A RATIONALE FOR THE EYES

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A RATIONALE FOR THE EYES

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A guide to the diagnosis of eye diseases, discharges, vision problems and damage.
CONTENTS

Introduction

SECTION ONE
Red Eye Diagnostic Flow Chart
A flow chart that leads the user through various symptoms and signs to possible diagnoses.

SECTION TWO
Diagnostic Algorithm for Eye Symptoms
Symptoms involving the eye and the conditions that may be responsible

SECTION THREE
Ophthalmological Conditions
The symptoms, signs, investigation and treatment of medical conditions that may cause an alteration in the eyes.

Appendices
Eye anatomy, physiology and investigation
INTRODUCTION

This book is designed for both the medical student and the doctor who is not a specialist in ophthalmology.

It will take the user through a logical rationale in order to diagnose, and then treat, virtually every eye condition likely to be encountered outside a specialist practice.

There are two ways to reach a diagnosis, using the flow chart in Section One, or the Diagnostic Algorithms in Section Two.

In Section One, a flow chart will guide the user through the presenting symptoms and signs of most eye conditions to a selection of possible diagnoses.

As an alternative, the algorithms in Section Two will indicate the diagnoses possible with a variety of ophthalmological presenting symptoms.

Once a diagnosis has, or number of differential diagnoses have been made, a detailed explanation of the various ophthalmological diagnoses can be found in the largest part of the book, Section Three. This has been written in a style that should be easy to understand by even junior medical students, with technical terms explained in each monograph, but should still be useful to the non-specialist doctor. The symptoms, signs, investigations and treatment of a very wide range of conditions are explained, along with pictures of the more common conditions.

I trust that you will find it useful.

Warwick Carter
Brisbane

OTHER BOOKS IN THIS SERIES

A Rationale for Rashes
A Rationale for the Brain
A Rationale for the Abdomen
A Rationale for the Chest
Section ONE

RED EYE
DIAGNOSTIC FLOW CHART
A RATIONALE FOR THE EYES

DIAGNOSTIC ALGORITHM FOR A RED EYE

1. Is the globe painful? YES
2. Is there a mucopurulent discharge? YES
3. Are the pupils reacting normally? YES
4. Is the undersurface of the lids cobblestone in appearance? YES

5. Is there a discharge? NO
6. Are the eyelids stuck together on waking? NO
7. Are eyelids sticky together during day? NO
8. Do the eyes feel or appear dry? NO
9. Are tears excessive? NO

10. Is there a corneal foreign body? NO
11. Is the vision blurred or impaired? NO
12. Is there a foreign body? NO
13. Is the pupil size and reaction normal? NO

14. Stain the eye
15. Is there a foreign body? NO
16. Is there an abrasion? NO
17. Is there a unilateral headache? NO
18. Is there a branching ulcer? NO
19. Is the anterior chamber shallow? NO
20. Is the eye itchy? NO

21. Perform tonometry
22. Is the iris swollen with blurred markings? NO

23. Refer for investigation

This algorithm is only a guide to diagnosis and causes other than those indicated by the algorithm must always be considered by the attending physician.

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SEE SEPARATE jpg FILE FOR MORE DETAIL
## Guide to Diagnosis of Eye Diseases

<table>
<thead>
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<th>Symptom</th>
<th>Red Eye</th>
<th>Painless</th>
<th>Itchy Eye</th>
<th>Pus</th>
<th>Watery Eye</th>
<th>Dry Eye</th>
<th>Vision Spots</th>
<th>Light Sensitive</th>
<th>Poor Vision</th>
<th>Tender Eye</th>
<th>Ulcer</th>
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Number of x determines severity of symptom
A RATIONALE FOR THE EYES

Section Two

DIAGNOSTIC ALGORITHMS FOR EYE SYMPTOMS
A RATIONALE FOR THE EYES

DIAGNOSTIC ALGORITHMS FOR EYE SYMPTOMS AND SIGNS

FORMAT

Presenting Symptom (Alternate Name)
Explanation of terminology
System or other group of symptoms
Diagnoses that may present with this symptom [alternate name of diagnosis] (other symptoms of each diagnosis, or a discussion of the diagnosis)
Other entries to consider

Clinical Sign (Alternate Name) [Abbreviation]
Exp: An explanation of the sign, with its methodology described in sufficient detail to enable the practitioner to perform the test.
Int: The interpretation of the sign.
(+): The diseases, syndromes etc. that should be considered if the test is positive
(++) The interpretation of an exaggerated or grossly positive test
(−): Ditto for a negative test result
(AB): Ditto for an abnormal test result
Phys: The pathophysiology of the sign to enable its significance to be better understood
Other entries to consider

Adie’s pupil
See Aniscoria

Amaurosis Fugax
See Vision Loss

Amblyopia
Decrease in vision sense
Strabismus (squint)
Double vision of any cause
Cataract (cloudy lens)
Severe refractive errors [eg. hyperopia or astigmatism]
Vitamin B deficiency
Strachan syn. (orogenital dermatitis, neuropathy)

Aniscoria
Exp: Pupils are different sizes
Int: (+) Physiological (20% of population), raised intracranial pressure, abnormal migraine, third nerve palsy, Adie’s pupil (ciliary muscle paresis), Horner syndrome, Pancoast tumour, penetrating trauma, medications (eg. atropine)
Phys: Causes may be physiological, structural, pharmacological or neurological

Arcus Senilis
Exp: An opaque ring in the peripheral cornea with a clear zone separating it from the limbus
Int: (+) Occurs invariably with advancing age, hypercholesterolaemia
Phys: Deposition of lipids in periphery of cornea

Argyll Robertson Pupil
Exp: Small, irregular, unequal pupils that do not react to light, but do react to convergence
Int: (+) Neurosyphilis, tabes dorsalis, diabetes
Phys: Damage to the midbrain section of the optic tract

Arlt’s Line
Exp: Cicatricial scarring of upper eyelid causing entropion

See Vision Loss
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Int: (+) Chronic trachoma
Phys: Chronic inflammation of subepithelial tissue of the tarsus
See also Entropion

Arteriovenous Nipping, Retinal
Exp: Ophthalmoscopic or slit lamp examination of the retina reveals narrowing of venules where they are crossed by arterioles
Int: (+) Hypertension, arteriosclerosis
Phys: Increased pressure on the venule in the shared adventitial sheath where vessels cross

Black Eyes, Bilateral
Exp: Both eyelids spontaneously blackened after trauma excluded. May be precipitated by sigmoidoscopy
Int: (+) Amyloidosis
Phys: The Valsalva manoeuvre of sigmoidoscopy may be a precipitant

Black Spots in Visual Field
See Vision, Black Spots in Field of

Blindness
See Vision, Loss of

Blue Sclera
See Sclera, Blue

Blurred Vision
See Vision, Blurred

Buphthalmos
See Exophthalmos

Cataract
Senile cataract due to age
Genetic and chromosomal causes (eg. Down syn., Patau syn., X-linked recessive disorders)
Alport’s disease (nepropathy, deaf)
Rubella in utero
Diabetes mellitus (polyuria, polydipsia)
Hypocalcaemia (tetany)

Infantile hypoxia
Starvation and malnutrition
Radiation (ultraviolet, x-ray, infrared)
Uveitis (inflammation)
Retrolental fibroplasia

Syndromes
Conrad syn. (deaf, limb contractures)
Fuchs syn. (anterior uveitis, different coloured irises)
Hallerman-Streiff syn. (face anomalies, dwarf)
Lowe syn. (retarded, epicanthal folds)
Marinesco-Sjögren syn. (ataxia, retarded)
Maroteaux-Lamy syn. (bone dysplasia, cardiac lesions)
Morquio syn. (bone dysplasia, deaf)
Scheie syn. (recurrent respiratory infections, kyphosis)
Sly syn. (recurrent respiratory infections, kyphosis)

Other
Drugs (eg. glucocorticoids, phenothiazines, heavy metals)
See also Lens Opacity; Vision, Loss of

Conjunctivitis
See Eye Pain; Eye, Inflamed or Red

Consensual Reflex
Exp: Light shone into one eye causes the pupils of both eyes to contract
Int: (–) (only opposite eye contracts)
Phys: Afferent path of reflex arc is interrupted, but efferent path remains intact

Cornea
See Corneal Anaesthesia; Corneal Reflex; Eye Pain; Eye, Dry; Eye, Inflamed or Red; Eye, Watery

Corneal Anaesthesia
Exp: Loss of corneal sensation to light touch
Int: (+) Riley-Day syn.

Corneal Reflex
Exp: Lightly touching one cornea with a piece of cotton wool produces blinking in both eyes
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Diplopia
Paralytic squint due to 3rd, 4th or 6th cranial nerve palsy (limited movement of one eye)
Cerebrovascular accident (neurological changes)
Concussion (trauma, headache, nausea)
Orbital trauma
Migraine (nausea, headache, photophobia)
Botulism (dry mouth, dysphagia, paralysis)
Cerebral tumours (headache, neurological signs)
Myasthenia gravis (weakness, ptosis)
Thyroid diseases
Multiple sclerosis (weakness, abnormal sensation)
Gradenigo syn. (headache, facial pain)
Wernicke-Korsakoff syn. (ataxia, demented)

Exophthalmos
(Proptosis)
Protrusion of eyeballs within sockets. Marked amount of sclera visible above iris in normal forward vision

Int: (+) Hyperthyroidism, cerebral tumour, optic or orbital tumour, Cushing’s disease, cavernous sinus thrombosis, Hand-Schuller-Christian disease, pituitary tumours, osteomas, neurofibromatosis, Wegener's granulomatosis, metastatic carcinoma, xanthomas, malignant hypertension, uraemia, cellulitis, vascular malformation, lacrimal tumours, mucocele, rhabdomyosarcoma, Apert syn., Crouzon syn., Sturge-Webber syn.

Phys: Increase in the volume of orbital contents.

Exudates, Retinal
See Retinal Exudates

Eye Discharge
Bacterial conjunctivitis (purulent, red, pain)
Viral conjunctivitis (serous, red, slight pain)
Allergic conjunctivitis (mucoid, red, itch)
Herpetic ulcer (serous, severe pain, photophobia)
Foreign body (serous, irritation, severe pain)
Arc damage (serous, red, severe pain)
Iritis (serous, depressed vision, pain)
Acute glaucoma (serous, severe pain, poor vision)
Scleritis (serous, severe pain, partial redness)
See also Eye, Watery

Eye Pain
Ophthalmic
Iritis (red, sluggish pupil, blurred vision)
Glaucoma (blurred vision, small pupil)
Conjunctivitis (red, discharge, photophobia)
Scleritis (red, photophobia, serous discharge)
Arc or UV damage (red, photophobia)
Herpetic conjunctivitis (photophobia, serous discharge)
Foreign body (history, sensation)
Keratitis and corneal ulcer (discharge, blurred vision)

Other
Sinusitis (fever, catarrh, rhinorrhoea)
Ankylosing spondylitis (back pain, arthritis)
Cluster headache (sudden, unilateral, lacrimation)
Migraine (headache, photophobia, nausea)
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Ramsay Hunt syn. (blistering rash)
Ulcerative colitis (diarrhoea, rectal blood)
Hyperthyroidism (sweaty, fatigue, exophthalmos)
Yellow fever (generalised pain, jaundice, fever)
Syphilis (varied symptoms)
Sjögren syn. (dry mouth, dysphagia)
Reiter syn. (urethritis, arthritis)
Leptospirosis (fever, myalgia)
See also Photophobia

Eye, Black Spots in Visual Field
See Vision, Black Spots in Field of

Eye, Dry
(Xerophthalmia)
Vitamin A deficiency (dry skin, loss of night vision, corneal ulcers)
Primary biliary cirrhosis (pruritus, dry mouth, cholestasis)
Rheumatoid arthritis (joint pain and swelling)
Idiopathic keratoconjunctivitis sicca (skin eczema)
Diabetes mellitus (polydipsia, polyuria)
Viral conjunctivitis (chronic)
Erythema multiforme
Mucus pemphigoid
Trachoma
Sjögren syn. (dry mouth, dysphagia)
Other autoimmune disorders
Dry eye syn. (see Syndromes section 6)
Facial nerve paresis
Riley-Day syn. (see Syndromes section 6)
Trauma
Irradiation damage
Drugs (eg. antihypertensives, psychotherapeutics, sympathomimetics)

Eye, Inflamed or Red
Bacterial or viral conjunctivitis (diffuse injection, discharge, pain)
Iritis (circumcorneal injection, pain, blurred vision)
Glaucoma (pain, blurred vision)
Corneal trauma or foreign body (unilateral, blurred vision, pain)
Allergic conjunctivitis (itch, watering, oedema, slight pain)
Keratitis (corneal ulcer, pain, discharge)
Acute glaucoma (severe pain, photophobia)
Trachoma (pain, discharge, lymphoid hypertrophy)
Cluster headaches (lacrimation, unilateral)
Leptospirosis (fever, myalgia)
Chemical irritation (eg. pool chlorine)
Excess alcohol
Drug abuse (eg. cocaine)

Syndromes
Louis-Bar syn. (mental deterioration, telangiectasia)
Reiter syn. (urethritis, arthritis)
Stevens-Johnson syn. (erythema, stomatitis)
Toledo-Hunt syn. (ptosis, unilateral ophthalmoplegia)
Toxic shock syn. (fever, diarrhoea, vomiting)
Behçet syn. (arthritis, genital and mouth ulcers)
Heerfordt syn. (sarcoiditis, adenitis)
Richner-Hanhart syn. (tyrosinaemia)
Rieger syn. (iris dysgenesis, small teeth)
Uveoparotid syn. (parotid hypertrophy, facial paralysis)
Vogt-Koyanagi-Harada syn. (vertigo, blind)
See also symptoms listed under Eye

