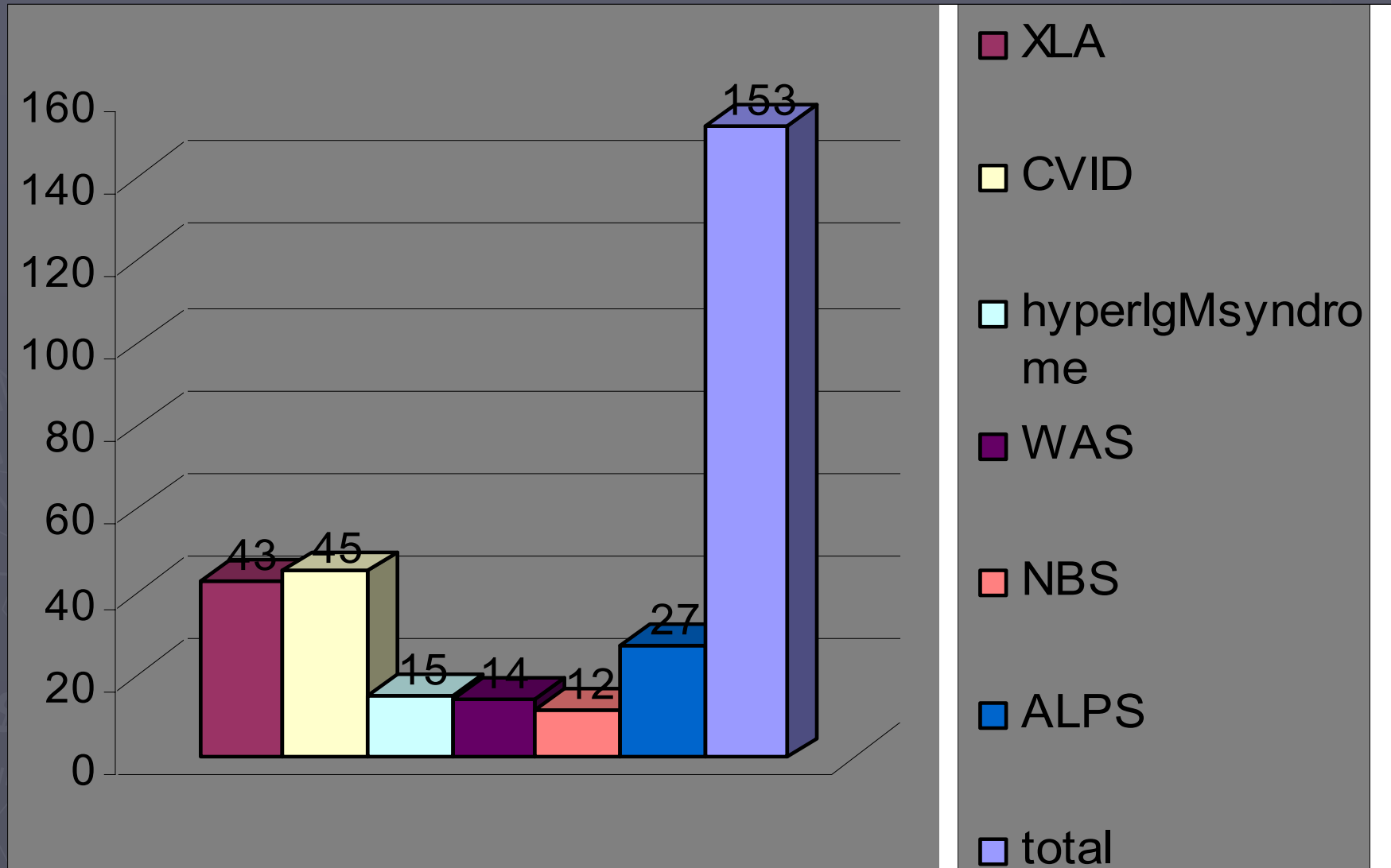


Hematological manifestations of primary immunodeficiencies.

