

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

1. The Scheme

The ECVO has a Hereditary Eye Disease Scheme (further referred to as the Scheme) as mentioned in the Bylaws of the ECVO, Article 4.9, used for the diagnosis and the control of Known and Presumed Hereditary Eye Diseases (KP-HED) in animals.

The main purposes of the Scheme are to set standards for the diagnosis and provide advice for the control of KP-HED of dogs and cats. The diseases included are either disabling, painful or perturbing to the wellbeing of animals, or diseases that necessitate surgical, or otherwise physiologically altered intervention or lifelong medication.

The Scheme provides definitions, guidelines, advice and information concerning KP-HED, as described in the ECVO Manual for KP-HED of dogs and cats. Additional information is given in the appendices of the scheme, and through illustrations. Current versions of these documents are published on the ECVO website www.ecvo.org.

The Scheme guarantees that:

- all examiners are able to recognize and to differentiate relevant and irrelevant clinical signs of KP-HED and establish their significance in dogs and cats;
- rules and guidelines are provided for the examination and validation of specifically trained European Specialists in Veterinary Ophthalmology (Diplomates) and specifically trained veterinarians (European Eye Scheme Examiners or ESE) to provide a service for owners, breeders, the breed and kennel clubs and the public in general, by providing an appropriate level of expertise for the diagnosis of KP-HED.

Examinations under the Scheme are performed in accordance with the ECVO Constitution and Bylaws and include a general screening examination of the eye and its adnexa. Based on the examination, a certificate is issued. The certificate is valid for 12 months. It is recommended that animals used for breeding are examined annually. Animals not used for breeding may be examined at longer intervals, in accordance with the ECVO Manual. If the examination findings are inconclusive, it may be necessary for the animal to be re-examined earlier than recommended in this paragraph. This shall be stated on the certificate.

2. Organization and panels

The ECVO Diplomates and the national ESEs (when recognized) shall form a National Panel in countries where this has been agreed upon, and the necessary rules and regulations set. These rules and regulations shall not contain any provision that violates the Constitution or Bylaws of the ECVO or the rules of the Scheme.

Each National Panel shall have a Board, elected by its members. The Board shall consider all business and policies pertaining to the affairs of the National Panel. In the absence of a National Panel, the HED-Committee of the ECVO can act in its place.

For countries where an established National Panel has been in existence for more than 5 years, on the advice of the HED-Committee, this Panel can apply to the Executive Committee of the ECVO for recognition under the ECVO Scheme.

The National Panel must meet at least once per year to discuss the Scheme and its operation.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

3. Panellists

Panellists approved by the ECVO to perform examinations under the Scheme are:

- Diplomates of the ECVO
- Eye Scheme Examiners (ESEs), being veterinarians, specifically trained and examined in countries where this has been agreed. This agreement may be extended ad infinitum by the ECVO.
- Affiliate members as defined in the ECVO Constitution & Bylaws.

Training and examination of Eye Scheme Examiners (ESEs)

Before training commences

- The candidate must confirm normal stereoscopic vision and colour vision (binocular, with a minimum visual acuity of 0.7 corrected for refractive errors).
- An education and training plan based on the requirements of the ECVO Eye Scheme must be organised by the candidate.
- It is recommended to have a main supervisor, who could be an ECVO Diplomate or an ESE from an ECVO approved national Panel.
- The candidate should have access to the basic ophthalmic instruments; direct and indirect ophthalmoscopes, a slit-lamp biomicroscope and a gonioscopy lens.

Practical training

- The candidate must document the ophthalmic examination performed according to the ECVO rules (See ECVO Manual) of at least 500 dogs under supervision.
- For these examinations, the supervisor is a recognised panel member and must be present.
- Panellists from non-ESE national panels and acknowledged by the ECVO HED Committee, may in exceptional circumstances be approved to serve as supervisors.
- At least 50 of the dogs shall be examined under direct supervision of an ECVO Diplomate. Up to 200 dogs can be examined under direct supervision of an ACVO Diplomate.
- The candidate must document the examination of certain specific breeds and diseases, the minimum numbers for each as defined in Appendix A.
- The candidate must document the examination of at least 100 cats. At least 10 of these cats shall be examined under direct supervision of an ECVO Diplomate or ESE.
- The trainee must perform and record gonioscopy on at least 20 dogs under supervision.
- A record shall be kept for all animals examined. The record should include breed, sex and age of the animal examined, ophthalmic findings and diagnosis/-es and should be signed by the supervisor(s).

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

Theoretical training

Before sitting the examination, the candidate should acquire a sufficiently high level of current knowledge of

- Ocular embryology and anatomy
- Ocular physiology
- Basic examination techniques
- Breed-related diseases and genetic testing
- Neuro-ophthalmology

For this, the candidate

- Must document participation in at least 3 ECVO-recognized continuing education courses in ophthalmology (each of a duration of minimum 1,5-2 days), which include the use of ophthalmic equipment, diseases of the anterior and posterior segments, and basic genetic principles
- Should attend the annual ECVO congress, especially the HED session
- Must have studied the literature concerning known and presumed hereditary eye diseases (KP-HED) in animals, including relevant published scientific articles and the ECVO Manual for KP-HED in Dogs and Cats

Details about number of KP-HEDs to be seen and a list of relevant literature are indicated in Appendix A and B respectively.

The requirements defined by ECVO will be considered minimum requirements. National or regional requirements for training and examination may exceed the demands of the ECVO.

Examination

An examination is necessary to qualify as an ECVO-Eye Scheme Examiner (ESE). The full training programme should be fulfilled before the candidate is qualified to sit the ESE examination for the scheme. Candidates may apply to an exam planned by any active ECVO Panel. The documentation will be reviewed by the ECVO/HED committee according to an agreement with the involved Panel.

