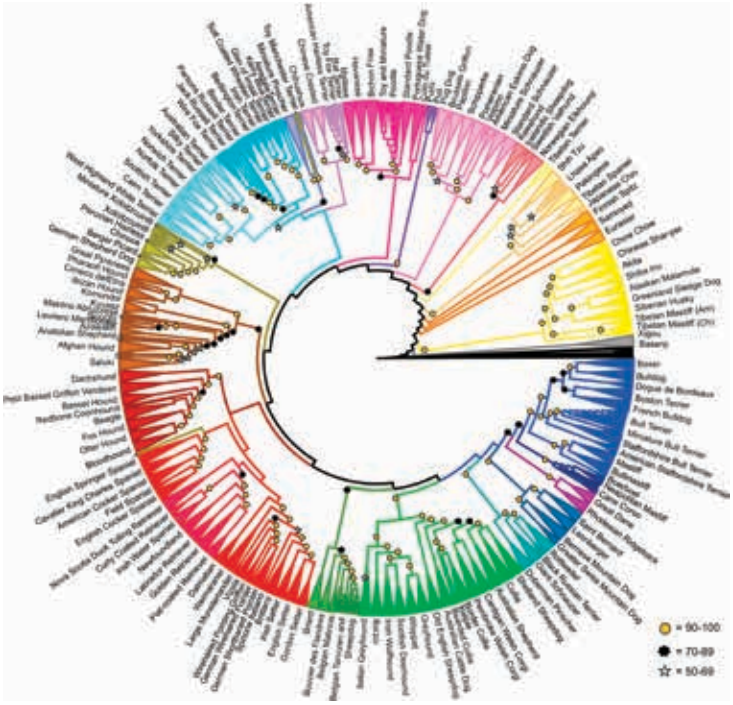
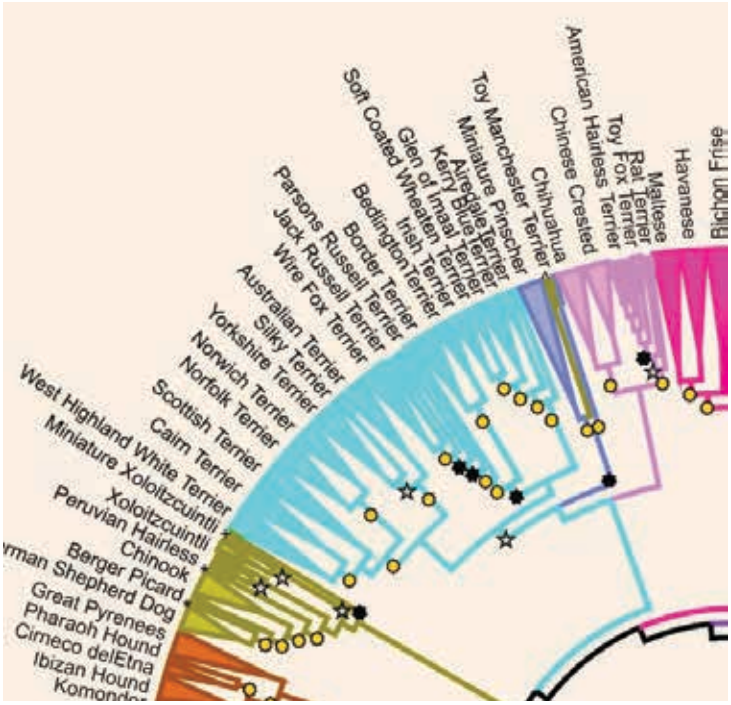
Why a Cairn is not a Westie – And vice versa!

Part Two: Genomic and Disease Susceptibility Differences

by Pat Joyce





The first part of this article described the historic origins of the two breeds and the current conformational differences as delineated in their breed standards. The second part will describe the genomic and health issues of the West Highland White Terrier (WHWT) and the Cairn Terrier (Cairn) and further demonstrate how they are distinctly different breeds at this time.

