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## Malignant Histiocytosis

### Issue Description

*Malignant histiocytosis is a rare hereditary disease found in the Bernese Mountain Dog characterized by histiocytic infiltration of the lungs and lymph nodes. The liver, spleen, and central nervous system can also be affected. Histiocytes are a component of the immune system that proliferate abnormally in this disease.*

### Histiocytic Disorders

A histiocyte is a differentiated tissue cell that has its origin in the bone marrow. The source for histiocytes is the monocyte/macrophage line. Monocytes (found in the blood) and macrophages (found in tissue) are responsible for phagocytosis (ingestion) of foreign material in the body. Langerhans cells are dendritic cells found in the skin and function by internalizing antigens (foreign particles) and presenting them to T cells. They arise from monocytes. Histiocytic disorders refer to diseases that are caused by abnormal behavior of these cells.

### They include the following:

#### Reactive diseases of Langerhans cells

- Histiocytomas
- Cutaneous histiocytosis
- Systemic histiocytosis

#### Reactive diseases of macrophages

- Hemophagocytic syndrome - a condition where macrophages phagocytose myeloid and erythroid precursors (similar to hemophagocytic lymphohistiocytosis in humans)

#### Malignant diseases of Langerhans cells

- Malignant histiocytosis - a condition found in Bernese Mountain Dogs
- Diffuse histiocytic sarcoma
- Localized histiocytic sarcoma

#### Malignant diseases of macrophages

- Histiocytic lymphoma

### Breed Disposition

Up to 25 percent of Bernese Mountain Dogs may develop malignant histiocytosis in their lifetime. Other breeds with a possible genetic tendency toward malignant histiocytosis include Rottweilers, Flat-Coated Retrievers, and Golden Retrievers

### Symptoms

The disease in the lungs is characterized by enlargement of the tracheobronchial lymph nodes and infiltration of the lungs, sometimes leading to lung lobe consolidation and pleural effusion. Signs and symptoms include cough, loss of appetite, weight loss, anemia, and difficulty breathing. Seizures and rear limb weakness can be seen. Invasion of the bone marrow can cause pancytopenia.

## **Diagnosis**

Diagnosis requires a biopsy.

## **Treatment**

Treatment with chemotherapy has been used with some success, particularly using lomustine, prednisone, doxorubicin, and cyclophosphamide. Because of the rapid progression of this aggressive disease, the prognosis is very poor

## **Other Histiocytic Diseases In Bernese Mountain Dogs**

A similar disease is diffuse histiocytic sarcoma, a term used to designate a localized histiocytic sarcoma that has spread throughout the body.

Another disease of histiocytic origin that affects Bernese Mountain Dogs is systemic histiocytosis. This condition generally begins as lesions on the eyelids, nasal mucosa, and skin, especially the scrotum. It progresses to a more generalized disease affecting the lymph nodes, bone marrow and spleen. Other signs and symptoms include weight loss and loss of appetite. It also has a very poor prognosis.

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