HED SESSION 2021 - KAHOOT



Dr. Marianne Richter (Switzerland) Dr. Claus Bundgaard Nielsen (Denmark)

ECVO	new	ECVO	O Certificate	
ant.	Photogr		/temp. med./nas.	Photographs: ant. post. med./nas. lat./temp.
Descriptive comments:			15. Catara	ct other: punctata 8. ICAA: PLA mild
Eye disease no.			. 🖂 severe	Image: matrix ing nuclear ring nuclear fiberglass/pulverulent ICA narrow (moderate) (width) closed (severe)
Results for the known or presumed	*		*	Results valid for 12 months
1. Persistent Pupillary Membrane (PPM)			iris Cornea	11. Entropion / Trichiasis
 Persistent Hyperpl. Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 			grade 1 grade 2-6	12. Ectropion / Macroblepharon
3. Cataract (congenital)			(multi)focal	13. Distichiasis / Ectopic cilia
4. Retinal Dysplasia (RD)			geographical	14. Corneal dystrophy
5. Hypoplastic-/Micro-papilla			choroid, hypoplasia	15. Cataract (non-congenital)
6. Collie Eye Anomaly (CEA)			coloboma	16. Lens luxation (primary)
7. Other:				17. Retinal degeneration (PRA)
8. IridoCorneal Angle Abnormality. (ICAA)			moderate severe	18. Other:

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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



Case 1



Border Collie, m, 4y, bilateral findings; no clinically noticeable visual disorders Slides: M.Richter





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Border Collie, m, 4y, bilateral findings; no clinically noticeable visual disorders Slides: M.Richter



retinopathy (CMR)

hered. retinal degeneration

	ECVO CERT ew printe			Border Collie, m, 4 bilateral findings;	Y,	12	
Aight eye (OD)	Photographs:	/temp. med./nas.		ant.	Photograp		ed,/nas. lat,/temp.
Descriptive comments:			15. Catara	act other: punctata	8. ICA		— mild
				suture line		```	severe
				\ nuclear ring			marrow (moderate)
Eye disease no.		severe		nuclear fiberglass/pulver		(width)	closed (severe)
Results for the known or presumed	* **	*		Results valid for 12 month	*	***	*
		D AFFECTED	cornea		INAFFECTED S	SUSPICIOUS	AFFECTED
1. Persistent Pupillary Membrane (PPM)		lens	amina	11. Entropion / Trichiasis			
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1		12. Ectropion / Macroblepharon			
3. Cataract (congenital)		(multi)fo	cal	13. Distichiasis / Ectopic cilia			
4. Retinal Dysplasia (RD)		geograp		14. Corneal dystrophy			
5. Hypoplastic-/Micro-papilla			. hypop l asia	15. Cataract (non-congenital)			cortical post. pol.
6. Collie Eye Anomaly (CEA)			21 1	16. Lens luxation (primary)			nuclear other
7. Other:		│		17. Retinal degeneration (PRA)			
8. IridoCorneal Angle Abnormality. (ICAA))	moderal severe	te	18. Other:			

- * "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.
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FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

- Retinal dysplasia: KP-HED; abnormal development of the retina with ophthalmoscopic changes observed early in life, characterized by neuroretinal folding (s), rosettes and partial or total retinal detachment; non-progressive and generally recognized to have three forms: (multi)focal, geographic and total.
- 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.



HED Manual Chapter 6 Guidelines

- *["]* Retinal Dysplasia (RD):
- Tick at "4: Retinal dysplasia" and "(multi)focal" "affected"



HED Manual Chapter 8 Vet Advice

- (Multi)focal form in any breed: OPTIONAL. Note: different advice may be given for specific breeds by the breeding clubs



