HED SESSION 2024 - KAHOOT



In cooperation with the Dutch Panel

Prof. Dr. Frans Stades Dr. Roswitha van de Sandt

This session and the information on how to correctly issue the ECVO certificate is based on the ECVO Manual 2023

Comments to Chapter 8 Vet Advice:

["]In Chapter 8, we choose the term « Veterinary Ophthalmologists' **advice** relating to HED control » and **intentionally avoid** the words "pass", "fail", "certifiable" and "registerable". The **ECVO does not prescribe breeding rules**, nor does it serve as a registry organization.

"Breed clubs and registry organizations prescribe breeding rules.



Case 1



English Cocker Spaniel, 2 years, m, bilateral slide: FS





English Cocker Spaniel, 2 years, m, bilateral slide: FS



PSENP= Posterior segment examination not possible

Comm=descriptive comments

15. Cataract (later onset)- mature, 7&18 PSENP; Comm: iris atrophy	 15: Cataract (later onset) cortical + nuclear, 7. Other: iris hypoplasia
 15. Cataract (later onset)-mature, 7. other: uvea hypoplasia + PSENP 	 15. Cataract (later onset) cortical + nuclear, 7. Iris hypoplasia, 7&18 PSENP; 7 severe



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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions:

Hypoplasia iris: KP-HED; characterized by congenital **thinning and/or absence** of iris (sphincter) tissue or **colobomatous** defects due to failure in closure of the optic fissure. It may be a separate disorder or associated with other ocular malformations. See and use iris hypoplasia.



HED SESSION

HED Manual Chapter 5 Definitions:

Cataract: is an opacity (generally whitish) in different shapes and sizes in the lens nucleus, and/or cortex and/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 KP- HEDs (except secondary cataracts)



HED Manual Chapter 6 Guidelines

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



HED Manual Chapter 6 Guidelines

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.

In a **total/mature cataract**, in case of cataract (later onset): tick at 15. Cataract (later onset)" the boxes affected and the specifying boxes **"cortical" and "nuclear"**.



Case 2



Pug, 7 y OU Slide: Djajadiningrat Utrecht University clinic



OU



Pug, 7 y OU Slide: Djajadiningrat Utrecht University clinic



ESO=

exophthalmus due to shallow orbit

 7. Other: Exophthalmos due to shallow orbit (ESO), 12. Ent/Tri, 18. Ocular melanosis 	 7. Other: ESO, 12. Ect/macrobl; 7. Severe Comm: Corneal scar
• 7. Other: ESO, 11. Ent/Tri, 12. Ect/	7. Other: ESO, 11. Ent/Tri, 12. Ect/
Macrobl; 7, 11, 12 severe	Macrobl; 11, 12 severe



"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.

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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED SESSION

HED Manual Chapter 5 Definitions:

Exophthalmos due to shallow orbit:

KP-HED in some brachycephalic breeds;

As part of the facial and skull bone deformities, the bony cavity is not deep enough to accommodate a normal sized globe. In combination with macroblepharon this leads to an anatomically **pathological exposure of the globe**, **recognizable by the visible sclera when the eye is directed straight ahead**



HED Manual Chapter 5 Definitions:

Macroblepharon:

KP-HED; an exceptionally large palpebral fissure. Macroblepharon in conjunction with laxity of the lateral canthal structures may lead to lower lid ectropion in combination with lateral and medial entropion and upper lid entropion and trichiasis. This may in severe cases result in diamondshaped eyes. Either of these conditions may lead to conditions associated with corneal exposure.



HED Manual Chapter 5 Definitions:

Trichiasis:

KP-HED or acquired abnormality of deviated hairs on a normal place around the lid fissure, irritating the conjunctiva, the free lid margin of the opposite lid and/or the conjunctiva and/or the globe. It is predominantly due to facial folds or due to misalignment of the eyelids.



Exophthalmos due to shallow orbit

is usually seen in combination with macroblepharon. If the sclera is visible in two or three quadrants or all around 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.



HED Manual Chapter 6 Guidelines

Only if the **sclera is visible all around** (with a normal-sized globe), also the box "**severe**" in the comments area is ticked.