Similar appearance but different genomes

It is said that humans have produced over 350 different breeds of dogs since we first bonded with canines, estimated to have occurred some 30,000 years ago by archeologic findings. Certain factors favored the development of specific dog breeds. The original humans undoubtedly began contact with canids in their environment, then selected for animals amenable to domestication and to serve work and companionship functions. Whether in the distant past or more recent time, humans chose a specific animal to foster and breed. From that moment and locality, the human also created what is termed the "founder effect." Genetic information, for good or for bad, in that original choice of a specific animal to keep, became the basis for inheritance in all future generations.

The first complete mapping of a canine whole-genome sequence was done in 2005 and performed on the DNA of a Boxer. By 2015 descriptions of the dog genes included 2.41 billion base pairs across 38 autosomal chromosome pairs, the X chromosome, and the mitochondrial DNA. (Parker 2015) Specific markers for canine diseases have been identified such as the GALC gene for GCL/Krabbe disease (Cairn and WHWT), the DLA-DRB and DLA-DQA genes for insulin-dependent diabetes (Cairn, Samoyed, and Tibetan Terrier), the SLC37A2 gene for craniomandibular osteopathy (Cairns, WHWT, Scottish Terrier, and Boston Terrier), and the COMMDI gene for copper toxicosis (Bedlington).

A major description of the canine genetic spectrum was published in 2017 by Parker et al., in which 161 clades (genetic groups coming from a common ancestor) corresponded to specific dog breeds. While the methods of genetic analysis are beyond this article, the authors depicted the genetic relatedness of the breeds studied. Their work showed that Cairns and WHWT have a 90-100% similarity on "bootstrap" genetic analysis while maintaining that they are separate clades. These two breeds are next most related to

the Scottish Terrier clade, then to the Norfolk and Norwich Terriers. Of other Terriers, these breeds are more closely related to the Australian Terrier (a breed blended from terriers historically exported to that continent along with their humans). Of interest is that the Bedlington Terrier demonstrates a markedly different genetic background from Cairns or WHWT, despite the Bedlington's susceptibility to copper storage liver disease shared with the WHWT. (Parker 2017)

Disease Susceptibilities in Cairns and WHWT

The decisions made by Colonel Malcolm and The Duke of Argyll began the separation of the two breeds. Whatever genetic information was carried by their original chosen dogs to keep, and by every breeding decision since, WHWT and Cairn breeds have carried more than just a difference in coat color. Unbeknownst to those men as they sorted dogs by color, they invoked a founder effect with whatever other genetic information and disease susceptibility was within those dogs at that specific time. Since then the breeds have carried certain diseases in common and other diseases that are quite dissimilar.

To illustrate the genetic differences that currently separate the breeds, I will briefly summarize five diseases that are unique to each breed.

Copper toxicosis (WHWT)

Copper toxicosis is a chronic hepatopathy with a well-known genetic predisposition in Bedlington terriers and WHWT, but also reported in Skye and Airedale Terriers. An accumulation of copper stored in the liver cells causes a chronic hepatitis. The disease is caused by failure to excrete excess copper via the biliary system. An affected dog will progress over several years from being asymptomatic, to having a general "failure to thrive" (weakness, weight loss, and poor appetite), and finally to overt liver failure with cirrhosis by age four to seven years. Blood tests show general liver dysfunction but definitive diagnostic testing is by liver biopsy showing copper in the cells.

Copper toxicosis is known to be hereditary but the mode of transmission is unclear. The COMMDI gene has been identified as a genetic marker in Bedlingtons but not all dogs with the disease have the gene. It is said that in the past as much as fifty percent of Bedlingtons had copper toxicosis, but that the incidence now is greatly reduced due to genetic screening and breeding from the non-affected gene pool.

Copper toxicosis may be treated with the medication penicillamine given orally before meals and by feeding special prescription low-copper diets. Penicillamine is a chelator drug which binds copper in the blood and is then removed through the urine.

