BVA/KC/ISDS Primary Glaucoma

What is primary glaucoma?

Primary glaucoma is a painful and blinding disease associated with high intraocular pressure (high pressure inside the eye). It is an inherited condition and is subdivided into two types: primary open angle glaucoma (POAG) and primary angle closure glaucoma (PACG)/primary closed angle glaucoma (PCAG). In both forms, glaucoma results from reduced drainage of the fluid (aqueous humour) that is produced within the eye, resulting in a build-up of intraocular pressure which, in turn, leads to pain and blindness. For closed angle glaucoma (but not open angle glaucoma), a screening technique called gonioscopy can identify dogs at risk.

Breeds at risk – open angle glaucoma

In the UK, the Petit Basset Griffon Vendeen, Basset Hound and Shar Pei are the breeds certified for primary open angle glaucoma (POAG) under the Eye Scheme. The genetic mutation responsible for the disease in all these breeds was discovered by geneticists at the Kennel Club Genetics Centre at the Animal Health Trust and DNA tests are available. The DNA tests will be invaluable in eliminating the disease from these breeds as there is no predisposing factor that can reliably be screened for by eye examination before the onset of disease. POAG is silent in onset and the usual clinical features of glaucoma are not present initially, or are so subtle as to be easily missed. Whilst tonometry (measurement of intraocular pressure) and regular examination of breeds at risk when they are 3 years of age and older may be helpful, it is DNA testing that is the essential strategy.

Breeds currently certified for primary open angle glaucoma (POAG) under Schedule A of the Eye Scheme are as follows:

- Petit Basset Griffon Vendeen
- Basset Hound (under Schedule A for POAG and goniodygenesis)
- Shar Pei
Breeds at risk – primary angle closure/closed angle glaucoma

Primary Angle Closure Glaucoma (PACG)/Primary Closed Angle Glaucoma (PCAG) is significantly associated with defective development of the drainage angle which is termed goniodysgenesis (gonio = angle, dysgenesis = defective development), also known as Pectinate Ligament Dysplasia (PLD) or Pectinate Ligament Abnormality (PLA). Goniodysgenesis is inherited in complex fashion in several breeds and is tested for using a technique called gonioscopy. It was originally believed that the degree of goniodysgenesis did not progress after birth and so a ‘one-off’ test before breeding was advised for dogs of certified breeds. However, recent research has provided evidence of progression of goniodysgenesis with age in several breeds, namely the Flat Coated Retriever, Welsh Springer Spaniel, Dandie Dinmont Terrier, Basset Hound and Leonberger. In consequence, the advice on gonioscopy has been updated for all breeds in which gonioscopy is performed. It is advised that for Schedule A breeds gonioscopy should be carried out every 3 years, unless any evidence to the contrary emerges. The first test can be performed in dogs from 6 months of age onwards and current advice is that gonioscopy is performed at approximately 1, 4 and 7-8 years of age. Repeat testing should provide much needed longitudinal information about the risk of developing glaucoma in later life and, in conjunction with Breed Health Coordinators and assistance from the Kennel Club Health Team, will enable breed-specific recommendations to be developed.

Gonioscopy grading

A simple grading scheme (0-3) for gonioscopy was agreed by the Eye Panel Working Party in 2016; a pilot study started in July 1st 2017 with the aim of being formally adopted, with or without any revisions, if analysis of the results supports this approach.

Previously gonioscopy was recorded as either ‘CLINICALLY AFFECTED’ or ‘CLINICALLY UNAFFECTED’. While this is clearly a binary choice, the grading of PLA utilises a continuous scale and so grading can be regarded as more nuanced and helpful when deciding the breeding strategy to adopt.

Initial analysis took place at the Eye Panel Working Party (EPWP) meeting in November 2017 and, as the Kennel Club subsequently produced a means of recording grades on its database, it was agreed that results would be based on the grade only from the initiation of the grading process on July 1st 2017. ‘Clinically Unaffected’ and ‘Clinically Affected’ results under Goniodysgenesis (G) prior to July 1st 2017 have already been published by the KC.

Gonioscopy breeding advice

The aim of grading gonioscopy is to allow for more informed breeding decisions. Under the pilot scheme, dogs are classified as Grade 0 (unaffected), Grade 1 (mildly affected), Grade 2 (moderately affected) or Grade 3 (severely affected).

In general it is recommended that dogs affected by known inherited eye conditions should not be bred from. With regard to goniodysgenesis, it is preferable to breed only from dogs graded as 0 or 1. Dogs graded as 2 (moderately affected) have a greater risk of developing and passing on the condition to offspring and any breeding decisions must take other factors into account, such as significant concerns relating to maintenance of genetic diversity and the prevalence of the condition in the breed. The KC advises that in dogs graded as 2, only those in excellent health and with good results from other screening schemes may be used for breeding, taking particular care to use mates...
with the best possible gonioscopy results (preferably grade 0). Breeding guidelines for Breed Health Coordinators and breeders are available on the KC website https://www.thekennelclub.org.uk/media/1159316/gonioscopy-breeding-advice.pdf. Further research may lead to the updating of this advice.