In case of macroblepharon also tick at 12. Ectropion/ Macroblepharon

In case of additional Trichiasis also tick at 11. Entropion/Trichiasis



Case 3



Dachshund 8 y, bilateral , forget reflex on cornea slide: FS



OS



Dachshund 8 y, bilateral slide: FS forget reflex on cornea



	15. Cataract (later onset): juvenile- nuclear	15: Cataract(later onset)- other lens opacity - nuclear ring
•	15. Cataract (later onset)- mature nuclear	Nuclear sclerosis



Interpretation

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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED SESSION

HED Manual Chapter 6 Guidelines

Cataracts Classifications:

- **Refraction discontinuity zones**: fine (light-grey) regular circular/bend lines due to different refractive indices of the fibers of the embryonic, fetal, juvenile
- "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;



Case 4



Sussex Spaniel, 3 y, m, bilateral, slitlamp photo of the posterior lens capsule slide: FS



OS



Sussex Spaniel, 3 y, m, bilateral, slitlamp photo posterior lens capsule slide: FS



▲ 3. Cataract-(congenital) – post pol.	 3. Cataract (congenital) + 7. Persistent hyaloid artery
Comm: Persistent hyaloid artery	 3. Cataract (congenital) + 7. Persistent hyaloid artery; 7. severe



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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions

Persistent hyaloid artery (PHA):

KP-HED; congenital defect resulting from abnormalities in the development and regression of the hyaloid artery. The blood vessel remnant can be present in the vitreous as a small patent vascular strand (PHA) or as a non-vascular strand that appears grey-white (persistent hyaloid remnant).



HED Manual Chapter 6 Guidelines

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 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.



HED Manual Chapter 6 Guidelines

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.



Case 5



Lapponian Shepherd 2y, OS Slide: FS



OS



Lapponian Shepherd 2y, OS Slide: FS



 18. Other: Retinopathy Canine multifocal (CMR) 	◆ 4. Retinal Dysplasia- Geographical
• Comments: (Chorio-) retinopathy, inheritance under investigation	18. Other: Chorioretinopathy, primary retinal disease



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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.


HED Manual Chapter 5 Definitions

Canine multifocal retinopathy (CMR):

KP-HED; 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 Chapter 6. Guidelines

18. Other: Retinopathy, Canine multifocal (CMR) Breeds: Great Pyrenees, Coton de Tulear, Eng-, French-, Bull mastiff, Lapponian Shepherd

Also see ECVO Retina Atlas 2022



HED SESSION

Not to be confused with e.g. Retinal Dysplasia: a. Focal/Multifocal





b. Geographical



Case 6









 3. Cataract-(congenital) – post pol. 	 2. PHTVL/PHPV-Grade 2-6 ; Comm: TVL ant-persist.
• 2. PHTVL/PHPV-Grade 2-6 + Lens	2. PHTVL/PHPV- Grade 2-6; Comm: TVL
hypoplasia; Comm: PPM	ant-persist.+ Cataract + lens hypoplasia



Interpretation

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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions

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

KP-HED; 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 of fibrous tissue 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



HED Manual Chapter 5 Definitions:

Tunica vasculosa lentis (TVL): embryonic vascular network which surrounds the lens; The TVL, vasa hyaloidea propria and the pupillary membrane normally fully regress between 2 to 4 weeks after birth, starting from post to anterior, except a minor pig-tail-like remnant attached just below the center of the posterior lens capsule, extending into the vitreous. Persistence of one or more portions is seen. The most common is persistence of parts of the anterior portion, the pupillary membrane, which is referred to as persistent pupillary membrane (PPM).



TVL persistens ant.: on iris connecting to the collarette







HED Manual Chapter 6 Guidelines

PHTVL/PHPV grades 2-6:

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".



HED Manual Chapter 6 Guidelines

PHTVL/PHPV grades 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".



HED Manual Chapter 6. Guidelines

Fibrovascular tissue + dots









HED Manual Chapter 6 Guidelines: (PHTVL/PHPV): 2-6

lens hypoplasia



elongated ciliary processes microphakia













HED Manual Chapter 6. Guidelines:

Extreme Hyaloid Art. Persist.: Artery + Vein





HED Manual Chapter 8 Vet Advice

2. Persistent hyperplastic tunica vasculosa lentis/persistent hyperplastic primary vitreus (PHTVL/PHPV):

- Grade 1: OPTIONAL (NL: grade 1 x unaffected)
- Grade 2-6: NO BREEDING from the affected animal

(In NL PHTVL/PHPV in Dobermann is eradicated)



Case 7



Bobtail, 12 weeks, m, unilateral , fundus no abnormalities.

slide: FS





Bobtail, 12 weeks, m, unilateral, fundus no abnormalities.

slide: FS



	2. PHTVL/PHPV-Grade 2-6	◆ 15. Cataract (later onset) – post polar
•	3. Cataract (congenital)	7. Other: Cataract (congenital)- post polar



Interpretation

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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED SESSION

HED Manual Chapter 5 Definitions

Cataract

is an opacity (generally whitish) in different shapes and sizes in the lens nucleus, and/or cortex and/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 KP-HEDs (except secondary cataracts)



HED Manual Chapter 6 Guidelines

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)".



