White Cats and Deafness

by Roy Robinson

The completely white cat, especially the Long-haired, is rightly regarded as one of the most beautiful of breeds. The eye colour may be orange or blue, or even one eye orange and the other blue on occasion (the Odd-eyed). These cats are healthy and fertile except for one problem, there is a propensity for impaired hearing.

The genetics of the white cat is simple enough. The colour is produced by a dominant gene ***W*** which is responsible for several features. These are:

- 1) white coat
- 2) blue iris to the eyes, and
- 3) deafness.

The white coat is invariably manifested but the blue irises and deafness are produced in only a proportion of cats but always in association with the white coat.

The Dominant White Syndrome

When a variety of effects are consistently produced, these are termed a syndrome. It is not unusual for one feature of a syndrome to be regularly expressed while the others are less so. This is the case for the dominant white syndrome. This is conventionally interpreted as **variable expression of the syndrome**. Simply put, the mildly-affected cat would have a white coat but normal eye colour and normal hearing, while the extreme expression would be a white coat with two blue eyes and deaf in both ears.

The white coat is produced by an absence of melanin pigment granules in the hair. Consequently, the hairs are translucent and appear white to human vision. The blue iris colour is due to a partial deficiency of the pigment granules. The defective hearing is due to a progressive degeneration of vital hearing organs of the inner ear.

When a gene produces a consistent syndrome of effects, this implies a common cause, rooted in the early stages of embryonic development. The present syndrome of pigment granule deficiency in the hair and eye, coupled with anomalies of the inner ear, has been described for several species of mammals (cat, dog and mouse) and has been extensively studied. This research indicates the origin of the syndrome can be traced back to an important feature of the embryo, namely the neural crest.

At a certain stage of development, a folding arises in the embryo to create the neural tube which has a very active region known as the **neural crest**. Active because, as development proceeds, the region supplies cells which are distended to become constituents of a great variety of tissues, organs and the nervous system (Hall 1988).

Abnormal Functioning of the Neural Crest

Included among these cells and of immediate relevance are specialised cells termed **melanoblasts** which, after arising from the specific sites in the neural crest, migrate between the dermal layers of the skin to take up positions at the base of the hair follicles where they are known as **melanocytes**. The function of the melanocytes is to synthesise and feed melanin granules into the growing hair. This is how the hairs are coloured.

Normally, the melanoblasts are able to reach all parts of the body so that the skin receives a full complement of melanocytes and the coat is completely coloured. However, this is not invariably the case and the migration may fail to reach some areas of the skin. Consequently, these areas are devoid of melanocytes and the hairs are colourless because they lack melanin granules. When the loss is partial, so that some areas of the skin have melanocytes and other areas do not, the bi-coloured or piebald pattern is produced, *i.e.* coloured cats with grades of white areas. A total absence of melanocytes results in a completely white coat.

The eye may be normally or near normally pigmented even when the coat is completely white. The reason appears to be that the eye tissues do not rely exclusively on the above migration but receive melanocytes by a more direct route, as in the case of the retina cells layer. However, the eye does not always escape and, when it is involved, the iris is blue indicating a general deficiency of pigmentation of the eye. One or both eyes may be affected. In particular, the ***tapetum lucudum***, which is responsible for the characteristic 'eye shine' of cats' eyes, may be partially or completely missing (Bergsma and Brown 1971, Thibos *et al*, 1980).

Similarly, the organs of the inner ear may be sufficiently well formed for hearing to be normal or near normal. In affected cats, the degeneration is progressive and involves both **cochlea** and **saccule** structures. The delicate hair cells of the **organ of Corti** disintegrate and the **saccule** collapses. The former are essential for reception of sound waves and sending impulses to the brain. The result of the degeneration is deafness. The anomaly may affect one or both ears, hence the animal is described as either unilateral or bilateral deaf (Bosher and Hallpike 1965, Mair and Elverland 1977, Elverland and Mair 1980).

The white coat could be regarded as the primary effect of the ***W*** gene because it is regularly expressed. However, this is another way of saying that the prime cause is a failure of the melanoblast migration. This implies the sites in the neural crest from which the melanoblasts originate are either deficient or malfunctioning. It is likely that these are not the only cells to be affected. The normal migration of those responsible for normal development of the eye and organs of the inner ear may be disrupted. It has been proposed that these could be either the melanoblasts or melanocytes (Steel and Barkway 1989).

In other words, the action of the gene ***W*** is not to induce a white coat. This happens to be incidental. The prime function of the gene is an anomaly of the normal functioning of the neural crest. The exact nature of the anomaly is unknown but the consequences cannot be other than profound because of the importance of cells from the neural crest in embryonic development.

Available Statistics

White kittens may have a small spot or patch of coloured fur on the head which disappears with maturity. Observations have revealed that cats with a patch are less likely to be deaf than those without (Table 1). The implication is that the presence of a patch is an indication that these cats received more functional melanocytes (even if few in number) compared with their fellows - that is, they were less severely affected.

The situation is quite different if the incidence of deafness is examined in conjunction with blue eyes.

The association between blue eye colour and deafness is well documented and is shown by Table 2. Cats with blue eyes are more prone to deafness than those with orange eyes. Furthermore, cats with two blue eyes are more likely to be deaf than cats with one blue eye (Table 3). The data is meagre but there is evidence that in doubly unilateral affected individuals, blue eye colour and deafness will occur on the same side of the head.

