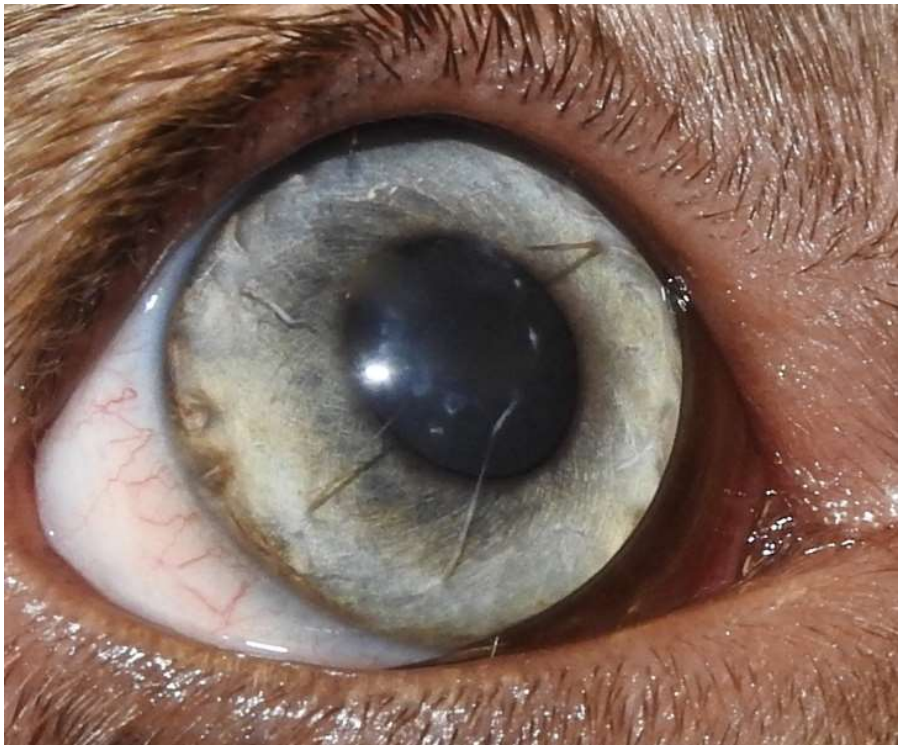
HED SESSION – Florence 2018

In cooperation with the Italien panel

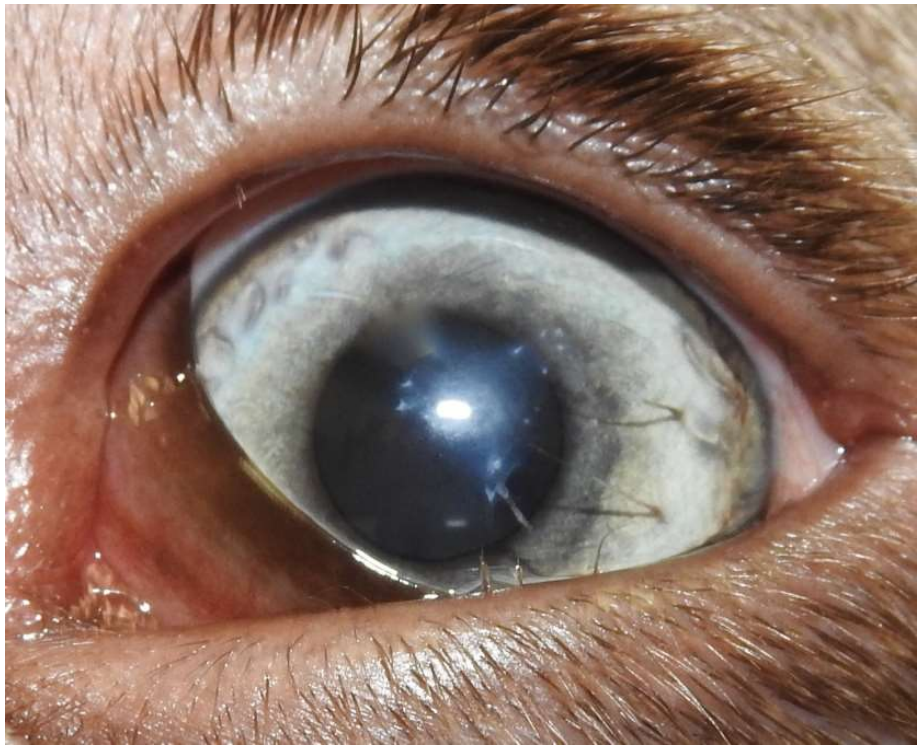
Case1: Weimaraner, 7 months, female, OU

(abnormalities distinctly visible after pupil dilation)

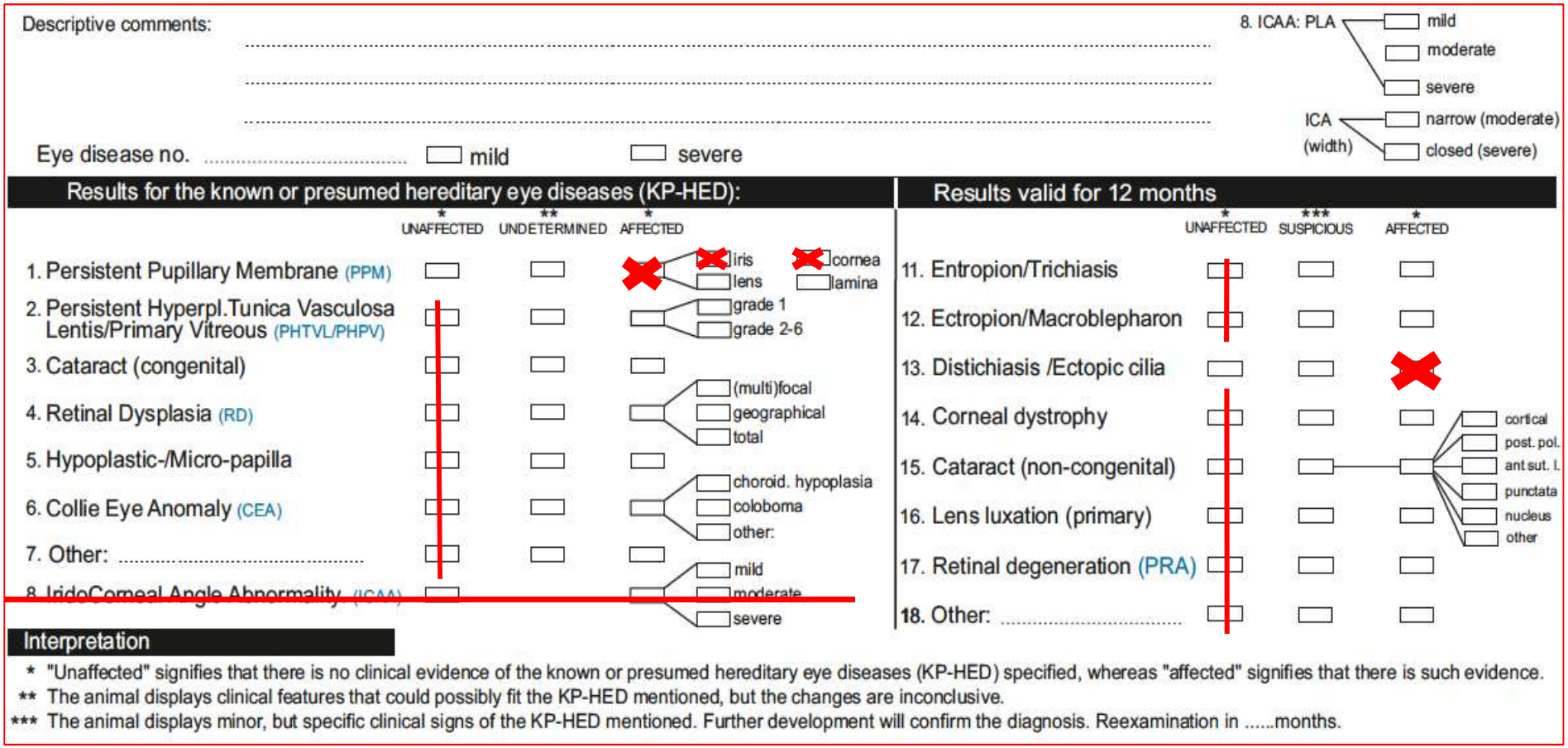
OD



OS



Case1: Weimaraner, 7 months, female, OU
(abnormalities distinctly visible <u>after</u> pupil dilation)



HED Manual 2017-04: Ch. 5 Definitions

- **Distichiasis:presumed hereditary eye disease**; single or multiple hairs (cilia) from an abnormally located hair follicle in the eyelid margin, usually growing from or in between the **Meibomian glands,** and arising from the Meibomian duct openings, which may cause ocular irritation.
- **Persistent pupillary membrane (PPM):presumed hereditary congenital eye disease** in which blood vessel remnants of the embryological vascular network in the **anterior chamber** of the eye fail to regress which normally occurs during the first 4 to 5 weeks of life.These remnants may be found on the surface of the iris at the **collarette**, the lens capsule or against the corneal endothelium or strands may bridge from **iris** to iris, iris to **cornea**, iris to **lens**, with or without sheets of tissue in the **anterior chamber**. The last three forms pose the greatest threat to vision and, when severe, vision impairment may occur.

HED Manual 2017-04: Ch. 6 Guidelines

- **Distichiasis**: only if there are clinical signs of corneal irritation such as detritus on the distichia, corneal edema, corneal vessels, defects or pigmentation at the location of the distichia; hard stiff distichia and/or ectopic cilia, the examiner will also tick the box: “severe’ in the comment area.

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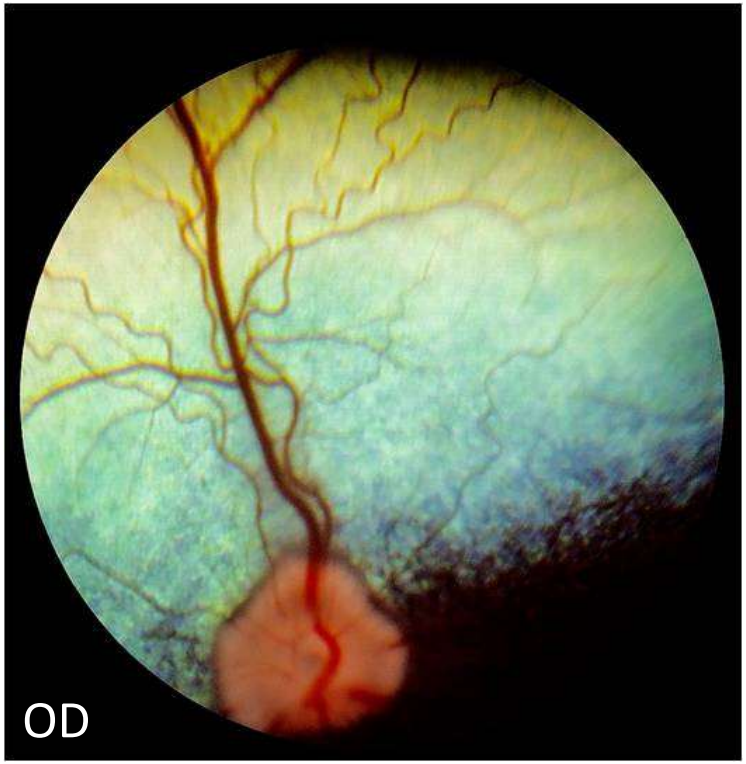
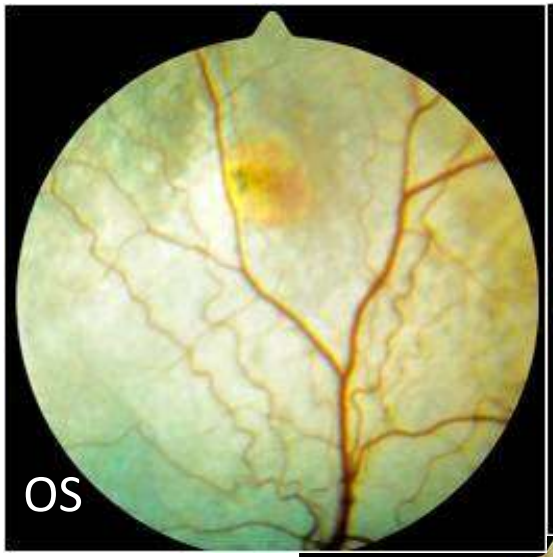
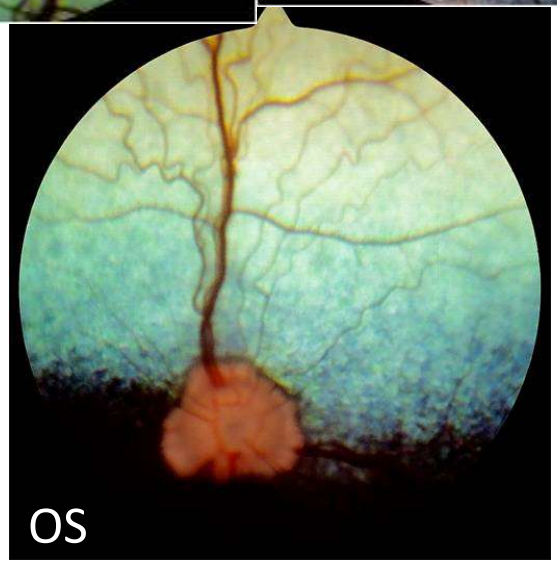
- **Persistent Pupillary Membrane**: Remnants, still distinctly present after pupil dilatation, crossing the pupil, corneal, or with lens involvement, are ticked in the box for 1. PPM: “affected” and the respective box of other parts involved. Areas which can be involved are: retrocorneal (boxes PPM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

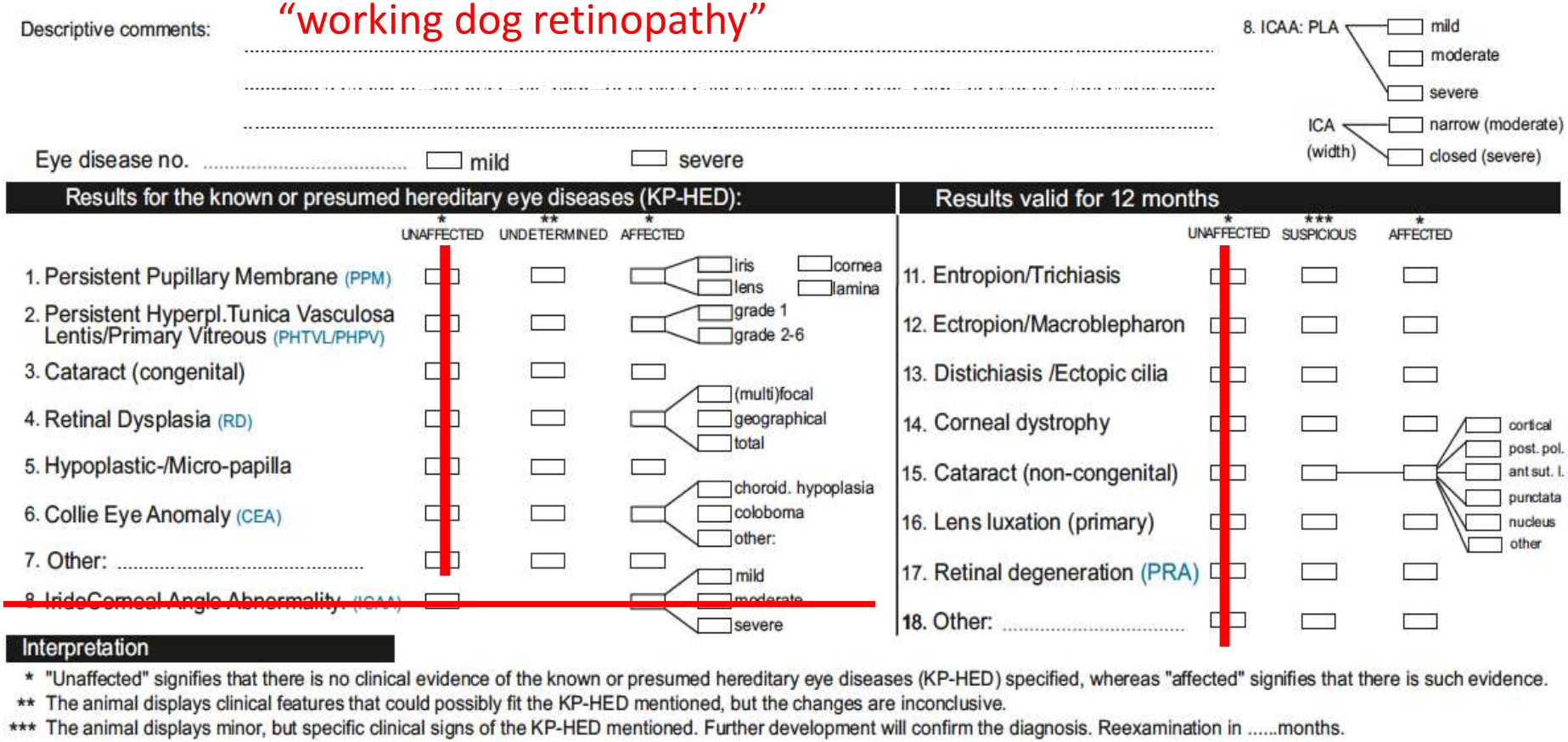
Case 3: Malinois, 4 years, male OU







Case 3: Malinois, 4 years, male OU



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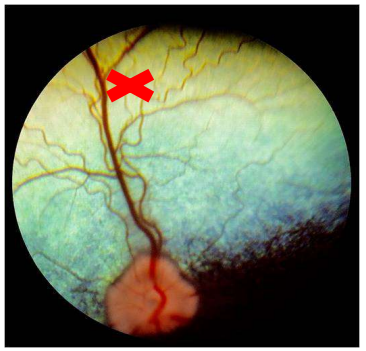
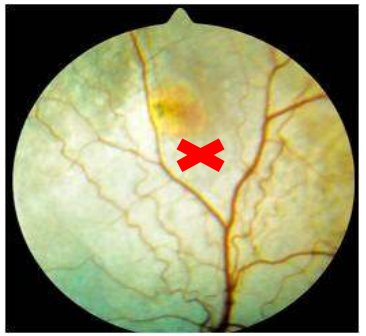
Comments