Case 8



Belgian shepherd Tervueren, 4 y, f, bilateral

slide: FS



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Belgian Shepherd Tervueren, 4 y, f, bilateral slide: FS

	18. Other: Plasmoma/plasmacellular conjunctivitis, Keratitis pigmentary	18. Other: Keratitis, pigmentary
•	18. Other: Keratitis, chronic superficial CSK/Pannus	18. Other: Plasmoma/plasmacellular conjunctivitis



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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED SESSION

HED Manual Chapter 5 Definitions:

Chronic superficial keratitis(CSK)/Pannus

KP-HED; bilateral inflammatory disease of the cornea which usually starts as a greyish haze at the inferior or inferio-temporal cornea, followed by the formation of a vascularized subepithelial opacity that begins to spread towards the central cornea; pigmentation follows the vascularization. Vision impairment occurs, if severe.

The disease can be seen with concurrent **plasmoma**/ plasmacellular conjunctivitis and/or medial canthus erosion



Case 9





OS



slide: FS



	14. Corneal dystrophy; 14. severe; comm: C. dys. macular	 14. Corneal dystrophy; 14. severe - comm: C. dys. stromal
•	14. Corneal dystrophy; 14. severe; comm: C. dys. endothelial	 14. Corneal dystrophy; comm: C. dys. Stromal



Interpretation

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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions

Corneal dystrophy:

KP-HED; non-inflammatory corneal opacity in one or more of the corneal layers (epithelium, stroma, endothelium). It is usually bilateral but not always symmetrical.

Corneal dystrophy, epithelial/stromal: KP-HED; non-inflammatory corneal opacity (white to grey with crystalline appearance) in one or more of the corneal layers. Often it is associated with deposits of cholesterol and other lipids (or fats) within the cornea.





HED Manual Chapter 5 Definitions

Corneal dystrophy, epithelial/stromal:

KP-HED; non-inflammatory corneal opacity (white to grey with crystalline appearance) in one or more of the corneal layers. Often it is associated with deposits of cholesterol and other lipids (or fats) within the cornea.





HED SESSION

HED Manual Chapter 5 Definitions

Corneal dystrophy, endothelial:

KP-HED; abnormal loss of the inner lining (endothelium) of the cornea causing progressive fluid retention (edema) leading to increased corneal thickness, keratitis, corneal clouding and decreased vision.




HED SESSION

HED Manual Chapter 5 Definitions

Corneal dystrophy, macular:

KP-HED; there is a bilateral diffuse haziness of the cornea, and there are multiple whitish/grey maculalike lesions throughout the corneal stroma. The periphery appears slightly less affected. Density and size of the lesions progresses throughout life leading to quite severe visual impairment.

DNA-tests for specific breeds are available.

Macular C dystrophy Courtesy: Dr. Claudia Busse



E1 Eigenaar; 10.03.2024



HED Manual Chapter 6 Guidelines

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)
- The box "severe" is to be ticked in the descriptive comments area.



Case 10



Great Dane, 6 years

Slide: Frans Stades



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Great Dane, 6 years



Slide: Frans Stades

14. Cataract: cortical	♦ 18. Other: Uveal Cysts; 18. severe
• Comm: synechia posterior	18. Other: Uveal cysts



Interpretation

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Uveal cyst:

ECV



KP-HED; 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 more or less pigmented spheres in the anterior or posterior chamber or occasionally in the vitreous.

When reaching maximal size, cysts tend to adhere to the endothelial surface in the center of the cornea, thus causing visual impairment.



HED Manual Chapter 6 Guidelines

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 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.



Case 11







Dobermann, 9 wk, m, bilateral,

slide: FS

 7. Other: Persistent Hyaloid artery; 7. severe; TVL post + Vasa Hyaloid prop. 	 2. PHTVL/PHPV Grade 2-6; Comm: lenticonus post + TVL post
 2. PHTVL/PHPV Grade 2-6; Comm: PHA 	7. Other: Persistent Hyaloid artery; 7. severe; Comm: TVLpost+Vasa Hyaloid prop.



"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. "Undetermined" : The animal displays clinical features that could possibly fit the KP-HED mentioned, but the changes are inconclusive. "Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions

Persistent hyaloid artery (PHA):

KP-HED; congenital defect resulting from abnormalities in the development and regression of the hyaloid artery. The blood vessel **remnant** can be present **in the vitreous as a small patent vascular strand (PHA) or as a non-vascular strand that appears grey-white** (persistent hyaloid remnant).



