

# Lines, dots, spots and rings in ophthalmology: understanding eponyms

Eponymous names are familiar to all who have undergone undergraduate and postgraduate training in medicine. The ability to name a few allows one to stand out among your peers and rare, or not so rare, eponymous syndromes are a favourite of college exams and MCQ question books. There are numerous eponyms in medicine and their value in postgraduate training in my experience is similar to the undergraduate years. In modern medicine opinion is divided as to whether new discoveries should have an eponymous title with current trends favouring a move towards descriptive nomenclature. Some eponymous conditions in medicine with ophthalmic manifestations such as Wegner's granulomatosis and Reiter's syndrome are now filled with controversy due to the association between these doctors and the unfortunate events of the Second World War in Germany. Interestingly no studies have ever evaluated whether eponyms aid or hinder learning.

## Eponyms – friend or foe?

Some arguments for eponyms might include the fact that they represent an important part of the history of medicine, they can be a convenient shorthand; indeed some thrive on remembering them and there is a good chance you will need a few before the exams. The debate

against them includes the fact that they are outdated, descriptive terms allow you to determine some aspects of the disease or syndrome such as pathophysiology, scientific discoveries usually involve a team, there is regional and continental differences for example Morbus Horton's disease in Germany vs. maladie de Horton in France; Paget's disease could imply disease of the bone, breast, vulva or penis. There is also overlap in the interpretation of the eponyms with authors describing the same eponym differently.

I recall my first job in ophthalmology where during my first few weeks in theatre my consultant, who was a brilliant teacher, showed me a small haemorrhage occurring in the angles of the eye during cataract surgery and he proceeded to ask me "What is Amsler's sign?" I responded with a minute of silence, hoping the answer would miraculously become apparent to me. I then responded it had something to do with the Amsler chart although the humiliation of not knowing was such that I now know what the Amsler sign is and think of this episode every time I hear it mentioned. Would a descriptive name have saved me embarrassment that day? I determined to make a list of all these eponymous syndromes so that other ophthalmic trainees would be spared the same treatment. I am confident this list will also help with exams. I determined to do this through a prospective review of

medical literature and major ophthalmic text books.

## Aim

- To determine the total number of historical eponymous described as lines, dots, spots and rings or the equivalent in the ophthalmic literature.
- To determine the total number of these eponymous signs in each ophthalmic subspecialty.

## The nature of eponyms

Thirty-six eponyms are described as lines, dots, spots and rings. Seventeen (47.2%) eponyms are lines, four (11.2%) are dots, eight (22.2%) are spots, seven (19.4%) are rings. A majority of the eponyms are located on the anterior segment (81%). Fourteen (39%) of all eponyms are described on the cornea. The retina and the conjunctiva have five (14.0%) eponyms each. The lens has four (10%), the angles of the eye two (5.5%), the choroid two (5.5%), the orbit one (3%), sclera one (3%), periorbit one (3%) and iris one (3%). They are described in the table below.

It is important to note these may not be all the eponyms as only major general ophthalmic textbooks and the opportunistic ones are included. There are also likely geographic differences of the same findings.

## Eponyms and their characteristics.

| Eponym             | Location | Description / context / pathophysiology   | Relevance                                |
|--------------------|----------|---|--|
| Hudson-Stahli line | Cornea   | Brown-green, horizontal line. Middle to inferior 1/3 cornea. Significance: iron deposition, usually with age. Enhanced by chloroquine or hydroxychloroquine.                            | Not usually visually significant         |
| Waring line        | Cornea   | Stellate corneal epithelial iron deposition. After radial keratotomy (RK). Present in up to 80% of RK eyes.   | Not usually visually significant         |
| Khodadoust line    | Cornea   | Line on the corneal endothelium in an eye with a corneal graft. Keratoplasty / corneal graft surgery. Mononuclear cells / keratic precipitates accumulating on the corneal endothelium. | Corneal graft endothelial cell rejection |

