

Retrospective case studies of selective IgAD patients

Ellenes J. Zoltán MD

Childrens' Hospital Oradea, Romania

Prof. Dr. Maródi László

Dept. of Infectology and Pediatric Immunology,
University of Debrecen, Hungary

s IgAD

- most frequent PID
- co-morbidities:
 - atopic diseases
 - autoimmune diseases
 - malignancies

OR

asymptomatic, diagnosed by fortune

Goals of the study

to detect those factors that can influence +/-
the clinical presentation and prognosis of
the sIgAD

Methods

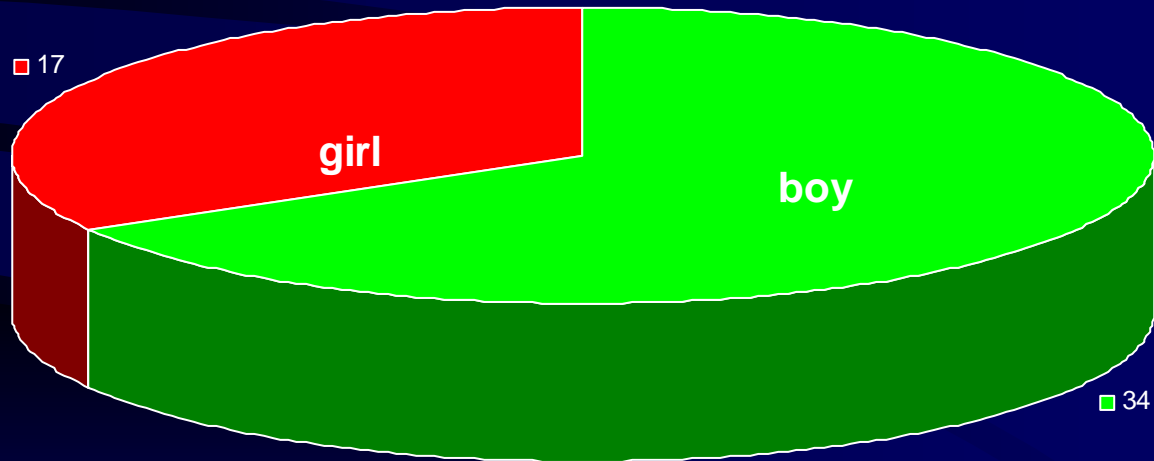
- Analysis of the clinical and laboratory data of the sIgAD patients treated at the Dept. of Infectology and Pediatric Immunology of the Debrecen University, period 1998-31.03.2005
- diagnostic criteria of the sIgAD (ESID):
 - age > 3 yrs
 - se IgA < 0.05 g/l at least 2 times , measured in more than 3months
 - secr IgA not detectable
 - other PID and secondary ID excluded

Studied parameters

- Family history:
 - PID, AID, neo. in relatives of I. and II. degree
 - atopic diseases in relatives of I. degree
- Personal history:
 - atopy, AID, malignancies
 - infections
- clinical presentation and lab parameters of the sIgAD during the follow-up period

Results 1.

IgAD patients



Results 2.

- Age : 5,6- 24,9 yrs. (average 12,4)
- Follow-up period: 2 months- 8 yrs.

