

**SPEAKER:** Alpha-galactosidase deficiency that occurs in Fabry disease leads to accumulation of Gb3. The Gb3 then goes on to deposit within the actual Schwann cells of peripheral nerves and therefore impairs myelination. It also can be found in the vascular endothelium for the vessels that are supplying the nerve fibers.

It is thought that due to the accumulation of this substrate, it leads to a vaso spasm of the nerve fiber bundle and therefore leading to pain. It also leads to impaired myelination. It affects both myelinated unmyelinated fibers for this reason. However, in particular, it does cause a small fiber neuropathy with a predisposition to the A-delta fibers.