6. Guidelines

Guidelines for the use of the ECVO certificate in the Known and Presumed Hereditary Eye Disease scheme (KP-HED)

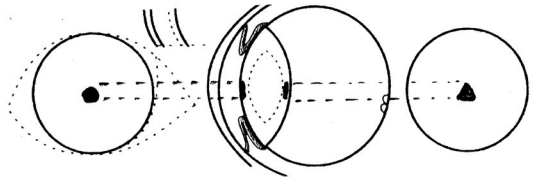
Section **Animal**:

Breed club: In countries where there is more than one society for one breed, the name of the society to which the results are to be reported is registered.
Previous examination: When reports from previous examinations are available, and the animal was recorded as "undetermined", "suspicious" or "affected", the date, the certificate number and the registration number of the examiner are noted.

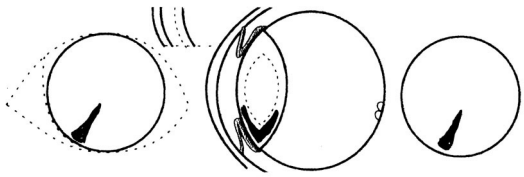
Section **Examination**:

The drawings in the middle of the form are used to draw and position any changes found. The circles on the left can be used for the cornea, e.g. to position corneal dystrophy, or for the anterior capsule of the lens. The dotted lines around the first circle represent contours of the lids and nictitating membrane. These can be used to indicate the presence and position of e.g. aplasia/coloboma of the lids, dermoid etc. The depth of corneal disease can be shown in the corneal section drawings. The position and contour of cataracts in the anterior part of the lens are marked on the circle to the left for each eye and posterior cataracts on the circle to the right for each eye. In the transverse section of the lens the position of the cataract is drawn, e.g. cortical, nuclear, capsular.

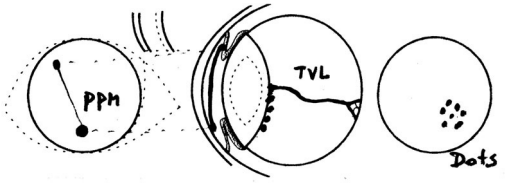
Examples of how to draw cataracts and PPM's are given below:



Anterior, polar cataract, diameter approx 2 mm, and posterior, polar, subcapsular (=cortical), triangular cataract.



Anterior and posterior, spoke-shaped, cortical cataract, from pole to pole, via the equatorial area, at seven-o-clock.



Group of (retrolental) dots on the posterior capsule of the lens at 5-o-clock and a persistent hyaloid artery from a Mittendorf's dot on the posterior capsule to a Bergmeister papilla. A PPM from the endothelium, 2 mm from the limbus at 6-o-clock to the iris collarette at 11-o-clock.

It is strongly recommended to give conclusive comments in English to easier enable translation into other languages.

Section Examination, part: **Descriptive comments**:

The number of the relevant eye disease is noted, especially for abnormalities, e.g. pectinate ligament abnormality. Tick boxes are provided for mild, moderate or severe, enabling the examiner to indicate the degree/severity/significance of involvement. The assessor will know which result to include in the computerized data collection system for each country. Such data can then be used for research purposes, such as proving the mode of inheritance of a specific eye disease.

Section Results:

'Unaffected' means that there is no evidence of the KP-HED specified. 'Affected' signifies that there is clinical evidence of the KP-HED . When the animal displays clinical features that could possibly fit the KP-HED mentioned, but the features are not specific enough, the result of the examination is: 'undetermined'. If the animal displays minor, but specific clinical signs of the specific KP-HED, the result of the examination is: 'suspicious'. Further changes may confirm the diagnosis and re-examination in at least 6 to 12 months is then recommended.

The box for the eye disease found and the specifying box, if available (e.g. for type or grade) are ticked.

The available name of the disease in the list of Definitions of this Manual (see chapter 5) is used

For number **"7. Other**", on the certificate, **known and presumed hereditary eye anomalies** (congenital/developmental, non-progressive) are mentioned. These disease names are also used in "roll down" menus in the computerized forms. These are:

Anophtalmos
Choroidal hypoplasia
Choroidal hypoplasia in Non-Collie breeds
Dermoid
Eyelid coloboma
Hyaloid artery, persistent: severe (e.g. causing vision impairment)
Iris coloboma: use iris hypoplasia
Iris hypoplasia
Lacrimal punctum atresia/micropunctum
Lens hypoplasia
Lenticonus
Lentiglobus
Macrophthalmos
Microphthalmos
Microblepharon
Microphakia
Nictitating membrane, eversion of the cartilage of the
Nictitating membrane, prolapse of the gland of the
Multiple ocular anomalies for 2 or more anomalies, to be specified in the comment field
Optic disc coloboma
Retinal coloboma
Retinal dystrophy/ RPE65 mutation (include ERG results in non Briard breeds)
Scleral coloboma

