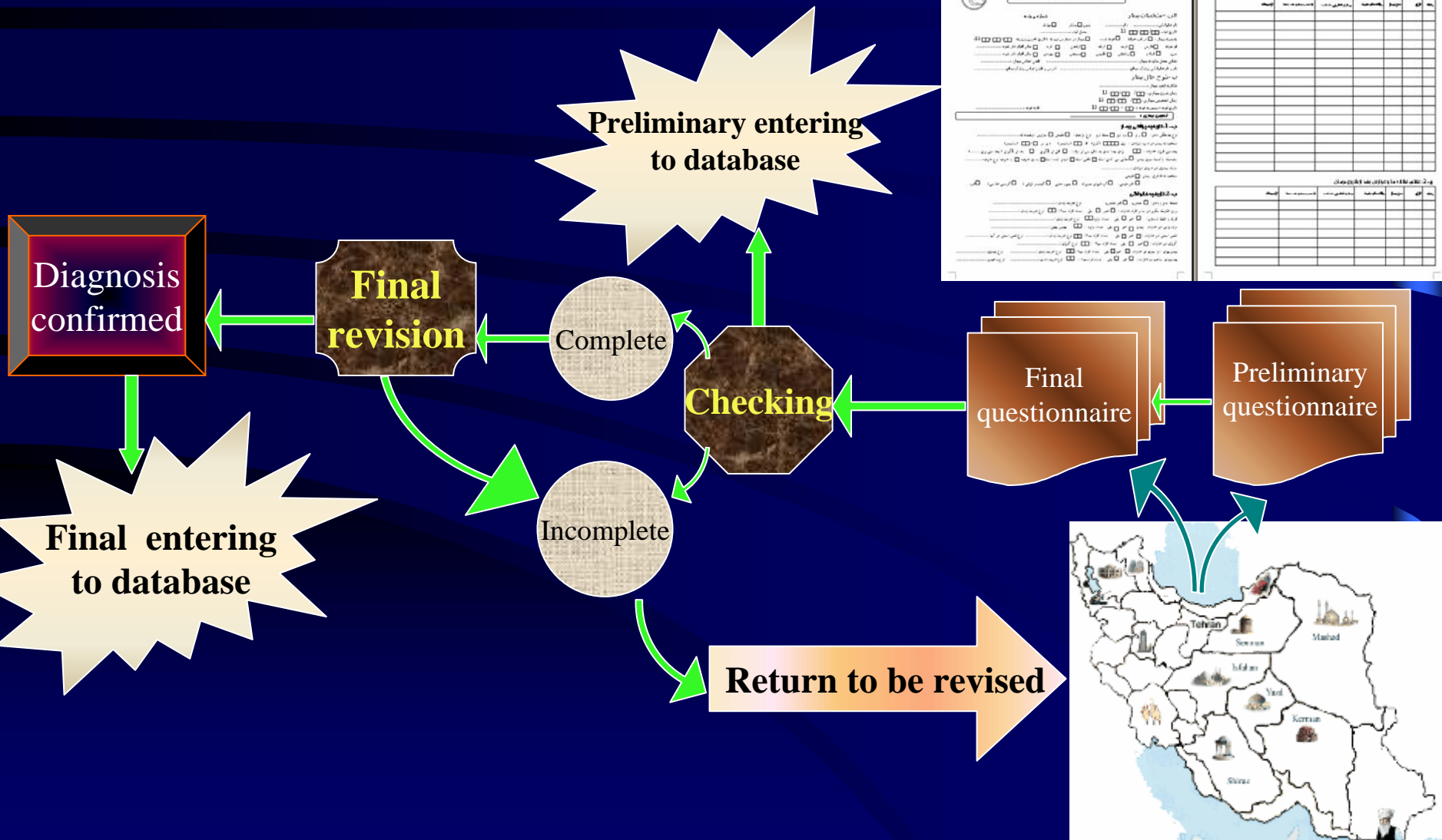


IPIDR was established in August 1999.

The clinical files of the 930 patients with
PiD were reviewed during three decades
(before 1980- 2006).



