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Bronchiectasis, Bulla and Ciliary Dyskinesia
(Pulmonary Structural/Functional Airway Abnormalities)

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BRONCHIECTASIS

Bronchiectasis is a pathological destruction of the elastic and muscular components of the bronchial wall leading to chronic abnormal dilation and distortion of the bronchi. Bronchiectasis may be congenital or may be acquired secondary to other respiratory disease. Acquired bronchiectasis is much more common than the congenital form. Rarely, reversible or pseudobronchiectasis may develop in dogs and must be distinguished from true bronchiectasis. True bronchiectasis is a permanent structural change in the bronchus.

Congenital causes of bronchiectasis include primary ciliary dyskinesia, bronchial cartilage aplasia, bronchial hypoplasia and common variable immunodeficiency. Acquired causes include collapsing trachea, eosinophilic bronchitis, chronic bronchitis, bronchiolitis or bronchopneumonia. The cylindrical form (dilated larger bronchi with nontapering ends) is much more common than the saccular form (circumscribed sacculations of intermediate sized bronchial walls at their terminal end). Cystic bronchiectasis (similar to saccular but in the terminal bronchi) has been described in the dog but is rare; where as varicose bronchiectasis (beaded, widened bronchi with irregular contours) has not been described. Bronchiectasis may be focal or diffuse in nature. Focal lesions tend to be secondary to a focal underlying disease (e.g., neoplasia, aspiration pneumonia, foreign body).

Since bronchiectasis is typically acquired, it is not surprising that most dogs with bronchiectasis are middle age or older with no sex predilection. The American cocker spaniel appears to be an overrepresented breed. Dog breeds that are predisposed to conditions that may result in bronchiectasis, like collapsing trachea, may be more likely to develop bronchiectasis. Clinical signs typically reflect the underlying disease process. These signs may include cough, retch, tachypnea and dyspnea.

The diagnosis of bronchiectasis consists of discovering dilated airways and then investigating the underlying etiology. Dilated airways may be detected via thoracic radiography, bronchography, CT, bronchoscopy or histology. Thoracic radiography may not be a sensitive indicator of bronchiectatic changes and bronchiectasis has been documented in dogs with normal thoracic radiography. Other modalities such as bronchography, CT, bronchoscopy or histology may be necessary to detect lesions, especially early lesions, in some patients. Visual examination of bronchiectatic airways should reveal pronounced dilation of the airway and accumulation of purulent exudate.

Bronchiectasis is an irreversible structural change in the airway so there is no therapy that can reverse this process. The goal of therapy, instead, is to control clinical signs and avoid further damage to the airways. The exception may be patients with focal bronchiectasis, where a lung lobectomy may be curative. Focal bronchiectasis treated with lung lobectomy has a good prognosis. For dogs with diffuse bronchiectasis, prognosis relates to the underlying cause, severity of lesions and their clinical manifestations.

PULMONARY BLEBS AND BULLAE

Pulmonary blebs are accumulations of air within the layers of the visceral pleura. They are most commonly found at the lung apices. Blebs may be up to several centimeters in diameter and develop when air escapes from the lung and is trapped between the layers of the visceral pleura. Bullae are air filled spaces within the pulmonary parenchyma that form because of destruction of the alveoli. The size of bullae can be variable with the largest bullae consuming the majority of the lobe. Most blebs or bullae are discovered in patients with spontaneous pneumothorax. Dogs with bleb or bulla-induced pneumothorax are most often middle aged, large breed dogs. Often dogs have no clinical signs associated with the respiratory system until there is rupture of the bleb or bulla with subsequent development of a pneumothorax. The diagnosis of blebs or bullae can be difficult. Radiographic evidence of blebs or bullae may be lacking unless the lesions are quite large. Many dogs that develop spontaneous pneumothorax secondary to blebs or bullae will have no radiograph changes aside from those consistent with pneumothorax. CT scan is recommended in these patients as it is a more sensitive imaging modality for the detection of blebs or bullae. Definitive treatment consists of surgical resection of the affected areas. Prognosis is variable depending upon the extent of the lesions and the inciting disease process.

PRIMARY CILIARY DYSKINESIA

Primary ciliary dyskinesia (ie immotile cilia syndrome and congenital ciliary dysfunction) is a congenital disorder associated with defective ciliary motility with or without ultrastructural abnormalities resulting in the impairment of the mucociliary apparatus. Kartagener's syndrome is a triad of situs inversus, rhinosinusitis and bronchiectasis. In healthy animals, the mucociliary apparatus moves inhaled debris and mucous from the lung to the pharynx for expectoration. Dogs with primary ciliary dyskinesia have chronic mucous plugging, inflammation and infection.

Primary ciliary dyskinesia is most commonly diagnosed in young dog and has been reported in many breeds of dog including the English pointer, English springer spaniel, English setter, Border collie, Old English sheepdog, Doberman pinscher, Chihuahua, golden retriever, Newfoundland and bichon frise. This condition is rare in cats. Dogs with primary ciliary dyskinesia present for clinical signs associated with rhinosinusitis, bronchitis and bronchopneumonia. Clinical signs are typically antibiotic responsive but return with the cessation of antibiotics. Some dogs will have clinical signs associated to dyskinetic cilia in other organs like the brain, ear or reproductive tract. Diagnosis is made based on clinical signs and evidence of ciliary dysfunction. Evidence of ciliary dysfunction

includes: 1.) identification of immotile sperm; 2.) mucociliary scintigraphy detecting abnormalities in mucociliary transport; 3.) ultrastructural abnormalities of the cilia from the nasal and or tracheal mucosa; 4.) abnormal ciliary beat frequency and synchrony identified via computerized microscope photometry; and/or 5.) abnormal ciliogenesis.

There is no definitive treatment for primary ciliary dyskinesia. Recurrent infections should be treated with antibiotics. It is imperative that antibiotic selection is based on culture and sensitivity results in these patients to avoid induction of antibiotic resistance. Cough suppressants should never be used in a patient with primary ciliary dyskinesia as coughing is the only method of removing debris and mucous from the airways. Minimizing environmental pulmonary irritants is also helpful for many patients. Prognosis varies based on the severity of the ciliary dysfunction. Although many dogs with primary ciliary dyskinesia will have recurrent pulmonary infections, with appropriate aggressive care dogs may live several years after diagnosis.

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