- Why do not tick “Other” and put in comments “presumed hereditary retinal degenerations ” ?
- **“18. Other”**, on the certificate for KP-HED**,** which are considered not to be congenital/developmental or which are progressive, and not yet named on the form, are mentioned. The available name of the disease in the list of ‘Definitions’ of this Manual (see chapter 5) is used.
- “Working dog retinopathy” is not available in definitions as a presumed hereditary retinal degeneration

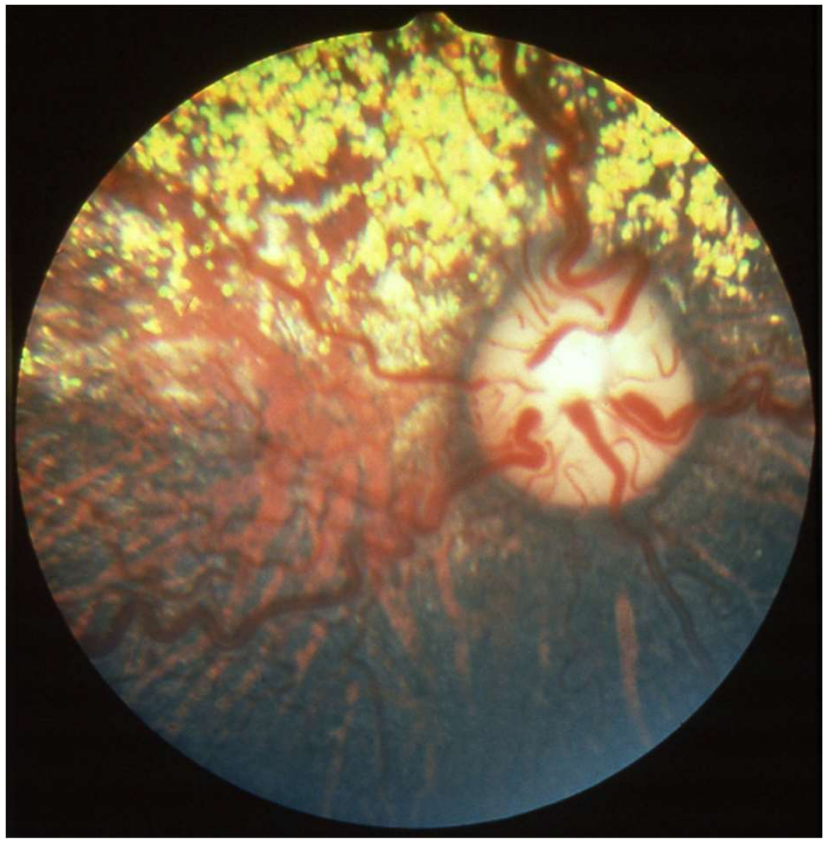
→ Write in the descriptive comments area: Working dog retinopathy





Case 4: Rough Collie, 4 years, male

OD



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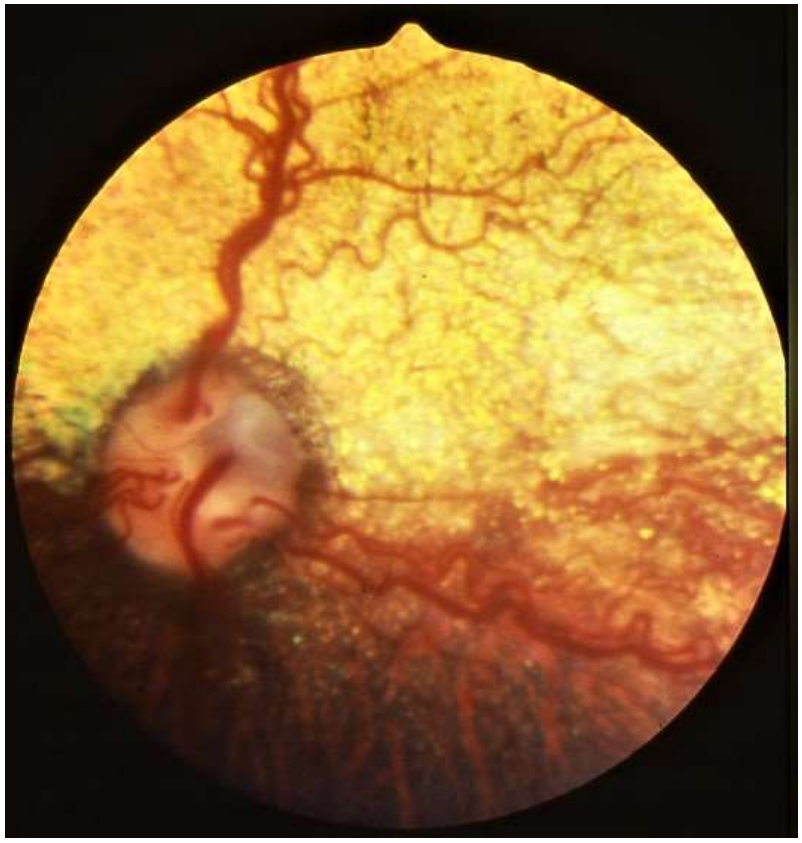
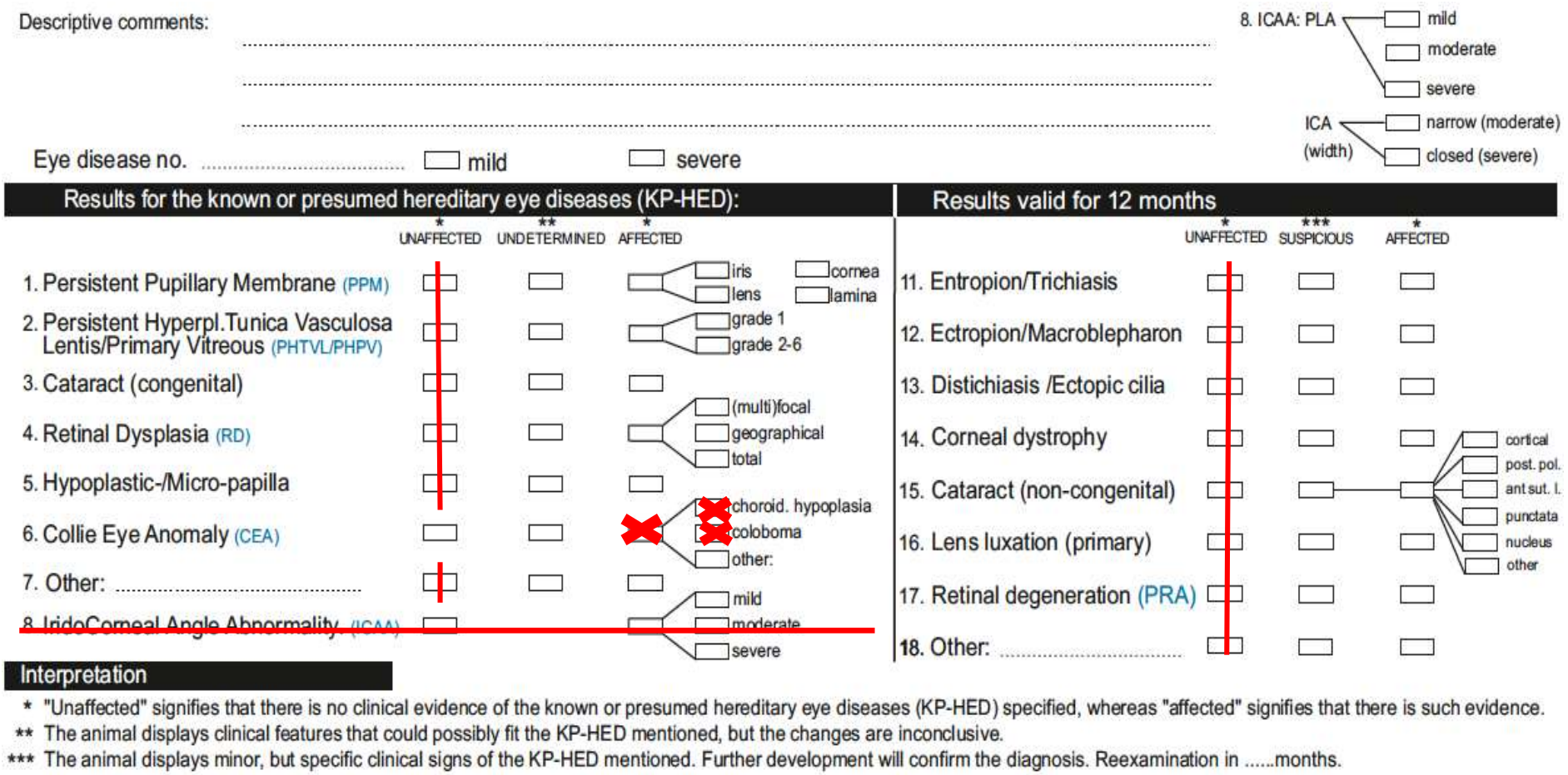


Case 4: Rough Collie, 4 years, male



Interpretation

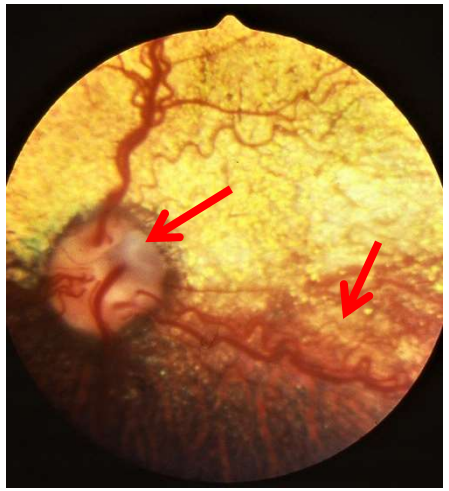
* "Unaffected" signifies that there is no clinical evidence of the known or presumed hereditary eye diseases (KP-HED) specified, whereas "affected" signifies that there is such evidence.

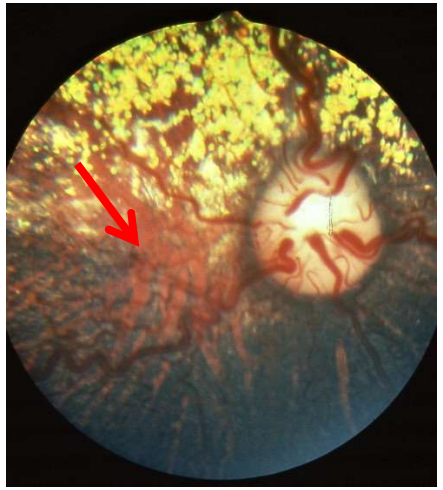
** The animal displays clinical features that could possibly fit the KP-HED mentioned, but the changes are inconclusive.

*** The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis. Reexamination in .....months.

HED Manual 2017-04: Ch. 5 Definitions

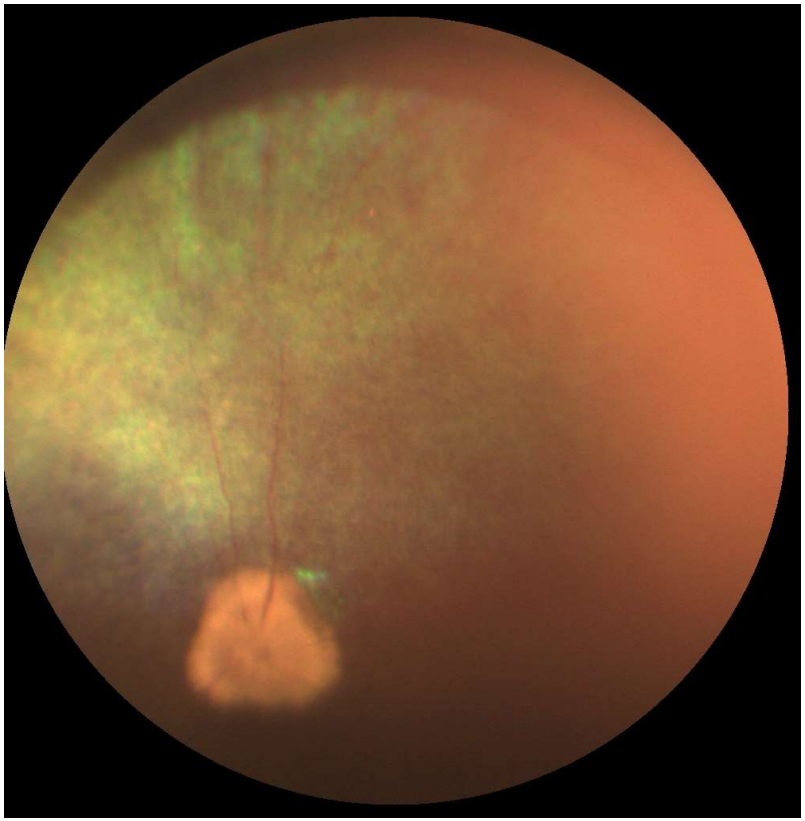
- **Collie Eye Anomaly (CEA):** known hereditary congenital eye disease; a **congenital** syndrome of ocular anomalies mainly in Collie breeds affecting the choroid and sclera and indirectly the retina and optic disc. It is characterized by bilateral and often symmetrical defects including **choroidal hypoplasia (CH, CRD)** with or without **coloboma**, **retinal detachment and intraocular hemorrhage**. Vision varies with the degree to which an individual is affected and may be minimally compromised to having severe visual impairment or blindness. DNA-tests for choroidal hypoplasia in specific breeds are available.





Case 6 Miniature Schnauzer, male, 4 years, OU

OD



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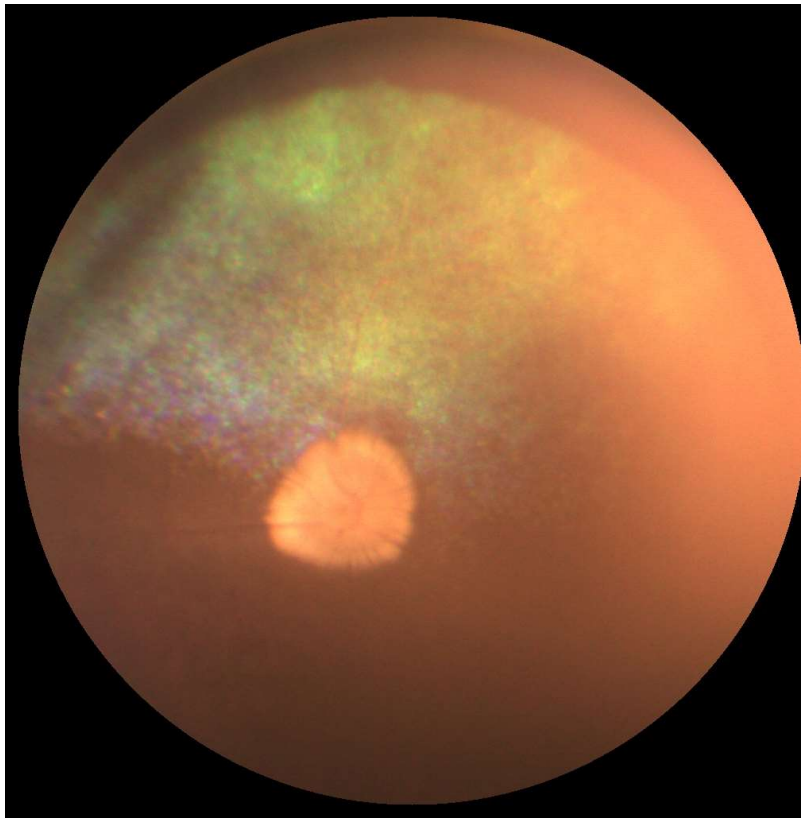
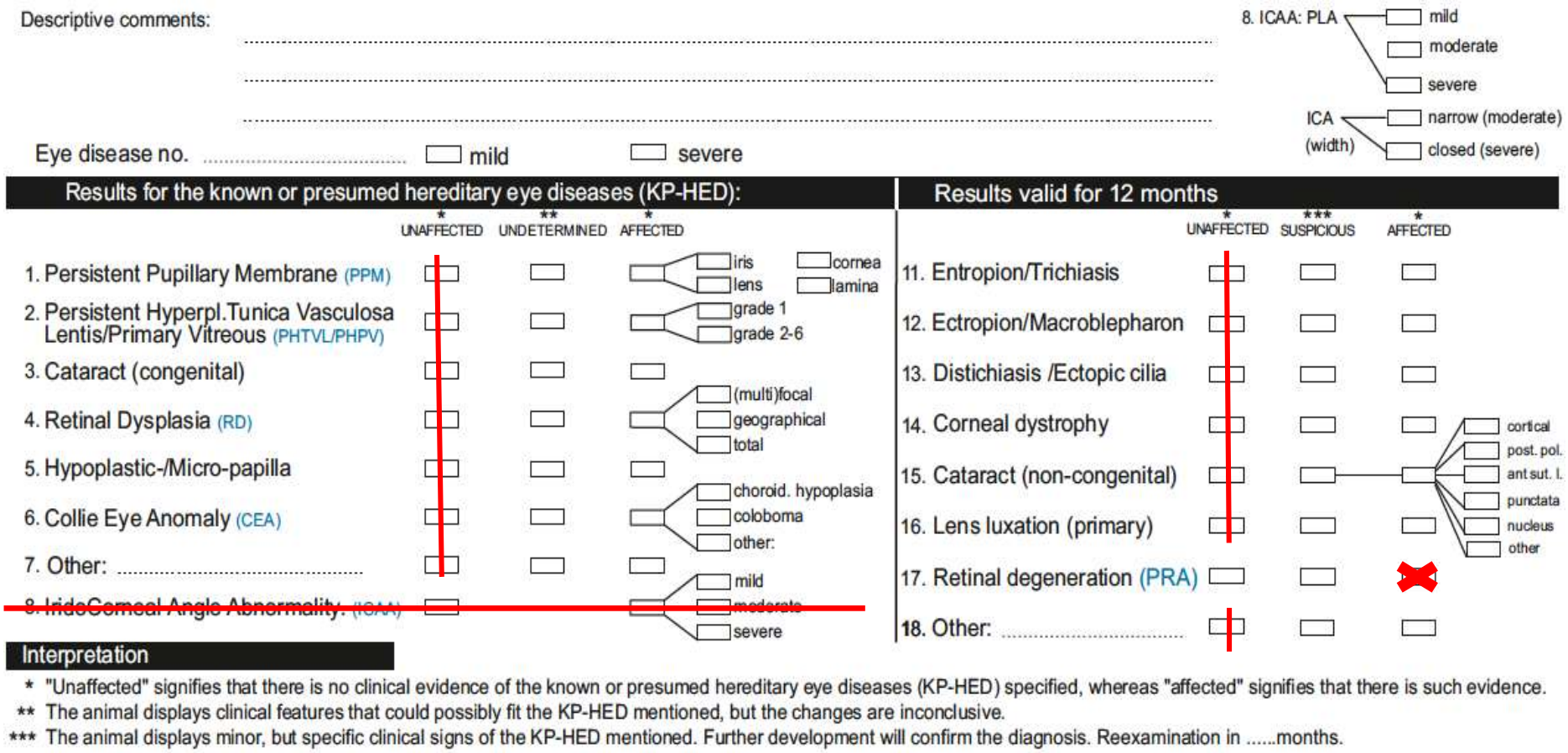