PROJECT OUTLINE



The first screenshot shows a form with a header in Persian and a table with columns for 'Name', 'Sex', 'Age', 'Date of Birth', 'Referral Source', 'Referral Date', 'Referral Hospital', 'Referral Doctor', 'Referral Address', 'Referral Phone', 'Referral Email', 'Referral Fax', 'Referral Website', 'Referral Other'. The second screenshot shows a similar form with a table for 'Referral Source' and 'Referral Date'.



PARTICIPATING CENTERS

IPIDR covered 12 universities of medical sciences from 10 major states of Iran, including;

- Tehran
- Fars (Shiraz)
- Khorasan (Mashhad)
- Isfahan
- Mazandaran (Babol)
- Azarbaijan (Tabriz)
- Zanzan
- Kerman
- Semnan
- Sistan & Balouchestan





COMPUTER DATABASE PROGRAM

A computerized database program was designed, based on our questionnaire, written with visual Basic language programming and using Access Database software.



PATIENTS

The diagnosis of PiD was based on standard criteria.

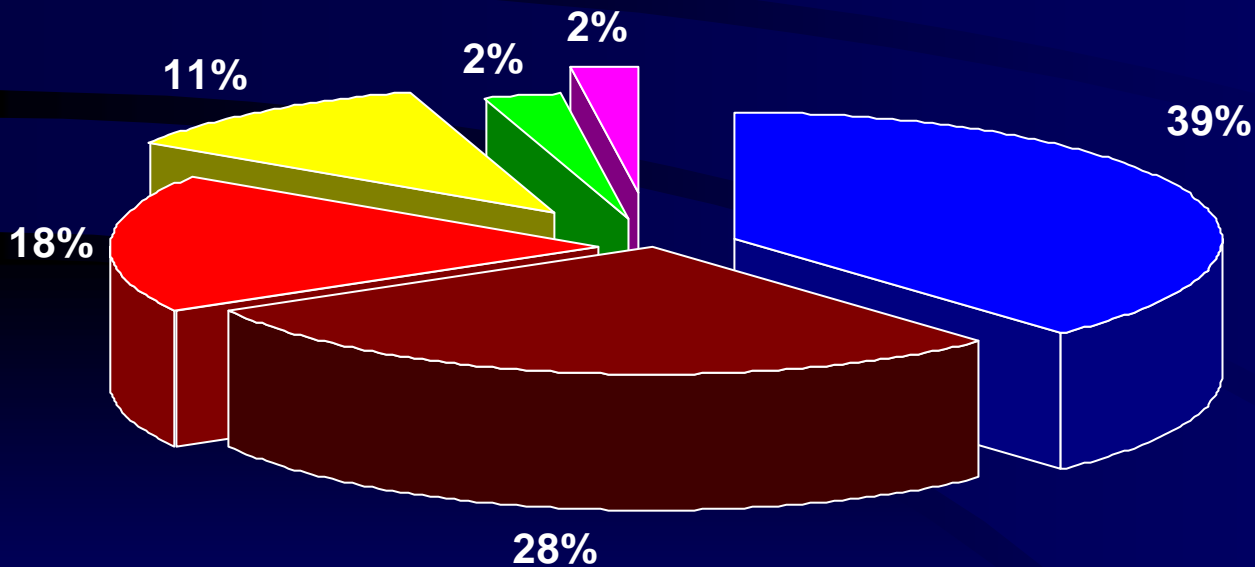
Only patients with well-established immunodeficiency and the clinical manifestations, compatible with their diagnosis, were included in our registry.





RESULTS

Nine hundred and thirty patients with PiD have been registered in IPI DR during three decades (573 male and 357 female).



