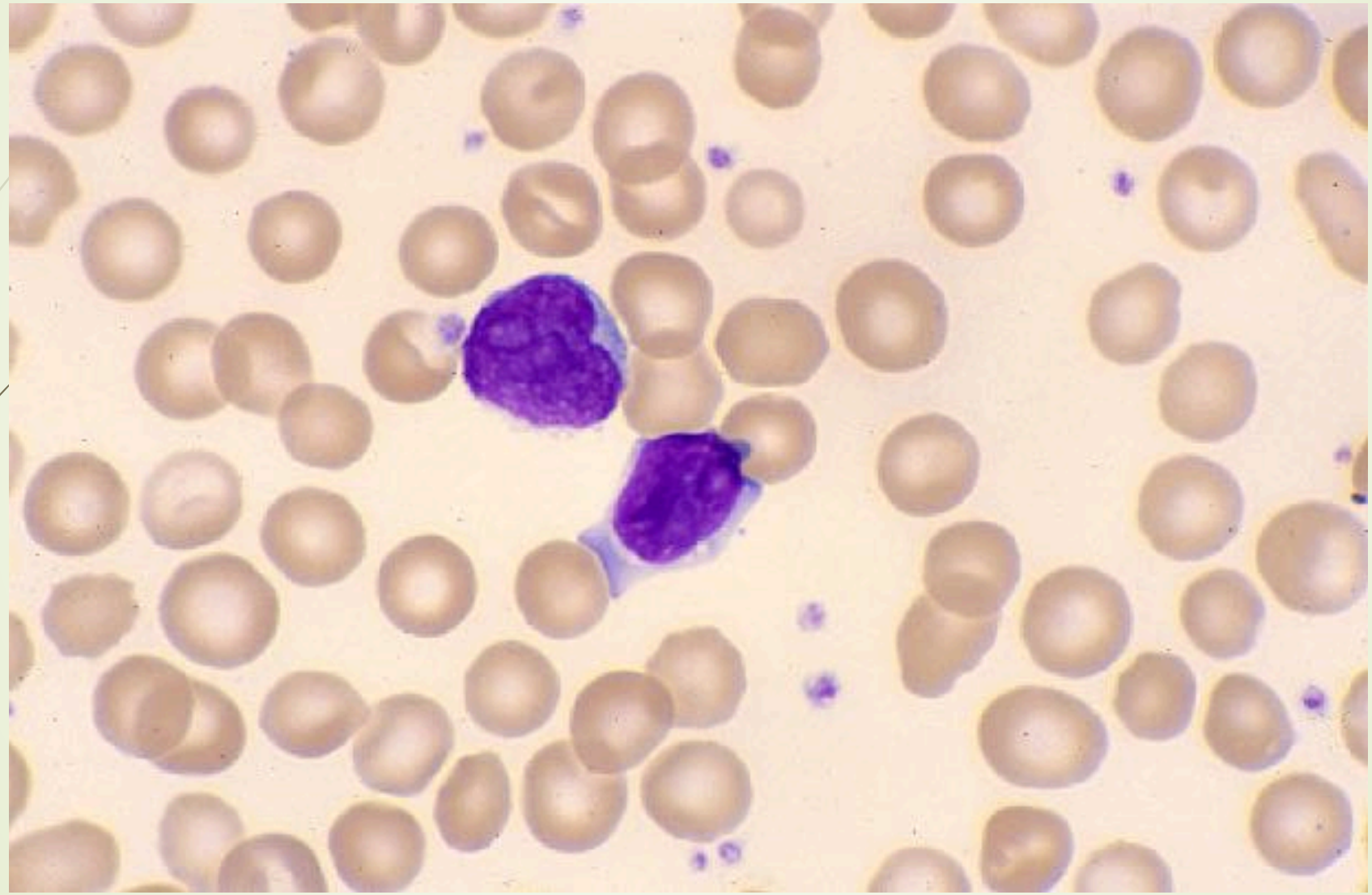
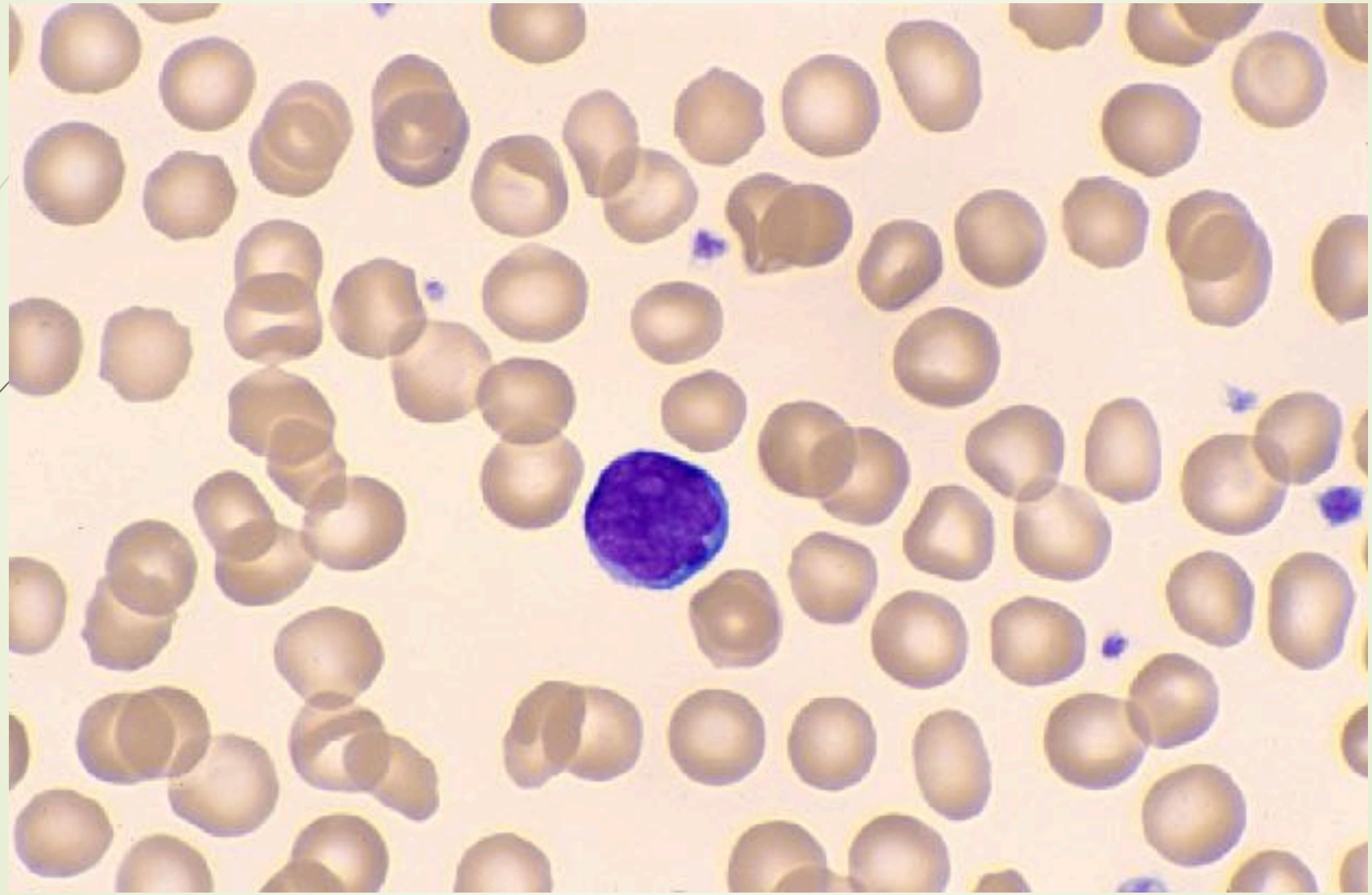
PAEDIATRIC LYMPHOCYTES