Case 2







Labrador Retriever, m, 1y, menace response OU

Slides: M.Richter



ECVO	ECVO CER ew printe	TIFICATE ed form		rador Retriever, m, menace response (-	
ant.	Photographs:	at./temp. med./nas.		ant.	Photogra	med./r	nas. lat,/temp.
Descriptive comments:			15, Cataract		8. ICAA: I		mild
Menace	response po	sitive OU		suture line tip			moderate severe narrow (moderate)
Eye disease no.		🖂 severe		nuclear fiberglass/pulver		(width)	closed (severe)
Results for the known or presume	d hereditary eye dise	ases (KP-HED):		Results valid for 12 month	าร	***	*
	UNAFFECTED UNDETERMIN			U	NAFFECTED SUS	PICIOUS A	AFFECTED
1. Persistent Pupillary Membrane (PPM)			cornea	11. Entropion / Trichiasis			
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1 grade 2-6		12. Ectropion / Macroblepharon			
3. Cataract (congenital)		(multi)foca	J	13. Distichiasis / Ectopic cilia			
4. Retinal Dysplasia (RD)		geographic total		14. Corneal dystrophy			
5. Hypoplastic-/Micro-papilla		choroid, h	vnenlesie	15. Cataract (non-congenital)			cortical
6. Collie Eye Anomaly (CEA)		coloboma	· · ·	16. Lens luxation (primary)			nuclear other
7. Other:		mi l d		17. Retinal degeneration (PRA)			
8. IndoCorneal Angle Abnormality. (ICAA	A)	severe	-	18. Other:			

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 ** 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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

- Hypoplasia-/ optic 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.
- Micropapilla: KP-HED; small optic disc which is not associated with vision impairment. It may not be differentiated from hypoplastic papilla/optic disc on a routine, dilated ECVO-eye examination.



HED Manual Chapter 6 Guidelines

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".



HED Manual Chapter 8 Vet Advice

Micropapilla/Optic Nerve Hypoplasia: NO BREEDING from the affected animal



Case 3



Slides: M.Richter





Slides: M.Richter



3. Cataract - congenital

ECVO	ECVO CER ew printe		Basenji-Mix; f bilateral findi		
Align eye (OD)	Photographs:	at/temp. med./nas.	ant.	Photographs:	med_/nas. lat./temp.
Descriptive comments:		15. Catar	act other: punctata	8. ICAA: PL	A mild
			suture line		severe
			nuclear ring		A narrow (moderate
Eye disease no.		severe	nuclear fiberglass/pulver	ulon (ridth)
Results for the known or presumed	* **	eases (KP-HED):	Results valid for 12 mont	NAFFECTED SUSPIC	
1. Persistent Pupillary Membrane (PPM)		iris Cornea	11 Entropion / Trichiasis		
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1 grade 2-6	12. Ectropion / Macroblepharon		
3. Cataract (congenital)			13. Distichiasis / Ectopic cilia		
4. Retinal Dysplasia (RD)		(multi)focal geographical	14. Corneal dystrophy		🖌
5. Hypoplastic-/Micro-papilla		choroid, hypoplasia	15. Cataract (non-congenital)		cortical post. po
6. Collie Eye Anomaly (CEA)		coloboma	16. Lens luxation (primary)		nuclear
7. Other:			17. Retinal degeneration (PRA)		

Interpretation

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FOR FUTHER INFORMATION: P.T.O.	Examiner

Explanation:

If the opacity on the lens is limited to the insertion of the PPM on the capsule, do NOT tick the box for cataract

Comment: The lens opacity is NOT limited to the attachment of the PPM; adjacent to the PPM there are whitish opacities (cataract) extending into the lens cortex which are likely congenital; there are also opacities in the posterior lens cortex which may be non-congenital.



Slides: M.Richter



HED Manual Chapter 5 Definitions

Persistent pupillary membrane (PPM): KP-HED; in which blood vessel remnants of the embryological vascular network in the anterior part of the lens fails 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 colarette, 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 Chapter 6 Guidelines

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 lens: boxes PPM, iris and lens are ticked; * New text

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



HED Manual Chapter 6 Guidelines

Section Results:

- ⁷⁷ The box for the KP-HED (1-6, 11-17) on the certificate and the specifying box, if available (e.g. for type or grade) are ticked.
- If there is no specific box available on the certificate for the KP-HED, the box at number "7. Other" and/or at number "18. Other" is to be ticked and the definition name of the disease (in the list in chapter 5) is written (online: is used). <u>Only if there are more than one KP-HEDs</u> <u>present which are not listed in the results field under no 1-6 and no 11-17, the box "affected" at "7. Other" and/or at "18. Other" is ticked and the term "Multiple other KP-HEDs" is written (online: is used); the KP-HEDs must also be specified in the comment field using the definition name in the list in chapter 5.</u>
 - e.g. "7. Other": Persistent hyaloid artery (PHA) + Iris hypoplasia