- Predominantly antibody deficiencies
- Congenital defects of phagocyte number and/or function
- Other well-defined immunodeficiency syndromes
- Combined T- and B-cell immunodeficiencies
- Complement deficiencies
- Diseases of immune dysregulation



The most common PiD in our registry are:

Common variable immunodeficiency (CVID)	193 (20.8%)
Chronic granulomatous disease (CGD)	166 (17.8%)
Ataxia telangiectasia (AT)	94 (10.1%)
Btk deficiency (XLA)	69 (7.4%)
Selective IgA deficiency (IgAD)	55 (5.9%)
T-B-severe combined immunodeficiency	55 (5.9%)

The rest of PiD disorders were less than 50 in number (<5%)



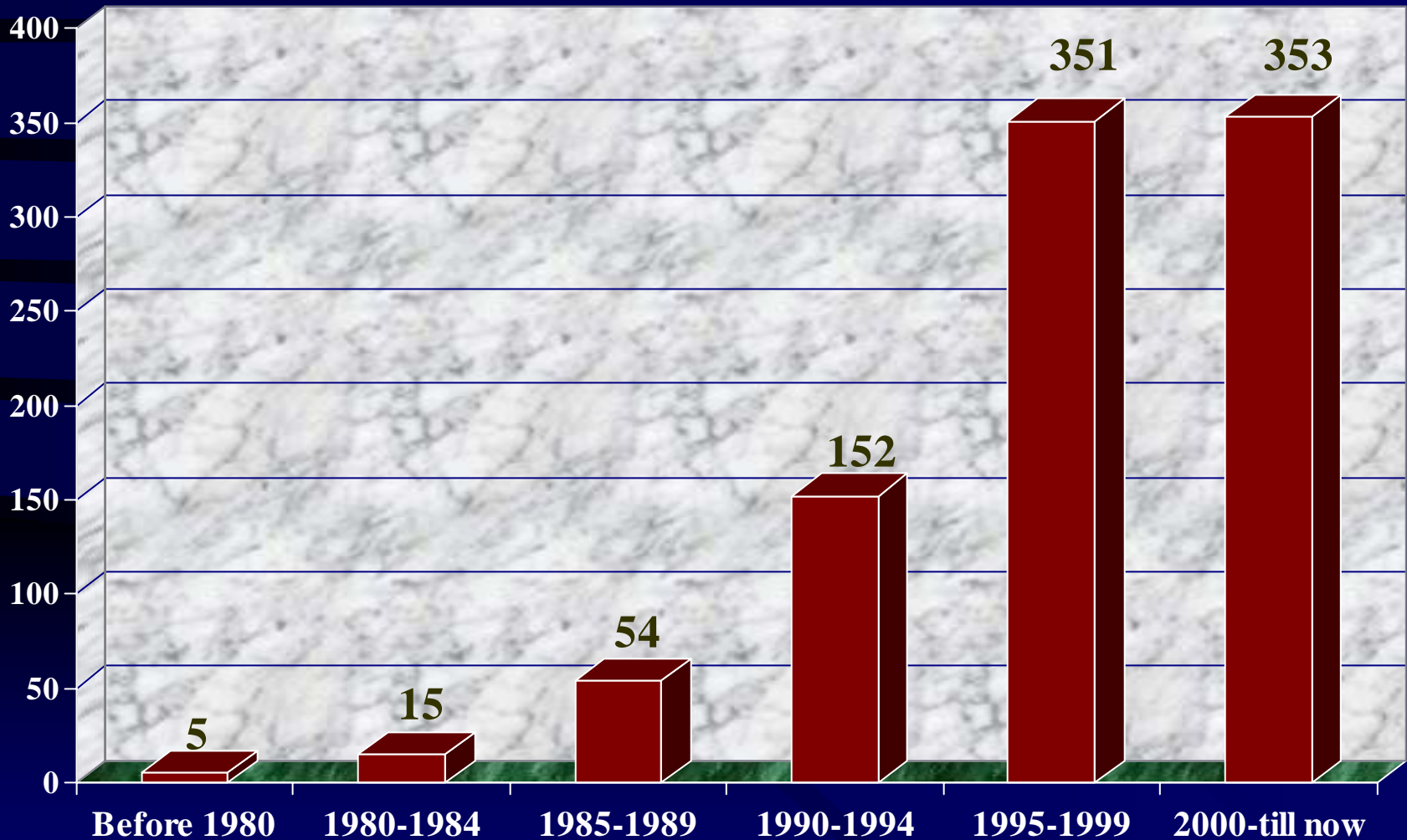
More than half of our PiD patients were in pediatric age range (59.3% of patients < 14 years).

The mean age of our patients at the time of study was 13.98 ± 9.3 years, with the youngest patient, referred to our registry, being 1 month old and the oldest having 82 years.