The examination committee for the examination to qualify as Eye Scheme Examiners consists of at least three members of ECVO Panels, of which at least two should be ECVO Diplomates.

The examination will consist of

- A written section of 3 hours' duration on multiple choice (50 minutes; 3 min/question) and/or essay questions on known and presumed hereditary eye diseases and relevant ophthalmology topics
- A combined practical and written section of 45 minutes' duration based on 45 images of known and presumed hereditary eye diseases as well as normal ocular variations
- A final section which will consist of live case evaluation (at least 5 cases) with the issue of a correct ECVO Certificate for each case. The candidate is allowed 15 minutes for examination of each animal, plus 5 minutes to fill in the certificate. The certificate should be completed in detail, including the "Conclusions" section.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

This final section can only be taken once the candidate has passed the first two sections of the examination.

The different parts of the education programme (courses, practical training, etc.) are valid for ten calendar years.

Candidates may sit each part of the examination (MCQ, slide recognition and practical/ oral) up to a maximum of four times.

A reasonable transition period will apply to changes made to the requirements. Major changes (i.e. significant increase in numbers of animals to be examined/cases to be seen, additional courses etc), apply only to aspirants who have started after the change has been made.

Appeals procedure

Appeals should be directed in writing to the Board of ECVO that will handle it according to normal appeals procedures.

Requirements for maintaining Panellist status

The Board of the National Panel is responsible for the compliance of their Panellists with the following rules. If no National Panel exists, the HED-Committee of the ECVO can act in its place. The Panellists, including both Diplomates and ESE are obliged to work under the rules of both the ECVO Scheme and the National Scheme.

- If work is conducted by an examiner in serious violation of these rules, the Board of the National Panel or, if no national panel exists, the HED-committee is entitled to expel the person from the panel.
- If there is proof of continuing misdiagnoses by a Panellist, this individual shall be re-examined, or the Board of the National Panel is entitled to expel the examiner from the Panel. If expelled, the former Panellist is neither allowed to issue ECVO certificates, nor issue documents that imply the same status as the ECVO certificate.
- The Panellist shall complete the minimum number of examinations for the Scheme of 100 per year or 300 per 3 years; if less, re-qualification is necessary. Recently qualified Panellists are exempted from this requirement during their first full year as Panellist. Panellists on maternal leave or a documented shorter period of work debilitating disease can apply the Board of the National Panel for exemption.
- If The Panellist wants to perform gonioscopy as part of the examination under the ECVO Scheme, the Panellist shall complete a minimum number of gonioscopy examinations for the Scheme of 10 per year or 30 per 3 years (which may be part of the general examinations for the Scheme); if less, gonioscopy should not be performed. Recently qualified Panellists are exempted from this requirement during their first full year as Panellist. Panellists on maternal leave or a documented shorter period of work debilitating disease can apply the Board of the National Panel for exemption.
- The Panellist shall attend the annual meeting of the national panel. If absent from 3 consecutive meetings, without dispensation of the Board of the National Panel, the Panellist can be expelled from the national panel. If the panel holds other related meetings (e.g. arbitrary or appeal cases) the Panellist shall attend these meetings. If absent of more than half of these meetings over 3 consecutive years, without written dispensation of the Board of the National Panel, the individual can be expelled by the Board of the national Panel.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

- The Panellist shall attend at the least one annual scientific meeting during 3 years. If absent without dispensation of the Board of the National Panel (or the HED-Committee), the individual can be expelled by the Board of the national Panel ((or the HED-Committee of the ECVO)
- The Panellist shall contribute to the education of new panel members through practical training and instruction of aspirants.
- The Panellist must possess good eyesight with a normal stereoscopic, colour vision (binocular, with a minimum visual acuity of 0,7 corrected for refractive errors). A certificate of visual acuity is to be presented to the Board of the National Panel or the HED-Committee of the ECVO every 5 years until the age of 70, and after that age, every second year.
- Under exceptional circumstances the Board of the National Panel, or if there is no Panel, the HED Committee of the ECVO may grant a Panellist exemption from the minimum requirements for number of eye examinations conducted, and meeting attendance.
- A decision to expel a Panellist from the Panel is taken by the Board of the National Panel. The Panellist has the right to appeal to the National Panel at the next annual meeting or alternatively to the HED Committee of the ECVO.
- A Panellist who is expelled from the panel may be allowed to sit a new examination in order to requalify. The decision is taken by the Board of the National Panel (or the HED-Committee of the ECVO) which will also decide on the extent of the examination.

4. Arrangements for the Eye Examination

Individual animals can be presented for ophthalmic examination, or group examinations can be arranged. Minimum facilities for performing an ophthalmic examination include darkened surroundings, an examination table and suitable electrical power supply.

5. Procedure for the Eye Examination

Examination for the Scheme is performed according to the current ECVO rules (See ECVO Manual). An ECVO certificate is issued upon completion of the examination.

- Partial or preliminary examinations are not permitted.
- Gonioscopy may be performed as an additional examination.
- An ECVO certificate shall be issued for each animal. Breed specific forms issued by breed clubs may be filled and signed by an ECVO examiner in addition. The decision to charge for any extra costs incurred is at the discretion of the Panellist.
- The following documents should be available before the examination:
 - the animal's registration document or other identifying document
 - any previous eye certification

The owner and/or his agent will present the animal for examination at the appropriate time together with the documents referred to above. If the animal's registration or other identifying documents are not available, examination can be undertaken, but the Certificate will not be issued until the Panellist has been provided with the relevant documentation. Prior to the examination, the owner or their appointed agent, must sign an agreement to the examination procedure, recording of results, submission of results to the appropriate National Kennel Club and that the results will be made available in the public domain, as outlined in the Scheme. This will complete the first part of the Certificate, verifying that the

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

details given in that section are correct. Details included in this section relate to the identification of the animal being submitted for examination and the date of the last eye examination. Microchip number or tattoo should be verified under direct control of or by the examiner, or his/her designated assistant, prior to the ophthalmic examination.

