

Investigation of the metabolism of porphyrins

- porphyria is classically divided into "hepatic", "erythropoietic" and "erythrohepatic"
- according to the tissue that produces porphyrins.
- liver diseases are acute and chronic

 For more information see *Porphyria*.

Porphyria cutanea tarda

File:Porphyria.png
Heme biosynthesis with marked enzyme deficiencies

- chronic hepatic porphyria, the most common
- uroporphyrinogen decarboxylase disorder
- patients usually come with skin problems (solar localization)
- without treatment, however, they are most at risk of cirrhosis
- acute attacks do not occur
- the precise distinction of cutaneous porphyrias can only be made in the laboratory
- in untreated patients, there is an increased level of porphyrins in the urine, ALA and PBG are normal
- uroporphyrin predominates in urine
- chromatography is done - typical peaks
- fluorescence is also used for determination - *Soret band*

Acute hepatic porphyria and lead poisoning

- most often ``acute intermittent porphyria (*AIM*), then *p. variegata*, *hereditary coproporphyria*
- AD, common - in attacks - form abdominal, neurological, psychiatric
- they are often operated on for an acute abdominal event
- it is enough to examine the values of **ALA, PBG** " *and total porphyrins' in urine*
- **Pb** blocks *ALAd hydratase* - **ALA** accumulates, PBG is normal
- main examination - "waste of total porphyrins in 24 h", "ALA" and "PBG"

Links

Related Articles

- Porphyria

References

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