For number **"18. Other"**, on the certificate for KP-HED**,** which are considered not to be congenital/ developmental or which are progressive, and not yet named on the form, are mentioned. The available name of the disease in the list of 'Definitions' of this Manual (see chapter 5) is used. These disease names are also used in the "roll down" menus in the computerized forms. These are:

Retinal degenerations, other presumed hereditary, to be specified in the comment field
Canine multifocal retinopathy (CMR)
Ceroid lipofuscinosis (CLN)
Chorioretinopathy, pigmentary (e.g. in Chinese crested)
Keratitis: Chronic superficial keratitis (CSK)/Pannus

Keratitis, Punctate (in specific breeds e.g. Dachshund)
Keratoconjunctivitis sicca (KCS; in specific breeds e.g. WHWT, Cavalier King Charles spaniel, Chinese crested, LH Dachshund
Ocular melanosis (do not use Glaucoma – pigmentary; e.g. Cairn Terrier)
Uveal cyst
Uveitis, pigmentary (e.g. in Golden retriever)
Vitreous degeneration and – prolapse (without any sign of lens luxation)

General guidelines and considerations:

For litter examinations, a separate Certificate should be issued for each animal examined. The examination can only be performed after permanent identification of the examined animals (see chapter 3, The Scheme). It is possible to use a litter form as long as the data can easily be transferred on the European database..

1. If a dog is exported, all results of former examinations are sent together with the pedigree to the new registry.
2. Gene testing for eye diseases does not replace clinical eye examination.
3. When a dog is found 'affected' for a KP-HED by a panel member or the local appeals authority and the dog is transferred to another registry, the result 'affected' for this KP-HED will not be changed, unless the dog has been re-examined by the appeals authority of the new registry. Exceptions to this are conditions that are changed artificially with surgical correction. In those cases the previous results are definitive (e.g. distichiasis, entropion).
4. If a dog is transferred from one registry to another, the "exporting" registry provides all results of former examinations in regards to KP-HED and the "importing" registry includes them in their official data.

For an ophthalmic screening examination in accordance with the ECVO Scheme, evaluation of the entire eye is recommended. This examination includes the adnexa and the anterior and posterior segments. Visual function should also be noted if abnormal.
It is recommended to examine every animal before dilation.

List of breeds in special concern for the below listed KP-HED to be examined before dilation:

1. **Pectinate ligament abnormality (PLA) and/or angle with defects:**

American Cocker Spaniel
Bouvier des Flandres
Bassets (all)
Bloodhound
Chow Chow
Border Collie
Dandy Dinmont Terrier
Dutch Shepherd (Rough Hair)
English Springer Spaniel
Entlebucher Mountain Dog
Flat Coated Retriever
Golden retriever
Husky Siberian
Leonberger
Magyar Viszla
Samoyed
Tatra Mountain dog

2. **Persistent Pupillary Membrane using slitlamp biomicroscopy**

Basenji
Chow Chow
English Cocker Spaniel
Petit Basset Griffon Vendéen

3. Iris hypoplasia using slitlamp biomicroscopy

Australian shepherd
Rottweiler
Dalmatian

4. **Ocular melanosis/Lens luxation/vitreous degeneration/KCS using slitlamp biomicroscopy**

Small Terrier breeds (ocular melanosis: Cairn Terrier)
Chinese crested dog (also for KCS)
Dachshund (KCS)
English Bulldog (KCS)
Lancashire heeler
WHW terrier (also for KCS)

Some recommendations and details in regards to ticking of the ECVO certificate of eye examination
The given figures are found on the ECVO website.

