ARYLSULFATASE A-DEFICIENT MICE

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HINTERGRUND

Metachromatic leukodystrophy (MLD) is a lysosomal storage disorder caused by the deficiency of arylsulfatase A (ASA). This results in accumulation of sulfated glycosphingolipids, mainly 3-O-sulfogalactosylceramide (sulfatide), in the nervous system and various other organs. In patients, lipid storage causes a progressive loss of myelin leading to various neurological symptoms. The sulfatide storage in ASA-deficient mice is comparable to humans, but the mice do not mimic the myelin pathology.

LÖSUNG

Therefore, in addition to the sole transgenic ASA-deficient (tg/ASA(-/-)) knock-out mice, a second model was generated overexpressing the sulfatide-synthesizing enzyme galactose-3-O-sulfotransferase.

These mice displayed a significant increase in sulfatide storage in the brain and peripheral nerves. Mice older than one year developed severe neurological symptoms. Nerve conduction velocity was significantly reduced due to hypomyelinated and demyelinated axons of the nerves.

Thus, increasing sulfatide storage in ASA-deficient mice leads to neurological symptoms and morphological alterations that are reminiscent of human MLD.

VORTEILE

- Mouse models with sulfatide storage, neuropathy, neurological symptoms
- Valuable model for metachromatic leukodystrophy
- Matching models of symptomatic and asymptomatic mice
SERVICE

On behalf of the University Bonn, PROvendis offers access to the mouse models under a Material License Agreement. The two models are available in combination or separately.

PUBLIKATIONEN & VERWEISE
