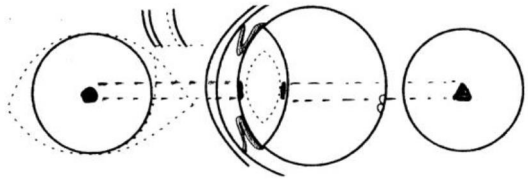
ECVO Manual: Chapter 6 - Guidelines (2024)

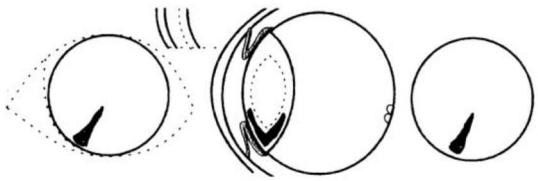
Guidelines for the use of the ECVO certificate in the Known and Presumed Hereditary Eye Disease scheme (KP-HED)

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 or these data are given by the on-line registry.
In the third section of the certificate (Fig. 2), the date of the examination and the microchip/tattoo check is specified. All animals presented for examination under the Scheme undergo a general examination of the eye and its adnexa. A mydriatic is used for the examination of the deeper structures. The minimum equipment to be used for the examination is a slit-lamp biomicroscope and a binocular indirect ophthalmoscope. If additional examination techniques (photography, tonometry without mydriatics, direct ophthalmoscopy, electroretinography, ocular ultrasound, STT and OCT) are used, this must be ticked and details of the device or measurement technique and measurement values must be provided in the online form. If the paper certificate is issued the same terminology is to be used, and specifications, and measurements have to be written in the descriptive comments area. Gonioscopy is an optional, additional examination and is to be ticked in the specifying box and must be performed before mydriasis. It is strongly recommended to examine every animal before dilation; lists of breeds with special concern for specific diseases e.g. persistent pupillary membrane, iris hypoplasia, lens luxation etc. are given below.

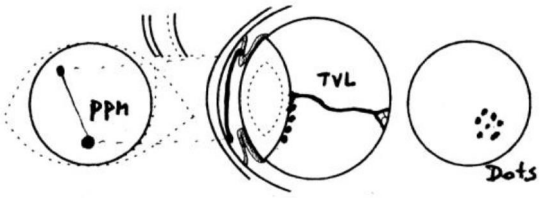
Section Examination:
The drawings in the middle of the form are used to draw and position any changes found. The circles on the left can be used for the cornea, e.g. to position corneal dystrophy, or for the anterior capsule of the lens. The dotted lines around the first circle represent contours of 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**:
In this section, the examiner should describe any findings in the eye and adnexa, either KP-HED or other. Details of all lesions and conditions are drawn and/or written in the descriptive comments. A standardized terminology (drop-down menu in the comments area of the on-line form, see below) has to be used. If the paper certificate is issued, the same terminology is to be used. An individual description of a finding should only be used rarely as it cannot be used for statistics.

- tiny pigment dots central ant. lens capsule
- tiny pigment dots near/on post. lens capsule
- minor whitish irregularity on post. lens capsule
- linear or round juvenile retinal folds (in puppy)
- **4. RD: not visible but affected, as diagnosed during puppyhood**
- 6. CEA: other: retinal detachment, haemorrhage
- **6. CEA: not visible but affected, as diagnosed during puppyhood**
- 11. Entropion nasal
- **11. Entropion/trichiasis: not visible but affected based on previous diagnosis**
- 11. Trichiasis nasal/nasal fold
- 11. Trichiasis upper eyelid temporal
- **12. Ectropion/macroblepharon: not visible but affected based on previous diagnosis**
- 12. Macroblepharon 41-44 mm
- 12. Macroblepharon ≥ 45 mm
- **13. Distichiasis/ectopic cilia: not visible but affected based on previous diagnosis**
- 13. Ectopic cilia
- 14. Corneal dystrophy, stromal
- 14. Corneal dystrophy, macular
- 14. Corneal dystrophy, endothelial
- minor lens imperfection – not visible with the naked eye in retro illumination
- Iris: pigmented lesion – to be observed
- Choroid: pigmented lesion – to be observed
- (Chorio-) retinopathy, inheritance under investigation
- chorioretinal scar/lesion
- eyelid scar
- corneal scar
- menace response reduced or absent
- Keratoconjunctivitis sicca (in non-specific breeds)
- **Undetermined**: Re-examination after 3 months (in puppy)
- **Suspicious**: Re-examination after 3 months (in puppy)
- **Undetermined**: Re-examination after 12 months or if earlier, then by the Chief Panellist/panel
- **Suspicious**: Re-examination after 12 months or if earlier, then by the Chief Panellist/panel

Other: "open text" (important: No breeding permit or breeding ban must be noted on the ECVO certificate)

"severe": The number of the relevant eye disease is noted. A tick box is provided for "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.