HED Manual Chapter 8 Vet Advice

- ["] Persistent Pupillary Membrane (PPM):
 - Strands iris to lens: NO BREEDING from the affected animal
- **Cataract (congenital):** NO BREEDING from the affected animal
- ["] Cataract (hereditary, non-congenital):
 - Cataract "cortical": NO BREEDING from the affected animal



Case 4



Cane Corso, m, 6m, OS (unilateral)

Slides: M. Richter





Cane Corso, m, 6m, OS (unilateral)



 3. Cataract (congenital) 7. Other: Persist. Hyaloid Artery 	 2. PHTVL/PHPV grade 2-6
• 2. PHTVL/PHPV grade 2-6	7. Other: Lenticonus
3. Cataract (congenital)	28

****	ECVO C ew prii			n	Cane Corso, m, 6m, OS (unilateral)		1	
ant.	Photographs	s:	np. me	ed./nas.	ant.	Phote post.	med.	/nas. lat./temp.
Descriptive comments:				15, Catara	ct other: punctata	8. ICAA:		mild moderate
					suture line		\backslash	severe
					nuclear ring			narrow (moderate)
Eye disease no.			sever	-	nuclear fiberglass/pulver		(width)	closed (severe)
Results for the known or presumed	*	**	*):	Results valid for 12 mont	*	***	*
	UNAFFECTED UNDE	TERMINED A		liris cornea		NAFFECTED SUS	SPICIOUS	AFFECTED
1. Persistent Pupillary Membrane (PPM)				lens amina	11. Entropion / Trichiasis			
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 				grade 1 grade 2-6	12. Ectropion / Macroblepharon			
3. Cataract (congenital)				(multi)focal	13. Distichiasis / Ectopic cilia			
4. Retinal Dysplasia (RD)			\square	geographica	14. Corneal dystrophy			
5. Hypoplastic-/Micro-papilla				total	15. Cataract (non-congenital)			cortical post. pol
6. Collie Eye Anomaly (CEA)				choroid, hypop l asia co l oboma	16. Lens luxation (primary)			nuclear other
7. Other:				other: mi l d	17. Retinal degeneration (PRA)			
8. IridoCorneal Angle Abnormality. (ICAA)				severe	18. Other:			

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

FOR FUTHER INFORMATION: P.T.O.	Examiner



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 6 Guidelines

Persistent hyperplastic tunica vasculosa lentis/persistent hyperplastic primary vitreus (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 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".



HED Manual Chapter 8 Vet Advice

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

-Grade 1: OPTIONAL -Grade 2-6: NO BREEDING from the affected animal



Case 5



Collie, puppy 8 weeks, bilateral finding

Slides: S.Borer





Collie, puppy 8 weeks, bilateral finding

Slides: S.Borer



4. Retinal dysplasia
(multi)focal

Comments: Chorioretinitis

• Comments: retinal folds

6. CEA – choroid. Hypoplasia +Comments: retinal folds³⁵

ECVO	ECVO CERT ew printe		Collie, puppy 8 weeks, bilateral	
Alight eye (OD)	Photographs:		ant.	post. med./nas. lat./temp.
Descriptive comments: OU: retinal folds			ract other: punctata a	B. ICAA: PLA mild
00.100			suture line	severe
			nuclear ring	ICA narrow (moderate)
Eye disease no.			nuclear fiberglass/pulverulent	(width) Closed (severe)
Results for the known or presumed		Results valid for 12 months	TED SUSPICIOUS AFFECTED	
1. Persistent Pupillary Membrane (PPM)				
 Persistent Hyperpl. Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1 grade 2-6	12. Ectropion / Macroblepharon	
3. Cataract (congenital)			13. Distichiasis / Ectopic cilia 🛛 🗆	
4. Retinal Dysplasia (RD)		(multi)focal	14. Corneal dystrophy	
5. Hypoplastic-/Micro-papilla		└── total └── choroid. hypop l asia	15. Cataract (non-congenital)	cortical post. pol.
6. Collie Eye Anomaly (CEA)		coloboma	16. Lens luxation (primary)	nuclear
7. Other:		mild	17. Retinal degeneration (PRA)	
8. IndoCorneal Angle Abnormality. (ICAA)			18. Other:	

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

FOR FUTHER INFORMATION: P.T.O.	Examiner


HED Manual Chapter 5 Definitions

- Retinal dysplasia: KP-HED; abnormal development of the retina with ophthalmoscopic changes observed early in life, characterized by neuroretinal folding (s), rosettes and partial or total retinal detachment; non-progressive and generally recognized to have three forms: (multi)focal, geographic and total.
- Retinal dysplasia- (multi)focal: seen ophthalmoscopically as linear (vermiform), triangular, curved or curvilinear foci of retinal folding that may be single or multiple. Its significance to vision is unknown. When seen in puppies this condition may partially or completely resolve with maturity. 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.



