

## Acute Intermittent Porphyria

- Enzyme defect- Uroporphyrinogen I synthase / HMB synthase/ PBG deaminase.
- Reaction blocked: step number 3 of heme synthesis pathway.
- For conversion of Porphobilinogen to Uroporphyrinogen I, Uroporphyrinogen I synthase enzyme is required.
- PBG and ALA levels accumulated in Blood, tissues and urine.
- Urinaryporphobilinogen on photo oxidation convert to Porphobilin