| Eponym                               | Location            | Description / context / pathophysiology  | Relevance  |
|--------------------------------------|---------------------|--|--|
| Paton's lines                        | Retina / optic disc | Vertical, circumferential retinal folds especially temporal to the optic disc.<br>On direct ophthalmoscopy they are better visualised when the light on ophthalmoscope is moved back and forth.<br>Optic nerve head swelling, protrusion and eventual corrugation of the retina.<br>Blockage of the axoplasmic transport at the lamina cribrosa. | Optic disc swelling  |
| Sampaoli's line                      | Drainage angles     | Pigment deposition along Schwalbe's line.  | Pseudoexfoliation, pigment dispersion syndrome, iris melanoma, trauma  |
| Scheie's line                        | Lens                | Also see Zentmayer ring.<br>Pigment accumulated at the zonular attachments to the lens.<br>Although some report it as pigment on peripheral posterior lens capsule.  | Pigment dispersion syndrome  |
| Arlt's line                          | Conjunctiva         | Horizontal scarring of the upper tarsal conjunctiva at the junction of the anterior one third and posterior two thirds of the conjunctiva.   | Typically seen in trachoma.<br>Chronic inflammation of tarsal conjunctiva  |
| Stocker's line                       | Cornea              | Yellow or brown deposition in epithelium.<br>Iron deposition can be seen adjacent to the leading edge of pterygium.  | Pterygium  |
| White lines of Vogt                  | Retina              | Sheathed or sclerosed vessels in lattice degeneration.   | Lattice degeneration   |
| Vogt striae                          | Cornea              | Vertical stromal / Descemet's membrane lines.<br>Stress lines due to stretching and thinning.<br>Disappear with globe pressure.  | Keratoconus  |
| Haab striae                          | Cornea              | Horizontal or concentric breaks in Descemet's membrane.<br>Similar to posterior polymorphous dystrophy (PPMD). However on histopathology: the edge of Haab's striae are thickened, curled, with the area between the edge being smooth and thin. This helps differentiate from PPMD.   | Congenital glaucoma  |
| Ohngren's line                       | Orbit               | An x-ray description from 1930.<br>Delineates the limits of resectability of maxillary sinus tumours.<br>If superoposterior it is more likely to invade orbit, ethmoids and paterygopalatine fossa.  | Its use is less certain due to a difference in surgical techniques and treatments  |
| Ferry line                           | Cornea              | Corneal epithelial line at the edge of trabeculectomy blebs.<br>Iron deposition.   | No visual significance   |
| Ehrlich-Turck line                   | Cornea              | Linear deposition of KPs on corneal endothelium.<br>Uveitis.   | Uveitis  |
| Schwalbe's line                      | Drainage angles     | Clinical: gonioscopic view of the drainage angles.<br>Delineates anterior edge of trabeculum and termination of Descemet's membrane.   | Recognition, last to angle structure to disappear in narrow angles   |
| Siegrist streaks                     | Choroid             | Hyper-pigmented flecks that are arranged in a linear fashion along the choroidal blood vessels.<br>Hypertensive choroidopathy.<br>Fidrinoid necrosis.  | Hypertension<br>Giant cell arteritis   |
| Linear naevus sebaceous of Jadassohn | Periorbit           | A congenital hairless plaque that is usually found on the scalp, face or neck.   | Schimmelpenning syndrome: triad of: sebaceous nevi, seizures and learning difficulties.<br>Ocular: coloboma, choristomas, e.g. posterior scleral cartilage |
| Kayes dots                           | Cornea              | Subepithelial infiltrates seen in corneal graft rejection. Could be an elevated line.  | Corneal graft rejection  |
| Gunn's dots                          | Retina              | These are visible reflections of the internal limiting membrane, created by the footplate of the Muller cells. Described in 1918. Note Marcus Gunn was using an ophthalmoscope based on a mirror and a solid flame as an illuminating source to find them.   | Significance: Uncertain.<br>Described as a cause for photosensitivity  |
| Horner-Trantas dots                  | Cornea              | Gelatinous dots at the corneal limbus.<br>Chalky collections of eosinophils at limbus.   | Vernal Keratoconjunctivitis  |
| Mittendorf's dot                     | Lens                | Whitish spot (on direct illumination) at posterior lens surface.<br>Black in retroillumination.<br>Usually nasal / inferonasal.<br>Embryological remnant of the hyaloid artery.  | Associated posterior polar cataract, care as associated with posterior capsule (PC) rupture risk during hydrodissection                                    |