Family immunological history

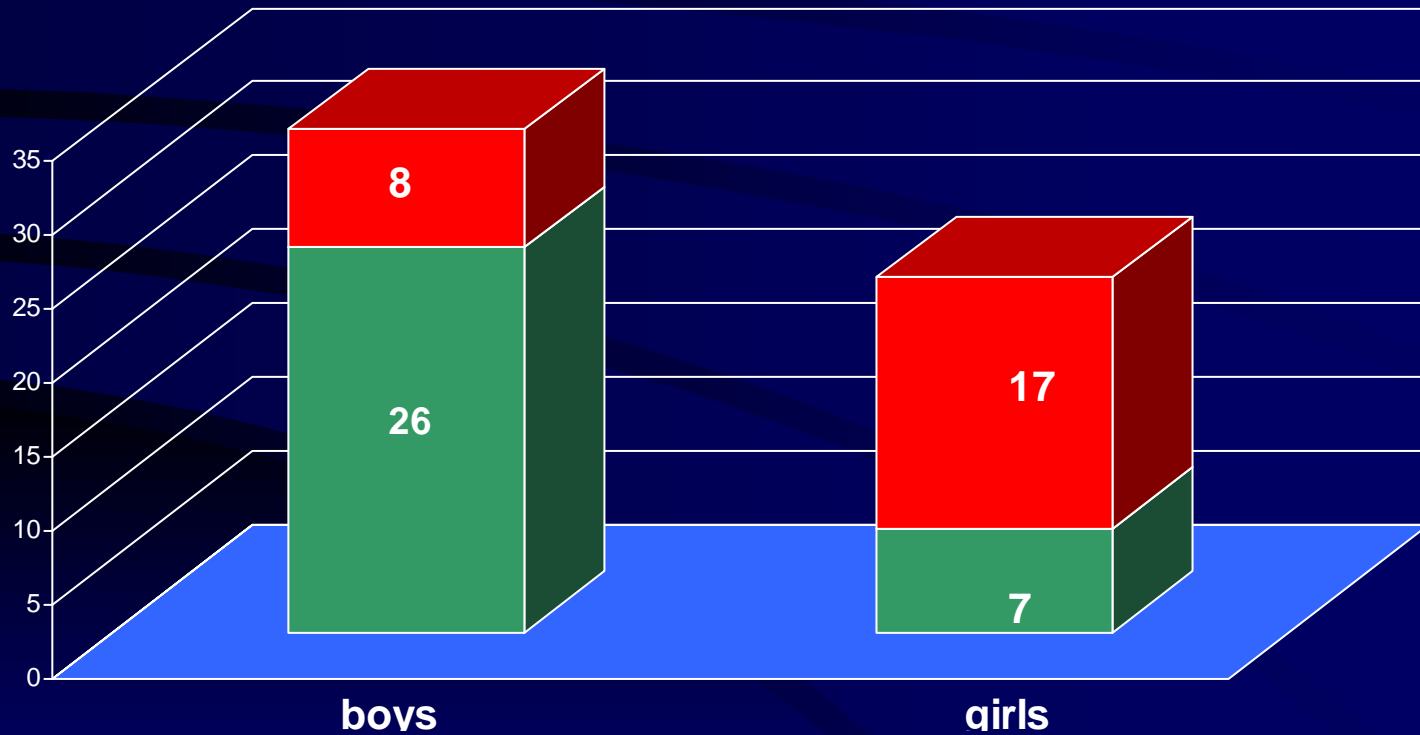
- PID: sIgAD 1 case mother, 2 cases brothers
- AID: 1-1 ITP, RA, colitis ulcerosa in one of the parents
- Neo:
 - I. degr. relatives.: 1 case
 - I and II. deg. Rel.: 1 case
 - II. deg.. Rel.: 9 cases, 2 cases cumulated
- Atopic diseases I. d.r.: 10 cases
- Neo and AID: 1 family

Personal immunological history

- Atopic diseases:
 - 33 cases (64,7%): M 26 (76,4%), F 7 (41,1%)
 - asthma: 11 patients
 - rhinitis allergica: 15 pts
 - asthma and rhinitis: 4 pts
 - dermat. atopica, urticaria, food allergies
 - 10 families: one atopic parent → 9 atopic children (90%)