HED Manual Chapter 6 Guidelines

Retinal Dysplasia (RD):

- Linear (vermiform), triangular, curved or curvilinear foci of retinal folding that may be single or multiple seen ophthalmoscopically, the boxes at "4: Retinal dysplasia" and "(multi)focal" "affected" are ticked.
- In puppies, linear or round juvenile folds, usually in the peripapillary area, may be observed as a result in inequity in the relative growth rates of the optic cup and these folds resolve as the animal matures. These folds are not accurately referred to as dysplasia and should be ticked "unaffected" but have to 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".



HED Manual Chapter 8 Vet Advice

Retinal folds (in puppies or as sequelae post inflammation or retinal reattachment): no restriction



Case 6



Boerboel, f, 10 months, bilateral finding Slides: Birgit Lohmann





Boerboel, f, 10 months, bilateral finding Slides: Birgit Lohmann



Comments: iris naevus	18: Other: uveal cysts
• 7. Other: iris hypoplasia	 18. Other: iris melanoma - suspicious

ECVO		RTIFICATE ted form		Boerboel, f, 10 mo bilateral finding	nths,		
ant.	Photographs:	lat./temp. med./nas.		ant.	Photographs.	med./na	as. lat./temp.
Descriptive comments:			15. Catarac	ct other: punctata	8. ICAA: I		mild moderate
				suture line			severe
				\ nuclear ring			narrow (moderate)
Eye disease no.		severe		nuclear fiberglass/pulver	rulent	(width)	closed (severe)
Results for the known or presumed	* **	*		Results valid for 12 mont	* *		*
			cornea			PICIOUS AF	FECTED
1. Persistent Pupillary Membrane (PPM)		lens grade 1	amina	11. Entropion / Trichiasis			
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 2-6		12. Ectropion / Macroblepharon			
3. Cataract (congenital)			al	13. Distichiasis / Ectopic cilia			
4. Retinal Dysplasia (RD)		(multi)foca geographi		14. Corneal dystrophy			
5. Hypoplastic-/Micro-papilla			menlesia	15. Cataract (non-congenital)			cortical post. pol
6. Collie Eye Anomaly (CEA)		coloboma		16. Lens luxation (primary)			nuclear other
7. Other:				17. Retinal degeneration (PRA)			
8. IndoCorneal Angle Abnormality. (ICAA)		severe	-	lriš ^{18. Other:} melanoma			

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FOR FUTHER INFORMATION: P.T.O.	Examiner



New text

HED Manual Chapter 5 Definitions

Iris melanoma: KP-HED; a neoplasm caused by malignant transformation of melanocytes in the iris. Occurs with a higher than normal incidence in the Labrador Retriever. Left untreated it may result in secondary glaucoma and/or metastasis. Without a histological examination, an iris melanoma cannot be differentiated from a benign melanocytoma in every case. A distinction may become possible depending on the further development.



HED Manual Chapter 6 Guidelines

New text

Ílris melanoma

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



HED Manual Chapter 8 Vet Advice

Iris melanoma: NO BREEDING from the affected animal



Case 7



Boston Terrier; m, 5y, bilateral findings

Slides: M.Richter







Boston Terrier; m, 5y, bilateral findings

Slides: M.Richter





Comments: corneal edema	Comments: lipid keratopathy
• 14. Corneal dystrophy, severe	 18. Other: chronic. superf. Keratitis (CSK) 49



Interpretation

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

FOR FUTHER INFORMATION: P.T.O.	Examiner



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. The onset in one eye may precede the other.
- 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 Manual Chapter 6 Guidelines

- Corneal dystrophy is to be ticked "affected" at "14. Corneal dystrophy", and the details described in the field Descriptive comments.
 - In cases of <u>endothelial dystrophy</u> (bilateral progressive diffuse, deep corneal edema, e.g. in Chihuahua, Boston Terrier etc.) or <u>macular dystrophy</u> (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 <u>severe forms of stromal dystrophy</u> (e.g. in Siberian Husky) is recognized, also the box <u>"severe"</u> is to be ticked in the comment area.



