

Presentation on
CranioMandibular Osteopathy (CMO)
in Scottish Terriers

to Members of
The Scottish Terrier Club of Chicago

on

January 16, 2005

by

George Procento, MD

STCC Member



from:
*Off
The
Mark*
by
Mark
Parisi

CRANIOMANDIBULAR OSTEOPATHY

“ **CMO** “

- INHERITED / RECESSIVE
- VERY PAINFUL PUPPY
- EUTHANIZED IF NOT PROPERLY RECOGNIZED
- TREATMENTS CAN ALLEVIATE PAIN
- COMPLETE REMISSION



NERA

1992 - 2004

Scottish Terrier Club of America

WEBSITE

<http://clubs.akc.org/stca/geneticissues.htm>



CRANIOMANDIBULAR OSTEOPATHY (CMO) is an inherited disorder characterized by an abnormal growth of the bone of the lower jaw. **CMO** usually appears between four and seven months. A puppy with **CMO** usually pulls away, flinches or screams with pain when his mouth is examined, depending on the severity of the disease. Other early symptoms are lethargy, fever and unwillingness to eat. An acutely affected puppy may be unable to open his mouth but mild cases may be misdiagnosed as teething problems or virus symptoms. An accurate diagnosis of **CMO** requires X-ray confirmation.

CMO is nearly always treatable. Mild cases respond to aspirin or other non-steroidal anti-inflammatory drugs such as ibuprofen or while acute cases may require the use of steroids such as prednisone or prednisolone. Fortunately, as the dog matures, the abnormal bony growth abates and is often undetectable in the adult dog, even by radiography.

- First mention of CMO in veterinary literature - 1958

Littlewort, M.: *Tumor-like Exostoses on the Bones of Head in Puppies* Vet. Rec. 70:977

- United States, Canada and Europe

- CARRIER RATES:

Scottish Terriers - 4.7% (1995)

- West Highland White Terriers - 22.1% (1999)

- Cairn Terriers - 13.59% (1999)

- 11 Terrier breeds

- 31 breeds affected *

including: Great Danes, Doberman Pinschers,
Bull Mastiffs, Labrador Retrievers

* *Control of Canine Genetic Diseases* (1998)

George Padgett, DVM – MSU (Professor Emeritus)

1995 STCA Health Survey *

<u>DISEASE</u>	<u>Mode of Inheritance</u>	<u>Frequency of Occurrence</u>	<u>Est. Carrier Frequency</u>
CMO	R	~ 1 / 1700	4.7%
Hip Dysplasia	Poly	~ 1 / 1700	4.7%
von Willebrand's	R	~ 1 / 500	8.4%
Cerebellar ataxia	R	~ 1 / 200	13.0%
Patellar Luxation	R?, Poly?	~ 1 / 80	19.5%
Underbite	Und	~ 1 / 50	24.7%
Scottie cramp	R	~ 1 / 30	29.3%
Atopic dermatitis (skin allergies)	Und	~ 1 / 18	35.8%

* <http://clubs.akc.org/stca/healthsurvey.htm>

Control of Canine Genetic Diseases, G.Padgett, DVM (1998)

1995 STCA Health Survey *

CMO

Frequency of
Occurrence

~ 1 / 1700

* <http://clubs.akc.org/stca/healthsurvey.htm>

Control of Canine Genetic Diseases, G.Padgett, DVM (1998)

1964-1991 Purdue University * *

CMO

Frequency of
Occurrence

~ 1 / 7000

* Purdue U. Veterinary Medical DataBase, 1964-1991,

153 listings of CMO among **1,080,396** dogs;

The Compendium, Small Animal, Vol.17, No.7, July 1995, CMO in Dogs,

Watson, Adams, Thomas, pp.911-922

CMO ON THE RISE ?

- Breeders are more willing to admit problems and talk about them
- After 70-odd years of breeding the gene pool in Cairns is becoming smaller and smaller and odds of running into any problem are rising
- Breeders tend to use only a few (top-winning) studs and are concentrating the gene pool even further
- (CMO)Dogs represent a broad cross section of the top bloodlines in this country
- “ CMO HAS BECOME A BREED PROBLEM ”
- (to Cairn Breeders): “ Are you dedicated to the protection of the breed or the protection of your ego? ”

Ken Kauffman, Potomac Cairn Terrier Club Newsletter, January, 1987

- Lion Jaw , Scottie Jaw , Westie Jaw
 - Mandibles (jaws)
 - Skull bones
 - Occasionally temporomandibular joints
 - Rarely long leg bones (radius / ulna)
- Usually Bilateral & Symmetric
- Male & Female equally affected
- 4-7 months (3wks. - 11mo.)
- Misdiagnosis: teething / glands / virus

“ CMO is a disease no breeder wants to produce ! “

Carole Owen, STCA - Health Trust Fund,
CMO Research Project Chairperson

NEUTERING:

Associated with **REDUCED RISK** of manifestation of SYMPTOMATIC CMO
(& EARLIER *RECOVERY* from CMO SYMPTOMS?)

“ODDS RATIO”: **NEUTERED** vs. **INTACT** DOGS = **0.09**

SUGGESTS:

INTACT dogs (with genetically positive CMO)

ELEVEN TIMES more likely to have SYMPTOMS of CMO
than **NEUTERED** dogs

The Compendium, Small Animal, Vol.17, No.7, July 1995, CMO in Dogs,
Watson, Adams, Thomas, pp.911-922

(1958) . . .(CMO) in five West Highland White Terriers . . . thought to be neoplastic (CANCER) and the affected dogs were destroyed. Histologically, however, it was diagnosed as abnormal BENIGN osseous and chondromyxomatous (BONE & CARTILAGE) proliferation . . .

Archibald, J., and Cawley, A. J.: *The Locomotor System. Mandibular Periostitis Associated with Dysfunction of the Temporomandibular Joint.*
Hoskins, H.P., Lacrois, J.W., and Meyer, K. (Eds.): *Canine Medicine, Second Edition.* Am. Vet. Pub., Santa Barbara, Calif., 1959, p. 475

Alexander and Kallfeiz reported a case of CMO in a 6-month-old Labrador retriever in 1975. They injected the dog with ^{99m}Tc -MDP (methylene diphosphonate), which is a *bone-seeking radio-pharmaceutical*. A subsequent bone scan demonstrated that *uptake of the drug was increased significantly in the mandibular rami, indicating increased new-bone formation.*

W.Riser & C.Newton

Symptoms of CMO

- Pain during chewing or jaw manipulation
- Swollen jaws / Mouth may not open well
- Drooling
- Intermittent fever
- Lethargy & Inactivity ; “Depression”
- Lack of appetite
- Cyclical pain / fever (10-14 days)
- Severity varies
(Severe cases: Mandible fuses to Skull !)

Diagnosis of CMO

- SYMPTOMS

- BLOOD TESTS:

INCONSISTENT
OFTEN NORMAL
NOT DIAGNOSTIC

- XRAYs

“ The dog may need to be anesthetized or sedated to obtain proper positioning and radiographic detail “

CMO, The Basics of a Painful Disease
2002, Carole Fry Owen

MacDuffie (Maureen Drummond)

"It's a horrifying scream when you touch the jaw"

. . . noticed him drinking water very awkwardly. He was trying to sip through the side of his mouth. His jaw was not opening normally

. . . MacDuffie screaming in pain when Drummond touched his head . . .

"TAD...A STUDY IN CMO"

Nancy Ann Schoch

January, 1992, one of a litter of three (Westies) - a repeat breeding which had produced lovely pups the year before. . .

Tad began having 'episodes.' He would be fine one day and the next - listless, ouch-y, and have difficulty eating.

His eyes looked a bit strange, almost as though he had a severe headache. His temperature was 103.8.

He would spend much of his day in his crate or under a table, shifting frequently to find a comfortable position. By now his entire lower jaw was swollen to three times its normal size.

. . .could not stand or walk and moaned in pain when I lifted him to change his bedding. He could not eat. (continued)

"TAD...A STUDY IN CMO" (continued)

Nancy Ann Schoch

. . .devastating news that Tad's sister had just been diagnosed positive with CMO.

Tad's brother had just been diagnosed CMO-positive, after nearly three weeks of exhaustive and expensive testing by veterinarians who had never heard of the disease.

CMO may be far more widespread than most of us realize. There are sub-clinical cases which do not product the severe manifestations obvious with Tad. Radiographs at the appropriate age are required to determine that a puppy does not have the disease.

Treatment of CMO

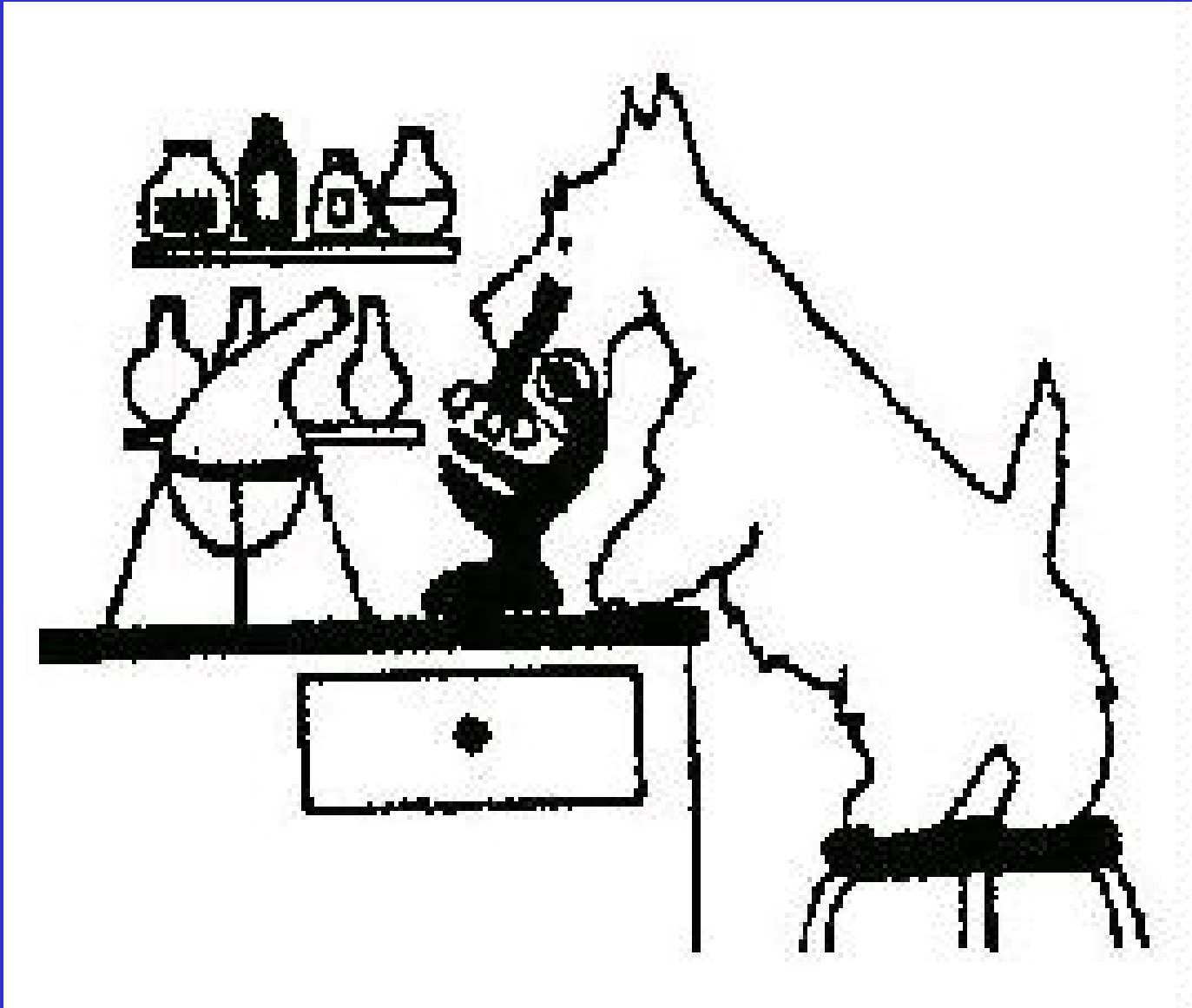
- **SYMPTOM RELIEF** (*disease spontaneously resolves*)
 - **NSAIDS** (*CAUTION / gastric ulcers*) :
buffered or enteric-coated aspirin, caroprofen, etodolac, phenylbutazone, meclofenamic acid, and piroxicam
 - **STEROIDS** (prednisone / prednisolone)
 - **PAIN RELIEVERS** (tylenol with CODEINE)
- **SOFT DIET** (high calorie, protein rich, gruel)
- **ACTIVITY RESTRICTIONS**

Side Effects of Corticosteroids

Systemic side effects generally dependent on dose and duration of treatment. Short-term use is unlikely to cause adverse effects. Adverse effects are more common in animals on high and/or long term doses.