One hundred and sixty out of these patients have died because of recurrent infections (17.2%).



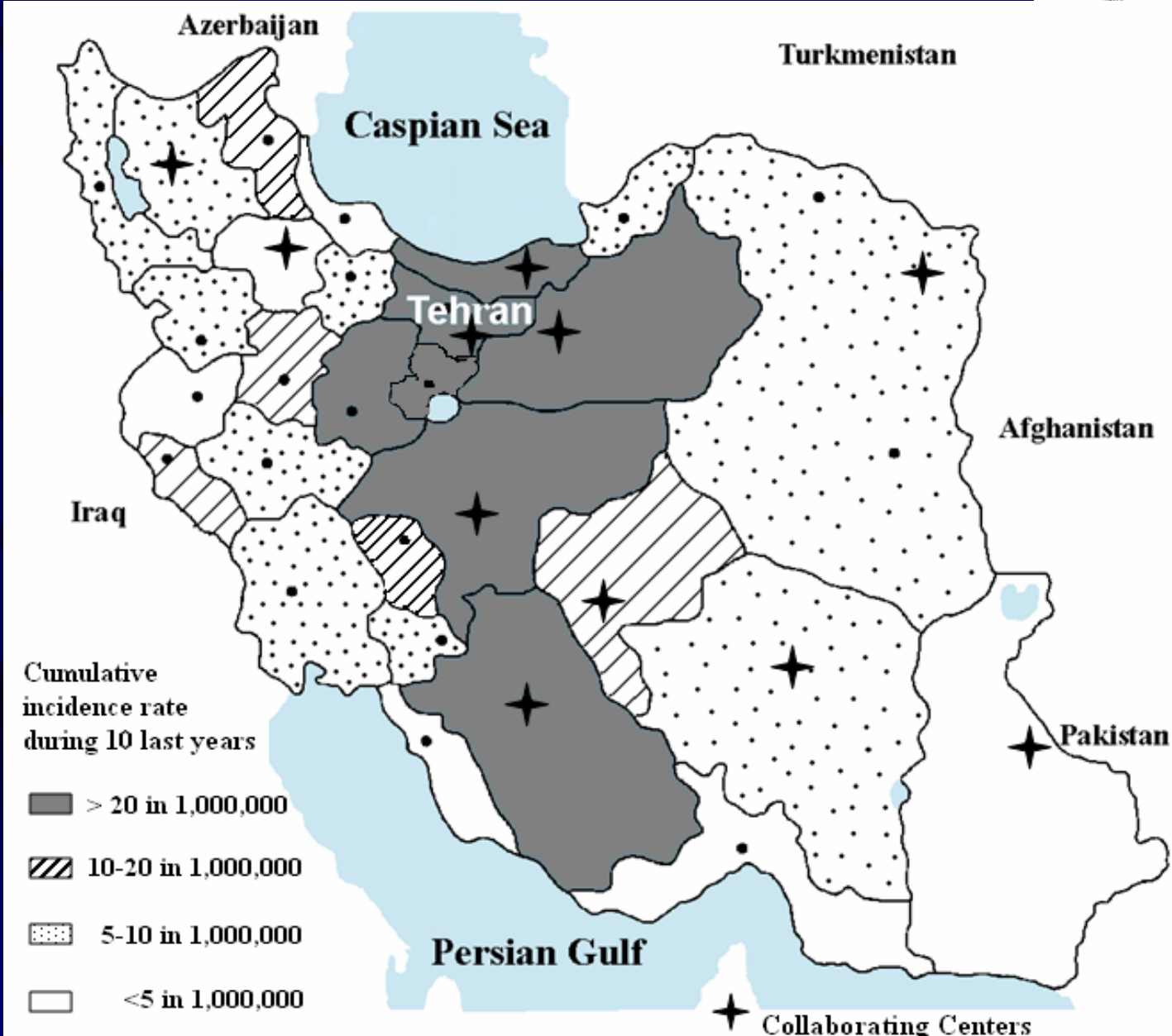
The number of patients diagnosed in 5-year intervals





The estimated occurrence of PID is about 6 per 100,000 live births.