Atopic patients

Atopic patients



AID

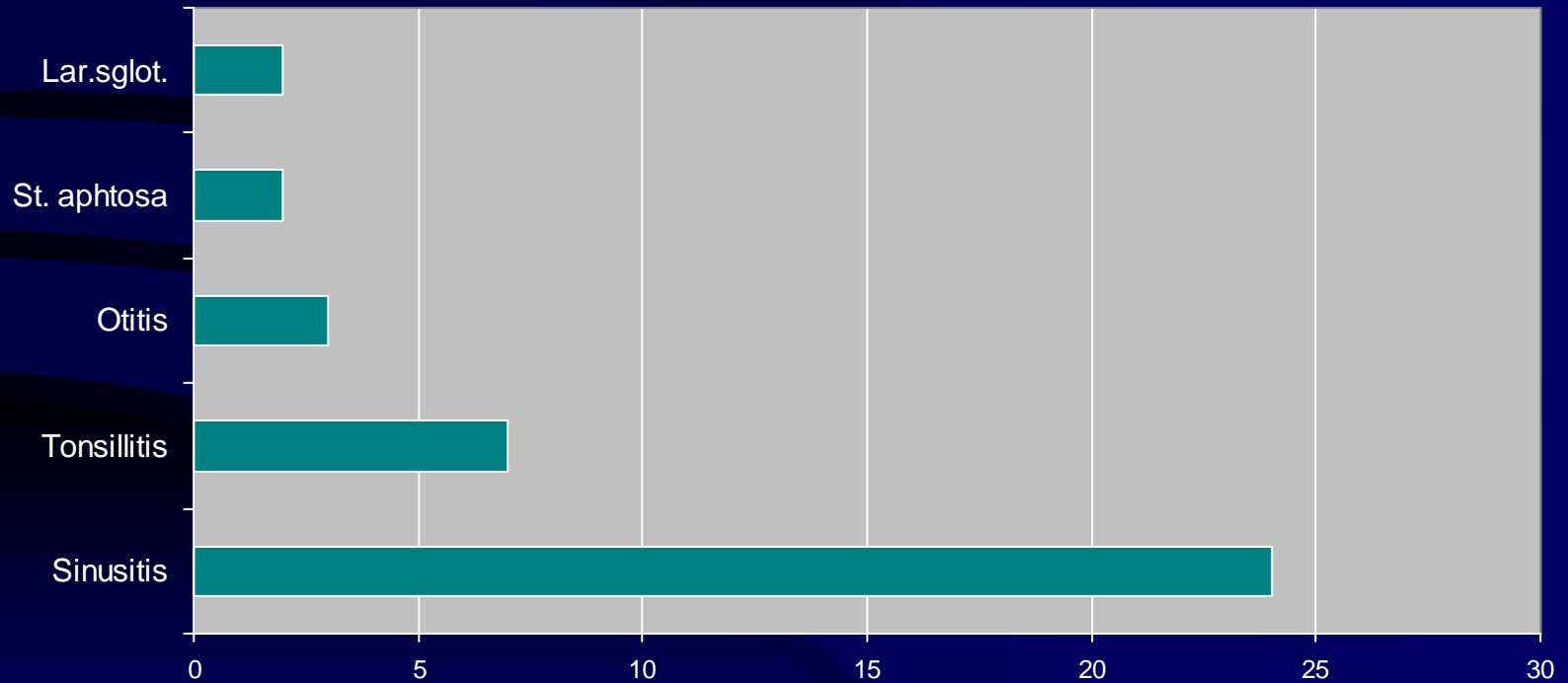
- JRA 3 pts
- co-morbidities:
 - 2 pts sIgAD + JRA + Rhinitis allergica
 - 1 pt sIgAD + JRA + Sp AD

Infections

- Recurrent purulent upper respiratory tract inf: 42 pts
- frequent infections: sinusitis, pharyngitis, tonsillitis
- rare inf.: otitis, stomatitis aphthosa
- frequency of the URTI related with aging:
 - decreased - 29 pts
 - increased - 4 pts
 - same- 9 pts
- LRTI: 3 bronchitis, 3 pneumonia, not recurrent

Infections 2.

URTI



Infections 3.

- O-R-L interventions: 30 pts
 - 2 tonsillectomia
 - 14 pts adenectomy
 - 16pts tonsilloadenectomy
- decrease of respiratory infections after the surgical interventions: 3 pts !

Labor 1.