The implication of the above observations is the obverse of that for patch and deafness. It is apparent that those cats with two blue eyes are more severely affected than those with one. The former cats could have received fewer functional embryonic neural crest cells than the latter. That is, the extent of the underlying neural crest anomaly may vary and this accounts for the variation of expression of the syndrome.

The data is insufficient to establish statistically that kittens with a patch are less likely to have blue eyes (Table 4) although the trend is in this direction.

In view of the results described in previous paragraphs such an association would be anticipated and could be confirmed by additional data.

Many blue-eyed white cats, whether partially or completely deaf, lack a tapetum. Insufficient data is available to indicate the proportion of these animals but it would seem to be high. Absence would suggest a severely affected eye, hence it is assumed that it represents an increase in the severity of the anomaly.

Selective Breeding Difficult

The conclusion to be drawn from the analysis is that deafness in white cats is an aspect of a syndrome of effects of the dominant gene *W* and is inherent in all cats with the gene. Selective breeding against the incidence of deafness is likely to be difficult if not ineffective but if such a policy is to be adopted it would mean taking advantage of any inherited variation in the severity of the neural crest anomaly. That is by a selection of polygenes which could ameliorate the condition. In order to achieve this, it is necessary to have an idea of the variation of the degree of severity of the syndrome. From the data to hand, it is possible to illustrate the variation in the terms of increasing severity in the manner of Table 5.

This will mean that only matings between orange-eyed white cats of sound hearing be allowed. Those with spots or patches as kittens should be especially favoured. Blue-eyed as well as deaf cats (in both cases, unilateral as well as bilateral) would have to be removed from breeding. Mating between white and coloured cats should not be permitted because the genetic status of the latter as regards the postulated polygenes which could ameliorate the severity of the neural crest anomaly cannot be detected.

The objection to breeding cats with the ***W*** gene centres on the deafness and this appears to be mainly aesthetic. Deaf cats do not appear to be truly disadvantaged. It is possible that they could have a greater likelihood of being involved in road accidents. The lack of tapetum could mean that the vision is impaired in dim illumination compared with that of a normal cat. A deaf cat kept in a caring environment would be expected to live a normal life.

All breeds of cat which have the ***W*** gene will have a propensity to produce deaf individuals. This includes both the Shorthaired and Long-haired breeds, as well as Foreign White, Manx and Rex.

To investigate the problem of deafness in white cats it will be desirable to collect data on the incidence. Simple collection to ascertain whether a cat is

deaf or is not deaf could reveal differences between breeds in the incidence. It would be interesting and probably significant if such differences could be adequately established.

However, concentration on one feature is not sufficient: if new information is to be discovered. Especially, an endeavour should be made to collect data on the probable close association between absence of tapetum and deafness. From a genetic viewpoint, to gain greater insight on the pattern of incidence of deafness and other features, data should be collected on complete litters from parents of known status. Each kitten should be as fully classified as possible.

It should be mentioned that the blue eyes of the Birman, Colourpoint and Siamese breeds are produced by a completely different genetic mechanism which is not associated with deafness. This includes the blue-eyed albinos which are bred on the Continent. The eyes of the Siamese are deficient in pigmentation but a tapetum is present.

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TABLE 1: Dissociation between patch and deafness.

	Patched	No patch
Normal hearing	53	57
Deaf	22	62
Incidence(%)	29.3	52.1

Chi square = 9.71 for 1 degree of freedom, highly significant.

TABLE 2: Association between blue iris and deafness.

	Orange iris	Blue iris
Normal hearing	46	55
Deaf	13	68
Incidence(%)	22.0	55.3

Chi square = 17.85 for 1 degree of freedom, highly significant.

TABLE 3: Association between number of blue irises and deafness.

	Orange iris	1 blue iris	2 blue irises
Normal hearing	46	30	27
Deaf	13	19	50
Incidence(%)	22.0	38.8	64.9

Chi square = 55.00 for 2 degrees of freedom, highly significant.

TABLE 4: Lack of association between patch and eye colour.

	Patched	No patch
Orange iris	31	39
Blue iris	52	97
Incidence(%)	62.6	71.3

Chi square = 1.78 for 1 degree of freedom, not significant.

The figures for Table 1 to 4 are a distillation of data presented by Bergsma and Brown (1971).

TABLE 5: A depiction of increasinq severity of various affected eye colours and deafness of white cats.

Eye colour	Kitten patch	Deafness	Tapetum
Orange	Present	Absent	Present
One blue	Present	Absent	Present
One blue	Absen	Absent	Present
One blue	Present	Absent	Absent
One blue	Absent	Absent	Absent
Two blue	Present	Absent	Present
Two blue	Absent	Absent	Present
Two blue	Present	Absent	Absent
Two blue	Absent	Absent	Absent
One blue	Present	Present	Present
One blue	Absent	Present	Present
One blue	Present	Present	Absent
One blue	Absent	Present	Absent
Two blue	Present	Present	Present
Two blue	Absent	Present	Present
Two blue	Present	Present	Absent

Two blue	Absent	Present	Absent

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