HED Manual Chapter 6 Guidelines

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 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.









Case 12



Brit. Shorthair, 2 y, OS before dilation Slide: FS



OS



Brit. Shorthair, 2 y, OS before dilation Slide: FS



 7. Other: iris coloboma, 13. Distichiasis/Ect. Cilia; 13. severe 	◆ 13. Distichiasis/Ect. Cilia
• 13. Distichiasis/Ect.cilia; comm: 2 Hairs	7. other: aniridy; 7. severe



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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.





Distichiasis:

KP-HED; 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. 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.



HED Manual Chapter 6. Guidelines:

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.



Shetland Sheepdog 9 wks, OS Slide: FS





Interpretation

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HED Manual Chapter 6. Guidelines

Distichiasis/ectopic cilia

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.



Case 13



Miniature Pinscher, 4 month, M

Slide: Frans Stades





Miniature Pinscher, 4 month, M, stretched fissure 19 mm Slide: Frans Stades





	7. Other: Microblepharon	 7. Other: Microphthalmos
•	11. Entropion/Trichiasis	7. Other: Microblepharon; 7. severe



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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions

Microblepharon:

KP-HED; an exceptionally short palpebral fissure. Microblepharon may lead to upper lid entropion and trichiasis.



HED Manual Chapter 6 Guidelines

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.



Case 14



Rottweiler, 1.5 Y, m, OD, unilateral, before mydriasis slide: FS



OD



Rottweiler, 1.5 y, m, OD, unilateral, before mydriasis slide: FS



 7. Iris hypoplasia; 7. severe; Comm: iris thinning 	 7. Other: iris hypoplasia
• 7. Other: aniridy- severe	Comment: Iris atrophy



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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED Manual Chapter 5 Definitions:

Hypoplasia iris:

KP-HED; characterized by congenital **thinning** and/or absence of iris (sphincter) tissue or colobomatous defects due to failure in closure of the optic fissure. It may be a separate disorder or associated with other ocular malformations. See and use iris hypoplasia.


HED Manual Chapter 6 Guidelines

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



Case 15



Persian cat, 7 months, OS, Slide: FS





Persian cat, 7 months, OS, Slide: FS



	5. Hypoplastic-/Micropapilla	 7. Optic disc coloboma
•	Comments: (Chorio-) retinopathy, inheritance under investigation	7. Optic nerve head hypoplasia



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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



Optic papilla/optic disc/optic nerve head: the part of the optic nerve which is visible, by ophthalmoscopic examination, in the fundus.

Optic nerve head hypoplasia: see and use hypoplastic papilla/optic nerve head/disc.



Hypoplasia-/ optic nerve head/disc hypoplasia:

KP-HED; congenital failure of development of the optic nerve which causes visual deficit or blindness and abnormal pupil response in the affected eye. It can often not be differentiated from micropapilla on a routine (dilated) ECVO eye examination



HED Manual Chapter 6. Guidelines

Other 7. Optic "disc"/Optic nerve head 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 fullthickness, 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



Case 16







Boomer, 2 years, OS

Slide: Frans Stades

13. Distichiasis/ Ectopic cila	 11: Entropion/Trichiasis. Comm: 11 Trichiasis nasal fold.
 11. Entrop./ Tri; 11.severe Comm: 11. Entrop. nasal + 11. Tri. nasal/nasal fold 	11: Entrop./Trichiasis. Comm: 11 Entrop. Nasal + other: Tri. caruncle



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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



Entropion:

KP-HED; a conformational defect resulting in "inrolling" of one or both of the margins of the eyelids which may cause ocular irritation. It is likely that entropion is influenced by several genes (polygenic), defining the skin and other structures which make up the eyelids, the amount and weight of the skin covering the head and face, the orbital contents and the conformation of the skull.



Trichiasis:

KP-HED or acquired abnormality of **deviated** hairs on a normal place around the lid fissure, irritating the conjunctiva, the free lid margin of the opposite lid and/or the conjunctiva and/or the globe. It is predominantly due to facial folds or due to misalignment of the eyelids.



HED Manual Chapter 6 Guidelines

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:

- 11. Trichiasis nasal/nasal fold
- 11. Trichiasis upper eyelid temporal

No further details are to be mentioned on the form 122



HED Manual Chapter 6 Guidelines

Entropion/trichiasis

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.



