

Code of Practice for the Responsible Breeding of Animals with Heritable Defects that cause Disease



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PREVENTION OF CRUELTY TO ANIMALS ACT 1986 CODE OF PRACTICE FOR THE RESPONSIBLE BREEDING OF ANIMALS WITH HERITABLE DEFECTS THAT CAUSE DISEASE

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PREFACE

The *Prevention of Cruelty to Animals Act 1986* ('the Act') came into force on 20 May 1986 and is administered by the Department of Primary Industries (DPI). It has the purpose of protecting animals, encouraging the considerate treatment of animals and improving the level of community awareness about the prevention of cruelty to animals.

This Code of Practice for the Responsible Breeding of Animals with Heritable Defects that cause Disease is made under Section 7 of the Act. It was developed by the Bureau of Animal Welfare DPI in consultation with a working group comprised of persons who have knowledge and expertise in animal welfare, veterinary science, the commercial use and breeding of animals and the testing, diagnosis and control of heritable diseases in animals, and major stakeholders.

1 LEGAL REQUIREMENTS

The Prevention of Cruelty to Animals Act 1986 sets out offences for intentionally or recklessly breeding an animal with a heritable defect that causes disease as listed in the Schedule ('the Schedule') of the Act.

It is a cruelty offence to permit an animal to suffer from a heritable disease.

The code requires that animals with disease caused by a heritable defect must not be disposed of to another person without advice of the animal's heritable defect status.

The advice provided by the breeder must include:-

- 1. Permanent identification details e.g. number of microchip implant, brand or ear tag or tattoo, depending on the accepted method of permanent identification for the species of animal; and
- 2. Veterinary certificate with details of the diagnosis linked to that permanent identification.

2 PURPOSE OF THE CODE

The purpose of the Code is to set standards for the prevention and spread of heritable defects and the expression of disease caused by them.

The Code aims to educate animal breeders how to best minimize or avoid the development of heritable disease in progeny caused by inappropriate selection and mating of animals with heritable (genetic) defects. It also outlines breeding practices that will assist the reduction of the prevalence of the heritable defect in the animal population.

The standards set by the Code should be practiced by owners and custodians of animals used for breeding that are affected by any heritable defect that causes disease and must be observed for breeding of animals with heritable (genetic) defects causing the diseases listed the Schedule of the Act.

A person breeding animals in a program that conforms at least to the principles in this code is not considered to be breeding animals recklessly or intentionally as defined as an offence in Section 15C(1) of the Prevention of Cruelty to Animals Act 1986.

3 DEFINITIONS

ACES: AVA-ANKC Australian Canine Eye Scheme, a national certification system conducted by registered veterinary eye specialists to internationally recognised standards.

ANKC: Australian National Kennel Council.

Approved Organisation: for any species this is an organisation approved by reference in this Code of Practice in Section 6. Only this organisation may approve breeding programs for that species.

Approved Breeding Program: program developed in consultation with **veterinary specialists**, **geneticists** and **organisations** breeding the species, with support given by their respective professional bodies and associations of members. While the program must meet the standards of this code, there is no constraint on developing a higher standard of breeding management to reduce the prevalence of defective genes and heritable disease in the breeding population.

Approved collection officers: breed association designated collection officers nominated and approved to collect samples for testing at shows or specific testing days or a veterinary practitioner.

AVA: Australian Veterinary Association.

Veterinary practitioner: means a registered veterinary practitioner.

Affected: refers to the homozygous affected state, where the animal in question is abnormal in both phenotype and genotype and is diseased.

Clear: refers to the homozygous unaffected state, where the animal in question is normal in both phenotype and genotype. This can be proven by testing or evidence that both parents are Clear.

Collie Eye Anomaly: a complex of potentially blinding congenital eye defects, of which choroidal hypoplasia is the simplest and least threatening to vision.

