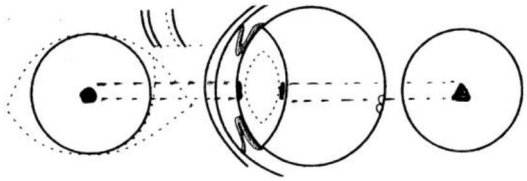
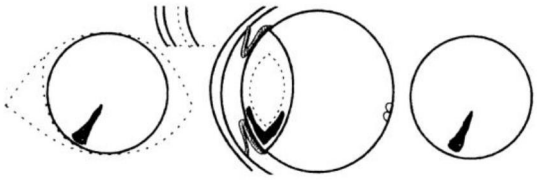
## Guidelines for the use of the ECVO certificate in the Known and Presumed Hereditary Eye Disease scheme (KP-HED)

**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.

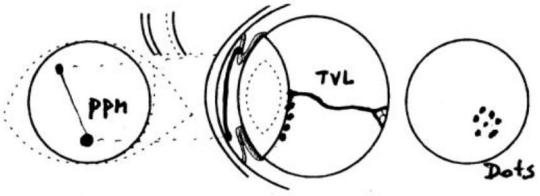
**Section Examination**:
The drawings in the middle of the form are used to draw and position any changes found. The circles on the left can be used for the cornea, e.g. to position corneal dystrophy, or for the anterior capsule of the lens. The dotted lines around the first circle represent contours 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, and 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 ophthalmologist’s 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(es) for the KP-HED (1-6, 11-17) on the certificate and the specifying box, if available (e.g. for type or grade) are ticked.
*If there is no specific box available on the certificate* for the KP-HED, the box at number “7. Other” and/or at number “18. Other” is to be ticked and the definition name of the disease (in the list in chapter 5) is written (online: is used). *Only if there are more than one KP-HEDs present which are not listed* in the results field under no 1-6 and no 11-17, the box “affected” at “7. Other” and/or at “18. Other” is ticked and the term "Multiple other KP-HEDs" is written (online: is used); the KP-HED must also be specified in the comment field using the definition name in the list in chapter 5.

For number “**7. Other”**: **known and presumed hereditary eye anomalies** (congenital/developmental, non-progressive) that are not yet mentioned on the form are mentioned here. The terminology for the diseases can be found in "Definitions", Chapter 5, which are to be used (and are listed in the drop-down menu in the computerized forms). These are:

- Anophthalmos
- Choroidal coloboma
- Choroidal hypoplasia in Non-Collie breeds
- Congenital stationary night blindness (CSNB)
- Dermoid
- Eyelid coloboma
- Exophthalmos due to shallow orbit
- Persistent hyaloid artery (PHA)
- Iris hypoplasia
- Lacrimal punctum atresia/micropunctum
- Lens hypoplasia
- Lenticonus
- Lentiglobus
- Macrophthalmos
- Microphthalmos
- Microblepharon

- Microphakia
- Nictitating membrane, eversion of the cartilage
- Nictitating membrane, prolapse of the gland
- Multiple other KP-HED (2 or more KP-HED anomalies, to be specified in the comment field)
- Optic disc coloboma
- Posterior segment exam not possible
- Retinal coloboma
- Retinal dystrophy/*RPE65* null mutation (e.g. Briard)
- Scleral coloboma

Note: If there is a congenital lack of tissue in the iris and lens, the term "hypoplasia" is used: iris hypoplasia, lens hypoplasia. Reason: Iris tissue can be absent in full-thickness, but also only partially (hypoplastic); The lens equator may have a flattened curvature due to abnormal development of zonular fibers or ciliary processes.
For congenital absence of tissue of the eyelid, retina, choroidea, sclera or optic nerve/papilla use the term “coloboma”, e.g. eyelid coloboma, retinal coloboma, choroidal coloboma, scleral coloboma and/or optic nerve coloboma.