Section Results:
"Unaffected" means that there is no clinical evidence of the KP-HED specified. "Affected" signifies that there is clinical evidence of the KP-HED. If the clinical signs are not specific enough, "undetermined" is ticked; If clinical signs are specific but very minor, "suspicious" is ticked. Details see below:

"Undetermined" and "Suspicious"

If an **adult* animal** displays clinical features that possibly fit the KP-HED, but changes are inconclusive (not specific), **"undetermined"** is ticked for the relevant disease and it is **recommended** that the animal be **re-examined**.
If an animal displays minor, but specific clinical signs of the KP-HED mentioned, **"suspicious"** is ticked for the relevant disease. Further development will confirm the diagnosis. It is **required** that "suspicious" cases are re-examined.

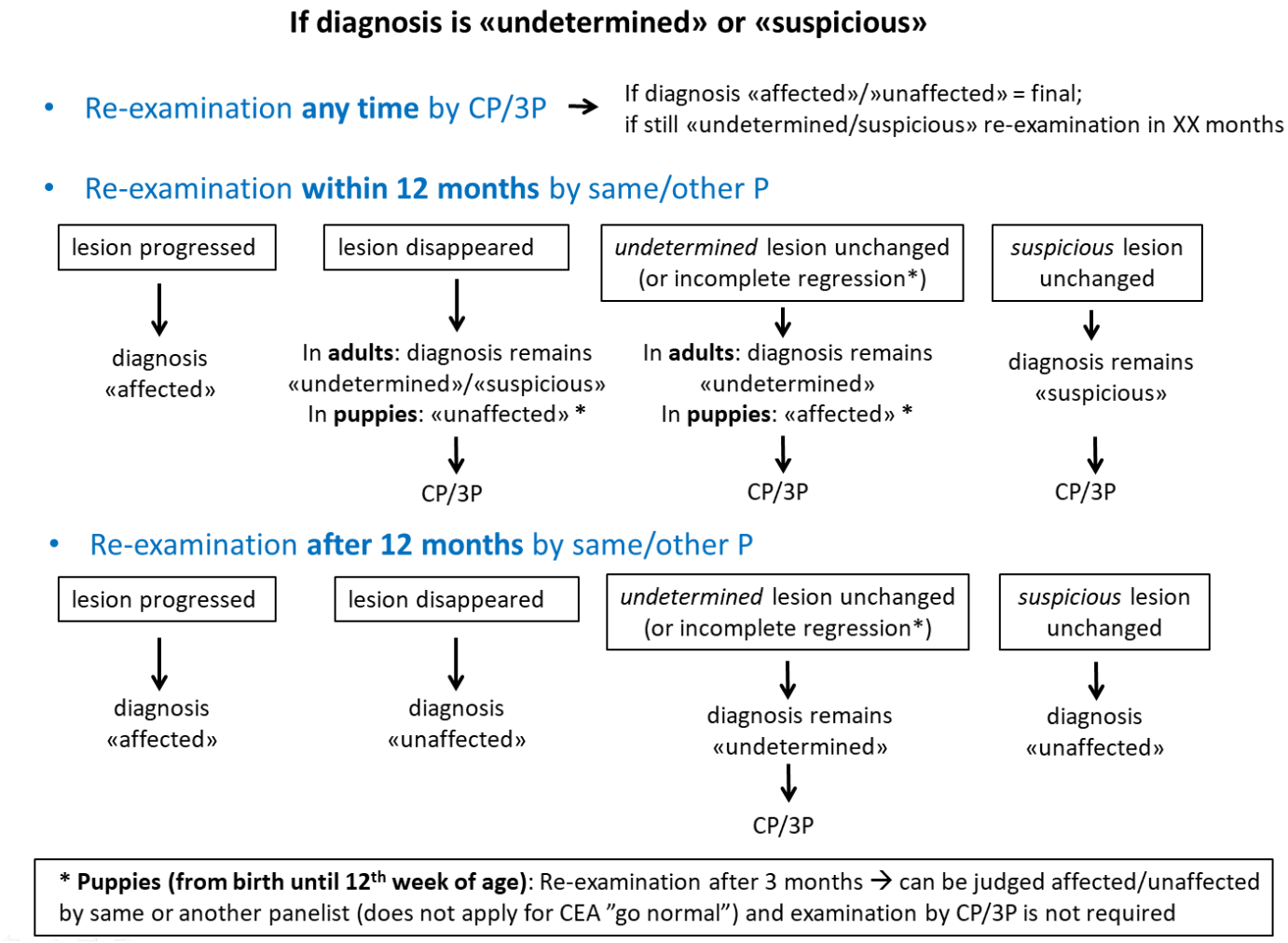
Re-examination can be done

- **Within 12 months of the last examination** by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist (3P/CP/DCP), whose decision is final, except the following alternative: The same or another Panellist (P) can do the re-examination within 12 months of the last examination, but if the Panellist (P) determines that the animal previously judged "undetermined" or "suspicious" is "affected", this "affected" judgement is valid. If the owner appeals this decision, the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist (3P/CP/DCP), whose decision will be final. In the event that the first examiner or another Panellist (P) determines that the animal previously judged "undetermined" or "suspicious" is "unaffected", the initial judgement "undetermined" or "suspicious" remains valid until the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist (3P/CP/DCP), whose decision is final.
- If a period **of at least 12 months has passed** by the same or by another Panellist (P) (examination by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist (3P/CP/DCP) is possible but *not* required).
  - In the event that the "undetermined" finding in an **adult* animal** has *not changed* at the subsequent examination, it will be judged "undetermined".
  - In the event that the "suspicious" finding has *not changed* in an animal at the subsequent examination, it will be judged "unaffected"
  - In the event that the "undetermined" or "suspicious" finding has *disappeared* at that examination, it is judged "unaffected"
  - In the event that the "undetermined" or "suspicious" finding has *progressed*, the judgment will be "affected".

***Special note for findings in puppies (birth until 12th week of age)**:
If a puppy displays clinical features that possibly fit the KP-HED, but changes are inconclusive (not specific) or if its regression with development is expected (e.g. PPM, lens opacity, PHA, retinal folds), then **"undetermined"** is ticked at the relevant KP-HED and and a re-examination indicated in the descriptive comments field using the drop-down menu: "**Undetermined**: Re-examination after 3 months (in puppy)". Findings seen **in puppies** which are typical for the KP-HED and have not changed/not regressed with development (e.g. PPM, retinal "folds", lens opacities) or have progressed (e.g. lens opacities) until re-examination will be judged "affected". If the lesion has