Carrier: (or Normal) refers to the heterozygous unaffected state, where the animal in question is normal in phenotype but abnormal in genotype. For simple autosomal recessive conditions the animal is not diseased. With simple autosomal dominant conditions the Carrier state is diseased.

Desexing: means a scientifically accepted method that permanently prevents reproduction in the species e.g., surgical ovariohysterectomy or castration, fallopian tube ablation or vasectomy. An exception for large animals such as livestock might be achieved by distinctively and permanently identifying the animals and preventing breeding of them with appropriate barriers endorsed by the approved breeding program.

Dominant: a genetic disease that appears when the progeny has received one copy of the defective gene, from either parent.

Heterozygous: possessing two different forms of a particular gene, one inherited from each parent.

Homozygous: possessing two identical forms of a particular gene, one inherited from each parent.

Intentional breeding: is breeding of animals done or made or performed with purpose and intent.

Recessive: a genetic disease that only appears when the progeny has received two copies of the defective gene, one from each parent.

Reckless breeding: is highly unreasonable conduct that is an extreme departure from ordinary care outlined in this Code.

Test: is the recommended method of diagnosing the carrier or affected status of an animal. It may include DNA tests or other tests or physical examinations recommended by veterinary practitioners and scientists. Some conditions may require additional examinations as required by an approved breeding program e.g. CEA in some dog breeding programs may require an ACES Panellist examination before eight weeks of age. Such programs may require that only approved collection officers collect samples for testing in order to ensure high standards of identification of animals and record keeping. Test results must be recorded and kept by the breeder in a manner that permanently identifies the result to the animal tested.

Unknown: refers to an animal of a breed or cross-breed that is known to be at risk from the condition and the animal has not been tested for it. There is reason to suspect the animal for the condition due to a diagnosis in the progeny or parents. Such animals must be tested before use in a breeding program.

4 HERITABLE DISEASE GROUPS AND PERMITTED BREEDING PRACTICES

Breeding programs must consider the effects and ethics of high risk mating combinations that may, based on the principles of genetic inheritance, in theory produce animals with heritable disease. Where such heritable disease has potential to cause severe welfare issues for affected progeny such breeding programs must be justifiable. Affected progeny must be assessed and humanely destroyed if they suffer. Such animals must not be used for breeding.

Heritable diseases may be grouped by the manner in which they are inherited:

- **4.1** *Dominant diseases* only require one defective gene to be present for the disease to be caused i.e. both the heterozygous and homozygous states for the defective gene each develop the disease. This also includes dominant conditions that can only show partially or some conditions that only show in a particular sex. For example:
 - Cats

Polycystic Kidney Disease.

Folded ears associated with osteochondrodystrophy (e.g. Scottish Folds). Aplasia or hypoplasia of long bones (e.g. Munchkin cats or Twisty Cats).

Dogs

Progressive Retinal Atrophy (in those breeds where dominant inheritance has been scientifically established).

Hereditary cataract (in those breeds where dominant inheritance has been scientifically established).

- **4.2** *Simple recessive diseases that result in severe signs of disease* in the homozygous state for the defective gene. For example:
 - Dogs

Neuronal Ceroid Lipofuscinosis. Von Willebrand's Disease type 3.

- **4.3** *Simple recessive diseases that may take years to develop signs of the disease* in the homozygous state for the defective gene. For example:
 - Dogs

Progressive Retinal Atrophy (in those breeds affected by the *prcd* form, also *rcd* 1,2,3).

Hereditary Cataract (in breeds where a simple recessive mode has been scientifically established).

- 4.4 Simple recessive diseases that are sex linked or show weak penetrance and limited expression of the disease resulting in only a few affected individuals. While the following diseases are not listed in the Schedule of the Act some examples of this grouping are Haemophilia A, X-linked PRA type 1, X-linked PRA type 2 (described in crossbred dogs) and goniodysgenesis as an established risk factor for canine glaucoma.
- 4.5 Simple recessive diseases that are also dependant on over-riding or modifying genetic effects for full expression, before they pose a threat as a debilitating condition. This includes conditions where the vast majority of genetically affected individuals fail to exhibit the full range of clinical signs unless modifying factors are present factors that directly influence the degree to which the disease is ultimately expressed. For example:

Dogs
 Collie Eye Anomaly.
 Von Willebrand's Disease type 1 and 2.