- IgG: 7 pts 18-20 g/l, extremely rare infections
- IgG subclasses (20): 13 NV, 3 IgG2 ↓, 2 IgG4 ø, 1 pt IgG2↓+ IgG4 ø - recurrent infections
- SpAB (42): 32 NV, 10 ↓
 - recurr. Inf.: - a-HIB ø + IgG2↓+ IgG4 ø
 - » a-HIB ø + a- SPn negative test vaccination
- IgM↑ (IgG- NV): 3 pts: rare inf.
- IgE ↑: 18 pts- 16 atopic diseases (also in other 17 pts, with normal IgE values!)

Labor 2.

- Complement (CH50, C3, C4): 24 pts
- Ly-subpopulations: 29 pts
- T-cell activity: 4 pts

NV

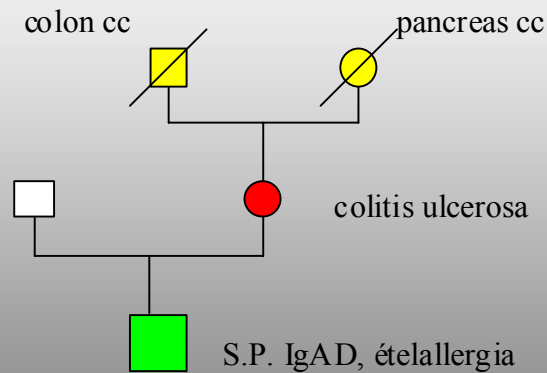
- JRA pts: RF negative, ANA gran. pos.
- Viral serology:
 - EBV, CMV 3 pts: IgM neg, IgG pos.
 - HBs-Ag, anti-HCV Ab: 2 pts, negative

Comments 1.

- M/F = 2:1 (N = 51) gender as risk factor ?
- Familial risk factors:
 - neo. I dr. 1 family (2%) → not RF for IgAD
 - AID I dr 3 pts (6%) → relevancy ?
 - IgAD 1 family ; IgAD inheritance TACI-R ?
 - Atopic parents: not RF for IgAD, but RF for associated atopic disease
 - cumulated immunological disturbances: RF for IgAD ?

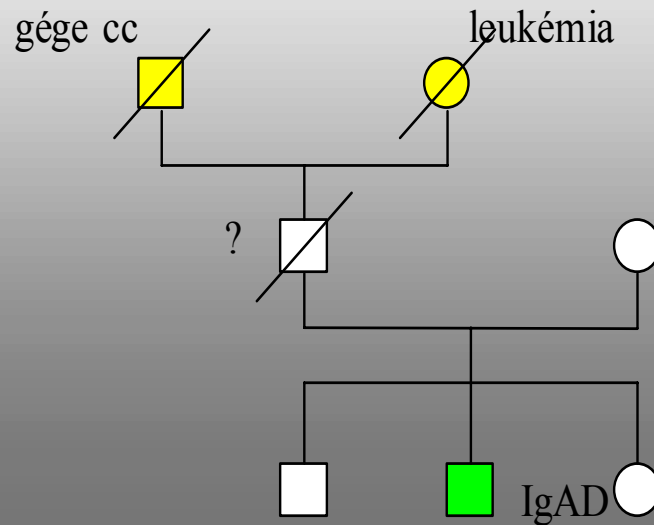
Families with aggregated immunological disturbances 1.

. S.P., fiú, szül. 1997. október



Families with aggregated immunological disturbances 2.

2. E.Z., fiú, szül. 1987. július



Personal immunological history

- Atopic diseases: 64,7% of pts, M 76,4 %, F 41,1 % ↑general population → IgAD: RF for atopic diseases
- JRA 3 pts (11,76%) ↑ general population → IgAD: RF for AID ?
- IgAD and IgG subclass deficiencies (IgG2 and IgG4): which one is the cause of the recurrent URTI ?
- AID, atopy associated with IgAD does not increase the frequency of URTI!
- The frequency of URTI ↓ aging
- tonsillotomy does not ↓ the frequency of URTI!!
- ↑ total IgG-protection against URTI?