Topic: oxidation of fats

Class: B.Sc Part –III (Hons.)

Paper- V

Group – A

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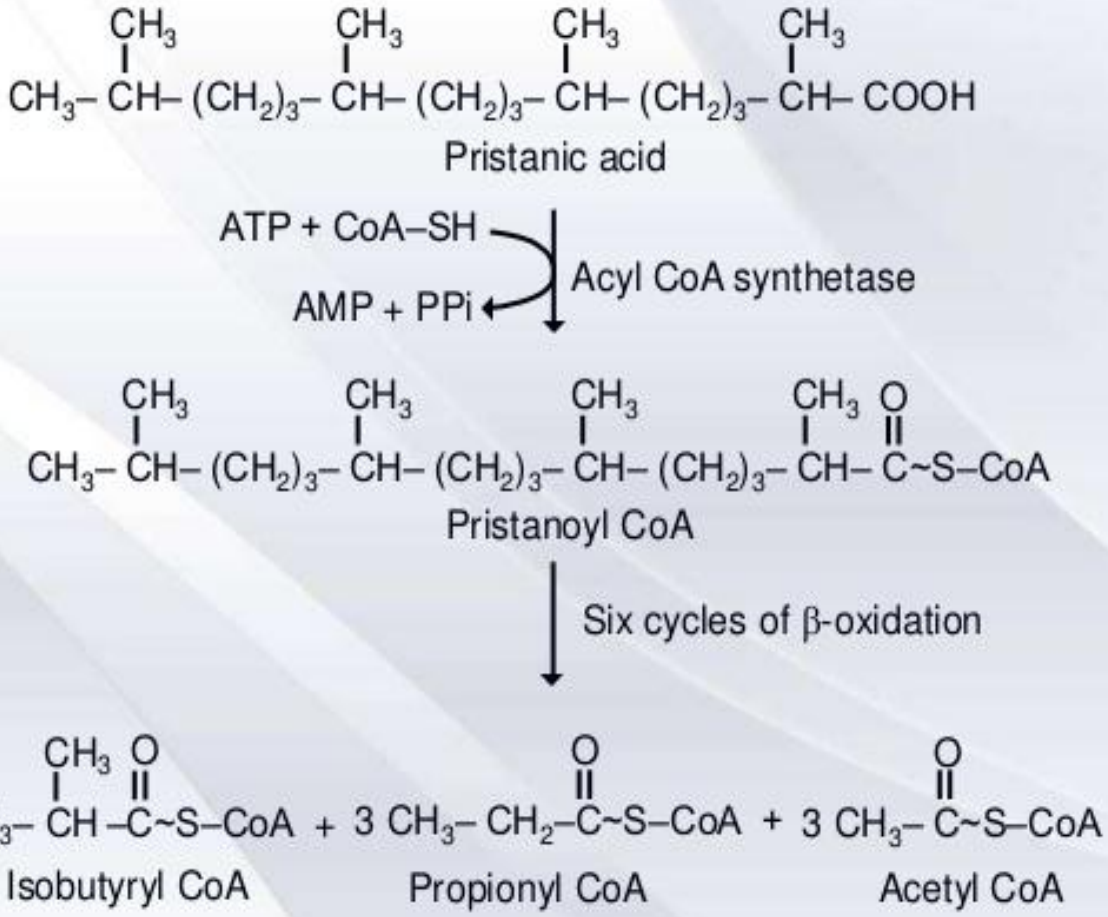
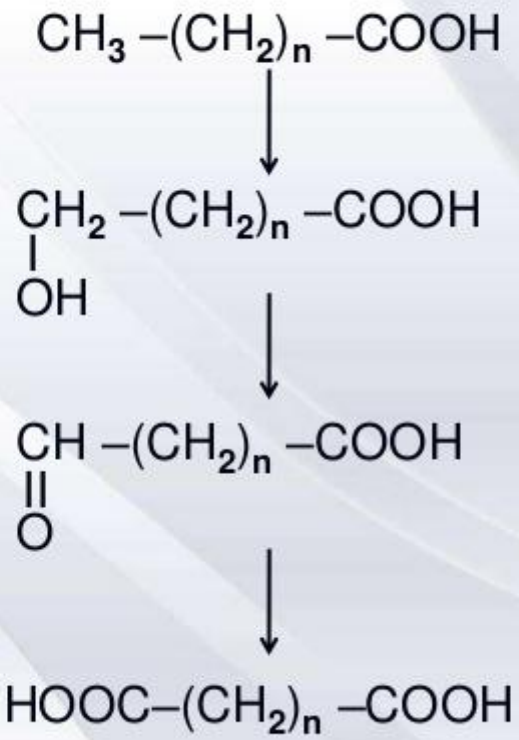
- Pristanic acid is activated to pristanoyl CoA, which then undergoes beta-oxidation
- Six cycles of b-oxidation produce iso- butyryl CoA and three molecules each of acetyl CoA and propionyl CoA


CH3 CH3 CH3 CH3
CH3– CH– (CH2)3– CH– (CH2)3– CH– (CH2)3– CH– COOH
Pristanic acid
ATP + CoA–SH
AMP + PPi
Acyl CoA synthetase
CH3 CH3 CH3 CH3 O
CH3– CH– (CH2)3– CH– (CH2)3– CH– (CH2)3– CH– C~S–CoA
Pristanoyl CoA
Six cycles of β-oxidation
CH3 O O O
CH3– CH –C~S–CoA + 3 CH3– CH2–C~S–CoA + 3 CH3– C~S–CoA
Isobutyryl CoA
Propionyl CoA
Acetyl CoA

- An inherited defect in a-oxidation results in Refsum's disease This is most commonly due to deficiency of phytanoyl CoA hydroxylase Some times, it is due to deficiency of peroxin-7, a peroxisomal receptor Peroxisomal enzymes fail to reach peroxisomes in peroxin-7 deficiency
- Large amounts of phytanic acid accumulate in brain in Refsum's disease This causes neurological damage, cerebellar degeneration, and peripheral neuropathy Patients are advised to take a phytanic acid-restricted diet
- This is another minor pathway for oxidation of fatty acids It becomes important when b-oxidation is defective It is located in endoplasmic reticulum of liver and kidney cells w-Oxidation
- The first reaction is introduction of a hydroxyl group onto the w-carbon The reaction is catalysed by the microsomal hydroxylase system Alcohol dehydrogenase then oxidizes the hydroxyl group to an aldehyde group Aldehyde dehydrogenase oxidizes the aldehyde group to a carboxyl group


CH3 –(CH2)n –COOH
CH2 –(CH2)n –COOH
OH
CH –(CH2)n –COOH
O
HOOC–(CH2)n –COOH

- The fatty acid now has a carboxyl group at each end
- The dicarboxylic acid is activated and enters the mitochondria
- beta-Oxidation starts from both the ends
- Two carbon atoms are removed in one cycle from both the ends
- This continues until a 6-carbon or 8-carbon dicarboxylic acid is left
- The 6-carbon product is adipic acid, and the 8-carbon product is suberic acid
- These are excreted in urine