

OCULAR MELANOSIS IN CAIRN TERRIERS

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An emerging problem affecting Cairn Terriers, initial description by Covitz at the ACVO meeting in 1984, first published report by Petersen-Jones in 1991. This problem is of great concern to Cairn Terrier breeders and has been the focus of much attention in recent years.

Ocular melanosis is a progressive pigment proliferation within the eye that leads to secondary glaucoma and blindness. The condition is inherited, both eyes are affected and pigment proliferates and deposits in several areas within the eye. This is a slowly progressive disease and may not be noticed by the owner until the onset of glaucoma at least in one eye. The age of onset (often from about 4 years onward) and the rate of progression appear to vary. I have had the opportunity to monitor several affected dogs with no current signs of glaucoma and have found the progression indeed to be slow.

As pigment proliferates and disperses the cells may eventually obstruct the filtration angle which results in glaucoma. Signs of glaucoma seldom occur before about 7 years of age and most that I have seen are 10 years of age or more.

With the emergence of this condition the incidence of glaucoma in Cairn Terriers has increased from 0.51% of Cairns examined from 1974 to 1983 according to the Veterinary Medical Database, to 1.82% from 1994 to 2003.

The progression of this disease has been ranked into four stages of physical findings:

- I. Donut-shaped thickening of iris root (dark colour), scleral or episcleral pigment is not apparent. IOP normal and gonioscopic findings are unremarkable.
- II. Thickened iris root with appearance of small scleral pigment deposits particularly ventrally. Pigment plaques initially spicule shaped progressing to circular spots. Gonioscopy reveals pigment coating pectinate fibers.
- III. The scleral and episcleral patches become more extensive. Some patients have a “lumpy” appearing iris, others have donut-shaped iris root thickening. There may be reduced pupil constriction or dyscoria. Pigment particles are found in the aqueous. Gonioscopy shows pigment deposition coating the ventral drainage angle and extends onto the ventral corneal endothelium.

- IV. Further increase in pigment in the drainage angle, larger scleral patches. Glaucoma occurs and you may see changes typical of chronic glaucoma – buphthalmos, lens subluxation, optic disc cupping, vision loss, scleral staphyloma (area of thinned sclera) or sometimes phthisis.

The condition is bilateral and often quite symmetrical in progression. Glaucoma is not always bilateral or at least usually does not occur in both eyes at the same time. The posterior segment also shows progressive pigment deposition gradually encroaching on the tapetum and sometimes on the optic nerve head.

Pedigree analysis: autosomal dominant seems most likely. Heterozygous dogs might have a later clinical onset. Work is in progress at Michigan State University to identify the gene mutation. Abnormal ocular pigment dispersion (ocular melanosis) has been described in the Boxer and Labrador Retriever. The condition in these two dogs appeared histologically different, with the main cell being a melanophage in contrast to melanocytes in Cairn Terriers.

Three of 114 Cairn Terriers reported with ocular melanosis also developed ocular melanomas. It was speculated that the increased pigmentation in the anterior uvea may have increased the chance of developing melanomas. Such has been speculated at other sites and in other species.

Treatment: no usual treatment prior to the onset of glaucoma. Once glaucoma has developed standard topical glaucoma medications may be used – xalatan, dorzolamide, timolol. The condition may prove refractory to medications and if blind and painful then enucleation is a reasonable option. Affected dogs should not be bred. The goal of the genetic research is to develop a genetic test for ocular melanosis. With such a test affected dogs could be excluded from breeding at an earlier age.

Histopathology: diffuse infiltration of plump pigment laden cells in iris, ciliary body, sclera/episclera and peripheral deep cornea. The posterior segment is involved to lesser extent. The increased cellularity along with infiltration into the sclera, corneal stroma and optic nerve meninges and periphery of optic nerve is more suggestive of neoplasia. Large round cells packed with melanosomes are similar in microscopic appearance to those seen in uveal and limbal melanocytoma of dogs. However, in contrast to the classic limbal and uveal melanomas of dogs the cellular infiltration was diffuse rather than resulting in an expanding mass. Immunohistochemistry and transmission electron microscopy suggest pigment laden cells consist of two populations – melanocytes and melanophages. The primary increase in uveal pigmented cells is due to an abnormal proliferation of melanocytes. Inflammatory cells also occur and engulfed melanosomes. Pigmentary glaucoma in human beings appears to be different to ocular melanosis in Cairn Terriers.

Data collection: Contact Dr Simon Petersen-Jones at MSU: telephone 517 353 3278. They are interested in collecting more DNA samples from families with ocular

melanosis, both affected dogs and relatives. Blood sample preferred (purple top – EDTA). You can also send enucleated globes for histology and cell culture.