

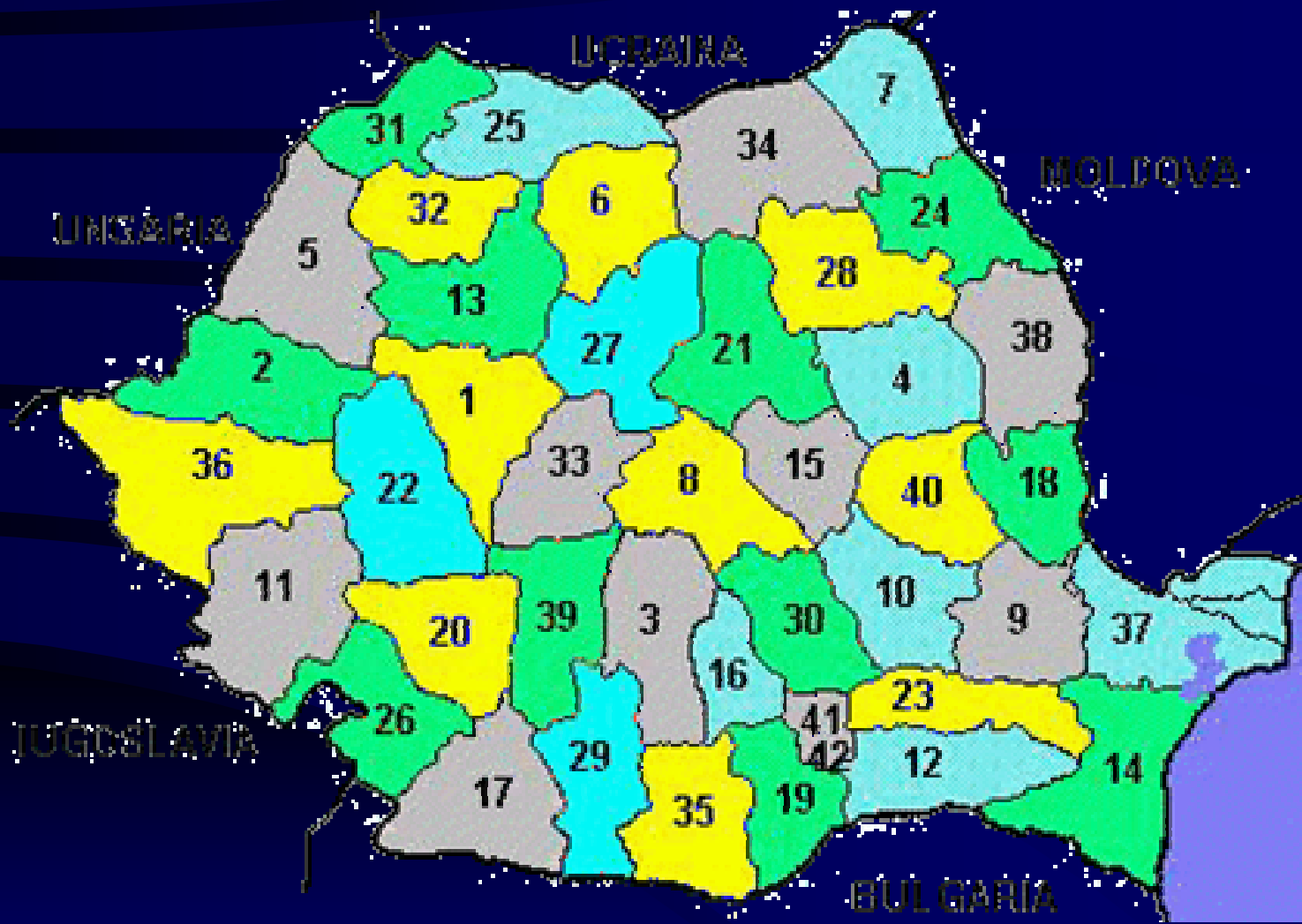
The Pediatric PID Register of Bihor County 1992-2004

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Spectrum of diseases

- Immunoglobuline class deficiencies: 49 patients
- Transitory hypogammaglobulinemia of the infant: 6 pts
- Hyper IgE syndrome: 1 pt
- Complement system deficiencies: 4 pts
- TOTAL : 60 patients (33 males, 27 females)