Gillian Rozenberg

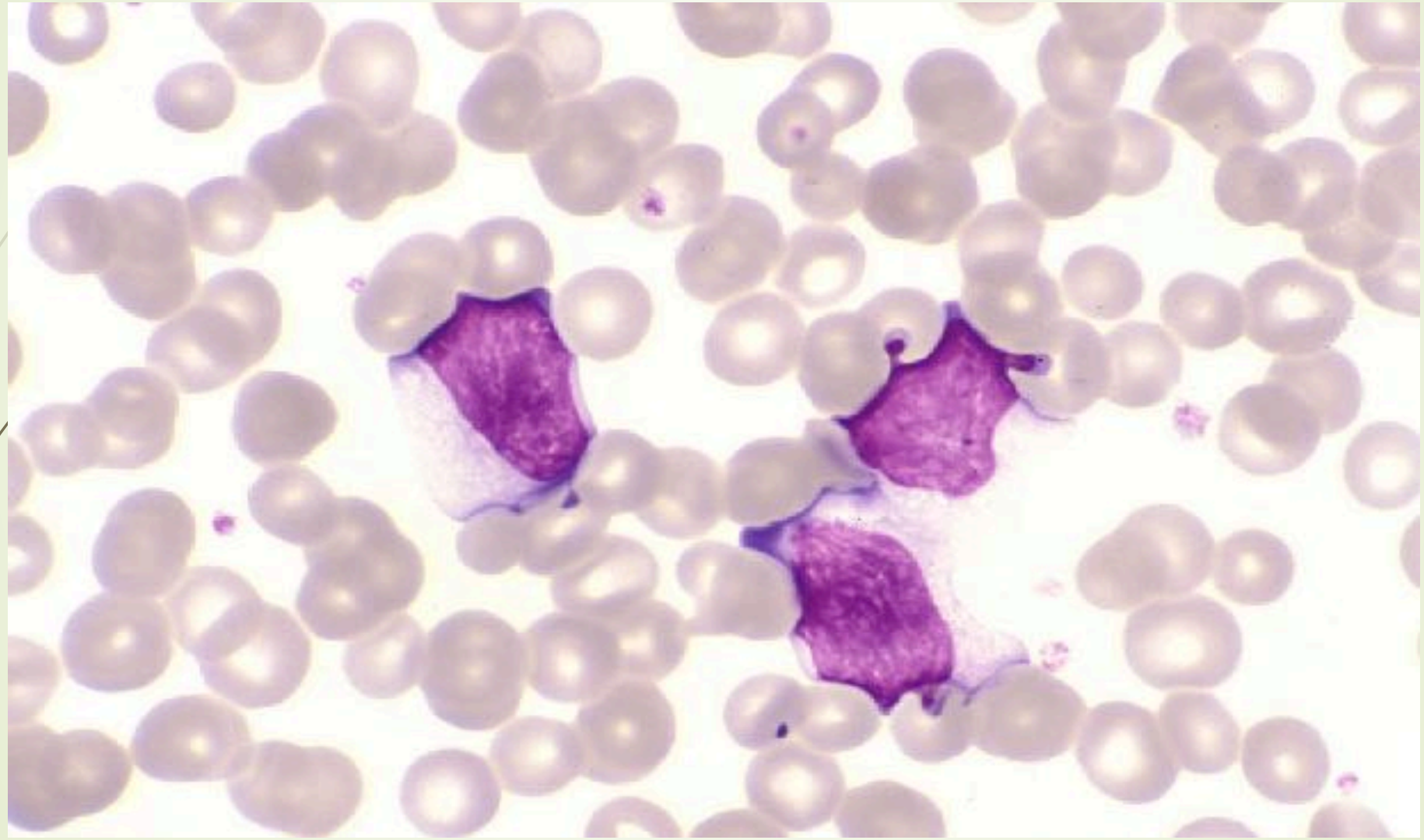
Lymphocytes in a 1 day old neonate



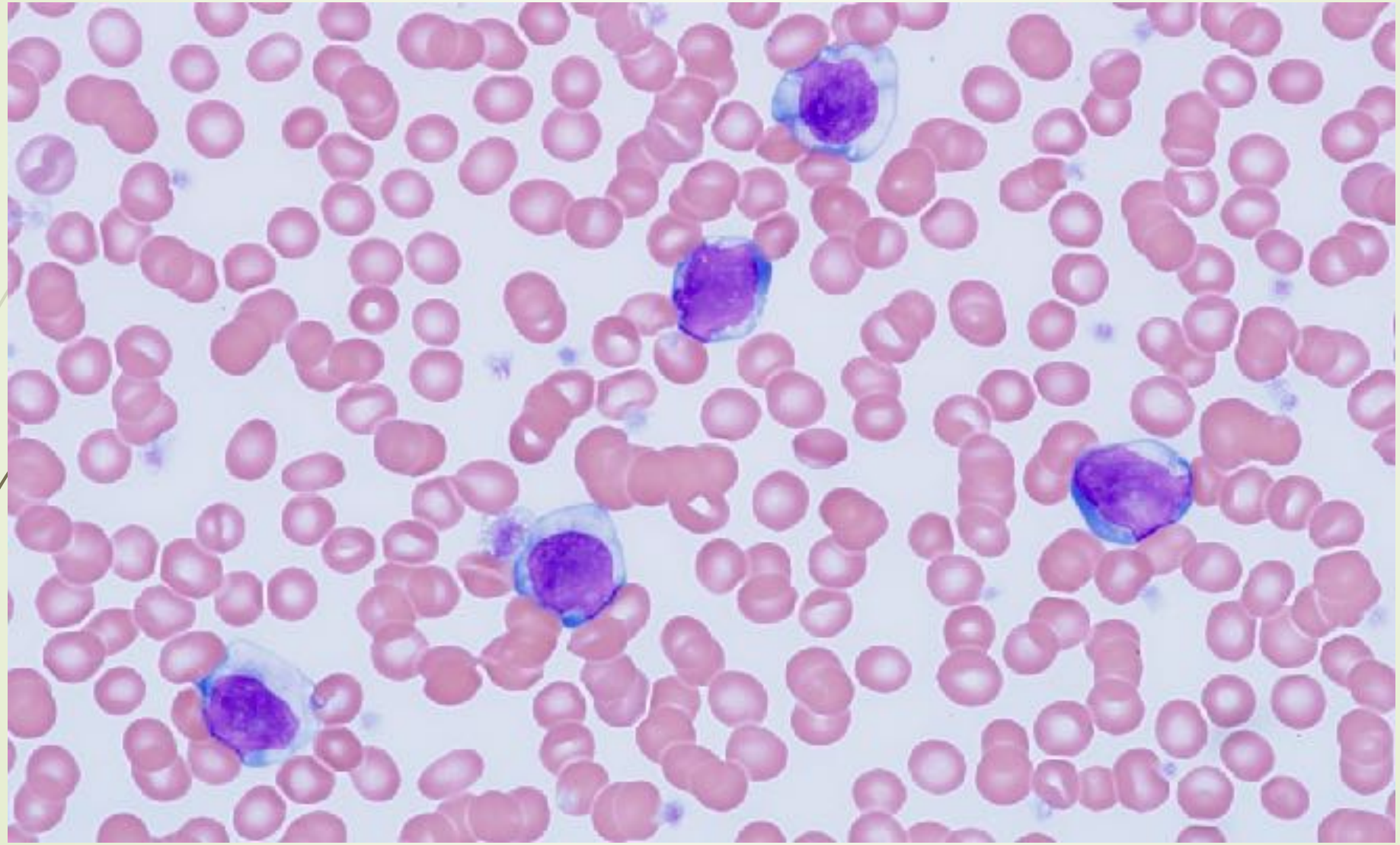
Lymphocyte in a 1 day old neonate



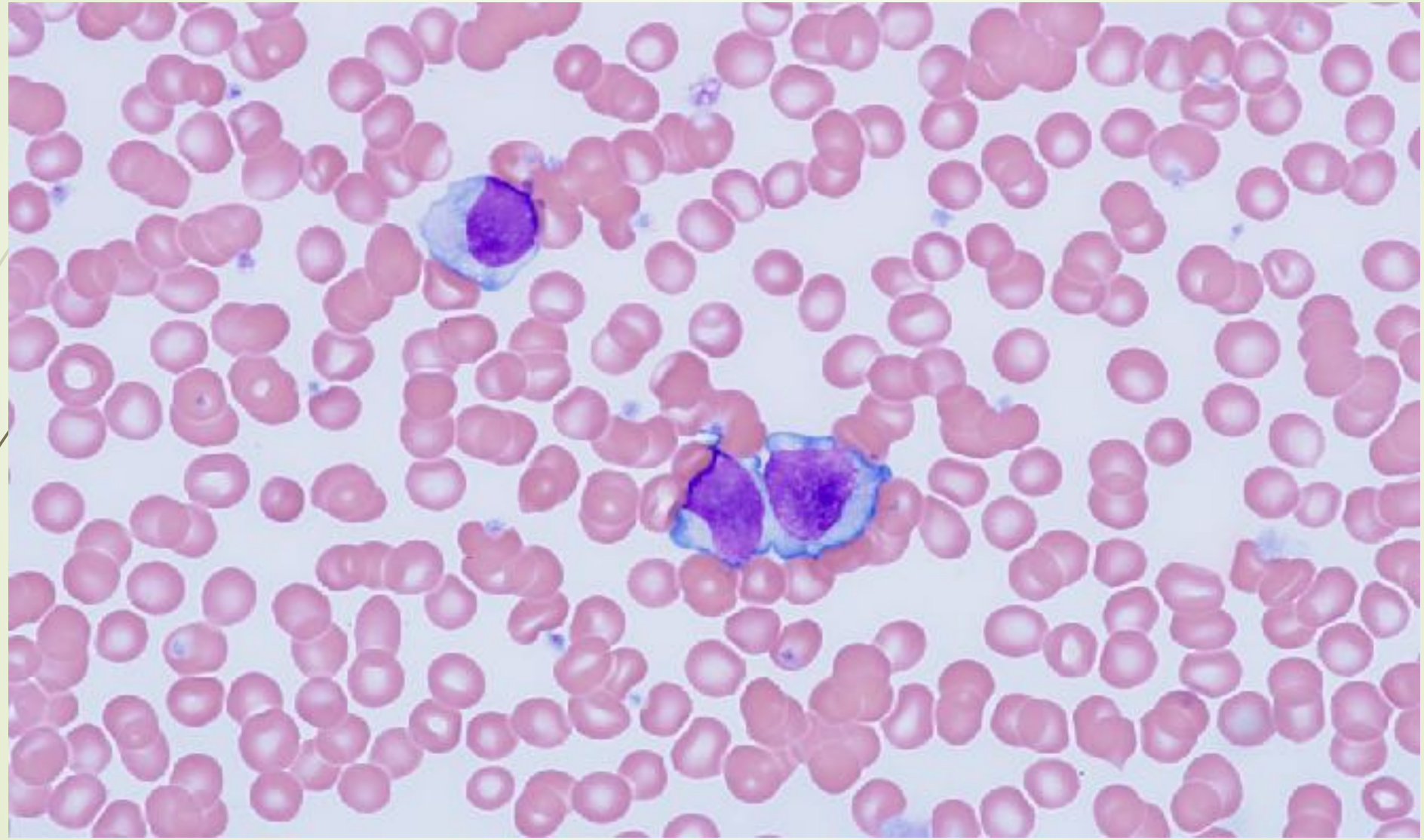
Reactive lymphocytes (Infectious Mononucleosis)