Eye, Red
See Eye, Inflamed or Red

Eye, Watery
(Epiphora)
Excess Tear Production
Trauma and emotion
Infections (inflammation, pus)
Foreign bodies (pain, inflammation)
Dendritic ulcers
Congenital glaucoma
Congenital cataract
Iritis (pain, inflammation)
Entropion (infolding of lower lid)
Trichiasis (ingrown eye lash)

Obstructed Tear Drainage
Congenital tear duct obstruction
Trauma to medial canthus
Ectropion (everted lower lid)
Proptosis
Foreign body in tear duct
Radiation
Dacryocystitis
Sinusitis (pain, fever)

Eyelid Disease
Stye (tender red swelling)
Chalazion (Meibomian gland infection)
Blepharitis (generalised inflammation of lid margins)
Entropion (inward rolling of lower lid)
Ectropion (outward rolling of lower lid)
Allergic dermatitis (swollen excoriated lids)
Ptosis (see separate entry)
**Halos, Visual**
- Cataracts (cloudy cornea)
- Glaucoma (subacute angle closure type)
- Allergic conjunctivitis
- Conjunctival oedema
- Corneal disease
- Contact lenses
*See also Rainbow Halo*

**Hoagland’s Sign**
- **Exp:** Eyelid oedema
- **Int:** (+) Infectious mononucleosis

**Horizontal Light Test**
- **Exp:** Shine a bright narrow beam of light horizontally onto the eye from its temporal aspect. Observe the iris shadow pattern
- **Int:** Iris evenly illuminated or shadow on temporal side – Deep anterior chamber (glaucoma unlikely)
  - Shadow on nasal side – Shallow anterior chamber (potential for acute angle closure glaucoma)
- **Phys:** The iris is normally concave, but when convex it narrows the anterior chamber and casts a shadow on the nasal side
*See also Swinging Torch Sign*

**Inflamed Eye**
- **See Eye, Inflamed or Red**

**Iris Abnormality Associated Syndromes**
- Gillespie syn. (aniridia, mental retardation)
- Miller syn. (aniridia, Wilm’s tumour, mental retardation)
- Pseudoexfoliation syn. (glaucoma)
- Rieger syn. (iris dysgenesis, small teeth)
- WAGR syn. (Wilms’ tumour, mental retardation)
- Williams syn. (blue iris, prominent lips)

**Kayser-Fleischer Ring**
- **Exp:** Greenish-brown ring at the outer edge of the cornea
- **Int:** (+) Wilson’s disease
- **Phys:** Deposition of copper compounds in cornea (diagnostic sign)
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Lacrimation, Abnormal
Facial nerve palsy
Crocodile tears syn. (lacrimation with eating after Bell’s palsy)
Mikulicz syn. (parotitis, enlarged lacrimal glands)
Riley-Day syn. (Jewish, excess sweating, fever)
Sicca syn. (dry eyes, adenitis)
Sjögren syn. (dry eyes and mucous membranes)
SUNCT syn. (severe brief headache)

Phys: Damage to or inhibition of the iris and its musculature or innervation

Myosis

Nystagmus
Exp: Involuntary rhythmic movement of eyeball. Two types:
    Pendular (oscillating) – with regular movements
    Jerk (rhythmic) – with movement faster in one direction than the other

Int: (+) Normal with acute lateral vision and watching a moving object, barbiturates, labyrinthine and vestibular disease, brain stem lesions (often vertical nystagmus), demyelinating diseases (eg. multiple sclerosis), during epileptic fit (eg. petit mal), brain tumours, syringobulbia, Dandy-Walker syn., Parinaud syn., diencephalic syn., pinealoma, central vision loss (eg. albinism, retinal disease), other visual disturbances, cerebral abscess, coma, Friedreich’s ataxia, congenital, alcohol, some normal infants

Phys: Jerk form more common and is neurological in aetiology. Pendular is due to a visual defect. Direction of nystagmus can give further clue to localise lesion

Oculogyric Crisis
Exp: Varies from mild cases with abnormal uncontrolled random eye movements, to severe cases with fixed elevated gaze associated with painful extension of the neck which may be so severe that the occiput nearly touches the thoracic vertebrae and the airway may be compromised

Int: (+) Rare side effect of prochlorperazine, encephalitis or Parkinson’s disease

Phys: Effect rapidly reversed by IV benztropine

Oedema, Eyelid
Angioneurotic oedema (pruritus, diffuse) Insect bites and trauma
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Thyrotoxicosis (fatigue, sweating, weight loss)
Superior vena cava obstruction (cyanosis, venomegaly)
Hypothyroidism (dry skin, myalgia, deaf)
Trichinosis (myalgia, nausea, diarrhoea)

Opacity of Lens
See Lens Opacity

Optic Atrophy
Exp: On ophthalmoscopic examination of retina, a pale optic disc with blurred margins is noted. Disc cupping may also occur, and patient complains of reduced visual acuity. Field defects may be found
Int: (+) Glaucoma, arteriosclerosis, retinal ischaemia, optic neuritis, Paget's disease, tumour pressing on optic nerve, retinitis pigmentosa, vitamin B deficiency, methanol poisoning
Phys: Ischaemia or chronic inflammation of optic disc

Optic Cup/Disc Ratio
Exp: Ratio of the vertical diameter of the optic cup to that of the optic disc, measured on ophthalmoscopic or slit lamp examination of the retina
Int: Normal Optic Cup/Disc Ratio = 0.2
<0.5 – Probably normal
>0.5 – Suspicious of glaucoma
>0.8 – Usually diagnostic of glaucoma

Optic Disc, Abnormal
Exp: Characteristics noted during ophthalmoscopic examination of retina
Int: (+) Extra vascularity – Optic neuritis
(+) Papilloedema (bulging of disc) – Increased intracranial pressure
(+ Atrophy (reduced vascularity) – Optic nerve disease (see Optic Atrophy)
(+ Pale disc – Central retinal artery occlusion
(+ Haemorrhages – Central retinal vein occlusion
See also Optic Cup/Disc Ratio

Optic Paralysis
Cogan syn. (no horizontal eye movement)
Duane syn. (deficient horizontal eye movement)
Moebius syn. (ptosis, fixed facies)
Parinaud syn. (loss of upward gaze)
Steele-Richardson-Olszewski syn. (rigidity, dementia)
Tolosa-Hunt syn. (unilateral, ptosis, mydriasis)
Wernicke-Korsakoff syn. (dementia)

Papilloedema
Exp: On ophthalmoscopic or slit lamp examination, the optic disc is noted to be flattened, swollen or protruberant bilaterally with blurred edges. Absent venous pulsation, dilated retinal veins, and flame shaped haemorrhages may also be noted
Int: (+) Increased intracranial pressure (eg. haemorrhage, tumour, meningitis, cerebral abscess, emphysema, hypoparathyroidism), optic neuritis, hypertension, multiple sclerosis, Guillain-Barré syn.
Phys: Increased CSF pressure, due to an increase in CSF volume (caused by haemorrhage or increased protein content) or blocked CSF circulation, is transmitted along the sheath of the optic nerve to the optic disc. Vision remains unimpaired in early stages

Photophobia
Eye pain with bright light
Migraine (headache, nausea, vertigo)
Meningitis (headache, neck stiffness, fever)
Iritis (pain, blurred vision, small pupil)
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Corneal inflammation (eg. herpetic ulcer) 
(discharge, blurred vision) 
Corneal foreign body (red, pain) 
Acute glaucoma (severe pain, poor vision) 
Episcleritis (pain, discharge) 
Hypoparathyroidism (tetany, wheeze, convulsions) 
Rickettsial and viral infections (eg. measles) 
Trichinosis (myalgia, nausea, diarrhoea) 
Richner-Hanhart syn. (tyrosinaemia, mental retardation)

Pupil, White 
(Leucocoria) 
Exp: Pupil of eye is white and reflects light 
Int: (+) Cataract, retinoblastoma, retinal fibroplasia, corneal scarring, persistent tunica vasculosa lentis 
See also Lens Opacity

Rainbow Halo 
Exp: Rainbow coloured halo seen around lights, particularly at night 
Int: (+) Acute angle closure glaucoma, corneal oedema, incipient cataract, allergic conjunctivitis 
Phys: Distortion of light, as in a prism, by unequal alignment of inner and outer surfaces of cornea 
See also Halos, Visual

Red Eye 
See Eye, Inflamed or Red

Retinal Arteriovenous Nipping 
See Arteriovenous Nipping, Retinal

Retinal Exudates 
Exp: Ophthalmoscopic examination of retina reveals white fluffy patches 
Int: (+) Diabetes mellitus, hypertension, increased intracranial pressure, massive blood loss 
Phys: Occlusion of retinal capillaries

Retinal Haemorrhages 
Exp: Red spots and patches adjacent to blood vessels are noted on ophthalmoscopic examination of the retina. Various types described as punctate, splinter and flame 
Int: (+) Pernicious anaemia, leukaemia, aplastic anaemia, hypertension, diabetes mellitus, bacterial endocarditis, anticoagulants, haemorrhagic disease 
Phys: Damaged retinal capillaries 
See also Roth’s Spots

Pupil Changes 
See Aniscoria; Leucocoria; Miosis; Mydriasis; Lens Opacity; Pupil, Irregular; Pupil, White 
Pupil, Dilated 
See Aniscoria

Pupil, Irregular 
Exp: Pupil of eye is irregular in outline 
Int: (+) Iritis, surgery, trauma, pupillary membrane, congenital

Proptosis 
See Exophthalmos

Ptosis 
Drooping eyelid(s) 
Bell’s palsy (unilateral, spontaneous, painless) 
Myasthenia gravis (generalised weakness) 
Third cranial nerve palsy from any cause 
Pseudoptosis (fat deposits in lid)

Syndromes 
Dubowitz syn. (reduced growth, mental retardation) 
Eaton-Lambert syn. (myasthenic symptoms) 
Guillain-Barré syn. (progressive palsy) 
Horner syn. (myosis, exophthalmos, anhidrosis) 
Marcus Gunn syn. (lid twitch with jaw movement) 
Moebius syn. (ophthalmoplegia, dysphagia, drool) 
Pancoast syn. (arm and chest pain, lung cancer) 
Parinaud syn. (loss of upward gaze) 
Smith-Lemli-Opitz syn. (mental retardation, hypospadias) 
Tolosa-Hunt syn. (pain, mydriasis) 
Uveoparotid syn. (facial paralysis, uveitis) 
Wernicke-Korsakoff syn. (ataxia, dementia) 
See also Eyelid Disease; Eyelids, Abnormal

Pupil, White 
(Leucocoria) 
Exp: Pupil of eye is white and reflects light 
Int: (+) Cataract, retinoblastoma, retinal fibroplasia, corneal scarring, persistent tunica vasculosa lentis 
See also Lens Opacity

Rainbow Halo 
Exp: Rainbow coloured halo seen around lights, particularly at night 
Int: (+) Acute angle closure glaucoma, corneal oedema, incipient cataract, allergic conjunctivitis 
Phys: Distortion of light, as in a prism, by unequal alignment of inner and outer surfaces of cornea 
See also Halos, Visual

Red Eye 
See Eye, Inflamed or Red

Retinal Arteriovenous Nipping 
See Arteriovenous Nipping, Retinal

Retinal Exudates 
Exp: Ophthalmoscopic examination of retina reveals white fluffy patches 
Int: (+) Diabetes mellitus, hypertension, increased intracranial pressure, massive blood loss 
Phys: Occlusion of retinal capillaries

Retinal Haemorrhages 
Exp: Red spots and patches adjacent to blood vessels are noted on ophthalmoscopic examination of the retina. Various types described as punctate, splinter and flame 
Int: (+) Pernicious anaemia, leukaemia, aplastic anaemia, hypertension, diabetes mellitus, bacterial endocarditis, anticoagulants, haemorrhagic disease 
Phys: Damaged retinal capillaries 
See also Roth’s Spots

Pupil Changes 
See Aniscoria; Leucocoria; Miosis; Mydriasis; Lens Opacity; Pupil, Irregular; Pupil, White 
Pupil, Dilated 
See Aniscoria

Pupil, Irregular 
Exp: Pupil of eye is irregular in outline 
Int: (+) Iritis, surgery, trauma, pupillary membrane, congenital

Proptosis 
See Exophthalmos

Ptosis 
Drooping eyelid(s) 
Bell’s palsy (unilateral, spontaneous, painless) 
Myasthenia gravis (generalised weakness) 
Third cranial nerve palsy from any cause 
Pseudoptosis (fat deposits in lid)

Syndromes 
Dubowitz syn. (reduced growth, mental retardation) 
Eaton-Lambert syn. (myasthenic symptoms) 
Guillain-Barré syn. (progressive palsy) 
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Uveoparotid syn. (facial paralysis, uveitis) 
Wernicke-Korsakoff syn. (ataxia, dementia) 
See also Eyelid Disease; Eyelids, Abnormal
A RATIONALE FOR THE EYES

Retinal Pigmentation
(Retinitis Pigmentosa)
Exp: Cells filled with epithelial pigment congregate beside retinal blood vessels
Int: (+) Bassen-Kornzweig syn., Laurence-Moon-Biedl syn., other genetic conditions
Phys: Night blindness and tunnel vision early symptoms

Roth’s Spots
Exp: Superficial retinal haemorrhages with pale or white centres
Int: (+) Retinal infarct, leukaemia, retinal haemorrhage with central resolution
Phys: In leukaemia, due to extravasation of excess white corpuscles from a retinal haemorrhage
See also Retinal Haemorrhages