For number "18. Other ": KP-HEDs that are not considered to be congenital / developmental or progressive and are not yet mentioned on the form are mentioned here. The terminology for the diseases can be found in "Definitions", Chapter 5, which are to be used (and are listed in the drop-down menu in the computerized forms). These are:

- Other presumed hereditary retinal degenerations
- Retinopathy, Canine multifocal (CMR)
- Ceroid lipofuscinosis (CLN):
- Chorioretinopathy, pigmentary (e.g. in Chinese crested)
- Glaucoma, primary
- 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)
- Multiple other KP-HED (2 or more acquired KP-HED, to be specified in the descriptive comment field)
- Ocular melanosis (do not use Glaucoma –pigmentary; e.g. Cairn Terrier)
- Posterior segment exam not possible
- Retinal Pigment Epithelial Dystrophy (RPED)
- Uveitis, pigmentary (e.g. in Golden retriever)
- Uveal cyst(s)
- Uveodermatologic Syndrome (UDS)
- 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

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Some recommendations and details in regards to ticking of the ECVO certificate of eye examination:

***Cataracts***

1) **Classifications:**
   - **Refraction discontinuity zones:** fine (light-grey) regular circular/bend lines due to different refractive indices of the fibers of the embryonic, fetal, juvenile and adult nucleus and cortex;
     clinical significance: none
   - **Nuclear sclerosis**: is a translucent optical turbidity of the lens nucleus due to aging; appearance: blue-gray shade of the central area of the lens; translucent: the fundus can be viewed without restriction using ophthalmoscopy;
     clinical significance: myopia
   - **Cataract:** is an opacity (generally whitish) in different shapes and sizes in the lens nucleus, cortex or capsule; it is resulting from pathologic changes in lens protein composition or disruption of lens fiber arrangement;
     clinical significance: The clinical significance is influenced by the extent, density and location of the opacity, as well as its potential to progress, which leads to scattering of incident light, reduced illumination, reduced contrast sensitivity, increased glare, degraded color vision, and loss of visual acuity and visual function.

     **Classification according to aetiology:**

   - **Primary cataracts:** all bilateral or unilateral cataracts and especially cortical cataracts are known or presumed hereditary eye diseases (KP-HED; except secondary cataracts)
   - **Secondary cataracts:** cataracts known to be caused by physical influences (trauma, electric, irradiation), ocular inflammation, metabolic diseases, nutritional deficiencies, age, intoxication or another KP-HED (i. e. attachment point of PPM, PHA or as part of PHTVL or sequelae of PRA) should NOT be ticked as KP-HED «cataract» but should be mentioned in the comment field (as it is not a primary cataract, but a result/ consequence of another disease).

**Classification according to age of onset:**

- **Congenital cataracts:** If cataracts are observed in the period between birth and the 8th week of age the entity is ticked "affected" at "3. Cataract (congenital)"; if diagnosed later in life but there is distinct indication the cataract is congenital in origin (e. g. in microphthalmos, in the lens cortex adjacent to PPM, or PHA) the entity is ticked "affected" at "3. Cataract (congenital)", except in PHTVL/PHPV, where the cataract is part of the entity. If there are also signs of juvenile or adult cataract (e. g. post. pol. or cortical cataract not adjacent to the insertion of the PPM or PHA) also tick "affected" at "15. Cataract (non-congenital)". If lens opacities are seen in puppies (up to 8 weeks of age) which are not described as a known or presumed hereditary congenital cataract, tick at "3. Cataract (congenital)" "undetermined" and recommend re-examination in 1-3 months. *See also Chapter 3, paragraph 9. 1. "Undetermined" (no. 1-7 on the certificate) cases.*
- **Juvenile and adult cataracts:** cataracts developed at older age (after 8$^{th}$ weeks of age) are ticked "affected" at "15. Cataract (non-congenital)".
- **Senile cataracts*:** are parts of the aging process. These lesions are frequently preceded by the formation of a dense nuclear sclerosis. Opaque streaks extend from the nucleus toward the cortical equator like spokes of a wheel. Senile cataracts are ***not*** ticked as KP-HED "cataract"
  * in large breeds after about 7 years, in medium breeds after about 9 years and in small breeds after about 11 years of age;
- This also means that, <u>if no previous ECVO-eye examination reports are available</u> from the period before that year it is not always possible to distinguish these senile cataracts from hereditary cataracts. In case of doubt, the dog should be given "suspicious" for KP-HED cataract and a final decision should be made by a minimum of three members of the National Panel or the (deputy) Chief panellist. *See also Chapter 3, paragraph 9. 2. "Suspicious" (no. 11-18 on the certificate) cases.*

**IMPORTANT:** typical <u>KP hereditary cataracts</u> (e. g. posterior polar) and <u>pre-existing cataracts are not changed into a senile cataract in old age or with age</u>.