- **4.6** *Polygenic disease* where more than one gene is involved and environmental effects can add to the severity of the condition. While the following diseases are not listed in the Schedule of the Act they are examples of diseases in this grouping that have widely divergent signs canine hip dysplasia and elbow dysplasia. These are also conditions where simple and/or effective DNA tests are unlikely to be developed.
- **4.7** *Recognised inherited diseases* that produce significant potential health risks in small numbers of affected individuals, but where there is no advance warning mechanism offered through the early onset of signs or the availability of a reliable genetic test, to be able to predict the development of debilitating disease in later life. For example:

Dogs

Hereditary Cataract (where late onset is characteristic of the condition).

5 HERITABLE DEFECT BREEDING STANDARDS

5.1 Heritable disease caused by a simple <u>dominant</u> defective gene

Carrier (is affected) = heterozygote (i.e. one clear gene and one defective gene), displays degrees of disease.

Affected = homozygous for heritable defect genes (i.e. two defective genes) displays severe form of disease.

Clear = homozygous for clear genes (i.e. two clear genes) and is free of the disease.

The breeding of animals with the following diseases caused by a heritable defect must be conducted in accordance with the Code.

• Cats

Polycystic Kidney Disease. Folded ears associated with osteochondrodystrophy. Aplasia or hypoplasia of long bones

Dogs

Progressive Retinal Atrophy (In those breeds where dominant inheritance has been scientifically established).

Hereditary cataract (in those breeds where dominant inheritance has been scientifically established).

Animals with a heritable disease of this type that causes suffering and disability in the animal must not be used for breeding. Where the carrier state will produce lethal outcomes in the progeny they must not be used for breeding.

In the following table breeding of animals with the heritable disease may be permitted but only under the specified conditions and only if approved by a veterinary practitioner as suitable for breeding.

Parent combination	Theoretical status of progeny	Heritable disease requirements	
Clear x Clear	100% Clear	No restriction	
Clear x Carrier	** 50 % Clear 50 % Carrier	 Progeny must be tested for the heritable defect**. 	
	(that may be diseased to some degree)	2. The severity of the disease in the Carrier progeny must be assessed by a veterinary practitioner and the animal managed in accordance with the instructions of a veterinary practitioner.	
		3. A diseased (Carrier) animal's must not be disposed of to another person without advice of the animals heritable disease status.	
		4. A diseased (Carrier) animal must be de-sexed unless they are to be used in an approved breeding program, must not be permitted by their owner to suffer from their condition and must be under the supervision and monitoring of a veterinary practitioner.	
Carrier x Carrier	** 25% Clear	1. Breeding prohibited unless as part of an approved breeding program.	
Currier	50% Carrier (diseased to some degree) 25% Affected	 All progeny must be tested for the heritable disease. ** 	
	(usually seriously diseased)	 The severity of the disease in Affected and Carrier progeny must be assessed by a veterinary practitioner and the animal managed in accordance with the instructions of a veterinary practitioner. 	
		4. A diseased (Carrier and Affected) animal must not be disposed of to another person without advice of the animal's heritable disease status.	
		5. Diseased (Carrier and Affected) animals must be de-sexed unless they are to be used in an approved breeding program, must not be permitted by their owner to suffer from their condition and must be under the supervision and monitoring of a veterinary practitioner.	