disappeared at that examination, it is judged "unaffected"(this does not apply for CEA-CH, “go normals”) and examination by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist is *not* required.

Rationale:
In *adult animals* “undetermined” means that the finding is not specific/not typical for the listed KP-HED (as opposed to suspicious = minor typical signs). In *puppies* “undetermined” is also used if findings are expected to regress with development. If the lesion a) has disappeared at the next examination, it is judged "unaffected"; b) remains unchanged in *adult animals* and is still not typical for the KP-HED, "undetermined" is ticked again (breeding advice = optional/low priority); c) has been seen *in the puppy* and is typical for the KP-HED and has not regressed, it is judged “affected”; d) has progressed and/or become typical for the KP-HED listed, it is judged "affected".



Graphic: serves as an overview of the procedure if a clinical diagnosis is "undetermined" or "suspicious".

For the KP-HEDs listed under numbers 1-6 and 11-17, the relevant box and, if available, the specification box (e.g. for type or class) must be ticked on the certificate.
KP-HEDs that do not have their own number on the certificate are listed in the drop-down menu under the number “7. Other” and/or under the number “18. Other” and must be ticked there. Paper certificate: For KP-HEDs that do not have their own number on the certificate, the box “7. Other” and/or “18. Other” will be ticked as “affected” and the definition names of the disease(s) must be written (see below: List of KP-HEDs, also in Chapter

5). Only if there are more than two KP-HEDs that need to be written at “7. Other” and/or “18. Other” the term “multiple other KP-HEDs” is written and the relevant KP-HEDs must be specified in the comment field using the definition names listed below (also 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 (online: listed in the drop-down menu). These are:

- Anophthalmos
- Choroidal coloboma
- Choroidal hypoplasia
- Dermoid
- Eyelid coloboma
- Exophthalmos due to shallow orbit
- 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
- Optic disc coloboma
- Persistent hyaloid artery (PHA)
- Posterior segment exam not possible
- Retinal coloboma
- Retinal dystrophy/*RPE65* null mutation (e.g. Briard)
- Scleral coloboma
- Strabismus/Esotropia (congenital)

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 are 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 (online: listed in the drop-down menu). These are:

- Retinopathy, Canine multifocal (CMR)
- Ceroid lipofuscinosis (CLN)