**Classification according to location:**

- **Cortical cataracts:** any opacity in the anterior and/or posterior cortex unilateral or bilateral (except the posterior polar cataract and those listed under "other opacities")
- **Posterior polar cataract:** is a subtype of the cortical cataract, it presents as a distinctive triangular (sometimes discoid) plaque situated in the central posterior cortex, in general adjacent to the posterior capsule. Sometimes there is a smaller satellite rosette lesion adjacent to the central opacity. It can be stationary as well as progressive (progression may begin at any age). In the progressive type, whitish opacification changes take place in the posterior cortex in the form of radiating rider opacity.
- **Nuclear cataracts:** any whitish opacity in the nucleus (embryonal, fetal, juvenile, adult); exceptions: fiberglass like and pulverulent cataracts (see Other lens opacities)

- **Other lens opacities (cataracts – other):**
  Certain lens opacities/cataracts can occur frequently in a certain breed of dog (therefore presumed hereditary), but are considered regarding breeding "optional" or of low priority because they usually remain clinically less relevant. These opacities vary in size, location and transparency: some opacity is whitish but very small (e. g. punctate, suture tips, suture line), others are almost transparent but more extensive (e. g. fiberglass like or pulverulent, nuclear ring).
  Clinical significance: these lens opacities usually remain unchanged or limited and have no clinically relevant effect on vision. These lens opacities are summarized under "15. Cataract (non-congenital)" – "other" and specified in the comment field.
    - Punctate: one or more *clearly defined* whitish dot like opacities located in the cortex or nucleus
    - Suture line tips: *clearly defined* whitish small linear opacities at the ends of the suture lines
    - Suture line: *clearly defined* whitish line or dots in the cortex that form an upright or inverted Y; sometimes faint dotted circular opacities can be seen in its center.
    - Nuclear ring: *delicate semi-translucent irregular shaped* more or less circular structure in the nucleus
    - Nuclear fiberglass-like/pulverulent: Fiberglass or crystal-like opacities in the nucleus or scattered fine pulverulent granules parallel to the suture lines in the posterior nucleus and later with fibrillary opacities in the entire fetal nucleus, which may become dense and extending into the adult nucleus. These nuclear opacities are generally bilateral and do not impair vision significantly.

**2) Further instructions for filling out the form**

- To describe the type of cataract, the general box for cataract "affected" and, the specifying box for the type of cataract are to be ticked.
  - congenital cataracts (any location): tick at "3. Cataract (congenital)" "affected"
  - non-congenital cataracts: tick at "15. Cataract (non-congenital)" "affected" and the specifying box
    - "cortical" for cataracts in the cortex (including subcapsular, perinuclear, equatorial
    - "nuclear" for cataracts in the nucleus
    - "post. pol" for distinctive triangular (sometimes discoid) cataracts in the central posterior cortex, in general adjacent to the posterior capsule
    - "other" for the following lens cataracts: punctate, suture line tip, suture line, nuclear ring or nuclear fiberglass/pulverulent; tick at "15. Cataract (non-congenital)" "affected" and the box "other"; in addition, the type (name) of cataract must be ticked (or written) in the field "Descriptive Comments" at "15. Cataract – other"
- If a dog has **2 (or more) different types of cataract**, all relevant types of cataracts are ticked: e. g. a cataract in the nucleus and in the cortex, tick at "15. Cataract (non-congenital)" the boxes "affected" and the specifying boxes "nuclear" **AND** "cortical"; the same is true, if a dog has a posterior polar cataract and a punctate cataract: tick at "15. Cataract (non-congenital)" the boxes "affected" and the specifying boxes "post. pol."