Case 6 Miniature Schnauzer, male, 4 years, OU



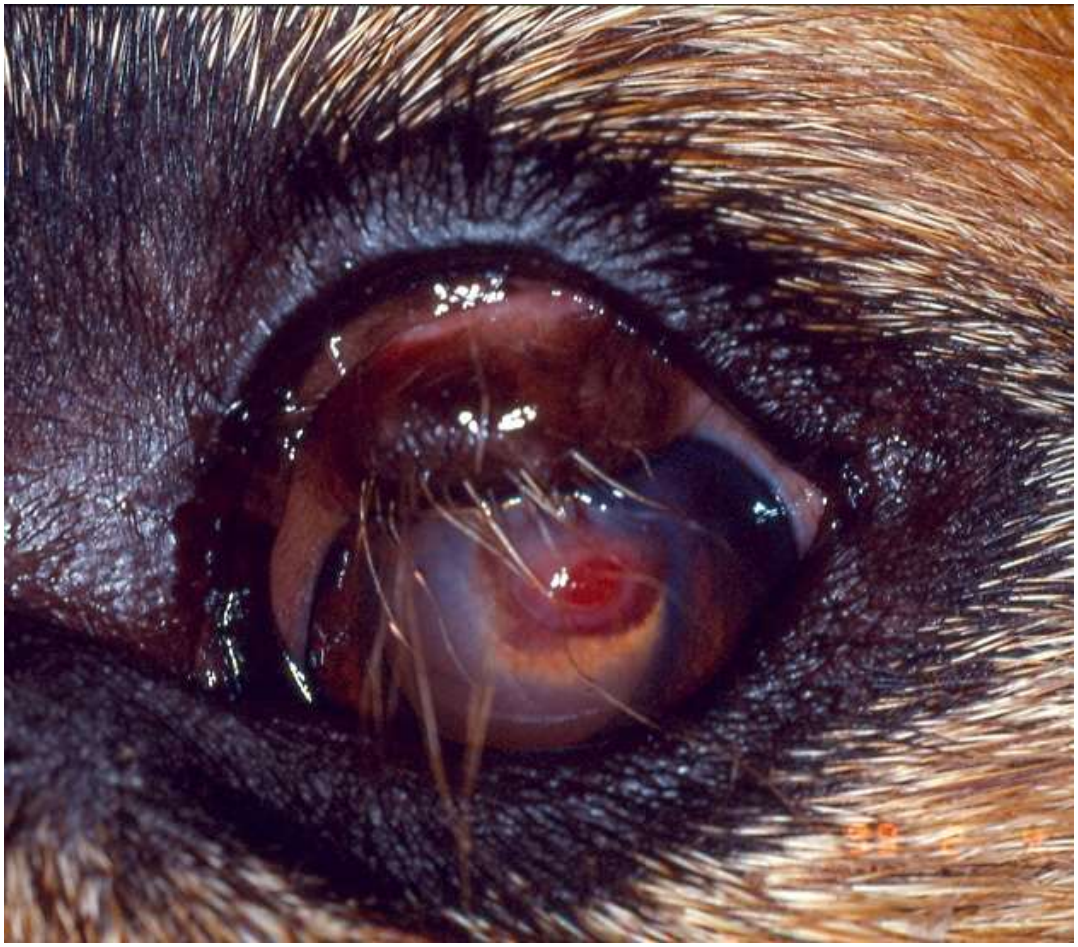
HED Manual 2017-04: Ch. 5 Definitions

- **Retinal degeneration/Progressive Retinal Atrophy (PRA):** known hereditary eye disease; 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 Scheme eye examination) before there are detectable **fundus** changes observed by ophthalmoscopy.

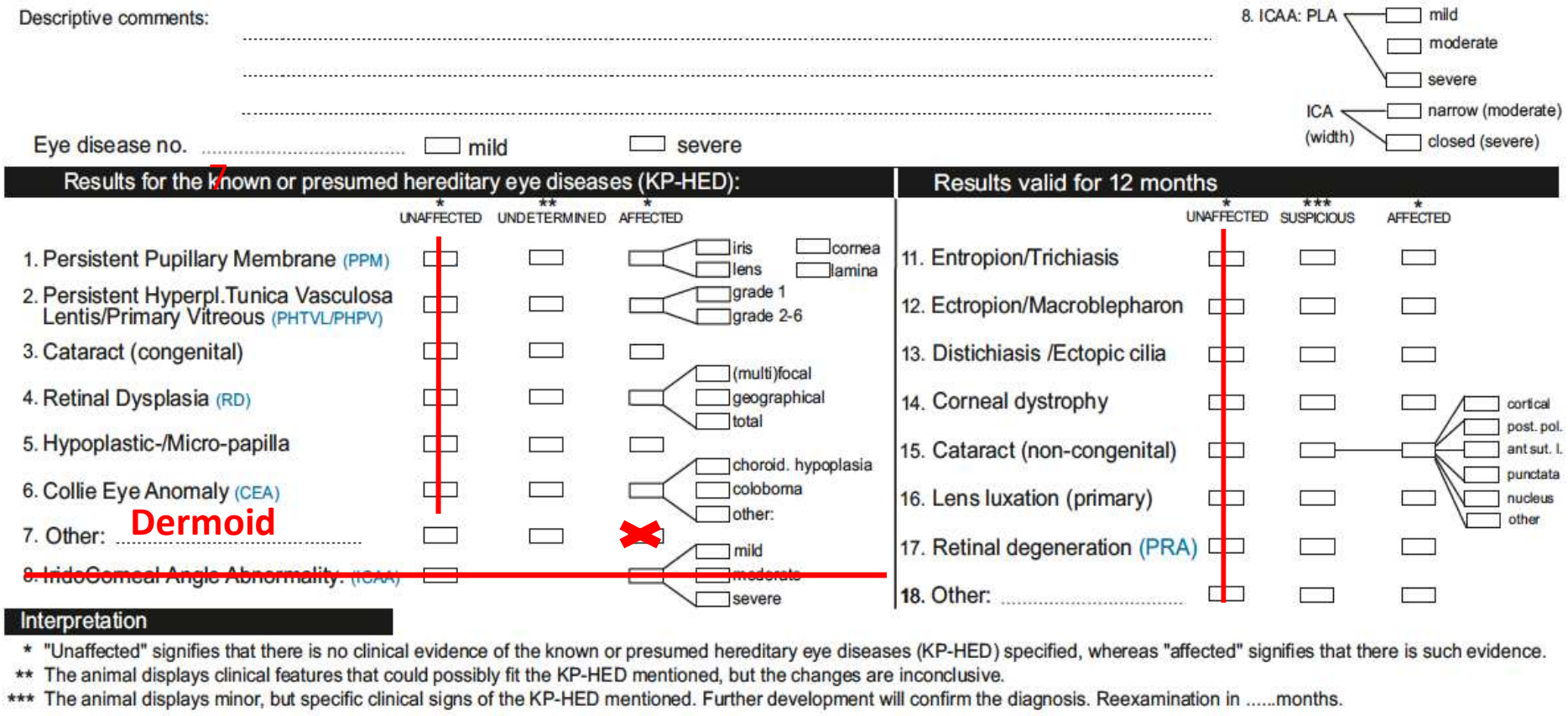
HED Manual 2017-04: Ch. 5 Definitions

- These funduscopic changes consist in the early disease of a change in reflectivity with greyish discoloration mainly in the periphery and midperiphery 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**, severe vascular attenuation and a pale optic disc. There are multiple genetic types of PRA including different forms of **rod-cone dysplasia and degeneration (rcd1-4)** and **progressive rod cone degeneration (prcd)**. DNA-tests for specific forms and breeds are available.

Case 7: German Shepherd, 6 months, female, OS



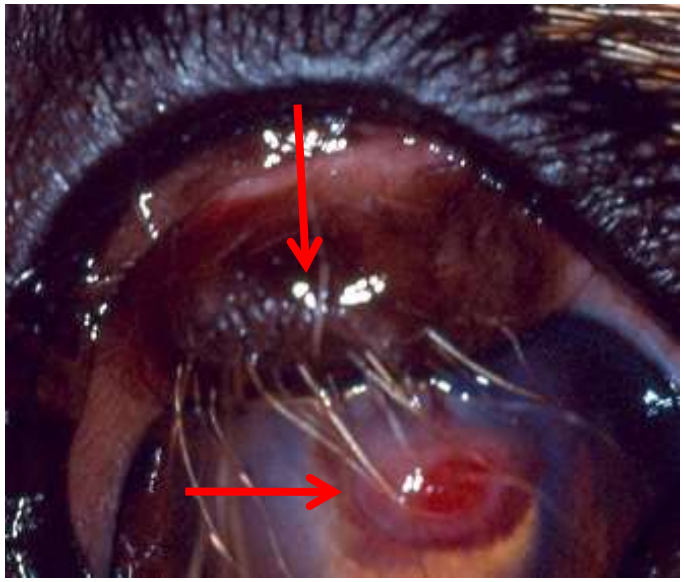
Case 7: German Shepherd, 6 months, female, OS



HED Manual 2017-04: Ch. 5 Definitions

- **Dermoid:** <u>presumed hereditary eye disease;</u>

  a congenital patch of skin in an abnormal location. Most ocular dermoids affect the **cornea** or adjacent **conjunctiva**, and its presence usually causes ocular irritation

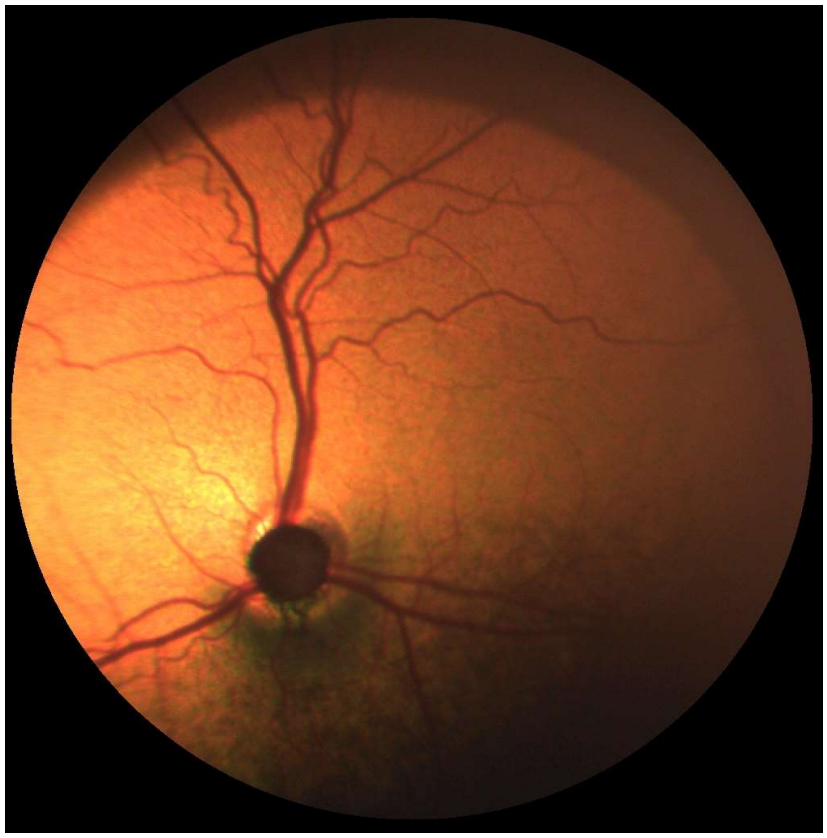


HED Manual 2017-04: Ch. 6 Guidelines

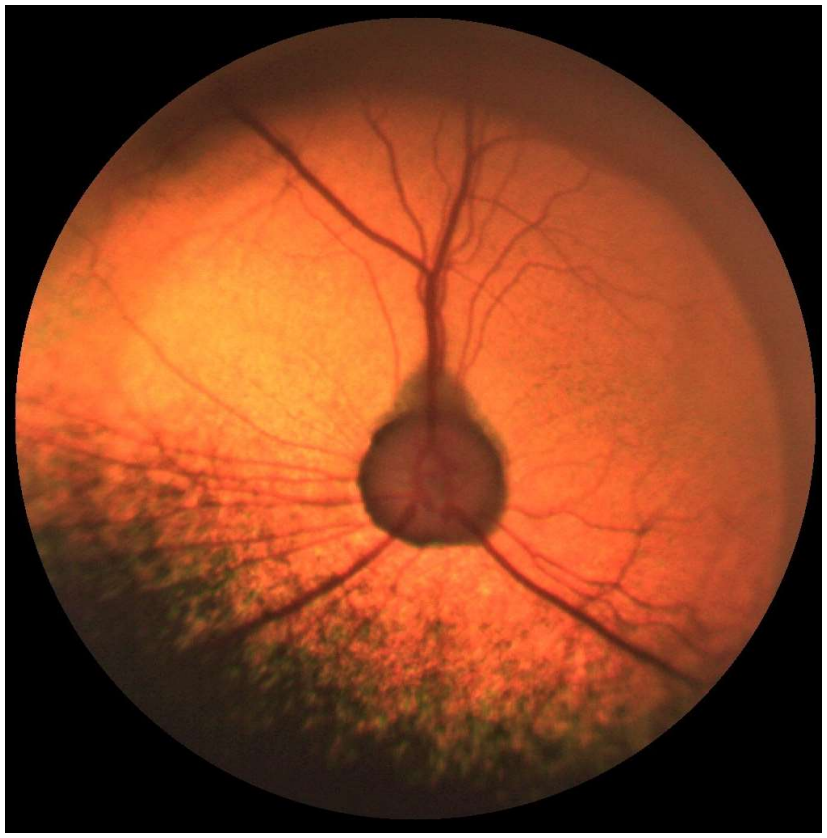
- "**7. Other"**, on the certificate, known and presumed hereditary eye anomalies (congenital/developmental, non-progressive) are mentioned. The terminology for the diseases as given in chapter 5. Definitions of this manual are to be used. These disease names are also used in "roll down" menus in the computerized forms.

Case 8: Bernese Mountain Dog, 9 months, male

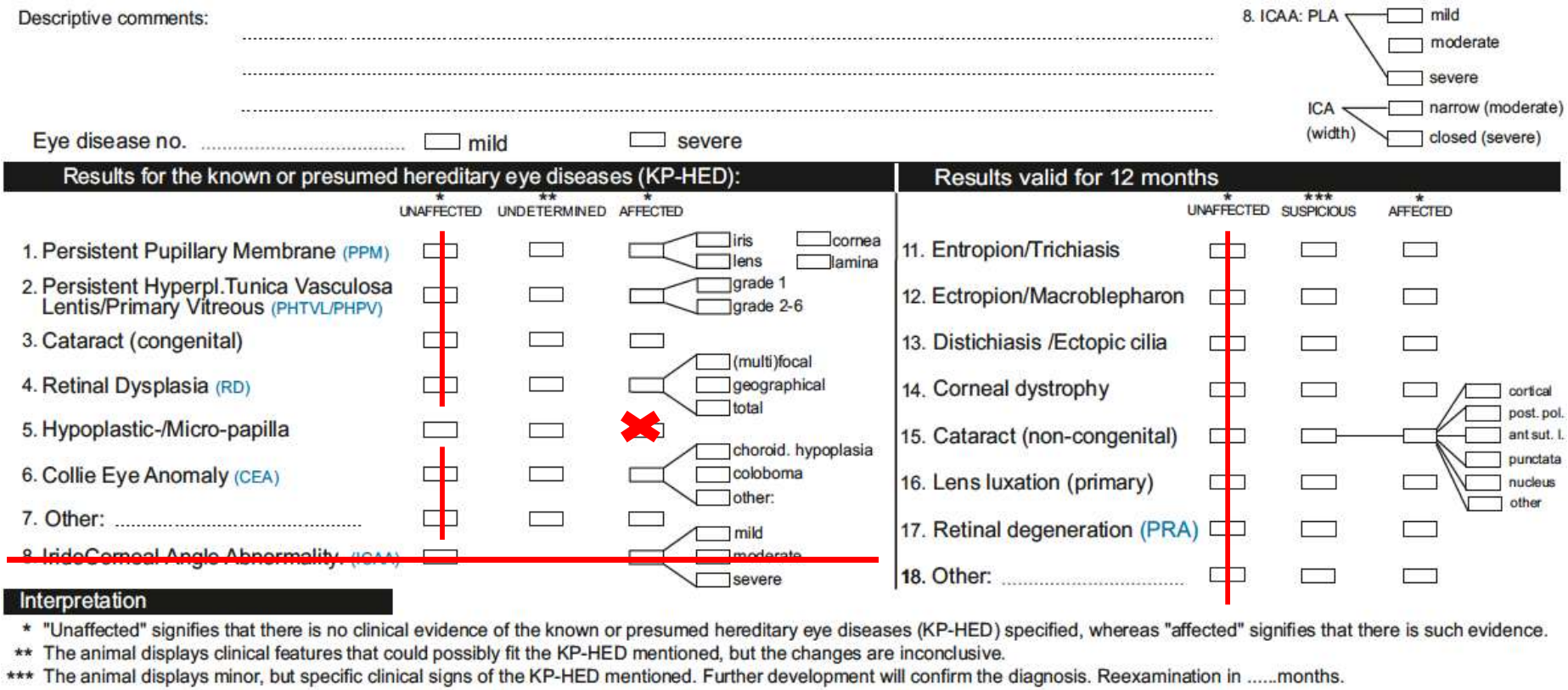
OD (blind)



