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. A tick box is provided for “mild” and “severe”, enabling the examiner to indicate if the expression of the respective KP-HED is severe (see detailed description of KP-HEDs below).

Cautious use of the grading is recommended as for certain diseases, the indication of severity will influence the veterinary ophthalmologists advice. Guidelines for using the grading, where applicable, can be found in conjunction with the description of a given diagnosis in this chapter.

In this section, the examiner should describe any findings in the eye and adnexa, either KP-HED or other.

**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. The terminology for the diseases as given in chapter 5. Definitions of this manual are to be used. These disease names are also used in “roll down” menus in the computerized forms. These are:

- Anophthalmos
- Choroidal coloboma
- Choroidal hypoplasia in Non-Collie breeds
- Congenital stationary night blindness
- 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
Lenticularus
Lentiglobus
Macrophthalmos
Microphthalmos
Microblepharon
Microphakia (Nanophthalmos): use Microphthalmos
Nictitating membrane, eversion of the cartilage
Nictitating membrane, prolapse of the gland
Multiple ocular anomalies for 2 or more anomalies, to be specified in the comment field
Optic disc coloboma
Retinal coloboma
Retinal dystrophy/ RPE65 null mutation (include ERG results)
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:

Other presumed hereditary retinal degenerations
Canine multifocal retinopathy (CMR)
Ceroid lipofuscinosis (CLN):
Chorioretinopathy, pigmentary (e.g. in Chinese crested)
Iris melanoma
Keratitis: Chronic superficial keratitis (CSK)/Pannus
Keratitis, Punctate (in specific breeds e.g. Dachshund)
Keratoconjunctivitis sicca (KCS; in specific breeds e.g. WHWT, Chinese crested, LH Dachshund, Cavalier King Charles Spaniel)
Ocular melanosis (do not use Glaucoma – pigmentary; e.g. Cairn Terrier)
Uveitis, pigmentary (e.g. in Golden retriever)
Uveal cyst
Vitreous degeneration (without any sign of lens luxation)
Vitreous strands/vitreous prolapse (without any signs of lens luxation)

General guidelines and considerations:

For litter examination a separate Certificate should be issued for each animal examined. The examination can only be performed after permanent identification, e.g. by microchip implantation, 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 to 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 mandatory. 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 possible breeds with special concern for the below listed KP-HED to be examined before dilation:

1. **Iridocorneal angle abnormality using gonioscopy**:
   - American Cocker Spaniel
   - Bouvier des Flandres
   - Bassets (all)
   - Bloodhound
   - Border Collie
   - Chow Chow
   - Dandie Dinmont Terrier
   - Dutch Shepherd (Rough Hair)
   - English Springer Spaniel
   - Entlebucher Mountain Dog
   - Flat Coated Retriever
   - Golden Retriever
   - Siberian Husky
   - Leonberger
   - Magyar Viszla
   - Samoyed
   - Tatra

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
Dalmatian
Rottweiler

4. Lens luxation (PLL)/KCS/vitreous degeneration/ocular melanosis using slitlamp biomicroscopy

Chinese Crested dog (PLL, vitreous degeneration, KCS)
English Bulldog (KCS)
Lancashire Heeler (PLL)
Longhaired Dachshund (KCS)
Lhasa Apso (KCS)
Pug (KCS)
Shih Tzu (KCS)
Cairn Terrier also (ocular melanosis)
Small Terrier breeds (PLL)
West Highland White terrier (KCS)
Other breeds listed in Chapter 10 for PLL

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)]

CH in non-Collie breeds: At number “7: Other”: Choroidal hypoplasia is written (online: is used), and the box affected is ticked. In cases where the Non-Collie 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. The box “Affected – other” has to be specified in the comment area of the ECVO certificate (retinal detachment or –haemorrhage).

**Corneal dystrophy**

Corneal dystrophy is to be ticked affected at no. 14 Corneal dystrophy, and the details described in the field Descriptive comments.

Only if endothelial dystrophy (bilateral progressive diffuse, deep corneal edema, e.g. in Chihuahua, Boston Terrier etc) or macular dystrophy (bilateral diffuse haziness of the cornea with multiple whitish/grey macula like lesions throughout the corneal stroma, periphery slightly less affected, e.g. in Labrador Retriever) or severe forms of stromal dystrophy (e.g. in Siberian Husky), is recognized , the examiner will also tick the box: “severe’ in the comment area.:  

**Distichiasis/ectopic cilia**

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.