Portal Shunt Syndrome (Cairn)

Although reported very rarely in WHWT, this spectrum disorder is extremely well-known in Cairns. A functional blockage of the circulation of blood and bile acids occurs either outside the liver (extra-hepatic), within the liver (intra-hepatic) or on a cellular level (also known as micro-vascular dysplasia). Dogs affected may be asymptomatic as puppies but start to show failure to thrive by 6-12 months of age. Similar to liver cirrhosis in humans, severe portal shunt in canines results in a backup of the body's waste toxins into the blood. This affects the brain and results in general neurologic dysfunction, and then to seizures, coma, and death. Unrecognized portal shunt should be considered whenever there is a new-onset of seizures or brain dysfunction in a previously healthy Cairn of any age.

Inheritance of Portal Shunt Syndrome is thought to be autosomal but no clear patterns have been identified. Screening may be done by performing paired bile acid blood levels (done fasting and two-hours post-prandial) or by serum ammonia level (the screening test preferred in Europe). Severely affected dogs with highly elevated bile acid levels can be identified early and their owners warned. Dogs with less severely elevated bile acids may be identified as carriers of the syndrome. They may lead relatively healthy and asymptomatic lives but should be removed from the breeding population. Surgical decompression of the liver portal vessel system has been performed in severe extrahepatic cases. While hepatic diets have been recommended, there is no specific treatment for portal shunt.

White Shaker Syndrome (WHWT)

This disorder is known in white-coated dog breeds, particularly in Maltese, Samoyeds, and WHWT. Affected dogs develop a unique shaking. Fine, rapid, whole-body tremors are seen although other non-specific neurologic behaviors may include abnormal eye movements and seizures. The disorder has an onset at five months to three years of age. The onset and severity of shaking appears to be triggered by the dog becoming excited or stressed, or by simply moving or being handled. The shaking may cause a wobbly, uncoordinated gait with overreaching of the legs (hypermetria).

White Shaker Syndrome may be associated with a mild lymphocytic encephalitis. This will be known only if the animal undergoes examination of brain fluid. The differential cause of tremors includes toxin exposures and various metabolic disorders. This disease is not fatal by itself. Treatment is provided by giving steroid medications orally. Response with a decrease in tremors is said to be very rapid, perhaps within less than one week, and this is an important clinical clue that the diagnosis is correct. Steroid treatment needs to be tapered very slowly over many months. Recurrence of the tremors may result if the steroids are tapered too quickly. Benzodiazepine medications also may have some benefit.

Ocular Melanosis (Cairn)

Also known as Pigmentary Glaucoma, this disease occurs almost exclusively in the Cairn Terrier. Ironically the disease has been reported in humans. Pigment-containing cells (melanocytes) increase in the iris, choroid and surrounding eye structures. The cells eventually cause a blockage of the normal flow of fluids within the eye. This increased fluid pressure causes

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disruption of eye function and is known as glaucoma. Untreated ocular melanosis with glaucoma will lead to blindness. Medical or surgical treatment for severe glaucoma is the only treatment.

Inheritance of Ocular Melanosis is thought to be autosomal dominant in Cairns. No genetic screening test is yet identified. The disorder is diagnosed by ophthalmologic examination with dilation of the eyes. Unfortunately, affected dogs may not show abnormal eye exams until they are five to six years old. The dogs may have been bred already before the disease is apparent. Regular eye examinations, and ideally before a Cairn is bred, may identify specific dogs carrying the disease. Affected dogs should not be used for any further breeding. Knowing that the gene may be carried within a pedigree line and identifying carriers early are currently the only methods to prevent transmission.

Idiopathic Pulmonary Fibrosis (WHWT)

Also known as "Westie Lung Disease" this syndrome is unique to this breed and involves fibrotic changes in the lungs. The walls (septum) between alveolar air sacs become thick and stiff due to excessive collagen formation in the extracellular matrix. In addition to disrupting air exchange in the lungs, the condition causes secondary pulmonary hypertension of blood pumped from the right side of the heart and through the lungs.

This condition presents in older WHWT, with the majority of cases in animals over ten years old. The initial symptoms are dyspnea (shortness of breath), chronic cough, easy fatiguability, and intolerance of exercise. Heart sounds may suggest pulmonary hypertension. A chest radiograph will show lung changes due to thickening of the lung tissue and right heart enlargement. Low oxygen levels can be measured in arterial blood. Definitive diagnosis is made by biopsy of the lung tissue showing the fibrotic changes in the lung tissue.

