VETERINARY TECHNICAL DATASHEET

X-Linked Hereditary Nephropathy, (XLHN); mutation originally found in Samoyed

Mutation Found In :Samoyed

Disorder Type

- Urinary

Disease Severity

- Severe

Background

X-linked hereditary nephropathy is a kidney disorder observed in Samoyeds. XLHN affects the glomeruli in the renal corpuscle and is caused by defective type IV collagen in the basal lamina which causes proteinuria and results in juvenile-onset renal failure. It shows an X-linked mode of inheritance, as indicated in the name, and so the severe form of the disease is encountered only in males. In some cases, female carriers can develop a renal disease with milder symptoms.

Key Signs

- Excessive drinking and frequent urination
- Proteinuria
- Diarrhea
- Vomiting
- Weight loss
- Reduced growth
- Renal failure

Clinical Description

The main clinical sign of XLHN is proteinuria, which occurs around the age of 3 to 6 months. The disease leads to renal failure and eventually to death of affected males by the age of 9 to 15 months. Clinical signs of renal failure include excessive drinking, and frequent urination. Other possible signs are reduced growth, vomiting, diarrhea, and weight loss. Female carriers might exhibit proteinuria as a sign of mild renal disease, but the condition usually does not lead to renal failure before the age of five.

Mode of Inheritance

- X-linked

Gene Name

- COL4A5

Next Steps

Affected male puppies are usually euthanized on welfare grounds because of the severity of the condition. In carrier females, the condition can be partly managed through diet therapy. Dietary options and the importance of diet management should be emphasized to clients.

References

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