Topic #5: Craniomandibular osteopathy (CMO) in Westies and other Scottish terrier breeds

The ability to eat and to drink is essential to staying alive! Eating and drinking are complex activities, dependent on coordination of the processes of hunger and thirst by the brain and also on the function of nose, mouth, esophagus and gastrointestinal tract. The bones of the skull (the temporal bone, temporomandibular joint, tympanic bullae, and mandible), muscles of chewing (temporal muscles, primarily), attached ligaments, and tendons, allow dogs to hold and chew their food effectively. Malfunction in any part of the “eating/drinking” system is serious. When disease develops, it can affect the dog’s ability to eat, gain weight, grow, and in extreme cases, to live.

West Highland White Terriers are very highly affected with craniomandibular osteopathy (CMO), a non-neoplastic (not a tumor) disease altering form and function of the bones of the skull and mandible (jaw bone). This disease is known by several synonyms, such as 'mandibular periostitis', 'Westie jaw', 'Scottie jaw' and 'lion’s jaw' (Alexander, 1983). Other Scottish terrier breeds (Scottish Terriers and Cairn Terriers) are affected more commonly than other breeds of dogs, such as Irish Setters (Tronwald-Wigh, G, et al, 2000), Labrador Retrievers (Alexander, et al, 1975), Doberman Pinschers (Watson, ADJ, et al, 1975), English Bulldogs (Hathcock, 1982), Pyrenean Mountain Dogs (Franch, et al, 1998), and Shetland Sheepdogs (Taylor, et al, 1995).

CMO was first recognized in England in 1958 ((Littlewort, MC, 1958) and the pathology of the disease was studied extensively by Wayne Riser and his colleagues at the University of Pennsylvania. Drs. Riser and Newton have written a very extensive review of CMO. You can read this review on-line at http://www.ivis.org/special_books/ortho/chapter_54/ivis.pdf (Riser and Newton, 1985). In addition to this Web citation, there have been excellent reviews of this subject, and these are listed in References for this Topic (Alexander, JW, wt al, 1983; Watson, ADJ, et al, 1995; LaFond, et al, 2002, Schwarz, T, et al, 2002;).

The incidence of CMO: Not rare in Westies!

In a review of literature on CMO, Watson and colleagues (Watson, et al, 1995) found reports of 81 affected dogs: 44 of 81 dogs were West Highland White Terriers, 22/81 were Scottish Terriers, 2/81 were Cairn Terriers. Taken together, these three Scottish terrier breeds comprised about 84% of cases described in literature reviewed. In a very large study of information on developmental orthopedic diseases in dogs contained in medical records, and collected over a ten-year period of time, LaFond and colleagues (LaFond, et al, 2002), found that Westies were the only breed with CMO. They identified 35 cases of CMO among 24,373 records of dogs of all breeds with developmental orthopedic diseases. It is clear that the incidence of CMO among dogs is low, but a significant problem for owners of Westies (see more below).
Onset and clinical signs

CMO is a developmental disease, primarily affecting the form and function of bones of the skull of young dogs. When we say the disease is “developmental” we mean that there is a genetic predisposition to the disease in some dogs, and it usually becomes a problem at a particular stage of life. In this case, CMO usually begins to “develop” beginning around 6 months of age and may be fully developed in affected dogs by 18 months of age (see the Table 1, found below, from Watson, et al., 1995, modified slightly for this Topic). Data from this Table show that slightly more than 90% of dogs with CMO develop the disease by 1 year of age.

Table 1. Age at diagnosis of 153 dogs with craniomandibular osteopathy included in the Veterinary Medical Data Base (VMDB) at Purdue University

<table>
<thead>
<tr>
<th>Breed category</th>
<th>Age 2-6 months</th>
<th>Age 6-12 months</th>
<th>Age 1-2 years</th>
<th>Older than 2 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Terriers</td>
<td>37.9%</td>
<td>33.3%</td>
<td>3.3%</td>
<td>3.9%</td>
</tr>
<tr>
<td>Other breeds</td>
<td>9.2%</td>
<td>10.5%</td>
<td>1.3%</td>
<td>0.6%</td>
</tr>
<tr>
<td>Total</td>
<td>47.1%</td>
<td>43.8%</td>
<td>4.6%</td>
<td>4.5%</td>
</tr>
</tbody>
</table>


In considering the data contained in Table 1, it is important to understand that while young West Highland White Terriers and other Scottish terriers are predisposed to developing CMO, the disease cannot be considered common in dogs as a species. The data summarized in this Table included 153 affected dogs, but this disease represented a very small fraction (0.014%) of the 1,080,396 canine entries in the VMDB at the time the table was compiled. Similarly, LaFond and colleagues only found CMO in 35 Westies in a study of 300,122 VMDB records (1986-1995) (an incidence of 0.01%). The fact that this particular disease is very uncommon has implications for research, discussed later (See Research and Additional Information, below).

Individual dogs may vary widely in the severity of disease, the time of onset, and the rate of progression (more on this below). In one review, there was no sex predisposition noted, with males and females being represented in approximately equal numbers (Watson, et al., 1995). This lack of gender predisposition has been confirmed by others (LaFond, et al., 2002).