Sclera, Blue
Exp: Markedly blue sclera (not iris) noted
Int: (+) Fragilitas ossium (osteogenesis imperfecta)
Phys: Congenital disease transmitted by a dominant mutant gene

Spots in Vision
See Vision, Black Spots in Field of

Squint
(Strabismus)
Deviation of eye axes
Paralytic (limited movement of affected eye caused by trauma, vascular disease, cerebrovascular accident, tumour, multiple sclerosis, hyperthyroidism, etc.)
Concomitant (deviation constant at all angles of gaze due to congenital cataract, infantile lazy eye, etc.)
Brown syn. (limited eye elevation in adduction)
Crouzon syn. (facial distortion, exophthalmos)
See also Diplopia

Strabismus
See Squint

Swinging Torch Sign
Exp: In a darkened room, shine a torch in normal eye and there is immediate bilateral constriction of pupils. Swinging torch to affected eye causes initial bilateral dilatation with subsequent constriction. Swinging torch back to normal eye results in immediate pupil constriction again
Int: (+) Glaucoma (more advanced in affected eye), optic nerve demyelination, tumour or disease
Phys: As the light shifts from the less to the more diseased eye, the direct afferent stimulus passes along the more damaged optic nerve; it is now no longer sufficient in intensity to keep the pupils as small as they had been when the better eye was illuminated, both pupils thus dilate

Tears
See Lacrimation, Abnormal

Thyroid Glitter
Exp: ‘Glitter’ of light in the eyes
Int: (+) Thyrotoxicosis
Phys: Conjunctival oedema

Trichiasis
Exp: Ingrown eyelash causes irritation of conjunctiva
Int: (+) Trachoma, eyelid trauma
See also Entropion

Uveitis
See Eye, Inflamed or Red

Vision, Black Spots in Field of
Migraine (headache, nausea, photophobia)
Vitreous floater
Epilepsy (convulsions, absences)
Detached retina (fixed spot)
Cataract (small central opacity)
Optic nerve tumour
Poor cerebral blood flow (eg. before faint)
Noticing blind spot in visual field
A RATIONAL FOR THE EYES

Vision, Blurred

Ophthalmic
- Refractive error
- Cataract (progressive, lens opacity, painless)
- Retinal detachment (no pain or redness)
- Iritis (pain, red eye)
- Glaucoma (pain, red eye, small pupil)
- Conjunctivitis (discharge, red eye, no pain)
- Corneal ulcer (discharge, pain)
- Optic vascular disease
- Arc or UV eye (pain, red)
- Herpetic keratitis (pain, photophobia)
- Optic neuritis (pain)

Other
- Diabetes mellitus (fatigue, polyuria, polydipsia)
- Cerebrovascular accident (unilateral, sudden)
- Anaphylaxis
- Migraine (headache)
- Uraemia (fatigue, headache, pruritus)
- Hypoparathyroidism (tetany, wheeze, convulsions)
- Phaeochromocytoma (sweating, hypertension, headache)
- Fuchs syn. (anterior uveitis, different coloured irises)
- Strachan syn. (amblyopia, neuropathy)
- Drugs (eg. atropine, cocaine, nicotine)
  See also Amblyopia; Vision, Loss of

Vision, Double

See Diplopia

Vision, Loss of

Total (may be total)
- Glaucoma (rapid, pain)
- Cerebrovascular accident (confusion, paralysis)
- Transient ischaemic attack (brief, variable)
- Temporal arteritis (headache, jaw claudication, malaise)
- Retinal detachment (sudden, unilateral)
- Optic neuritis (eg. in multiple sclerosis)
- Head trauma (eg. optic canal haematoma)
- Amaurosis fugax (sudden, painless, transient)
- Malignant hypertension (sudden, unilateral)
- Acute chorioretinitis
- Optic nerve compression
- Retinal migraine (unilateral, painless, sudden)
- Vitreous haemorrhage (sudden, flashes)
- Retinal or optic nerve vascular disease (eg. thrombosis)
- Epilepsy (seizure not necessarily obvious)

Partial (may be total)
- Welding flash burn (pain, red)
- Conjunctival ulcer or trauma
- Migraine (headache, photophobia, nausea)
- Cataract (gradual, opaque lens) (see separate entry)
- Diabetes mellitus (polyuria, polydipsia)
- Senile macular degeneration (gradual, elderly)
- Keratitis (pain, red eye)
- Uveitis (pain, photophobia)
- Hypotension (faint, lightheaded)
- Thyrotoxicosis (gradual onset, painless)
- Anaphylaxis
- Acute glomerulonephritis

Syndromes
- Behçet syn. (arthritis, mouth ulcers)
- Devic syn. (multiple sclerosis)
- Down syn. (typical facies, mental retardation)
- Hurler syn. (corneal clouding, dwarf, arthralgia)
- Lissencephaly syn. (seizures, corneal opacity)
- Lowe syn. (retarded, cataracts, epicanthal folds)
- Marfan syn. (lens dislocation, kyphoscoliosis)
- Nelson syn. (postadrenalectomy, field defects)
- Pseudoexfoliation syn. (glaucoma)
- Stargardt syn. (adolescent, inherited)
- Uhthoff Phenomenon (demyelinating optic neuritis, heat exposure)
- Vogt-Koyanagi-Harada syn. (uveitis, vertigo)
- Von Hippel-Lindau syn. (retinal hamartoma)
  See also Amblyopia; Hemianopia; Vision, Black Spots in Field of; Vision, Blurred

Visual Halos

See Halos, Visual

White Pupil

(Leucocoria)

See Pupil, White
Xanthelasma
(Xantheloma Palpebrarum)
Exp: Yellow-brown nodules in soft tissues around eye
Int: (+) Primary biliary cirrhosis, elderly, hyperlipidaemia, cholestasis

Xerophthalmia
See Eye, Dry
Section Three

OPHTHALMOLOGICAL CONDITIONS
OPHTHALMOLOGICAL CONDITIONS

Common diseases that cause eye symptoms

**ACTINIC CONJUNCTIVITIS**
See FLASH BURN TO EYE

**ADIE’S PUPIL**
See ANISCORIA; HOLMES-ADIE SYNDROME

**ALBINISM**
Albinism is an uncommon condition in which there is a total lack of pigment in the skin and eyes. The skin is white, regardless of the race of the parents, and the iris (coloured part of the eyes) is pink. Both eyes and skin are very susceptible to damage by sunlight. Albinism is a defect of genes that occurs from the moment of conception, but it is not inherited, and an albino person will usually have normally pigmented children. There is no treatment for the condition other than carefully protecting skin and eyes from the sun.

Albinism occurs in 1:36,000 Caucasians, 1:10,000 Negroes, and the highest incidence in the world is 1:200 in the Hopi Indians of Arizona.

Chediak-Higashi syndrome is an inherited condition that can pass to subsequent generations. It causes recurrent skin and lung infections, partial albinism and sometimes liver, spleen and lung damage.

**ALLERGIC CONJUNCTIVITIS**
Allergic (vernal or atopic) conjunctivitis is an allergy reaction involving the surface of the eye.
A RATIONALE FOR THE EYES

If a pollen, dust or other substance to which a person is allergic lands on the eye, an allergy reaction will occur. Allergic conjunctivitis is often associated with hay fever and often only occurs at certain times of the year.

The symptoms include redness, itching, blurred vision and watering of the eye. In severe cases the white of the eye may swell dramatically and balloon out between the eyelids. There may be a clear, stringy discharge from the eyes, as well as excessive tears, and if the lower eyelid is turned down it appears to be covered with a large number of tiny red bumps. Rarely, ulceration of the eye surface may occur.

Blood and skin tests can be undertaken to identify the responsible substance in some patients who are repeatedly affected.

It can be prevented by the regular use of sodium cromoglycate drops throughout the allergy time of year. Attacks can be treated by antihistamine tablets and eye drops such as levocabastine and olopatadine. Simple eye drops available over the counter from chemists and containing artery-constricting (vasoconstrictor) medications can be used in milder cases. Appropriate treatment usually settles the symptoms rapidly.

See also CONJUNCTIVITIS; THIRD EYELID OF MORGAN-DENNY

AMAUROSIS

Amaurosis is a loss of vision caused by a central disease not directly related to the eye. It includes damage to the optic nerve and brain (due to a tumour, abscess or stroke etc.), diabetes mellitus, kidney failure, poisons or even severe emotional shock.

See also AMAUROSIS FUGAX; LEBER CONGENITAL AMAUROSIS

AMAUROSIS FUGAX

Amaurosis fugax is a sudden, painless, temporary loss of vision due to a disruption to the blood supply to the optic nerve or brain. It may be associated with narrowing of the carotid artery in the neck, or a tiny blood clot in the arteries supplying the retina at the back of the eye.

See also AMAUROSIS

AMAUROTIC FAMILIAL IDIOCY

See TAY-SACHS DISEASE

AMD

AMD is an abbreviation used in medicine for age-related macular degeneration.

See also MACULAR DEGENERATION

ANIRIDIA

Aniridia is a rare inherited genetic eye developmental abnormality, which results in lack of the iris.
A RATIONALE FOR THE EYES

There are two subtypes, Miller syndrome (associated with Wilms tumour and subnormal mentality) and Gillespie syndrome (associated with intellectual disability and poor coordination). The WAGR syndrome is a variation on the Miller syndrome. Glaucoma is very common and clouding of the cornea (outer surface of the eye) and lens (cataract) may occur. The iris (coloured part of the eye) is missing and vision is poor. Both eyes usually affected and the defect is obvious on simple eye inspection. There is no cure and very poor vision usually occurs.

See also CATARACT; GLAUCOMA; WAGR SYNDROME

ANISCORIA

Aniscoria is the medical term to describe pupils in the eyes that are of different sizes. The cause may be physiological (a variation of normal affecting up to 20% of the population) or due to raised pressure in the skull (eg. after a head injury), an abnormal migraine, third cranial nerve paralysis, Holmes-Adie syndrome (ciliary muscle paralysis), Horner syndrome, a Pancoast tumour, a penetrating injury to the eye or medications (eg. atropine). It is always a significant sign and the cause must be determined.

See also HOLMES-ADIE SYNDROME; HORNER SYNDROME

APHAKIA

Aphakia is a lack of the lens in the eye, either due to surgery, injury or very rarely a birth defect. A primitive form of surgery for a cataract is to push the lens back into the eye and correct vision with powerful spectacles.

See also CATARACT

ASTIGMATISM

People with astigmatism have an uneven curve to the refractive surfaces at the front of the eye (ie. the lens and cornea), so that some parts of the vision are clear while other parts are blurred at the same time. The problem can be corrected by appropriately manufactured spectacles.

BACTERIAL CONJUNCTIVITIS

See CONJUNCTIVITIS

BEHÇET SYNDROME

Behçet syndrome is a serious condition of unknown cause that results in widespread apparently unconnected symptoms such as recurrent severe mouth and genital ulcers, inflammation of the eye, arthritis and brain abnormalities such as convulsions, mental disturbances, partial paralysis and brain inflammation. Other symptoms may include rashes (eg. erythema nodosum), skin ulcers, inflamed veins and blindness. Treatment is often unsatisfactory. Steroids and immune suppressant medications are used, but the condition usually follows a long course with spontaneous temporary remissions. It is often seriously disabling and sometimes fatal.

BELL’S PALSY

Facial muscles are controlled by the facial nerve, which comes out of a hole in the skull just below and in front of the ear. From there, it spreads like a fan across the face to each of the tiny muscles that control facial expressions. Inflammation of the nerve at the point where it
leaves the skull causes the facial muscles to stop working. The exact reason for this inflammation is unknown.

Patients with Bell’s palsy (idiopathic facial paralysis) experience a sudden paralysis of the facial muscles on one side only. They can no longer smile or close the eye properly. There may be some mild to moderate pain at the point where the nerve leaves the skull beside the ear, but this settles after a few days. There may also be a disturbance to taste sensation.

No treatment is necessary for most patients, but in the elderly, if the paralysis is total, or if there is severe pain, treatment with high doses of prednisone (a steroid) may be tried, provided it is started within five days of onset.

10% of patients are significantly affected long term by facial paralysis, but two thirds of patients recover completely within a few weeks with no treatment. Most of the others obtain almost complete recovery.

See also CROCODILE TEARS SYNDROME

BLEPHARITIS

Blepharitis is a common inflammatory condition of the eyelid edges caused by a bacterial infection, allergy or a reaction to an environmental factor.

Both eyelids become red, covered with scales, sore and itchy. In advanced cases the eyelashes may fall out, and ulcers form on the lid margins. Treatment involves cleaning away the scales several times a day with moist cotton wool which may be dipped in baby shampoo, and applying antibiotic ointment to the affected areas of the eyelids. It is often difficult to cure and often recurrent.

The Greek word for eyelid is blepharon.
BLEPHAROEDEMA
Blepharooedema is a swollen eyelid due to allergy, injury, infection or excess fluid.

BLOCKED TEAR DUCT
See CONJUNCTIVITIS

BROWN SYNDROME
The Brown syndrome is a rare abnormality of eye movement from damage to the tiny muscles that move the eye. It is usually a congenital condition, but may be acquired due to damage to the eye and/or face during surgery, injury, inflammation or infection.

The affected eye is unable to move normally in all directions, and the patient appears to have a variable squint. The head may be held abnormally (tilted to side and with chin up) to compensate and vision in one eye may be suppressed. It is diagnosed by an ophthalmologist (eye specialist) after carefully examining all eye movements.

Surgery is the usual treatment, but in some acquired cases, steroids may assist by reducing inflammation. Treatment is reasonably effective, otherwise the condition persists lifelong.