OS

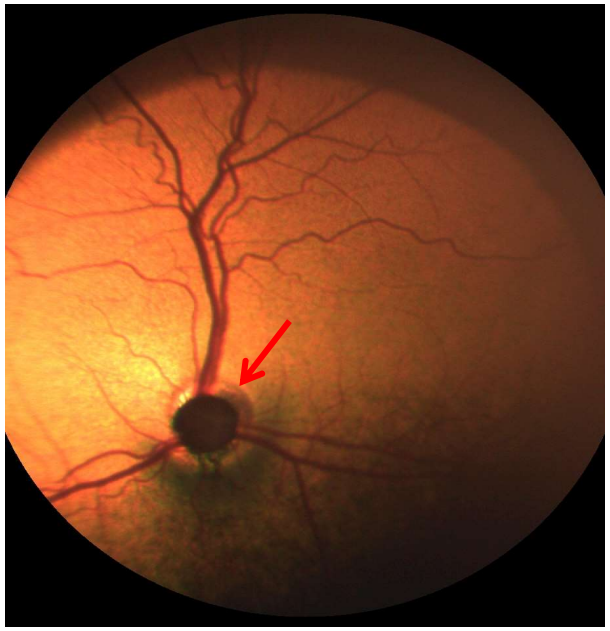


Case 8: Bernese Mountain Dog, 9 months, male



HED Manual 2017-04: Ch. 5 Definitions

- Hypoplasia-/ optic disc hypoplasia: presumed hereditary eye disease; congenital failure of development of the optic nerve which causes blindness and abnormal pupil response in the affected eye. Can often not be differentiated from micropapilla on a routine (dilated) ECVO eye examination

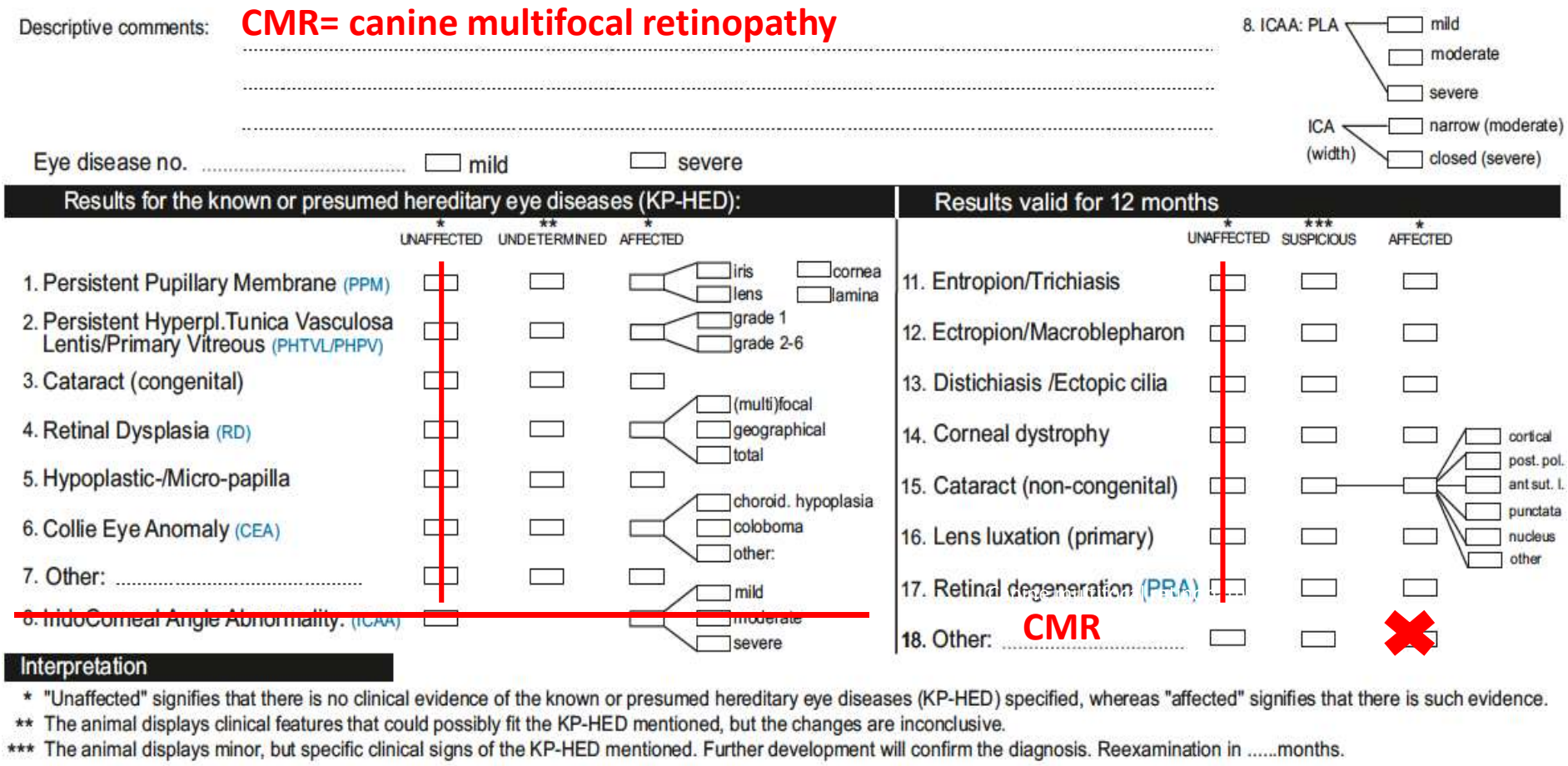


HED Manual 2017-04: Ch. 6 Guidelines

- **OD: Optic nerve hypoplasia:** see and use **hypoplastic papilla/optic disc**

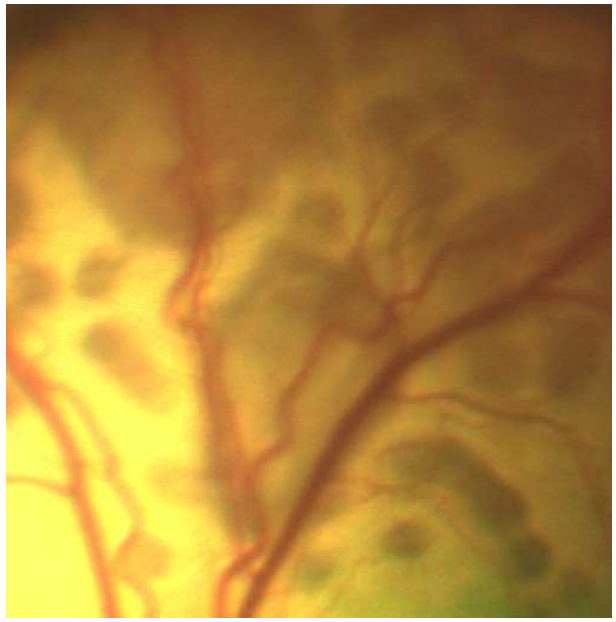
Case 9: Cane Corso, female, 8 months, OU

Case 9: Cane Corso, female, 8 months, OU



HED Manual 2017-04: Ch. 5 Definitions

- **Canine multifocal retinopathy (CMR):** known hereditary eye disease; autosomal mode of inheritance suspected. DNA-tests for specific breeds are available. Recognized as barely progressive, grey to tan bulging areas of circumscribed retinal detachments, generally more or less up to one optic disc diameter

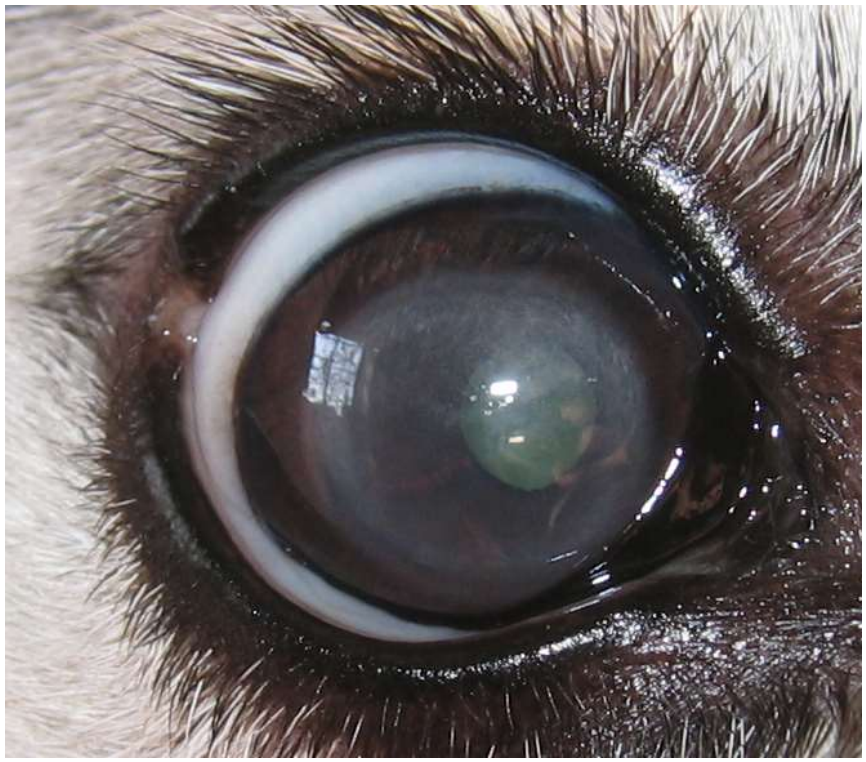


HED Manual 2017-04: Ch. 6 Guidelines

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

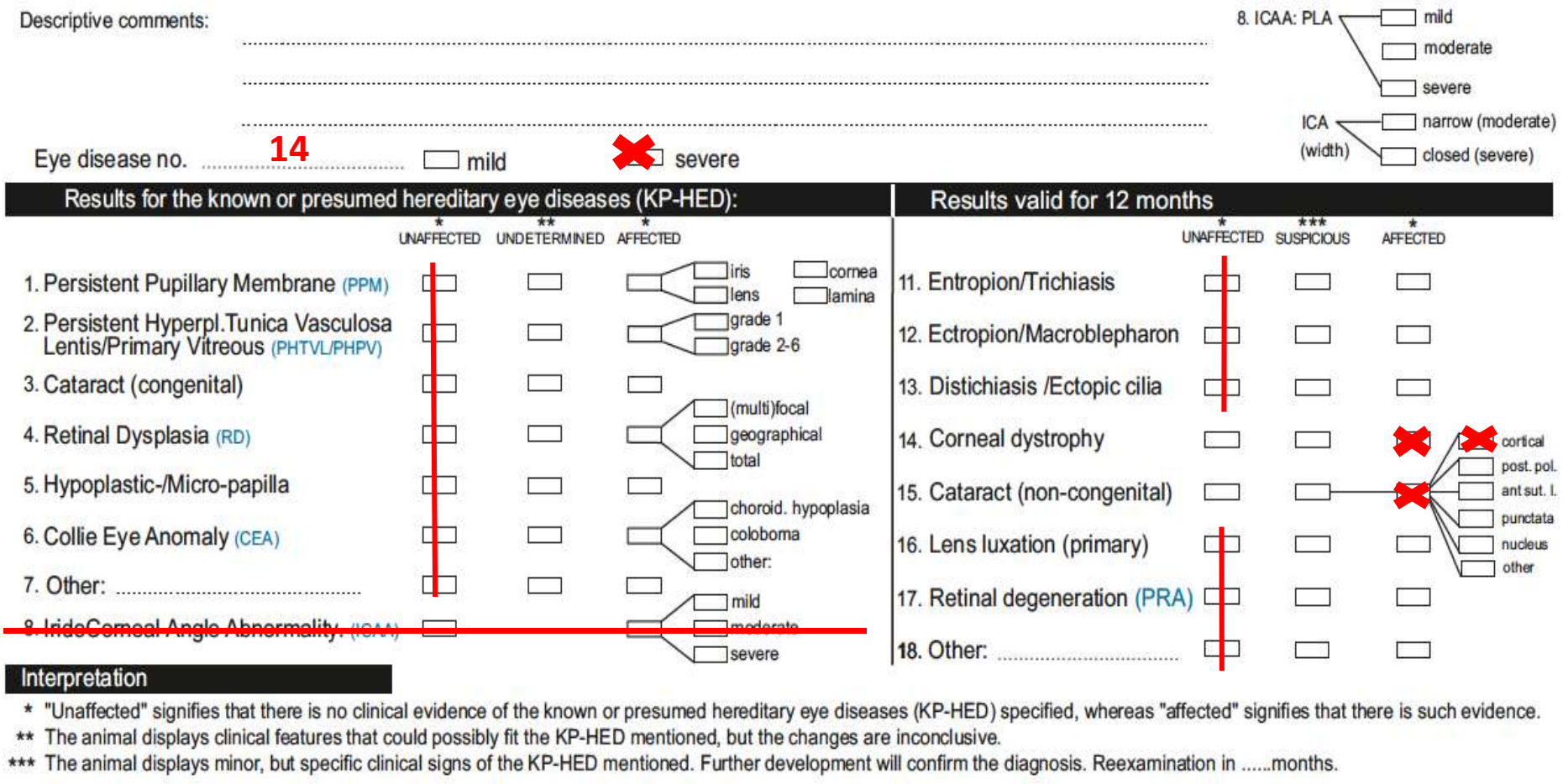
Case 10: Siberian Husky, 3 years, female, OU

OD



OS

Siberian Husky, 3 years, female



HED Manual 2017-04: Ch. 5 Definitions

- **Corneal dystrophy:** presumed hereditary eye disease; non-inflammatory **corneal** opacity in one or more of the **corneal** layers (epithelium, stroma, endothelium). It is usually bilateral but not always symmetrical. The onset in one eye may precede the other
- **Cataract:** any hereditary or non-hereditary, congenital or acquired, non-physiological opacity of the **lens** and/or its capsule. The defect may result in blindness if complete and bilateral. All bilateral or unilateral cataracts and especially cortical cataracts are known and presumed hereditary eye diseases except in cases known to be associated with trauma, other causes of ocular inflammation, metabolic disease, nutritional deficiencies, persistent pupillary membrane, persistent hyaloid artery or old age.
  DNA-tests for specific breeds are available.

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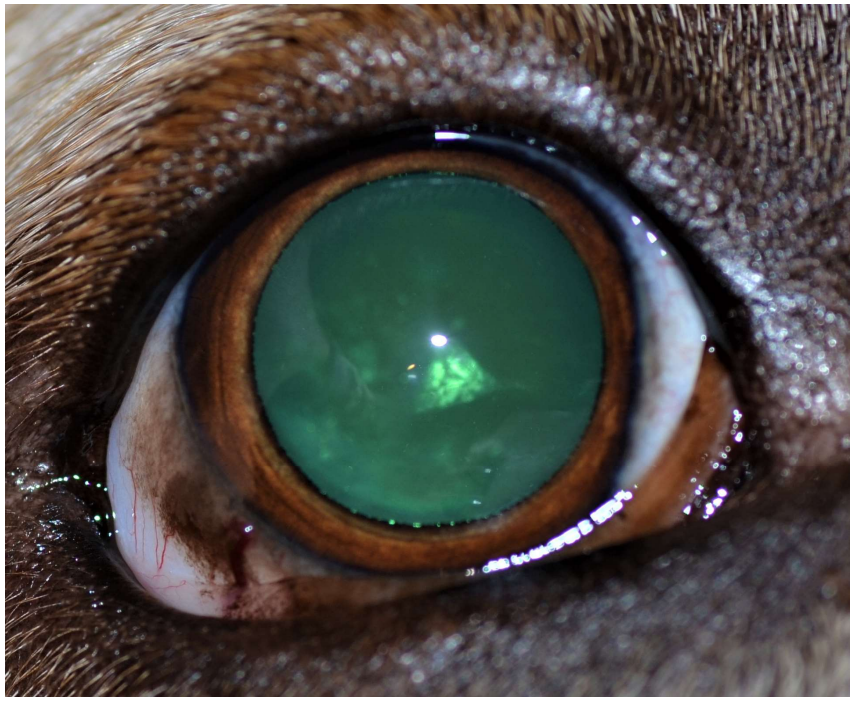
- **Corneal dystrophy** is to be ticked affected at no. 14 Corneal dystrophy, and the details described in the field Descriptive comments.

  Only if endothelial dystrophy or macular dystrophy or severe forms of stromal dystrophy (e.g. in Siberian Husky), is recognized, the examiner will also tick the box: “severe’ in the comment area.
- **Cataracts:** if observed in the period between birth and the 8th week of age the entity is ticked as congenital. Cataracts diagnosed at older age are ticked as non-congenital (acquired). If there is distinct proof the cataract is congenital in origin (e.g. associated PPM), the boxes for congenital and non-congenital cataracts can be ticked. It is strongly recommended to draw the cataract in the "pre-drawings" on the certificate, as seen from the anterior lens capsule (see separate instructions for drawing and filling the form). For the Scheme it is advised all bilateral or unilateral cataracts and especially cortical cataracts are presumed hereditary.

