Disorders of glycogen metabolism

Glycogen

Glycogen is a storage polysaccharide (branched glucan - main and secondary chains of α-D-glucose molecules connected by α(1–4) glycosidic bonds, secondary chains are connected to the main chain by α(1–6) bonds. The main stores of glycogen are in the liver and in the muscles. The breakdown of glycogen (glycogenolysis – breakdown into glucose molecules) is stimulated by glucagonem and catecholamines, glucose is released from the hepatocytes into the bloodstream and blood glucose rises ,it cannot be released from the muscles into the circulation and is consumed during muscle activity. It is found in the form granules in the cytoplasm (β-granules, their accumulation creates α-granules).

Glycogen is the only significant carbohydrate that can be observed microscopically (other, soluble carbohydrates are dissolved during the histological processing of the sample). In hematoxylin-eosin glycogen is not stained and its accumulation is manifested in the cells by conspicuously bright (water-clear cytoplasm), otherwise it is demonstrated:

- **Best carmine** - red lumps
- **PAS reaction - red lumps**, PAS reaction with control (A-PAS) is performed , when glycogen is cleaved with diastase in the control sample

Macroscopy

Macroscopically, no specific changes are visible on the organs in whose cells glycogen accumulates during disturbances in its metabolism. Their enlargement may occur (hepatomegaly, cardiomegaly, etc.).

Etiology

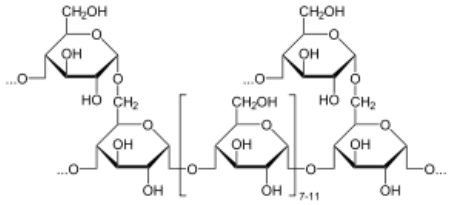
Pathological accumulation of glycogen occurs in the following situations:

1. with dystrophic glycogen infiltration in cells damaged by toxins, viruses, etc. (e.g. accumulation of glycogen in hepatocytes after persistent hepatitis, in damaged keratinocytes, etc.)
2. in diabetes mellitus in the cells of the proximal tubules of the kidneys (Armani's zone at the border of the cortex and medulla) and in the nuclei of hepatocytes
3. in some tumors – e.g. Grawitz's kidney cancer, clear cell lung tumor (sugar tumor)
4. in the projections of astrocytes subpially (cerebral cortex) in the form of spherical bodies (corpora amylacea)
5. in glycogenoses – genetically determined disorders of glycogen metabolism, glycogen accumulates in the cytoplasm of cells in the form of granules, depending on the topic we distinguish several groups of glycogenoses:
    - **hepatic** - accumulation of glycogen in the liver - e.g. von Gierke's disease, Anderson's disease
    - **myopathic** - accumulation of glycogen in the muscles - eg McArdle's disease
    - **others** – involvement of other organs, e.g. Pompe's disease

links

Related Articles

- Glycogenosis
- Dystrophy



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Source

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