Gonioscopy summary

<table>
<thead>
<tr>
<th>Grade</th>
<th>Gonioscopic findings</th>
<th>Advice</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal iridocorneal angle (ICA) with no/minimal (0%-&lt;1%) pectinate ligament abnormality (PLA)</td>
<td><strong>Advice:</strong> Normal iridocorneal angle - highly unlikely to develop primary glaucoma Suitable for breeding</td>
</tr>
<tr>
<td>1</td>
<td>1-25% of ICA affected by PLA</td>
<td><strong>Advice:</strong> mildly affected - unlikely to develop primary glaucoma Suitable for breeding</td>
</tr>
<tr>
<td>2</td>
<td>26-75% of ICA affected by PLA</td>
<td><strong>Advice:</strong> moderately affected - low risk of developing primary glaucoma Breed specific advice required if breeding considered</td>
</tr>
<tr>
<td>3</td>
<td>&gt;75% of ICA affected, and/or severe narrowing of ICA</td>
<td><strong>Advice:</strong> severely affected - highest risk of developing primary glaucoma Not recommended for breeding</td>
</tr>
</tbody>
</table>

**Grade 0** – Unaffected (suitable for breeding);

**Grade 1** - mildly affected (unlikely to develop glaucoma and suitable for breeding);

**Grade 2** - moderately affected (low risk of glaucoma; Breed Clubs and Breed Health Coordinators probably best placed to provide breed specific advice, with assistance from the Kennel Club);

**Grade 3** - severely affected (highest risk of glaucoma - not recommended for breeding).
Protocol to be followed for the Pilot Grading Project

On the old Certificate of Eye Examination

INHERITED EYE DISEASE STATUS – SCHEDULE A BREEDS ONLY

Draw a horizontal line through the two tick boxes for (G) Goniodysgenesis.

Record as R grade, L grade and Result immediately next to (G) Goniodysgenesis.

Should there be disparity between the grade assessments in each eye, it is the higher value that is recorded as the Result.

For example: R 1  L 2  Result 2

On the new Eye Examination Certificate

INHERITED EYE DISEASE STATUS – SCHEDULE A BREEDS ONLY

Simply fill in the grade in the table provided and record the grade for each eye – if there is disparity between the Right and Left eye readings it is the higher value that is recorded under Result.

For example: R 1  L 2  Result 2

Breeds currently certified for goniodysgenesis under Schedule A of the Eye Scheme are as follows:

- Basset Hound (under Schedule A for POAG and goniodysgenesis)
- Dandie Dinmont Terrier
- Japanese Shiba Inu
- Leonberger
- Retriever (Flat Coated)
- Siberian Husky
- Spaniel (American Cocker)
- Spaniel (Cocker)
- Spaniel (English Springer)
- Spaniel (Welsh Springer)
- Spanish Water Dog

There are also several breeds where goniodysgenesis is suspected of being inherited, and these breeds are listed under Schedule B. The current list is as follows and is kept under constant review.

- Border Collie
- Great Dane
- Hungarian Vizsla
- Retriever (Golden)
- Welsh Terrier
How do I go about having my dogs examined?

Performing gonioscopy requires certain expertise and specialised equipment and it is for these reasons that gonioscopic examinations are not a routine part of the eye scheme and are not available from every member of the BVA/KC/ISDS Eye Panel. A list of the BVA/KC/ISDS eye panellists is available from the British Veterinary Association or The Kennel Club. However, when telephoning a panellist to book an appointment, owners of the breeds listed above, who wish to have gonioscopy performed should check whether this is available.

How is gonioscopy performed?

Gonioscopy is generally performed without dilating the pupil. After application of local anaesthetic drops to the eye, a special lens (goniolens) is placed on the surface of the cornea to enable the drainage angle to be examined. The test is then repeated on the other eye. Some dogs require sedation for the procedure to be carried out effectively. The fee for sedation is in addition to the cost of the test.

The panellist who performs gonioscopy on your dog should be able to answer any questions that you may have about the findings. There is a set procedure for appealing against the results of an eye examination should you wish to do so and the panellist will supply the requisite leaflet, which is also available on the BVA Website entitled Information for Owners Leaflet. Appeals must be lodged in writing with the BVA within 30 days of the examination.

What is the cost of the test?

Gonioscopy is treated as a separate examination. The scale of fees, as well as a detailed leaflet on all of the conditions covered by the BVA/KC/ISDS Eye Scheme, is available from the British Veterinary Association https://www.bva.co.uk/Canine-Health-Schemes/Eye-scheme

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Telephone: 01234 352672