Case 11: Labrador Retriever, 14 months, female, OU

OD



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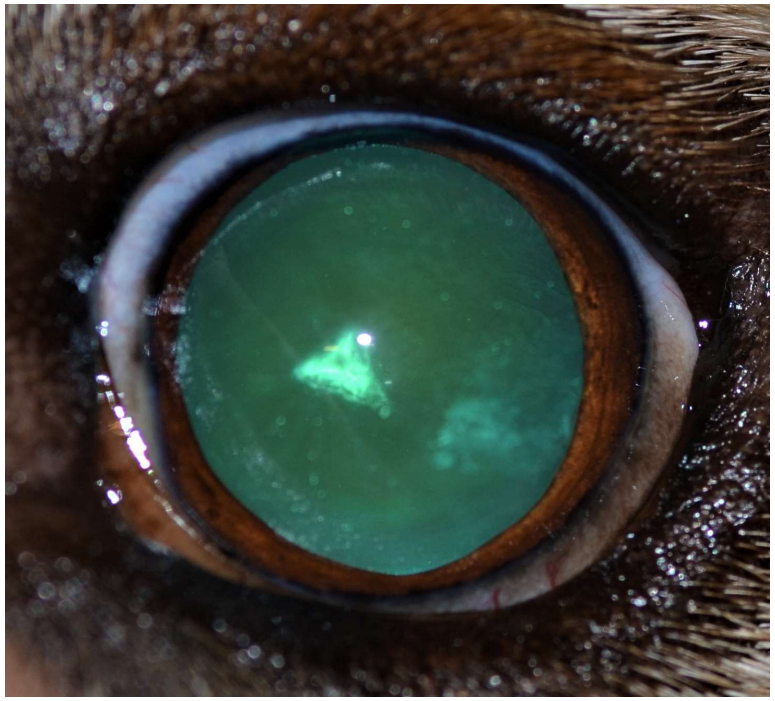
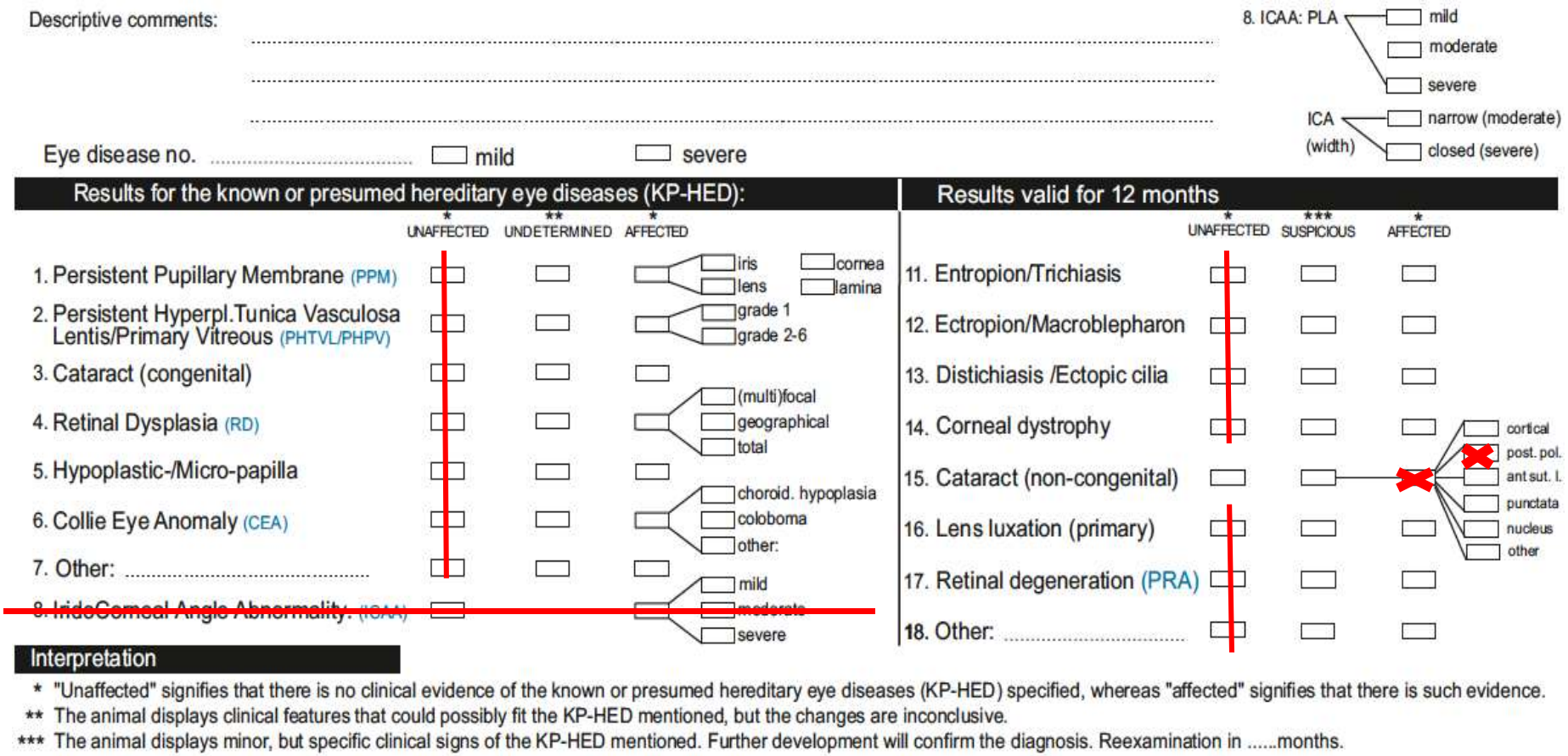
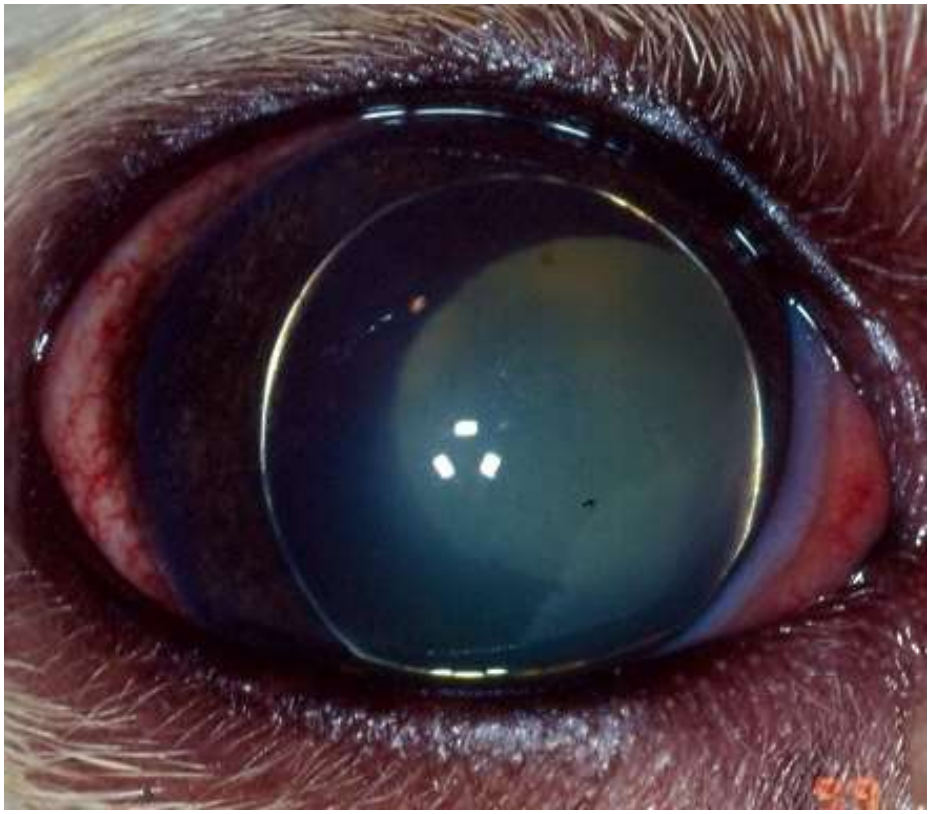


Case 11: Labrador Retriever, 14 months, female, OU



Case 12: Volpino Italiano, 4 years, male, OU

OD



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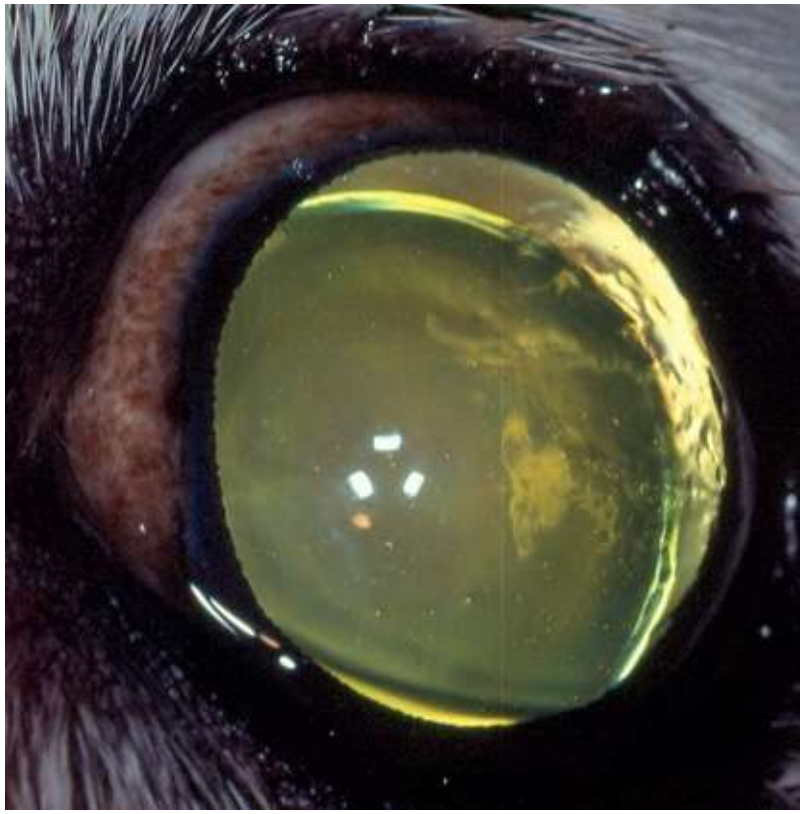
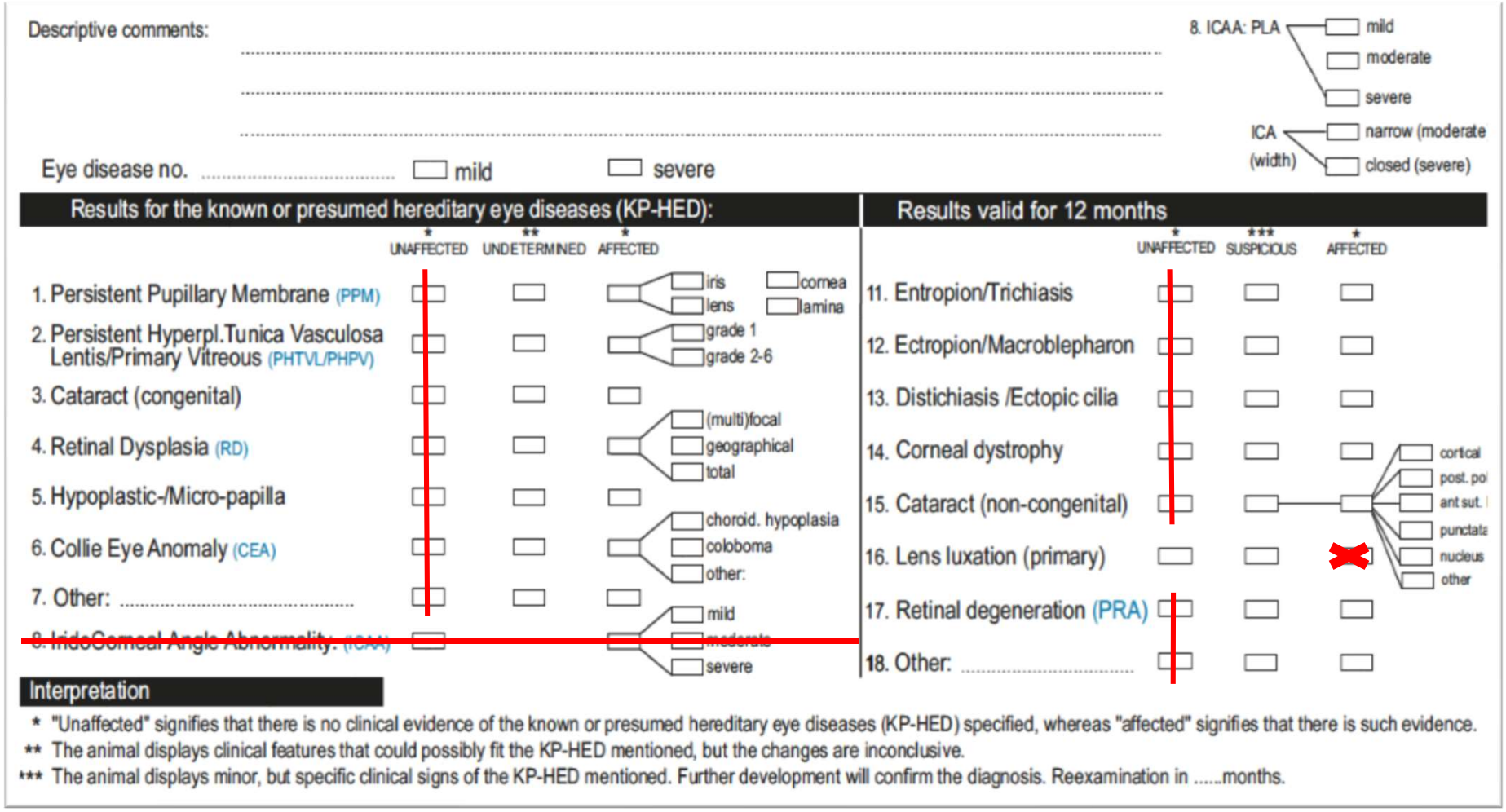


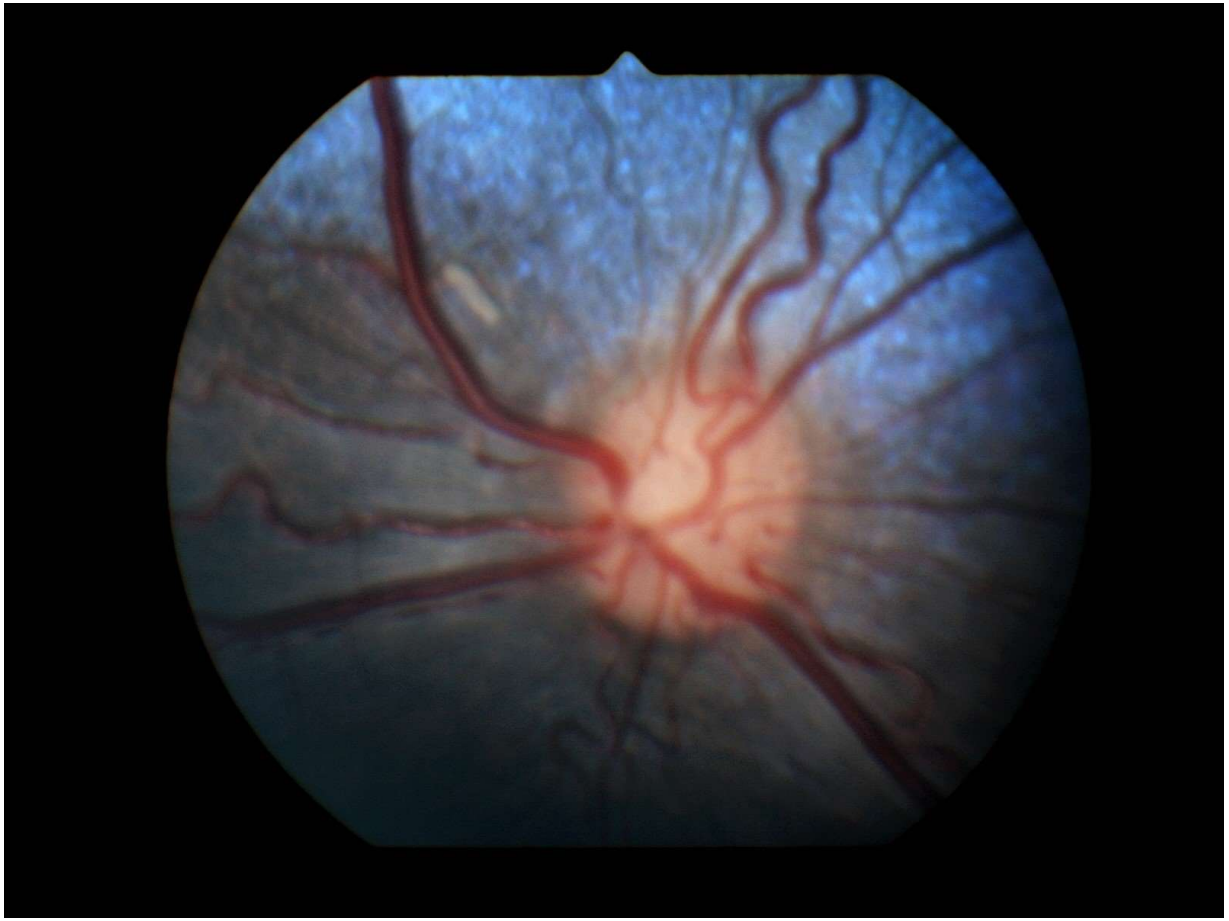
Case 12: Volpino Italiano, 4 years, male, OU



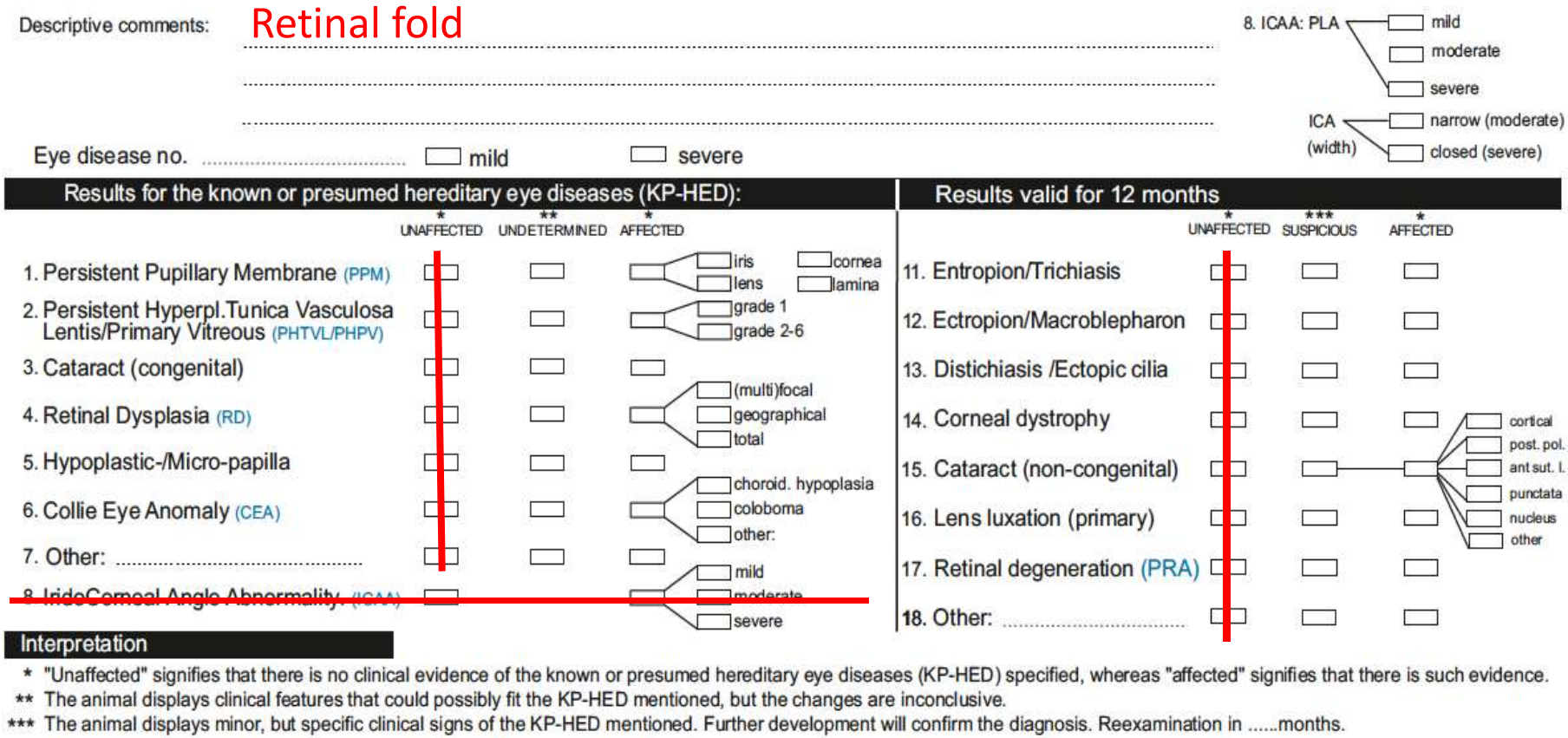
HED Manual 2017-04: Ch. 5 Definitions

Lens luxation (primary): known hereditary eye disease; partial (subluxation) or complete displacement of the **lens** from the normal anatomic site, in the fossa patellaris, behind the **pupil**. Lens luxation may result in elevated intraocular pressure (**glaucoma**) causing vision impairment or blindness. DNA-tests for specific breeds are available.