Immunoglobuline class deficiencies

- XLA: 2 ± 3 pts
- IgA: - selective deficiency: 4 pts
 - seric and secretory def.: 8 pts
 - seric def.: 13 pts
 - secretory def.: 7 pts
- IgG deficiency: 9 pts
- IgA and IgG def (susp. CVID) : 3 pts

XLA

Name, gender	Birth date	Comments	Treatment
• D. V., M	15.08.1991	Moved to Timisoara	Octagam
• K. L., M	3.11.1996	Dg confirmed in Timisoara; IVIG recommended but not done; died at age 4 yrs ; Three brothers from the same mother but two fathers, died in early infancy (purulent meningitis, bronchopneumonia)-possibly	
XLA, not investigated			

Selective IgA deficiency (IgA= 0 g/l)

Name, gender	Birth date	Comments	Treatment
• A. T., F	24.06.1998	Recurrent inf. resp. infections	
• B. G., M	9.07.1993		
• C. N., M hypotrophia	28.11.1996	Polydactylia, cavernous haemangioma,	
• U. B., F	29.07.1999	Coeliakia	

IgG deficiency (IgG < N.V.)

Name, gender	Birth date	Comments	Treatment
• M. A., M	3.04.1997	Testvaccine Pneumo 23	
• R. T., F	13.12.2000	Rec. inf. resp. infect	
• L. A., F	1993	Congenital teleangiectasia but not AT ; 4 sisters, 1 brother and mother : the same dg.	

IgG and IgA def. (IgG, IgA < N.V), susp. CVID

Name, gender	Birth date	Comments	Treatment
• C. D., F	7.10.1996	Rhinitis allergica	Flixonase, loratadine
• P. M., F	2.04.1999		
• Ş. A., F	25.07.2000	Rec. inf. resp. infect	

Transitory Hypogammaglobulinemia of the Infancy

• Name, gender	Birth date	Comments	Treatment
• G. K., M	20.12.2001	Def. of C4	
• K. D., M	6.09.2001		
• M. C., M	8.02.2001		
• C. A., M	28.05.2001		
• C. T., M	1.07.2003		
• N. B., M	10.07.2002	Dermatitis atopica	Loratadine, Elidel

Hyper IgE syndrome

Name, gender	Birth date	Comments	Treatment
• P. Á., M	22.08.1993	Dermatitis atopica, asthma bronchiale, rhinitis allergica, toxico-septic pyodermitis	
			– Trt: Flixotide, loratadine, Elidel

Complement system deficiencies

Name, gender	Birth date	Diagnosis	Comments	Treatment
• F. E., F	2.06.1993	HANO	Virilism, iatrogenic Cushing s.	Danazol, predn.
• B. I., F	1989	C4 deficiency	L.E.S, Antiphospholipidic s.	
• A. A., M	21.08.1999	C4 deficiency	Atopic angioedema	Loratadine
• L. O., M	28.02.1988	C3, C4 def.	Asthma bronchiale	Flixotide

Associations: PID and other immunological disorders

- Different PID: THI and C4 def- 1 pt
- PID and autoimmune diseases:
 - IgAD and IDDM: 3 pts (1 M, 2 F)
 - C4 def and LES and APL: 1 pt
 - secr IgAD and coeliakia: 1 pt
- PID and allergic diseases:
 - IgAD and asthma+allergic rhinitis+rec. urticaria: 1 pt
 - secr IgAD and asthma: 1 pt
 - THI and atopic dermatitis: 1 pt
 - IgA and IgG def and allergic rhinitis: 1 pt

Intrafamilial cumulation of PID

- XLA: 4 brothers
- IgAD (seric and secr): 2 twins
- secr IgAD: 1 brother and 1 sister
- IgG def. and congenital teleangiectasia: 5 sisters, 1 brother and mother

PID and genetical disorders

- IgG def and congenital telangiectasia: 5 sisters, 1 brother, mother
- selective IgA def and polydactylyia, cavernous hemangioma, hypotrophia- 1 pt
- secr IgAD and Ehlers- Danlos syndrome- 1 pt