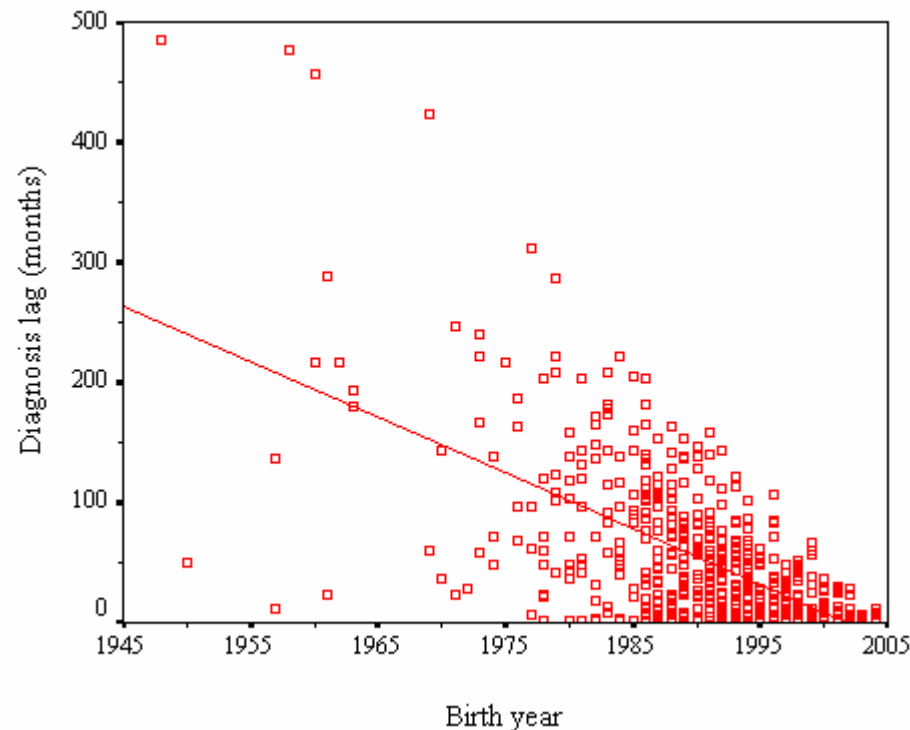
The cumulative incidence of PID is about 11.9 per 1,000,000 population during last 10 years, with a regional variation from 3 to 28 per 1,000,000 population.



The first infectious manifestation had occurred at a median age of 7 months (range: <1 month- 49 years).

