→ develop when > 20 y.

__/__/____

HEMOCHROMATOSIS:

defined as a group of genetic diseases that predispose to iron overload, potentially leading to fibrosis and organ failure.

PATHOGENESIS:

1° = - Mutation of gene encoding HFE, HJV, TfR2
↓
lead to Absence of Hepcidin
(Hepcidin is needed for iron Absorption)

2° = • Parenteral iron overload
• Ineffective erythropoiesis.
• Chronic liver disease.
• β thalassemia.

Mechanism of tissue damage:

1) Lipid peroxidation → Free Radicals formed.
2) stimulate collagen formation → Activate hepatic stellate cells.
3) DNA damage by Reactive O_2 species.

MORPHOLOGY:

Gross → Appear grossly normal or slightly decrease in colour → chocolate brown color.
cirrhosis → micronodular → macronodular

__/__/____

Microscopy: Iron deposit as finely granular golden yellow hemosiderin pig in cytoplasm.

Stain → Prussian blue stain.

WILSON DISEASE.

Hepatolenticular degeneration is an Autosomal Recessive disorder of copper metabolism.

Risk factors

→ impaired excretion of copper into bile And a failure to incorporate copper into ceruloplasmin

↓

major Method of elimination of copper from the body by ATP7B gene excreted as copper in bile

↓

loss of mutation of ATP7B gene

lead to → ↓ in circulating level of ceruloplasmin

Accumulation of copper within the liver

__/__/__

MORPHOLOGY:

Liver → . Steatosis ⟶ Lead to macronodular cirrhosis.

MICROSCOPY:

→ Fatty change
→ Acute liver failure
→ chronic hepatitis.
→ Cirrhosis
→ Massive liver necrosis.

Eye lesions – Kayser - Fleischer Ring due to green to brown deposits of copper in descement membrane

Biochemical findings

→ ~~↑ se~~ ↓ed serum ceruloplasmin
→ ↑↑ copper content
→ ↑↑ urinary excretion