Affected x	100% Carrier	1.	Breeding prohibited unless as part of an
Clear	(will have a		approved breeding program and only with
	degree of the		the purpose of establishing sufficient breeding
	disease)		stock for the breeding program to develop
			Clear animals.
		2.	The severity of the disease in progeny must
			be assessed by a veterinary practitioner and
			the animal managed in accordance with the
			instructions of a veterinary practitioner.
		3.	A diseased (Carrier and Affected) animal's
			must not be disposed of to another person
			without advice of the animals' heritable disease
			status.
		4.	Diseased (Carrier and Affected) animals must
			be de-sexed unless they are to be used in an
			approved breeding program, must not be
			permitted by their owner to suffer from their
			condition and must be under the supervision
			and monitoring of a veterinary practitioner.
Affected x	50% Carrier,	1.	Breeding prohibited .
Carrier	50% Affected		
	(all will be	2.	Use of these combination is an offence under
and	diseased		the Act.
	to some		
	degree or		
	be seriously		
	diseased)		
Affected by	100%		
Affected	Affected		
	(usually		
	seriously		
	diseased)		

**Testing is required as in practice the unpredictable nature of the process of gene inheritance in these combinations may cause variation in the actual % outcomes per generation. As carriers may express varying degrees of the heritable disease they must be tested, assessed and monitored by a veterinary practitioner experienced with the disease to determine the impact on the animal.

5.2 Heritable disease caused by a simple <u>recessive</u> defective gene resulting in severe disease

Carrier = heterozygote (i.e. one clear gene and one defective gene) and does not exhibit the disease.

Affected= homozygous for heritable defect genes (i.e. two defective genes) and is affected by the disease.

Clear = homozygous for clear genes (i.e. two clear genes) and is free of the disease.

The breeding of animals with the following diseases caused by a heritable defect must be conducted in accordance with the Code.

Dogs

Neuronal Ceroid Lipofuscinosis (CL). Von Willebrand's disease Type 3.

Parent breeding combinations of animals with a heritable defect of this type that result in a disease that causes suffering and disability in progeny must not be used outside of an approved breeding program and only where the approved organisation believes it to be justifiable in the short term to establish breeding stock that are Clear.

Parent combination	Theoretical status of progeny	Heritable disease requirements
Clear x Clear	100% Clear	No restriction.
Carrier x Clear	** 50 % Clear 50 % Carrier (not diseased)	 Progeny to be used for <i>breeding purposes</i> must be tested for the heritable defect**. Progeny should all be tested for the heritable defect. Carrier animals must be desexed if not to be used for breeding purposes.

	1	1	
Carrier x	**	1.	Breeding not recommended. Must only occur
Carrier	25% Clear		as part of an approved breeding program.
	50% Carrier		
	25% Affected	2.	All progeny must be tested for the heritable
	(diseased)		defect**
	(uiseaseu)		delect .
		3.	Diseased (Affected) and Carrier progeny must
			not be disposed of to another person without
			advice of the animal's heritable defect status.
		4.	Diseased (Affected) and Carrier animals must
			be de-sexed if not to be used for breeding
			nurnoses
			purposes.
		_	Discourse description of the second state of t
		5.	Diseased animals must not be permitted by
			their owner to suffer from their condition
			and must be under the supervision and
			monitoring of a veterinary practitioner.
Affected x	100% Carrier	1.	Breeding not recommended. Must only occur
Clear			as part of an approved breeding program
			and only with the purpose of establishing
			sufficient broading stock for the broading
			sufficient breeding stock for the breeding
			program to develop Clear animals.
		2.	Progeny must not be disposed of to another
			person without advice of the animal's
			heritable defect status.
		3.	Carrier animals must be de-sexed unless to be
			used in an approved breeding program.
Affected x	50% Carrier	1.	Breeding prohibited .
Carrier	50% Affected		· ·
	(diseased)	2	Intentional or reckless use of these
and	(abcubcu)	<u> </u>	combinations is an offence under the Act
and			combinations is an onence under the Act.
Affected	100%		
Affected x	100%		
Affected	Affected		
	(diseased)		

**Testing is required as in practice the unpredictable nature of the process of gene inheritance in these combinations may cause variation in the actual % outcomes per generation.