Reactive lymphocytes (viral infection)



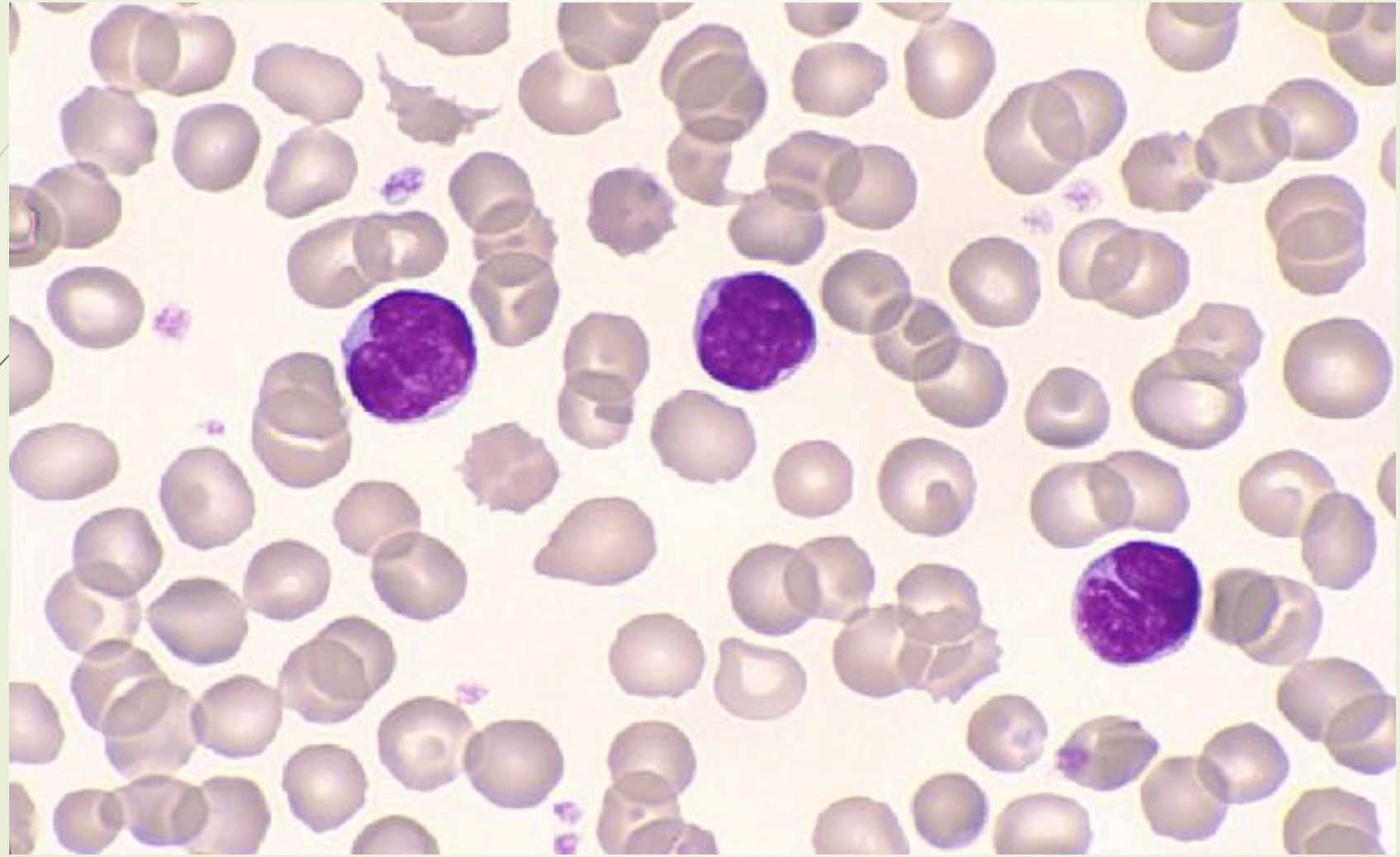
Reactive lymphocytes (viral infection)



Reactive lymphocytes

- Reactive lymphocytes are CD8+ cytotoxic T lymphocytes that are attracted towards the antigen target and commence production of cytokines
- Activated CD8+ cytotoxic T lymphocytes are increased in size and have flowing basophilic cytoplasm
- The nucleus is enlarged and the chromatin fine and often contains one or more nucleoli

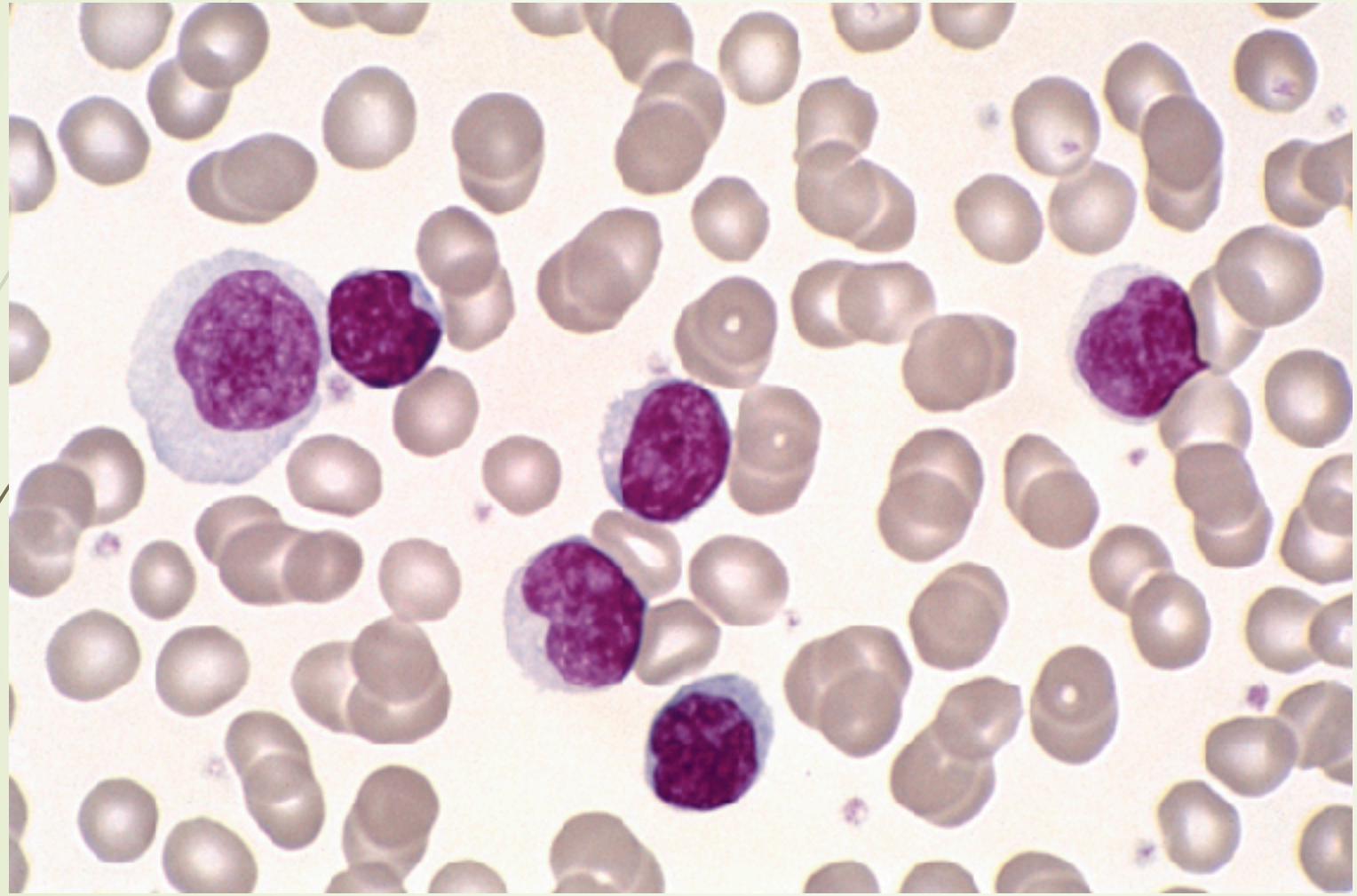
Reactive lymphocytes: *Bordetella pertussis*