- * POLYURIA(excessive urination)
- * POLYDIPSIA(excessive thirst/drinking)
- * POLYPHAGIA(excessive appetite/eating)
- POOR HAIRCOAT
- GI DISTURBANCE (diarrhea, vomiting, weight gain, ulcers)
- PANCREATITIS ; ELEVATED LIVER ENZYMES
- DIABETES MELLITUS
- MUSCLE WASTING
- BEHAVIORAL CHANGES

* may be seen in dogs even on short-term therapy



SCIENTIFIC RESEARCH

Tumor-like Exostoses on the Bones of Head in Puppies
Littlewort, M.: Vet. Rec. 70:977 (1958)

Archibald, J., and Cawley, A. J.: *The Locomotor System. Mandibular Periostitis Associated with Dysfunction of the Temporomandibular Joint.* Hoskins, H.P., Lacrois, J.W., and Meyer, K. (Eds.): *Canine Medicine, Second Edition.* Am. Vet. Pub., Santa Barbara, Calif., p. 475, (1959)

CANINE CRANIOMANDIBULAR OSTEOPATHY

W. Riser, DVM, L. Parkes, DVM, & J. Shirer, BS, MA
J. Amer. Vet. Rad. Soc., Vol., 8, (1967)

ANIMAL MODEL: THE MODE OF INHERITANCE OF CRANIOMANDIBULAR OSTEOPATHY IN WEST HIGHLAND WHITE TERRIER DOGS

George A. Padgett and Ulreh V. Mostosky,
Dept. Pathology, College Vet. Med., MSU (1986)

Control of Canine Genetic Diseases
George Padgett, DVM (1998)

George Padgett, DVM (MSU/emeritis)
& Barbara Stoll (Westie breeder)

- Autosomal Recessive Trait
- Both parents of an CMO affected puppy are CARRIERS
 - Two carriers bred together:
 - 25% affected / afflicted with CMO
 - 50% carriers
 - 25% non-carriers
- Siblings of affected puppies: 66 $\frac{2}{3}$ % carriers
- Offspring of known carriers: 50% carriers

Affected dogs should be bred only in test matings to "clear" breeding stock. Breed known carriers only with great caution (preferably AVOID)

- Barbara J. Stoll
- West Highland White Terrier Club of S.E. Michigan
- Compiled data from 12 different breeders who had produced one or more CMO puppies over a time span covering 1965 through 1982
- Purchased a CMO affected female; club raised until she was of breeding age; subsequently bred to a CMO affected male, and in the spring of 1983 she whelped a litter of four, two males and two females, all of whom were verified as affected by Dr. Ulreh Mostosky, head of the Radiology Department at Michigan State University
- Test breeding confirmed that the mode of inheritance of CMO is an autosomal recessive genetic trait

□ MALE

○ FEMALE

○ SEX UNKNOWN

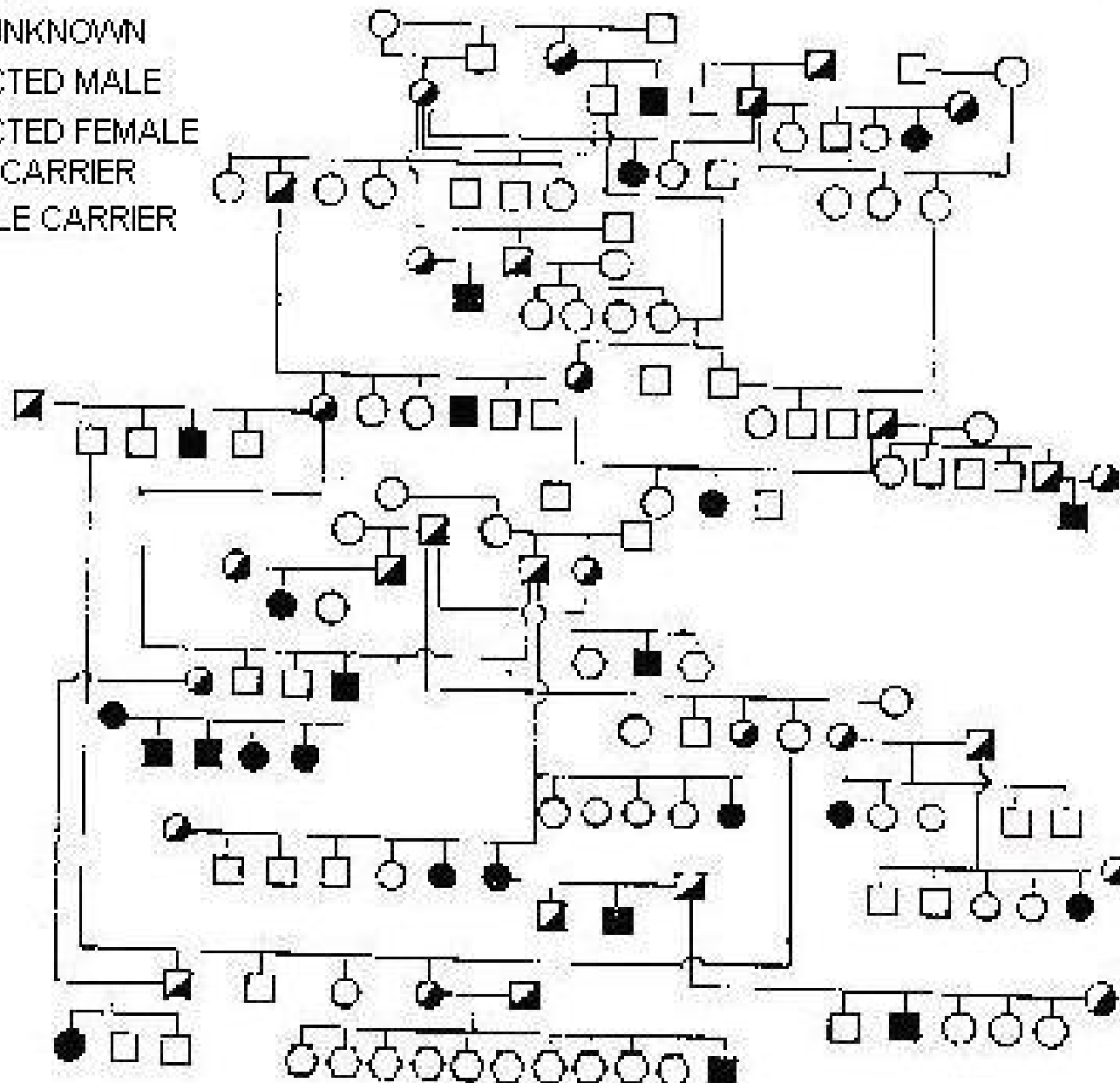
■ AFFECTED MALE

● AFFECTED FEMALE

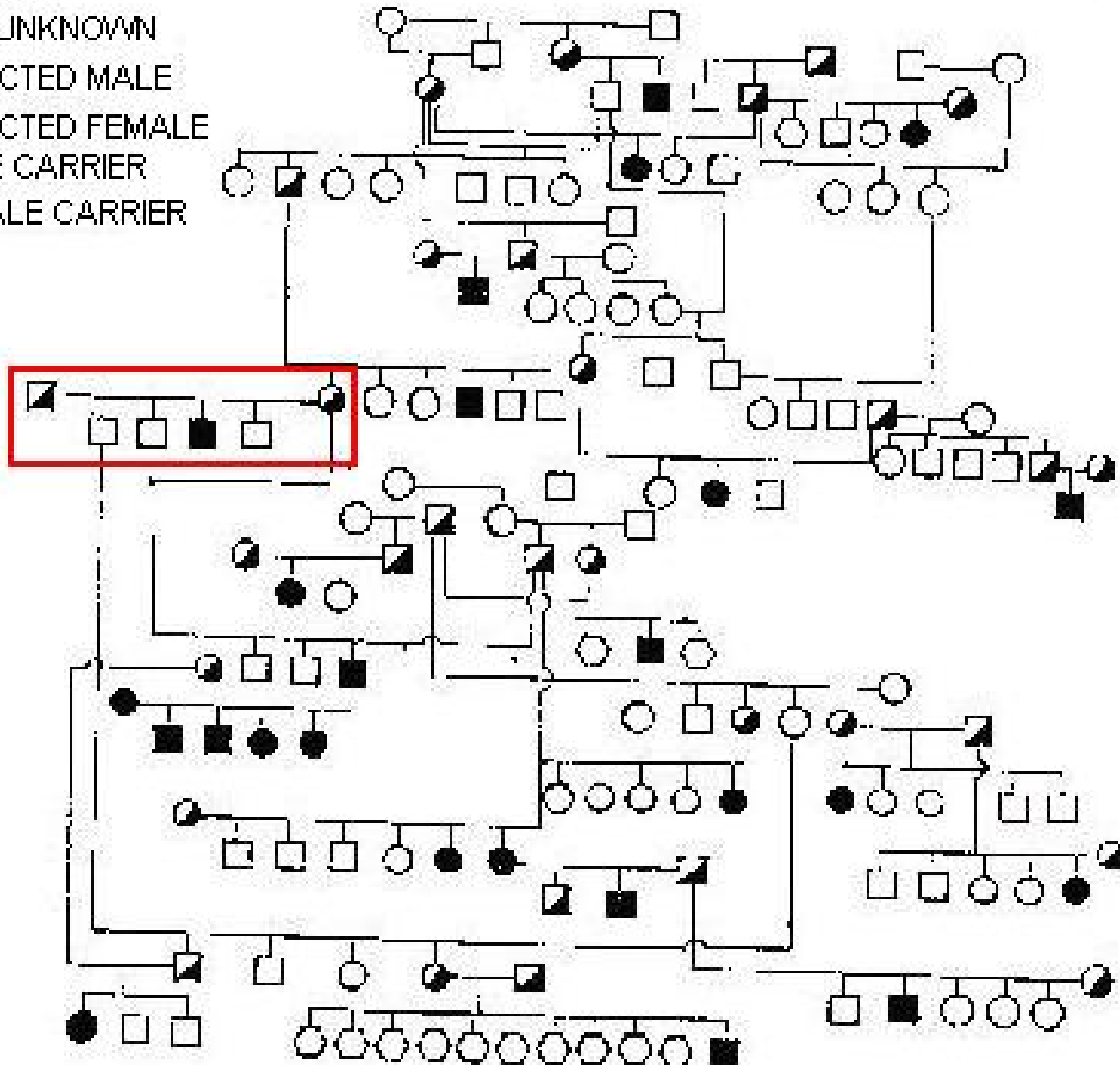
◻ MALE CARRIER

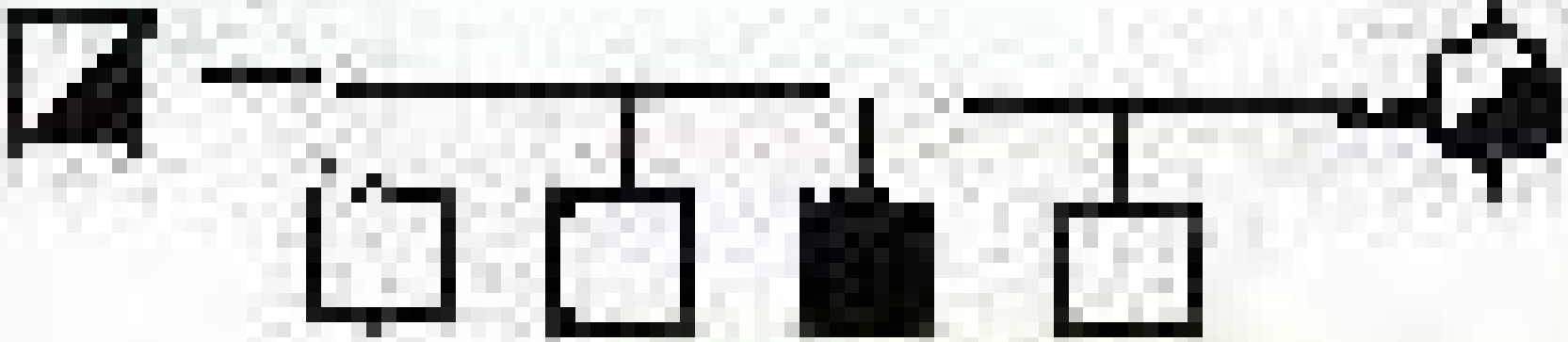
◐ FEMALE CARRIER

CMO in a Westie pedigree (Barbara J. Stoll)