5.3 Heritable disease caused by simple recessive gene that may take years to develop symptoms of the disease

All progeny will initially appear to be unaffected by the disease. Depending on the severity of the disease and time of onset of the breeding program the animal may be bred before it is known they carry the heritable defect.

• Dogs

Progressive Retinal Atrophy (where affected by the *pcrd* form, also *rcd* 1,2,3). Hereditary Cataract (where a simple recessive mode has been scientifically established).

Parent	Theoretical	He	ritable disease requirements
combination	status of		
	progeny		
Clear x Clear	100% Clear	No	restriction.
Clear x Carrier	**	1.	Progeny to be used for breeding purposes
	50 % Clear		should be tested for the heritable defect**.
	50 % Carrier		
		2.	All progeny should be tested for the heritable
			defect.
		3.	Carrier animals should be de-sexed if not to
			be used for breeding purposes.
Carrier x	**	1.	Breeding not recommended. Must only occur
Carrier	25% Clear		as part of an approved breeding program.
	50% Carrier		
	25% Affected	2.	All progeny must be tested for the heritable
	(may develop		defect.
	disease)		
		3.	A diseased (Affected) animal must not be
			disposed of to another person without advice
			of the animal's heritable defect status.

		4.	Affected progeny (or any juvenile offspring
			confirmed as 'Affected' on test) should be
			de-sexed unless they are to be used in an
			approved breeding program, must not be
			permitted by their owner to suffer from their
			condition if it develops and should be under
			the supervision, advice and monitoring of a
			veterinary practitioner.
Affected x	100% Carrier	1	Breeding not recommended. Should only
Clear			occur as part of an approved breeding
			program and only with the purpose of
			establishing sufficient breeding stock for the
			breeding program to develop Clear animals.
		2.	Progeny must not be disposed of to another
			person without advice of the animal's
			heritable defect status.
		3.	Carrier animals should be de-sexed unless to
			be used in an approved breeding program.
Affected x	50% Carrier	1.	Breeding prohibited.
Carrier	50% Affected		
and	(may develop	2.	Intentional or reckless use of these
	the disease)		combinations is an offence under the Act.
and			
		3.	Under exceptional circumstances the Affected
Affected x	100% Affected		x Carrier combination may occur as part of
Affected	(may develop		an approved breeding program but only
	the disease)		with the purpose of establishing sufficient
			breeding stock for the breeding program to
			develop Clear animals. Progeny must not be
			disposed of to another person without advice
			of the animal's heritable defect status.

**Testing is required as in practice the unpredictable nature of the process of gene inheritance in these combinations may cause the actual % outcomes per generation to vary from the theoretical outcomes.

5.4 Heritable disease caused by simple recessive genes that are sex linked (or show weak penetrance or limited expression resulting in only a few affected individuals).

See section 4.4. Advice from a veterinary practitioner and approved organisation should be sought before considering a breeding program.

5.5 Heritable disease caused by a simple recessive defective gene that is dependant on over-riding or modifying genetic effects for full expression of disease.

This includes conditions where the vast majority of genetically affected individuals do not exhibit the full range of clinical signs of the disease unless modifying factors are present i.e. factors that directly influence the degree to which the disease is ultimately expressed. All progeny may initially appear to be unaffected by the disease.

• Dogs Collie Eye Anomaly.

Von Willebrand's Disease type 1 and 2.

Parent	Theoretical	Heritable disease requirements
combination	status of	
	progeny	
Clear x Clear	100% Clear	No restriction.
Clear x	50 % Clear	1. Progeny to be used for <i>breeding purposes</i>
Carrier	50 % Carrier	must be tested for the heritable defect.
		2. All progeny should be tested for the heritable defect.
		3. Carrier animals should be de-sexed if not to be used for breeding purposes.

