**Table III. Peroxisome enzyme functions**

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| Enzyme pathways | Features | Relevance to disease |
| Straight, very long chain (≥C22) fatty acid β-oxidation | Chain shortening of very long chain fatty acids (VLCFA), synthesis of docosahexaenoic acid (DHA)- the last step in DHA synthesis is a β-oxidation step inside the peroxisome). | Tissue accumulation of VLCFA causes brain, nerve and adrenal damage. Deficiency of docosahexaenoic acid affects brain function and vision. |
| Methyl-branched chain fatty acid β-oxidation | Chain shortening of pristanic, and di- and tri-hydroxycholestanoic acid utilizes a different oxidase than that used in straight chain shortening. | Accumulation of pristanic acid affects brain. Increased bile acid intermediates, di- and tri-hydroxycholestanoic acid, cause liver toxicity. |
| Dicarboxylic fatty acid β-oxidation | Chain shortening of dicarboxylic acids utilizes a different dehydrogenase/hydratase than that used in straight chain shortening. | Unknown |
| Fatty acid α-oxidation | Degradation of methyl-branched phytanic acid requires an additional α-oxidation step before entering the β-oxidation pathway as pristanic acid. | Tissue accumulation of phytanic acid causes retinal degeneration, cerebellar ataxia, peripheral neuropathy. |
| Ether phospholipid (plasmalogen) biosynthesis | Crucial for the formation of plasmalogens, a specialized class of membrane phospholipids. | Deficiency causes problems in bone development, growth, psychomotor retardation and cataracts. |
| Glyoxylate detoxification | Prevents the conversion of glyoxylate into the toxic metabolite, oxalate. | Accumulation of oxalate results in calcium oxalate renal stones. |
| Pipecolic acid oxidation | Major pathway for lysine degradation in the brain. | Pipecolic acid is an important peroxisome biomarker, that accumulates in ZSD and also in B12-dependent seizures. |
| Catalase activity | Required for catabolism of hydrogen peroxide, produced in peroxisomes as a by product of β-oxidation reactions. | Increased oxidant damage |