Certificates for the Scheme can only be issued for permanently identified animals (e.g. by microchip or tattoo). Exceptions for mixed-breed animals that are permanently identified and animals outside the scheme are given in § 6.

Subject to the registration documents being available as stated above, the Certificate should be signed and issued by the Panellist and distributed according to the instructions.

If online-registration of results is available, this process should be completed immediately or at the latest within three days of the examination.

If paper copies are issued, this distribution will involve:

- 1) Top copy (white) will be sent as soon as possible (within 14 days of the date of examination) to the appropriate Kennel club or National Scheme authority, which will undertake to make the result publically available within a maximum of 10 weeks after the date of examination,
- 2) One copy (yellow) can be sent by the owner to the appropriate Breed club, or is sent by the Panellist to the Breed club within a maximum of 10 weeks after the date of the examination,
- 3) One copy (pink) will be retained by the Panellist as their record,
- 4) One copy (white) will be given to the owner or agent,
- 5) One (optional) copy (blue) may be distributed to the referring veterinarian by the Panellist or owner.

All animals presented under the Scheme will have a general screening examination of the eye and adnexa in darkened surroundings. This will include the necessary use of a short-acting mydriatic.

For an ophthalmic screening examination in accordance with the ECVO Scheme, evaluation of the **entire eye** is recommended. This examination includes the adnexa, and the anterior and posterior segments. Visual function should also be noted if abnormal.

It is recommended initially to **examine the visual processes** by visual behaviour, followed by examination of the ophthalmic reflexes and reactions, e.g. menace reaction, dazzle-, palpebral- and pupillary light reflexes (PLRs). **Biomicroscopy** of the adnexa and anterior segment is then performed. Evaluation of the iridocorneal angle is done by **gonioscopy** (see gonioscopy below). These procedures are followed by dilation of the pupils using short acting mydriatics. Ophthalmoscopy and biomicroscopy (slit lamp examination) are then performed with focus on the posterior and anterior segments of the eye.

The minimum equipment to be used for the examination is a slit-lamp biomicroscope (at least 10 x magnifications) and a binocular indirect ophthalmoscope with appropriate lenses. The use of other equipment is optional.

If **electroretinography** is used as an early diagnostic test for hereditary retinal degeneration, the following standardized protocol of the ECVO has to be followed: Ekestén B, Komáromy AM, Ofri R, Petersen-Jones SM, Narfström K. Guidelines for clinical electroretinography in the dog: 2012 update. Doc Ophthalmol DOI 10.1007/s10633-013-9388-8, published online: 01 June, 2013.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

The Panellist should evaluate all conditions specified on the certificate, and all boxes referring to diagnoses should be ticked. The appropriate boxes should be left open if gonioscopy has not been performed or if diagnoses from the drop-down menus, boxes No 7 and 18, "Other" are not relevant. Details of all lesions and conditions found at the time of examination, whether relating to hereditary eye disease or not, should be concisely and legibly recorded, in English or in the national language in the descriptive comments section in the middle of the Certificate, using drawings and/or written remarks. Photographic documentation of conditions should be done whenever possible.

Gonioscopy

Gonioscopy is not included in the basic ophthalmoscopic examination but may be performed as an additional examination, and performed prior to pupillary dilation (see Procedures for eye examinations above). A standard procedure for examination and technique in regard to performing gonioscopy is recommended. This includes the use of a focal light source *and* magnification (e.g. slit-lamp microscope) as well as special gonioscopy lenses (e.g. Barkan, Koepe, 4-mirror lens). Technical errors (due to position of the goniolens, avoidance of unintentional indentation etc) should be avoided. If performed, the result of gonioscopy shall be recorded on the ECVO Certificate.

It is recommended that gonioscopy is done in breeds in which primary glaucoma is known to occur. A list of relevant breeds is published in the ECVO Manual, chapter 6. Guidelines, and revised regularly by the ECVO HED committee.

The evaluation of the iridocorneal angle (ICA) for iridocorneal angle abnormality (ICAA) is ticked as defined in the ECVO Manual, chapter 6. Guidelines, present at the time of examination.

If any additional (or alternative) method of examination, not mentioned on the form, is used, the box in the examination section, "other" is ticked and specified, and the Certificate is only valid if accompanied by an additional document specifying the method(s) used.

Litter examination

For litter examination a separate Certificate should be issued for each animal examined. The examination can only be performed after permanent identification, e.g. by microchip implantation, of the examined animals. It is, however, possible to use a litter form as long as the data can easily be transferred to the European database.

A registration document may not be available, but any certification can be completed using the details of the dam and sire, together with the date of birth of the puppies and, the microchip numbers (alternatively, a readable tattoo). The distribution of copies is the same as for the Certificate of Eye Examination.

6. Examination outside the Scheme

- If a permanently identified dog or cat is examined outside the Scheme by an ECVO Panellist and a KPHEd is recognized, the Panellist is strongly advised to issue an ECVO Certificate.
- If no certificate is issued, the Panellist is obliged to keep record of such cases and report the number of cases per disease to the ECVO HED-Committee annually (due at the annual HED-committee meeting).
- Mixed-breed dogs or cats that can be identified by microchip or tattoo can be examined within the Scheme, and an ECVO Certificate, with the first part signed by the

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

owner/agent, can be issued. If there is an appropriate system for registration of results, a copy of the certificate should be sent to the relevant body. The Panellist is obliged to keep records of these examinations.

7. Publication of Results

The name of the registered animal examined under the Scheme, the registration and identification numbers, together with the results of the examination shall be made public. They will be sent to the appropriate National registration office. The results will be published in accordance with National regulations.

An ECVO certificate from one country should be accepted by all European National registries.