Only if there are clinical signs of corneal irritation such as detritus on the distichia, corneal edema, corneal vessels, defects or pigmentation at the location of the distichia; hard stiff distichia and/or ectopic cilia, 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.

Only if there are clinical signs of corneal irritation such as detritus on the lid hairs, corneal edema, corneal vessels, defects or pigmentation at the location of the entropionised lid margin, the examiner will also tick the box: “severe’ in the comment area.

**Ectropion/macroblepharon**

No further details such as e.g. deleting or encircling ectropion or macroblepharon are to be mentioned on the form. Fissure length (stretched) in dog over 40 mm.

Only if the stretched fissure length is over 45 mm, or there are signs of corneal changes due to the exposure or chronic irritation caused by the ectropion/macroblepharon, the examiner will also tick the box: “severe’ in the comment area. **Hyaloid artery, persistent**

If the AHP is distinctly visible by the naked eye (thus not by microscope), at number “7: Other”; Hyaloid artery, persistent is written (online: is used) and the box “affected” is ticked. Only if there is a Mittendorf’s dot with signs of capsular cataract and/or a Bergmeister papilla with a patent
vascular or non-vascular fibrous strand in between them, at number “7”: Other: Persistent hyaloid artery is written (online: is used) and the box “affected” plus the box: “severe” in the comment area are ticked.

**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.

**Iridocorneal angle abnormality (ICAA):**

Two predominant types of involvement of the angle are distinguished. The pectinate ligament (PL) and the iridocorneal angle (ICA) width are evaluated by gonioscopy in its extent of 360 degrees, thus giving the owner and/or the breed club/society the opportunity to select animals on severity of the defect (See figures 11-13).

**Pectinate Ligament (PL)** consists of thin/filamentous fibres from iris base to its insertion at the cornea.

Fibrae latae (FL): fibres with a confluent (broad) base and shortened thin insertions at the cornea or thick fibres (<5 fibres)

Laminae (LA): plates or sheets of continuous tissue (>5 fibres), with or without flow holes

**Iridocorneal angle (ICA) width:**

Open: PL length (A) is equal to or more than 1/3 of B; A ≥ 1/3 of B

Narrow: PL length (A) is smaller than 1/3 of B; A < 1/3 of B (visible length of PL is severely reduced)

Closed: collapsed/closed angle - PL not visible

A = length of PL; B = distance from the origin of the PL to the anterior surface of the cornea at the transection area

**Grading of PLA (FL = fibrae latae, LA = laminae):**

- 0-50% FL = unaffected
- >50-100% FL and/or <25% LA = affected mild
- 25-50% LA = affected moderate
- >50% LA = affected severe

**Grading of ICA width:**

- Open = normal
- Narrow = affected moderate
- Closed = affected severe
• Grading of the ICAW

Terminology:

<table>
<thead>
<tr>
<th>Closest to the Angle</th>
<th>Ratio A/B:</th>
<th>PL Length</th>
</tr>
</thead>
<tbody>
<tr>
<td>Closed</td>
<td>PL not visible</td>
<td>A ≥ 1/3 of B</td>
</tr>
<tr>
<td>Narrow</td>
<td>A &lt; 1/3 of B</td>
<td>A ≤ 1/3 of B</td>
</tr>
<tr>
<td>Open</td>
<td>A ≥ 1/3 of B</td>
<td>A = 1/3 of B</td>
</tr>
</tbody>
</table>

Open: PL length (A) is equal to or more than 1/3 of B; A ≥ 1/3 of B
Narrow: PL length (A) is smaller than 1/3 of B; A < 1/3 of B (visible length of PL is severely reduced)
Closed: PL not visible = collapsed/closed angle


**Iris hypoplasia**

Congenital, uni- or bilateral thinning or absence of iris (sphincter) tissue or colobomatous defects due to failure in closure of the optic fissure. At number “7: Other” Iris hypoplasia is written (online: is used), and the box affected is ticked. Only if uni- or bilateral iris tissue is missing or failed to develop (developmentally colobomatous) e.g. in one of the specific breeds Australian Shepherd, Dalmatian, Rottweiler, the examiner will also tick the box: “severe” in the comment area.

**Iris melanoma**

If there are clinical signs of an iris melanoma, at 18. Other: Iris melanoma is written (online: is used), and the box affected is ticked.

