VETERINARY TECHNICAL DATASHEET

Protein Losing Nephropathy, (PLN); NPHS1 gene variant

Mutation Found In :Airedale Terrier, Soft Coated Wheaten Terrier

Disorder Type

- Urinary

Disease Severity

- Moderate

Background

Protein losing nephropathy (PLN) is a renal disorder in which affected dogs lose protein through their kidneys, resulting in protein excretion in their urine. Protein losing nephropathy has a complex background and several different genes and environmental factors can contribute to the onset of symptoms. Two risk alleles associated with PLN have been identified in Soft Coated Wheaton Terriers, NPHS1 and KIRREL2. These were also found in a single affected Airedale Terrier. Dogs homozygous for both risk alleles have an increased risk of developing protein losing nephropathy.

Key Signs

- Proteinuria
- Hypoalbuminemia
- Hypercholesterolemia
- Polyuria
- Polydipsia
- Anorexia
- Vomiting
- Diarrhea

Clinical Description

Protein losing nephropathy is characterized by high levels of protein in the urine. Clinical signs include weight loss, fatigue, vomiting, and diarrhea. Increased drinking and urination are also typically observed. Accumulation of fluid in the abdominal cavity (ascites) and chest cavity (pleural effusion), high blood pressure, and high cholesterol can be associated with the disorder as well. A characteristic sign of protein losing nephropathy is low albumin concentration in the blood (hypoalbuminemia) and presence of excess albumin in the urine (proteinuria). Protein losing nephropathy is an adult-onset disorder with onset of signs at the age of 4 to 8 years. Severity of signs can vary from mild to severe. Affected dogs can be treated with medication and controlled diet and those with mild signs can usually have a normal lifespan. Dogs suffering from severe signs have a shorter life expectancy.

Mode of Inheritance

- autosomal recessive

Gene Name

- NPHS1

Next Steps

The condition can be partly managed through diet therapy. Dietary options and the importance of diet management should be emphasized to clients.

References

Littman MP, Wiley CA, Raducha MG, Henthorn PS. Glomerulopathy and mutations in NPHS1 and KIRREL2 in soft-coated Wheaton Terrier dogs. Mamm Genome 24:119-26, 2013.