8. Conflicting results of eye examinations conducted on the same animal

- If the results of two eye examinations of the same animal conflict, the most adverse judgement is valid until the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision will be final.
- When an animal is determined to be “affected” for a known or presumed inherited eye disease (KP-HED) by a panel member or the local appeals authority, and the animal is transferred to another registry, the result “affected” for this KP-HED will not be changed, unless the animal has been re-examined by the appeals authority of the new registry. The latter results, with the exception of conditions that necessitate surgical, or otherwise physiologically altered intervention, (e.g. distichiasis, entropion, etc.), are definitive.
- When an animal is determined to be “affected” for distichiasis by a panel member, the decision is final. When an animal is judged “affected” for another presumed inherited eye disease (KP-HED) which necessitates surgical, or otherwise physiologically altered intervention e.g. entropion, ectropion or macroblepharon, an appeal must be lodged within fourteen (14) days after the initial examination. The animal is then examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision will be final.
- Eye examinations in case of conflicting results are performed at the owner’s expense.

9. 1. “Undetermined” (no. 1-7 on the certificate) cases

If an **adult animal** displays clinical features that possibly fit the KP-HED, but changes are inconclusive, “**undetermined**” is ticked for the relevant disease (no 1-7 on the certificate). It is **recommended** that the animal be **re-examined** by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist **in 6-12 months**, whose decision is final.

- Alternatively, re-examination of cases previously judged “undetermined” can be performed by the first examiner or by another Panellist.
- In the event that the first examiner or another Panellist determines that the animal previously judged “undetermined” is “affected”, this “affected” judgement is valid. If the owner appeals this decision, the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision will be final.
- In the event that the first examiner or another Panellist determines that the animal previously judged “undetermined” is “unaffected”, the “undetermined” judgement remains valid until the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision is final.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

- Alternatively, **if a period of at least 1 year has passed** after the initial examination and the “undetermined” finding in an **adult animal** has not changed at the subsequent examination, it will be judged “undetermined”. If the lesion has disappeared at that examination, it is judged "unaffected" and examination by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist is *not* required. If the finding has progressed, the judgment will be "affected".

Special note for findings in puppies:

Some findings can regress with development (e.g. PPM, lens opacity, PHA, retinal folds); if it is to be expected that the finding will regress, then “undetermined” is ticked at the relevant KP-HED and the animal **re-examined in 1-3 months**.

Findings seen **in puppies** which are typical for the KP-HED and have not changed/not regressed with development (e.g. PPM, retinal “folds”) or have progressed (e.g. lens opacities) until re-examination will be judged “affected”. If the lesion has disappeared at that examination, it is judged "unaffected" (this does not apply for CEA-CH, “go normals”) and examination by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist is *not* required.

Rationale:

In *adult animals* “undetermined” means that the finding is not specific/not typical for the listed KP-HED (as opposed to suspicious = minor typical signs). In *puppies* “undetermined” is used if findings are expected to regress with development.

If the lesion a) has disappeared at the next examination, it is judged "unaffected"; b) remains unchanged in *adult animals* and is still not typical for the KP-HED, "undetermined" is ticked again (breeding advice = optional/low priority); c) has been seen *in the puppy* and is typical for the KP-HED and has not regressed, it is judged “affected”; d) has progressed and/or become typical for the KP-HED listed, it is judged "affected".

Eye examinations in “undetermined” cases as mentioned before, are performed at the owner’s expense.

9. 2. “Suspicious” (no. 11-18 on the certificate) cases

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.

- Alternatively, re-examination of cases previously judged "suspicious" can be performed by the first examiner or by another Panellist.
- In the event that the first examiner or another Panellist determines that the animal previously judged "suspicious" is "affected", this “affected” judgement is valid. If the owner appeals this decision, the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision will be final.
- In the event that the first examiner or another Panellist determines that the animal previously judged "suspicious" is "unaffected", the "suspicious" judgement remains valid

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

until the animal is examined by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist, whose decision is final.

- Alternatively, if a period of at least 1 year has passed after the initial examination and the “suspicious” finding has not changed at the subsequent examination, it will be judged “unaffected” and examination by a minimum of three members of the National Panel or by a Chief or deputy Chief Panellist is not required. If the finding has progressed, the judgment will be “affected”.

Rationale: “suspicious” means that the clinical signs are minor but specific (e.g., a tiny cataract only barely visible with the naked eye in retro-illumination); If the lesion a) has progressed at the next examination - tick “affected”; b) remains unchanged (still “minor”) until the next examination - tick “unaffected” (Note: For KP-HEDs with the advice “NO BREEDING”, the advice applies as long as the lesion is classified as “suspicious”).

Eye examinations in “suspicious” cases as mentioned before, are performed at the owner’s expense.

10. Appeal Procedures

- An owner has the right to appeal against the results of an eye examination.
- Evaluations of appeal cases are performed by a minimum of three members of the National Panel together or by a Chief or deputy Chief Panellist.
- Any appeal against the result of an eye examination should be lodged with the National Panel or National Registry within 60 days after the examination. In the case of choroidal hypoplasia and retinal dysplasia re-examination must be completed before 12 weeks of age or within 5 days if the puppy was aged less than 12 weeks at the time of the initial examination.
- The owner will present the animal, and the Certificate issued by the first Panellist, for examination at the owner’s expense. The most adverse judgement is valid until the animal is examined by a minimum of three members of the National Panel or by a Chief or Deputy Chief Panellist, whose decision will be final.

11. Further details

Further details of the Scheme are issued by the HED-Committee of the ECVO (see ECVO Manual for KP-HED of Dogs and Cats). Unclear rules or regulations, new disease problems or other matters of concern are to be reported by the Panellist to the HED-Committee Chair. The HED-Committee, on the advice of the Advisory Committee, will provide further details and inform the Panellists by Guidelines in the ECVO Manual.

