Craniomandibular Osteopathy in Westies and other Scottish Terrier Breeds

Craniomandibular osteopathy is a non-neoplastic disease that primarily affects the mandible and tympanic bullae in terriers, particularly Scottish, West Highland White and Cairn terriers. In one study, 66 of affected dogs were either West Highland White or Scottish Terriers. The disease also has been reported to occur in other breeds, including Irish Setters, English Bulldogs, Shetland Sheepdogs, Great Danes, Boxers, Labrador Retrievers, Doberman Pinschers, Pit Bull Terriers, Bullmastiffs and Akitas. It is important to recognize that the incidence of the disease is low; in one study, LaFond et al identified 35 cases of craniomandibular osteopathy in a study of more than 300,000 case records from 10 veterinary teaching hospitals, an incidence of 0.01%. Nineteen of the 35 cases occurred in Westies.

Craniomandibular osteopathy is known by several synonyms, such as 'mandibular periostitis', 'Westie jaw', 'Scottie jaw' and 'lion's jaw'. Thickening of the mandible and the bullae results in pain, particularly when the dog chews its food. In many cases, enlargement of the angular processes of the mandible and the bullae prevents the dog from fully opening its mouth. Typically, signs of pain first become evident when the dog is between 4 and 7 months of age, and may be associated with intermittent episodes of fever. The pain associated with the disease adversely affects the dog's ability to eat, gain weight and grow. In some cases, the disease may become self-limiting when the dog reaches 12 to 18 months of age. At this time, abnormal bone growth slows or even may cease, coinciding with completion of regular endochondral bone growth and ossification.

The severity of the disease, time of onset, and rate of progression vary considerably among affected dogs. A lack of a gender predisposition has been documented in two studies (Watson et al, 1995; LaFond et al, 2002). It also has been reported that the disease can affect several dogs in the same litter (TronwaldWigh et al, 2000). Although an early study of the genetics of this disease, using retrospective pedigree analysis (Padgett et al, 1986), indicated that the disease is inherited as a simple autosomal recessive characteristic, this has recently been proven to be false. A more recent study at the University of Bern determined that a mutation in chromosome 5 causes the disease. In that study, approximately 85% of affected dogs had two copies of the mutation, 10% had one copy, and the remainder did not carry it. Thus, while the presence of the mutation strongly influences the presence of the disease, other genetic and/or environmental factors must be involved. The investigators also reported that incomplete penetrance can occur, meaning that dogs with one copy of the mutation might develop clinical signs, while others will not. The Westie Foundation coordinated the gathering of frozen samples for that research project, which were shipped to Switzerland. A genetic test now is available through Optimal Selection **(www.https://optimal-selection.com).** The results of a study comparing three canine models of human rare bone diseases included craniomandibular osteopathy in Westies were published recently and are available online at the following site: **http://journals.plos.org/plosgenetics/article?id=10.1371/journal.pgen.1006037**

Diagnosis of Craniomandibular Osteopathy

The signs of craniomandibular osteopathy first identified by owners of affected dogs are difficulty grasping, holding and chewing food, drooling, and swelling of the face around the jaw. Some affected dogs may appear to be painful when the mouth is opened, and it may not be possible to open the dog's mouth fully. It has been reported that a painful reaction may be elicited when the dog's jaw and the joint between the jaw and skull (temporomandibular joint) are palpated (Padgett

Common Clinical Findings
4 – 7 Months of Age
Mandible and Tympanic Bullae
Pain When Eating

> Affected dogs have difficulty grasping, holding and chewing food, drooling, and swelling of the face around the jaw.

et al, 1986). Some affected dogs may be febrile, and some appear swollen over the jaw, temporomandibular joint, and at the base of the skull. This swelling is due to excessive growth of disorganized bone. Common laboratory tests typically are not useful in diagnosing this disease, and inconsistent results are obtained when indicators of bone remodeling or proliferation (e.g., serum inorganic phosphorus or alkaline phosphatase) are evaluated.

A definitive diagnosis of craniomandibular osteopathy is usually made by identifying the characteristic changes seen on radiographs (x-ray images) of the temporal bone of the skull and the mandible of affected dogs (Schwarz et al, 2002). These changes include a disorderly proliferation of bone on the surfaces of the affected bones (mandible and temporal bone) that limits the function of the temporomandibular joint between these bones (**Figures 4.4-4.6**). In most affected dogs, the changes occur bilaterally and in about one-third of dogs the changes are limited to the mandible. It is important to know that this proliferation of bone is not a tumor (neoplasm).