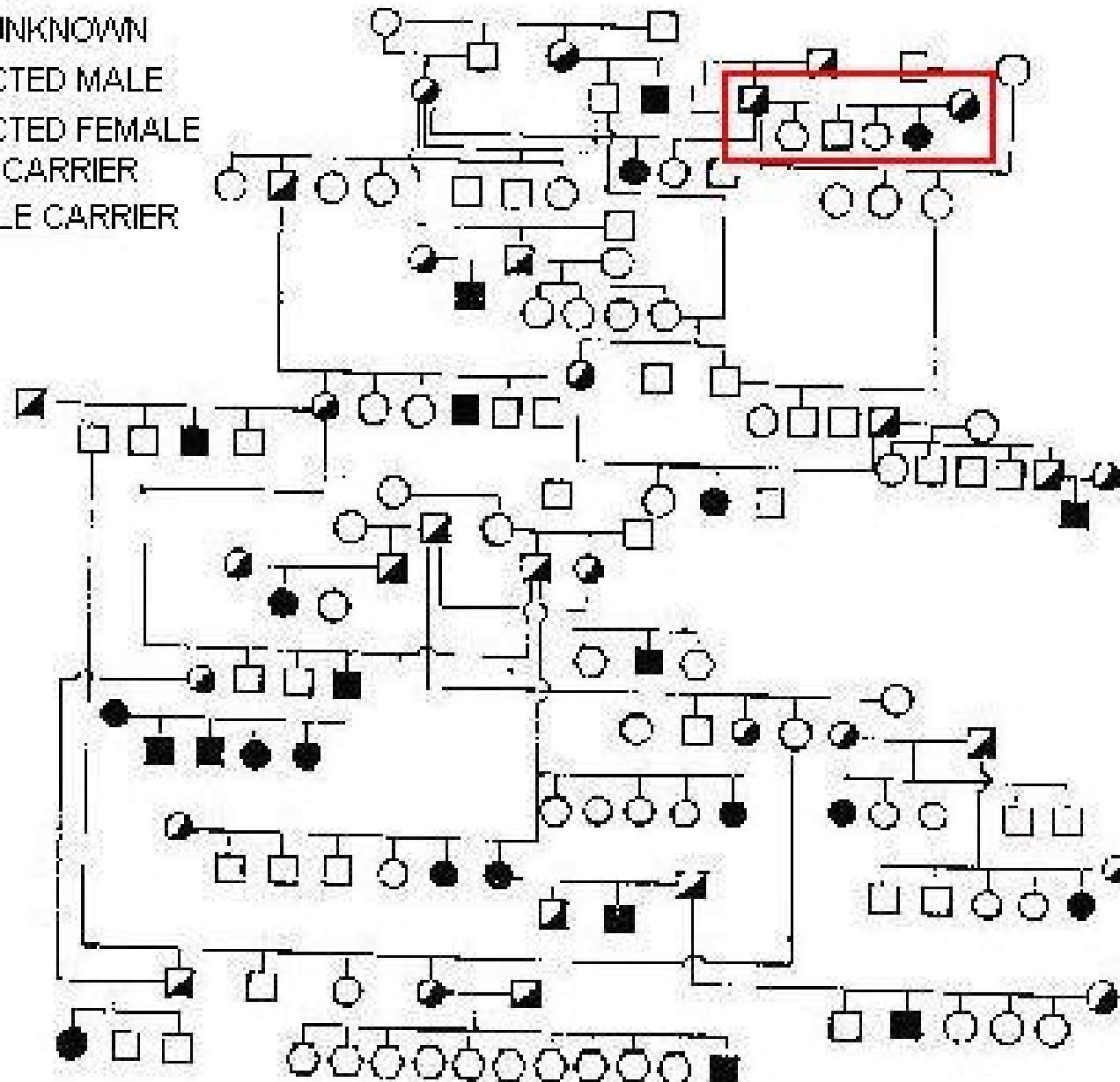


- MALE
- FEMALE
- SEX UNKNOWN
- AFFECTED MALE
- AFFECTED FEMALE
- ◻ MALE CARRIER
- ◐ FEMALE CARRIER



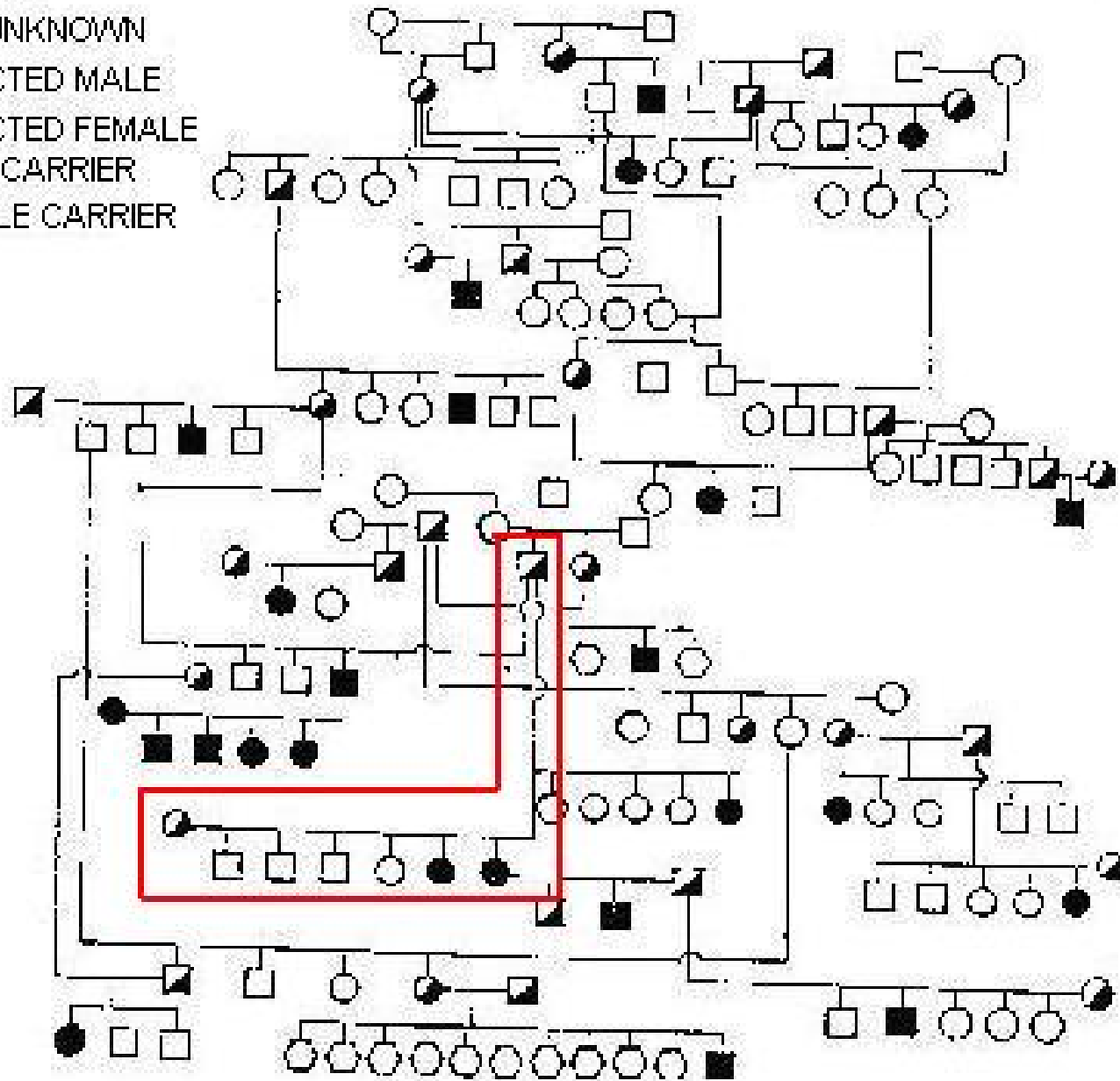


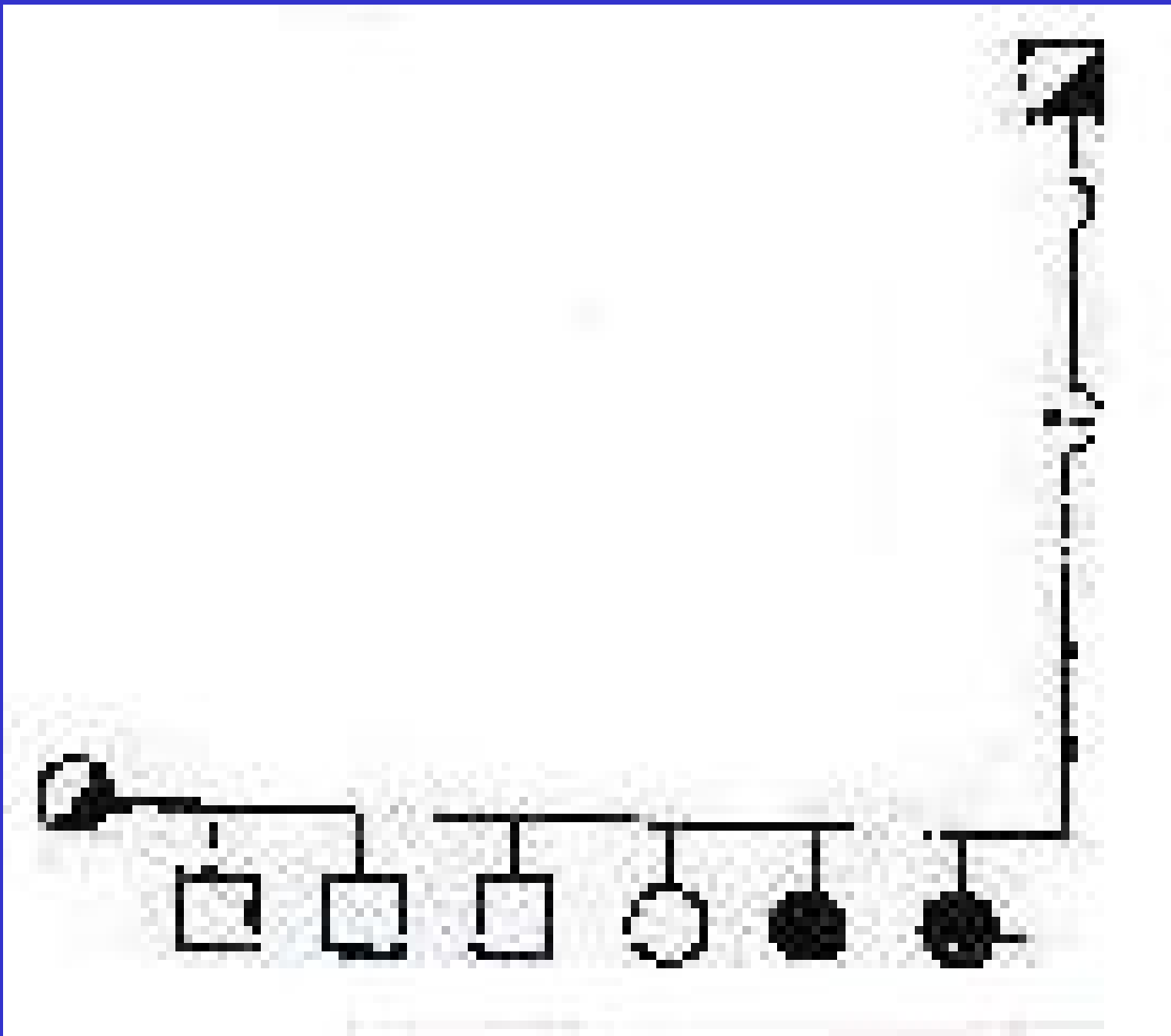
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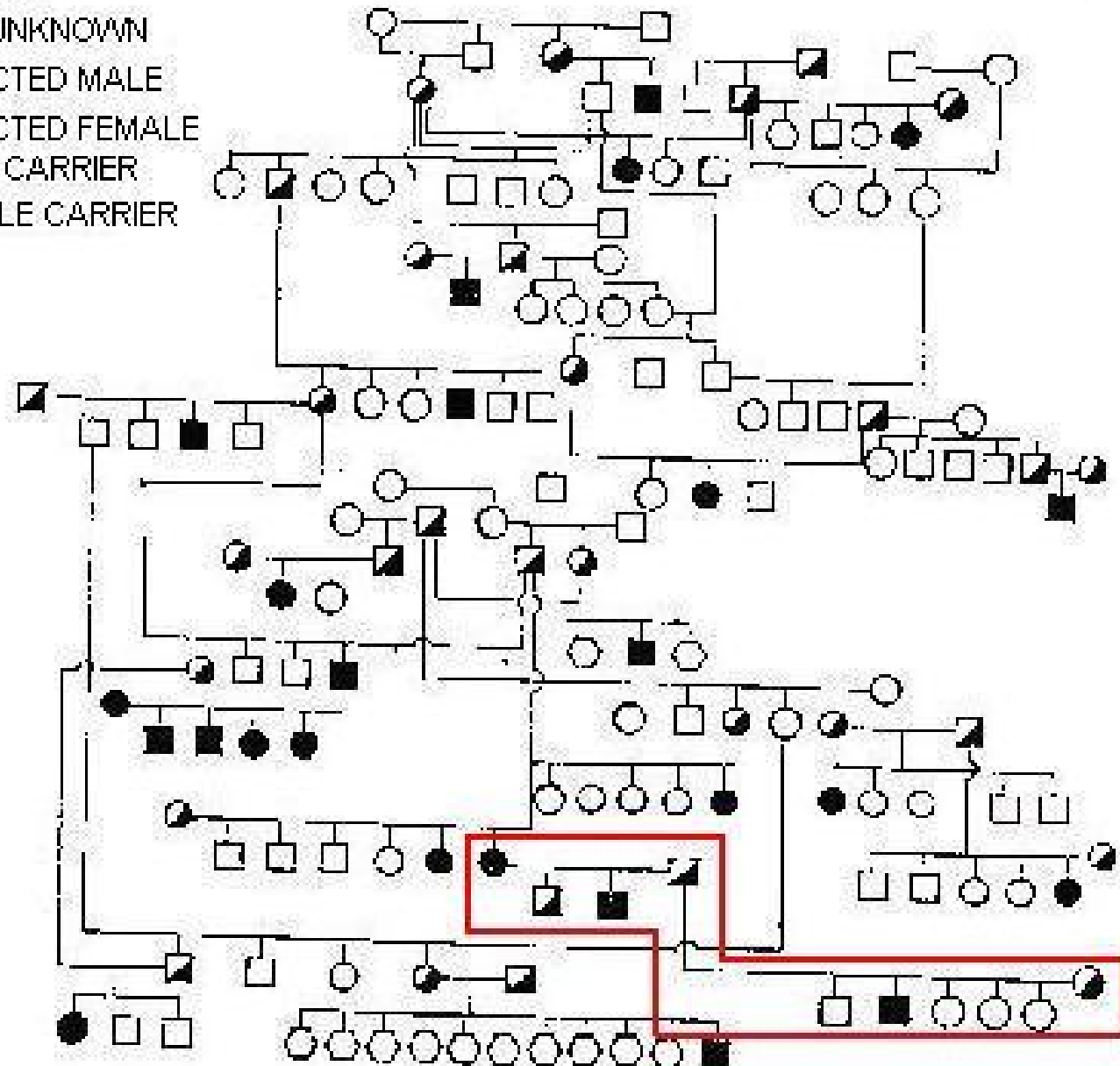