HED Manual Chapter 8 Vet Advice

Corneal Dystrophy:

Endothelial dystrophy (e.g. Chihuahua, Boston Terrier, Dachshund): NO BREEDING from the affected animal



Case 8



Labrador Retriever; m, 8y6m, bilateral findings Slides: M.Richter





ECVO

Labrador Retriever; m, 8y6m, bilateral findings Slides: M.Richter



***	ew print	RTIFICATE ted form		brador Retriever; . 8y6m, OU			
ant.	Photographs:	lat/temp. med./nas.		ant.	post	. me	d./nas. lat./temp.
Descriptive comments:			15. Catarac	t other: punctata	8. ICA		— mild — moderate
				suture line			severe
Eye disease no. 18		🔀 severe		nuclear ring	erulent	ICA (width)	narrow (moderate closed (severe)
Results for the known or presumed	hereditary eye di	•		Results valid for 12 mon	ths		
	UNAFFECTED UNDETERI	* MINED AFFECTED			* UNAFFECTED	*** SUSPICIOUS	AFFECTED
1. Persistent Pupillary Membrane (PPM)		iris lens	cornea	11. Entropion / Trichiasis			
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1		12. Ectropion / Macroblepharor	ו 🗖		
3. Cataract (congenital)		(multi)fa	ocal	13. Distichiasis / Ectopic cilia			
4. Retinal Dysplasia (RD)				14. Corneal dystrophy			
5. Hypoplastic-/Micro-papilla			, hypop l asia	15. Cataract (non-congenital)			cortical post. po
6. Collie Eye Anomaly (CEA)				16. Lens luxation (primary)			nuclear other
7. Other:				17. Retinal degeneration (PRA	.) 🗖		
8. IridoCorneal Angle Abnormality. (ICAA)			ite	18. Other: Uveal cysts			

- * "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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

["] Uveal cyst: 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 chamber. When reaching maximal size, cysts tend to adhere to the endothelial surface in the center of the cornea, thus causing visual impairment. Severe cases which occur with a higher than normal incidence in the Great Dane (Deutsche Dogge) and in the Golden Retriever and may lead to secondary glaucoma.



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 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.



HED Manual Chapter 8 Vet Advice

Uveal Cysts: OPTIONAL, Note: In severe cases the advice may be: NO BREEDING from the affected animal



Case 9



Before dilating the pupils (bilateral finding)







Australian Shepherd (red merle); f, 3y, bilateral findings

Before dilating the pupils (bilateral finding)





 6. CEA – choroid. Hypoplasia 7. Other: Iris hypoplasia, severe 	7. Other: lens hypoplasia
 6. CEA – choroid. Hypoplasia 7. Other: microphakia 	7. Other: Iris hypoplasia, severe 63

		RTIFICATE ited form	(red m	lian Shepherd erle); f, 3y, OU, dilating pupils	1	::
ant) () post.	lat./temp. med./nat	s, 2		post	med./nas. lat./temp.
Descriptive comments:			15, Cataract		8. ICAA: PLA	mild moderate
				1/ 411 / 412 3	1	severe
Eye disease no.		severe		nuclear ring nuclear fiberglass/pulver	ICA - ulent (width) narrow (moderate)
Results for the known or presumed	I hereditary eye			Results valid for 12 month	าร	
	WNAFFECTED UNDETE	* * RMINED AFFECTED		U	* *** NAFFECTED SUSPICIOU	s AFFECTED
1. Persistent Pupillary Membrane (PPM)			cornea	11. Entropion / Trichiasis		
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade	I	12. Ectropion / Macroblepharon		
3. Cataract (congenital)			i)foca	13. Distichiasis / Ectopic cilia		
4. Retinal Dysplasia (RD)			,	14. Corneal dystrophy		
5. Hypoplastic-/Micro-papilla			oid, hypop l asia	15. Cataract (non-congenital)		cortical post. pol.
6. Collie Eye Anomaly (CEA)			ooma	16. Lens luxation (primary)		nuclear other
7. Other: Iris hypoplasia				17. Retinal degeneration (PRA)		
8. IndoCorneal Angle Abnormality. (ICAA)		re	18. Other:		

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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

- Iris hypoplasia: 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.
- Lens hypoplasia: KP-HED; characterized by congenital incomplete formation of the lens equator, previously called lens coloboma.



