

## D

According to the U.S National Library of Medicine and the Medline Encyclopedia, the white matter of the brain speeds up the electrical nerve impulses along nerve fibers called axons. White matter is made up of axons and neurons that are surrounded by a protective covering of gray matter, which is largely comprised of myelin. Myelin is a protective layer enclosing nerve fibers and the spinal cord and protecting them from injury. Myelin is made up of proteins and fats and is what gives the white matter its identifying color.<sup>1</sup> Myelin plays a critical role in the proper cognitive function and health of the brain. However, there are many diseases and disorders that affect the growth and survival of myelin in certain individuals. There are two main branches of these diseases: inherited genetic abnormalities and autoimmune disorders.

Leukodystrophies are an example of inherited genetic disorders. The term Leukodystrophy refers to over fifty hereditary neurological disorders that affect the white matter of the brain and the protective sheath of myelin around the nerves. Currently, most leukodystrophies cannot be cured, but some can be clinically managed. As leukodystrophies cause abnormal white matter growth, they can affect neurological function in many different parts of the brain. For example, a leukodystrophy can cause problems with balance, sight, hearing, cognition and thinking, swallowing, and many other important processes.<sup>2</sup> Another inherited disorder that can affect myelin function is Lysosomal Storage Disorder (LSD). LSD is an inherited metabolic disease that is characterized by an excessive build-up of toxic substances in the cells resulting from various enzyme deficiencies. These toxic substances prevent the cell's

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<sup>1</sup> <https://medlineplus.gov/ency/article/002344.htm>

<sup>2</sup> <https://my.clevelandclinic.org/health/articles/6034-leukodystrophy>

lysosomes from functioning as normal. There are many distinct LSDs that are characterized by the enzyme that is in deficit, and they can affect the skeleton, skin, heart, and central nervous system, among other body organs and organ systems. Signs and symptoms of LSDs vary based on which enzyme(s) are affected.<sup>3</sup>

Krabbe disease is also an inherited disease process that is both a Leukodystrophy and an LSD that affects the myelin in the body. It is also referred to as Globoid Cell Leukodystrophy and is a genetic deficiency of the enzyme Galactosylceramidase (GALC).<sup>4</sup> Krabbe disease affects the central and peripheral nervous systems, resulting in a loss of control of the body's voluntary and involuntary movements and functions. Demyelination is the gradual breakdown of the protective myelin sheath surrounding nerve fibers. The beginning of the demyelination process marks the onset of Krabbe disease and results in various disease symptoms like loss of previously attained milestones in early life development. Examples of this regression in maturity are unexplainable crying or irritability, difficulty feeding, and decreased muscle control. The onset can occur from infancy into adulthood, but is most commonly seen in infants and young children. Early Infantile Krabbe Disease (EIKD) and Later Onset Infantile Krabbe Disease are most commonly encountered.

In addition to inherited genetic disorders, myelin function is also sometimes impaired by autoimmune disorders. In contrast to Leukodystrophies and LSDs, autoimmune conditions are disorders in which the body's immune system attacks existing tissues, rather than being affected

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<sup>3</sup> <https://rarediseases.org/rare-diseases/lysosomal-storage-disorders/>

<sup>4</sup> <https://www.huntershope.org/family-care/leukodystrophies/krabbe-disease/>

by a deficit in the production of said tissues. Two examples of autoimmune disorders which affect the body's myelin are Multiple Sclerosis (MS) and Neuromyelitis Optica (NMO).

Multiple Sclerosis (MS) is a disorder in which the immune system attacks the nervous system's protective myelin covering which disrupts electrical impulse communication within the brain, and between the brain and the body. It can result in permanent debilitating symptoms like ongoing paresthesia, diplopia, poor coordination, and tremors among many other things. There is currently no cure for MS despite ongoing research. Neuromyelitis Optica, also known as Devic's disease, is a disorder similar to MS, but it is more concentrated in the optic nerves. NMO is often misdiagnosed as MS because of the clinical similarities, despite being a distinctly different disease. There are many possible symptoms, a few of which are blindness, confusion, muscle spasms, and paralysis.

## References:

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