Bordetella pertussis

- The persistent lymphocytosis present in blood of a patient with *Bordetella pertussis* is due to a 'lymphocyte promoting factor' produced by the bacterium *Bordetella pertussis*
- This factor inhibits lymphocyte migration from the blood to the lymphoid tissues
- *Bordetella pertussis* (whooping cough) is a highly infectious disease occurring primarily in infants younger than 2 years of age
- Infection results from inhaling droplets contaminated with *Bordetella pertussis*
- The blood film shows a marked increase in the number of T lymphocytes
- The total count ranges from 15 – 50 by 10^9/L with 70 to 90% lymphocytes

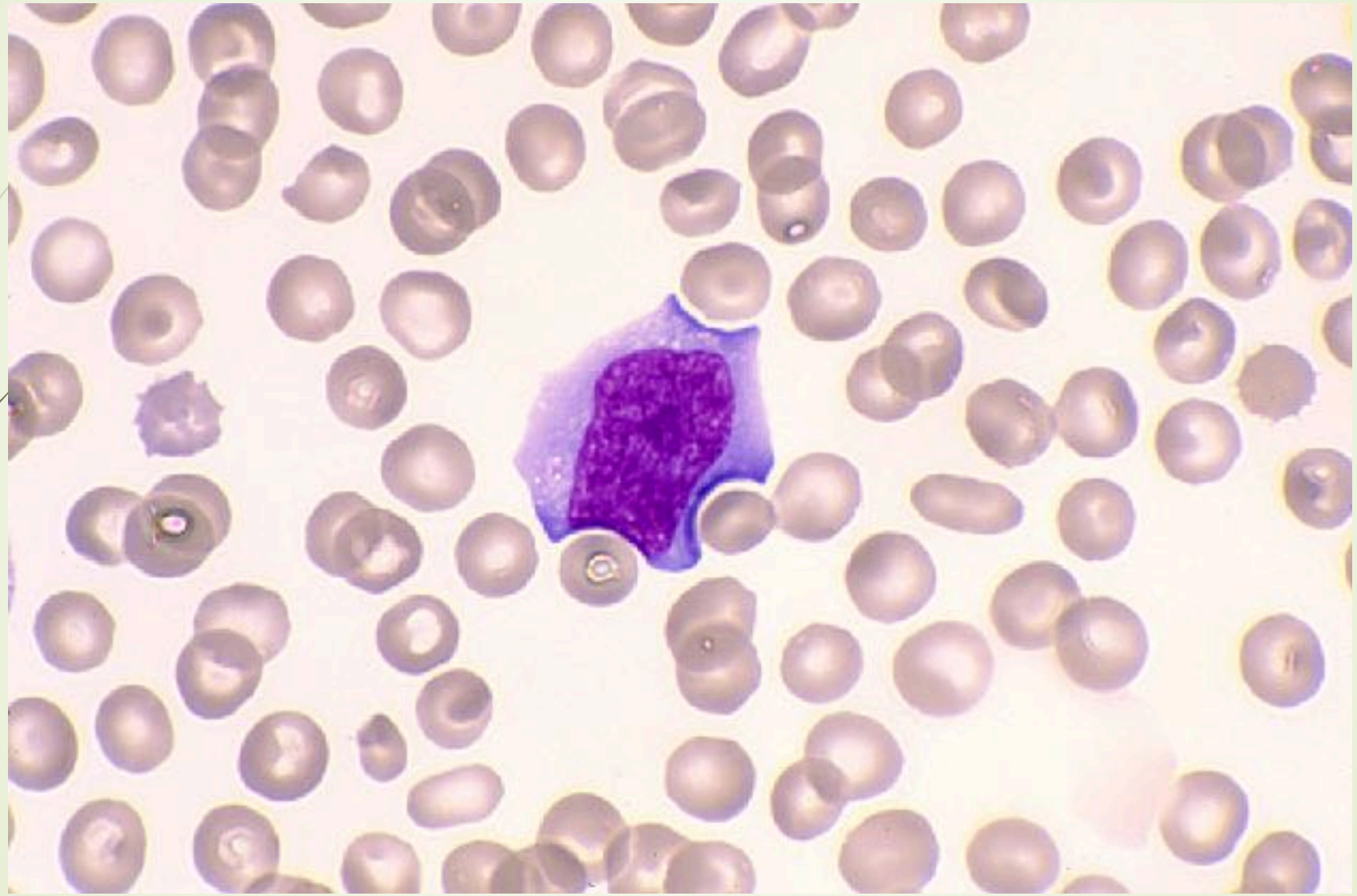
Acute infectious lymphocytosis



Acute infectious lymphocytosis

- Acute infectious lymphocytosis occurs in children between the ages of 1and 14 years with the highest incidence in the first 10 years of life
- It may be associated with a low-grade fever and diarrhoea
- The absolute lymphocyte count is very high; it can reach 50 x 10^9/L
- The majority of lymphocytes are CD4+ T lymphocytes
- The condition resolves in 2-4 weeks without treatment

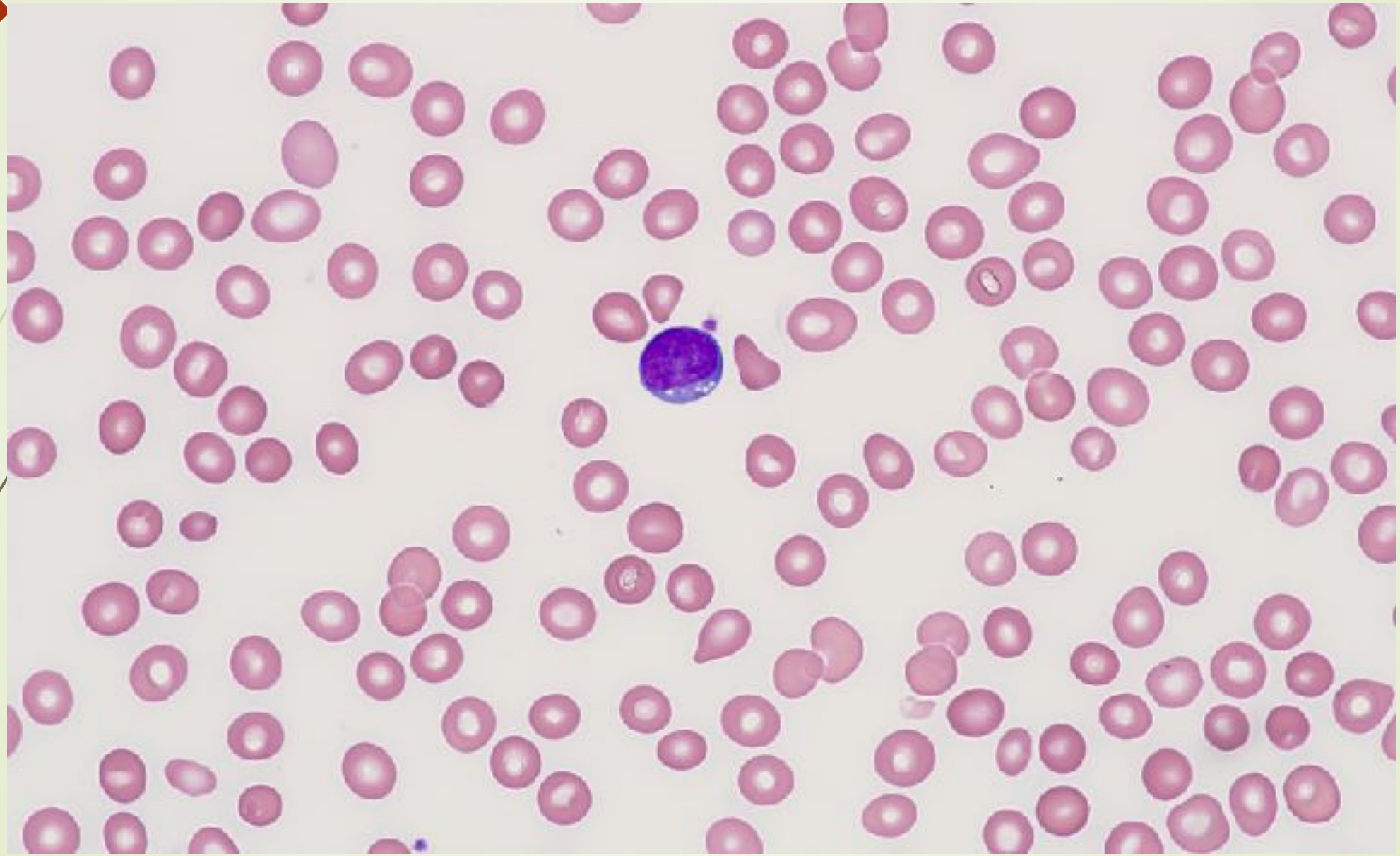
Immune thrombocytopenic purpura (post viral infection)