HED Manual Chapter 6 Guidelines

Iris hypoplasia: At "7. Other": "Iris hypoplasia" is written (online: is used), and the box "affected" is ticked. Only if uni- or bilateral iris tissue is missing (full thickness) or failed to develop (developmentally colobomatous) e.g. in one of the specific breeds Australian Shepherd, Dalmatian, Rottweiler, also the box "severe" is to be ticked in the comment area.

Note: if there is a congenital lack of tissue in the iris or lens, the term "hypoplasia" is used: iris hypoplasia, lens hypoplasia. Reason: iris tissue can be absent full-thickness or partially (hypoplastic); the lens may have a flattened curvature due to abnormal development of zonular fibers or ciliary processes.



HED Manual Chapter 8 Vet Advice

Hypoplasia:

• Iris: OPTIONAL, Note: In severe cases: NO BREEDING from the affected animal



Case 10



Collie (blue merle); f, 2y, bilateral findings Slides: R.Eördögh





Collie (blue merle); f, 2y, bilateral findings Slides: R.Eördögh



6: CEA – choroid. hypoplasia	 6: CEA – choroid. hypoplasia & coloboma 		
• 18: Other:	6: CEA – coloboma		
optic disc coloboma	70		

	ECVO CERT ew printe			lie (blue merle); f, bilateral findings	E		
ant.	Photographs:	at/temp. med./nas.		ant.	Pho post.	med	d./nas. lat./temp.
Descriptive comments:			15. Catarac	t other: punctata	8. ICAA:		— mild — moderate
				suture line			severe
Eye disease no.		🖂 severe		nuclear ring	ulent	ICA (width)	narrow (moderate) closed (severe)
Results for the known or presumed	hereditary eye dise	ases (KP-HED):		Results valid for 12 month	ıs "	***	*
	UNAFFECTED UNDETERMIN			UN	AFFECTED SU	SPICIOUS	AFFECTED
1. Persistent Pupillary Membrane (PPM)			cornea	11. Entropion / Trichiasis			
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1	6	12. Ectropion / Macroblepharon			
3. Cataract (congenital)		(multi)foc		13. Distichiasis / Ectopic cilia			
4. Retinal Dysplasia (RD)		geograph		14. Corneal dystrophy			
5. Hypoplastic-/Micro-papilla			hypop l asia	15. Cataract (non-congenital)			cortical post. pol.
6. Collie Eye Anomaly (CEA)				16. Lens luxation (primary)			nuclear other
7. Other:		mi l d		17. Retinal degeneration (PRA)			
8. I ridoCorneal Angle Abnormality. (IGAA)		moderate severe	9	18. Other:			

- * "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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

- 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"
- Collie Eye Anomaly (CEA): KP-HED; 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 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. <u>DNA-tests</u> for choroidal hypoplasia in specific breeds are available.


HED Manual Chapter 5 Definitions

 Coloboma: congenital defect of a portion of the eye due to a failure in closure of the body halves; most frequently affecting the iris* or the optic nerve at the 6 o'clock position. The latter is a presumed hereditary congenital eye disease that if large, may cause retinal detachment resulting in blindness or visual impairment.

*use the term "iris-hypoplasia"



HED Manual Chapter 6 Guidelines

Collie eye anomaly (CEA):

At no 6. Colly Eye Anomalie (CEA) tick boxes "affected" and "choriod.
 hypoplasia" + "coloboma"

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 – other" has to be specified in the comment area of the ECVO certificate (retinal detachment or –haemorrhage)



HED Manual Chapter 8 Vet Advice

- **Collie Eye Anomaly (CEA):**
 - Choroidal hypoplasia (CH)/chorioretinal dysplasia (CRD): OPTIONAL
 - Coloboma and other defects (retinal detachment, haemorrhage): NO BREEDING from the affected animal