- Chorioretinopathy, primary retinal disease (in specific breeds: Chinese crested, pigmentary chorioretinopathy)
- Chorioretinopathy, secondary retinal disease with genetic predisposition ("working dog retinopathy" in specific breeds: Border collie, Flat coated Retriever, Borzoi)
- Glaucoma, primary
- Keratitis, chronic superficial (CSK)/Pannus
- Keratitis, punctate (in specific breeds: Dachshund)
- Keratoconjunctivitis sicca (KCS; in specific breeds: WHWT, Chinese Crested Dog, LH Dachshund, Cavalier King Charles Spaniel, English/American Cocker Spaniel, Miniature Schnauzer, English Bulldog, Pug, Shi Tzu, Lhasa Apso, Pekingese, Samoyed)
- Ocular melanosis (do not use Glaucoma –pigmentary; e.g. Cairn Terrier)
- Posterior segment exam not possible
- Retinal Pigment Epithelial Dystrophy (RPED; in specific breed: English Cocker Spaniel)
- Retinopathy, primary retinal disease (in specific breeds: Shetland sheepdog, Swedish Vallhund, Basenji, Labrador Retriever)
- Uveal cyst(s)
- Uveal melanoma (in specific breeds: Labrador Retriever)
- Uveitis, pigmentary (e.g. in Golden Retriever, Great Dane)
- Uveodermatologic Syndrome (UDS)
- Vitreous degeneration (without any signs of lens luxation)
- Vitreous strands/vitreous prolapse (without any signs of lens luxation)

General guidelines and considerations:

- The examination of the animal can only be performed after permanent identification, e.g. by microchip implantation (see chapter 3, The Scheme).
- **Puppies: dogs from birth until 12th week of age;** in puppies some findings exceptionally can regress with development (e.g. PPM, lens opacity, PHA, retinal folds); if it is to be expected that the finding may regress, then "undetermined" is ticked at the relevant KP-HED and and a re-examination after 3 months (in puppy) is indicated. (For details see section: "Undetermined" and "Suspicious")
- **Findings in dogs older than 12 weeks are assessed as in adult dogs**
- Gene testing for eye diseases does not replace clinical eye examination.
- When a dog is found "affected" for a KP-HED by a panel member or the local appeals authority (final decision) 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. In general, a final diagnosis/decision by a national panel (Chief Panellist or 3 Panellists together) should not be overruled by another panel. Exceptions to this are conditions that are changed artificially with surgical correction. In those cases, the previous results are definitive (e.g. distichiasis, entropion).

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 breeds (not complete) with special concern for primary glaucoma **to be gonioscopically examined before dilation**:

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
Finnish Lapponian Dog
Flat Coated Retriever
French Bulldog
German Longhaired Pointers
Golden Retriever
Siberian Husky
Leonberger
Magyar Viszla
Mini Australian Shepherds
Old English Sheepdog
Samoyed
Spanish Waterdog
Tatra

Examination using slit-lamp biomicroscopy before dilation of the pupil is highly recommended for all breeds. List of **breeds (not complete)** in which slit-lamp examination before dilation has to be done always for the below listed KP-HED:

1. Persistent Pupillary Membrane
Basenji
Chow Chow
English Cocker Spaniel
Petit Basset Griffon Vendéen
2. Iris hypoplasia
Australian Shepherd
Dalmatian
Rottweiler
Tibetan Spaniel
3. **Lens luxation (PLL)/KCS/vitreous degeneration/ocular melanosis**
Chinese Crested dog (PLL, vitreous degeneration, KCS)
Cavalier King Charles Spaniel, American Cocker Spaniel, English Cocker Spaniel (KCS)
English Bulldog (KCS)
Lancashire Heeler (PLL)
Longhaired Dachshund (KCS)
Lhasa Apso (KCS)
Miniature Schnauzer (KCS)
Pug (KCS)
Pekingese (KCS)
Samoyed (KCS)
Shih Tzu (KCS)

Cairn Terrier also (ocular melanosis)
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:

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 (using the drop-down menu and write at "other:" secondary cataract – non-hereditary).

   Classification according to age of onset:
   - **Congenital cataracts:** If cataracts are observed in the period between birth and the 12th 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 (later onset)". If lens opacities are seen in puppies (up to 12 weeks of age) which are not described as a known or presumed hereditary congenital cataract, at "3. Cataract (congenital)" "undetermined" is ticked and a re-examination indicated in the descriptive comments

field using the drop-down menu: **Undetermined**: Re-examination after 3 months (in puppy). *See also page 3 "undetermined" and "suspicious".*

- **Juvenile and adult cataracts:** cataracts developed at older age (after 12th weeks of age) are ticked "affected" at "15. Cataract (later onset)" and tick further specification as given below.
- **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 page 3 "undetermined" and "suspicious".*

  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:**
  Certain lens opacities 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 and specified in the comment field under "15. "other lens opacity":
    - <u>Punctate</u>: one or more *clearly defined* whitish dot like opacities located in the cortex or nucleus
    - <u>Suture line tips:</u> *clearly defined* whitish small linear opacities at the ends of the suture lines
    - <u>Suture line:</u> *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.
    - <u>Nuclear ring:</u> *delicate semi-translucent irregular shaped* more or less circular structure in the nucleus
    - <u>Nuclear fiberglass-like/pulverulent:</u> 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.

