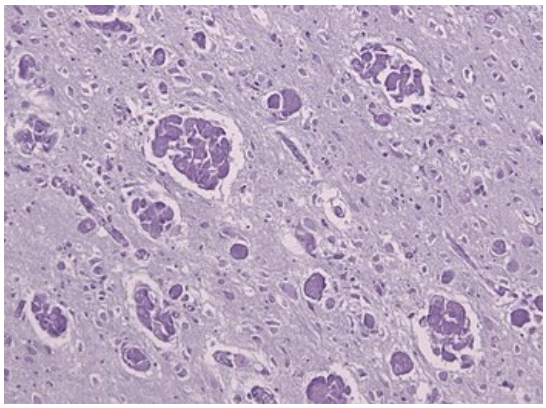
Krabbe disease



Multinucleated macrophages with PAS-positive inclusions, so-called *globoid cells*

Krabbe disease (also Krabbe **globoid leukodystrophy**) is a *leukodystrophy* . It is one of the inherited diseases caused by a congenital defect of lysosomal enzymes. The result is an unrestricted, non-tumor accumulation of mature adipose tissue in places where it does not normally occur. This is a galactocerebroside β-galactosidase deficiency. It affects CNS cells.

Links

related articles

- Leukodystrophy
- Lipidosis
- Hereditary metabolic disorders
- Hereditary small molecule metabolic disorders
- Hereditary metabolic disorders of complex molecules

Source

- - VOKURKA, Martin and Jan HUGO, et al. *Great Medical Dictionary* [online] . 8th edition. Prague: Maxdorf, 2009. 1144 pp. Also available from < http://lekarske.slovniky.cz/ >.

Hereditary metabolic disorders (DMPs)	
In general	DMP of complex molecules • DMP of small molecules • Neonatal screening • Screening of hereditary diseases • Examination methods at DMP
DMP amino acids	Alkaptonuria
Organic aciduria	-
DMP urea cycle	Alcaptonuria • Ornithine transcarbamylase deficiency • Prolidase deficiency • Phenylketonuria • Glutaric aciduria • Hyperphenylalaninemia • Hyperornitinemia • Isovaleric aciduria • Leucinosis • Non-ketotic hyperglycemia • Cystinosis • Tyrosinemia
DMP propionate, biotin and cobalamin	Biotinidase deficiency • Methylmalonic acidemia • Propionic acidemia
DMP purines and pyrimidines	Liver porphyria • Skin porphyria • Mitochondrial neurogastrointestinal encephalomyopathy
DMP sugars	Glycogenoses • Fructosealdolase deficiency • Fructose-1,6-bisphosphatase deficiency • Essential fructosuria • Galactokinase deficiency • Galactose-1-phosphate uridyltransferase deficiency
DMP mitochondria	Phosphoenolcarboxykinase Deficiency • LCHAD Deficiency • MCAD Deficiency • Pyruvate Dehydrogenase Deficiency • Pyruvate Carboxylase Deficiency • SCAD Deficiency • Chronic Progressive External Ophthalmoplegia • Leber's Hereditary Optic Neuropathy • Leigh Syndrome • Maternally Hereditary Diabetes and Deafness • SayLC Syndrome
DMP peroxisomes	Neonatal adenodystrophy • Refsum's disease • Rhizomelic chondrodystrophia punctata • X-linked adrenoleukodystrophy • Zellweger syndrome
DMP of lysosomes	Fabry disease • Gaucher disease • **Krabbe disease** • Danon's disease • Mucolipidosis II • Metachromatic leukodystrophy • Mucopolysaccharidosis III • Niemann-Pick disease • Cystinosis • Tay-Sachs disease
Portal: Pathobiochemistry	