See also SQUINT

BUPHTHALMOS
See GLAUCOMA
CHALAZION

A chalazion is a bacterial infection of an oil gland (Meibomian gland) deep within an eyelid due to blockage of the duct draining the gland. Patients develop a painful, red, tender swelling in an eyelid.

Treatment involves antibiotic eye ointment, and sometimes either cutting open or cutting out the infected gland. The condition may be recurrent, and rarely infection can spread further into the eyelid, but there is a good response to treatment.

See also MEIBOMIAN CYST

CLUSTER HEADACHE

A cluster headache is a severe, intermittent one-sided headache that occurs in clusters lasting from days to weeks. Attacks may be triggered by alcohol, stress, exercise, certain foods and glare. They are more common in middle-aged men.

Patients experience severe, one-sided pain around the eye that occurs daily for weeks and then subsides, only to flare again months later. The pain may be quite disabling, and are often accompanied by a congested nostril on the same side as the headache, a watery red eye and weakness on the affected side of the face. Unfortunately, there are no specific diagnostic tests available, and the diagnosis rests on the clinical acumen of the doctor.

Once present, these headaches are very difficult to control. Normally it is a matter of trial and error to determine the most effective treatment regime in any individual. The inhalation of pure oxygen may settle an otherwise intractable attack in a few minutes. Prevention is far
A RATIONALE FOR THE EYES

better than cure, and medications such as propranolol, ergotamine, lithium and amitriptyline can be used on a regular basis to prevent further attacks. In severe cases prednisone is prescribed.

COCKAYNE SYNDROME

The very rare Cockayne syndrome is a congenital condition characterised by dwarfism, blindness from failure of the retina to develop, deafness, intellectual disability, light sensitive skin and jaw abnormalities.

COGAN MICROCYSTIC CORNEAL DYSTROPHY

See CORNEAL DYSTROPHY

COLOBOMA

A coloboma is a congenital defect in the iris (coloured part of the eye) so that it has a cleft or hole in it. They almost invariably occur in the lower part of the iris unless caused by an injury to the eye.

COLOUR BLIND

Colour blindness is an inherited condition that nearly always occurs in men, and is an inability to differentiate between colours, usually red and green. Rarely all colour vision is lost with the patient seeing only black and white.

Ishihara test-cards covered in coloured dots, with numbers hidden amongst the dots, are used to diagnose the form of colour blindness.

No treatment or cure is available. Usually it is merely a nuisance, and most patients live happily with the problem, adapting so completely that they are not aware of its existence. Those planning a to work as an electrician or commercial pilot, or in some other areas where colour differentiation is vital, may not be allowed to undertake these careers.

CONJUNCTIVITIS

Conjunctivitis is an inflammation of the outer surface (cornea) of the eye, due to an allergy, or a viral or bacterial infection.

A bacterial conjunctivitis is the most common form, and is due to bacteria infecting the thin film of tears that covers the eye. It is very easily passed from one person to another (eg. a patient rubs their eyes with a hand, then shakes hands, and the second person then rubs their eyes). Babies suffering from a blocked tear duct may have recurrent infections. Tears are produced in the lacrimal gland beyond the outer edge of the eye, move across the eye surface and then through a tiny tube at the inner edge of the eye that leads to the nose. If the duct is too small in an infant, or is blocked by pus or phlegm, the circulation of tears is prevented and infection results.
A RATIONALE FOR THE EYES

Any one or more of a number of viruses may infect the cornea to cause conjunctivitis. This form is not quite as easily transmitted as bacterial conjunctivitis.

Bacterial conjunctivitis causes the formation of yellow or green pus in the eyes, which may stick the eyelids together. The eyes are bloodshot and sore, and almost invariably the infection involves both eyes. If allowed to persist, it may cause scarring of the eye surface and a deterioration in sight.

Viral conjunctivitis causes slight pain or an itch, redness of the eye and often a clear sticky exudate.

Rarely, resistant infections make it necessary to take a swab from the eye to determine the exact bacteria or virus responsible, but in most cases, no investigations are necessary.

Bacterial conjunctivitis is easily treated with antibiotic drops or ointment on a regular basis until the infection clears, usually in two to four days. Children must be excluded from school until all eye discharge has ceased. A blocked tear duct may be probed and cleared if conjunctivitis persists in a baby for several months, but most grow out of the problem.

Viral conjunctivitis is the more difficult form to treat as there is no cure for most viral infections, but Herpes virus infections can be cured by antiviral drops. Soothing drops and ointment may be used, but time is the main treatment, and the infection may persist for several weeks until the body’s own defences overcome it.

Allergic (vernal) conjunctivitis is a reaction on the surface of the eye to a pollen, dust, chemical or substance to which the patient has an allergy. The eye becomes red, itchy and watery. Vasoconstrictor or antihistamine eye drops can be used to control the condition.

See also ALLERGIC CONJUNCTIVITIS; TRACHOMA

CONRAD SYNDROME

The Conrad syndrome is a congenital developmental abnormality. It varied symptoms include cataracts in the eyes, limb contractures, deafness and intellectual disability. Abnormal
A RATIONALE FOR THE EYES

long bone ends (stippled epiphyses) are seen on x-ray. Surgery can be performed for the cataract and limb contractures. Although there is no cure, life expectancy is reasonable. See also CATARACT

CORNEAL DYSTROPHY

Corneal dystrophy is a degeneration of the cornea, the clear dome over the pupil and iris (coloured part of the eye), with formation of dots, cysts, cracks or other deformities in the normally clear membrane. Many different subtypes are known (eg. Cogan microcystic dystrophy, Reis-Bücklers dystrophy, Meesmann dystrophy, Schnyder dystrophy).

It is usually an inherited characteristic and causes deterioration of vision due to damage to, and distortion of, the cornea. Significant deterioration in vision may occur. The eye is examined with a magnifying light (ophthalmoscope) to see corneal damage.

Treatment involves surgical replacement of the cornea with a transplanted cornea, or laser destruction of abnormalities in the cornea. Good results are usually obtained from treatment but the disease may recur in a transplanted cornea. Progressive sight deterioration occurs without treatment.

CORNEAL ULCER

See EYE ULCER

CROCODILE TEARS SYNDROME

The crocodile tears syndrome is a complication of Bell’s palsy. It is possibly due to regenerating nerves that normally control salivary glands being misdirected to the tear gland during recovery from Bell's palsy. The only symptom is tears pour from the affected eye when eating. Surgically cutting the responsible abnormal nerve fibre gives good relief.

See also BELL’S PALSY

CROUZON SYNDROME

The Crouzon syndrome is a familial (runs in families) developmental abnormality of the face. Those affected have abnormal growth and shape of their face and skull, protruding eyes, a squint and loss of vision in one or both eyes. From the side, the face appears flattened. A skull X-ray is abnormal. There is no cure, but reasonable results are obtained from major facial surgery, depending on the severity of deformities.

CYSTINOSIS

Cystinosis is a very rare familial (runs in families) disorder that results in the deposition of crystals of cystine in the eyes, bone marrow, lymph nodes, white blood cells, kidneys and other organs. Two forms of the disease are known - infantile which is widespread, and adult which affects only the eyes.

Affected infants are very small, feverish, vomit constantly, pass excess urine and become dehydrated. Adults have eye pain, intolerance to bright light and headaches. Kidney failure may be a late complication in adults.

The diagnosis is difficult and is confirmed by seeing crystals in white blood cells when a blood sample is examined under a microscope.

There is no effective treatment and death from kidney failure before the age of ten is usual with the infant form. Some reduction in life expectancy occurs with the adult form.
DACROCYSTITIS
An infection of the tear (lacrimal) gland is called dacrocystitis. It is far more common in babies than adults and usually only occurs on one side. Patients have watery eyes with a painful swelling above the outer part of the upper eyelid, where the lacrimal gland is located. It is treated with antibiotics given by mouth.

DIABETIC RETINOPATHY
Damage to the light sensitive cells in the retina at the back of the eye from poorly controlled diabetes is called diabetic retinopathy. It is caused by damage to the tiny capillaries supplying blood to the retina. The abnormality can be detected by examining the retina with a magnifying lens (ophthalmoscope). Patchy blindness occurs, which cannot be reversed, but further damage to the retina can be prevented by laser coagulation of damaged arteries to prevent them from bleeding. Untreated, the condition progresses to total blindness. Good control of diabetes prevents the condition in most patients.

DRUSEN
Drusen is a degenerative condition of the retinal macular (central area of light sensitive cells at the back of the eye) in late middle age and the elderly. Patients experience steadily worsening central vision, while edge (peripheral) vision is often normal.

The condition is diagnosed by examining the eye through a magnifying light (ophthalmoscope) and injecting a bright dye (fluorescein) into the artery supplying the eye.

No treatment is available, and the condition is slowly progressive.
See also MACULAR DEGENERATION

DRY EYE SYNDROME
The dry eye syndrome (xerophthalmia) is a very common problem, with the incidence increasing with age, and is due to reduced tear production. Patients experience dry scratchy irritated eyes.

A small piece of blotting paper placed under lower eyelid remains dry (Schirmer tear test) in patients with the syndrome.

Treatment involves artificial tear drops or ointment, lubricating inserts under lower lids, and surgical blockage of tear duct at inner corner of eye to prevent tear drainage. Complications include eye ulcers and infections. There is no cure, but it can be reasonably controlled with treatment. Severe forms of the syndrome are known as keratoconjunctivitis sicca.

See also KERATOCONJUNCTIVITIS SICCA

DUANE SYNDROME
Duane syndrome is a rare congenital abnormality of nerve supply to eye muscles due to absence of the 6th nerve, which supplies some eye movement muscles. Vision remains
A RATIONALE FOR THE EYES

normal. Patients are unable to move the eye horizontal resulting in a variable squint. There is no cure, but the severity of squint may be reduced by surgery.

DUBOWITZ SYNDROME

A rare developmental abnormality, that is familial, but both parents must be carriers. The Dubowitz syndrome is characterised by a low birth weight baby with drooping eyelids (ptosis), small jaw, sparse hair, short stature, mild intellectual disability and eczema. No treatment is available, and there is no cure.

ECTROPION

Ectropion is the out turning of the lower eyelid, due to slackness of tissue in the eyelid with ageing or injury. The tears cannot be retained in the eye and trickle down the cheek, and there is an increased risk of eye surface infection or ulceration. Surgery can be performed to tighten up the lower lid.

EPISCLERITIS

Episcleritis is a common inflammation of the outer surface of the eye in young adults that causes discomfort, tenderness and redness of one section of the white of the eye. No treatment is usually necessary, as most cases settle without treatment in one or two weeks, but persistent cases may become lumpy and thickened and require nonsteroidal anti-inflammatory tablets or steroid eye drops.
A RATIONALE FOR THE EYES

EYE FLOATER
See FLOATER IN EYE

FLASH BURN TO EYE
A flash burn (actinic conjunctivitis) is a superficial burn to the surface of the eye (cornea) caused by looking at a welding arc or ultraviolet light for a prolonged time. The eye is very painful, red and sometimes swollen, but the pain may not develop until 6 to 12 hours after exposure. Severe or recurrent eye burns can cause scarring and blindness that can only be corrected by a corneal transplant.

Patients are prescribed eye drops and painkilling tablets, and it is necessary to cover the eye until it has recovered, and most settle completely within 24 hours. The only effective first-aid measure is a cold, wet compress.

See also KERATITIS

FLOATER IN EYE
A floater is a collection of cells or protein in the thick vitreous fluid that fills the eyeball, which casts a shadow on the light-sensitive retina at the back of the eye. The floater forms because of bleeding into the eye, a detached retina, posterior vitreous detachment, infection, or no cause may be found. Diabetes, leukaemia, high blood pressure, and rarer conditions may cause bleeding into the eye.

Patients notice a spot in the field of vision that may continue to move across the visual field after the moving eye comes to rest - thus the name floater. Because a serious condition may be responsible, all patients with floaters must be investigated to exclude any disease. A detached retina can be repaired by a laser in the early stages, but if left, may cause permanent blindness.

They are only treated if causing significant trouble, but if necessary, a laser can destroy the floater while a doctor uses a microscope to look into the eye. Most floaters dissipate with time.

See also POSTERIOR VITREOUS DETACHMENT; RETINAL DETACHMENT

FLOPPY EYELID SYNDROME
The floppy eyelid syndrome is a persistent drooping of the upper eyelid. Obesity and the deposition of excess fat in the eyelid structures are the usual causes.

There is an inability to fully open or close the eyelid, an inflamed red eye, and a spontaneous turning out of the upper eyelid during sleep. Ulceration of the eye surface may be a complication. Treatment involves weight loss, taping eyelid shut at night, and surgery to remove fat from the eyelid.

The prognosis is often poor, as most patients do not lose sufficient weight, and the eye problem becomes chronic.

FUCHS HETEROCHROMIC CYCLITIS
See FUCHS UVEITIS SYNDROME

FUCHS UVEITIS SYNDROME
Fuchs uveitis syndrome (also known as Fuchs heterochromic cyclitis) is a gradually worsening form of iritis (inflammation of the iris - coloured part of the eye) that usually affects young adults and only one eye. The cause is a degeneration of the cells responsible for colour in the iris that may be congenital.
A RATIONALE FOR THE EYES

The irises gradually become different colours and the vision is irregularly blurred. Cataracts and glaucoma may subsequently occur. No treatment is available except for the cataracts and glaucoma.