In the case of "other lens opacities": indicate in the comment field at “15. Other lens opacities” the corresponding subtype. At “15 Cataract (later onset)” the box "unaffected" is ticked.

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”
  - later onset cataracts: tick at “15. Cataract (later onset)” “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 lens opacity”: punctate, suture line tip, suture line, nuclear ring or nuclear fiberglass/pulverulent; indicate in the comment field at “15. Other lens opacities” the corresponding subtype. At “15. Cataract (later onset)” the box "unaffected" is ticked.

- 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 (later onset)” 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 (later onset)” 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. Other lens opacity”.
- If there is a <u>distinct (well defined)</u> **post. pol. cataract**, without signs of spreading into the remaining cortex, tick at “15. Cataract (later onset)” the box “affected” and the specifying box "post. pol";
- If the **post. polar cataract** <u>extends into the adjacent cortex</u> (and is therefore progressive), tick at “15. Cataract (later onset)” 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 (later onset)” the box “affected” and the specifying box “cortical”. However, if there are distinct delicate suture lines or suture line tips tick at “15. Cataract (later onset)” the box “affected” and the specifying box “other lens opacity”, and in the comment area at “15. Other lens opacity”: “suture line (tip)”.
- In a **total/mature cataract (defined as cortical and nuclear cataract):** in case of **congenital cataract, tick at "3. Cataract (congenital) the box “affected” and tick also “affected” at “7. And use: Other, Posterior segment exam not possible” in the drop-down menu**. In case of **cataract (later onset)** tick at “15. Cataract (later onset)” the boxes “affected” and the specifying boxes “cortical” **AND** “nuclear”; tick also “affected” **and** write (online: use) 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). <u>Only, if</u> 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 (later onset)".

- **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". **If the posterior segment examination in case of PHTVL/PHPV is not possible, tick also "affected" at "7. Other", and use: "Posterior segment exam not possible" in the drop-down menu.**
- **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).
- **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 imperfections and those visible are to be mentioned as cataracts.
  In case of doubt, 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 after 12 months or if earlier, then by the chief panelist/panel. *See also page 3 "undetermined" and "suspicious".*

Choroidal hypoplasia (CH) [or chorioretinal dysplasia (CRD)]

CH/CRD resembling CEA but seen in breeds other than those listed under "Collie eye anomaly (CEA)": At number "7. Other: Choroidal hypoplasia" is written (online: is used), and the box affected is ticked. In cases where the clinical features which could possibly fit this entity, but the changes are not specific enough, the result of the examination is: "undetermined".

Collie eye anomaly (CEA)

Collie eye anomaly is to be ticked "affected" at "6. CEA" in the following breeds known to have CEA (genetic test available): Australian Kelpie, Australian Shepherd, Bearded Collie, Border Collie, Boykin Spaniel, Collie (short hair, long hair), Hokkaido, Lancashire Heeler, Miniature American Shepherd, Nova Scotia Duck Tolling Retriever, Shetland Sheepdog (Sheltie), Silken Windhound, Silken Windsprite. The box "affected" and one of the specifying boxes have to be ticked. If the box "other" is applicable, this has to be specified using in the comment area the drop-down menu "6. CEA: other: retinal detachment, haemorrhage".
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 8th 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.

Corneal dystrophy

Corneal dystrophy is to be ticked "affected" at "14. Corneal dystrophy", and has to be specified in the comment area using the drop-down menu: 14. Corneal dystrophy, stromal, 14. Corneal dystrophy, macular or 14. Corneal dystrophy, endothelial.
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 descriptive comments 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, are to be written on the form. **Ectopic cilia can be noted in the Descriptive comments field using the drop-down menu.**
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". The entropion or trichiasis localisation can be specified using the drop-down menu (text) in the Descriptive comments field. No further details are to be mentioned on the form. Only if there are distinct 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". The macroblepharon can be specified using the drop-down menu (text) in the Descriptive comments field, but no further details 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 <u>in combination with macroblepharon</u>. 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 in the **examination field**, at "other". The values found are to be noted in the **descriptive comments** field using the drop down at: other.