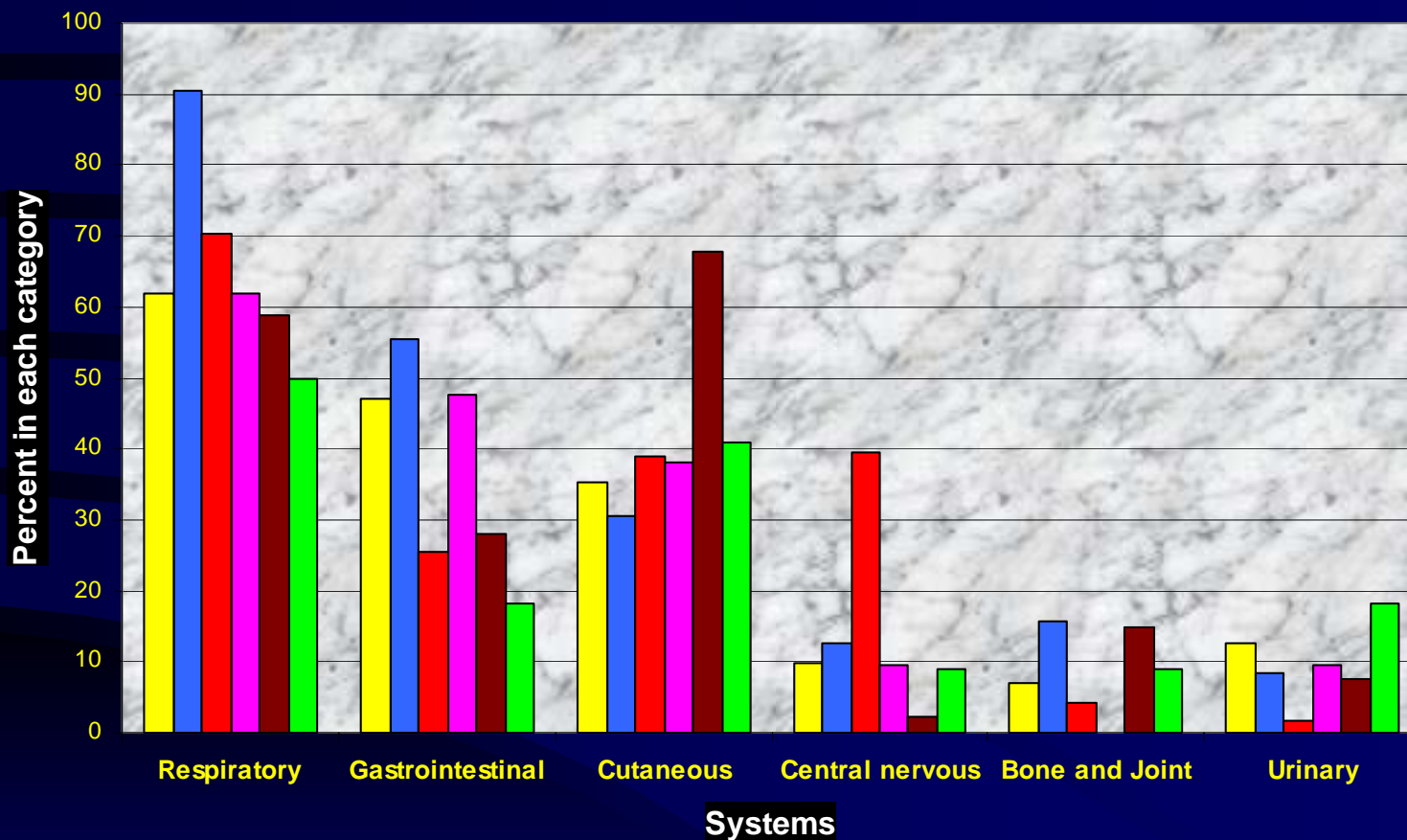
The median age of patients at the time of diagnosis was 57 months (range: 2 months- 54 years), with a median diagnosis delay of 31 months (range: 1 month- 40 years).

The diagnosis has increasingly been made at an earlier age in more recent years ($r = -0.625$, $F = 374.6$, $P\text{-value} < 0.001$)