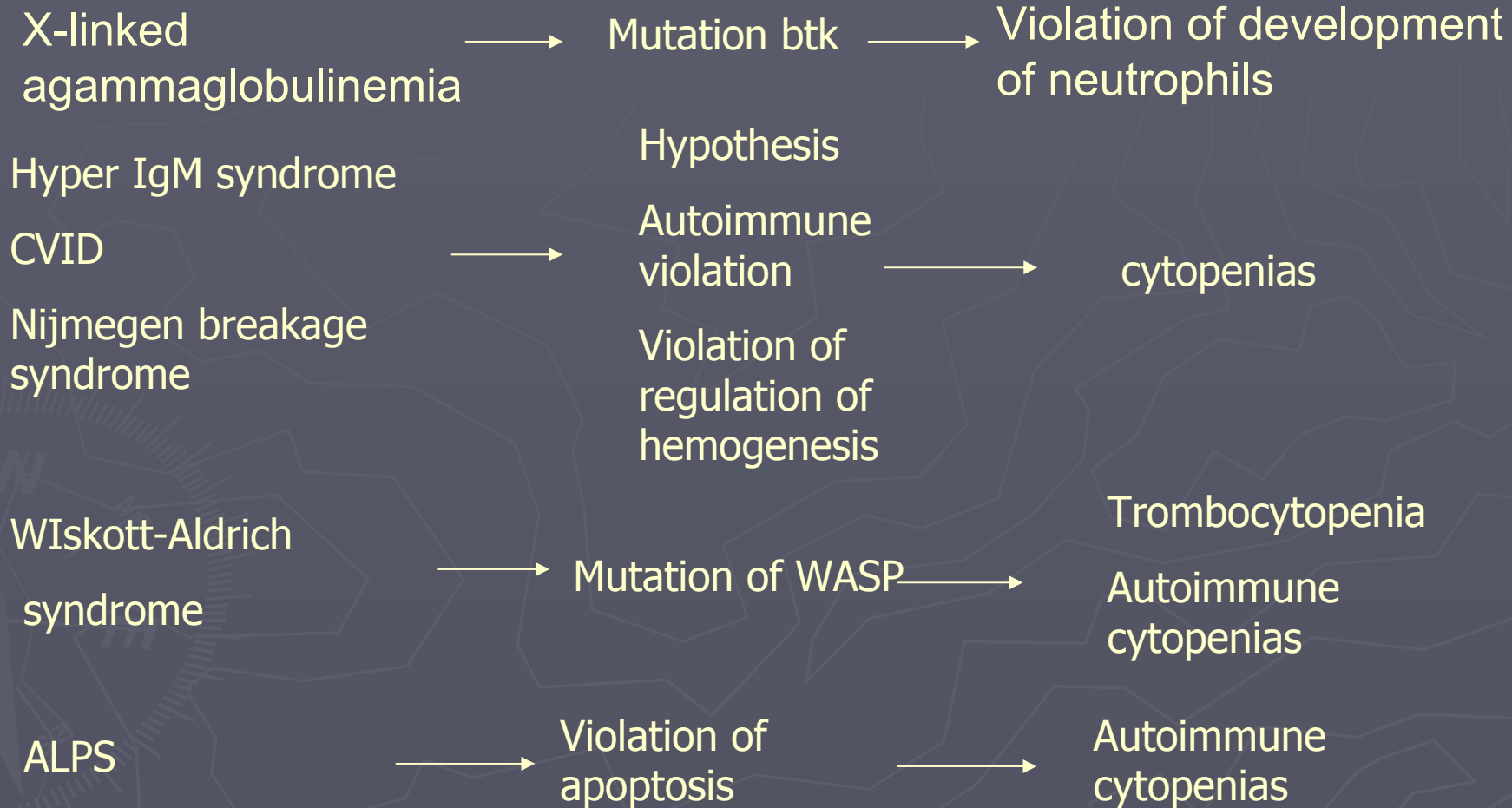
A.S. Yurasova, M.A. Kuznetsova, I.V. Kondratenko

Russian Children's Clinical Hospital
Moscow

Forms of primary immunodeficiencies



Mechanism of development of cytopenias



Cytopenias in primary immunodeficiencies.

Quantity of patients	Neutropenia	Trombocytopenia	Anemia
XLA(43)	17		12
CVID (49)	10	1	2(hemolytic anemia)
Hyper IgM syndrome (15)	10	2	3(hemolytic anemia)
Nijmegen breakage syndrome (12)	7	1	Aplastic anemia-2
Wiskott-Aldrich syndrome (14)	3	11	4+3 hemolytic anemia
ALPS (27)	17	17	9

Therapy of cytopenias in patients with Hyper IgM syndrome

Medication	Quantity of patients	Effects of therapy
Prednisolon 2 mg/kg → ↓dose	2	Good effect in 2 patients
G-CSF 3-5 mkg/kg	5	Good effect during the therapy
G-CSF 10 mkg/kg	1	Good effect during the therapy

Therapy of cytopenias in patients with NBS and CVID

Therapy	Form of immuno deficiencies	Cytopenias	Quantity of patients	Effect
Prednisolon 1-2 mg/kg	CVID	Different cytopenias	3	3-good
GM-CSF 3-5 mg/kg	CVID	Leucopenia, neutropenia	2	2-without effect
CsA +ATG	NBS	Aplastic anemia	2	2-without effect

Therapy of cytopenias in WAS

Method of therapy	Character of cytopenias	Quantity of patients	Effect
Splenectomy	Thrombocytopenia	6	6-Good effect
Methylprednisolone 20 mg/kg N3	Thrombocytopenia	1	Good effect
IVIg 2g/kg	Thrombocytopenia	1	Temporary effect
Prednisolone 7,5mg+azathioprine 5 mg/kg	Thrombocytopenia	1	Good effect
Prednisolone 2 mg/kg+azathioprine 3mg/kg	Hemolytic anemia	2	Good effect
G-CSF 3-5 mkg/kg	Neutropenia	1	Good effect

Therapy of cytopenias in patients with ALPS

Method of therapy	Character of cytopenias	Quantity of patients	Effect
Prednisolon 1-2 mg/kg		17	7-good 5-partial
Methylprednisolone 20 mg/kg IV N 3	Cytopenias resistant to therapy	5	3-good
IVIg 2 g/kg		7	1-good 1-partial
CsA 4-7 mg/kg per os	Trombocytopenia Neutropenia	2	Without effect
Vincristine		2	2-good
Splenectomy		6	2-good 3-partial
G-CSF 3-5 mkg/kg	Neutropenia	4	1-good