Iridocorneal angle abnormality (ICAA)

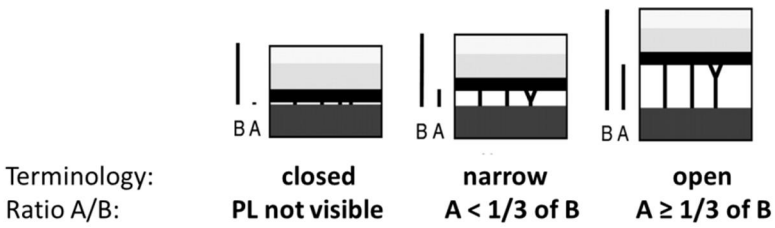
Two predominant types of involvement of the angle are distinguished. The pectinate ligament (PL) and the iridocorneal angle (ICA) width are evaluated by gonioscopy in its extent of 360 degrees, thus giving the owner and/or the breed club/society the opportunity to select animals on severity of the defect.

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) also the box "severe" is to be ticked in the comment area.

Iris melanoma, use Uveal melanoma

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. If the STT is below 10 mm and there are clinical signs of KCS in specific breeds (WHWT, Chinese Crested Dog, LH Dachshund, Cavalier King Charles Spaniel, American and English Cocker Spaniel, English Bulldog, Pug, Shi Tzu, Lhasa Apso, Pekingese, Miniature Schnauzer, Samoyed): 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 (in the non-specific breeds), the box "suspicious" is ticked. The STT values are indicated in the online form (or written in the descriptive comments field). Clinical re-examination is recommended according to clinical symptoms. Note: The time until the re-examination for the next ECVO certificate is indicated in the comment field (drop-down menu) as follows: "Re-examination after 12 months or if earlier, then by the chief panelist/panel"

Microblepharon

If the lid fissure in an adult dog is smaller compared to dogs of the same breed, at "7. Other": "Microblepharon" is written (online: is used), and the box "affected" is ticked. Only if a uni- or bilateral

microblepharon is diagnosed usually combined with a severe upper lid entropion, 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-HEDs

- Paper certificate: If there are more than two KP-HEDs that need to be written at "7. Other" and/or "18. Other" the term "multiple other KP-HEDs" is written and the relevant KP-HEDs must be specified in the comment field using the definition names listed at pages 5-6 (also in Chapter 5).
- Online form: at "7. Other" and/or "18. Other" all relevant diseases are to be used from the drop-down menus and are ticked "affected". The selected diseases are displayed in the comment field.

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 used (on paper written 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 used (on paper: written) 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: PHTVL-like 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 descriptive comments field, using the drop down: "2. tiny pigment dots near/on post. lens capsule". However, the dog is ticked "unaffected" for PHTVL/PHPV.
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: PPM-remainder: 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 descriptive comment field using the drop down menu: "1. tiny pigment dots central ant. lens capsule" These are not ticked "undetermined", "suspicious" 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 (later onset)".

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 thus 4. Retinal dysplasia in the result field should not be ticked as "affected". These folds have to be mentioned in the descriptive comments field using the drop-down menu: "linear or round juvenile retinal folds (in puppy)"
In the English Springer Spaniel, Golden Retriever, Labrador Retriever and Samoyed these juvenile folds are considered as retinal dysplasia (RD) and should be ticked "affected".
In puppies where clinical features do not allow a differentiation of juvenile folds and retinal dysplasia, at 4. Retinal Dysplasia: "undetermined" is ticked and a re-examination indicated in the descriptive comments field using the drop-down menu: **Undetermined**: Re-examination after 3 months (in puppy)

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".

