Bengal Progressive Retinal Atrophy (PRA-b)

About the disease

Bengal PRA-b causes loss of photoreceptors in the eye and ultimately results in blindness. Clinical signs typically become evident between 8 and 20 weeks of age and the disease progresses so that by around one year of age complete retinal degeneration is apparent in most cats[1]. At this age the affected cats also show behavioural signs of blindness. Affected cats can have more difficulty at night, their pupils are usually more dilated and they show marked tapetal hyperreflectivity[1]. As with blind cats in general, PRA-b affected Bengals can negotiate their home environment relatively easily and are mobile and active.

The mutation is inherited as an autosomal recessive trait, meaning that cats with only one copy of the mutant gene (Heterozygous or Carrier) have normal vision, but they can pass the mutation to their offspring. Cats with two copies of the mutant gene (Affected) will develop PRA.

We have screened nearly 2500 Bengal cats from across Europe and found Carriers in most European countries (see table below). The overall European Bengal PRA-b Carrier prevalence is around 18%.

Country	Normal	Carrier	Affected
Austria	21	6	0
Belgium	54	17	1
Denmark	28	5	0
Finland	64	10	1
France	254	60	0
Germany	79	40	3
Italy	136	28	1
Netherlands	21	13	0
Norway	9	1	0
Poland	54	8	2
Russia	92	42	5
Spain	2	0	0
Sweden	234	41	2
Switzerland	3	2	0
UK & Ireland	912	171	19
Ukraine	33	13	0

[1]Ofri R, Reilly CM, Maggs DJ, et al. Characterization of an early-onset, autosomal recessive, progressive retinal degeneration in Bengal cats. Invest Ophthalmol Vis Sci. 2015;56:5299–5308.

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About the test

At the Molecular Diagnostic Unit, we have developed a PCR-based pyrosequencing assay to quickly and accurately identify the PRA-b mutation that is known to cause PRA in Bengal cats.

This assay will allow owners and breeders to identify Affected and Carrier cats and will inform breeding programs to reduce the prevalence of PRA-b by selective breeding.

Interpretation of results

A **Normal** result on the PRA-b genetic assay means that the cat does not have the PRA-b genetic mutation. It is possible that some cats may go on to develop retinal atrophy due to other, as yet unidentified, genetic mutations.

A **Carrier** result on the PRA-b genetic assay means that the cat carries one copy of the PRA-b genetic mutation. This cat is a Carrier of PRA-b and will not develop retinal atrophy due to the PRA-b mutation, but can pass the mutation to its offspring. It is possible that some cats may go on to develop retinal atrophy due to other, as yet unidentified, genetic mutations.

An **Affected** result on the PRA-b genetic assay means that the cat has two copies of the PRA-b genetic mutation and will be affected by Bengal PRA.

Each certificate we issue will specify whether the cat is Normal, Carrier or Affected for the Bengal PRA-b mutation.

The gene test will help breeders decide whether or not to use cats for breeding. Generally Affected cats should not be used for breeding because they are certain to pass on the genetic mutation. There is a 25% probability of two Carrier cats producing Affected kittens. Breeding Carrier and Normal cats will produce around 50% Normal and 50% Carrier kittens.

This strategy can be used as part of a breeding programme to gradually eliminate the defective gene from the affected populations.

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FAQs

I have already tested my Bengal for PRA (rdAc), should I test for PRA-b?

Yes, PRA-b is a specific mutation that causes PRA in the Bengal breed. PRA (rdAc) is thought to have originated in the Abyssinian breed. Since Abyssinians were used in the foundation of the Bengal breed it is valid to test Bengals for PRA (rdAc). We have not tested enough Bengals for PRA (rdAc) to determine a prevalence, however, if the PRA (rdAc) mutation is in the Bengal breed the prevalence is likely to be very low. The Bengal PRA-b mutation is likely to account for the majority of Bengal PRA cases.

What are the genetics of breeding?

Carrier		+	-
Carrier	+	+/+ Affected	+/- Carrier
	-	+/- Carrier	-/- Normal

Autosomal Recessive

Bengal PRA-b is inherited as an autosomal recessive trait, meaning that two copies of the defective gene are required in a cat to produce the disease.

Carrier cats (those with a single copy of the defective gene; Heterozygous) are not affected and have normal vision. However, breeding from two Carrier cats has a 25% chance of producing Affected kittens, a 25% chance of producing Normal kittens and a 50% chance of producing more Carrier kittens.

Therefore, it is NOT recommended to breed two Carrier cats together, since this can produce Affected kittens.

What do I do with a Carrier?

Normal		+	-
	-	+/- Carrier	-/- Normal
	-	+/- Carrier	-/- Normal

Breeding is still possible

It is possible to continue to use Carrier cats in breeding programmes to retain important breeding lines and to avoid reducing the size of the gene pool.

As long as Carrier cats are mated to Normal cats no Affected kittens will be produced. This mating is likely to produce kittens that are Carriers, which can be identified by genetic testing and, if necessary, future matings arranged with Normal cats.