**AND** "other". In the field "Descriptive Comments" the box "punctate" must be ticked (or written) at "15. Cataract – other".

- If there is a distinct (well defined) **post. pol. cataract**, without signs of spreading into the remaining cortex, tick at "15. Cataract (non-congenital)" the box "affected" and the specifying box "post. pol";
- If the **post. polar cataract** extends into the adjacent cortex (and is therefore progressive), tick at "15. Cataract (non-congenital)" the box "affected" and the specifying boxes "post. pol." **AND** "cortical";
- If there are cataracts in the cortex associated with the anterior or posterior suture lines tick at "15. Cataract (non-congenital)" the box "affected" and the specifying box "cortical". However, if there are distinct delicate suture line cataracts tick at "15. Cataract (non-congenital)" the box "affected" and the specifying box "other", and in the comment area at "15. Cataract other": "suture line (tip)".
- In a **total cataract:** tick at "15. Cataract (non-congenital)" the boxes "affected" and the specifying boxes "cortical" **AND** "nuclear"; tick also "affected" **and** write at "18. Other: Posterior segment exam not possible"
- **The following opacities on the lens capsule are *not* to be ticked as cataract:**
    - whitish spiderweb like opacities on the posterior lens capsule (uni- or bilateral)
    - PPM, PHA, PHTVL grade1: If the opacity on the lens is limited to the insertion/attachment of the relevant structure on the capsule, do ***not*** tick the box for cataract (congenital). Only, if a whitish opacity extends into the lens cortex adjacent to this, also tick the box for cataract (congenital). If there are other lens opacities not adjacent to the relevant structure, which might not be congenital, tick the relevant box at "15. Cataract (non-congenital)".
- **PHTVL grade 2-6:** in PHTVL/PHPV grade 2-6 the cataract and other lenticular abnormalities are part of the entity and are ***not*** to be ticked at "3. Cataract" and/or "7. Other".
- **Drawings (or photographs):** It is strongly recommended to draw the cataract in the "pre-drawings" on the certificate (see separate instructions for drawing and filling the form).
- **Minor imperfections versus cataract:** The cut-off point is: those not visible with the naked eye in retro illumination using a slit lamp light beam are considered as minor imperfections and those visible are to be mentioned as cataracts.
  In case of doubt (e. g. very minor cataracts in the cortex, in the posterior pole etc.), if cataracts are only barely visible with the naked eye (thus not only with a microscope), in retro illumination using a slit lamp light beam, at least "cataract – suspicious" is given. This means the animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis. Re-examination in "…. months" is necessary. At least 6 months, but usually 12 months later, the animal is re-examined, or, preferably examined at a panel meeting or by the Chief panellist for further judgement. *See also Chapter 3, paragraph 9. 2. "Suspicious" (no. 11-18 on the certificate) cases.*

***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 dogs of a relevant breed that were not examined until after the 8$^{th}$ week of age, CEA can be masked ("go normal") later in life. 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 "14. Corneal dystrophy", and the details described in the field Descriptive comments.
In cases of 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, also the box "severe" is to be ticked 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.
Tick "affected" at "13. Distichiasis/Ectopic cilia". 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 recognized, also the box "severe" is to be ticked in the comment area.

***Entropion/trichiasis***

Tick "affected" at "11. 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, also the box "severe" is to be ticked in the comment area.

***Ectropion/macroblepharon***

If the fissure length (stretched) in dog is over 40 mm tick "affected" at "12. Ectropion/ Macroblepharon". No further details such as e.g. deleting or encircling ectropion or macroblepharon are to be mentioned on the form. Only if the stretched fissure length is over 45 mm and/or there are signs of corneal changes due to the exposure or chronic irritation caused by the ectropion/macroblepharon, also the box "severe" is to be ticked in the comment area.