See also CATARACT; GLAUCOMA; UVEITIS

GILLESPIE SYNDROME
See ANIRIDIA

GLAUCOMA
Glaucoma is an increase in the pressure of the half-set jelly-like fluid inside the eyeball that damages the eye and affects the vision. The eye is filled with a thick clear fluid (aqueous humour) that is slowly secreted by special cells within the eye, while in another part of the eye the fluid is removed, allowing a slow but steady renewal. If there is a blockage to the drainage of the fluid from the eye while new fluid continues to be secreted, the pressure inside the eye increases, and damage occurs to the light-sensitive retina at the back of the eye. Other conditions may also cause glaucoma including eye tumours, infections, injury, and in rare cases drugs (eg. steroids) may be responsible.

Three types of glaucoma occur - chronic, acute and congenital:
- Chronic glaucoma (open-angle glaucoma) is the most common type with a slow onset over years. It usually occurs in both eyes simultaneously and runs in families. Initially it affects the peripheral vision, which is how far can be seen to the sides and up and down while looking straight ahead, and patients develop tunnel vision. One in every 75 people over 40 years have this type of glaucoma.
- Acute glaucoma (angle-closure glaucoma) is the worst type, as it develops in a few hours or days, but usually involves only one eye. There is a rapid deterioration in vision, severe pain, rainbow-coloured halos around lights, nausea and vomiting. It may start after a blow to the eye, or for no discernible reason. Immediate treatment of acute glaucoma is essential if the sight of the eye is to be saved, but even with good treatment, permanent blindness can occur.
- Congenital glaucoma (buphthalmos) occurs in babies who are born with the condition. The earliest sign is the continual overflow of tears from the eye, and the baby turns away from lights rather than towards them as a normal.

Glaucoma is diagnosed in most cases by measuring the pressure of the fluid within the eye. This can be done by anaesthetising the eye surface with eye drops and then resting a pressure measuring instrument (tonometer) on the surface of the eye while the patient is lying down, or by using a machine that directs a puff of air onto the eye to measure the pressure. Glaucoma may also be detected by measuring deterioration in peripheral vision using a computerised device, charts or by following a white dot on a large black screen. More complex tests include examining the eye through a microscope to determine the nature and seriousness of the glaucoma.

The excessive pressure in the eye caused by glaucoma can be reduced by eye drops, which are usually beta-blockers, and/or tablets that remove some fluid from the eye. Apraclonidine may be used in severe cases.

Beta-blocker eye drops include betaxolol (Betoptic), bimatoprost (Lumigan), carteolol, latanoprost (Xalantan), levobunolol (Betagan), metipranolol and timolol. Their side effects may include blurred vision, headache and a small pupil. They should be used with caution in asthma and heart disease. The other commonly used eye drops for glaucoma are
brimonidine, brinzolamide, carbachol pilocarpine (Pilopt), dipivefrine, dorzolamide and travoprost.

The tablet used to treat glaucoma is acetazolamide (Diamox). Side effects may include pins and needles, excess urination and a poor appetite. It must be used with caution in pregnancy, and not in patients with liver disease.

In serious cases, laser microsurgery to the tiny drainage canals in the front of the eye is necessary. Congenital glaucoma always requires surgical treatment.

Without treatment, glaucoma progresses inexorably to total blindness, but if the disease is detected early, glaucoma in most patients can be successfully controlled but not cured.

See also OPTIC ATROPHY

GRADENIGO SYNDROME
The Gradenigo syndrome (also known as petrositis) is a bony abnormality of the skull that causes nerve compression.

Infection or a tumour of the petrous bone in the skull damages the 6th cranial nerve (abducent nerve) to cause a headache, double vision, facial pain and a middle ear infection. An infection may spread to the brain, ear or other areas. A skull X-ray or CT scan shows the abnormal bone.

Treatment involves potent antibiotics and surgery on the skull to drain any bone abscess or remove a tumour. The prognosis is good if diagnosed early, but permanent nerve damage may result.

HEERFORDT SYNDROME
Heerfordt syndrome (also known as Heerfordt-Waldenström syndrome and uveoparotid fever) is a complication of sarcoidosis involving the face, eyes and salivary glands. The cause is unknown.

The symptoms include painful inflammation of the salivary glands around the jaw, partial paralysis of one side of the face, inflammation of the eye (iritis) and sarcoidosis.

No treatment is necessary except for the sarcoidosis as the syndrome is a self-limiting condition that settles with time and rest, but the sarcoidosis may have serious consequences.

It is named after the Danish optician Christian Heerfordt (1872-1953).
stimulation and tendon reflexes in the arms and legs may be slower than normal. Patients may be adversely affected by bright light, or find it difficult to see in dim light. It is a harmless condition and no treatment is necessary.

See also ANISCORIA

HORNER SYNDROME
Horner syndrome is a bizarre combination of symptoms involving the eye and sweat glands caused by compression of a special network of nerves (autonomic nervous system) in the chest due to lung cancer or pneumothorax, or in the brain due to a tumour. It may be the first sign of a quite advanced lung cancer. The syndrome is characterised by a drooping eyelid, contracted pupil and a sunken eye, associated with reduced sweating.

Numerous investigations must be undertaken to find the cause, including x-rays, CT and MRI scans. It is necessary to correct the underlying cause of the nerve compression, usually by surgery.

It is named after the Swiss ophthalmologist Johann Horner (1831-1866).

IRITIS
Iritis is inflammation of the iris, the coloured part of the eye, for which there is often no obvious cause. The symptoms may include pain, excess tears, reduced vision, contracted pupil and dislike of bright lights. Permanent scarring may occur if treatment is neglected resulting in a permanently small and irregular pupil and cloudiness over the pupil.

See also UVEITIS

KAYSER-FLEISCHER RING
The Kayser-Fleischer ring is a greyish red to gold coloured ring around the outer edge of the cornea in the eye. It is caused by Wilson disease, an abnormality due to excess copper in the body.

See also WILSON DISEASE
KEARNS-SAYRE SYNDROME
Kearns-Sayre syndrome is a rare inherited cause of blindness caused by a metabolic abnormality transmitted through women from one generation to the next. Patients experience a rapid loss of central vision in both eyes in early adult life, gradual progressive paralysis of eye movement, pigmentation of the retina (light sensitive area at the back of the eye), and abnormal conduction of nerve signals in the heart. It is diagnosed by examining the retina with a magnifying light (ophthalmoscope). No treatment is available.

See also LEBER OPTIC NEUROPATHY

KERATITIS
Keratitis is an inflammation of the cornea (the outer layer of the eye), caused by ultraviolet lights, exposure to reflected sun (snow blindness), cement dust, and irritating chemicals (eg. alkalis) that may splash into the eye. It may also be due to a bacterial or fungal infection in contact lens wearers.

Severe burning pain starts in the eye some hours after exposure to bright light, but immediately with direct irritants. The eyes water and are very sensitive to bright light and any contraction or dilation of the pupil due to changes in light intensity causes pain. Scarring of the cornea may occur with chemical burns. Examination of the eye under magnification shows damage to cornea.
A RATIONALE FOR THE EYES

Treatment involves irrigating the eye to remove any irritant chemicals or substances, instilling drops to paralyse any pupil movement, and an eye patch. Recovery usually occurs within 24 to 48 hours.

See also FLASH BURN TO EYE; OPHTHALMIA

KERATOCONJUNCTIVITIS SICCA
Keratoconjunctivitis sicca is a severe form of the dry eye syndrome (xerophthalmia) that commonly occurs in elderly women whose lacrimal (tear producing) glands fail. It may also a complication of autoimmune diseases such as systemic lupus erythematosus, rheumatoid arthritis and Sjögren syndrome. Patients have continued eye irritation and discomfort from a dry eye surface, and rarely it may be a threat to sight.

The diagnosis can be confirmed by a simple test in which a thin strip of blotting paper is touched to the lower eye surface for a minute, and it remains abnormally dry with this condition.

Very regular and long-term use of lubricating drops and ointment in the eye are the main treatments. Inserts that are placed under the lower lid and ooze a lubricant constantly for many hours can also be used. Lodoxamide drops are another solution in some patients. There is no cure, it is difficult to control, and usually continues lifelong.

See also DRY EYE SYNDROME

KERATOCONUS
Keratoconus is thinning and a cone shaped protrusion of the cornea, which is the clear dome over the pupil and iris (coloured part of the eye). It is a condition that may be inherited or associated with Down syndrome, Turner syndrome, Marfan syndrome and numerous eye diseases, and causes distorted vision (astigmatism) in the affected eye that steadily worsens to the point where scarring or rupture of the clear corneal membrane may occur. It is diagnosed by examining the eye with a magnifying light (ophthalmoscope).

In early stages treatment merely requires spectacles or contact lenses to correct the vision, but in late stages, surgery to the surface of the cornea or replacement of the cornea with a transplant is necessary.

Patients get good results from treatment, but it is a progressive condition if left untreated.

KERATOMALACIA
Keratomalacia is ulceration and dryness of the cornea (front clear layer of the eye) due to a severe vitamin A deficiency. Early symptoms include night blindness, swollen itchy eyelids, avoidance of bright lights and a painful eye surface. It may progress to corneal softening, scarring and permanent blindness. Vitamin A supplements correct the condition.

KYASANUR FOREST DISEASE
Kyasanur Forest disease is characterised by headache, fever, muscle pains, cough, belly pains, eye pains and avoidance of bright lights. It is caused by a virus that is transmitted from small mammals to humans by the bite of a tick, and is found in southwest India. A vaccine is available.

LAURENCE-MOON-BIEDL SYNDROME
The Laurence-Moon-Biedl syndrome is an inherited condition of the eye, brain and genitals that causes night blindness due to excessive amounts of pigment in the retina at the back of
A RATIONALE FOR THE EYES

the eye, obesity, intellectual disability, extra fingers and toes and underdeveloped genitals. Examination of the back of the eye through an ophthalmoscope (magnifying light) will show the excess pigment. There is no treatment available and no cure.

See also RETINITIS PIGMENTOSA

LEBER CONGENITAL AMAUROSIS
Leber congenital amaurosis is a rare familial (runs in families) cause of blindness in children, but both parents must be carriers. Blindness occurs at birth or develops soon afterwards. There is no pupil contraction when bright light shone into eye, and may be associated with intellectual disability, deafness, epilepsy, kidney and bone abnormalities. No treatment or cure is available.

See also LEBER OPTIC NEUROPATHY

LEBER OPTIC NEUROPATHY
Leber optic neuropathy is a rare cause of blindness due to inflammation of the optic (eye) nerve. The cause is an inherited metabolic abnormality transmitted through women from one generation to the next. Patients develop a rapid loss of central vision of both eyes in early adult life. It is diagnosed by examining the light sensitive retina at the back of the eye with a magnifying light (ophthalmoscope). No treatment is available and the usual result is permanent vision loss, but 15% have a spontaneous recovery.

See also KEARNS-SAYRE SYNDROME; LEBER CONGENITAL AMAUROSIS

LEPTOSPIROSIS
Leptospirosis is a bacterial infection of the liver and other organs caught from infected cattle and pigs by abattoir workers, veterinarians and farmers. In third-world countries, dogs and rats may also be carriers. The spirochete bacteria *Leptospira interrogans* enter through minor abrasions or by being swallowed. The incubation period varies from three days to three weeks.

Patients develop a sudden high fever, headache, stomach pain, muscle aches and inflamed eyes. After a couple of days, these symptoms disappear, and the second stage of the infection commences which lasts for one to four weeks, and the patient complains of swollen lymph nodes, a generalised rash, eye pain, and in severe cases yellowing of the skin (jaundice). The second stage may cause permanent liver damage and Weil Syndrome. The diagnosis is confirmed by a specific immunoglobulin found on a blood test.

Antibiotics such as penicillin are prescribed as treatment, but sometimes they have remarkably little effect. Careful nursing is important. The disease can usually be prevented by taking a doxycycline antibiotic tablet once a week.

It is usually cured by correct treatment, but if jaundice develops, the death rate may be as high as 10%.

Leptospirosis is thought to be the oldest disease caused by civilisation as it probably first affected humans at the same time that they domesticated cattle and pigs about 5000 BC.

LISSENCEPHALY SYNDROME
The lissencephaly syndrome is a rare brain and developmental disorder. It is an inherited condition, but only if both parents are carriers of a defective gene. Epileptic seizures, poor muscle tone, jaundice, cataracts in the eye, and wrinkled forehead skin are the symptoms. No
A RATIONALE FOR THE EYES

Treatment is available, and seizures are difficult to control with normal epilepsy medication. An early death is normal.

See also CATARACT

LOIASIS

Loiasis is a disease of tropical central and West Africa that is caused by a filarial parasitic worm *Loa loa*. The larva of the worm is transmitted from one person to another by the deer fly. It enters the tissue under the skin and matures there and grows to between 25 and 70 mm. in length. It may migrate through the subcutaneous tissue for decades causing tender lumps known as Calabar swellings. The most significant symptoms occur when the worm migrates to the eye.

The diagnosis can be confirmed by a biopsy of a Calabar swelling to find the worm, or a blood test that detects the larvae. Allergy reactions to the worm are common and are treated with prednisone tablets. The infestation is cured by repeated doses of diethylcarbamazine.

LØKEN-SENIOR SYNDROME

The Løken-Senior syndrome is a rare inherited disorder that causes progressive deterioration in kidney function that leads to kidney failure, and progressive eye damage that results in blindness. There is a rapidly progressive juvenile form that has its onset at about 12 months of age that can also cause mental retardation and high blood pressure, and a slower developing adult form. The only treatment available is a kidney transplant.

LONG SIGHTED

Long sightedness (hyperopia) is a defect in visual acuity that is usually present from birth. The eyeball is too short, and close objects cannot be focused precisely on the retina (light sensitive cells) at the back of the eye. Distant objects can be seen clearly, while close objects are blurred.

