

# MEVALONATE KINASE DEFICIENCY

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- most cholesterol in the body is derived from its biosynthesis from saturated fatty acids
- cholesterol is essential as a component of membranes and for the synthesis of bile salts for fat digestion and of steroid hormones
- mevalonic acid
  - ✓ intermediate in the synthesis of cholesterol
  - ✓ formed by the enzyme HMG-CoA reductase
  - ✓ synthesis controlled, in part, by the amount of cholesterol in the cell