The most recent information regarding the ECVO Scheme can be found at: www.ecvo.org

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

Appendix A

The following list of diseases/variations must have been seen and recorded (at the discretion of the national panel). The protocol must be signed by the supervisor(s)

NOTE: The guidelines published in January of the examination year apply.

	Disease	Number
Globe	Microphthalmos	2
Eyelids	Distichiasis	5
	Lacrimal punctum, atresia	3
Iris	Persistent pupillary membrane (iris-cornea or iris-lens)	3
	Persistent pupillary membrane (iris-iris, crossing pupil)	2
	Iris hypoplasia	2
	Iris atrophy	2
Lens	Cataract, other: nuclear ring	2
	Cataract, other: fiberglass like, pulverulent, punctate	5
	Pigment on anterior lens capsule	5
	Cataract, complete (total)	2
	Cataract posterior cortical, including posterior polar	10
	Cataract, anterior cortical / subcapsular	5
	Cataract, other: anterior suture lines	5
	Cataract, other:suture tips	5
	Cataract, nuclear	5
	Lens (sub)luxation (can be seen without supervision – cats can be included)	5
Vitreous	Asteroid hyalosis	2
	Vitreous in anterior chamber	1
	PHTVL/PHPV grade 1	3
	PHTVL/PHPV grade 2-6	2
Fundus	Retinal detachment, complete (cats can be included) One can be seen without supervision	2
	Retinal detachment, partial (cats can be included) One can be seen without supervision	2
	RD focal/multifocal, retinal folds	5
	RD geographic	2
	CEA, CRD	20
	CEA, coloboma (3 has to be in collie-breeds)	5
	PRA early stage	2
	PRA late stage	3
	Micropapilla/Optic nerve hypoplasia	2
	Non-inherited focal retinopathies	5
	Other presumed hereditary retinopathies* (type described)	4
Other	Iridocorneal angle abnormalities (ICAA) as defined in Chapter 6 (Guidelines) as “affected”	5

*Examples of retinopathies, but not limited to: Canine Multifocal Retinopathy (CMR), Chinese Crested (CC) Pigmentary Chorioretinopathy, Working Dog Retinopathy (WDR), Västgötaspets Retinopathy (J175) (see Chapter 6: Guidelines).

Of the 500 dogs examined, at least 40 must be puppies <10 weeks age of any breed known to be affected by CEA. Of those at least 20 must be collie/sheltie puppies. At least 10 of the puppies must be merle dogs. Five of the 10 merle puppies can be of other breeds than collie/sheltie.

Appendix B

List of relevant literature

Anatomy, embryology and histology, physiology, immunology, pharmacology and vision
Veterinary Ophthalmology 5th ed. Vol I, 2013, Wiley-Blackwell

Pathology

1. Pathology of domestic animals (Jubb & Kennedy), 4th ed., 3 volumes, London Academic Press, 1993, (chapters on eye and related structures).
2. Veterinary Ocular Pathology. A comparative review. Eds.: Dubielzig RR, Ketring KL, McLellan GJ, Albert DM. Saunders Elseviers, 2010.

Neuro-ophthalmology

DeLahunta & Glass: Veterinary neuroanatomy and clinical neurology 3rd ed. Saunders/Elsevier 2009 (chapters related to the eye and adnexa and vision)

Clinical ophthalmology

1. Veterinary Ophthalmology 5th ed. Vol I and II, Wiley-Blackwell, 2013.
2. Slatter's Fundamentals of Veterinary Ophthalmology (Maggs, Miller & Ofri), 4th ed., Saunders/Elsevier, 2009
3. Ophthalmology for the veterinary practitioner (Stades F, Wyman, Boevé, Neumann, Spiess), Schlütersche Hannover, 2007
4. Small animal ophthalmology, a problem oriented approach (Peiffer R. & Petersen-Jones S.), 4th ed., Saunders/Elsevier, 2009.
5. Canine ophthalmology: an atlas and text (Barnett KC, Heinrich C & Samson J), 2002, WB Saunders.
6. Manual of small animal ophthalmology. 3rd ed., BSAVA. Editors: David Gould and Gillian McLellan, 2014.
7. Feline ophthalmology, an atlas and text (Barnett KC & Crispin SM), WB Saunders, 1998
8. Feline Ophthalmology the Manual (Mitchell N & Oliver J), Servet, 2015

Genetics

1. Inherited eye diseases in purebred dogs (Rubin L) Williams & Wilkins, 1989.
2. ACVO Genetics Committee: Ocular disorders proven or suspected to be hereditary in dogs. Vet.Pract.Publishing
3. <http://www.optigen.com>: testing for inherited eye diseases of purebred dogs

Recommended articles

1. Acland, G.M., and Aguirre, G.D.: Retinal degenerations in the dog: IV. Early retinal degeneration (erd) in Norwegian Elkhounds. *Exp. Eye Res.*, 44:491, 1987.
2. Aguirre, G.D., and Acland, G.M.: Variations in retinal degeneration phenotype inherited at the *prcd*.locus. *Exp. Eye. Res.* 46:663, 1988.
3. Aguirre, G.D., and Rubin, L.F.: Progressive retinal atrophy (rod dysplasia) in the Norwegian elkhound: *J. Am. Vet. Med. Assoc.*, 158:208, 1971.
4. Aguirre, GD; Rubin, LF; Pathology of hemeralopia in the Alaskan Malamute dog. *Invest Ophthalmol*; 13, 231-235, 1974.
5. Albert, D.M., et al: Canine herpes-induced retinal dysplasia and associated ocular anomalies. *Invest. Ophthalmol. Vis. Sci.*, 15:267, 1976.
6. Barnett KC, Curtis R. Lens luxation and progressive retinal atrophy in the Tibetan Terrier. *Vet Rec* 103: 160, 1978.
7. Barnett KC. Curtis R. Autosomal dominant progressive retinal atrophy in Abyssinian cats. *J Hered* 76: 168-70, 1985.
8. Bedford PGC. Gonioscopy in the dog. *J Small Anim Pract* 18: 615-29, 1977.
9. Bedford PGC. A gonioscopic study of the iridocorneal angle in the English and American breeds of cocker spaniel and the basset hound. *Journal of Small Animal Practice* 18: 631-642, 1977.
10. Bedford PGC. Collie eye anomaly in the United Kingdom *Vet Rec* 111: 263-70, 1982.
11. Beltran WA, Hammond P, Acland GM, Aguirre GD: A frameshift mutation in RPGR Exon