It is diagnosed by refractive tests using a number of different lenses until near objects can be seen as clearly as possible. Appropriate spectacles are then prescribed, and must be worn when reading or looking at close objects. Children may grow out of the problem at puberty, otherwise it is a lifelong problem.

See also ASTIGMATISM; PRESBYOPIA

LOUIS-BAR SYNDROME

The Louis-Bar syndrome is a rare form of rapidly progressive brain deterioration due to degeneration of the cerebellum (lower back portion of brain) and spinal cord. The symptoms include dilated capillaries on the whites of the eyes, the face and areas of skin flexion (eg. arm pit, behind knee); intellectual disability; recurrent infections of lungs and ears, and poor coordination that steadily worsens. Late symptoms include twitching movements of the eyes and abnormal writhing movements of the arms and legs. There is an above average incidence of cancer.

Numerous blood tests are abnormal, including very low immunoglobulin levels. No treatment is available and death in teenage years is usual.
LOWE SYNDROME

The Lowe syndrome (oculocerebrorenal syndrome of Lowe) is a rare, inherited, body chemistry disorder that is passed to males only through the female side of the family. These boys have intellectual disability, eye cataracts, clouding of the cornea (outer surface of eye), abnormal skin folds beside the eyes, and abnormal eye socket shape. Some patients have rickets and Fanconi syndrome. Abnormal levels of amino acids (protein breakdown products) found in urine. There is no cure, but they have a reasonable life span.

MACULAR DEGENERATION

Macular degeneration is a common form of vision deterioration in the elderly (age-related macular degeneration - AMD). The macular is the part of the retina at the back of the eye that is most sensitive to light. It degenerates because of a poor blood supply with advancing age, cholesterol build ups in arteries and diabetes, and patients experience a gradual loss of central vision while peripheral vision may be normal.

There are two types of AMD - dry and wet. The dry is a milder early stage of the disease, which causes patchy loss of small areas of the visual field. It often has a minimal effect on the person’s ability to see and function. The wet form occurs with an overgrowth of blood vessels that leak and cause wider and more rapidly progressive damage to the visual field. This form may cause loss of sharpness of images, distortion of images, a loss of colour differentiation and multiple blind spots in the vision.
A RATIONALE FOR THE EYES

There specific cause is unknown but the incidence is higher in smokers and there is probably a genetic tendency for the development of the condition.

It is diagnosed by examining the eye through an ophthalmoscope (magnifying light). There is no effective treatment for the dry form, although the deterioration in vision is usually very slow. The wet form progresses faster, but the damage may be slowed by laser destruction of the rapidly spreading blood vessels. A newer treatment is the injection of the substance Visudyne, which concentrates in the abnormal blood vessels and is activated by a specific light frequency shone into the eye to almost instantly destroy the abnormal blood vessels by coagulation. A number of other new treatments are under investigation and may lead to better management of the condition in the future.

See also DRUSEN

MACULAR DYSTROPHY

Macular dystrophy is any form of degeneration of the macular, the most light sensitive area in the retina at the back of the eye. The degeneration may be due to a genetic problem present from birth, but which does not become a problem until middle life when additional blood vessels grow into the macular, which bleed and damage it.

MARFAN SYNDROME

Marfan syndrome is an uncommon inherited condition that affects the skeleton, heart and eye, and occurs in all races but only in one out of every 20,000 people.

Its characteristics include very long thin bones in the arms, legs, fingers and toes (arachnodactyly), a tall skull, excessive joint movement, a high foot arch and a humped back. Half the patients have an eye lens that is in the wrong position, and they may develop keratoconus (protruding eye surface) and a detached retina (the light-sensitive area at the back of the eyes), which results in partial or total blindness. An abnormality in the elastic tissue of the heart valves and major arteries causes these to fail and the pumping of the heart to be inefficient. The main artery of the body, the aorta, becomes overly dilated and distorted and may eventually rupture, and heart infections (endocarditis) are common. Most patients do not have all these symptoms, as there is great variation between them. Some may be totally unaware that they are affected and just appear to be very tall and thin.

It is diagnosed by the characteristic appearance of the long bones on X-ray, and by assessing the heart abnormalities with echocardiograms. The problems in the heart and aorta are controlled and corrected by both medication and surgery, but death in middle age is common unless corrective surgery is successful.

See also KERATOCONUS; RETINAL DETACHMENT

MARINESCO-SJÖGREN SYNDROME

The Marinesco-Sjögren syndrome is a rare progressive degeneration of the cerebellum (lower back part of brain) that starts in early childhood. It is a familial (inherited) condition, but both parents must be carriers. Sufferers have poor coordination (ataxia), eye cataracts, intellectual disability, multiple bony abnormalities and underdeveloped testes or ovaries. Medication can be used to control ataxia, but there is no cure, and the symptoms steadily worsen to death.

MAROTEAUX-LAMY SYNDROME
A RATIONAL FOR THE EYES

The Maroteaux-Lamy syndrome is a congenital metabolic (body chemistry) disorder characterised by abnormal bone and heart formation, clouding of the cornea in the eyes, deafness, short body and neck from retarded growth, walk with a waddling gait, knock knees and flat feet. It is diagnosed by abnormal specific blood enzyme tests, and may be detected before birth by chorionic villus (placenta) sampling. No treatment is effective and survival beyond 40 years unusual.

MEESMANN CORNEAL DYSTROPHY
See CORNEAL DYSTROPHY

MEIBOMIAN CYST
The upper and lower eyelids each contain about twenty Meibomian glands, which secrete an oily substance that lubricates the surface of the eye. If the tiny tube leading out of one a gland becomes blocked, it will swell up into a cyst that is felt and seen as a lump in the eyelid. It may become infected by bacteria to form a chalazion. The problem is more common in those over 40 years of age, and may follow a period of eye irritation or conjunctivitis. A small cut into the cyst will drain out the contents.

See also CHALAZION

MIKULICZ DISEASE
Mikulicz disease (benign lymphoepithelial condition) is an inflammatory condition of salivary and tear glands, that has no known cause but is common in Scandinavia.

The symptoms are enlargement of the saliva producing glands under and behind the jaw, and enlargement of the lacrimal (tear) glands at the outer corners of the eyes. It may also be associated with tuberculosis, sarcoidosis, syphilis, actinomycosis, wasting of the mucous producing glands in the nose, throat and vagina, and dry eyes (keratoconjunctivitis sicca). Recurrent salivary gland infections may occur. Specific blood tests and biopsy of the affected glands are diagnostic.

No treatment is necessary unless a specific gland becomes painful when it can be surgical removed. No cure is available, but it is usually a benign condition.

See also SICCA SYNDROME
MÖBIUS SYNDROME
Möbius (Moebius) syndrome is a developmental abnormality of cranial nerves. Two nerves from the brain (numbers 6 and 7) fail to develop properly before birth. These children are born with drooping eyelids, inability to move their eyes normally, faces that cannot show expressions, difficulty in swallowing and speaking, constant drooling, but normal intelligence. There is no treatment or cure, but the lifespan is normal. Paul Möbius (1853-1902) was the German neurologist who first described the syndrome.

See MÖBIUS SYNDROME

MORGAN-DENNY EYELID
See THIRD EYELID OF MORGAN-DENNY

MORQUIO SYNDROME
Morquio syndrome is an abnormality of body metabolism that damages numerous organs, caused by excess accumulation of keratin sulfate in tissue. The symptoms include severe progressive bone damage, heart abnormalities and damage, cataracts in the eyes, deafness, retarded growth leading to a short neck and short trunk, flat feet and knock knees. Bones fracture easily, and heart damage may become significant. Specific blood enzyme tests are abnormal, allowing a diagnosis to be made. No treatment is available, but there is a normal life span expected in most patients. It is named after the Uruguayan physician, Luis Morquio (1867-1935).

See also CATARACT

MYASTHENIA GRAVIS
Myasthenia gravis is an uncommon condition characterised by varying weakness of the muscles that control the eyelids, the movement of the eyes and swallowing. Signals from the nerves that supply affected muscles are blocked, for which there may be an immunological cause when antibodies that normally fight off infection, actually attack nerve tissue (autoimmune response). It may occur at any age, but is most common in young women and
A RATIONALE FOR THE EYES

may be associated with rheumatoid arthritis, systemic lupus erythematosus, thymus and thyroid disease.

Drooping eyelids (ptosis), double vision and difficulty in swallowing are the main symptoms. In severe cases the muscles used in breathing and walking are also affected. Muscle weakness may vary in severity during the day and can disappear entirely for days or weeks before recurring, but over a period of months or years, the attacks become more severe. Unless adequate treatment is obtained, death eventually results from breathing difficulties.

The diagnosis is confirmed by the patient’s reaction to an anticholinergic drug, which immediately reverses all the muscle weakness or measurement of the anti-skeletal muscle antibodies or anti-acetylcholine receptor (Anti-AChR) antibody titre.

Treatment involves surgically removing the thymus gland, which is the source of most of the antibodies in the blood, and using anticholinesterase drugs (eg. distigmine, neostigmine) on a regular basis to control the muscle weakness. Steroids can be used in patients who respond poorly to other treatments.

There is no cure and patients require treatment for the rest of their lives, but some have lengthy periods when the disease is inactive, during which they may be able to cease their medication.

NAEVUS OF OTA

A naevus of Ota is a congenital (but not inherited) skin pigment disorder affecting Asians (particularly Japanese) and less commonly Negroes. They are rare in Caucasians. The condition usually starts in childhood or teenage years to cause a mottled, dusky blue/brown, disfiguring pigmentation of the face that may involve the inside the mouth and the whites of the eyes. Patients have an increased risk of malignant melanoma. The diagnosis can be confirmed by biopsy.

Laser treatment of the affected skin sometimes beneficial, but treatment is not particularly effective and there is no specific cure.

NEU-LEXOVA SYNDROME

The Neu-Lexova syndrome is a rare familial (runs in families) abnormality, but occurs only if both parents carry the abnormal gene. It is characterised by absent eyelids, underdeveloped nose and jaw, abnormal skin structure and texture, swelling of feet and hands, multiple skin contractures and intellectual disability. No treatment is available and the prognosis is poor.

NORRIE DISEASE

Norrie disease is a very rare congenital abnormality of the eye and brain caused by damage to the X chromosome. Patients have an abnormal growth on the retina at the back of the eye, cataracts in the lens of the eye, abnormalities of the iris (coloured part of the eye), hearing loss and intellectual disability. Specific chromosomal studies are diagnostic, but there is no specific treatment available.

OCHRONOSIS

Ochronosis is an inherited biochemical abnormality in which homogentisic acid accumulates to a high level in the body to cause arthritis, and bluish patches in the mouth, on the whites of the eyes, nose, ears, fingers, genitals and in the armpits. These blue to black patches may also be found within the body, and the urine is a darker than normal colour. There is no cure, but the arthritis can be controlled by appropriate medication.
A RATIONALE FOR THE EYES

**OCULAR LARVA MIGRANS**
See VISCERAL LARVA MIGRANS

**OCULOLOGYRIC CRISIS**
An oculogyric crisis is a rare complication of encephalitis (brain inflammation), Parkinson disease and some drugs (eg. the antiemetic prochlorperazine - Stemetil).

Patients develop a constant upward movement of the eyes, and painful extension of the neck so the head is tilted backwards as far as possible that last for minutes or hours. Very rarely it may be life threatening due to pressure on the throat from neck extension. Muscle spasms may cause tearing of muscles and persistent muscle pain that can take weeks to recover.

It is rapidly cured by injection of the drug benztropine into a vein.

**OCULOPHARYNGEAL MUSCULAR DYSTROPHY**
Oculopharyngeal muscular dystrophy is an inherited condition that may commence at any age. It is characterised by a very slowly progressive permanent weakness of the muscles that move the eye and enable swallowing. It is diagnosed by electrical studies of muscle action and a muscle biopsy. Choking, inhaling food and subsequent lung infections (eg. pneumonia) may be complications. There is no effective treatment and the diet may need to consist of thickened fluids.

**ONCHOCERCIASIS**
Onchocerciasis (river blindness) is caused by the nematode worm *Onchocerciasis volvulus*. The nematode is carried from one person to another by the bite of a small black fly that only lives along rivers. Larvae are deposited in the skin by the bite, mature after 6 to 36 months into adult worms, which are up to 60 cm. long and live tightly coiled under the skin. The adult worm releases tiny microfilariae into the blood, which spread throughout the body, particularly to the skin, eyes and lymph nodes. A biting fly can pick up the microfilariae when it sucks up blood, and there they develop into larvae. Adult worms can live up to 18 years. It only occurs in equatorial Africa, southern Arabia and Central America.

The symptoms include extremely itchy skin, generalised rash, lumps under the skin usually over the lower back and thighs, premature ageing and wrinkling of the skin, changes in skin pigmentation and grossly enlarged lymph nodes in the groin. Blindness occurs in 5% of patients when the microfilariae spread to the eye and damage the cornea (clear surface layer of the eye). Rarely, muscles and the intestine may be affected to cause weakness and weight loss.

Surgical removal and examination of a skin lump reveals an adult worm, and medication (eg. diethylcarbemazine, ivermectin) can be given to kill the microfilariae. The medication often must be repeated every six months for some years to give a cure. The death rate in untreated patients about one in a hundred.

**OPHTHALMIA**
Ophthalmia is a severe form of conjunctivitis with bacterial infection, copious pus production, keratitis and inflammation of the eye. It most commonly occurs in the newborn (ophthalmia neonatorum) whose eyes are exposed to bacterial such as *Chlamydiae* and *Gonnococcus* in the birth canal.