The disease can affect several individual dogs in the same litter (Tronwald-Wigh, et al., 2000). A study of the genetics of this disease, using retrospective pedigree
analysis, conducted by Padgett and co-workers (Padgett, et al, 1986) has shown that the disease is inherited as a simple autosomal recessive characteristic. As breeders know, this means that the breeding of dogs that may possess the genetic mutation causing CMO, but not showing signs of disease (carriers of a defective gene), are the source of disease. Diseases caused by the expression of recessive genes occur when dogs acquire mutated genes from both parents.

The diagnosis of CMO

The first signs of CMO noted by owners of affected dogs are difficulty eating (grasping, holding and chewing food), drooling, and swelling of the face around the jaw. Some affected dogs may display signs of pain when the mouth is opened, and there may be restriction in the amount the jaws can open. Padgett and Mostosky (Padgett, GA, et al, 1986) report that palpation of the jaw and temporomandibular joint, with pain on palpation, is a dependable method that can be used clinically to diagnose dogs with CMO. Fever may or may not be present. Affected dogs may show swelling of the face over the jaw, at the joint of the jaw and the skull (the temporomandibular joint), and at the base of the skull. This swelling is due to the proliferation of disorganized bone (see below). Laboratory tests for serum and hematologic (blood) abnormalities are not useful in diagnosing this disease. Analysis of indicators of bone remodeling or proliferation, such as serum inorganic phosphorus or alkaline phosphatase gives inconsistent results among affected dogs. Definitive diagnosis is usually made by characteristic radiographic changes, described below (Schwarz, et al, 2002).

Radiographs (x-ray images) of the temporal bones and mandible of dogs with CMO show disorderly proliferation on bone surfaces, compromising the function of the joint linking these bones (the temporomandibular joint) (Figures 1-3, courtesy of Drs. Greg Daniel and Martha Moon Larson, Virginia-Maryland Regional College of Veterinary Medicine [VMRCVM]). This proliferation of bone is not a tumor (neoplasm).

Other bones in the body, including long bones in legs (radius and ulna) may also show similar disorderly proliferations, although this is not common (Padgett, et al, 1986). In some dogs, bony changes may be present on radiographs, but dogs do not display typical clinical signs; this is uncommon.
**Figure 1.** Drawings of the skull of a dog with CMO, lateral and ventral views. Arrows and darkened lines show sites of body proliferation in mandible, temporal bones, tympanic bullae and temporomandibular joints. (Figure courtesy of Dr. Greg Daniels, VMRCVM; all rights reserved)

**Figure 2.** 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 3. 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)

Proliferating bone may decrease movement of the temporomandibular joint, limiting the ability of the affected dog to open its mouth, grasp and hold food and to chew. In very severe cases, dogs with CMO can only open their mouth to a limited degree and experience very significant pain when trying to grasp food and chew. Dogs experiencing pain and limited mobility of the jaw may develop secondary atrophy (shrinkage) of the muscles that allow chewing, due to disuse. This loss of muscle in the head and jaw may further compromise the affected dog’s ability to eat.

Riser and colleagues described histologic bony changes as bony proliferation, bone remodeling, increased connective tissue within and surrounding bone, and variable degrees of inflammation in and around bone (Riser, et al, 1967). The inflammation seen may play a role in the development of disease, perhaps supplying growth factors stimulating bone growth. It is pretty clear that treating dogs with anti-inflammatory agents may help control pain, fever, and perhaps the progression of disease (See Treatment of CMO).
Bony lesions progress at differing rates for different dogs. In some dogs, lesions are minimal and cause only minimal loss of function. In more severely affected dogs, bony changes may fuse the bones of the jaw to those of the skull. These severely affected dogs rarely show improvement clinically.

Your veterinarian needs to consider a variety of possible diseases when presented with a Westie with difficulty eating and with mouth pain. A good saying to remember is “Common things happen commonly” and that although Westies, as a breed are most likely to develop CMO, it is a relatively rare condition. Some possible causes of difficulty eating and/or mouth and jaw pain are (in no particular order):

- Bad teeth and bad gums
- Strains, sprains and fractures due to trauma
- Mouth, nose and throat infections
- Tonsillitis
- Tumors of soft tissues and bones of the skull and jaw
- Inflammation of the muscles of the head and neck
- Sprains, strains and fractures of the neck
- Palatability of food items offered
- Oral ulcers or ulcers elsewhere in the digestive tract
- Exposure to toxins in the diet or environment
- And many others….

It’s very important to see your veterinarian if you notice poor weight gain, trouble eating, pain, or drooling.