Immune thrombocytopenic purpura (post viral infection)

- In childhood ITP, males and females are affected with equal frequency
- Often there is a history of a viral illness prior to presentation
- In the majority of children, ITP is an acute self limiting disease, resolving within 6 months whether or not therapy is given
- One of two medical therapeutic options are chosen - corticosteroids or intravenous immunoglobulin (IgG)
- Either will increase the platelet count in the majority of patients and presumably decrease the risk of serious haemorrhage

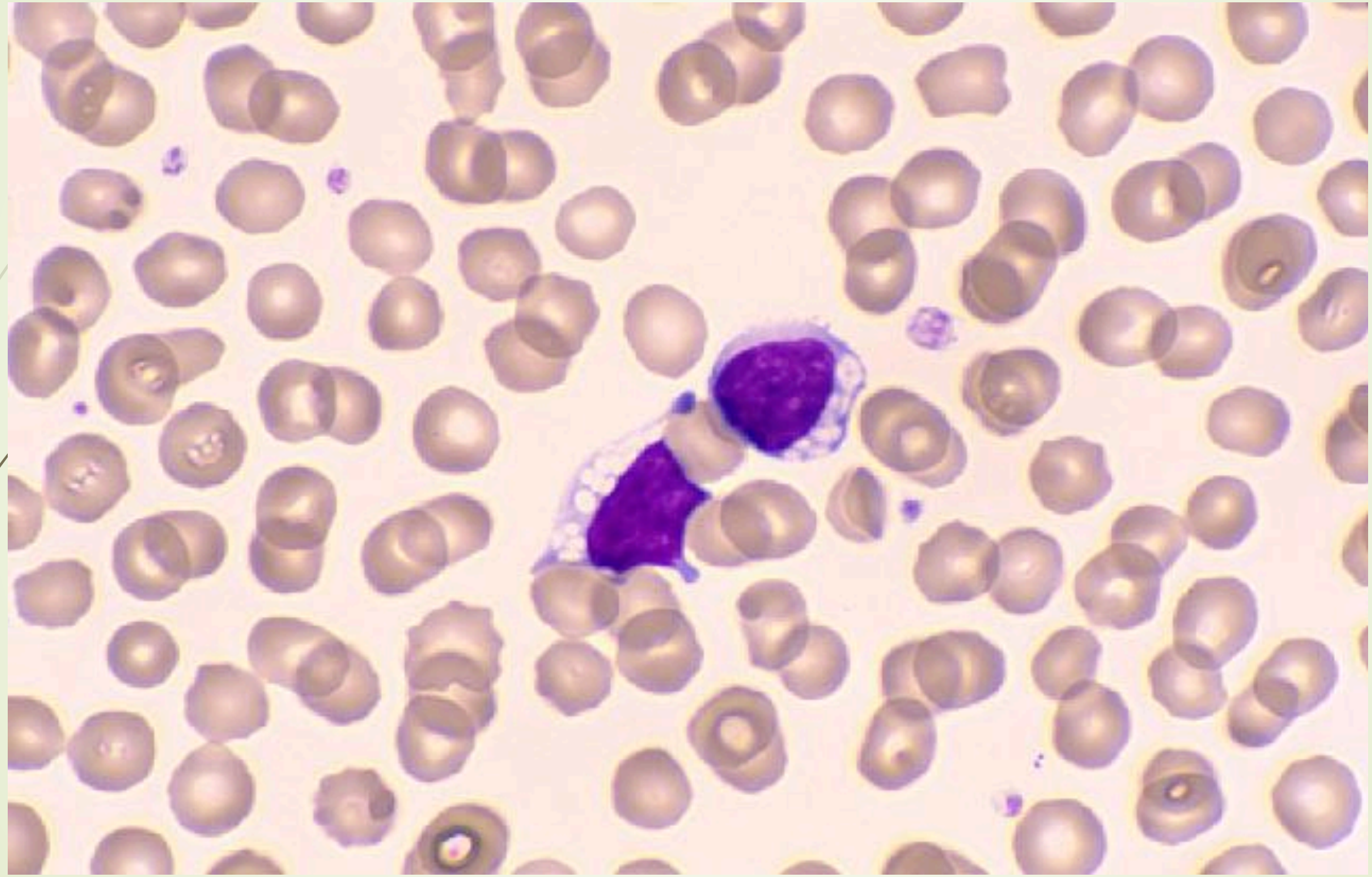
Storage disorders (Wolman Disease)



Wolman disease

- An autosomal-recessive disorder characterised by an abnormality in the lysosomal acid lipase (LIPA) gene resulting in a deficiency of the acid cholesteryl ester hydrolase enzyme
- Deficiency of this enzyme leads to an accumulation of cholesterol and triglycerides within the body organs and tissues
- The blood film is characterised by the presence of vacuolated lymphocytes in the peripheral blood and in the bone marrow by the presence of foamy macrophages
- The vacuoles in both the lymphocytes and bone marrow macrophages stain positively with oil red O stain and Sudan black B

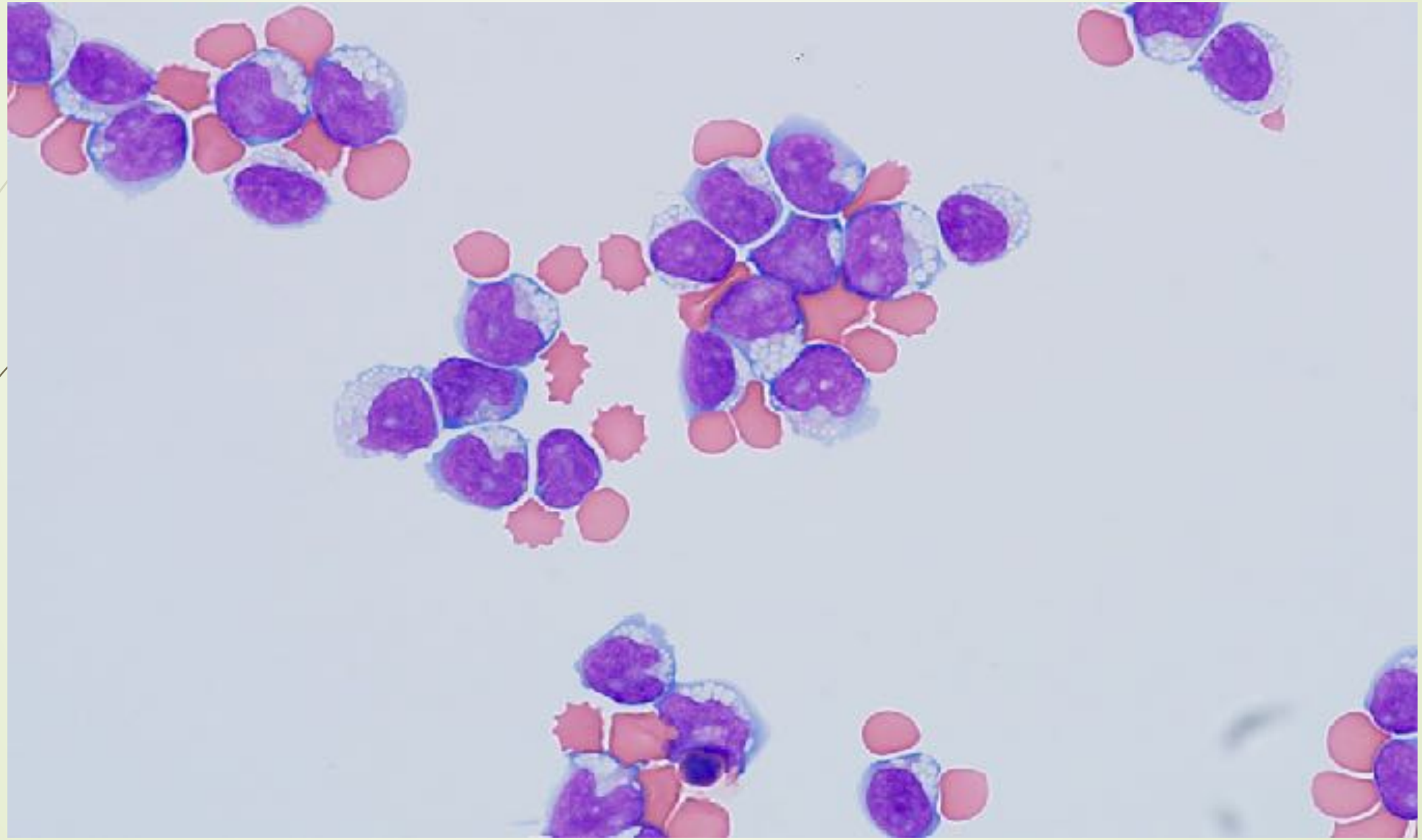
Storage disorders (Sialic acid storage disease)



