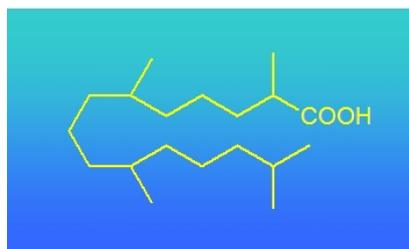


Pristanic acid



The occurrence of 2,6,10,14-tetramethylpentadecanoic acid in butterfat was reported by Hansen and Morrison in 1964 (1). Because of its structural similarity to the hydrocarbon pristane (2,6,10,14-tetramethylpentadecane), the acid was later given the trivial name "pristanic acid". Pristanic acid and its precursor, phytanic acid (3,7,11,15-tetramethylhexadecanoic acid), are also present in human and animal tissues including blood plasma (2). Phytanic acid is of dietary origin, the ultimate source being phytol (3,7,11,15-tetramethyl-2-hexadecenol), a common diterpene alcohol of plant origin.

Conversion of phytanic acid to pristanic acid in animal tissues takes place by peroxisomal α -oxidation (3). This stepwise process involves a) formation of phytanoyl-CoA, b) hydroxylation of the CoA ester into 2-hydroxyphytanoyl-CoA, c) decarboxylation of the latter into pristanal (2,6,10,14-tetramethylpentadecanal), and d) NAD⁺-dependent dehydrogenation of the aldehyde into pristanic acid.

The Norwegian neurologist Sigvald Refsum in 1945 described a hereditary disease presenting symptoms due to neural damage of cerebellum, sensory organs and peripheral nerves, as well as a skin abnormality (ichthyosis) (4; reviewed in ref. 5). The underlying biochemical abnormality was discovered in 1963 by Klenk and Kahlke, who found that blood and tissues from a patient with Refsum's disease contained high concentrations of phytanic acid (6). As shown later, accumulation of phytanic acid in Refsum's disease is due to deficient phytanoyl-CoA 2-hydroxylase activity required for degradation of phytanic acid and other 3-methyl-substituted fatty acids (reviewed in ref. 7).

Pristanic acid has been reported to serve as a ligand for the peroxisome proliferator-activated receptor α (PPAR α), and possibly reduced receptor activation contribute to the symptoms of Refsum's disease (8).

Pristanic acid (A-1500) supplied by Lipidox is chemically synthesized from phytol. Also available are phytanic acid (A-1600) and 2-hydroxyphytanic acid (O-1642).

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