The rate at which proliferation of bone occurs differs among dogs. In some dogs, the degree of proliferation is minimal and this results in minimal loss of function. In other dogs, however, the proliferating bone restricts movement of the temporomandibular joint, the dog's ability to open its mouth, grasp and chew food is reduced. In very severe cases, affected dogs can only open their mouth to a limited degree and experience very significant pain when trying to grasp food and chew. These severely affected dogs cease using the muscles that control these functions, and the muscles atrophy (shrink in size). This loss of muscle in the head and jaw may further compromise the dog's ability to eat. In the most severe cases, the proliferating bone may fuse the bones of the jaw to those of the skull. These severely affected dogs rarely show improvement clinically.

The changes that occur in this disease include bony proliferation, bone remodeling, increased connective tissue within and surrounding bone, and variable degrees of inflammation in and around bone (Riser et al, 1967). It is possible that the inflammation may play a role in the

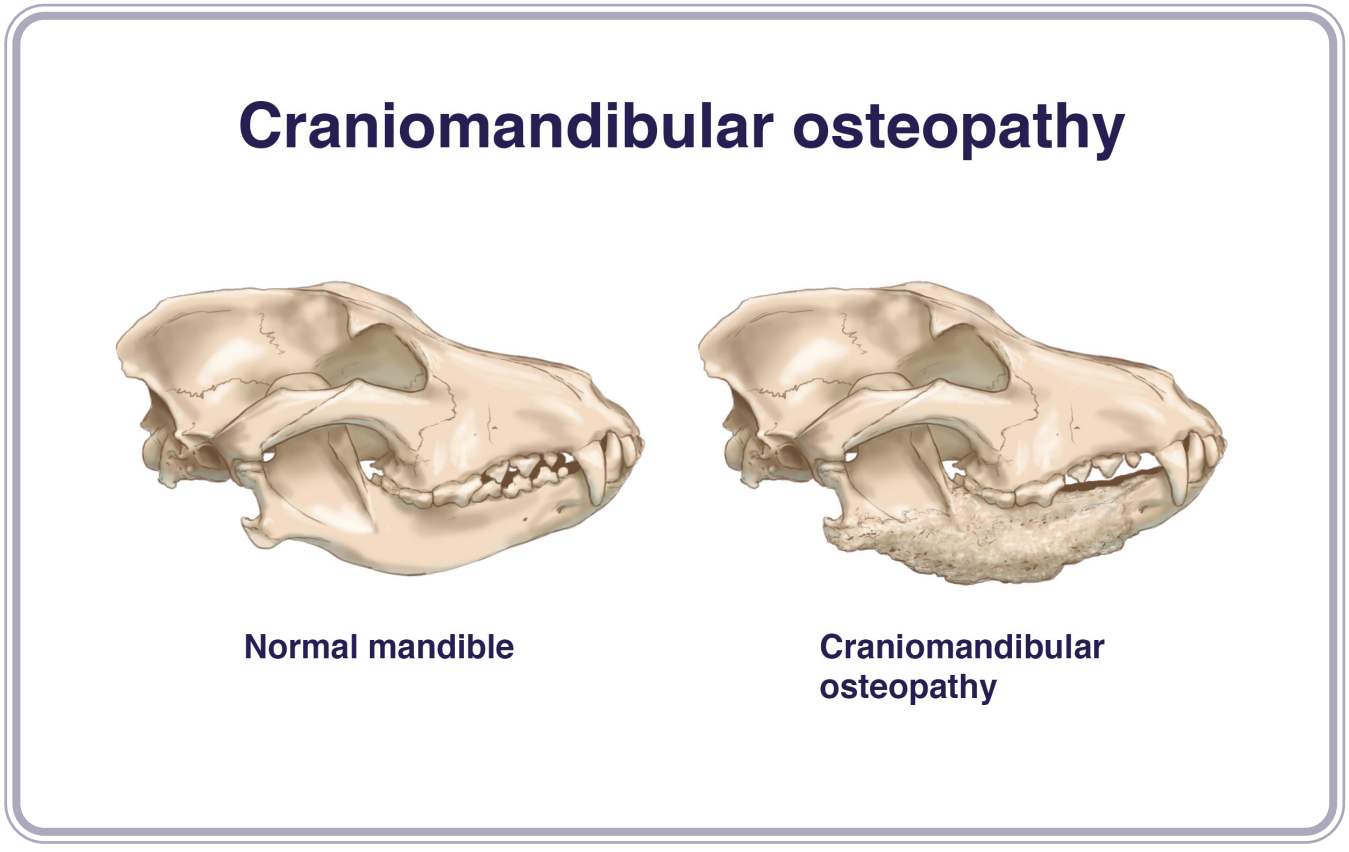

Figure 4.4 - An illustration depicting the abnormal thickening of the bones in a dog with craniomandibular osteopathy.