**Treatment of CMO**

There is some controversy regarding the progression of CMO. Riser and colleagues write that the disease is self-limiting (slows and stops with advancing age), often regresses and at times completely resolves (Riser, et al, 1967). Alexander noted a lot of variation in the progression of disease. He felt that some dogs might be minimally affected and the disease only seen with radiographs. Some dogs, unfortunately, had variable amounts of bony fusion of the temporomandibular joint and severely affected dogs were euthanized. Alexander, too, concluded that in most cases, CMO was self-limiting and that as dogs matured, the disease stopped progressing (Alexander, 1983). Padgett stated “With corticosteroid treatment (or sometimes spontaneously) remodeling occurs, the new bone is removed and the jaw line returns to a normal or near normal appearance” (Padgett, et al, 1986). In their review, Watson and colleagues note that bony proliferations may show smoothing and remodeling in some bones of some dogs (Watson, et al, 1995). Most authors agree that the use of anti-inflammatory agents can be helpful (see Treatment of CMO).

Many of the authors cited in this Topic advocate the use of anti-inflammatory drugs to help control pain, fever and swelling in dogs with CMO. The judicious
use of these drugs (both corticosteroids and non-steroidal drugs such as aspirin) allows affected dogs to eat and drink. Several articles have suggested the use of these drugs may interfere with progression of disease, but according to Riser and Newton "Most animals can be made comfortable using aspirin or corticosteroids; however, treatment does not result in cure" (Riser, et al, 1985).

There have been scattered reports of attempts to surgically free up bony fusions in the temporomandibular joint of severely affected dogs. This does not appear to help.

It is very clear that the best treatment for CMO in Westies is prevention by selective breeding.

**Current research and additional Information**

This disease attracted considerable attention between 1958, when it was first described, and 2002. Some of the features of CMO in Westies resembled skeletal diseases seen in other dog breeds and in people.

A disease of Bull Mastiffs, known as "idiopathic hyperostosis" or "calvarial hyperostotic syndrome (CHS)", causes asymmetrical thickening of some bones of the skulls in young, male dogs (Pastor, et al, 2000). McConnell and co-workers (McConnell, JF, et al, 2006) described magnetic resonance findings in two dogs, one of which had bony proliferation in the femur (sometimes seen in several skeletal diseases, including CMO). Huchkowsky (2002) successfully treated CMO-like syndrome in a 6-month old Bull Mastiff with anti-inflammatory drugs. While some of the proliferative bony lesions seen in CHS of Bull Mastiffs are similar histologically to CMO in Westies, there are important differences including the sex of affected dogs (only male Bull Mastiffs get CHS, while both sexes of Westies get CMO), and the bones affected (CHS in Bull Mastiff affects frontal, temporal and occipital bones asymmetrically; CMO in Westies affects the mandible and temporal bones, and usually symmetrically). A comparison of the underlying genomic mutations controlling each of these diseases (CHS, CMO) should be done to assess potential links and possible molecular therapies.

CMO was described as part of a complex pattern of disease in 12 dogs from 6 litters of Irish Setters (Tronwald-Wigh, et al, 2000). All of these dogs suffered from a primary disease condition known as “canine leukocyte adhesion deficiency (CLAD)”. All of the dogs were less than 15 weeks old when they developed a variety of infections, including bone (‘osteomyelitis’) and mouth (‘gingivitis’) infections. The susceptibility of these dogs to infection was due to the fact that their white blood cells (‘leukocytes’) did not function properly to protect against infectious agents like bacteria in the environment. Eleven of 12 dogs studied developed proliferative bony growth on mandibles, with the characteristic cobblestone appearance of CMO in 5/12 dogs. Histologic sections of bone showed increased activity of bone remodeling cells, bone death, and inflammation. The results of studying these dogs is of some interest, since
inflammation is a relatively common feature in CMO and one cannot exclude the possibility that one initial trigger for CMO might be exposure to infectious agents. This has relevance to observations of people with Padget’s disease (see next paragraph).

Some aspects of the development of CMO in Westies have interested researchers studying human bone diseases. For example, Padget’s disease is a relatively common non-neoplastic proliferative bone disease, generally seen in older people (Inzucchi, 2006). The radiographic and histologic appearance of newly-formed and disorderly bone in people with Padget’s disease resembles the appearance of newly-formed bone in CMO. In people, the bony growth occurs in the skull, pelvis and long bones of the legs and arms – somewhat similar in location to the location of CMO in Westies. Some research done on people with Padget’s disease has suggested, but not proven, that a viral infection might trigger the beginning of the disease. A role for viral infection in CMO has not been studied.

Another human disease, infantile cortical hyperostosis, resembles some aspects of the development of CMO in Westies. It is a genetic disease – an autosomal dominant disease, which results in non-neoplastic bony proliferation (Padgett, et al, 1986).

Food for thought

It’s pretty clear that CMO is a Westie problem. We understand the genetics of it and how to diagnose it. We know that selective breeding can reduce the incidence of the disease. What we don’t know is what if the key problem with the genome (one or more mutations) which trigger it and allow development. Understanding this could be important not only for diagnosis and treatment, but also to see if some of these same genomic mutations might be associated with other Westie and dog diseases. Unfortunately, the rarity of this disease among dogs and the absence of an identical disease in people mean that CMO is not likely to get studied much. Veterinarians and researchers may think they have the complete story. We think the book on CMO and other genetic disorders may only be open be on the first page!

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References