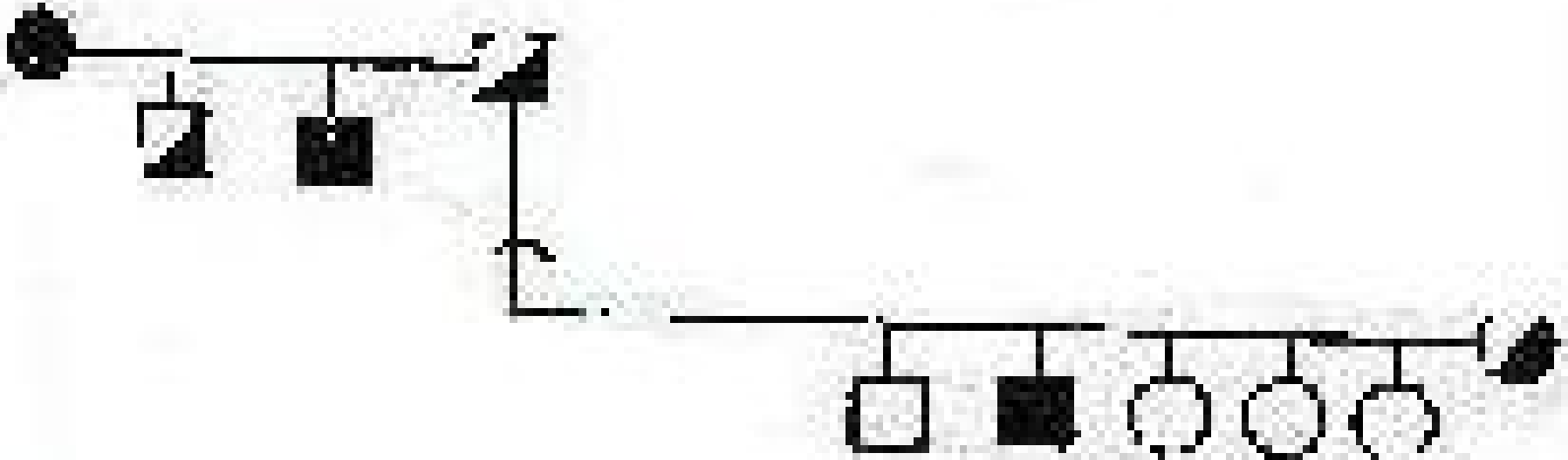
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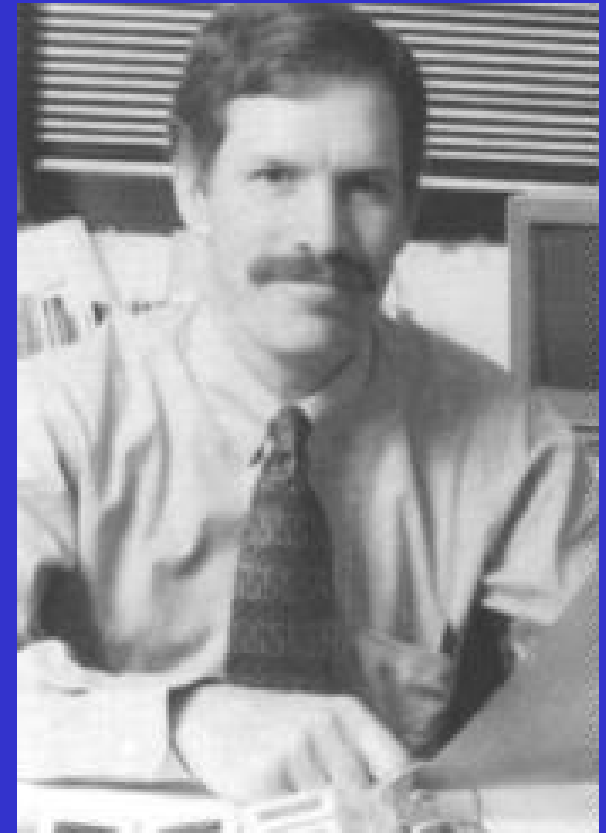




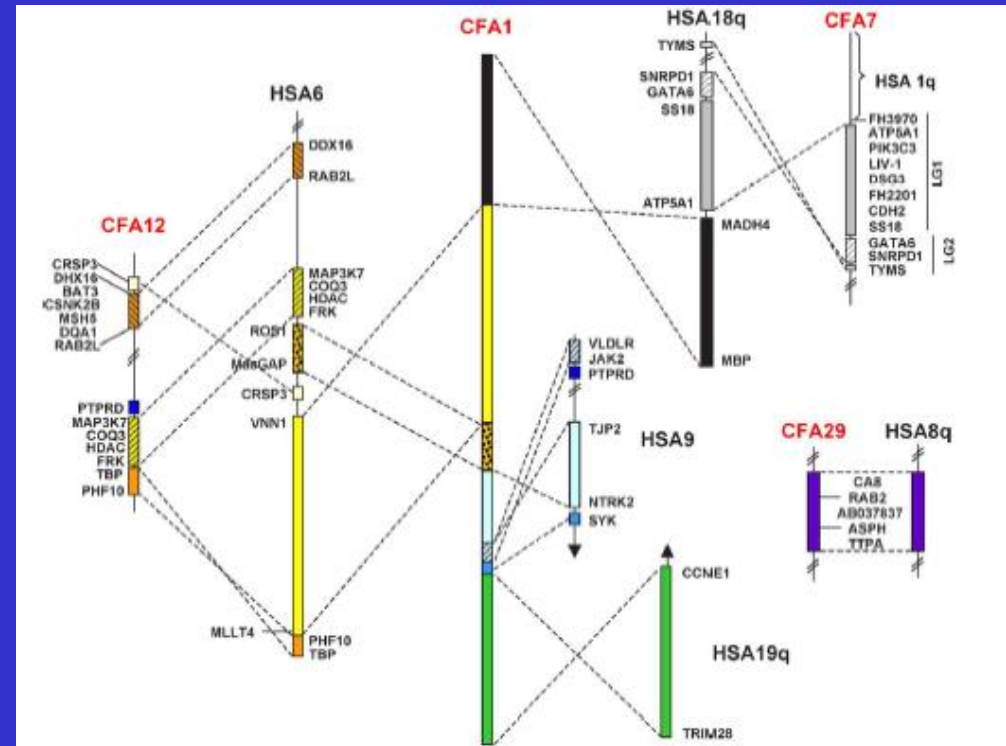
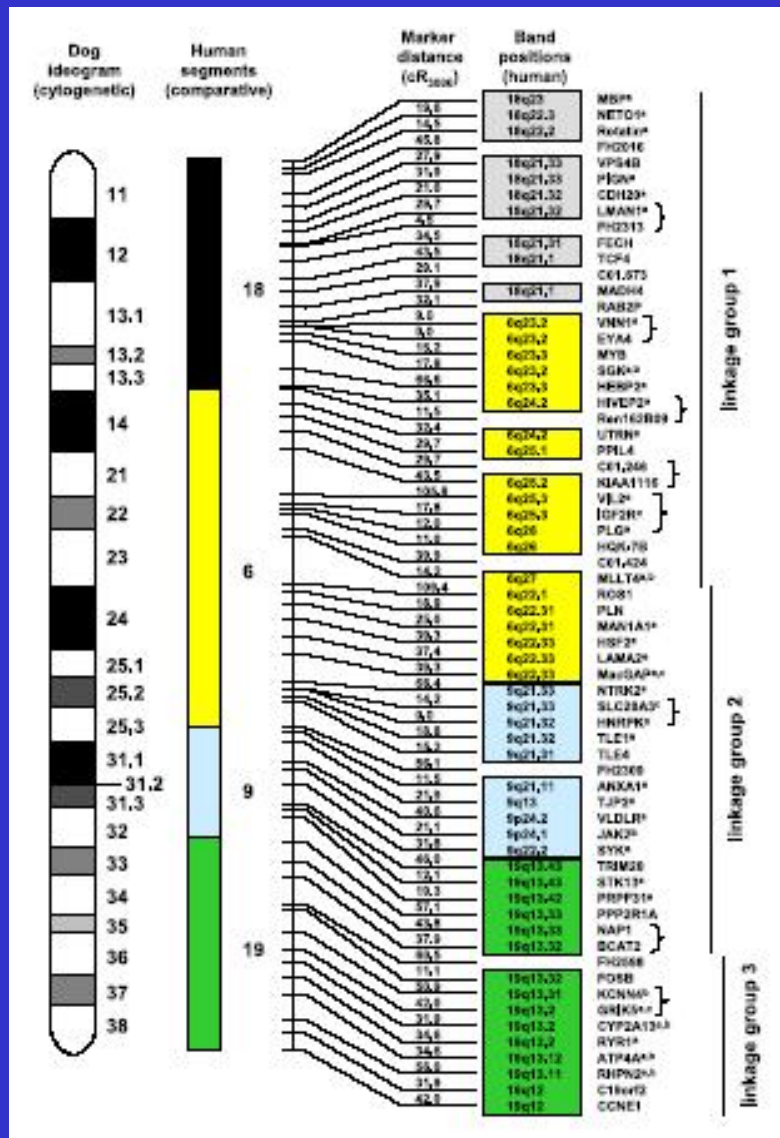
Patrick J. Venta, Ph.D., Associate Professor

Michigan State University, College of Veterinary Medicine

CMO occurs at a greater than average rate in the Scottish Terrier and West Highland White Terrier breeds. It is somewhat less prevalent in the Cairn terrier and Bull terrier breeds, and it is occasionally seen in many other breeds. It causes a non-cancerous proliferation of the bone, primarily in the region of the jaw. Animals experience tenderness of the jaw and can have difficulty eating. Although the disease often disappears after a year or so, it is a miserable time for both the dog and owner when it is active. Dr. Venta is in the early stages of research to develop a carrier test for this disease. He has collected a sufficient number of pedigrees in which the disease has been seen to begin the work.