***Exophthalmos due to shallow orbit***

Exophthalmos due to shallow orbit is usually seen in combination with macroblepharon. If the sclera is visible in two or three quadrants in the straight position of the globe, with or without strabismus divergens (without prior pathology) at "7. Other": **"**Exophthalmos due to shallow orbit" is written (online: is used) and the box "affected" is ticked. Only if the sclera is visible all around (with a normal-sized globe), also the box "severe" in the comment area is ticked. In case of macroblepharon also tick at "12. Ectropion/ Macroblepharon" the box "affected".

***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 to be mentioned on 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.

**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

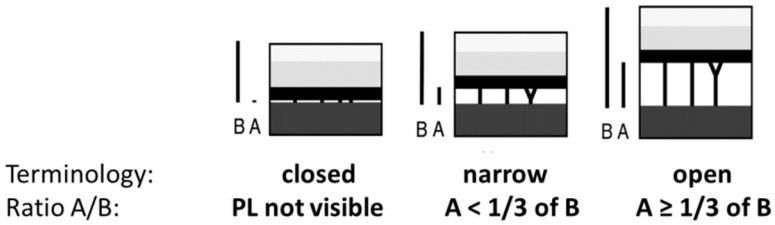
**Iridocorneal angle (ICA) width:** Open: PL length (A) is equal to or more than 1/3 of B; A ≥ 1/3 of B Narrow: PL length (A) is smaller than 1/3 of B; A < 1/3 of B (visible length of PL is severely reduced) Closed: collapsed/closed angle - PL not visible
*A = length of PL; B = distance from the origin of the PL to the anterior surface of the cornea at the transection area*

**Grading of PLA (FL = fibrae latae, LA = laminae):** tick at "8. ICAA" and the specifying box

– 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:** tick at "8. ICAA" and the specifying box

– Open = normal
– Narrow = affected moderate
– Closed = affected severe




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

Modified from publication: «Correlation of morphologic features of the iridocorneal angle to intraocular pressure in Samoyeds» Ekesten B, Narfström K. Am J Vet Res, vol 52, no. 11, November 1991, p 1875-1878.

***Iris hypoplasia***

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

***Iris melanoma***

If there are typical "clinical" signs of an iris melanoma (raised, black-brown lesion in the iris whose growth has been noted), at "18. Other": "Iris melanoma" is written (online: is used) and the box "affected" is ticked. If a small, non-raised pigmentation is noticed for the first time, and no information about an increase in size is available, at "18. Other": "Iris melanoma" is written (online: is used) and the box “suspicious” is ticked and re-examination in 6-12 months required; If not progressive, tick “unaffected” and write in “descriptive comments”: “Pigmented lesion on the iris – to be observed”; if the lesion is progressive, tick “affected”.

***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 “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, also the box “severe” is to be ticked in the comment area.

***Micropapilla***

Micropapilla is difficult to differentiate from hypoplasia with vision impairment. For this reason, on the Certificate, the entity is ticked as a KP-HED at “5. Hypoplastic-/Micropapilla” “affected”.

***Multiple other KP-HED anomalies (two or more)***

At "7. Other": "Multiple other KP-HED anomalies" is written (online: is used), and the box "affected" is ticked. The anomalies found can be e.g. microphthalmia, iris hypoplasia, lens anomalies, posterior segment colobomas or other developmental defects. The anomalies found are to be specified in the descriptive comments field. (See also page 2)

***Multiple other acquired KP-HEDs (two or more)***

At "18. Other": "Multiple other KP-HEDs" is written (online: is used), and the box "affected" is ticked. The acquired KP-HEDs found are to be specified in the descriptive comments field. (See also page 3)

***Persistent hyaloid artery (PHA)***

If the PHA is distinctly visible by the naked eye (thus not only by microscope) in retro illumination, at number "7. Other": "Persistent hyaloid artery" (PHA) is written (online: is used) and the box "affected" is ticked. Only if there is a Mittendorf's dot with signs of capsular cataract that goes beyond the insertion of the PHA 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.

***Persistent hyperplastic tunica vasculosa lentis/persistent hyperplastic primary vitreus (PHTVL/PHPV)***

Minor, yellow-brown hyperplastic dots of fibrous tissue remaining retrolentally, more or less centrally on the posterior capsule of the lens are ticked at "2. PHTVL/PHPV" "affected", and the specifying box as grade 1. If they are unilateral, and of minimal degree, "undetermined" is to be ticked.