Case 14: Rough Collie, female 7 weeks, OS



Case 14: Rough Collie, female 7 weeks, OS



HED Manual 2017-04: Ch. 5 Definitions

- **Retinal folds: hereditary or nonhereditary** changes in the retina, can be neuroretinal folding due to hereditary factors or as sequelae post inflammation

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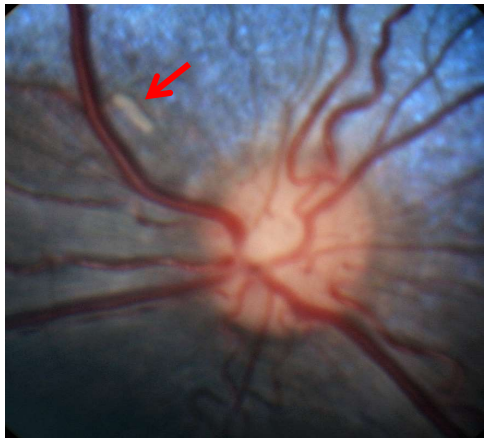
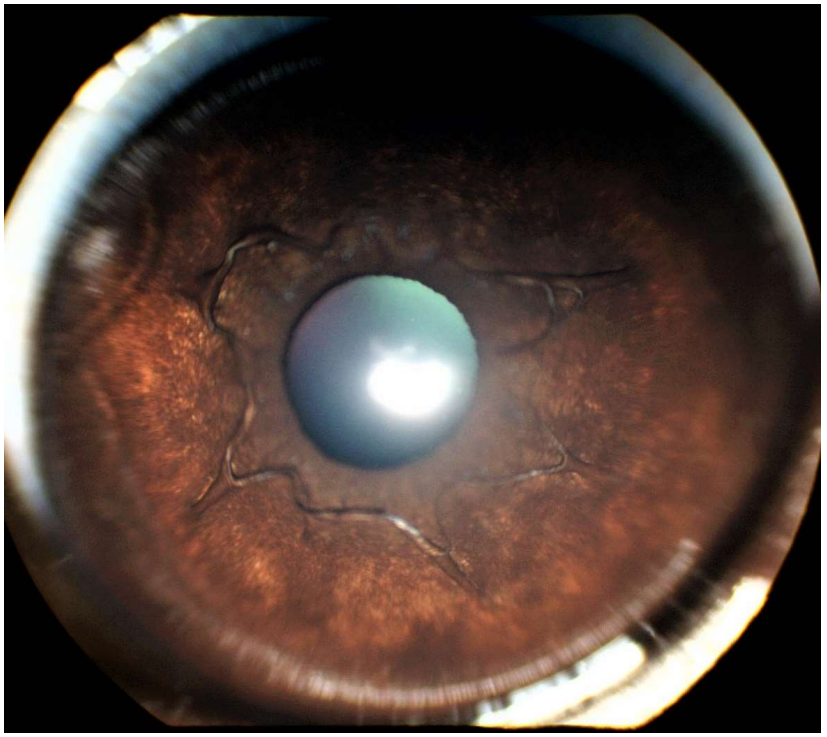
- In puppies, linear or round juvenile folds, usually in the peripapillary area, may be observed as a result in inequity in the relative growth rates of the optic cup and these folds resolve as the animal matures. These folds are not accurately referred to as dysplasia and should be ticked “unaffected”, 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”.



Case 15: Bouvier des Flandres, 2 months, male, OU

(abnormalities distinctly visible using 10 x magnification after pupil dilation)

OD



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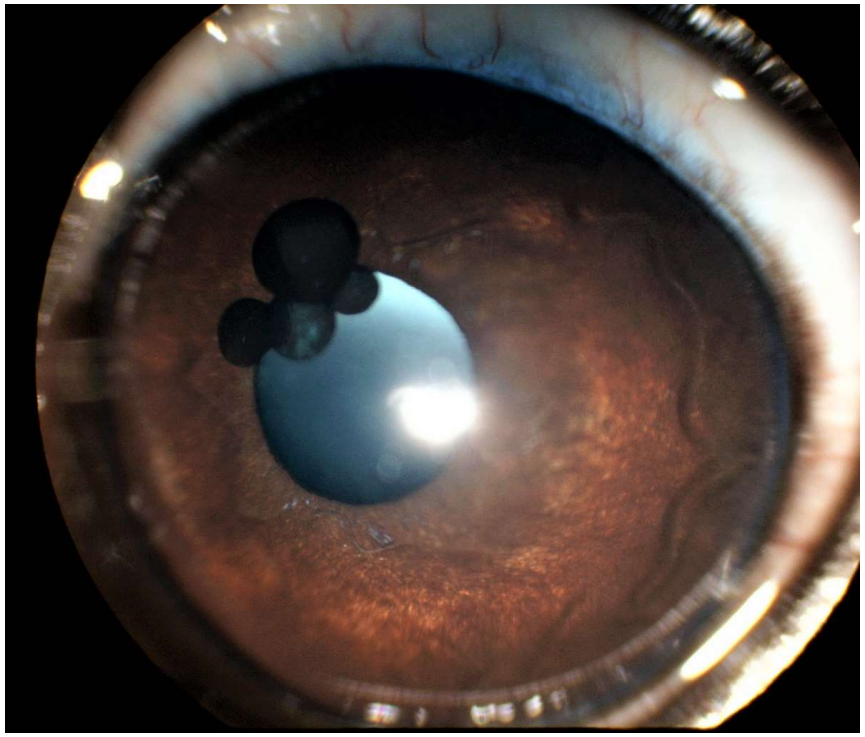
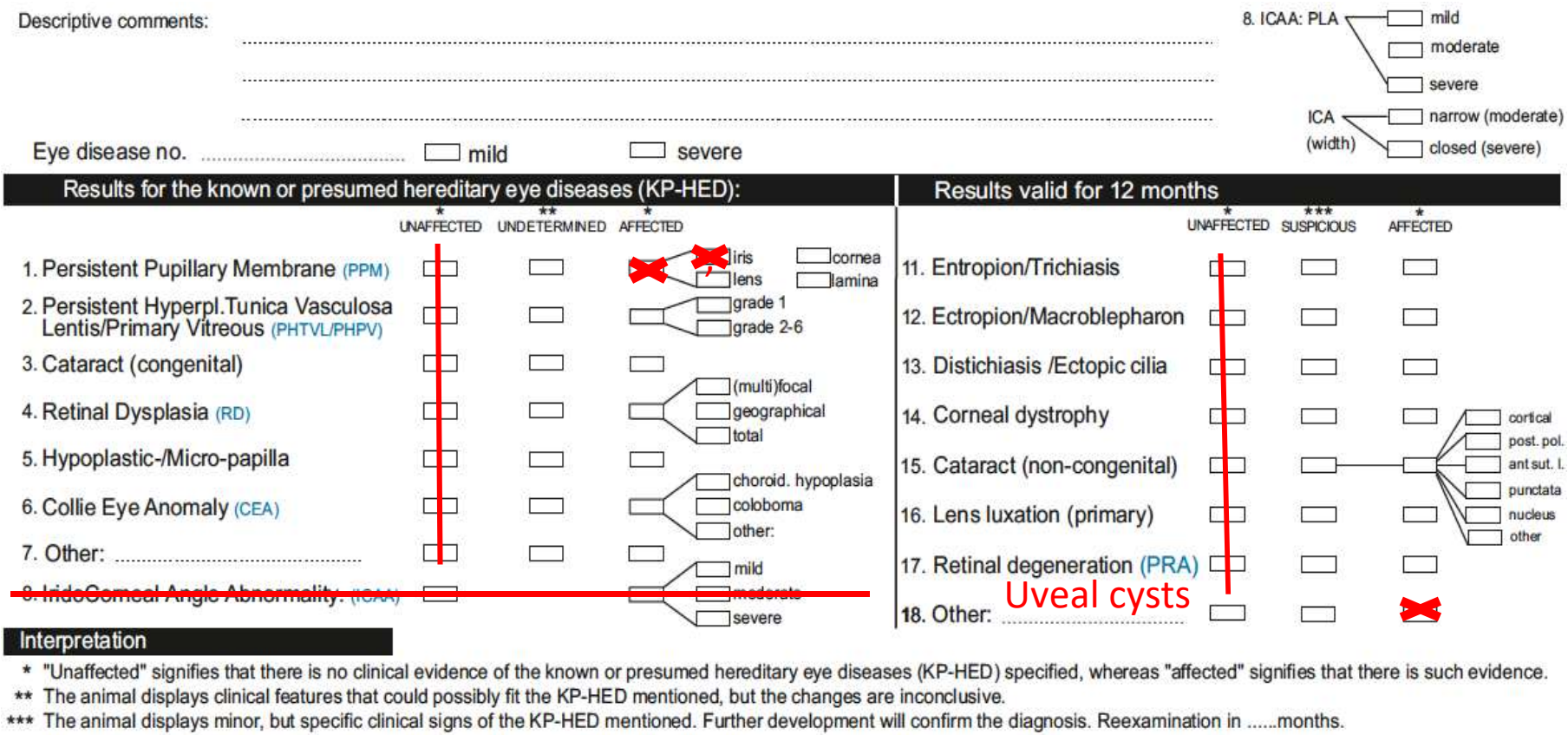


Case 15: Bouvier des Flandres, 2 months, male, OU

(abnormalities distinctly visible using 10 x magnification <u>after</u> pupil dilation)



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- **Persistent pupillary membrane (PPM):**presumed hereditary congenital eye disease in which blood vessel remnants of the embryological vascular network in the **anterior chamber** of the eye fail to regress which normally occurs during the first 4 to 5 weeks of life.These remnants may be found on the surface of the iris at the **collarette**, the lens capsule or against the corneal endothelium or strands may bridge from **iris** to iris, iris to **cornea**, iris to **lens**, with or without sheets of tissue in the **anterior chamber**. The last three forms pose the greatest threat to vision and, when severe, vision impairment may occur.
- **Uveal cyst:** presumed hereditary eye disease; usually pigmented membrane spheres of various sizes, arising from posterior pigmented epithelial cells of the iris/ciliary body and which remain attached, or break free floating as pigmented spheres in the **anterior chamber**. When reaching maximal size, cysts tend to adhere to the **endothelial** surface in the center of the **cornea**, thus causing visual impairment

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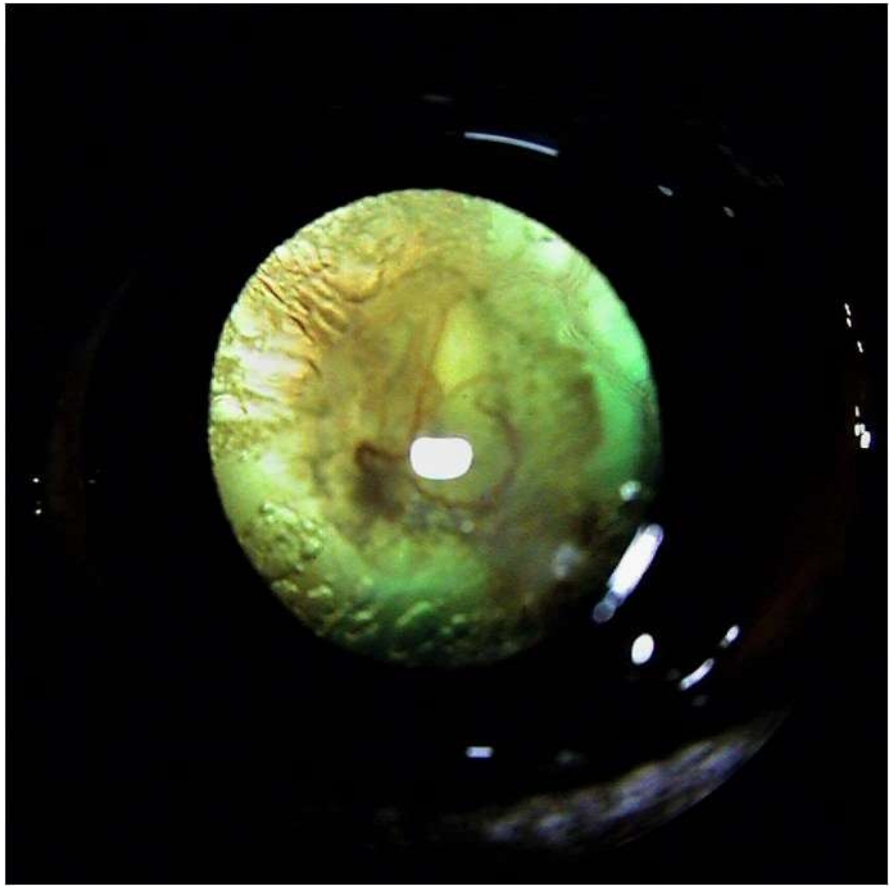
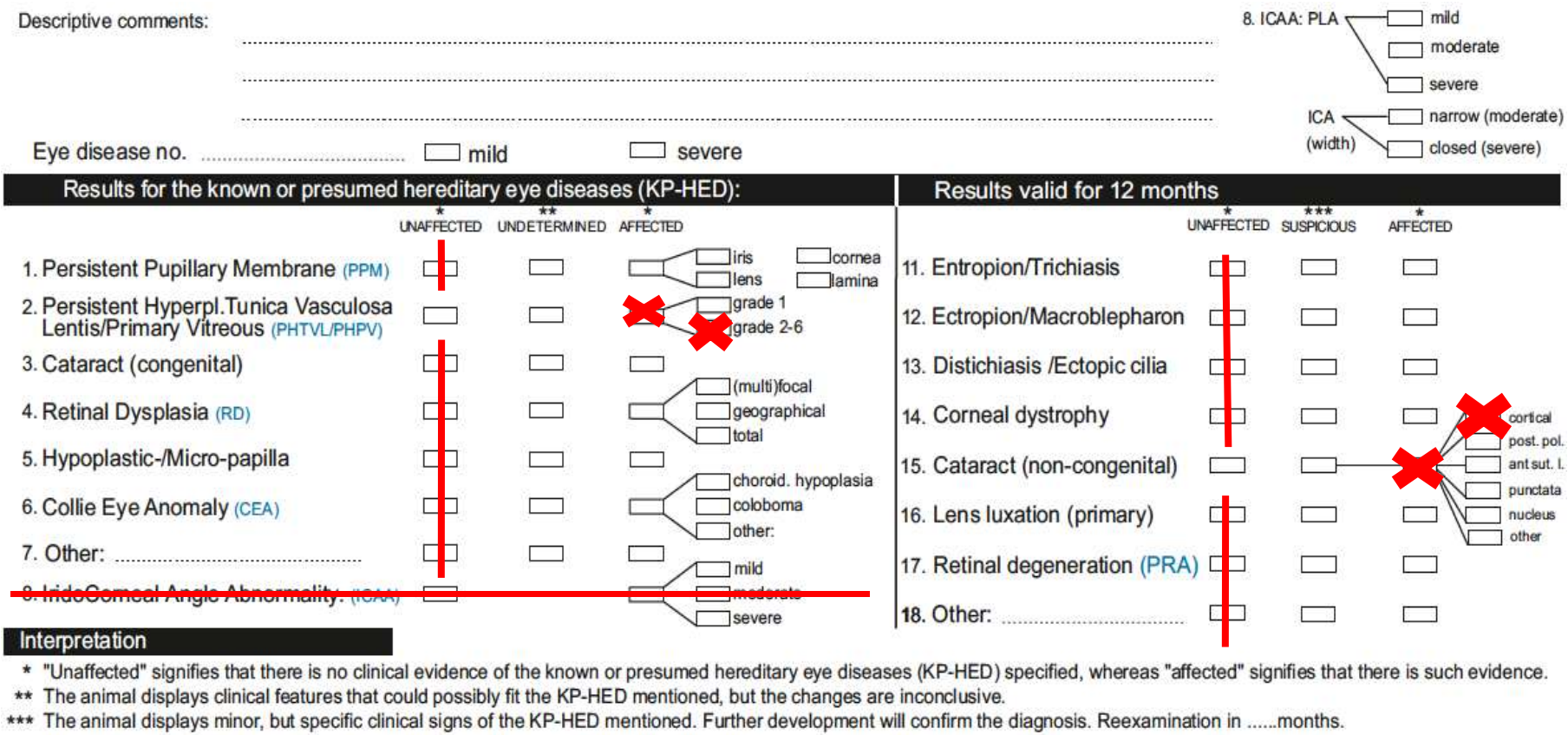
- **Remnants of the pupillary membrane**, still distinctly present after pupil dilatation, crossing the pupil, corneal, or with lens involvement, are ticked in the box for 1. PPM: “affected” and the respective box of other parts involved. Areas which can be involved are: retrocorneal (boxes PPM and cornea); strands from cornea to iris (boxes: PPM, cornea and iris); from iris to iris (boxes PPM and iris); iris to lens (boxes: PPM, iris and lens), connected to areas of cataract (also the box for congenital cataract is ticked); strands connected to a sheet/”spider web” of tissue in the anterior chamber (boxes PPM, lamina and other parts involved are ticked).
- **Uveal cysts:** “**18. Other”**, on the certificate for KP-HED**,** which are considered not to be congenital/developmental or which are progressive, and not yet named on the form, are mentioned. The available name of the disease in the list of ‘Definitions’ of this Manual (see chapter 5) is used.