**Keratoconjunctivitis sicca (KCS)**

The STT should be done to measure tear production in case of doubt or with clinical signs of KCS, especially in breeds known to be affected (e.g. Cavalier King Charles Spaniel, Chinese Crested Dog, Long-haired Dachshund, English Bulldog, West Highland White Terrier–If the STT is below 10 mm and there are clinical signs of KCS: at 18. Other: “Keratoconjunctivitis Sicca” is written (online: is used), and the box affected is ticked or in case of doubt if the abnormality is KP-HED, the box “suspicious” is ticked with re-examination in 6 months.
**Microblepharon**

Fissure (stretched) in the dog less than 25 mm in an adult dog. At number “7: Other”: Microblepharon is written (online: is used), and the box affected is ticked. Only if an uni- or bilateral microblepharon with a stretched fissure length of less than 20 mm is diagnosed, the examiner will also tick the box: “severe” in the comment area.

**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):** At number “7: Other”: Multiple ocular anomalies is written (online: is used), and the box affected is ticked. 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 abnormality**

See Iridocorneal angle abnormality (ICAA)

**PHTVL/PHPV**

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 PHTVL/PHPV affected, and the specifying box 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.

The severe forms (grades 2–6) usually occur bilaterally and may lead to visual problems. A plaque of white fibrovascular tissue can remain on the back of the posterior capsule, accompanied by grade 1 retrolental dots. In addition, other parts of the hyaloid system can persist: lenticonus, or even more severe malformations of the lens such as pigment or blood in the lens or behind it, lens hypoplasia, spherophakia, elongated ciliary processus etc.; and/or microphthalmia may be present. In the grade 2-6 forms, cataract develops, usually beginning centrally. ) are ticked as PHTVL/PHPV affected, Unilateral or bilateral grade 2-6 forms are ticked as PHTVL/PHPV ‘affected’ and the specifying box as grade 2-6.

**Persistent hyaloid artery**

See Hyaloid artery, persistent

**Persistent pupillary membrane (PPM)**

Remnants of the pupillary membrane, 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 in the anterior chamber (boxes PPM, lamina and other parts involved are ticked). 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. 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.

**Retinal dysplasia (RD)**

Linear (vermiform), triangular, curved or curvilinear foci of retinal folding that may be single or multiple seen ophthalmoscopically, the boxes 4: Retinal dysplasia and (multi)focal are ticked

In puppy, linear or round juvenile folds, usually in the peripapillary area, may be observed as a result in inequity in the relative growth rates of the optic cup and these folds resolve as the animal matures. These folds are not accurately referred to as dysplasia and should be ticked “unaffected”, but can be described in the comments area. In the English Springer Spaniel, Golden Retriever, Labrador Retriever and Samoyed these juvenile folds are considered as retinal dysplasia (RD) and should be ticked “undetermined” or “affected”.

Irregularly, horseshoe- or bladder-like shaped areas of abnormal retinal development, most often in the central part of the tapetal area of the fundus, in close association with the dorsal retinal vasculature, containing both areas of thinning and areas of elevation representing focal retinal detachment and areas of retinal disorganization seen ophthalmoscopically the boxes 4: Retinal dysplasia and geographical are ticked

Severe retinal disorganization associated with total separation (detachment) of the retina seen ophthalmoscopically associated with partial or complete vision impairment, the boxes 4. Retinal dysplasia and total are ticked

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’.

**Uveal Cysts**

If there are only 1-3 free separate floating cysts and no connected signs of glaucoma and/or uveitis at: no. 18 Other uveal cyst is written, and the box affected is ticked.

Only if there are more cysts and/or cysts developing from the back of the
iris/posterior chamber, and/or signs of uveitis and/or glaucoma, the examiner will also tick the box: “severe” in the comment area. Tonometry before dilation is recommended.

**Vitreous degeneration:**
Vitreous changes consistent with breakdown of the vitreous hydrogel (e.g. visible liquefaction, syneresis, asteroid hyalosis or synchysis scintillans). Presently, there is not sufficient scientific proof how to discriminate between mild and severe, thus at no 18, Other, Vitreous degeneration is written (used) and the box affected ticked.

**Vitreal strands/Vitreous prolapse**
To be recognized as vitreous degeneration or –prolapse 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 lens luxation is ticked and the animal is re-examined for lens luxation after a minimum of 3 months. Tonometry before dilation is recommended.

*Figures of the KP-HED are found on the ECVO website at [http://ecvo.org/inherited-eye-diseases/images-for-panellists](http://ecvo.org/inherited-eye-diseases/images-for-panellists)*