Sialic acid storage disease

- An autosomal recessive neurodegenerative disorder resulting from a block in sialic acid release from cell lysosomes
- The accumulation of sialic acid in many cells, including lymphocytes, leads to cytoplasmic vacuolation

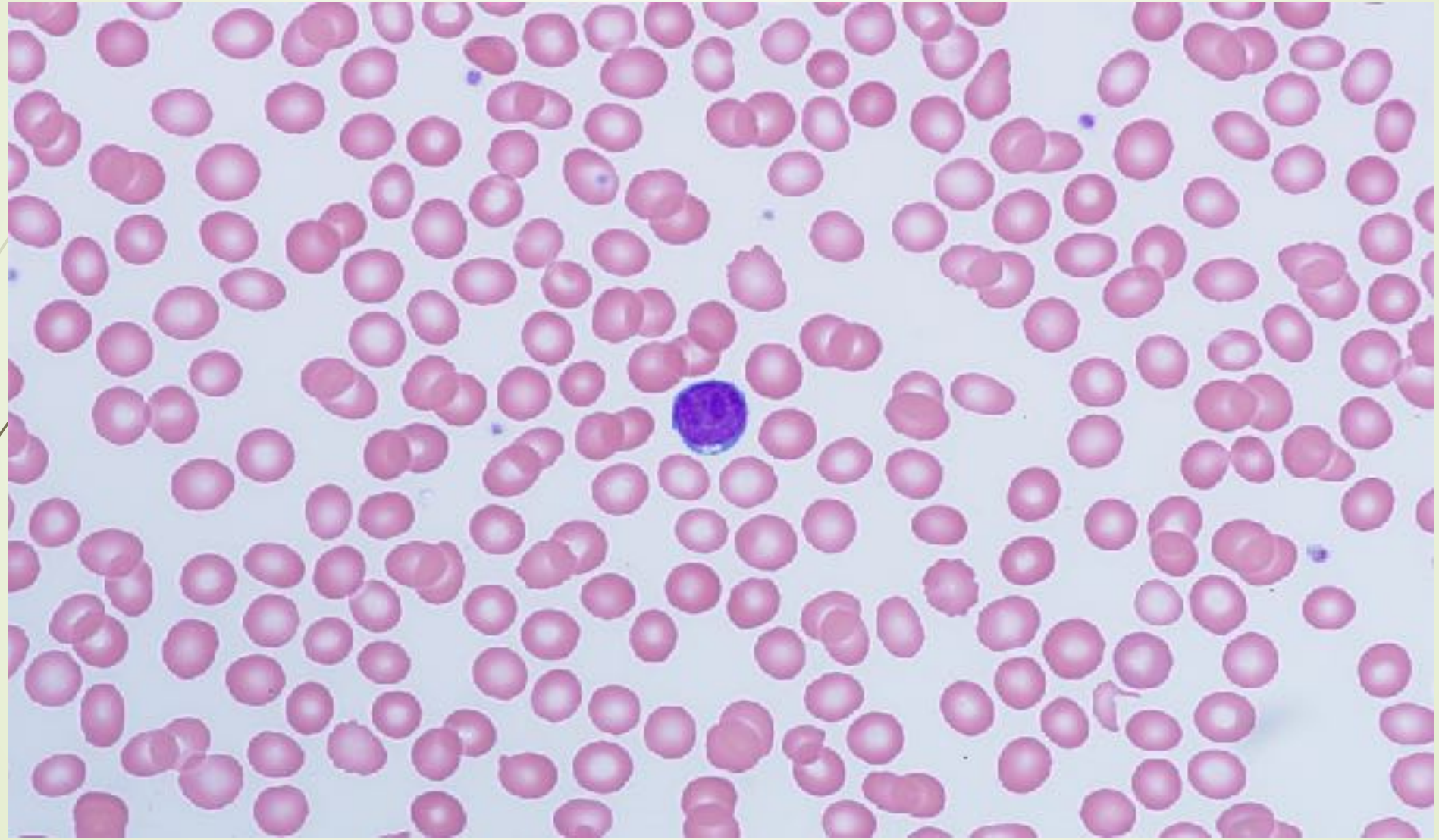
Storage disorders GM1 gangliosidosis



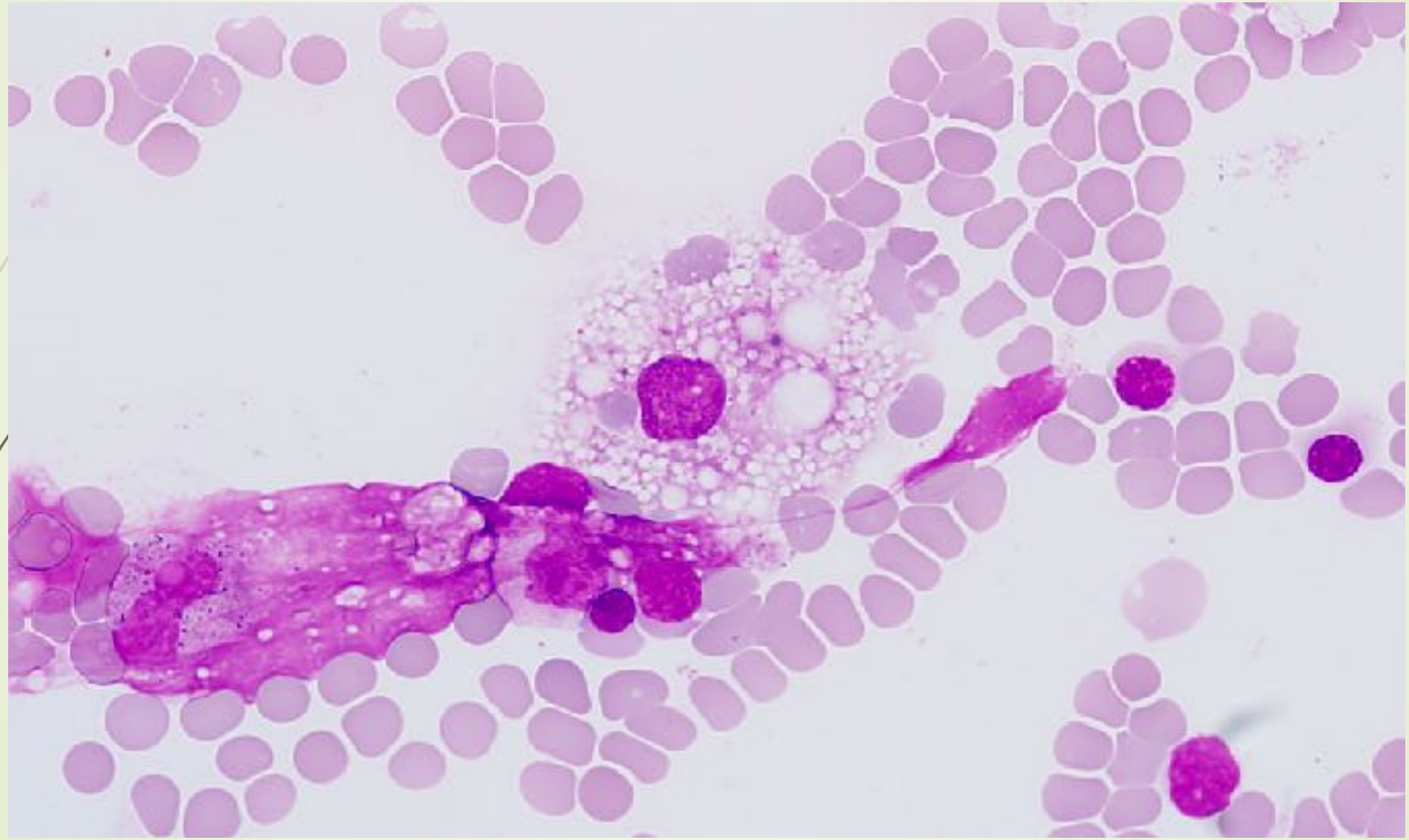
GM1 gangliosidosis

- GM1 gangliosidosis is an autosomal recessive lysosomal storage disease caused by deficiency of acid beta-galactosidase
- It is inherited as an autosomal recessive disorder
- GM1 gangliosidosis progressively destroys nerve cells in the brain and spinal cord
- The signs and symptoms of the most severe form, GM1, usually become apparent by the age of 6 months
- They develop a hepatosplenomegaly
- Infants appear normal until their development slows and muscles used for movement weaken