See also CONJUNCTIVITIS; KERATITIS
OPTIC ATROPHY
Optic atrophy (wasting) can be seen on ophthalmoscopic examination of the retina at the back of the eye. A pale optic disc with blurred margins is noted. Disc cupping may also occur, and the patient may complain of reduced visual acuity and reduced visual field (width of vision).

The causes of optic atrophy include glaucoma, arteriosclerosis, retinal ischaemia, optic neuritis, Paget's disease, tumour pressing on optic nerve, retinitis pigmentosa, vitamin B deficiency and methanol poisoning. These conditions all cause a poor arterial blood supply to, or chronic inflammation of, the optic disc.

PAPILLITIS
Papillitis is inflammation of the papilla at the back of the eye at the point where the optic nerve enters the eyeball. The red, inflamed and swollen papilla can be seen through an ophthalmoscope. It is a form of neuritis (nerve inflammation).

PARINAUD SYNDROME
The most common initial presentation of patients with the Parinaud or pretectal syndrome is that they are unable to look upwards without tilting their head back - the muscles that pull the eyes upwards are paralysed. Other symptoms include abnormal reaction of the pupils to light and other abnormalities of eye function. The cause is a tumour of the pineal gland (in the front of the brain) or brain that puts pressure on one of the nerves that control eye movement, or a stroke that affects the part of the brain that controls this nerve.

PATAU SYNDROME
Patau syndrome (trisomy 13-15) is a rare congenital defect affecting numerous parts of the body, caused by the presence of three copies of chromosomes 13 and 15 instead of two. The infant has extra fingers and toes, abnormal heart structure, cleft lip and palate, small eyes and brain malformations. Tests are performed on heart and brain function (eg. CT scan, echocardiogram) to confirm the diagnosis. No treatment is available, and the prognosis is poor.

The incidence of Patau syndrome increases with the age of the mother.

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PELIZAEUS-MERZBACHER DISEASE
Pelizaeus-Merzbacher disease is a rare X-linked inherited brain condition that is slowly progressive to death in early adult life. Symptoms include tremor, poor coordination, nystagmus (eye ball tremor), abnormal movements, difficulty in speaking and intellectual deterioration. It is caused by a worsening loss of myelin (insulating material) around the nerves in the brain and spinal cord.
PHACOMALACIA
Phacomalacia is a softening of the lens in the eye due to the presence of a cataract. It causes significant blurring of the vision. Surgical replacement of the lens is an effective form of treatment.
See also CATARACT

POLYARTERITIS NODOSA
Polyarteritis nodosa (PAN or periarteritis nodosa) is an inflammation of small to medium-sized arteries. The damaged artery may become weakened and balloon out to several times its normal diameter, it may scar and shrink down, or the blood passing through the inflamed section of artery may clot and completely block the artery (a thrombosis). The arteries affected may be anywhere in the body, but the gut, liver, heart, testes, kidney, and muscles are most commonly involved. The cause is unknown, but it is more common in drug abusers and in patients with hepatitis B. Rarely it may be a side effect of medication. Men are three times more likely to develop the disease than women, and it is most common in young adults.
The symptoms are very varied, depending on which arteries and organs are involved. The patient is usually feverish, and has pain in the area involved. Specific complaints may include muscle pain, palpitations, arthritis, skin ulcers, spots in the vision, abdominal pain, nausea, vomiting, diarrhoea and high blood pressure.
There are no diagnostic blood tests, and the diagnosis must be confirmed by a biopsy (sample) taken from an involved artery. Taking steroids (e.g. prednisone) in high doses for a long period of time is the main treatment, and immunosuppressive drugs may also be used.
The prognosis varies markedly from one patient to another, depending upon the areas and arteries involved. Some patients do recover, but the majority slowly deteriorate to die within a few months or years.

POLYCYTHAEMIA RUBRA VERA
Polycythaemia rubra vera (the “rubra” is sometimes omitted) is an excessive production of red blood cells that is most common in middle-aged to elderly, overweight men, but may occur in both sexes. It is rare under 40 years of age.
Red blood cells are made in the spleen and bone marrow, primarily of the breastbone (sternum), pelvis and thighbone (femur). If the marrow becomes overactive, excessive numbers of cells may be produced, and the patient develops a headache, dizziness, tiredness, blurred vision, generalised itching, noises in the ears, high blood pressure and an enlarged spleen. Blood clots may occur in vital organs (e.g. brain to cause a stroke), and some patients develop a form of chronic leukaemia.
The diagnosis can be confirmed by finding excess red blood cells in a blood test, and further tests on bone marrow determine the severity of the disease.
The disease can be controlled, but not cured, by draining large quantities of blood out of a vein initially, and smaller amounts on a regular basis long term. Medications (e.g. busulfan) to reduce the activity of the bone marrow may also be used. The average survival time after diagnosis is twelve years.

POSNER-SCHLOSSMAN SYNDROME
The Posner-Schlossman syndrome (glaucomatocyclitic crisis) is a rare condition characterised by recurrent attacks of glaucoma in one eye associated with uveitis. Young
A RATIONALE FOR THE EYES

adults are usually affected. The cause is inflammation of the drainage system for the fluid in the eye.

Patients experience halos around lights, blurred vision and sometimes pain. Repeated attacks may cause permanent eye damage.

It is diagnosed by measuring the pressure in the eye using a tonometer (device that rests on the anaesthetised eye, or a brief puff of air onto the eye) and treated with steroid eye drops, glaucoma drops and indomethacin tablets. There is a good response to treatment for each attack but future attacks cannot be prevented.

See also GLAUCOMA; UVEITIS

POSTERIOR VITREOUS DETACHMENT

The ball of the eye behind the lens and in front of the retina is filled with a jelly-like substance called vitreous humour. It is this that gives the eyeball its firmness and develops its spherical shape during foetal growth. The vitreous is attached to the retina at the back of the eye.

With ageing the structure of the gel that constitutes the vitreous humour slowly degenerates and the vitreous may detach from the retina at the back of the eye. Patients experience minimal symptoms in most cases but may see flashing lights at the edge of their vision or may see a “floater” in their vision. A floater is a collection of cells or protein in the thick fluid that fills the eyeball, which casts a shadow on the light-sensitive retina at the back of the eye. These symptoms usually settle in a few weeks but it is essential for patients to be assessed by an ophthalmologist to assess them for damage to the retina.
A RATIONALE FOR THE EYES

As the vitreous detaches from its adhesion to the retina it is possible in about 10% of patients for the retina to tear and also separate from the back of the eyeball. This causes much more significant visual disturbances that require urgent medical management.

If a posterior vitreous detachment is detected by examining the eye through a slit lamp or ophthalmoscope, and there is no sign of a tear in the retina or retinal detachment, and over the next few weeks the flashes and floaters disappear, then no treatment is necessary as it is a normal part of the ageing process and it is unlikely that there will be any further visual problems. If the floaters or flashes worsen or recur, further specialist assessment is necessary.

See also FLOATER IN EYE; RETINAL DETACHMENT

PRESBYOPIA

Presbyopia is a deterioration of close vision, which starts in middle age. The cause is stiffening of the lens with age, which prevents it focussing light rays from close objects accurately on the retina (light sensitive layer of cells at the back of the eye).

Patients experience steadily worsening difficulty in reading small print or seeing close objects in detail. Correct spectacles can be prescribed after suitable vision tests to assist with focusing.

See also LONG-SIGHTED

REITER SYNDROME

Reiter syndrome (reactive arthritis) is an inflammatory condition involving the eyes, urethra and joints. The cause is unknown, but it is more commonly in young men, and often follows a bacterial infection.

It has the unusual and apparently unconnected symptoms of conjunctivitis (eye inflammation), urethritis (inflammation of the urine tube - the urethra) and arthritis (joint inflammation). Other symptoms that may occur include mouth ulcers, skin sores, inflammation of the foreskin of the penis and a fever. Rarely, the heart becomes inflamed.

Blood tests are not diagnostic, but indicate presence of inflammation, and X-rays show arthritis in the joints of the back only after several attacks.

It heals without treatment after a few days or weeks, but the arthritis tends to last longer and recurrences are common. The disease course can be shortened by anti-inflammatory drugs such as indomethacin.

See also CONJUNCTIVITIS

RETINAL DETACHMENT

The light-sensitive retina at the back of the eye is loosely attached to the eyeball, but if it detaches from the back of the eye, full or partial blindness results. It may occur very slowly over a period of years, or be complete in a few minutes. The retina may detach if a blood vessel ruptures and bleeds behind the retina, if the fluid in the eye leaks behind the retina, it may follow an injury to the eye, or be caused by high blood
pressure or a tumour in the eye. Marfan syndrome is a rare cause, but frequently, there is no obvious cause.

The patient describes a black curtain slowly moving across the field of vision, as the retina progressively lifts away from the eyeball and causes at first partial, and later complete blindness. The detachment can be seen by examining the eye with an ophthalmoscope (small magnifying glass attached to a light).

Rapid treatment is essential to save the sight. A surgical procedures, or a laser that is shone in short, sharp, accurately aimed bursts into the eye, are used to seal the retina back onto the eyeball. 95% of retinal detachments can now be cured or controlled if treated immediately they occur.

See also POSTERIOR VITREOUS DETACHMENT; RETINOSCHISIS; STICKLER SYNDROME

RETINAL EXUDATES

Retinal exudates are white fluffy patches seen when the retina at the back of the eye is examined through an ophthalmoscope. The patches are caused by blockage of the capillaries in the retina that occurs in diseases such as poorly controlled diabetes mellitus, hypertension (high blood pressure), increased pressure in the skull (eg. after a head injury, tumour in the brain etc.) and after massive blood loss.

RETINAL HAEMORRHAGE

Retinal haemorrhages are red spots and patches adjacent to blood vessels that can be seen when the retina at the back of the eye is examined through an ophthalmoscope.

Various types of haemorrhage are described including punctate (points), splinter (lines) and flame (teardrop shape). They may be caused by diseases such as pernicious anaemia, leukaemia, aplastic anaemia, hypertension (high blood pressure), diabetes mellitus, bacterial endocarditis, anticoagulants (eg. warfarin) and other diseases that may increase the tendency to bleed (haemorrhage).
A RATIONALE FOR THE EYES

RETINAL PIGMENTATION
See RETINITIS PIGMENTOSA

RETINITIS
Retinitis is an inflammation of the retina at the back of the eye. The usual cause is a viral infection. It causes light sensitive eyes and blurred vision. Recovery is common, but rarely there may be loss of vision.
See also RETINITIS PIGMENTOSA

RETINITIS PIGMENTOSA
Retinitis pigmentosa is an inherited disease of the light-sensitive cells in the retina at the back of the eye that passes from one generation to the next. It may occur alone or in conjunction with the Bassen-Kornzweig syndrome (abetalipoproteinaemia) and Laurence-Moon-Biedl syndrome.

It starts with night blindness in childhood, followed by tunnel vision and slowly progresses to cause near total blindness in old age. The retinal cells steadily deteriorate, and pigmented cells replace them. The degeneration starts at the edge of the retina and progressively moves towards the centre. The field of vision slowly decreases until the patient can only see straight ahead as though through a tunnel, and has no peripheral vision.

The retina has a characteristic pigmented appearance when viewed by a magnifying light (ophthalmoscope).

No treatment is available, and the condition is slowly progressive over many years
See also LAURENCE-MOON-BIEDEL SYNDROME; RETINITIS

RETINOBLASTOMA
A retinoblastoma is cancer of the retina (light sensitive cells at the back of the eye) that usually occurs in children under three years of age. There is a familial (inherited tendency) in 40% of cases, but cause in others is unknown.
A RATIONALE FOR THE EYES

The pupil becomes white, a squint is noticed, the affected eye bulges forward, becomes reddened and the vision is affected. The cancer may spread (metastasise) from the eye along the optic (vision) nerve to the brain.

It is diagnosed by examining the eye with a magnifying light (ophthalmoscope) and a CT scan.

Small tumours may be treated by laser or chemotherapy (medication), but most are not diagnosed until large, and the eye must be removed. The prognosis depends on the size of the tumour at time of diagnosis. Survival rate with no spread is 85%, but this drops dramatically if cancer cells are found in the optic nerve. The overall five year survival rate is about 70%.

RETINOPATHY
A retinopathy is any disorder or disease of the retina at the back of the eye.
See also RETINITIS PIGMENTOSA; RETINOBLASTOMA

RETINOSCHISIS
A split in the light sensitive retina at the back of the eye is described as retinoschisis. It may be very small and barely noticed by the patient, or it may steadily extend and be a precursor to retinal detachment. It may appear as a black line in the vision of one eye and is corrected by sealing the tear with a laser.
See also RETINAL DETACHMENT

RICHNER-HANHART SYNDROME
The Richner-Hanhart syndrome is a congenital condition caused by excessive levels of the protein tyrosine in the blood because the liver lacks the necessary enzymes to break it down. The affected child suffers from eye damage, intellectual disability, convulsions and skin damage. Once diagnosed, the offending proteins can be removed from the diet, and no further deterioration of the patient’s condition should occur. Early diagnosis of this inherited syndrome is therefore vital, but may be very difficult due to the subtle nature and onset of symptoms in an infant.

RIEGER SYNDROME
Rieger syndrome is a familial (runs in families from one generation to the next) developmental abnormality of the teeth and eyes. Patients have an abnormal iris (coloured part) of the eye, and the teeth are reduced in number and smaller than normal. There is no cure, but patients have normal life expectancy.