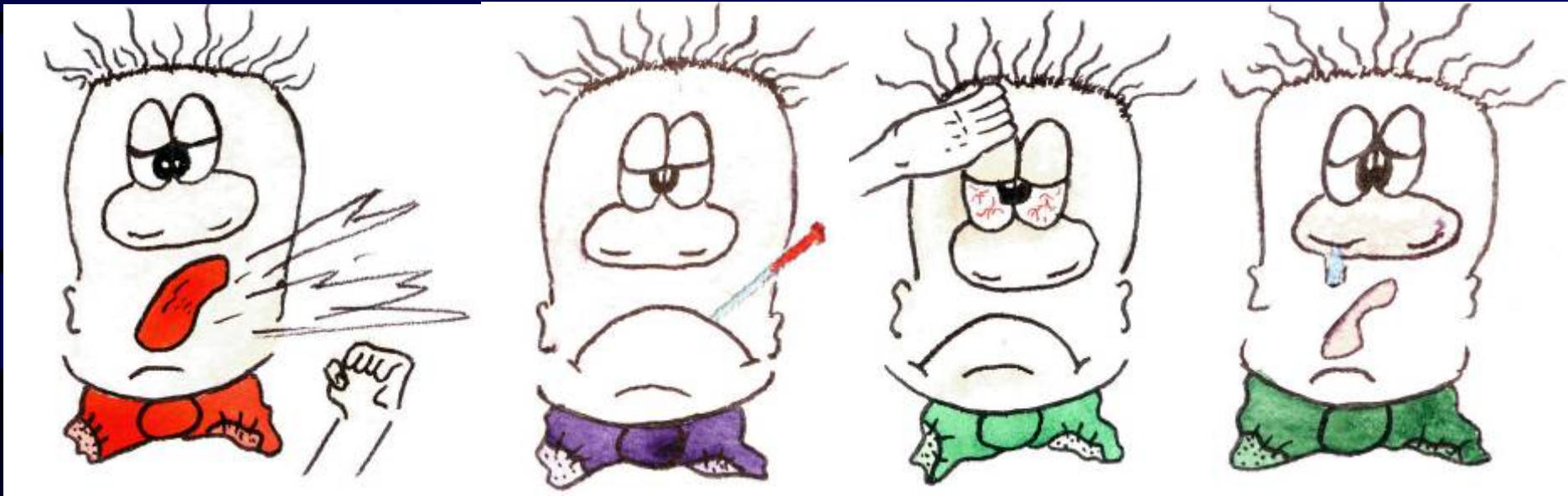


The infectious complications in PiD patients



- Combined T- and B-cell immunodeficiencies
- Predominantly antibody deficiencies
- Other well-defined immunodeficiency syndromes
- Diseases of immune dysregulation
- Congenital defects of phagocyte number, function, or both
- Complement deficiencies

All patients with recurrent infections should be screened for immunodeficiency.



Those patients with confirmed PiD should be registered as immunodeficient patients.

These definite PiD cases should be closely followed for development of infections and complications.

Their infections should be properly treated to prevent their further morbidity and mortality.



So, early diagnosis of PiD in suspicious patients should be considered to reduce the mortality and morbidity of these disorders.

In order to diagnose PiD patients earlier, the general knowledge of physicians should be increased.

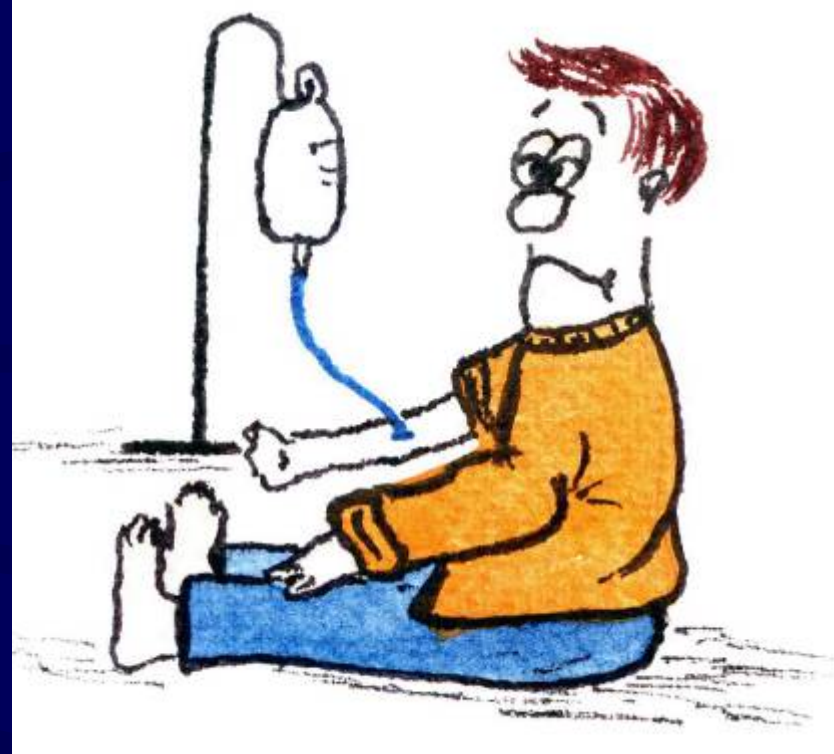
Such registries will increase the physicians' knowledge about such disorders.

Construction of such registry is much more important than merely for its epidemiological aspect; it can show the health impact of PiD and also increase the physician's awareness about such disorders.



We suggest that:

Iranian primary immunodeficiency registry to be further supported



1. to increase general knowledge of our physicians about the importance of PiD in Iran
2. to give a better care to the patients
3. to perform further researches on other aspects of immunodeficiencies

To keep the registry active, periodic contact must be maintained with all participants.

Thanks for your attention