Cataracts

If cataracts are observed in the period between birth and the 8th week of age the entity is ticked as congenital. Cataracts diagnosed at older age are ticked as non-congenital (acquired). If there is distinct proof the cataract is congenital in origin (e.g. associated PPM), the boxes for congenital and non-congenital cataracts can be ticked. It is strongly recommended to draw the cataract in the "pre-drawings" on the certificate, as seen from the anterior lens capsule (see separate instructions for drawing and filling the form). For the Scheme it is advised all bilateral or unilateral cataracts and especially cortical cataracts are presumed hereditary (see fig. 1 and 2), except:
1. Cases where there is evidence that the cataract is associated with trauma, inflammation, metabolic disease, nutritional deficiencies or old age (senile cataract; generally more localized and whiter densities than the normal, diffuse sclerosis of the lens). These senile cataracts generally start in large breeds after 7, in medium breeds after 9 and in small breeds after 11 years of age (large e.g.: Great Dane (Deutsche Dogge), Leonberger; medium e.g.: Labrador retriever, E. C. Spaniel; small e.g.: Dachshund, min. Schnauzer). This also means that, if no ECVO-eye examination reports are available from the period before that year it is not always possible to distinguish these senile cataracts from hereditary cataract. In case of doubt, the case should be examined at the next panel meeting or given “affected” for presumed hereditary cataract.
2. Cases with minor, clearly circumscribed cataracts e.g. located in the (posterior) suture lines (other than those specifically described to be hereditary), or distinctly in the nucleus e.g. fibreglass/crystal-like (see fig. 3) cataracts in

	the nucleus (not to be confused with pulverulent-like cataracts, see fig. 4), or located, in/on (the back) of the posterior capsule as whitish "scar-ghosts" of the tunica vasculosa lentis, or in/on the anterior capsule associated with persistent pupillary membrane. In case of doubt (e.g. very minor cataracts in the cortex, in the posterior pole etc., only barely visible by the naked eye (thus not with a microscope), using a slit lamp light beam, at least suspicious is given. This means the animal displays minor, but specific signs of the inherited disease(s) mentioned. Further change may confirm the diagnosis. Re-examination in .... months is advised. At least 6 months, but usually 12 months later, the animal is re-examined, or, preferably examined at a panel meeting for further judgement. 3. Unilateral minor, circumscribed cataracts located in the cortex, (such as e.g. punctate cataracts), developing at the age of 6 years, or later (with proof that the animal was unaffected at 5 years of age) are given “suspicious” and re-examination after 12 months. If there is no progression at that re-examination, the animal can be given unaffected for presumed hereditary cataract. To describe the type of cataract, the general box for cataract and, if available, the specifying box for the type of cataract should be ticked. If there is e.g. a cortical and a nuclear cataract, all three boxes are ticked. If there is e.g. a punctate cataract or a posterior polar cataract, (which both are generally cortical), the specifying box for that type is also to be ticked. For the remaining typical cataracts, e.g. like posterior suture line, pulverulent, etc., the box “other” is to be ticked, and further described in the “comments” area.
Choroidal hypoplasia (CH) *[or chorioretinal dysplasia (CRD)]*	In cases where the animal displays clinical features which could possibly fit this entity, but the changes are not specific enough, the result of the examination is: ‘undetermined’. In such cases the breeder/owner is advised to define the status of the animal by e.g. DNA testing.
Collie eye anomaly (CEA)	In cases where the animal displays clinical features that could possibly fit this KP-HED, but the changes are not specific enough, the result of the examination is: ‘undetermined’. In such cases the breeder/owner is advised to distinguish the status of the animal by e.g. DNA testing.
Corneal dystrophy	KP-HED. Known HED in the Siberian Husky (superficial) and in the Labrador retriever (macular).
Distichiasis/ectopic cilia	Presumed inherited eye disease Single or multiple hairs (cilia) from an abnormally located hair follicle in the eyelid margin, usually growing from or in between the Meibomian glands, and arising from the Meibomian duct openings, or emerging through the eyelid conjunctiva which may cause ocular irritation. The defect is due to abnormal differentiation of a tarsal gland. Distichiasis usually occurs at an early age (< 1-2 years), but may occur any time in life. No further details, such as e.g. mentioning the number of

hairs, or encircling distichiasis or ectopic cilia are to be written on the form.
In chapter 8, The veterinary ophthalmologists' breeding advice, the general advice for distichiasis/ectopic cilia is: "optional", but in severe cases: "no breeding". Thus in case of e.g. hard, stiff hairs, or ectopic cilia distinctly irritating the cornea, the examiner will also tick the box: "severe' in the comment area.

Entropion/trichiasis — No further details such as e.g. deleting or encircling entropion or trichiasis are to be mentioned on the form. The descriptive area can be used for details about the entity/-ies. In chapter 8, The veterinary ophthalmologists' breeding advice, the general advice for entropion/trichiasis is: "optional", but in severe cases: "no breeding".

Ectropion/macroblepharon — No further details such as e.g. deleting or encircling ectropion or macroblepharon are to be mentioned on the form. The descriptive area can be used for details about the entity/-ies. In chapter 8, The veterinary ophthalmologists' breeding advice, the general advice for ectropion/ macroblepharon is: "optional", but in severe cases: "no breeding".