Case 16 Dobermann, 2.5 years, male, OD



Case 16 Dobermann, 2.5 years, male, OD



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- **Persistent hyperplastic tunica vasculosa lentis/ persistent hyperplastic primary vitreous (PHTVL/ PHPV):**known or presumed hereditary, congenital eye disease which results from failure of regression of the embryologic vascular network, surrounding the developing **lens** and primary **vitreous**. The latter fails to regress within the first 2-3 weeks after birth. The defect is currently graded in 6 levels of severity, in which grade 1 is characterized by uni- or bilateral small, yellow to brown dots mainly centrally, retrolentally on the posterior capsule of the lens. These are stationary and do not affect vision. The more severe forms (2-6) usually occur bilaterally and cause visual impairment or blindness. Known hereditary e.g. in the Dobermann and the Staffordshire Bull terrier
- **Cataract:** any hereditary or non-hereditary, congenital or acquired, non-physiological opacity of the **lens** and/or its capsule. The defect may result in blindness if complete and bilateral. All bilateral or unilateral cataracts and especially cortical cataracts are known and presumed hereditary eye diseases except in cases known to be associated with trauma, other causes of ocular inflammation, metabolic disease, nutritional deficiencies, **persistent pupillary membrane, persistent hyaloid artery** or old age. DNA-tests for specific breeds are available.

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- ***PHTVL/PHPV:*** Minor, yellow-brown dots of fibrous tissue remaining retrolentally, more or less centrally on the posterior capsule of the lens (See fig. 21) are ticked as PHTVL/PHPV affected, and the specifying box as grade 1. These grade 1 dots are not to be confused with scattered pigment, retrolental near or on the posterior capsule of the lens. If they are unilateral, and of minimal degree, ‘undetermined’ is ticked.

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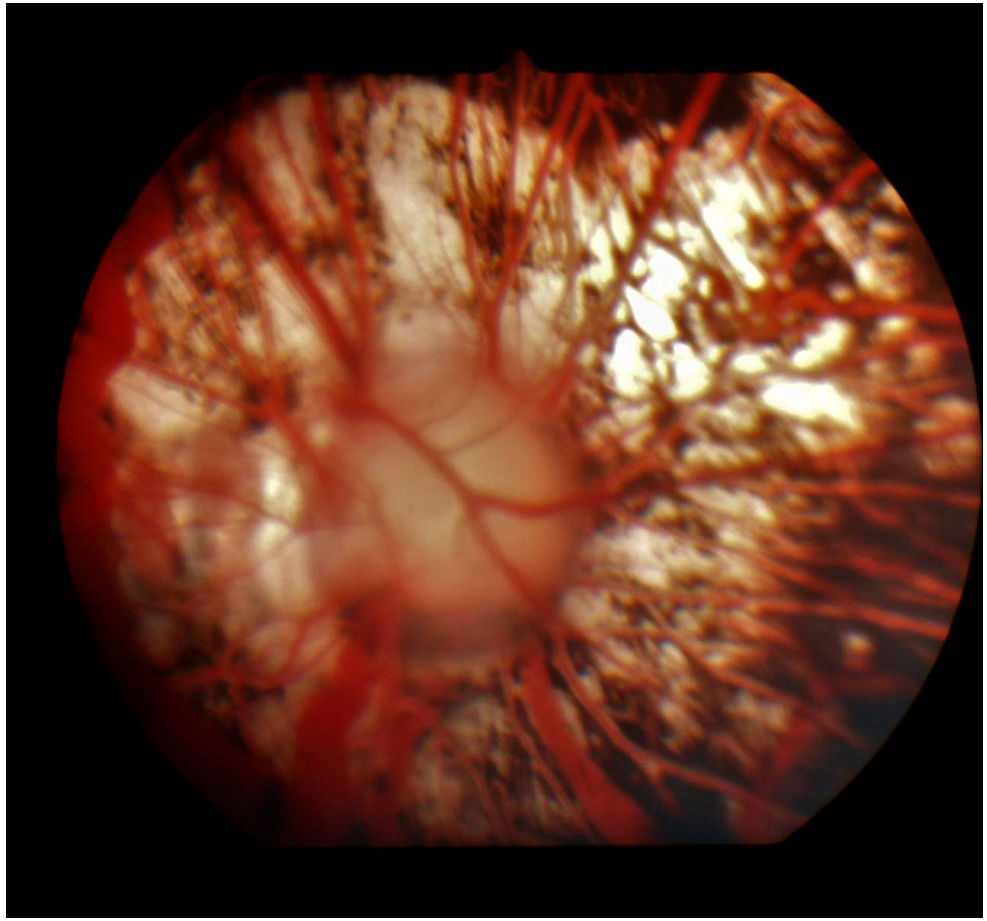
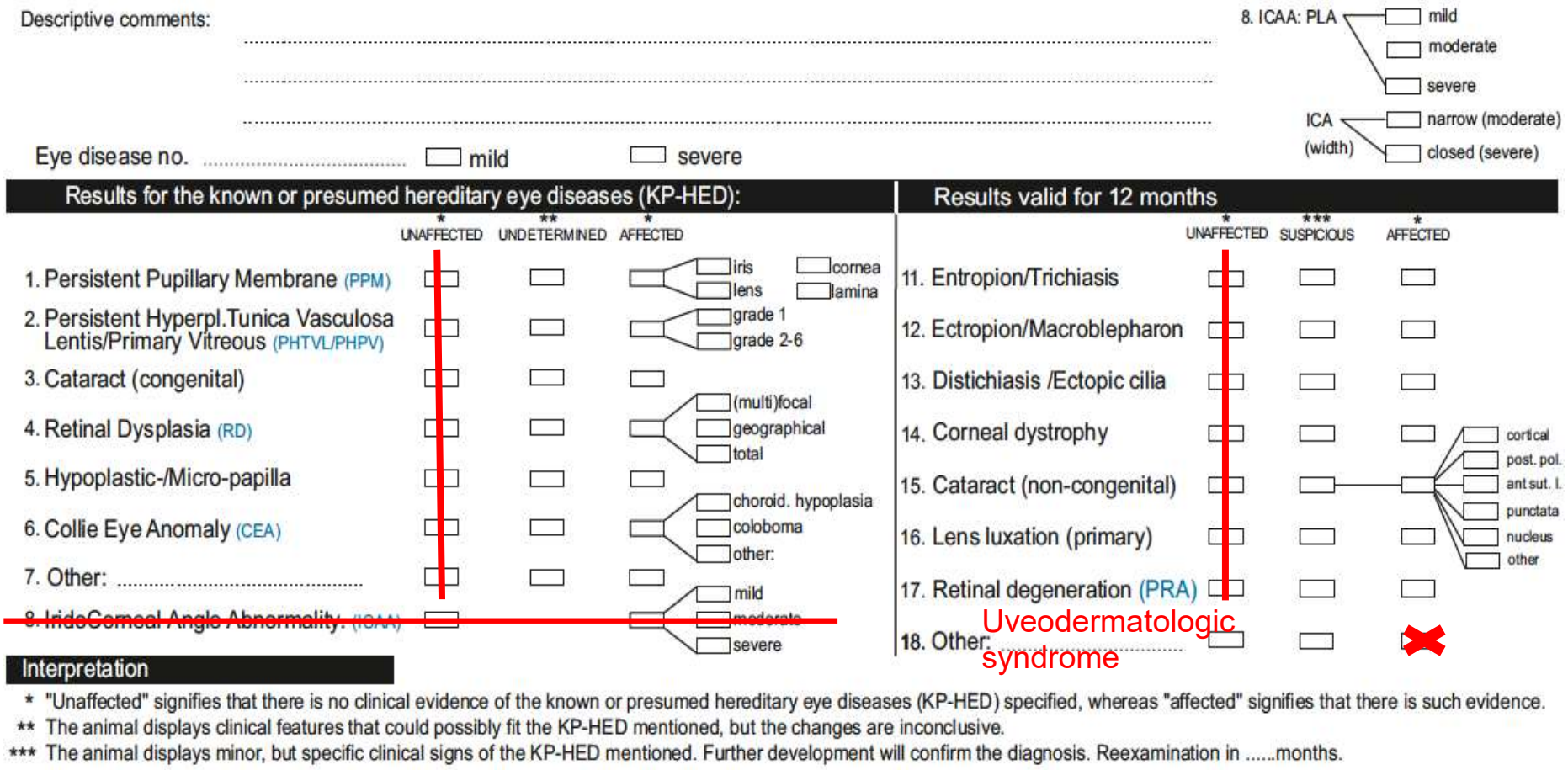
The severe forms (grades 2–6) usually occur bilaterally and may lead to visual problems. A plaque of white fibrovascular tissue can remain on the back of the posterior capsule, accompanied by grade 1 retrolental dots. In addition, other parts of the hyaloid system can persist: lenticonus, or even more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

Unilateral or bilateral grade 2-6 forms are ticked as PHTVL/PHPV ‘affected’ and the specifying box as grade 2-6.

Case 17: Dachshund, 3 years, male, OS



Case 17: Dachshund, 3 years, male, OS



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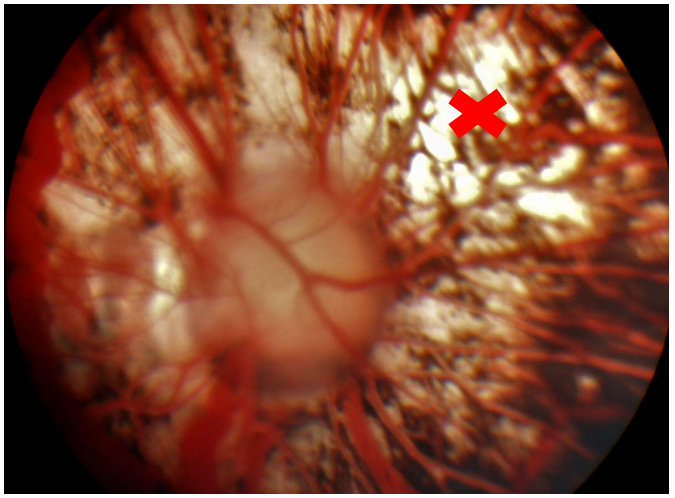
- **Uveodermatologic syndrome:** an **immune-mediated** syndrome of severe **uveitis** combined with dermal depigmentation (vitiligo) and hair depigmentation (poliosis). Secondary **glaucoma** and/or **retinal detachment** are frequent complications of this disease. Seen most commonly in the Akita Inu, Samoyed, Siberian Husky breeds. A similar syndrome is recognized in people and is called **Vogt-Koyanagi-Harada syndrome (VKH)**



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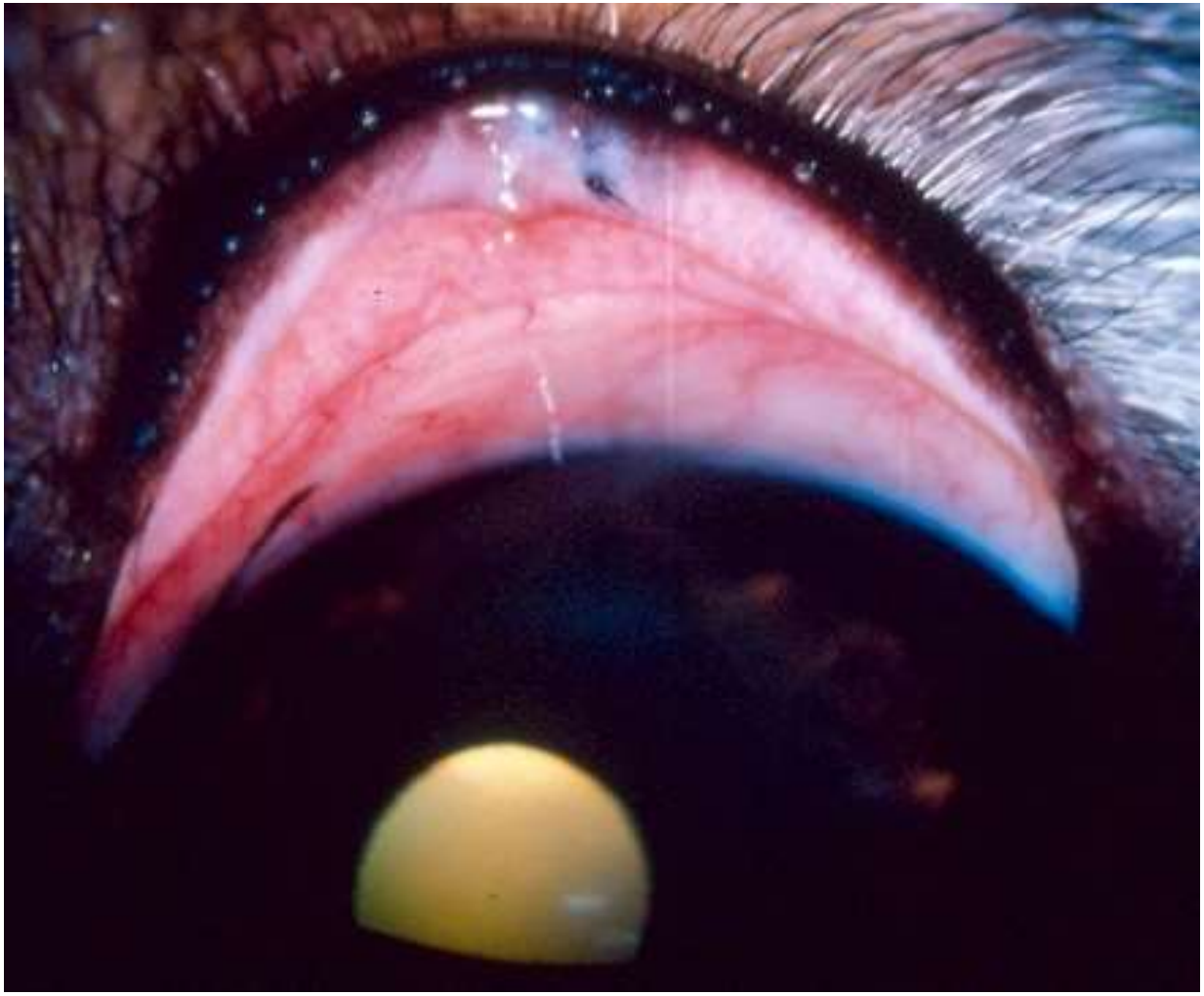
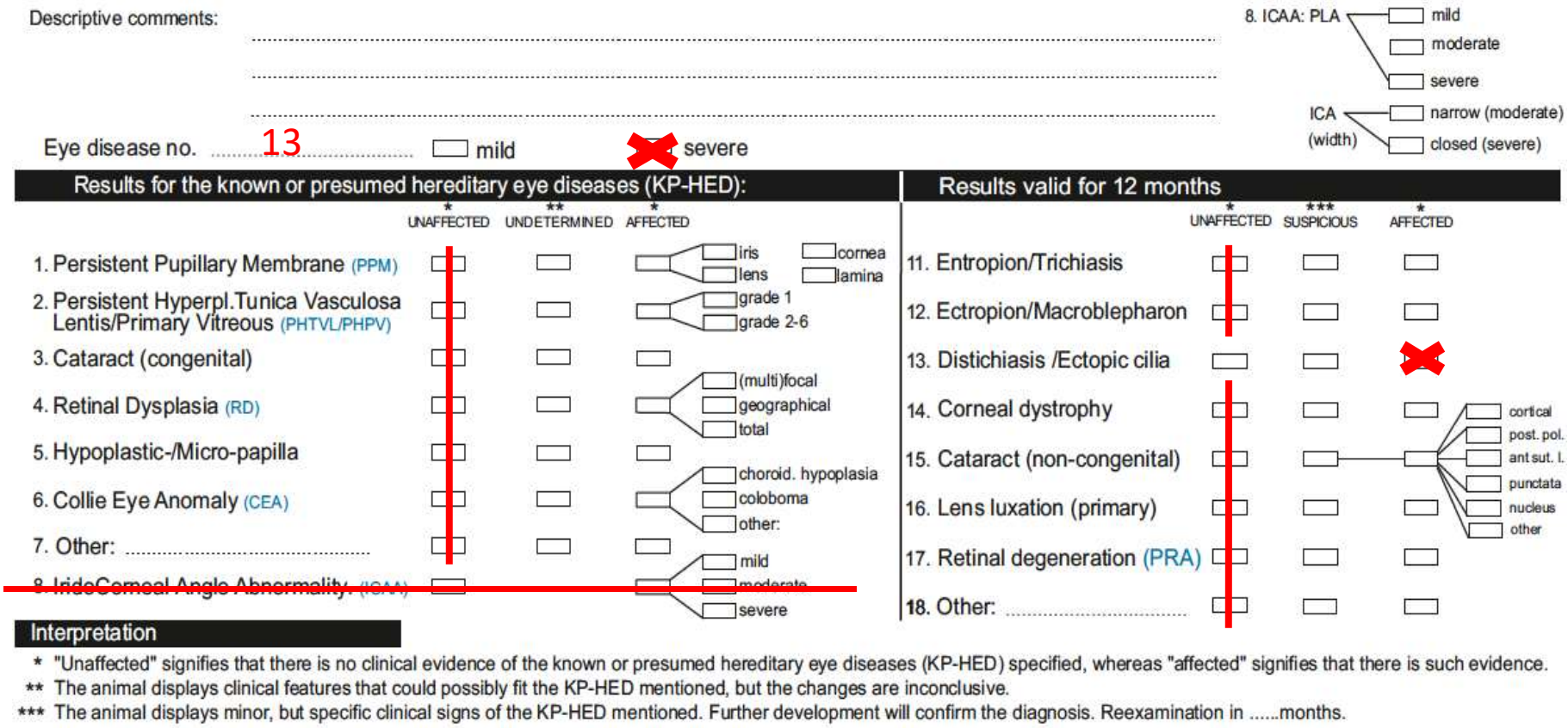
- For number “**18. Other”**, on the certificate for KP-HED**,** which are considered not to be congenital/developmental or which are progressive, and not yet named on the form, are mentioned. The available name of the disease in the list of ‘Definitions’ of this Manual (see chapter 5) is used.

Case 18: English Bulldog, 9 months, female



Case 18: English Bulldog, 9 months, female



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- **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.