**Testing is required as in practice the unpredictable nature of the process of gene inheritance in these combinations may cause variation in the actual % outcomes per generation.

5.6 Polygenic based heritable diseases

See section 4.6.

Generally, where these conditions affect large numbers of the breed, broadbased surveillance and assessment schemes have been developed. The worst affected individuals should be removed from the breeding population before they reach maturity. Development of reliable statistical results such as sire's statistics can further help breeds lower the incidence and severity of the disease. These control schemes have been shown to work over the longer term, by raising the standard.

5.7 Recognised Inherited Diseases that produce significant health risks in small numbers of affected individuals, where there is no advance warning mechanism offered through the early onset of signs or the availability of a reliable genetic test. Some of these diseases are recognised as 'breed predilections', i.e. a higher than normal incidence may be observed within that breed. Many of these conditions appear unpredictably in the older animal, with no apparent inheritance pattern.

It may be impossible to establish whether or not a debilitating condition that arises at mature age is controlled by inherited factors at all, and is therefore able to be predicted, selected against or detected in advance by an established genetic test.

General awareness of a possible breed predilection is the best protection that can be issued against these conditions in the longer term. Purchasers of animals likely to develop the disease should consider this when making their selection of an animal to keep or breed.

Dogs

Hereditary Cataract (where late onset is characteristic of the condition).

6 APPROVED BREEDING PROGRAMS AND APPROVED ORGANISATIONS

- **6.1** Approved breeding programs must be reviewed every 3 years by the approved organisation to evaluate progress in reducing the prevalence of the heritable defect and the disease it causes and to ensure that there is compliance by its members with this Code.
- **6.2** A breeder must be a member of an approved organisation to undertake its approved breeding program.
- **6.3** Organisations should aspire to develop breeding programs that reduce the prevalence of the heritable defect in their breeding stock.

Species	Approved organisation		
Cats and dogs	An 'applicable organisation' approved by the Minister for		
	Agriculture in accordance with the Domestic (Feral and		
	Nuisance) Animals Act 1994.		
Other species	A recognised professional body or association of breeders of		
	the species.		

6.4 Approved Organisations.

7

PROCESS FOR AMENDING THE HERITABLE DEFECTS SCHEDULE OF THE PREVENTION OF CRUELTY TO ANIMALS ACT

Recommendations to amend the Schedule may be made by approved organisations to the Minister. Submissions should provide evidence of consultation with **veterinary specialists**, **geneticists** and the **approved organisation** with support given by their respective professional bodies and associations of members.

Diseases to be listed should:-

- Be established, well-researched inherited diseases or defects known to be present in a local breed population (or likely to be imported from overseas); and
- ii. Have sufficient researched information to allow the condition to be correctly diagnosed and categorised.

In making a recommendation to the Minister the following information must be provided:-

- i. The severity of the disease;
- ii. The mode of inheritance (dominant, simple recessive, etc);
- iii. The proportion of 'affected', 'carrier', and 'clear' individuals within the breed;
- iv. The number of diseases being simultaneously tested/screened within a breed;
- v. Ease of access to a range of reliable and repeatable screening methods;
- vi. There should be a reliable test to diagnose the disease that is considered cost effective by the approved breeding program; and
- vii. Summary of consultation comments with breeders, veterinary specialists, geneticists and members of the approved organisation affected by the proposed amendment.

Where the number of affected individuals is very low, few affected animals will be bred from nor are needed in the gene pool for the breed. They should be prevented from breeding. When the percentage of affected and carriers is high, more time will be needed to manage the risk of producing affected individuals, so that other pressures present in any 'closed' population do not force the emergence of hitherto hidden diseases, as a result of disproportionate restrictions to the existing gene pool.

The more diseases being tested for or screened, the slower the overall progress will be in a breeding program. Some individuals may be shown to be genetically clear for one or two conditions under test, yet be affected by a third, perhaps milder condition.