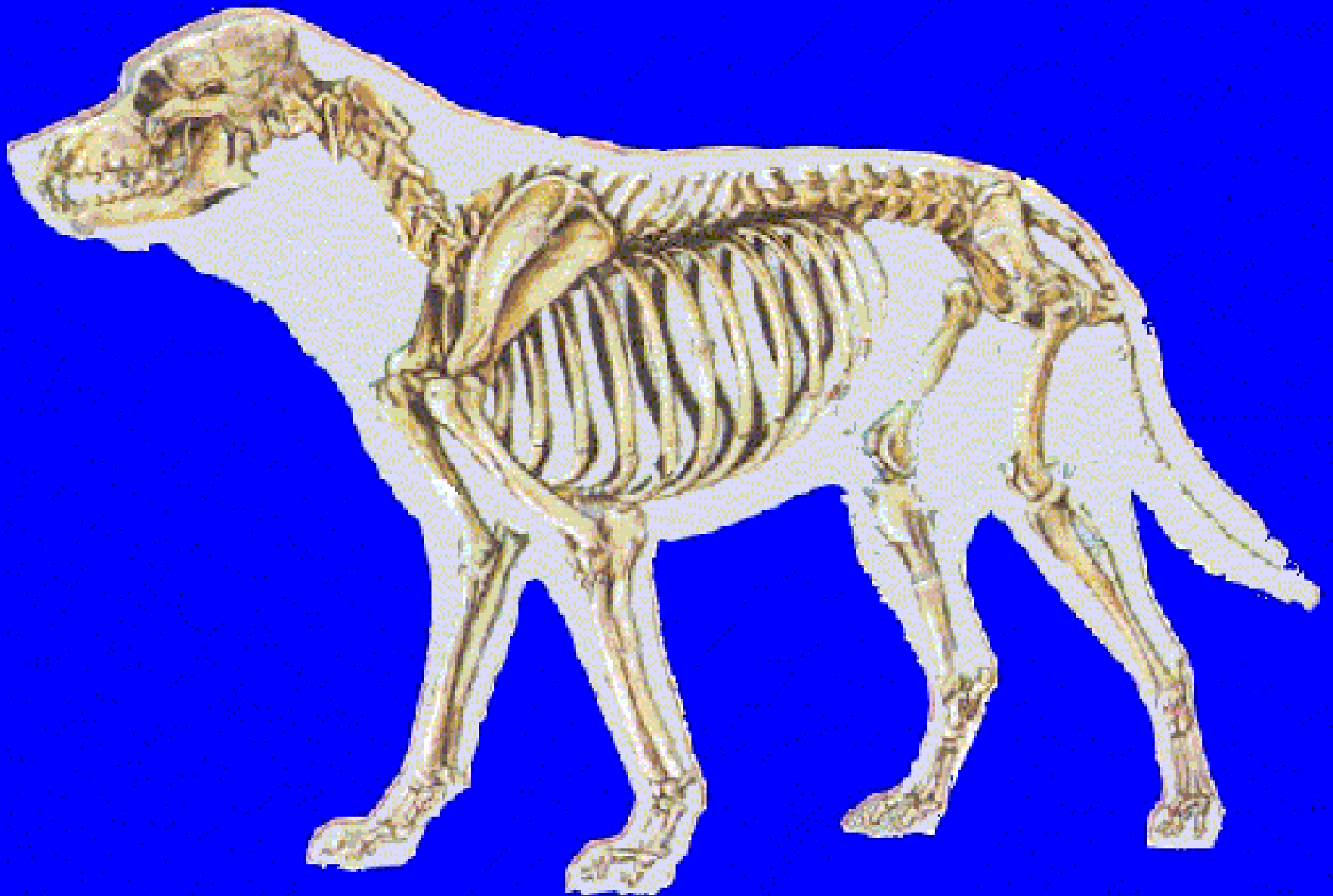


Canine Chromosome # 1

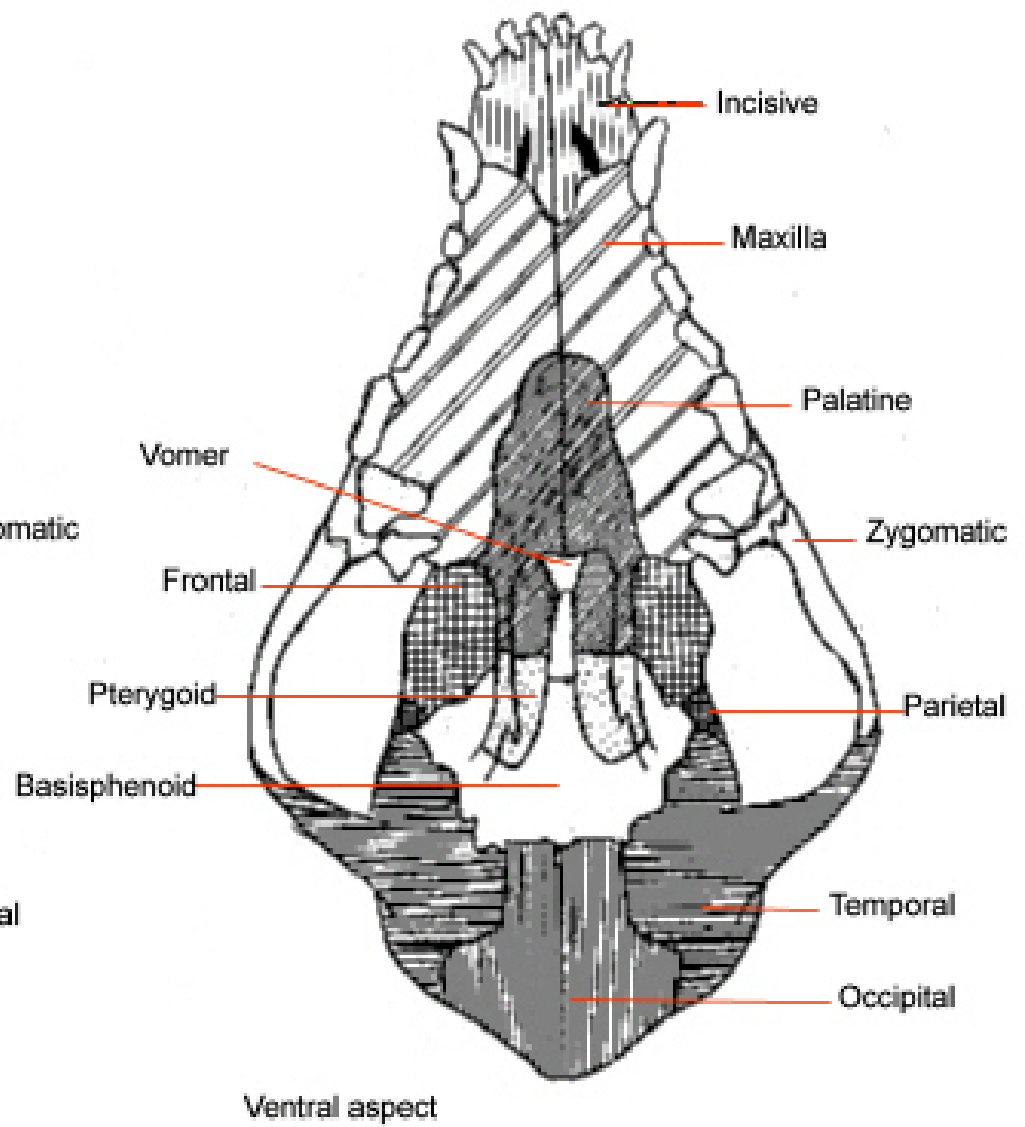
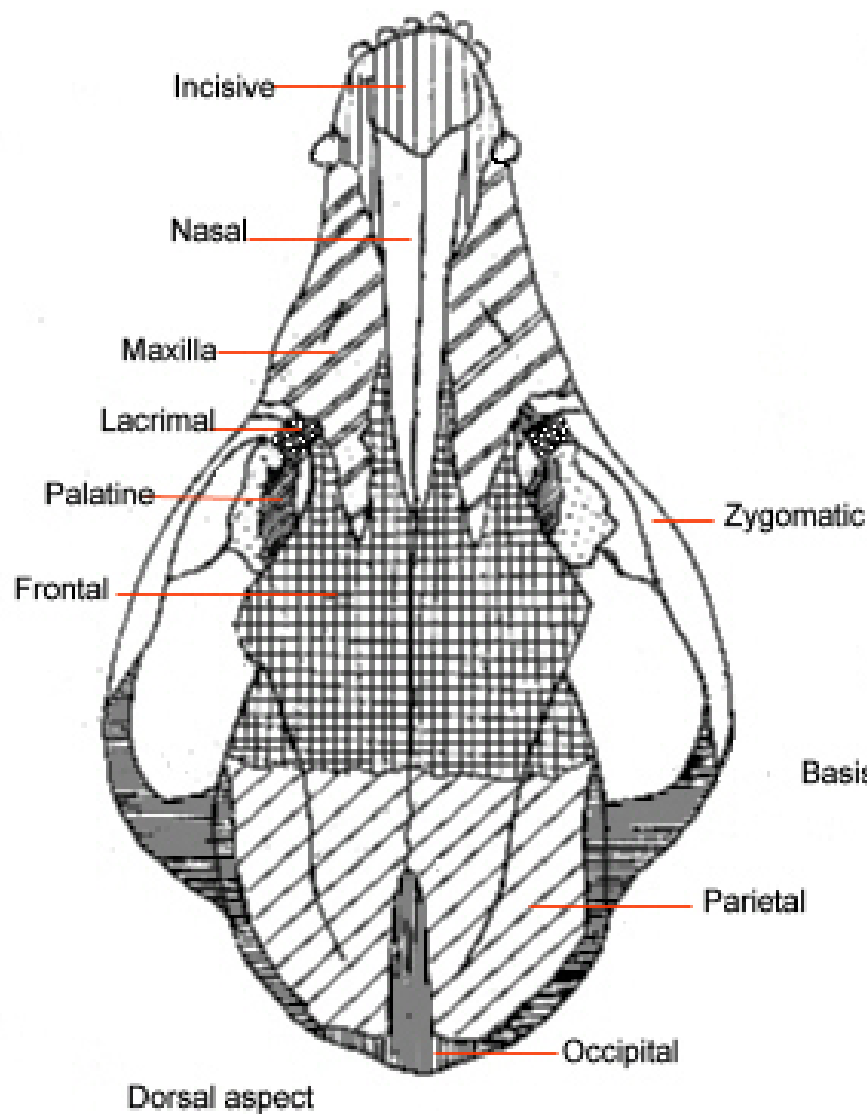


Comparative radiation hybrid map of canine chromosome 1 incorporating SNP and indel polymorphisms, Housley, Ritzert, & Venta, *Genomics*,(2004),84:248-264

