

## **Shar pei Mucinosis**

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Mucinosis is a skin condition that is characterized by formation of vesicles (blisters) in the epidermis. The vesicles are filled with a clear, viscous fluid called mucin. The condition is neither itchy, nor painful, except in the most severe cases. This condition occurs almost exclusively in Shar pei dogs.

Shar peis are known for their distinctive full muzzle and thick skin folds. The reason that Shar peis have such thick skin folds is that they have high dermal mucin content. The major constituent of mucin is hyaluronic acid, which is produced by keratinocytes, fibroblasts, endothelial cells and others. Hyaluronic acid is produced by all mammals and aids in the normal functions of cells. Typically, the cell produces hyaluronic acid, it becomes part of the extracellular matrix and is eventually degraded by enzymes. Then it is transported through lymphatics and the blood stream until it is eventually excreted in the urine.

Shar peis have increased levels of hyaluronan synthase 2 (HAS2), which enhances their ability to generate hyaluronic acid. This leads to increased levels of hyaluronic acid (and subsequently mucin) in the skin and in the blood.

The increased ability of Shar peis to produce hyaluronic acid, is also part of the pathogenesis of Shar pei autoinflammatory disease (Shar pei fever). Fragments of hyaluronic acid stimulate the innate immune system and induce fever. Shar peis with a mutation of the gene MTBP are at increased risk of developing Shar pei autoinflammatory disease.

Chronic inflammation due to Shar pei autoinflammatory disease leads to the development of reactive systemic amyloidosis. When large amounts of amyloid deposit in the kidneys or liver, organ failure can result.

Simply having Shar pei mucinosis does not guarantee that the dog will develop Shar pei fever or amyloidosis. However, the presence of mucinosis should alert the veterinarian and owners to observe closely and frequently for signs of fever.

Mucinosis alone, typically does not require treatment. Bathing with a gentle shampoo is helpful to prevent secondary infection in the event that vesicles burst. In cases where mucinosis is causing discomfort, systemic steroids can be used. Systemic steroids quickly deplete mucin in the skin. The result is that thick skin folds flatten, the full muzzle becomes thin and vesicles associated with mucinosis resolve.