| Eponym                | Location         | Description / context / pathophysiology  | Relevance  |
|-----------------------|------------------|--|--|
| Bitot's spot          | Conjunctiva      | White, foamy area (oval / triangular / irregular in shape) on conjunctiva.<br>Conjunctival squamous metaplasia of bulbar conjunctiva with keratin layer.   | Vitamin A deficiency<br>Xerophthalmia  |
| Elschnig spots        | Choroid          | Hyperpigmented patches in the choroid surrounded by a ring of hypopigmentation.<br>Choriocapillaris hypoperfusion.   | Hypertension   |
| Fuchs spot            | Retina           | Pigmented macular lesion.<br>Significance: retinal pigment epithelium (RPE) hyperplasia / degeneration.<br>Forster-Fuchs' retinal spot: subretinal neovascularisation.   | Pathological myopia  |
| Brushfield spot       | Iris             | White, grey spots in peripheral iris.<br>Iris stromal hyperplasia and surrounding hypoplasia.  | Down's syndrome present in around 78%  |
| Roth's spots          | Retina           | Haemorrhages with white centre.<br>Platelet and fibrin thrombus at the centre of a ruptured capillary network.<br>Also: immune complex mediated vasculitis.  | Numerous e.g.: <ul style="list-style-type: none"> <li>Subacute bacterial endocarditis</li> <li>Leukemias</li> <li>Anaemia</li> <li>Anoxia</li> <li>Carbon monoxide poisoning</li> <li>Eye decompression</li> </ul> |
| Koplik's spot         | Conjunctiva      | Occur on the conjunctiva and resemble specks of sand surrounded by a red areola.<br>Also curuncle lesion and semilunar fold (Hirschberg's sign).   | Measles  |
| Fischer-Khunt spot    | Sclera           | Blue grey plaque anterior to horizontal recti insertions.<br>Senile scleral plaque.<br>Area of hyalinised sclera.  | Seen in old age  |
| Krachmer spots        | Cornea           | Sub-epithelial opacities similar in appearance to adenovirus keratitis.<br>Present Bowman's layer.<br>Note: Stromal swelling can coincide leading to some calling it a stromal rejection.                                  | Corneal graft epithelial rejection   |
| Kayser-Fleischer ring | Cornea           | Deposition in peripheral cornea with a gold / brown / yellow / green hue.<br>Starts superior, then inferior and finally circumferential.<br>Copper deposition in Descemet's membrane.                                      | Wilson's disease present in about 95% with neurological Wilson's.<br>Whilst around 65% in those with hepatic disease   |
| Fleischer's ring      | Cornea           | Best seen with cobalt blue filter.<br>Basal epithelial iron deposition around base of cone.  | Keratoconus  |
| Vossius ring          | Lens             | Iris pigment imprinting on the anterior lens capsule.<br>Pigment after trauma.<br>Note force has to be high enough to flatten cornea.  | Significant traumatic force likely   |
| Wessley ring          | Cornea           | At the level of the stroma.<br>Corneal viral antigen precipitate.<br>Type 3 immune response involving antigen-antibody complex formation.  | Not specific as numerous causes possible e.g. trauma, infectious vs. sterile causes.<br>Microbial keratitis important cause.<br>Beware in contact lens wearers, etc.   |
| Soemmering's ring     | Lens             | Doughnut shaped ring at capsule.<br>In pseudophakia and also reported in Aphakia or ocular trauma.<br>Cortical regeneration and transformation into Elschnig pearls and an element of equatorial epithelial proliferation. | Incomplete cortex removal<br>Trauma  |
| Zentmayer ring        | Lens             | See Scheie line.<br>Pigment accumulated at the zonular attachments to the lens.  | Pigment dispersion syndrome  |
| Coat's white ring     | Cornea           | Granular, oval ring, patient asymptomatic.<br>Iron deposition at the level of Bowman's layer.<br>Usually associated with a previous corneal foreign body.  | Not usually visually significant   |
| Amsler's Sign         | Anterior chamber | A haemorrhage opposite a paracentesis site due to rupturing of small angle vessels in Fuchs' Heterochromic Iridocyclitis.  | Can cause problems during intraocular surgery  |

**Conclusion**

I hope this list of eponymous syndromes help those undertaking exams to answer those critical questions that are unanswerable using common sense alone. They are disproportionately important for this reason more than any other. Despite the pros and cons of abolishing the use of eponymous syndromes for the foreseeable future they are here to stay and thus knowledge of their meaning is a prerequisite for success.

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**SECTION EDITOR**

**Gwyn Samuel Williams,**  
ST7 Ophthalmology,  
University Hospital of Wales,  
Cardiff, UK.

E: [gwynwilliams@doctors.org.uk](mailto:gwynwilliams@doctors.org.uk)

**AUTHOR**

**Tafadzwa Young-Zvandasara,**  
ST4 Ophthalmology,  
Royal Glamorgan Hospital,  
Wales, UK.