**Exception:** Tiny scattered pigment dots (flat, not fibrotic or hyperplastic), retrolental near or on the posterior capsule of the lens (remnants of the posterior TVL): these are drawn in the figures in the "drawing area" and mentioned in the comment area and is not ticked "undetermined" or "affected" for PHTVL/PHPV grade 1.

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 and more severe malformations of the lens (such as lenticonus, pigment or blood in the lens or behind it, lens hypoplasia, spherophakia), elongated ciliary processes and/or microphthalmia may be present. Unilateral or bilateral forms of grades 2-6 are ticked at "2. PHTVL/PHPV" "affected" and the specifying box "grade 2-6". Cataract and/or other lenticular abnormalities are part of the entity and are therefore **not** ticked at "3. Cataract (congenital)" and/or at "7. Other".

***Pectinate ligament abnormality* – see *Iridocorneal angle abnormality (ICAA)***

***Persistent pupillary membrane (PPM)***

Remnants of the pupillary membrane, still distinctly present after pupil dilatation, from the iris collarette, with corneal, and/or with lens involvement, are ticked at "1. PPM" "affected" and the relevant box of other parts involved:

- **Strands from iris to iris:** boxes PPM and iris are ticked;
  Remnants of the pupillary membrane, which are not distinctly visible on the iris surface/collarette (using 10 x magnifications) after pupil dilatation, are not mentioned on the form.

- **Strands from iris to cornea:** boxes PPM, iris and cornea are ticked;
- **Retrocorneal remnants without strands, *only if substantial*** *(= visible with the naked eye)*, boxes PPM and cornea are ticked; minor (visible with 10x magnification only) retrocorneal remnants are drawn in the figures in the "drawing area" and are not ticked "undetermined" or "affected" for PPM.
- **Strands from iris to lens:** boxes PPM, iris and lens are ticked; *
- **Fibrotic (thickened, hyperplastic) more or less pigmented tissue remnants on the anterior capsule of the lens, without strands, *only if substantial*** *(= visible with the naked eye)*, boxes PPM and lens are ticked;*
  **Exception:** Tiny (flat, not fibrotic or hyperplastic) pigmented dots, centrally on the anterior capsule of the lens (PPM)**:** these are drawn in the figures in the "drawing area" and mentioned in the comment area as "pigmented dots, centrally on the anterior capsule of the lens" and are not ticked "undetermined" or "affected" for PPM.
- **Sheet/"spider web" of tissue in the anterior chamber with or without strands to the iris:** boxes PPM, lamina and other parts involved are ticked; *

* If the opacity on the lens is limited to the insertion of the PPM on the capsule, do NOT tick the box for cataract (congenital). Only, if a whitish opacity extends into the lens cortex adjacent to this, also tick the box "affected" for cataract (congenital). If there are other lens opacities not adjacent to the PPM, which might not be congenital, tick the relevant box at "15. Cataract (non-congenital)".

***Retinal dysplasia (RD)***

Linear (vermiform), triangular, curved or curvilinear foci of retinal folding that may be single or multiple seen ophthalmoscopically, the boxes at "4. Retinal dysplasia" and "(multi)focal" "affected" are ticked.
In puppies, 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 "affected" at "4: Retinal dysplasia" and "geographical" are ticked. Although it is a congenital disease, its manifestation might not be visible until after 8 weeks of age.

Severe retinal disorganization associated with total separation (detachment) of the retina seen ophthalmoscopically associated with partial or complete vision impairment, the boxes at "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 "18. Other": "uveal cyst(s)" is written (online: is used), and the box "affected" is ticked.

Only if there are several cysts and/or signs of uveitis and/or glaucoma also the box “severe” is to be ticked 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 “18. Other” “Vitreous degeneration” is written (online: is 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-panelists***

***Instructions (with pictures) for assessing specific KP-HEDs and completing the ECVO Certificate are available as PDF documents on the ECVO website***