Niemann-Pick disease - PB



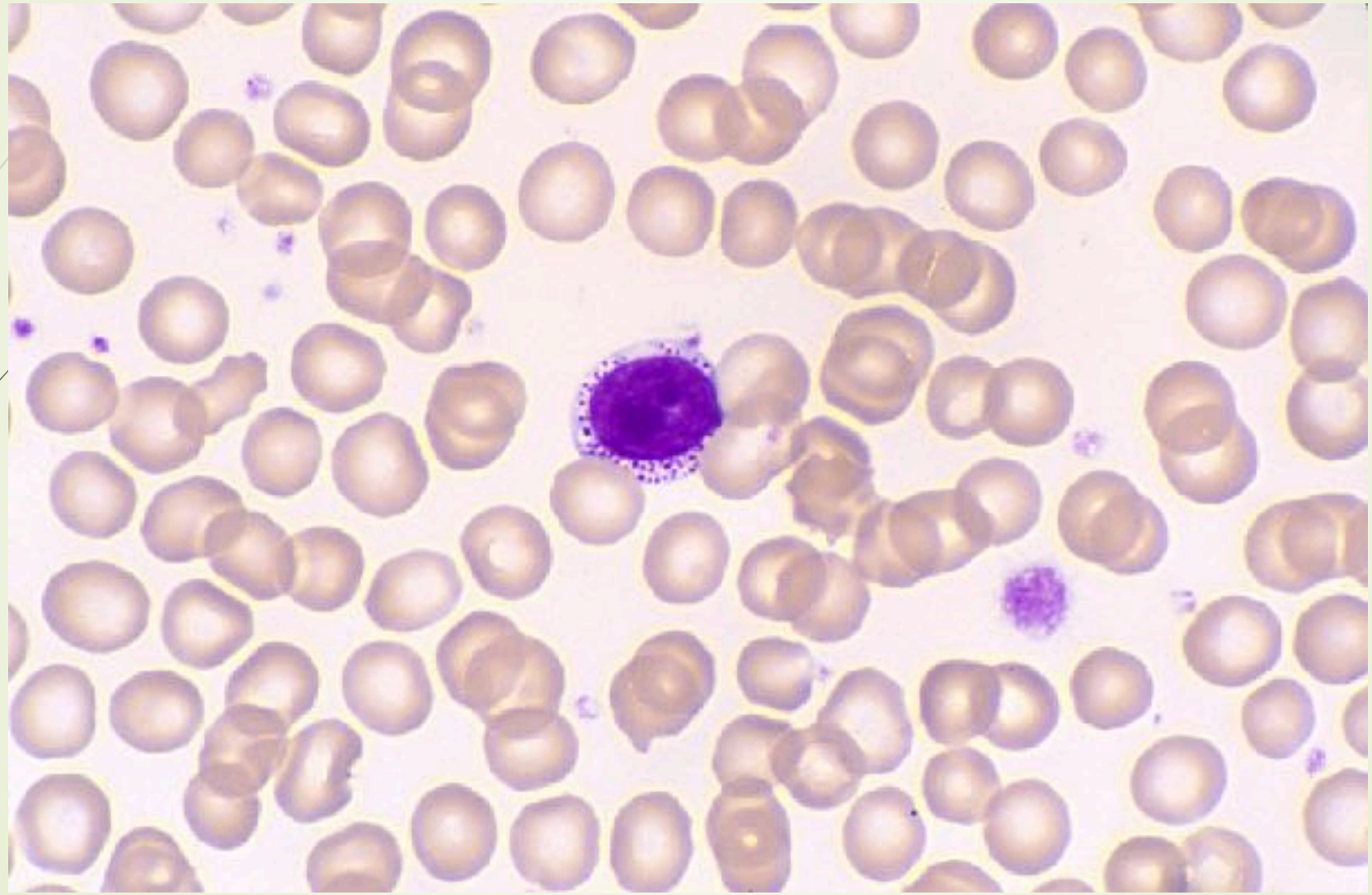
Niemann pick - BM



Niemann-Pick disease

- Characterised by a deficiency of the enzyme sphingomyelinase leading to a build up of sphingomyelin, cholesterol and other cell membrane lipids which accumulate within foam cells (macrophages) throughout the reticuloendothelial system and bone marrow
- On examination of the blood film, the majority of lymphocytes are noted to contain cytoplasmic vacuolation
- These vacuoles represent lipid-filled lysosomes

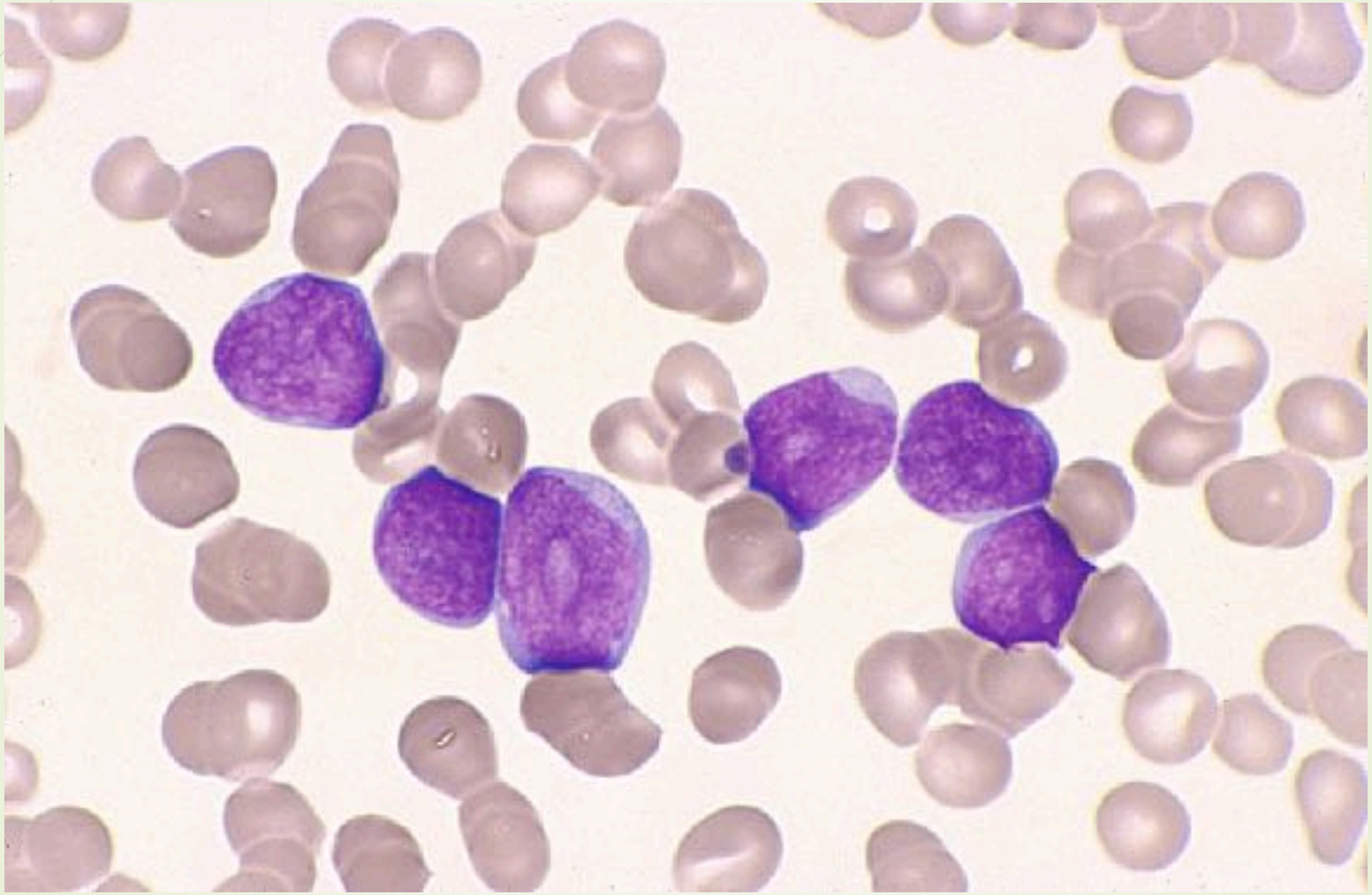
Hurler syndrome



Hurler syndrome

- Hurler syndrome, a mucopolysaccharide storage disorder, characterised by the presence of lymphocytes whose cytoplasm contains clear vacuoles filled with coarse metachromatic granules of mucopolysaccharide
- The lymphocytes have been described by Gasser and are known as Gasser lymphocytes