- 1998: **AKC-Canine Health Foundation** approved Dr. Venta's research proposal to find *genetic linkage marker for CMO*
- **GOAL**: Develop **SCREENING TEST for CMO**
- STCA-Health Trust Fund+ WFA + CTCA + AKC/CHF each funded \$10,000
- 2000: Third Year of research funding approved ; 2004 new grant proposal
- **GOAL SWITCHED**: from *LINKAGE MARKER* (85-90% accurate) to *DIRECT GENETIC MARKER* (100% accurate)
- 2004: Completion of Canine Genome Map – improves likelihood of finding chromosomal locations & developing **GENETIC MARKERS** for **CMO** & other canine genetic diseases
- Development of **AUTOMATED GENOTYPING TOOL** – much faster & less expensive than labor-intensive gel-based method
- **SNP** – Single Nucleotide Polymorphism: New type of genetic marker improves likelihood of success of finding direct genetic marker for CMO



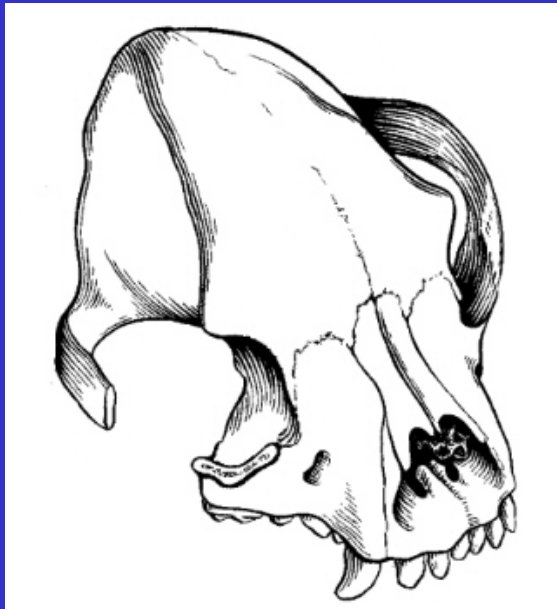
Dog Skeletal Anatomy



Dog Skull Anatomy



NORMAL DOG SKULL

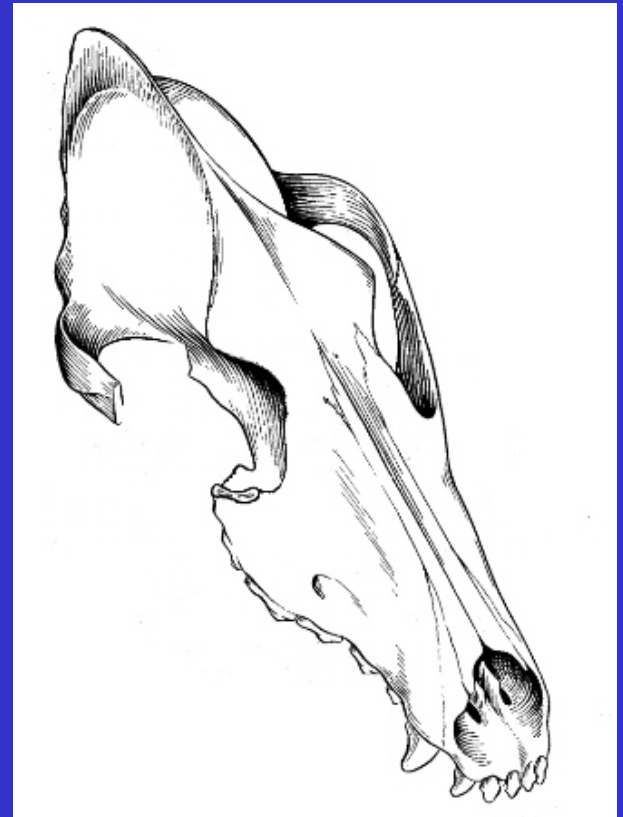


BRACHYCEPHALIC

Short and Wide
Boston Terrier,
Pekingese,
Bulldog

DOLICHOCEPHALIC

Long and Narrow
Collie, Sheltie,
Russian Wolf Hound



MESATICEPHALIC

Medium Proportion
German Shepherd,
Lab. Retriever,
SCOTTISH TERRIER





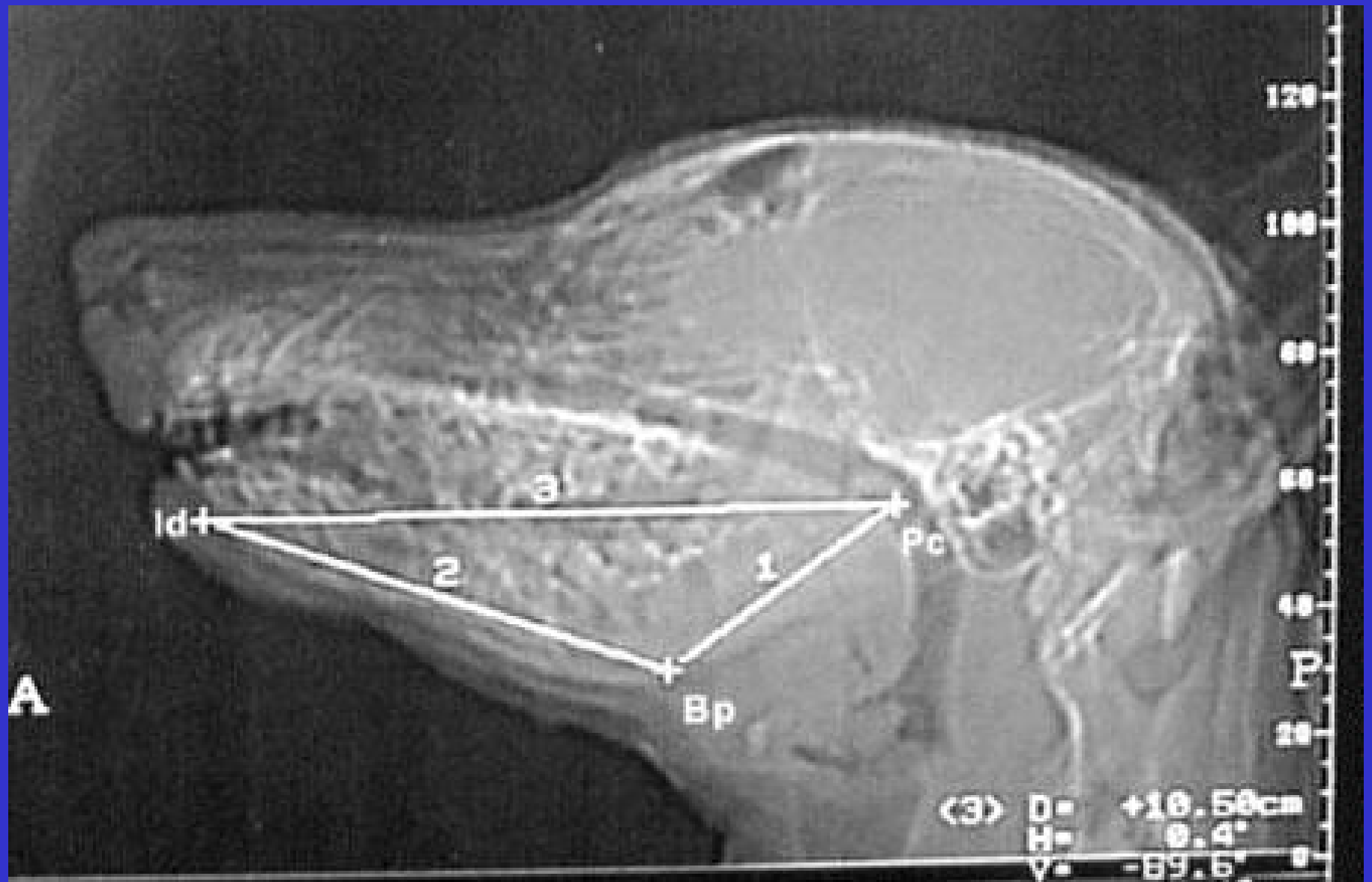


Angus "The Verminator" – STCC Howl-O-Ween 2004





NORMAL DOG SKULL



XRAY of NORMAL DOG SKULL with BioMetric Markings

XRAY OF
NORMAL
ADULT DOG
SKULL
(Chow Chow)