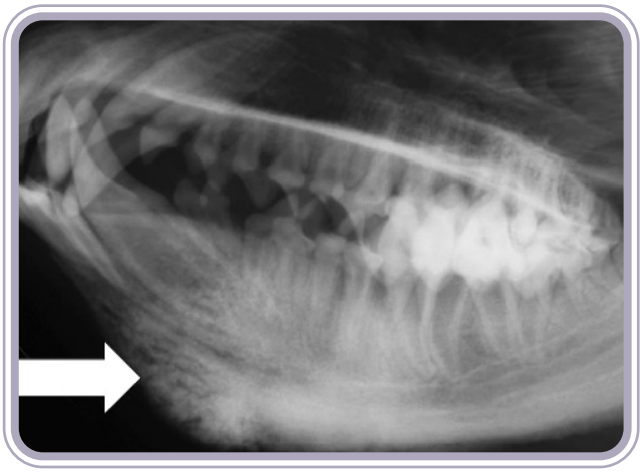

Figure 4.5 - Lateral radiographic view of the disorderly bony proliferation (white arrow) on the mandible (lower jaw) of a dog with CMO. The top of the skull is at the top of the figure, and the teeth/tooth roots clearly visible for reference. (Figure courtesy of Dr. Greg Daniel, VMRCVM; all rights reserved)

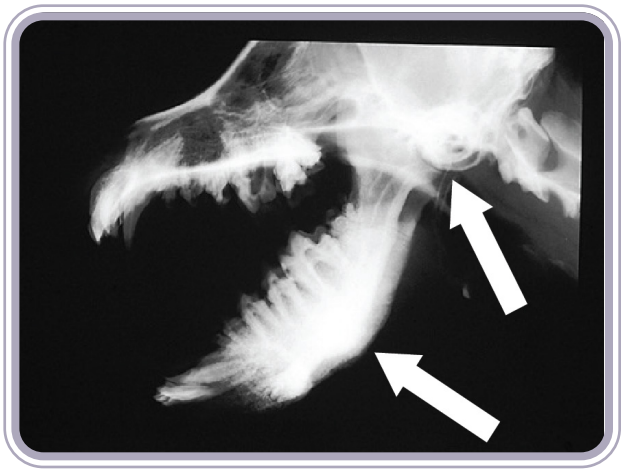

Figure 4.6 - Radiograph of anesthetized dog with CMO, with mouth open and view taken from the nose toward the back of the mouth. An endotracheal tube can be seen in the mouth (center, lower). Bilateral (both sides) disorderly bone growth on the mandibles and temporomandibular joints (white arrows) can be seen. The growth of this bone limits opening and closing of the mouth, ability to eat, is associated with pain and can be irreversible. (Figure courtesy of Dr. Martha Moon Larson, VMRCVM; all rights reserved)

development of the condition, perhaps by supplying specific growth factors that stimulate bone growth. Consequently, affected dogs are often administered antiinflammatory drugs in an effort to control pain, reduce the fever, and perhaps slow progression of disease.

In rare cases, dogs may have radiographic evidence of bone proliferation on the mandible and skull, but not have trouble eating. Equally uncommon, some dogs may have disorderly proliferation of bone on other bones in the body, including the bones of the legs (Padgett et al, 1986).

Other Things to Consider

Although Westies as a breed are more likely to develop craniomandibular osteopathy, the condition is relatively rare and there are several other reasons why a Westie might be having difficulty eating or a painful mouth. A good saying to remember is "Common things happen commonly", and the following possible causes for these clinical signs should be considered first (listed in no particular order):

- Abnormalities of the teeth and gums
- Mouth, nose and throat infections
- Oral ulcers or ulcers elsewhere in the digestive tract
- Strains, sprains and fractures
- Tumors of soft tissues and bones of the skull and jaw
- Inflammation of the muscles of the head and neck
- Palatability of food items offered
- Exposure to toxins in the diet or environment

It is very important to see your veterinarian if your Westie is failing to gain weight, has trouble eating, appears to be painful or is drooling.