Case 11



Bouvier des Flandres; f, 3y, bilateral findings Slides: R.Eördögh





Bouvier des Flandres; f, 3y, bilateral findings Slides: R.Eördögh

Comments: retinal edema	 17. Retinal degeneration (PRA) - suspicious
• 18. Other: other presumed hereditary retinal degenerations	Normal/"unaffacted" 78

ECVO	ECVO CERT ew printe		Bouvier des Flan f, 3y, OU	dres;	H
ant.	Photographs:	L/temp. med./nas.	ant	Photogra	med_/nas, lat,/temp.
Descriptive comments:			15. Cataract other: punctata	8. ICAA: PL	
ERG	suture line tip		moderate severe		
Eve disease no.		🖂 severe	nuclear ring nuclear fiberglass/		A narrow (moderate) idth) closed (severe)
Results for the known or presumed			Results valid for 12 m	-	
	* ** UNAFFECTED UNDETERMINE	*		WNAFFECTED SUSPIC	
1. Persistent Pupillary Membrane (PPM)		iris	□ ^{cornea} 11. Entropion / Trichiasis		
 Persistent Hyperpl.Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 		grade 1 grade 2-6	12. Ectropion / Macrobleph	aron 🗖 🗖	
3. Cataract (congenital)		(multi)focal	13. Distichiasis / Ectopic ci	lia 🗖 🗖	
4. Retinal Dysplasia (RD)					
5. Hypoplastic-/Micro-papilla		total	15. Cataract (non-congenit	al) 🗖 🗖	cortical
6. Collie Eye Anomaly (CEA)			16. Lens luxation (primary)		nuclear other
7. Other:		other:	17. Retinal degeneration (F	PRA) 🗖 🚺	
8. IndoCorneal Angle Abnormality. (ICAA)	severe	- 18. Other:	- i - i - i - i - i - i - i - i - i - i	
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 https://www.nonths.nonths.nonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

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 Scheme eye examination) before there are detectable fundus changes observed by ophthalmoscopy. These funduscopic changes consist in the early disease of 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, 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.

Retinal degeneration can also be due to non-hereditary causes, e.g. inflammation and/or infection, toxicity, etc., affecting retinal structures with degeneration of cells or entire cellular layers. The end-stage is often complete retinal atrophy, which may appear ophthalmoscopically similar to (hereditary) PRA.



HED Manual Chapter 6 Guidelines & Chapter 3 Eye Scheme

- Retinal Degeneration/Progressive Retinal Atrophy (PRA): At number
 17. Retinal degeneration (PRA) tick box "suspicious"
- If an animal displays minor, but specific clinical signs of the KP-HED mentioned, "suspicious" is ticked for the relevant disease (no 11-18 on the certificate). Further development will confirm the diagnosis. It is required that "suspicious" cases are re-examined after the period prescribed on the Certificate, by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision is final. (see Chapter 3 Eye Scheme)
- Electroretinography (ERG) using the standardized protocol of the ECVO (Ekesten B et al. "Guidelines for clinical electroretinography in the dog: 2012 update", Doc Ophthalmol, 2013 Oct;127(2):79-87)
- **DNA-tests** for specific forms and breeds are available.



HED Manual Chapter 8 Vet Advice

Retinal Degeneration/Progressive Retinal Atrophy (PRA): NO BREEDING from the affected animal, its parents or offspring.

In instances where a <u>DNA-based genetic test</u> for recessive PRA is available breeders may choose to breed from carrier animals that have outstanding characteristics while still avoiding production of affected offspring. All such matings should be carefully controlled and all offspring subjected to DNA-based testing.



Case 12



Slides: M.Richter







Slides: M.Richter



• Comments: lipid keratopathy

■ 14. Corneal dystrophy



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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



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. The onset in one eye may precede the other.
- Corneal dystrophy, epithelial/stromal: 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 6 Guidelines

Corneal dystrophy is to be ticked "affected" at "14. Corneal dystrophy", and the details described in the field Descriptive comments.

In case of endothelial dystrophy (bilateral progressive diffuse, deep corneal edema, e.g. in Chihuahua, Boston Terrier etc.) or macular dystrophy (bilateral diffuse haziness of the cornea with multiple whitish/grey macula like lesions throughout the corneal stroma, periphery slightly less affected, e.g. in Labrador Retriever) or severe forms of stromal dystrophy (e.g. in Siberian Husky) is recognized, also the box "severe" is to be ticked in the comment area.



