



Always consult with a veterinarian that you feel comfortable with before diagnosing or treating any disease on your own. This information is for reference only.

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Fanconi Syndrome

Issue Description

Fanconi syndrome is a disorder in which the proximal renal tubules of the kidney do not properly reabsorb electrolytes and nutrients back into the body, but instead "spill" them in the urine. Symptoms include excessive drinking (polydipsia), excessive urination (polyuria), and glucose in the urine (glucosuria.) If Fanconi is left untreated, muscle wasting, acidosis, and poor condition will also occur.

Causes

Fanconi's syndrome appears to have a hereditary predisposition for Basenjis as well as Norwegian Elkhounds. Other breeds that are predisposed (to a lesser degree) are Shetland sheepdogs and Schnauzers. The onset of the disease is not until later in life (3-11 yrs of age in Basenjis), and thus affected dogs may have been bred before diagnosis, passing on the genetic trait. Approximately 10% of adult Basenjis have Fanconi's syndrome. The acquired form of Fanconi's syndrome can be caused by heavy metal poisoning (lead, mercury, cadmium and uranium). Drugs such as a gentamicin, cephalosporins, outdated tetracycline, cisplatin, and streptozotocin can cause proximal renal tubule resorption abnormalities. Chemicals such as Lysol® and maleic acid also have been reported to cause the syndrome. Renal cystic disease and neoplasia, including multiple myeloma and monoclonal gammopathies, also have been found to cause acquired Fanconi's syndrome.

Fanconi's syndrome is a progressive disease, which, if not treated, ultimately results in transport system failure to the point where solute losses are significant enough to overwhelm other compensatory mechanisms and the dog can no longer maintain homeostasis. The most significant of these is the loss of bicarbonate (HCO_3^-). Proximal renal tubular acidosis subsequently develops and, if left uncorrected, will ultimately lead to death. In an unaffected dog with a normal acid-base balance, most of bicarbonate ions in the urine are converted to carbonic acid ($\text{HCO}_3^- + \text{H}^+ \rightarrow \text{H}_2\text{CO}_3$), which is then converted to H_2O and CO_2 with the aid of carbonic anhydrase found in the brush border of the renal tubular epithelial cell.

The carbon dioxide formed readily diffuses across the luminal membrane of the renal tubular cell. In this way, bicarbonate is conserved. The hydrogen ion needed to form carbonic acid is supplied by the sodium-hydrogen ion antiporter, which has a high enough T_m to conserve the needed bicarbonate in an unaffected dog. In Fanconi's syndrome, the T_m is reduced due to either the lack of a sufficient sodium concentration gradient or a defect in the transporter itself. As a result, fewer hydrogen ions are secreted and, thus, less bicarbonate is conserved. The loss of bicarbonate causes an acidemia and the plasma bicarbonate level decreases until it has reached a level which the impaired transport system can handle. Affected dogs can compensate somewhat for the acidemia through respiratory mechanisms (hyperventilation), shifts in intracellular potassium, and secreting hydrogen ions in the distal renal tubule.

Symptoms

A dog suffering from Fanconi's syndrome typically presents with polyuria and polydipsia, a history of weight loss, a poor hair coat and, sometimes weakness.

Diagnosis

Glucose (sugar), and other nutrients and buffers spill into the dog's urine from the faltering renal tubule system of the kidneys; however, the dog will be found to have a normal to low BLOOD glucose. This condition may be mistaken for diabetes and disastrously mistreated. Blood glucose would be HIGH in diabetes.

A dog's urine can easily and inexpensively be checked for glucose with glucose test strips found at any drugstore in the diabetic supplies section. This form of testing can pick up oncoming Fanconi Syndrome well before any of the overt symptoms appear. Overt symptoms are: frequent urination, excessive water-drinking, and loss of weight and condition. Early detection means easier treatment and less permanent damage.

Glucose test stripping at one-month intervals is highly recommended for Basenjis of any age, and for a dog of any breed suspected of having Fanconi Syndrome. Simply hold the glucose testing stick (or diabetic test tape) under the dog's urine, remove, and wait the appropriate amount of seconds. Report any abnormalities in color to your veterinarian and have him do a blood glucose test. If the dog is spilling glucose in the urine and the blood glucose test reading is normal to low, a presumptive diagnosis of Fanconi Syndrome may be made. A subsequent venous blood gas analysis should confirm this diagnosis and tell you the status of the disease process.

Treatment

Approximately every six to eight months, a multivalent blood workup, a urinalysis, and a venous blood gas test need to be done on a Fanconi-afflicted dog by your veterinarian. Based on the dog's individual tests, appropriate, individually-tailored, and easily-obtainable nutrients are included with the dog's daily meals. These oral supplements resupply the nutrients which the tests identify as being lost in the urine.

Prognosis

The outlook is exceptionally good with early detection and treatment. This condition can now be controlled but not cured.

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