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

ORF15 causes photoreceptor degeneration and inner retina remodeling in a model of X-linked retinitis pigmentosa. *Invest Ophthalmol Vis Sci* 47, 1669-1681 2006.

12. Bergsjø T, Arnesen K, Heim P, Nes N. Congenital blindness with ocular developmental anomalies including retinal dysplasia, in Doberman Pinscher dogs. *J Am Vet Med Ass* 184: 1383-86, 1984.

13. Bjerkås E, Bergsjø T. Hereditary cataract in the rottweiler dog. *Progr Vet Comp Ophthalmol* 1: 7-10, 1991.

14. Bjerkås E, Haaland M. Pulverulent nuclear cataract in th Norwegian buhund. *J Small AnimPract* 36: 471-74, 1995.

15. Bjerkås E, Ekesten B, Farstad W. Pectinate ligament dysplasia and narrowing of the iridocorneal angle associated with glaucoma in the English springer spaniel. *Vet Ophthalmol* 5: 49-54, 2002.

16. Boevé MH, Stades FC, van der Linde-Sipman, Vrensen GFJM. Persistent hyperplastic tunica vasculosalentis and primary vitreous (PHTVL/PHPV) in the dog: A comparative review. *Progr Vet Comp Ophthalmol*, 2: 163-72, 1992.

17. Carrig CB, Sponenberg DP, Schmidt GM, Tvedten HW. Inheritance of associated ocular and skeletal dysplasia in Labrador retrievers. *J Am Vet Med Assoc* 193: 1269-72, 1988.

18. Cideciyan AV, Jacobson SG, Aleman TS, Gu D, Pierce-Kelling SE, Sumaroka A, Acland GM, Aguirre GD: In vivo dynamics of retinal injury and repair in the Rhodosian mutant dog model of human retinitis pigmentosa. *Proc of the Nat Acad of Sci of the United Stated of America*; 102, 5233-5238, 2005.

19. Cooper AE, Ahonen S, Rowlan JS, Duncan A, Seppälä EH, Vanhapelo P, Lohi H, Komaromy AM: A novel form of progressive retinal atrophy in Swedish Vallhund dogs. *PLOS ONE*, 9, 9, e106610, 2014.

20. Cottrell BD, Barnett KC. Primary glaucoma in the Welsh springer spaniel. *J Small AnimPract* 29: 185-99, 1988.

21. Crispin S, Long SE, Wheeler CA. Incidence and ocular manifestations of multifocal retinal dysplasia in the golden retriever in the UK. *Vet Rec*, 145:669-672. 1999.

22. Curtis R. Histopathological aspects of inherited lens dislocation in the Tibetan Terrier. *J Comp Path* 93: 151-163, 1982.

23. Curtis R, Barnett KC. Primary lens luxation in the miniature bull terrier. *Vet Rec*; 112: 328-329, 1983.

24. Curtis R. Late-onset cataract in the Boston terrier. *Vet Rec*; 115: 577-78 1984.

25. Curtis R, Barnett KC. A survey of cataracts in Golden and Labrador Retrievers. *J Small AnimPract*, 30: 277-86, 1989.

26. Curtis R. Lens luxation in the dog and cat. *Vet Clin North Am: Small AnimPract* 20: 755-773, 1990.

27. Curtis R, Barnett KC. Progressive retinal atrophy in miniature longhaired dachshund dogs. *Br Vet J* 149: 71-85, 1993.

28. Dekomien G, Runte M, Gödde R, Epplen JT. Generalized progressive retinal atrophy of Sloughi dogs is due to an 8-bp insertion in exon 21 of the PDE6B gene. *Cytogenet Cell Genet* 90: 261-267, 2000.

29. Djajadiningrat-Laanen SC, Boevé MH, Stades FC, van Oost BA. Familial non-rcd 1 generalised retinal degeneration in Irish setters *J Sm AnimPract* 44:113-116, 2003.

30. Downs LM, Wallin-Håkansson B, Bournsnel M, Marklund S, Hedhammar Å, Truve' K, Hubinette L, Lindblad-Toh K, Bergström T, Mellersh CS: A frameshift mutation in golden retriever dogs with progressive retinal atrophy endorses *SLC4A3* as a candidate gene for human retinal degenerations. DOI: 10.1371/journal.pone.0021452, 2011.