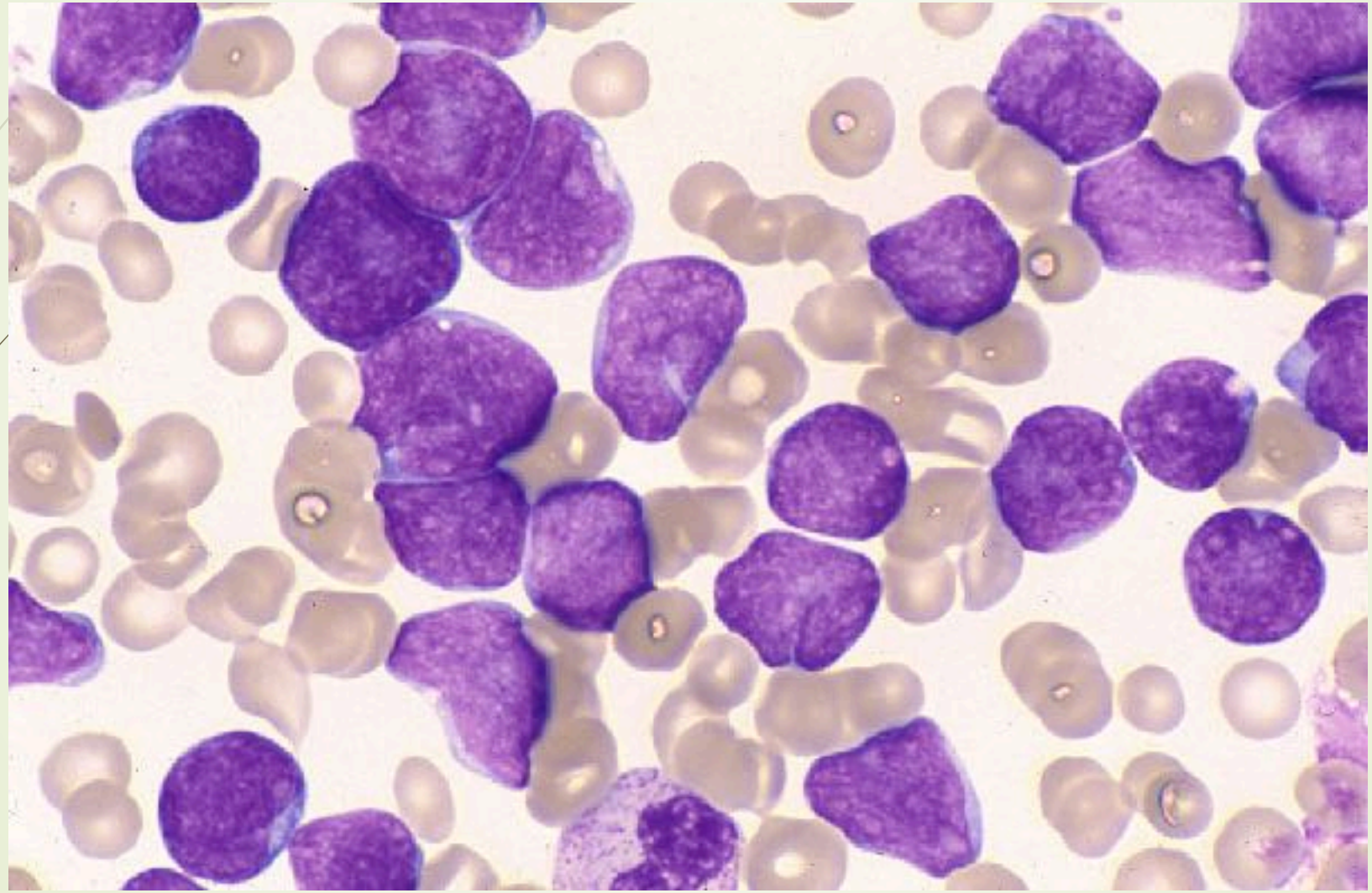
B lymphoblastic leukaemia/lymphoma



Precursor lymphoid neoplasms
B lymphoblastic leukaemia/lymphoma, not otherwise specified (NOS)

- The blasts range from those with a high N/C ratio, fine to clumped chromatin pattern with scanty cytoplasm and inconspicuous nucleoli (FAB L1) to those that are heterogeneous in size and shape, with fine to coarse chromatin pattern, often cleaved, indented and folded. Nucleoli are nearly always present, with variability in size and number. The amount of cytoplasm is variable and often abundant (FAB L2)
- The term 'B-lymphoblastic leukaemia' applies when there is involvement of the BM and PB and the term 'B-lymphoblastic lymphoma' when there is significant nodal or extra nodal involvement. The distinction between the two names is arbitrary

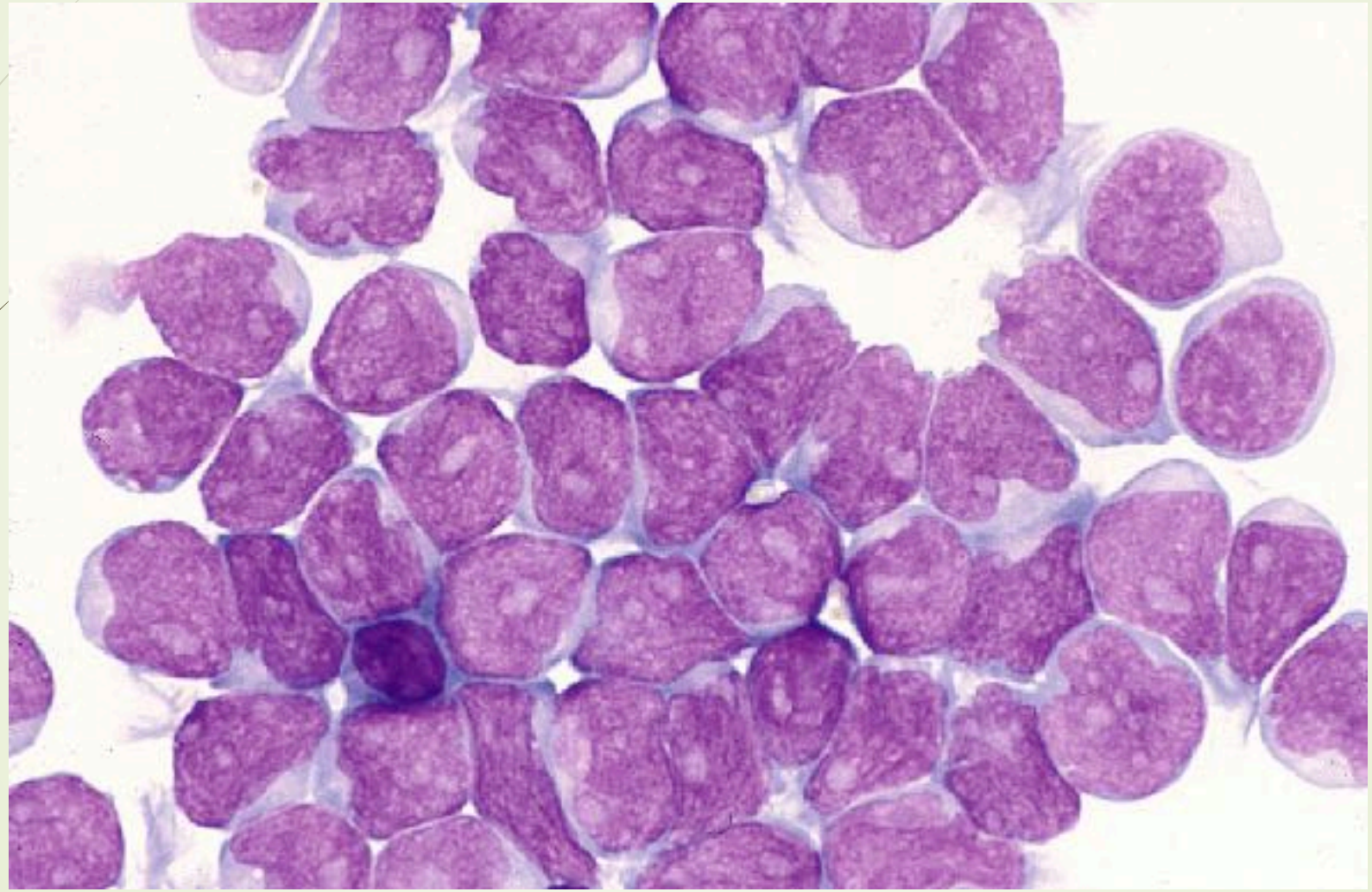
T lymphoblastic leukaemia/lymphoma



Precursor lymphoid neoplasms
T lymphoblastic leukaemia/lymphoma, not otherwise specified (NOS)

- T lymphoblastic leukaemia/lymphoma is a disease of precursor T lymphoblasts. It occurs more commonly in adolescents rather than in young children. The lymphoblasts are small to medium in size, with a high N/C ratio, moderately condensed chromatin pattern and inconspicuous nucleoli
- The nuclei are often folded and cleaved with scanty basophilic cytoplasm
- The term 'T-lymphoblastic leukaemia' applies when there is involvement of the PB and BM and the term 'T-lymphoblastic lymphoma' when there is significant nodal or extra nodal involvement
- The distinction between the two names is arbitrary

T lymphoblastic leukaemia/lymphoma (CSF)



PAEDIATRIC LYMPHOCYTES

- This has been a presentation of some of the most rare and the most commonly occurring disorders involving the lymphocyte lineage in the paediatric patient

- Thank you for your attention

- Gillian Rozenberg