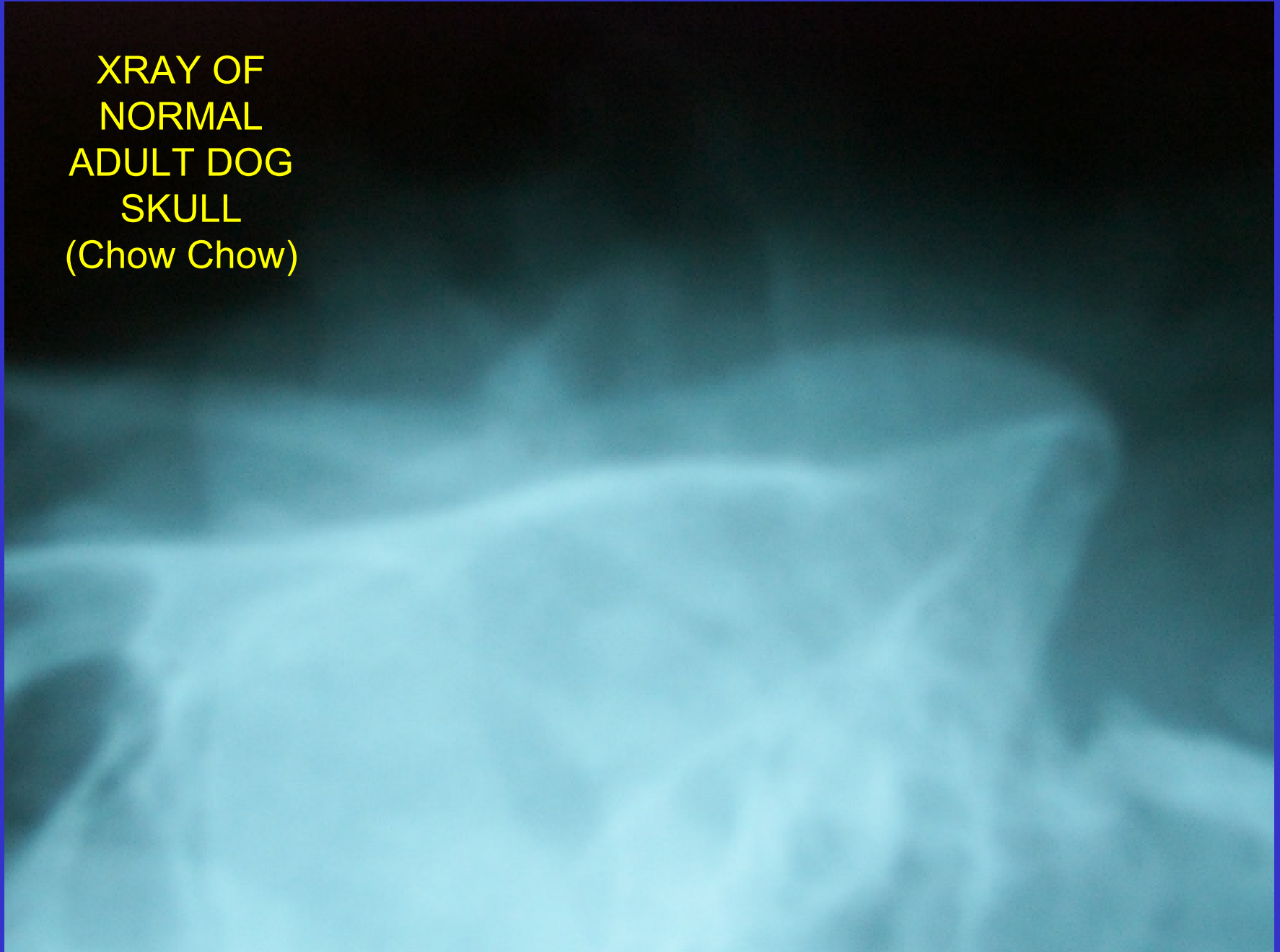


Courtesy of Sheree Block,DVM, Buffalo Grove, IL

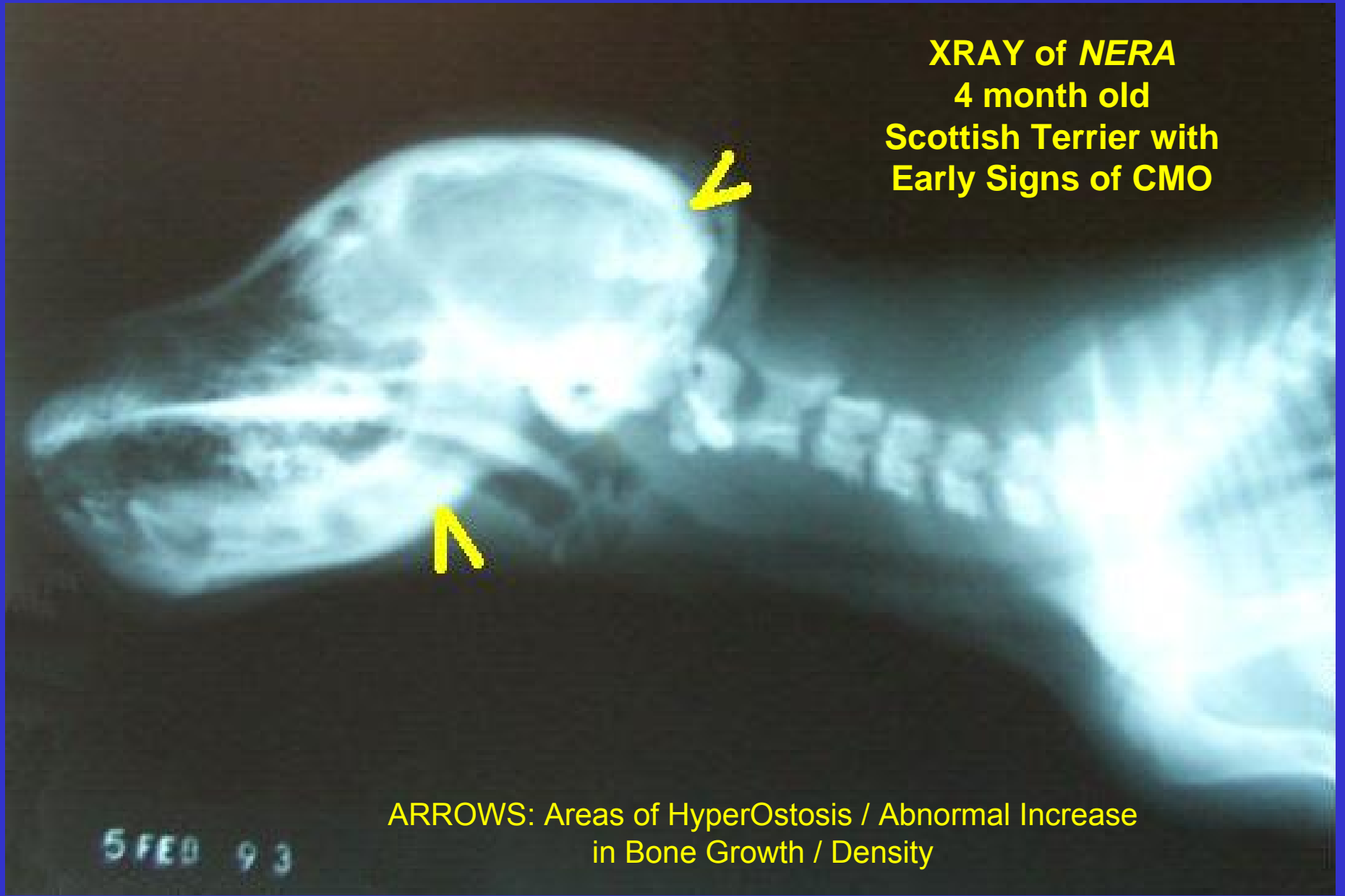


XRAY OF
NORMAL
ADULT DOG
SKULL
(Chow Chow)

XRAY OF
NORMAL
ADULT DOG
SKULL
(Chow Chow)



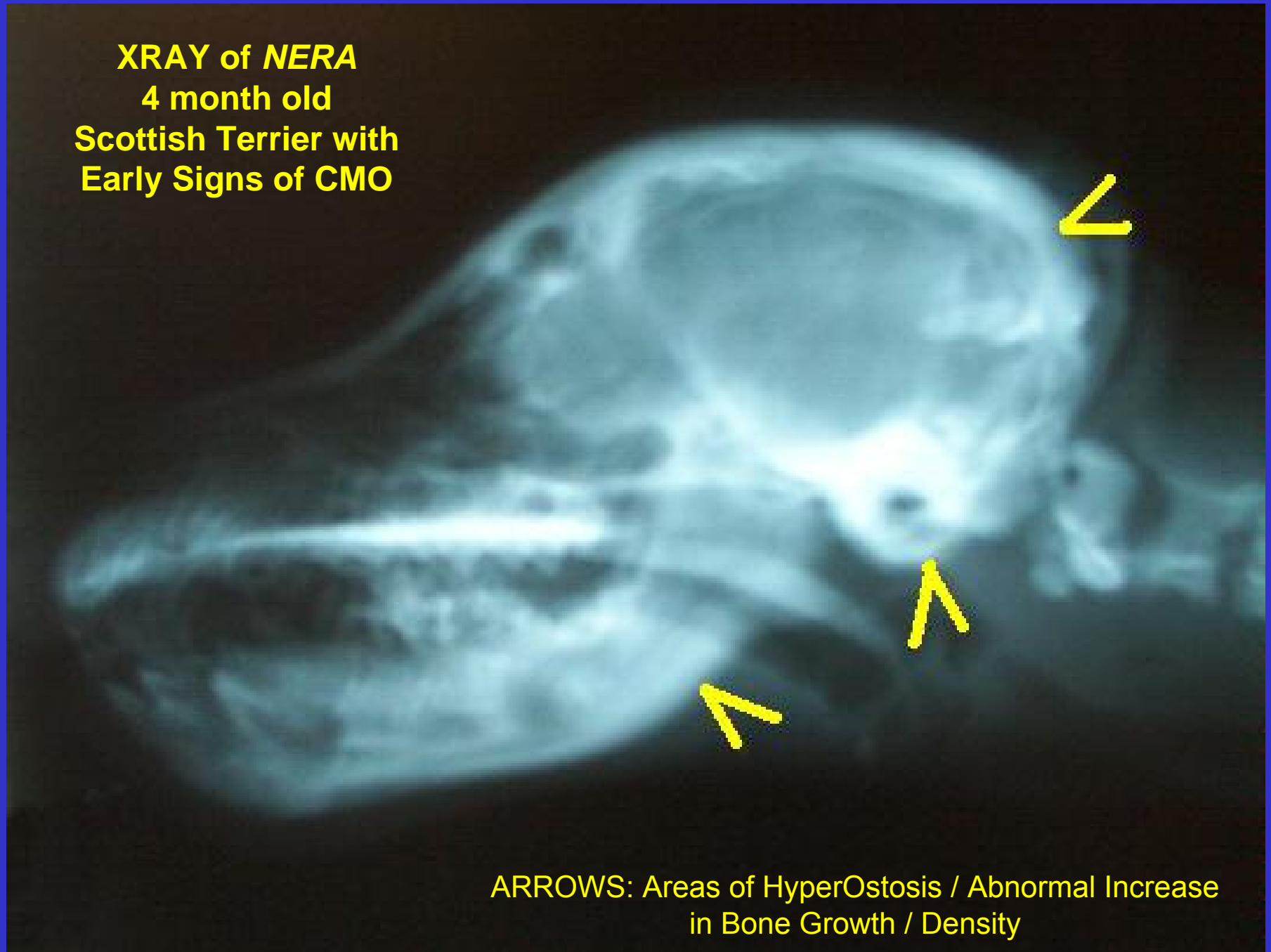
**XRAY of NERA
4 month old
Scottish Terrier with
Early Signs of CMO**



**ARROWS: Areas of HyperOstosis / Abnormal Increase
in Bone Growth / Density**

Courtesy of Sheree Block,DVM, Buffalo Grove, IL

XRAY of NERA
4 month old
Scottish Terrier with
Early Signs of CMO



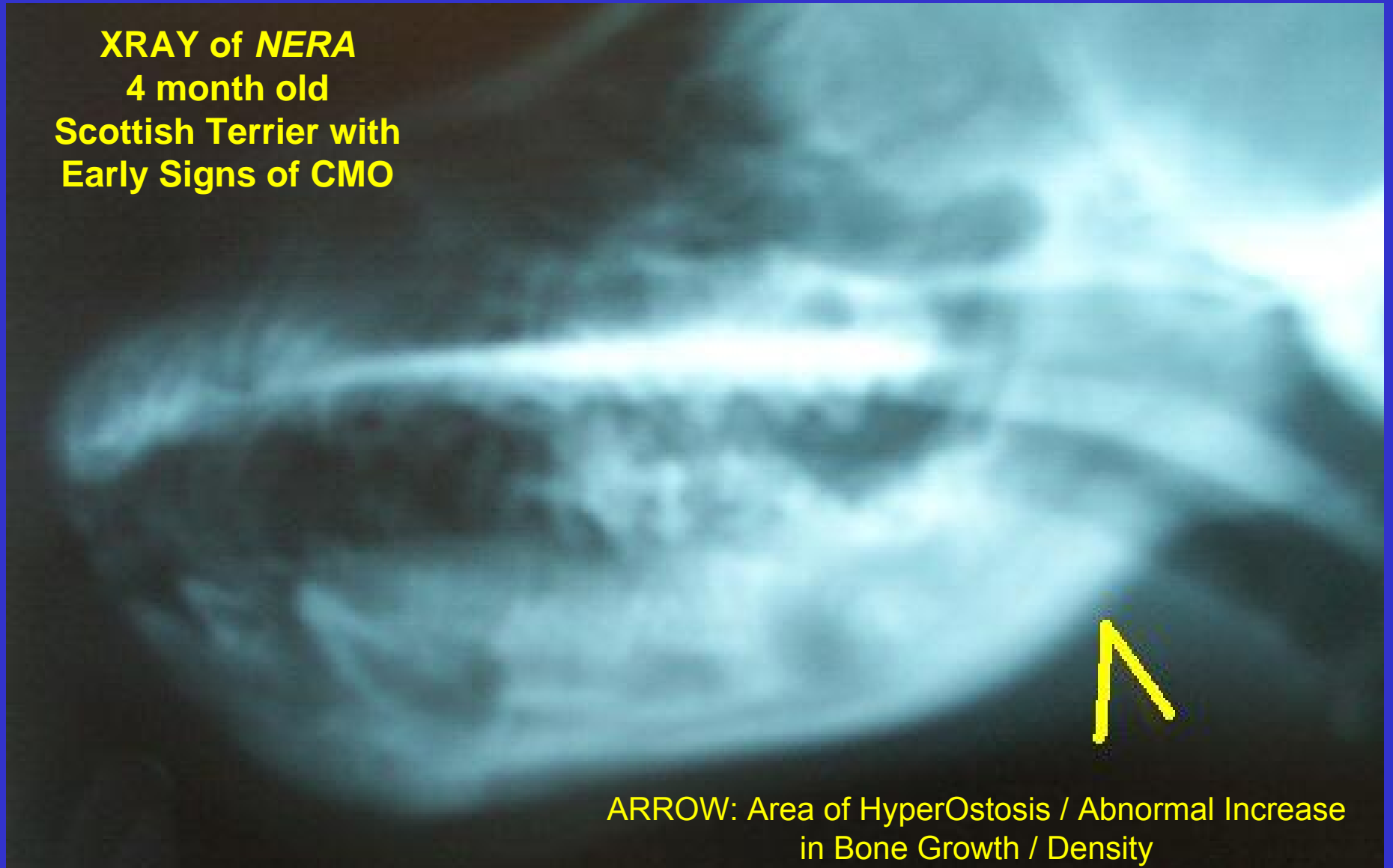
ARROWS: Areas of HyperOstosis / Abnormal Increase
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XRAY of NERA
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ARROWS: Areas of HyperOstosis / Abnormal Increase
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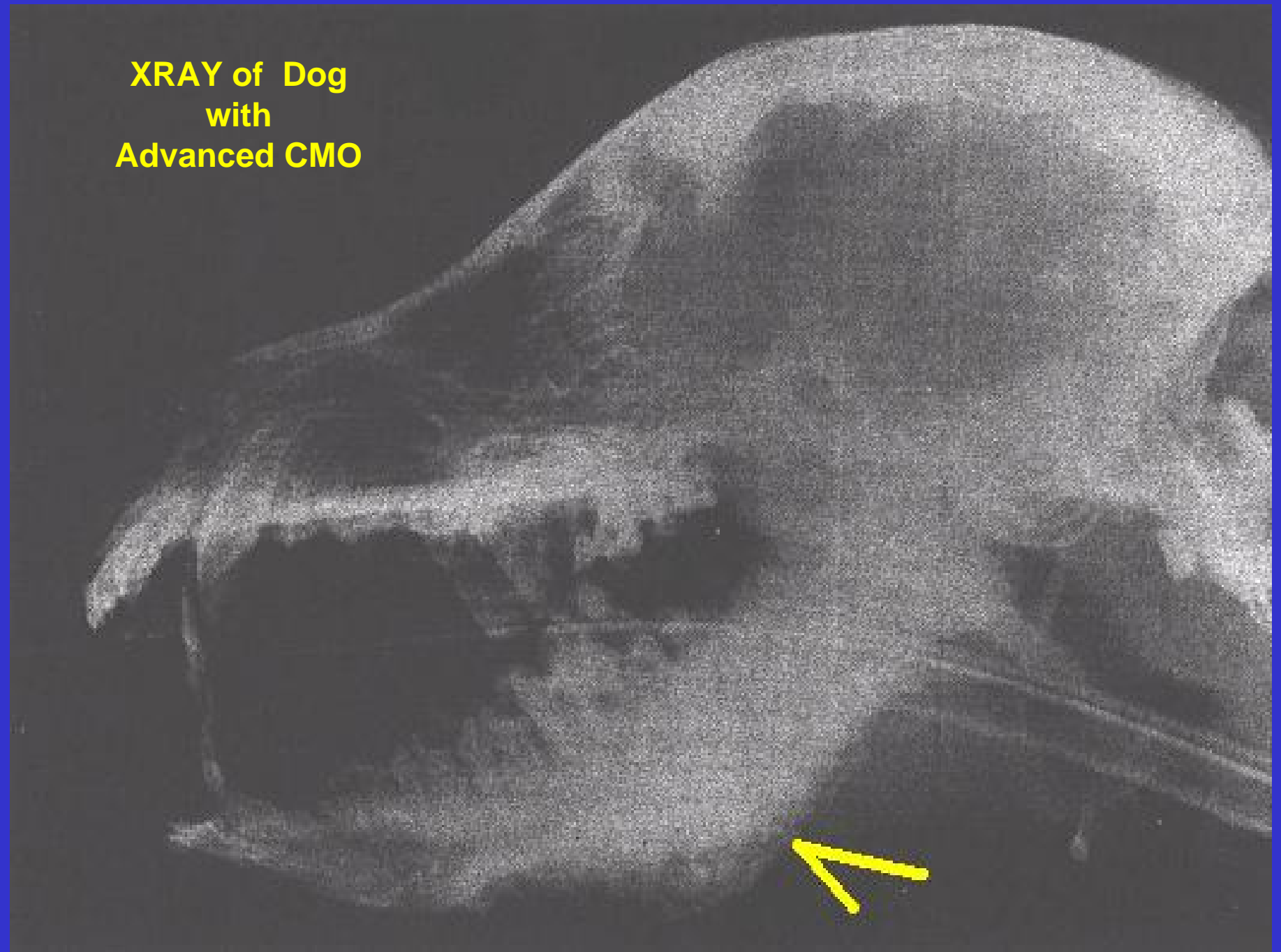