31. Ekesten B, Komaromy AM, Ofri R, Petersen-Jones SM, Narfström K: Guidelines for clinical electroretinography in the dog: 2012 update. *Doc Ophthalmol*, 127, 2, DOI 10.1007/s10633-013-9388-8, 2013.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

32. Ekesten B, Narfström K. Age-related changes in intraocular pressure and iridocorneal angle in Samoyeds. *Prog Vet Comp Ophthalmol* 2: 37-40, 1991.
33. Ekesten B. Correlation of intraocular distances to the iridocorneal angle in Samoyeds with special reference to angle-closure. *Prog Vet Comp Ophthalmol* 3: 67-73, 1992.
34. Ekesten B, Bjerkås E, Kongsengen K, Narfström K. Primary glaucoma in the Norwegian elkhound. *Vet Comp Ophthalmol* 7: 14-18, 1997.
35. Foster SJ, Curtis R, Barnett KC. Primary lens luxation in the Border Collie. *J Small Anim Pract* 27: 1-6, 1986.
36. Gelatt, K.N. et al.: Animal models for inherited cataracts: A review. *Curr. Eye Res.*, 3(5):765-778, 1984.
37. Gould D, Pettitt L, McLaughlin B, Holmes N, Forman O, Thomas A, Ahonen S, Lohi H, O'Leary C, Sargan D, Mellersh C. ADAMTS17 mutation associated with primary lens luxation is widespread among breeds. *Vet Ophthalmol*. 2011 Nov; 14(6):378-84.
38. Grahn BH, Cullen CL Retinopathy of Great Pyrenees dogs: fluorescein angiography, light microscopy and transmitting and scanning electron microscopy. *Vet Ophth* 4: 191-199, 2001.
39. Holle DM, Stankovics ME, Sarna CS, Aguirre GD. The geographic form of retinal dysplasia is not always a congenital abnormality. *Vet Ophth* , 2: 61-66, 1999.
40. Lazarus JA, Pickett JP, Champagne ES. Primary lens luxation in the Chinese Shar Pei: Clinical and hereditary characteristics. *Vet Ophthalmol*, 1: 101-107, 1998.
41. Leon A, Curtis R, Barnett KC. Hereditary persistent hyperplastic primary vitreous in the Staffordshire bull terrier. *J Am Anim Hosp Assoc* 22: 765-74, 1986.
42. Leppanen M, Mårtenson J, Mäki K. Results of ophthalmologic screening examinations of German Pinschers in Finland – a retrospective study. *Vet Ophth*; 4: 165-169, 2001.
43. Long, SE; Crispin, SM. Inheritance of multifocal retinal dysplasia in the golden retriever in the UK. *Vet Rec* 145: 702-204, 1999.
44. MacMillan, AD; Lipton, DE. Heritability of multifocal retinal dysplasia in American cocker spaniels. *J Am Vet Med Assoc* 172: 568-572, 1978.
45. Martin CL, Wyman M. Glaucoma in the basset hound. *Journal of the American Veterinary Medical Association* 153: 1320-1327, 1968.
46. Martin, C.L., and Chambreau, T.: Cataract production in experimentally orphaned puppies fed a commercial replacement for bitch's milk. *J. Am. Anim. Hosp. Assoc.*, 18:115, 1982.
47. Martin, C.L.: Development of pectinate ligament structure of the dog: Study by scanning electron microscopy. *Am. J. Vet. Res.*, 35:1433, 1974.
48. Martin, C.L.: Gonioscopy and anatomical correlations of the drainage angle of the dog. *J. Small Anim. Pract.*, 10:171, 1969.
49. Martin, C.L.: Scanning electron microscopic examination of selected canine iridocorneal angle abnormalities. *J. Am. Anim. Hosp. Assoc.*, 11:300, 1975.
50. Martin, C.L.: Slit lamp examination of the normal canine anterior ocular segment. Part I: Introduction and technique. *J. Small Anim. Pract.*, 10:143, 1969.
51. Martin, C.L.: Slit lamp examination of the normal canine anterior ocular segment. Part II: Description. *J. Small Anim. Pract.*, 10:151, 1969.
52. Martin, C.L.: Slit lamp examination of the normal canine anterior ocular segment. Part III: Description and summary. *J. Small Anim. Pract.* 10:163, 1969.
53. Martin, C.L.: The normal canine iridocorneal angle as viewed with the scanning electron microscope. *J. Am. Anim. Hosp. Assoc.*, 11:180, 1975.
54. McLellan GJ, Elks R, Lybaert P, Watte C, Moore DL, Bedford PG. (2002) Vitamin E deficiency in dogs with retinal pigment epithelial dystrophy. *Vet Rec* 151(22):663-7
55. Miyadera K, Kato K, Bournsnel M, Mellersh CS, Sargan DR: Genome-wide association study in RPGRIP1 (-/-) dogs identifies a modifier locus that determines the onset of retinal degeneration. *Mammalian Genome*, 23, 212-223, 2011.
56. Narfstrom, K.: Progressive retinal atrophy in the Abyssinian cat: Clinical characteristics.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

Invest. Ophthalmol. Vis. Sci., 26:193, 1985.

57. Narfström K, Dubielzig R. Posterior lenticonus, cataracts and microphthalmia; congenital ocular defects in the cavalier king charles spaniel. *J Small AnimPract*, 25: 669-77, 1984.

58. Narfström K. Cataract in the West Highland white terrier. *J Small AnimPract*, 22: 467-71, 1981.

59. Narfström K, Wrigstad A, Ekestén B, Nilsson SEG. Hereditary retinal dystrophy in the briard dog: Clinical and hereditary characteristics. *Prog Vet Comp Ophthalmol*, 4: 85-92, 1994.

60. Narfström K, Ekestén B: Electroretinographi evaluation of Papillons with and without hereditary retinal degeneration. *Am J of Vet Res*, 59, 221-226, 1998.

61. Narfström K, Jalomäki S, Mowat F, Samardzija M, Chaudieu G, Bergström T, Bragadottir R, Grimm C: Assessment of a novel pigmentary chorioretinopathy in the Chinese crested dog. *JSM Ophthalmol*, 2, 2: 1018, 2014.

62. Parshall CJ, Wyman M, Nitroy S et al. Photoreceptor dysplasia: An inherited progressive retinal dystrophy of miniature schnauzer dogs. *Prog Vet Comp Ophthalmol*, 1: 187-203, 1991.

63. Petersen-Jones SM, Clemens PJM, Barnett KC et al. Incidence of the gene mutation causal for rod-cone dysplasia type 1 in Irish setters in the UK. *J Small AnimPract*, 36: 310-14, 1995.