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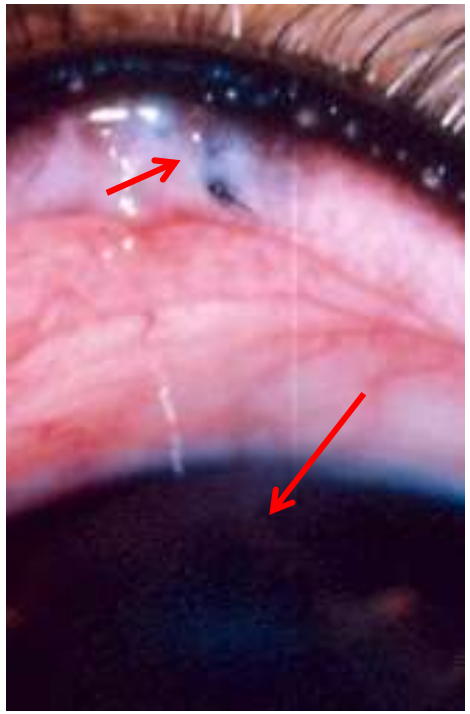
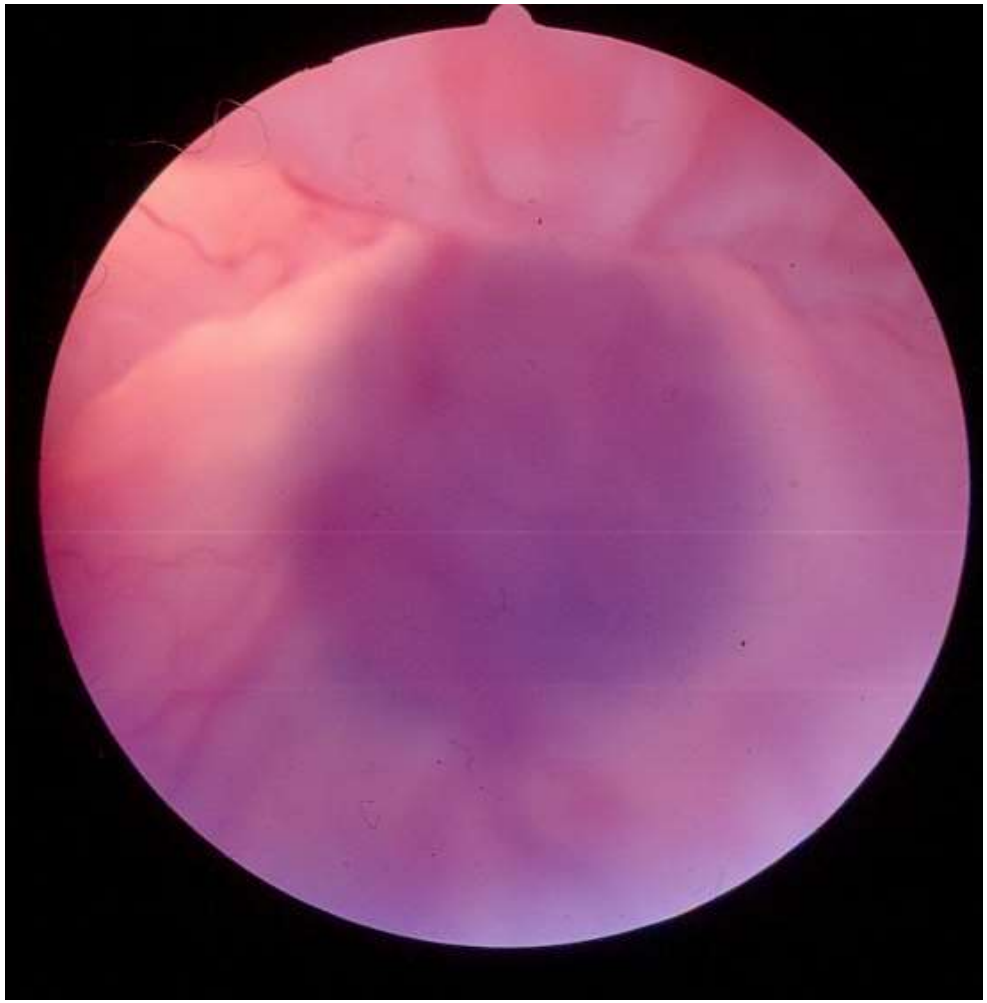
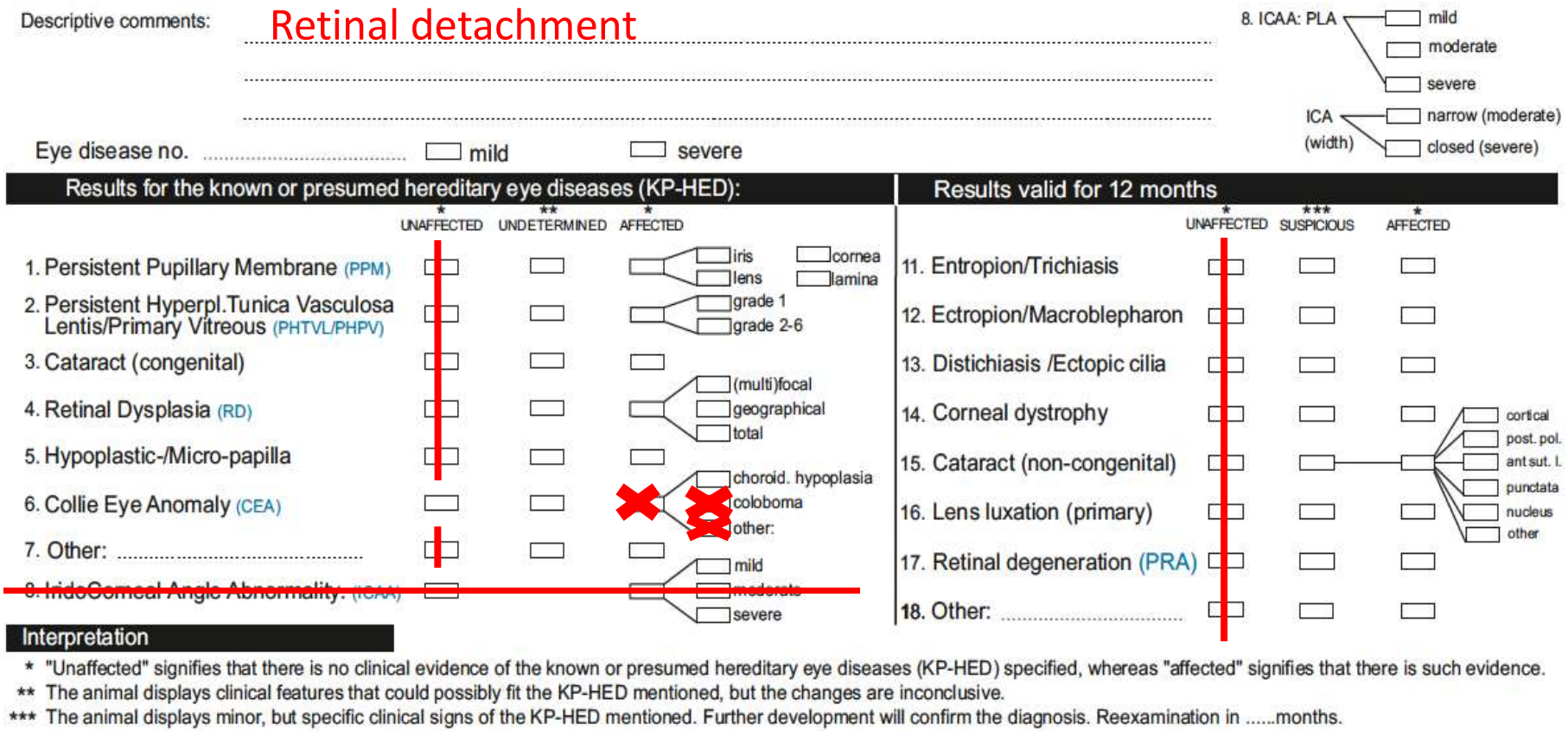
- Only if there are clinical signs of corneal irritation such as detritus on the distichia, corneal edema, corneal vessels, defects or pigmentation at the location of the distichia; hard stiff distichia and/or ectopic cilia, the examiner will also tick the box: “severe” in the comment area.



Case 19: Rough Collie, 3 months, male, OD



Case 19: Rough Collie, 3 months, male, OD



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- **Collie Eye Anomaly (CEA):** known hereditary congenital eye disease;a **congenital** syndrome of ocular anomalies mainly in Collie breeds affecting the choroid and sclera and indirectly the retina and optic disc. It is characterized by bilateral and often symmetrical defects including **choroidal hypoplasia (CH, CRD)** with or without **coloboma**, **retinal detachment** and intraocular hemorrhage. Vision varies with the degree to which an individual is affected and may be minimally compromised to having severe visual impairment or blindness. DNA-tests for choroidal hypoplasia in specific breeds are available.

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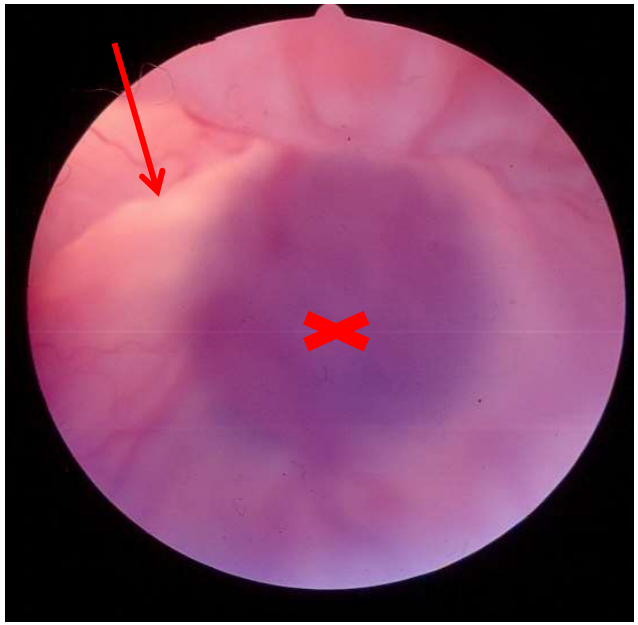
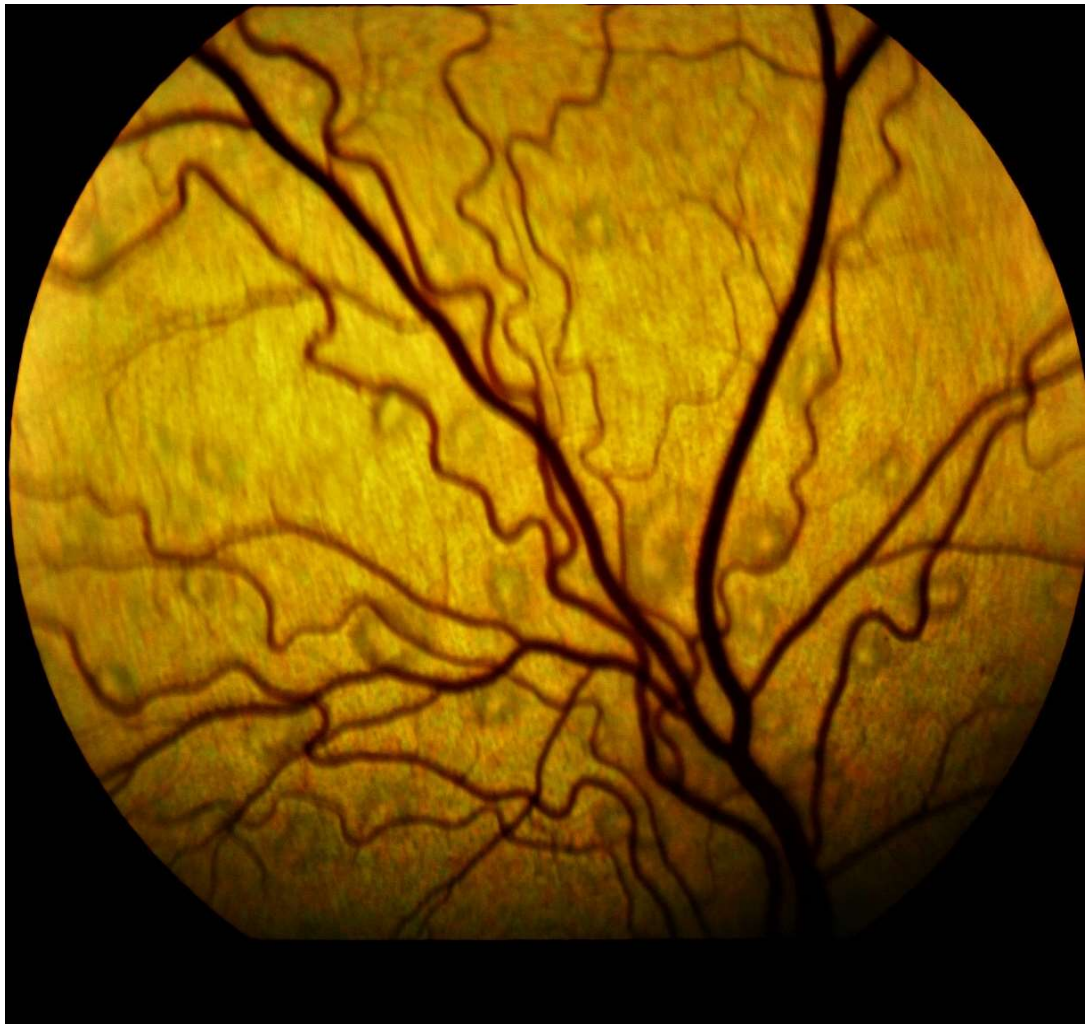
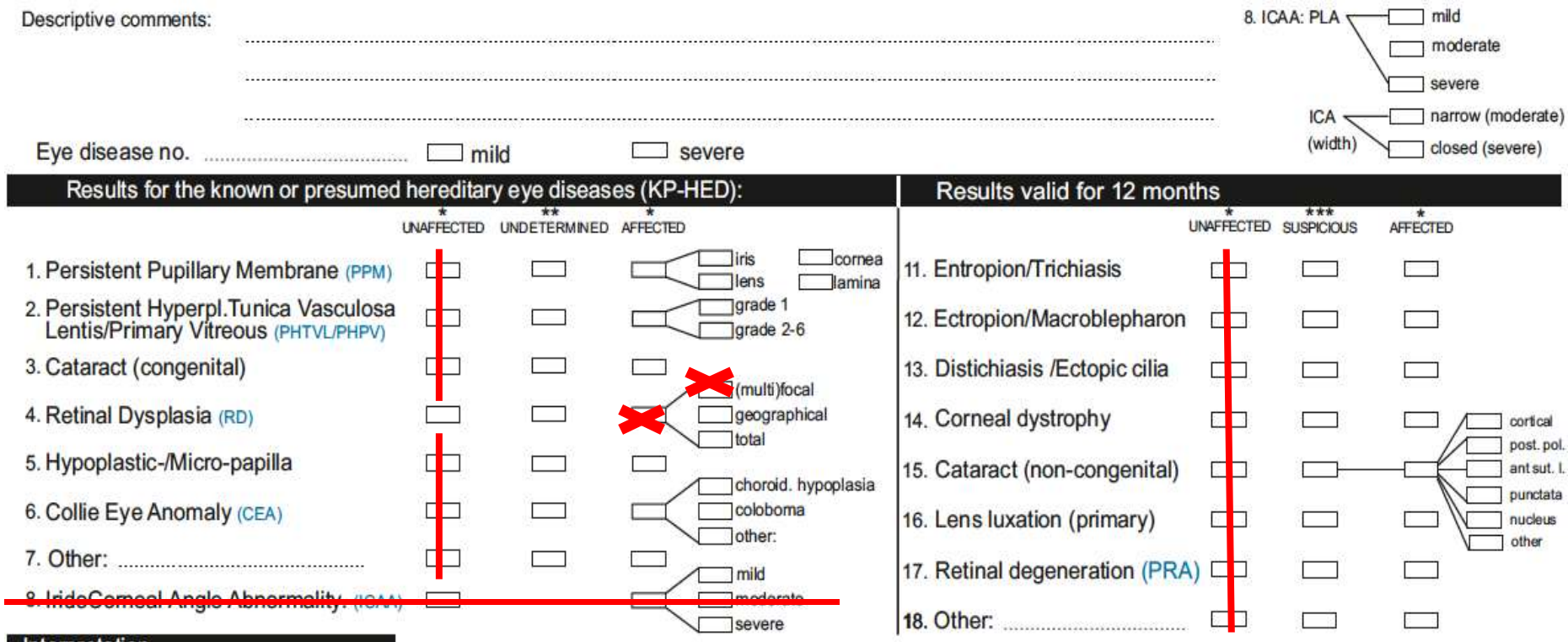
- In cases where the animal displays clinical features that could possibly fit this KP-HED, but the changes are not specific enough, the result of the examination is: ‘undetermined’. In such cases the breeder/owner is advised to distinguish the status of the animal by e.g. DNA testing. The box “Affected – other” has to be specified in the comment area of the ECVO certificate (retinal detachment or –haemorrhage).



Case 20: Australian Shepherd, 15 months, male, OD



Case 20: Australian Shepherd, 15 months, male, OD



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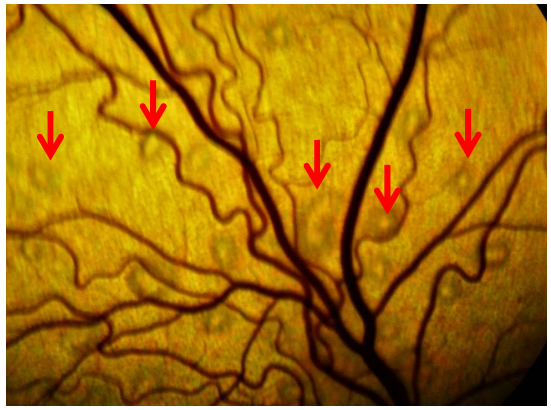
- **Retinal dysplasia- (multi)focal:** seen ophthalmoscopically as linear (vermiform), triangular, curved or curvilinear foci of retinal folding that may be single or multiple. When seen in puppies this condition may partially or completely resolve with maturity. Its significance to vision is unknown. The two other forms of retinal dysplasia (geographic and complete) which are known to be hereditary in some breeds and, in their most severe form, may cause blindness. The genetic relationship between folds and more severe forms of retinal dysplasia is undetermined



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