HED Manual Chapter 8 Vet Advice

- **Corneal Dystrophy:**
 - Epithelial and/or stromal: OPTIONAL; Note: In severe cases that cause visual problems and/or pain for the dog, e.g. in Siberian Husky or Shetland Sheepdog: NO BREEDING from the affected animal.



Case 13









 18. Other: uveodermatologic	 7. Other: Choroidal hypoplasia
syndrome	in Non-Collie breeds
• 7. Other: choroidal coloboma	7. Other: choroidal coloboma
17. Retinal degeneration (PRA)	92



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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

- Coloboma: congenital defect of a portion of the eye due to a failure in closure of the body halves; most frequently affecting the iris* or the optic nerve at the 6 o'clock position. The latter is a presumed hereditary congenital eye disease that if large, may cause retinal detachment resulting in blindness or visual impairment.
 - * use the term: "iris hypoplasia"



HED Manual Chapter 6 Guidelines

Coloboma:

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

Note: for congenital absence of tissue of the eyelid, retina, choroidea, sclera or optic nerve/papilla use the term "coloboma", e.g. choroidal coloboma



HED Manual Chapter 8 Vet Advice

Coloboma:

- Eyelid: NO BREEDING from the affected animal
- Papilla: NO BREEDING from the affected animal
- Retina: NO BREEDING from the affected animal
- Choroidea: NO BREEDING from the affected animal
- Sclera: NO BREEDING from the affected animal



Case 14



Australian shepherd, 2y, bilateral findings Slides: C. Bundgaard





OD

Australian shepherd, bilateral findings Slides: C. Bundgaard



Comments: ectopic pupil	 7. Other: uveal cysts
• 7. Other: iris coloboma	7. Other: iris hypoplasia, severe 99

	ECVO C ew pri	nteo	FICATE d form		ilian Shepherd efore dilating		· ···
ant) (post.	Iat./te	emp. med./na) s.	ant.	post.	med_/nas, lat,/temp.
Descriptive comments:				15, Catarao	ct other: punctata	8. ICAA: P	PLA mild
					suture line		severe
Eye disease no.			severe		nuclear ring		CA narrow (moderate width) closed (severe)
Results for the known or presumed	l hereditary ey	e disease			Results valid for 12 month		
	* UNAFFECTED UND	** ETERMINED	* AFFECTED		U	NAFFECTED SUSP	** * MCIOUS AFFECTED
1. Persistent Pupillary Membrane (PPM)				cornea	11. Entropion / Trichiasis		
 Persistent Hyperpl. Tunica Vasculosa Lentis/Primary Vitreous (PHTVL/PHPV) 			grade		12. Ectropion / Macroblepharon		
3. Cataract (congenital)			(mult	i)foca	13. Distichiasis / Ectopic cilia		
4. Retinal Dysplasia (RD)				raphica	14. Corneal dystrophy		
5. Hypoplastic-/Micro-papilla				oid, hypop l asia	15. Cataract (non-congenital)		cortical
6. Collie Eye Anomaly (CEA)				ooma	16. Lens luxation (primary)		nuclear
7. Other: Iris hypoplasia					17. Retinal degeneration (PRA)		
			mild		[17. Retinal degeneration (FRA)		

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 inmonths.

FOR FUTHER INFORMATION: P.T.O.	Examiner



HED Manual Chapter 5 Definitions

Iris hypoplasia: KP-HED; characterized by congenital 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.



HED Manual Chapter 6 Guidelines

Iris hypoplasia: At "7. Other": "Iris hypoplasia" is written (online: is used), and the box "affected" is ticked.

Only if uni- or bilateral iris tissue is missing (full thickness) or failed to develop (developmentally colobomatous) e.g. in one of the specific breeds Australian Shepherd, Dalmatian, Rottweiler, also the box "severe" is to be ticked in the comment area.

Note: if there is a congenital lack of tissue in the iris or lens, the term "hypoplasia" is used: iris hypoplasia, lens hypoplasia. Reason: iris tissue can be absent full-thickness or partially (hypoplastic); the lens may have a flattened curvature due to abnormal development of zonular fibers or ciliary processes.



HED Manual Chapter 8 Vet Advice

Hypoplasia:

• Iris: OPTIONAL, Note: In severe cases: NO BREEDING from the affected animal