64. Petersen-Jones SM. Abnormal ocular pigment deposition associated with glaucoma in the Cairn terrier. *J Small AnimPract*, 32: 19-22, 1991.

65. Read RA, Wood JLN, Lakhani KH. Pectinate ligament dysplasia (PLD) and glaucoma in flat coated retrievers. I. Objectives, technique and results of a PLD study. *Veterinary Ophthalmology*, 1: 85-90, 1998.

66. Roberts, S.R., and Dellaporta, A., and Winter, F.C.: The collie ectasia syndrome. Pathology of the eyes of young and adult dogs. *Am. J. Ophthalmol.*, 62:728, 1966.

67. Roberts, S.R., Dellaporta, A., and Winter, F.C.: The collie ectasia syndrome. Pathologic alterations of the eyes of pups one to fourteen days of age. *Am. J. Ophthalmol.*, 61:1458, 1966.

68. Roberts, S.R.: The Collie eye anomaly. *J. Am. Vet. Med. Assoc.*, 155:859, 1969.

69. Ropstad EO, Bjerkås E, Narfström K: Clinical findings in early onset cone-rod dystrophy in the Standard Wire-haired Dachshund. *Vet Ophthalmol*, 10, 2, 69-75, 2007.

70. Schmidt, GM; Ellersieck, MR; *et al.* Inheritance of retinal dysplasia in the English springer spaniel. *J Am Vet Med Assoc*, 174: 1089-1090, 1979.

71. Rubin, LF; Clinical features of hemeralopia in the adult Alaskan Malamute. *J Am Vet Med Assoc* 158: 1696-1698, 1971.

72. Silverstein, A.M.: The pathogenesis of retinal dysplasia. *Am. J. Ophthalmol.*, 72:13-21, 1971.

73. Sargan DR, Withers D, Pettit L, Squire M, Gould DJ, Mellersh CS. Mapping the mutation causing lens luxation in several terrier breeds. *J Hered*, doi:10.1093/jhered/esm029, 2007.

74. Smith RIE, Peiffer RL, Wilcock B. Some aspects of the pathology of canine glaucoma. *Prog Vet Comp Ophthalmol*, 3: 16-27, 1993.

75. Stades FC. Persistent Hyperplastic Tunica Vasculosa Lentis and Persistent Hyperplastic Primary Vitreous (PHTVL/PHPV) in 90 closely related Dobermann Pinschers, Clinical aspects. *J Amer Anim Hosp Assoc*, 16: 739, 1980.

76. Svensson M, Olse'n L, Winkler PA, Petersen-Jones SM, Bergström T, Garncarz Y, Narfström K: Progressive retinal atrophy in the Polski Owczarek Nizinny dog: a clinical and genetic study. *Vet Ophthalmol*, doi:10.1111/vop.12284, 2015.

77. Turney C, Chong NHV, et al. Pathological and electrophysiological features of a canine cone-rod dystrophy in the miniature longhaired dachshund. *Invest Ophthalmol Vis Sci* 48:4240-4249, 2007.

78. van der Linde-Sipman JS. Dysplasia of the pectinate ligament and primary glaucoma in the Bouvier des Flandres dog. *Vet Pathol*, 24: 201-6, 1987.

79. Wallin-Håkanson, B; Wallin-Håkanson, N; Hedhammar, Å; Influence of selective breeding on the prevalence of chorioretinal dysplasia and coloboma in the rough collie in Sweden. *J Small AnimPract.*, 41: 56-59, 2000.

80. Wallin-Håkanson, B; Wallin-Håkanson, N; Hedhammar, Å; Collie eye anomaly in the rough collie in Sweden: genetic transmission and influence on offspring vitality. *J Small AnimPract* 41: 254-258, 2000.

ECVO Manual: Chapter 3. The ECVO Hereditary Eye Disease Scheme (2021)

81. Whiting REH, Narfström K, Yao G, Pearce JW, Coates JR, Castaner LJ, Katz ML: Pupillary light reflex deficits in a canine model of late infantile neuronal ceroid lipofuscinosis. *Exp Eye Res*, 116, 402-410, 2013.
82. Whiting REH, Yao G, Narfström K, Pearce JW, Coates JR, Dodam JR, Castaner LJ, Katz ML: Quantitative assessment of the canine pupillary light reflex. *Vis Neurosci*, 13-12012, 2013.
83. Wood JLN, Lakhani KH, Read RA. Pectinate ligament dysplasia (PLD) and glaucoma in flat coated retrievers. II Assessment of prevalence and heritability. *Veterinary Ophthalmology* 1: 91-99, 1988.
84. Wrigstad A, Nilsson SEG, Dubielzig R, Narfström K. Neuronal ceroid lipofuscinosis in the Polish owczareknizinni (PON) dog. A retinal study. *Doc Ophthalmol*, ; 91: 33-47, 1995.
85. Zangerl B, Goldstein O, Philp AR, Lindaur SJ, Pearce-Kelling SE, Mullins RF, Grapodatsky AS, Ripoll D, Felix JS, Stone EM, Acland GM, Aguirre GD: Identical mutation in a novel retinal gene causes progressive rod-cone degeneration in dogs and retinitis pigmentosa in humans. *Genomics*, 88, 551-563, 2006.
86. Zangerl B, Wikström K, Slavik J, Lindaner SJ, Ahonen S, Schelling C, Lohi H, Guziwicz KE, Aguirre G: Assessment of Canine *BEST1* variations identifies new mutations and establishes an independent bestrophinopathy model (cmr3): *Molecular Vis*, 16, 2791-2804, 2010.

Journals

Veterinary Ophthalmology (last 7 years till 1st of January the year the exam is sat)

Relevant articles in (last 7 years till 1st of January the year the exam is sat):

Journal of Small Animal Practice

Veterinary Record