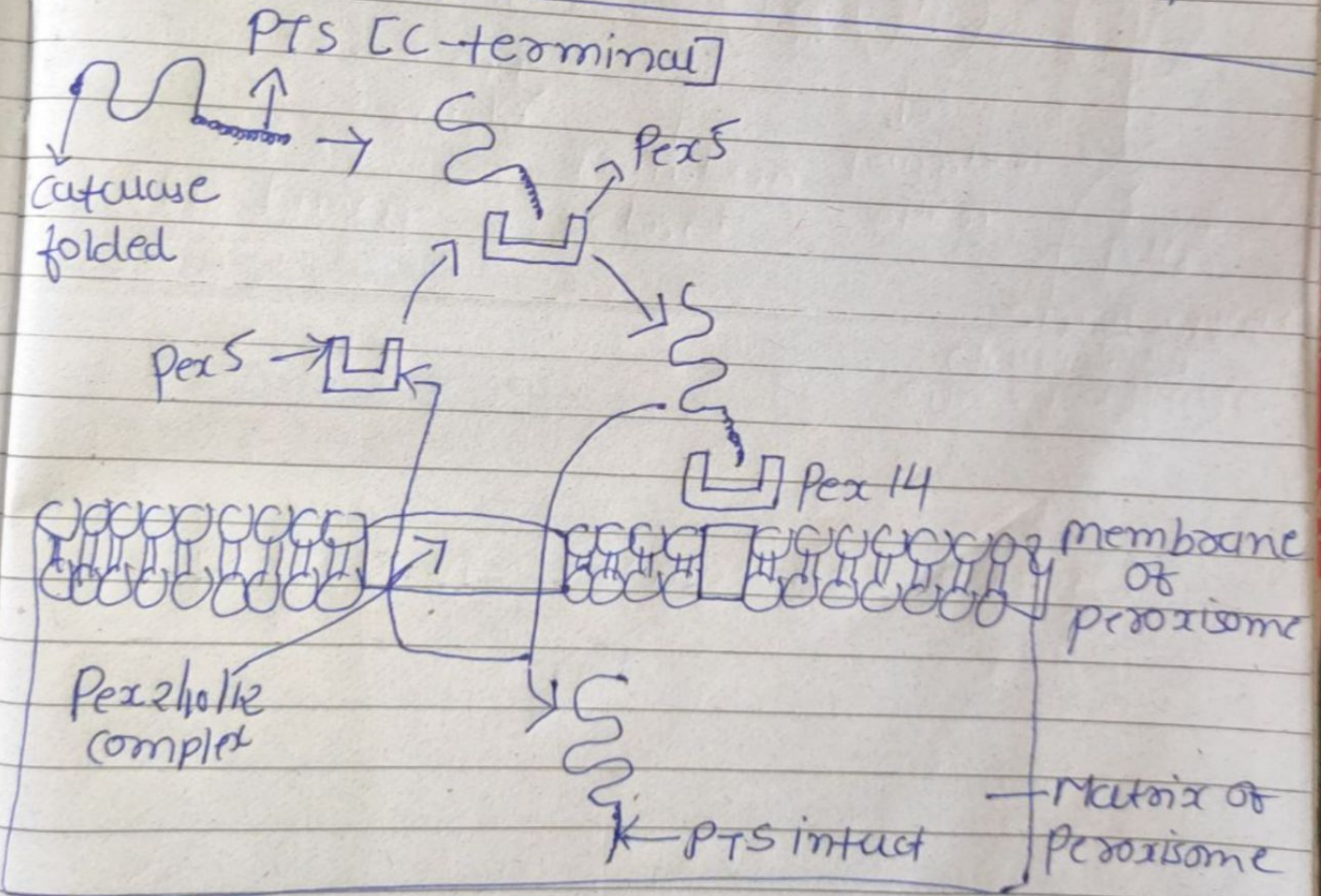


* Peroxisomal protein sorting & Zellweger Syndrome

→ Peroxisome - Involved in metabolism of Hydrogen peroxide [H₂O₂], fatty acids, Amino acids, purine, Cholesterol, bile acid, plasmalogens

→ Enzyme of peroxisome: Catalase

- It's protein synthesized on cytosolic polyribosome
- Fold prior to import
- Require ATP



PTS - peroxisomal matrix targeting sequence

Miscellaneous

→ Zellweger Syndrome :-

- Inherited disorder - autosomal recessive
- complete absence of peroxisomes in cells



due to defect in protein required for biogenesis of peroxisomes
[mutation in PEX family of gene]

- Accumulation of long chain fatty acid & Branched chain fatty acid
- Marked reduction in plasmalogen
- profound neurologic impairment
- Dye within 1 year
- most severe

→ Infantile Refsum disease - less severe