Case 17





Boykin Spaniel 9 wks, bilateral Slide: FS



	6. Collie Eye Anomaly - Choroid. Hypoplasia	 7. Other: Choroid. Hypoplasia - "go normal" 	
•	Comments: (Chorio-) retinopathy, inheritance under investigation	6. Collie Eye Anomaly - unaffected	



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"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



Choroidal (retinal) hypo- (dys-) plasia (CH, CRD):

KP-HED congenital eye disease which is characterized by inadequate development of the choroid present at birth which is non-progressive.

Most commonly identified in the Collie breed where it is a manifestation of "Collie Eye Anomaly" but can also be seen in other breeds.



HED SESSION

HED Manual Chapter 5 Definitions

Collie Eye Anomaly (CEA):

KP-HED; a congenital syndrome of ocular anomalies originally described in Collie breeds (short hair, long hair, Border Collie, Bearded Collie) and Shetland sheepdog 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 or CRD) with or without coloboma, retinal detachment and intraocular haemorrhage. Vision varies with the degree to which an individual is affected and may be minimally compromised to having severe visual impairment or blindness.



HED Manual Chapter 6. Guidelines

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.



HED Manual Chapter 6. Guidelines

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. 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".



HED Manual Chapter 6. Guidelines:

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".

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.



Case 18



Tervueren Shepherd, 1 y, OU

Slide: Frans Stades





Tervueren Shepherd, 1 y, OU

Slide: Frans Stades



18. Other: Keratitis, chronic superficial (CSK)/Pannus	11. Entropion/Trichiasis; 11. severe
11. Entropion/Trichiasis	 18. Other: Keratitis, chronic superficial (CSK)/Pannus; 18. severe



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.

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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



Entropion:

KP-HED; a conformational defect resulting in "in-rolling" of one or both of the margins of the eyelids which may cause ocular irritation. It is likely that entropion is influenced by several genes (polygenic), defining the skin and other structures which make up the eyelids, the amount and weight of the skin covering the head and face, the orbital contents and the conformation of the skull.



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.



Case 19



Vizsla, 2y, gonioscopy

Slide: Frans Stades



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Vizsla, 2 years, gonioscopy



Slide: Frans Stades

	8. ICAA: mild PLA: Unaff. ICA: narrow.	8. ICAA: unaff. PLA: unaff. + ICA: unaff.
•	8. ICAA: mild PLA: Mild, ICA: Unaff.	8. ICAA: moderate PLA: mild ICA: narrow



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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



Iridocorneal angle (ICA) width: the width of the ICA is evaluated (using gonioscopy) by comparison of the length of the pectinate ligament PL (A) and the distance from the origin of the PL to the anterior surface of the cornea at the transection area (B);


HED Manual Chapter 5 Definitions



Courtesy and modified by B. Ekesten from Ekesten B, & Narfström K.

Grading of ICA width:

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



HED SESSION

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

HED Manual Chapter 6 Guidelines







Case 20



Entlebucher Sennendog, 5 y, m, OS, Gonioscopy slide: FS



OS



Entlebucher Sennendog, 5 y, m, OS slide: FS

	8. ICAA: Severe: PLA: laminae >50%, Width: narrow	8. ICAA: Moderate: 25-50% LA, Width: narrow
•	8. ICAA: Severe- PLA: severe, Width: closed	8. ICAA: unaffected-PLA<50%



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

"Suspicious" : The animal displays minor, but specific clinical signs of the KP-HED mentioned. Further development will confirm the diagnosis.



HED SESSION

HED Manual Chapter 5 Definitions

I.C.A.A.:

- **Pectinate ligament** abnormality (PLA): KP-HED; characterized by an abnormal pectinate ligament that can be divided into 2 predominant types: 1. **Fibrae latae**, 2. **Lamina**; Diagnosis is by gonioscopy.

- Iridocorneal angle (ICA) width: the width of the ICA is evaluated (using gonioscopy) by comparison of the length of the pectinate ligament PL (A) and the distance from the origin of the PL to the anterior surface of the cornea at the transection area: opennarrow-closed

Both: part of the Irido Corneal Angle abnormality (ICAA)



Guidelines

Irido Corneal Angle abnormality (ICAA): KP-HED;

8. ICAA: unaffected-mild-moderate- severe Specifying in Comments:

8. ICAA" and the specifying box

- PLA:0-50% Fibrae Latae= "un"affected>50-100% FL and/or <25% LA</td>= affected mild25-50% Laminae (LA)= affected moderate>50% LA= affected severe
- Width:
 - Open
 - Narrow
 - Closed

- = normal
- = affected moderate
- = affected severe