Intraocular pressure (IOP) — In the ECVO certified examination, only the applanation/ rebound tonometric values of Tonopen, Tonovet and MacKay-Marg are currently accepted. The method used is mentioned in the certificate.

Macroblepharon — Fissure length (stretched) in dog over 40 mm.

Microblepharon — Fissure (stretched) in the dog less than 25 mm.

Micropapilla — Difficult to differentiate from hypoplasia with vision impairment. For this reason on the Certificate, the entity is ticked as a KP-HED.

Multiple ocular anomalies (two or more): To be ticked at number "7. Other", on the certificate. The anomalies found can be e.g. microphthalmia, iris hypoplasia, persistent pupillary membranes, lens anomalies, posterior segment colobomas or other developmental defects. The anomalies found are to be specified in the descriptive comments field.

Pectinate ligament — Three predominant types of involvement of the angle are distinguished, thus giving the owner and/or the breed club/ society the opportunity to select animals on severity of the defect (See figures 11-13).
1. Fibrae latae **(**FL; in which the normal part of the pectinate ligament fibre is too short and the abnormal part is broadened; also described as broad bands;
2. Laminae (LA; plates or sheets of continuous tissue, with very short remaining fibres in the angle);
3. Occlusio (OC; pectinate ligament completely closed, with flow holes, and narrowed angle and/or shallow anterior chamber;

If more than one type of abnormality is present, then both or all three relevant boxes are ticked.
If the pectinate ligament (P.L.) displays a few (less than 25% of the P.L.) very mild changes, that could possibly fit a pectinate ligament abnormality (P.L.A.), it is evaluated as 'unaffected';
If the pectinate ligament (P.L.) displays clinical features (over approximately 25-50% of the P.L.), that could possibly fit a pectinate ligament abnormality (P.L.A.) it is evaluated as: 'undetermined'; If more than 50% of the P.L. displays minor, but specific clinical signs of P.L.A. (fibrae latae or too short and/or broadened fibers) it is evaluated as 'affected' (mildly). If abnormalities like laminae or occlusio are found, it is evaluated as 'affected'. The most severe type of involvement in both eyes is used for clinical diagnosis. Severity is expressed in the section for descriptive comments. The severity of laminae or occlusio can never be less than moderate or severe. If "occlusio" is present over more than 25 % of the angle it is evaluated as: 'severe'.

PHTVL/PHPV KP-HED. Known hereditary disease in the Dobermann and the Staffordshire Bull Terrier. Minor, yellow-brown dots of fibrous tissue remaining retrolentally, more or less centrally on the posterior capsule of the lens (See fig. 21) are ticked as grade 1. These grade 1 dots are not to be confused with scattered pigment, retrolental near or on the posterior capsule of the lens. If they are unilateral, and of minimal degree, 'undetermined' is ticked. Unilateral or bilateral severe forms are ticked as 'affected'.

Persistent hyaloid artery — Severe, e.g. if causing visual impairment

Persistent pupillary membrane (PPM) — Tiny, more or less triangular shaped dots, centrally, on the anterior capsule of the lens. These are drawn in the figures in the "drawing area" and are not ticked in the 'undetermined' or 'affected' boxes in the Results area. Remnants of the pupillary membrane, which are not distinctly visible on the iris surface/collarette (using 10 x magnification) **after** pupil dilatation, are not mentioned on the form. Other forms of PPM, e.g. still distinctly present after pupil dilatation, crossing the pupil, corneal, or with lens involvement, are ticked in the box for 1. PPM: "affected" and the respective box of other parts involved. Areas which can be involved are: retrocorneal (boxes PPM and cornea); strands from cornea to iris (boxes: PPM, cornea and iris); from iris to iris (boxes PPM and iris); iris to lens (boxes: PPM, iris and lens), connected to areas of cataract (also the box for congenital cataract is ticked); strands connected to a sheet/"spider web" of tissue (lamina) in the anterior chamber (boxes PPM, lamina and other parts involved are ticked).

Retinal dysplasia (RD) — In cases where the animal displays clinical features that could possibly fit this specific KP-HED, but the changes are not specific enough, the entity is evaluated as: 'undetermined'.

Vitreal strands	To be recognized as vitreous degeneration only if there are no signs of lens luxation (less curving of the face of the iris, iridodonesis, etc.). In case of doubt, 'suspicious' for LLX is ticked and the animal is re-examined for LLX after a minimum of 3 months. Tonometry before dilation is recommended.

Figures of the KP-HED´s are found on the ECVO website at http://ecvo.org/inherited-eye-diseases/images-for-panellists