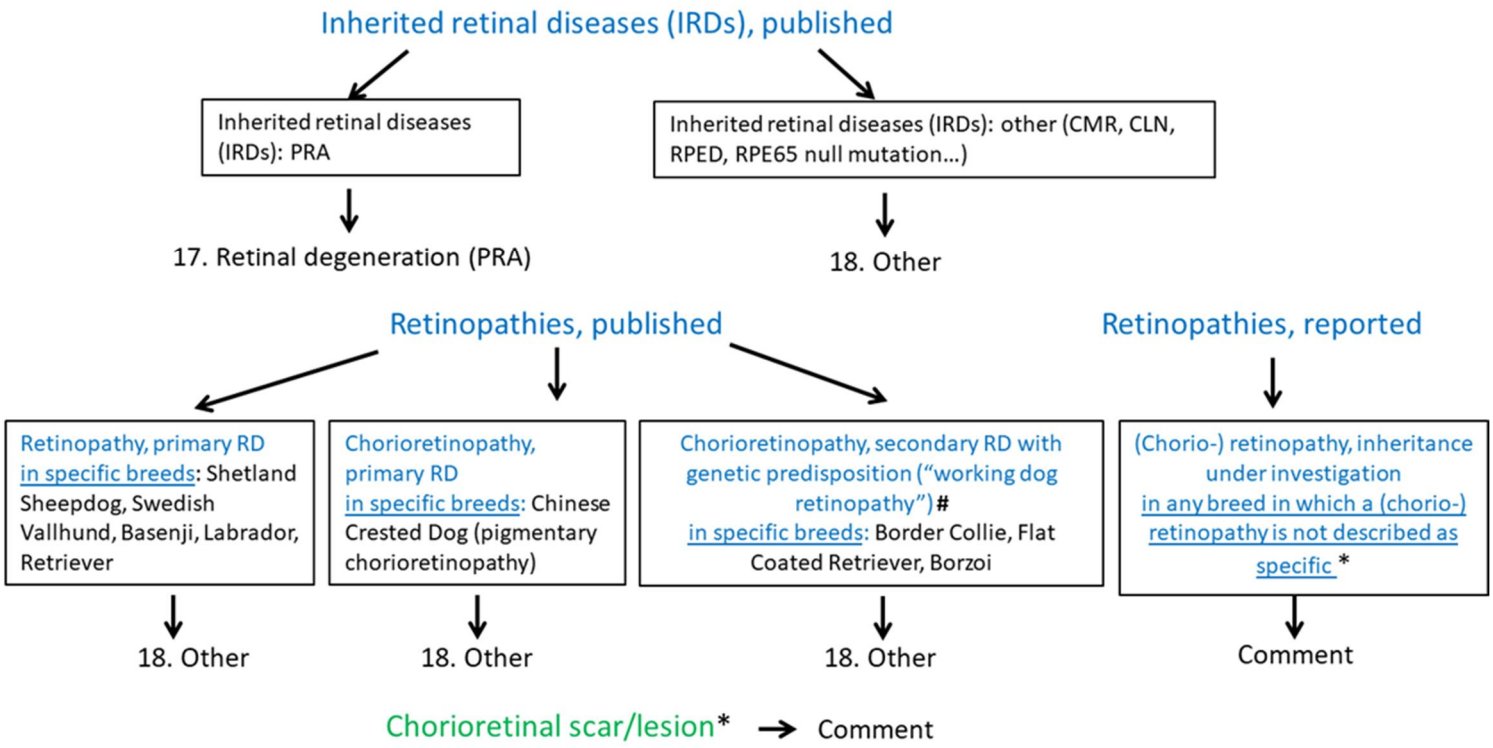
Retinal degeneration/Progressive Retinal Atrophy (PRA)

KP-HEDs; a group of bilateral, hereditary dysplastic and/or degenerative diseases of the photoreceptors primarily, progressing to blindness in both eyes simultaneously. The onset of the blindness depends on the affected breed and the type of process (dysplasia and/or degeneration). The photoreceptor abnormalities can be detected by an electroretinogram (not part of a routine ECVO HED-Scheme eye examination) before there are detectable fundus changes observed by ophthalmoscopy. If an ERG is

done, this is to be mentioned using the drop-down menu in the Examination field: “ERG”. The resulting values can be mentioned using the drop-down menu in the descripting comments field, at “other”.
The funduscopic changes consist in the early disease of a horizontal hyperreflectivity band just above the optic nerve head or a change in reflectivity, with greyish discoloration mainly in the periphery and mid-periphery in the tapetal area of the fundus accompanied by slight vascular attenuation. With progression of the disease there are more generalized changes with hyperreflectivity of the tapetal fundus, depigmentation and uneven pigment distribution in the non-tapetal fundus, ending in severe vascular attenuation and a pale optic disc. There are multiple genetic types of PRA including different forms of rod-cone dysplasia and degeneration (rcd 1-4) and progressive rod cone degeneration (prcd). DNA-tests for specific forms and breeds are available.
If early changes are recognized, in the Results field, at “17. Retinal degeneration (PRA)” the box “suspicious” is ticked and in the descriptive comments field using the drop down menu the following advice is given: “Suspicious: Re-examination after 12 months or if earlier, then by the chief panelist/panel” and at “other”: the dog should be examined by ERG and DNA test.
If there is clear evidence of retinal degeneration (PRA) the box “affected” is ticked.