Idiopathic Pulmonary Fibrosis progresses over one to two years after onset which is the life expectancy with the disease. There is no known cure. The dog may receive some symptomatic benefit from anti-inflammatory medications (corticosteroids and non-steroidals) and from bronchodilators and cough suppressants. There is no known genetic screening test to identify WHWT at risk. Westie Lung Disease is a particular problem for breeders as the illness presents in geriatric dogs with many generations of progeny but no way to screen the descendants.

The first chart above lists health conditions frequently associated with the two breeds.

The second chart above lists the recommended health tests for WHWT and Cairns as described by the national breed clubs (as of February 2022) and by the OFA.

	WHWT	**Cairns**
Genetic diseases more associated with either specific breed*	-Copper toxicosis of liver -White shaker syndrome -Idiopathic pulmonary fibrosis (Westie Lung Disease) -Atopic dermatitis/hyperplastic dermatosis -Keratoconjunctivitis sicca (dry eye syndrome) -Aggression, genetic based -Atrioventricular cardiac block, second degree	-Portal shunt/microvascular dysplasia of liver -Ocular melanosis/glaucoma -Renal dysplasia -Mitral valve prolapse with a murmur
Genetic diseases associated with both breeds*	-Craniomandibular osteopathy (CMO) -Globoid cell leukodystrophy (GCL or Krabbe disease) -Patellar subluxation -Avascular necrosis of femoral head (Legg-Calvé-Perthes disease) -Diabetes mellitus -Hypoadrenocorticism (Addison's disease) -Inflammatory bowel disease	

*Please see Westie Health Book and Gough and Tilley textbooks for extensive descriptions of specific disease entities and additional ailments common to both breeds.

	WHWT	**Cairns**
Recommended health testing by national breed club+	-Hip evaluation -Patella evaluation -Ophthalmologist examination	-Paired bile acid tests -Ophthalmologist examination -Renal ultrasound -Cardiac examination -Patella evaluation -GCL DNA test
OFA-CHIC requirements for breed to enroll in their program*	-OFA exam (hip dysplasia) -OFA exam (patellar luxation) -Eye exam – annually until 8 years old -CMO DNA test by approved lab	-OFA exam (patellar luxation) -Eye exam by approved ophthalmologist -Cardiac exam by approved cardiologist -GCL DNA test by approved lab

+per websites of CTCA and WHTCA and their Foundations
**https://www.ofa.org/browse-by-breed* (Accessed April 2022)

Recommended Health Screen Testing

Health issues of importance have been identified by the national breed clubs and given to the AKC-Canine Health Foundation and the Orthopedic Foundation for Animals-Canine Health Information Center (OFA-CHIC) which promotes a core of recommended testing. If an animal completes required testing for the breed (and if done following required protocols), a CHIC number and certification are given. The OFA-CHIC program maintains a database of health testing results, all voluntarily submitted by dog owners, including diseases not specifically required. Obviously, health data not voluntarily reported to OFA-CHIC are not included in their databases of breed information.

In conclusion

This two-part article arose from my desire to understand how Cairns and WHWT resemble each other so closely in appearance, but are in fact two separate and distinct breeds. While sharing a historic past, many changes have occurred over the years due to the founder effect which limited the genetic pools once the breeds were separated. Decades of breeding have defined their respective gene pools. While the breeds share a genomic similarity, they are indeed two separate entities at this time by conformational differences and by health susceptibilities.

I want to give my very special thanks to Marleen Burford, breeder-judge and member of the West Highland Terrier Club of America, for mentoring me in the Westie breed. I also wish to compliment the Westie Foundation for their outstanding *Westie Health Book*.

My conclusions do not represent any official positions of the Cairn Terrier Club of America or of the West Highland White Terrier Club of America. All interpretations of veterinary medical data are my sole opinions as a life-long student of the breeds.

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Editor's Note: The opinions expressed in this article are solely those of the author and do not express the views or opinions of the Cairn Terrier Club of America.

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