Treatment of Craniomandibular Osteopathy

There is some controversy regarding the progression of craniomandibular osteopathy. Most veterinarians indicate that the disease is selflimiting, often regresses (slows and stops with age) and at times completely resolves (Riser et al, 1967; Alexander, 1983), although there is variability in the progression of the disease. In their review, Watson and colleagues (1995) noted that bony proliferations may show smoothing and remodeling in some bones of some dogs.

Many veterinarians advocate the use of antiinflammatory drugs to help control pain, fever and swelling in dogs with the disease. The judicious use of these drugs (both corticosteroids and nonsteroidal drugs, such as aspirin, carprofen and meloxicam) allows affected dogs to eat and drink. While attempts have been made to surgically eliminate the bony fusions in the temporomandibular joint of severely affected dogs, this approach has not been successful. It is clear that the best approach is to prevent the development of the condition by selective breeding.

Current Research About Craniomandibular Osteopathy

Lopez LA, Ruiz JC, Steenkamp G, Holdsworth A. Computed tomographic characteristics of craniomandibular osteopathy in 20 dogs. Front Vet Sci 2024 Sep 30:11:1436356.

Craniomandibular osteopathy is a proliferative, self-limiting, non-neoplastic disease of growing dogs characterized by excessive new bone formation on the skull and mandible. This paper aims to characterize the spectrum of CT findings that can occur with this condition. The study is retrospective, descriptive, multicenter, and includes 20 cases, 80% of which were Terrier breeds. Mandibular osteoproliferation was present in all patients, cranial osteoproliferation was present in 90%, and tympanic bulla osteoproliferation was present in 60%. CT allows for detailed characterization of the bony changes associated with this condition, including the effects occurring secondary to osteoproliferation surrounding the tympanic bullae.

Guidi EE, Vercelli A, Corona A, et al. A case of drug-resistant staphylococcal para-aural abscess treated with photodynamic therapy in a West Highland White Terrier presenting with chronic otitis and craniomandibular osteopathy

A 10-year-old West Highland White Terrier with chronic unilateral otitis and a history of unsuccessful systemic and topical antibiotic treatments was examined. A computer tomography scan revealed unilateral chronic otitis with calcification of the ear canal, abscessation and fistula. On bacterial culture a Staphylococcus pseudintermedius sensitive to pradofloxacin was isolated. Systemic treatment with pradofloxacin, 3 mg/kg per os once daily, improved the infection and the dog had total ear canal ablation and bulla osteotomy performed. After one month, despite antibiotic treatment new fistulas developed in the same area. Bacterial culture revealed S. pseudintermedius sensitive only to rifampicin. Under general anesthesia, the area was cleaned and 1 mL of indocyanine green was injected into the fistulas. Treatment with diode laser of four cycles in the affected area was performed. No antibiotic was administered and one week later, 50% of the fistulas were closed with a significant improvement of the patient's clinical condition. Cytology and bacterial culture were negative 72 h after the treatment. Total recovery occurred after two weeks. Photodynamic therapy is a promising antibacterial technique in case of localized refractory bacterial infections.

Beever L, Swinbourne F, Priestnall SL et al. Surgical management of chronic otitis secondary to craniomandibular osteopathy in three West Highland white terriers. J Small Anim Pract 2019 Apr;60(4):254-260.

Three West Highland white terriers were presented for investigation of left-sided para-aural abscessation. CT revealed chronic otitis media with extensive osseous proliferation surrounding the horizontal and vertical ear canals contiguous with the expanded temporal bone, consistent with a unilateral variant of craniomandibular osteopathy. A left total ear canal ablation with lateral bulla osteotomy was performed in all dogs. An ultrasonic bone curette proved useful when removing the osseous proliferation in two dogs. Histopathological examination of the ear canals was consistent with craniomandibular osteopathy and the treatment led to resolution of the presenting clinical signs in all dogs. This is the first report of craniomandibular osteopathy engulfing the external ear canal, presumably leading to chronic otitis media and para-aural abscess formation, and the first reported use of an ultrasonic bone curette in canine otic surgery.

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