Retinopathies

- Retinopathy, primary retinal disease (in specific breeds: Shetland Sheepdog, Swedish Vallhund, Basenji, Labrador, Retriever): At “18. Other”: “Retinopathy, primary retinal disease” is used, and the box “affected” is ticked.
- Chorioretinopathy, primary retinal disease (in specific breeds: Chinese Crested Dog, pigmentary chorioretinopathy): At “18. Other”: “Chorioretinopathy, primary retinal disease” is used, and the box “affected” is ticked.
- Chorioretinopathy, secondary retinal disease with genetic predisposition (“working dog retinopathy”, in specific breeds: Border Collie, Flat Coated Retriever, Borzoi).
  If typical "clinical" signs of working dog retinopathy (several scar lesions or evidence of retinal degeneration) are seen at the first examination, at "18. Other”: “Chorioretinopathy, secondary retinal disease with genetic predisposition" is used and the box "affected" is ticked. If single or only a few minor scar lesions are noticed for the first time, and no information about progression is available, at "18. Other”: “Chorioretinopathy, secondary retinal disease with genetic predisposition" is used and the box “suspicious” is ticked and, in the descriptive comments field the “Suspicious: Re-examination after 12 months or if earlier, then by the chief panelist/panel” is required; If not progressive, tick “unaffected” and use in the descriptive comments field: “other: and mention “retinal lesions – to be observed”; if the lesions are progressive, tick “affected”.
- (Chorio-) retinopathy, inheritance under investigation:
  Any (chorio-) retinopathy in any breed NOT mentioned at “retinopathy, primary retinal disease”, “chorioretinopathy, primary retinal disease” or “chorioretinopathy, secondary retinal disease with genetic predisposition” = **in any breed in which a (chorio-) retinopathy is not described as specific** should be indicated in the descriptive comments field using the drop-down menu: “(Chorio-) retinopathy, inheritance under investigation”
  Important: (Chorio-) retinopathies, inheritance under investigation are multifocal progressive lesions and should not be confused with chorioretinal scar/lesions, which are single or a few typically non-progressive lesions (see also Chapter 5 Definitions). If the findings are inconclusive, the examiner can use "chorioretinal scar/lesion" from the drop-down menu in the comments area at the first examination and, if progressed, use "(Chorio-) retinopathy, inheritance under investigation" from the drop-down menu in the comments area at the next examination.



* Important: **(Chorio-) retinopathies, inheritance under investigation** are multifocal ***progressive*** lesions and should not be confused with **chorioretinal scar/lesions**, which are single or a few typically ***non-progressive*** lesions (see also Chapter 5 Definitions).
If the findings are inconclusive, the examiner can use "chorioretinal scar/lesion" from the drop-down menu in the comments area at the first examination and, if progressed, use "(Chorio-)retinopathy, inheritance under investigation" from the drop-down menu in the comments area at the next examination.

Important: If single or only a few minor scar lesions are noticed for the first time, and no information about progression is available, at "18. Other": "Chorioretinopathy, secondary retinal disease with genetic predisposition" is used and the box "suspicious" is ticked

Uveal Cysts

If there are only 1-3 cysts and no connected signs of glaucoma and/or uveitis at "18. Other": "uveal cyst(s)" is used (on paper: written), 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.

Uveal melanoma

In specific breed (Labrador Retriever): If there are typical "clinical" signs of an uveal melanoma (raised, black-brown, more or less circumscribed) lesion in the iris or choroidea whose growth has been noted), at "18. Other": "Uveal melanoma" is used (on paper: written) and the box "affected" is ticked. If a small, non-raised pigmentation in the iris or choroidea is noticed for the first time, at "18. Other": "Uveal melanoma" is used (on paper: written), the box "undetermined" is ticked and "re-examination after 12 months or if earlier, then by the chief panelist/panel" mentioned in the descriptive comments field (on paper: written); If the lesion remains unchanged or increases in size but still non-raised at following examinations, tick "undetermined" and use (on paper: write) in "descriptive comments": "iris or choroidea: pigmented lesion– to be observed"; if the lesion becomes raised, tick "affected".

In other breeds the heredity of uveal melanomas is at present not presumed: use (on paper: write) in "descriptive comments": "iris or choroidea: pigmented lesion– to be observed";

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" is ticked and "re-examination after 12 months or if earlier, then by the chief panelist/panel" mentioned in the descriptive comments field (on paper: written). Tonometry before dilation is recommended.

Figures of the KP-HED are found on the ECVO website at:
https://www.ecvo.eu/hereditary-eye-diseases/images-for-certified-examiners-panellists.html

Instructions (with pictures) for assessing specific KP-HEDs and completing the ECVO Certificate are available as PDF documents on the ECVO website
https://www.ecvo.eu/hereditary-eye-diseases/ecvo-manual.html
See Chapter 6