XRAY of *NERA*
4 month old
Scottish Terrier with
Early Signs of CMO

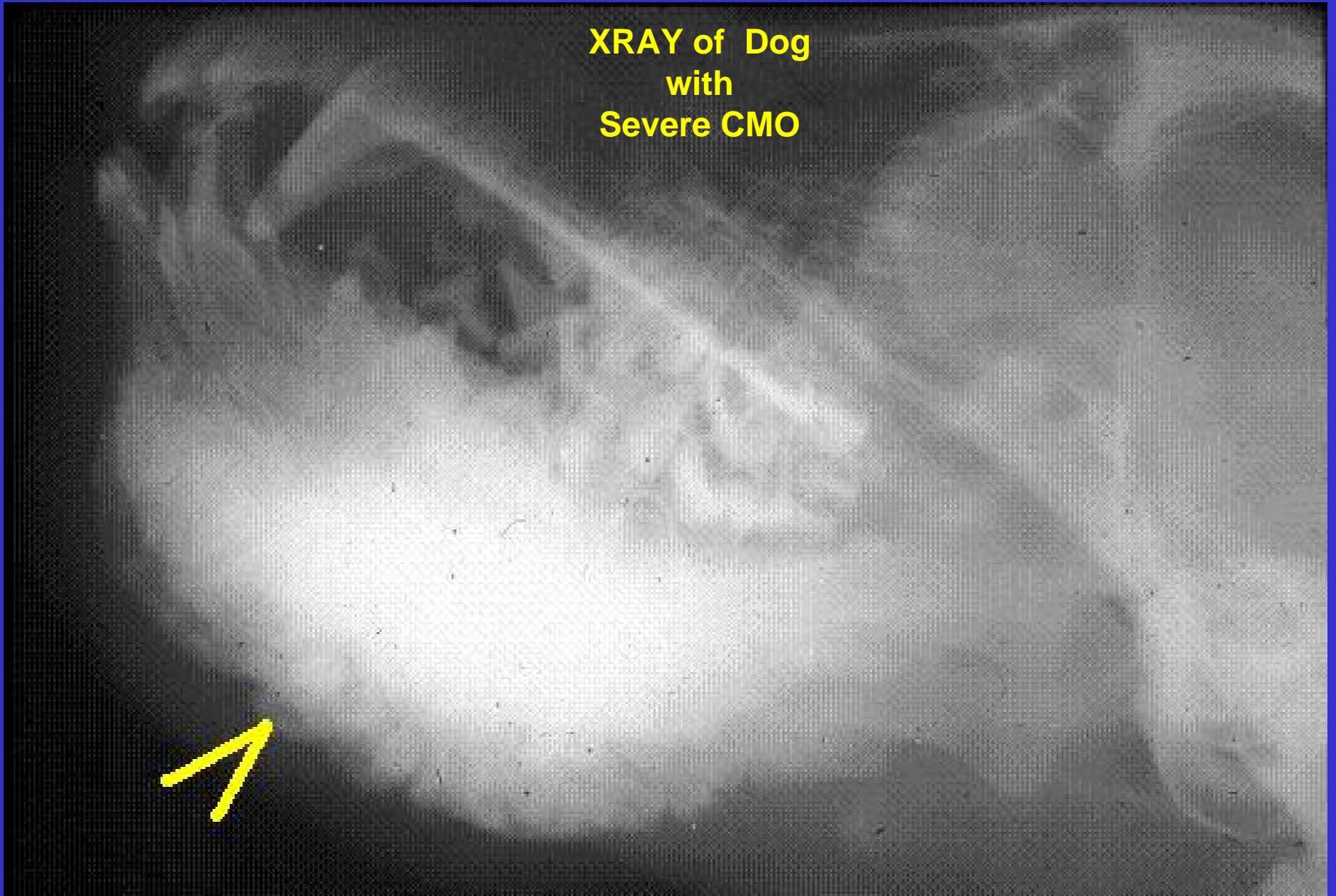


**ARROW: Area of HyperOstosis / Abnormal Increase
in Bone Growth / Density**

**XRAY of Dog
with
Advanced CMO**



**XRAY of Dog
with
Severe CMO**

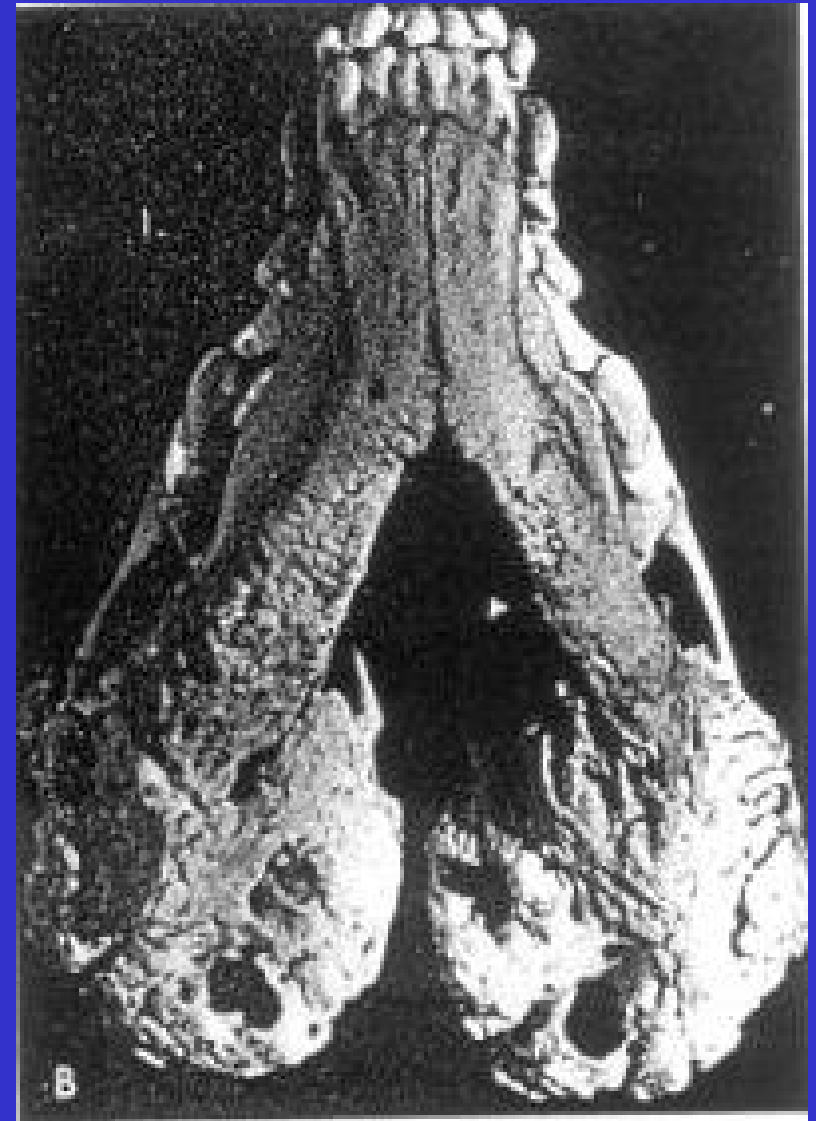


Courtesy of D. Rosenstein, DVM (MSU Vet. Radiology) from U. Mostosky, DVM (MSU)



Dog skull from an extremely severe case of CMO

From Leonard's Orthopedic Surgery of the Dog and Cat, 3rd ed.



HUMAN ANALOGUE?: CAFFEY'S DISEASE

(John Caffey, 1895–1978, American pediatrician), also called *infantile cortical hyperostosis*, a disease of infants.

Typically the affected infant is less than 5 months old and exhibits fever of abrupt onset, hyperirritability and soft tissue swelling, particularly over the mandible. The most prominent feature of the disease, cortical hyperostosis, may lead to doubling or tripling of the normal width of the bone. Sometimes, all evidence of hyperostosis can be lacking on follow-up examinations after 6 months to 1 year. Although the cause of Caffey's disease remains unknown, an infectious agent or a genetic factor (possibly with an autosomal dominant pattern of inheritance) may play a role in the pathogenesis of the condition.

CranioMandibular Osteopathy - “ CMO “

SUMMARY:

- VERY PAINFUL PUPPY (JAW & SKULL)
- TREATMENTS CAN ALLEVIATE PAIN
- COMPLETELY RESOLVES SPONTANEOUSLY
- NO RESIDUAL DEFORMITY OR IMPAIRMENT
- EUTHANIZED IF NOT PROPERLY RECOGNIZED
- INHERITED / RECESSIVE (NEUTER ! / SPAY !)
- RESEARCH HORIZON: DNA SCREENING TEST

“ THANK YOU ”

- Sheree A. Block, DVM
- Diana Rosenstein, DVM
- Marcia Dawson, DVM
- Patrick J. Venta, PhD
- George Padgett, DVM & Barbara Stoll
- Carole Owen
- Gail Gaines & Susan Morris

- Caryl Alten
- Sue Severtsen



NERA
1992 - 2004



Scottish Terrier

HAIKU

Sleeping here, my chin
on your foot - no greater bliss;
Except chasing squirrels.