RILEY-DAY SYNDROME
Riley-Day syndrome (dysautonomia) is an uncommon familial (runs in families from one generation to the next) syndrome that occurs in Jews of Middle Eastern extraction. Its symptoms include a lack of tears in the eyes, excessive sweating, intermittent fevers and episodes of low body temperature, blood pressure swings between being too high and too low, the surface of the eye may feel no pain, and generally patients feel only the most severe pains. As a result they may have fractures and other injuries of which they have no knowledge. Less commonly, they may have poor coordination, difficulty in swallowing, difficulty in talking, and extreme mood swings. They may suffer serious personal injury, particularly to the eye resulting in blindness. No treatment is available.
RIVER BLINDNESS
See ONCHOCERCIASIS

SCHILDER DISEASE
Schilder disease is caused by a progressive loss of myelin (insulating material) from around the nerves in the brain. This results in muscle spasms, loss of speech, deafness, eye disorders, dementia and even kidney damage. It is similar to multiple sclerosis but always occurs in children. No treatment is available.

SCLERITIS
Scleritis is an uncommon inflammation of the whites (sclera) of an eye. Numerous different types are recognised (eg. anterior scleritis, posterior scleritis, diffuse scleritis, nodular scleritis).

Scleritis
The cause is often unknown, but it may be associated with rheumatoid arthritis, polyarteritis nodosa, systemic lupus erythematosus and other autoimmune diseases.
Redness, discomfort, and sometimes painful ulceration, of the sclera occurs. The sclera may become thickened and affect vision, while cataracts and glaucoma may occur in severe forms.
Anti-inflammatory or steroid eye drops and tablets give a good response.
See also EPISCLERITIS
A RATIONALE FOR THE EYES

SENIOR-LØKEN SYNDROME
See LØKEN-SENIOR SYNDROME

SHORT-SIGHTED
Short-sightedness (myopia) is a developmental vision defect in which the eyeball is too long, and light rays from distant objects are focused in front of the retina (layer of light sensitive cells at back of the eye). Distant objects appear blurred while close objects are clearly seen. Reading is easy but moving around difficult.
Spectacles with accurately prescribed corrective lenses can correct the problem. In some people, laser keratotomy, in which the shape of the cornea (outer layer of the eye) is permanently reshaped, may cure the problem. This procedure is not carried out before the late teen years as vision can change with growth.
See also LONG-SIGHTED; ORTHOKERATOLOGY; PRESBYOPIA; REFRACTIVE SURGERY; STICKLER SYNDROME; VISION

SICCA SYNDROME
The Sicca syndrome is a functional failure of the salivary and tear (lacrimal) glands, which may be caused by a complication of Sjögren syndrome, a drug reaction or autoimmune disease. Patients have swollen salivary and lacrimal glands, a dry mouth, dry eyes, dry vagina and dry throat. A salivary gland biopsy is used to diagnose the condition.
It is a self-limiting benign condition and artificial tears, saliva and lubricants are used to ease the symptoms.
See also MIKULICZ DISEASE; SJÖGREN SYNDROME

SJÖGREN SYNDROME
Sjögren syndrome is a chronic widespread autoimmune inflammatory condition in which the body inappropriately rejects its own tissue. It is closely related to rheumatoid arthritis, but affects more organs.
Common symptoms include widespread arthritis, dry eyes, dry mouth, dry skin and dry throat. Other symptoms may include difficulty in swallowing, decaying teeth, loss of taste and smell, and a hoarse voice. Nearly all patients are women, and it usually commences in the fifth decade. Complications may involve inflammation of the pancreas, thyroid and other organs. It is diagnosed by specific blood tests.
Patients are prescribed anti-inflammatory drugs, steroids (eg. prednisone), and a number of unusual drugs such as gold by injection or tablet, antimalarial drugs (eg. chloroquine) penicillamine (not the antibiotic), and cell-destroying drugs (cytotoxics). Artificial tears and skin moisturisers, and good dental hygiene are also necessary.
There is no cure, but reasonable long-term control is usually possible.
See also SICCA SYNDROME
**SLY SYNDROME**

The Sly syndrome is a rare congenital condition in which children are unable to eliminate certain substances (mucopolysaccharides) from the body that starts at one to two years of age. The cause is an inherited abnormality of the metabolic system (mucopolysaccharidosis) that is closely related to Hurler syndrome.

The symptoms include recurrent respiratory infections, enlarged liver and spleen, excessive forward curvature of the spine, heart murmurs, eye cataracts and slow growth. Specific blood enzyme tests are abnormal and enable a diagnosis.

There is no effective treatment and the usual course is a steady progression to death by about 10 years of age.

**SMITH-LEMLI-OPITZ SYNDROME**

The Smith-Lemli-Opitz syndrome is an inherited genetic abnormality causing varied deformities. Both parents must be carriers for the syndrome to develop. The symptoms may include droopy eyelids, narrow forehead, intellectual disability, abnormal penis, malformed nostrils and webbed toes. Surgical correction of the deformities is possible, but there is no cure.

**SQUINT**

A squint (strabismus or a cast) occurs when the eyes appear to look in slightly different directions away from each other (cross-eyed). When the eyes are both turned inwards the condition is called esotropia. It is critical that this is detected and treated when it occurs in childhood, because if allowed to persist, the brain will permanently suppress the vision in one eye in order to overcome the double image it receives. Even if the good eye becomes blind later in life, the eye in which vision has been suppressed will not be able to see.

In children a squint is usually due to an inherited tendency with weakness or abnormal development of the tiny muscles within the eye socket, which move and align the eyes, or abnormal vision in one eye due to a cataract (cloudy lens).

In older patients a squint may be caused by damage to the muscles that control eye movement from a direct blow to the eye or surrounding skull, a poor blood supply to the muscles of one eye, a stroke that affects the nervous control of the eye muscles, a tumour or cancer in the eye socket, an over active thyroid gland (causes eyes to protrude slightly) or multiple sclerosis (affects nerves to eye muscles).

Special spectacles may be used long term to correct the problem by reducing the angle of the squint. In more severe cases an eye patch may cover the good eye to strengthen the poorer one and eye exercises may be added. In marked degrees of squint, it is necessary to operate to change the tightness of the tiny muscles that control eye movement, which is a
A RATIONALE FOR THE EYES

technically a difficult operation for the surgeon, but relatively minor surgery for the patient. Provided medical advice is followed, the long-term cosmetic and vision results are excellent.

See also BROWN SYNDROME; DUANE SYNDROME

STAGARDT SYNDROME

Stagardt syndrome is an inherited condition of adolescents in which there is a deterioration and eventual death of the cells in the centre of the light sensitive retina at the back of the eye where central vision is focused. Both parents must be carriers for the condition to occur.

Patients notice deteriorating central vision while peripheral vision remains. Eye examination with an ophthalmoscope is abnormal, and a special dye (fluorescein) injected into an artery shows the damaged blood supply to the retina. There is no treatment available and the condition is slowly progressive.

STEVENS-JOHNSON SYNDROME

The Stevens-Johnson syndrome is a severe complication of erythema multiforme that may be triggered by drugs or infection.

The characteristics of the syndrome include erythema multiforme; severe purulent ulcerating conjunctivitis; high fever; inflamed mouth (stomatitis); blisters or ulcers in nose, vagina, urethra, and anal canal; and ulceration, pain and swelling extending down the throat and into the lungs to give a form of bronchitis. The heart and lungs may become involved, and the scalded skin syndrome may develop. There is no specific diagnostic test.

Treatment involves intensive steroid therapy, and if possible, removing the cause of the erythema multiforme. Most patients recover slowly, but death is possible in the elderly and debilitated.

STICKLER SYNDROME

Stickler syndrome is an inherited condition that is characterised by short-sightedness (myopia), detachment of the retina from the back of the eye that results in blindness and premature arthritis in joints. There may be associated hearing problems.

STRACHAN SYNDROME

Strachan syndrome (also known as Jamaican neuritis) causes widespread nerve and skin damage due to poor nutrition, particularly a lack of vitamin B. It often occurs in alcoholics and with starvation, and is worse in smokers.

The symptoms include amblyopia (dim vision), dermatitis around the mouth and genitals, and painful and excessively sensitive areas of skin. Some patients have muscle weakness or spasms and permanent vision damage occurs if the syndrome is left untreated for a prolonged period. Abnormalities of the retina (light sensitive area at back of eye) can be seen when examined through an ophthalmoscope (magnifying light), and there are blood test abnormalities, and over active reflexes.

Vitamin supplements and a good diet cure the condition.

See also AMBLYOPIA

SUBCONJUNCTIVAL HAEMORRHAGE

A subconjunctival haemorrhage (or haematoma) may be seen as a red patch on the white of the eye (conjunctiva). It is caused by a small capillary in the conjunctiva leaking blood after an injury to the eye. This may be as mild as vigorously rubbing the eye. They are not serious
A RATIONALE FOR THE EYES

unless the bleeding appears to be coming from the back of the eyeball when a medical opinion should be sought.

Most cases settle slowly with no treatment over a week or two as the red becomes brown then yellow as it slowly fades.

**SUNCT SYNDROME**

The SUNCT syndrome is a variant form of cluster headache whose name is an acronym of its major symptoms (short lasting, unilateral, neuralgiform, conjunctival injection, tears). It may be triggered by alcohol, stress, exercise, certain foods and glare.

The symptoms are severe headaches that are short lasting (seconds to minutes), one sided (unilateral) and piercing (neuralgiform) with associated red eyes (conjunctival injection) and excess tear production. They may occur regularly or spasmodically.

Medication such as sumatriptan and ergotamine, nasal capsaicin spray, and inhaling pure oxygen are used as treatments. The headaches are very annoying but not serious.

**SYMPATHETIC OPHTHALMIA**

Sympathetic ophthalmia is a disastrous but very rare complication of eye injury or surgery. When the blood vessel carrying layer in the eyeball (uvea) on one side becomes inflamed, the uvea in the other eye also becomes inflamed and damaged resulting in both eyes becoming blind although only one was injured or operated on.

Treatment may involve steroid medications or removal of the eye originally injured, but the condition is often permanent.

**TAY-SACHS DISEASE**

Tay-Sachs disease, which was once known as amaurotic familial idiocy, is an inherited disorder that occurs mainly in Ashkenazi Jews of Eastern Europe. It affects the way in which fats (lipids) are metabolised due to the lack of an essential enzyme, and results in the accumulation of lipids in the brain. The victims suffer a progressively worsening deterioration in their mental capacity, convulsions, blindness, loss of the use of the limbs and death in childhood. The diagnosis can be made before birth by amniocentesis. There is no treatment available.
A RATIONALE FOR THE EYES

See also BLOOM SYNDROME; RILEY-DAY SYNDROME

TEMPORAL ARTERITIS
Temporal or giant cell arteritis, is an inflammation of medium to large arteries throughout the body, but most commonly the arteries in the temples at the side of the head. The cause is unknown but it may an autoimmune disease, and often follows a significant viral infection.

Involved arteries become extremely tender and swollen. Symptoms depend on which arteries are inflamed, but may include headache, scalp tenderness, pain in the jaw with chewing, throat pain and vision disturbances. Less commonly a cough, shoulder pain, weakness and a fever occur. Blindness due to involvement of the arteries in the eye, and aneurysms (dilations) of arteries are complications. About half of all patients also have polymyalgia rheumatica.

Blood tests are usually performed to detect the inflammation, and a biopsy of an artery will reveal the presence of characteristic giant cells. Treatment involves taking steroid tablets (eg: prednisone) for several months. It is usually well controlled and eventually cured, but recurrences when medication is ceased are common.

THIRD EYELID OF MORGAN-DENNY
The third eyelid of Morgan-Denny is an extra fold of skin above the upper eyelid. It is a sign of atopic eczema of the eyelid, allergic conjunctivitis or another chronic irritating conditions of the eye. It is caused by persistent rubbing of the eye and is more common in children.

See also ALLERGIC CONJUNCTIVITIS

TOLOSA-HUNT SYNDROME
In this syndrome, an aneurysm (ballooning) on the side of the internal carotid artery at the base of the skull puts pressure on nerves to cause a painful paralysis of one eye.

The diagnosis is made by angiography (x-ray of an artery after injection of a dye), CT or MRI scan. Surgical treatment of the cause, which may be difficult to find, usually cures the problem.

TRACHOMA
Trachoma is a type of conjunctivitis (superficial eye infection) caused by the bacteria-like Chlamydia organism, which is very common in areas of low hygiene where flies can transmit the infection from one person to another. It is particularly common among Australian Aborigines.

Mild infections may not be very noticeable, and in children may cause no symptoms. In more severe cases, eye pain, intolerance to bright lights, and a weeping swollen eye develop. Small bubbles on the underside of the eyelids are the earliest sign of the disease. Chronic trachoma causes scarring of the cornea (the outer surface of the eye) and subsequent blindness. Blood vessels grow into the scar tissue, and the coloured part of the eye and the pupil may be covered with a thick scar and obvious small arteries and veins. The gland that produces tears (the lacrimal gland) can also be damaged so that tears no longer form, the eye dries out, and is further damaged and scarred.

It is diagnosed by culture and examination of swabs from the eye.

A one to three month course of antibiotics and antibiotic eye ointment are required for treatment. Once blindness has occurred from corneal scarring, the only treatment is surgical replacement of the damaged cornea by one donated by a deceased person.
It is usually cured if treated within the first year, and the outcome is excellent, but if left longer some scarring of the eye surface may occur.

See also CONJUNCTIVITIS

TREACHER-COLLINS SYNDROME
The Treacher-Collins syndrome is a rare developmental disorder of the face that is transmitted within a family by an irregularly dominant gene. Patients have an under developed lower jaw, large mouth, absent angle between nose and forehead, abnormal eye slant, notched lower eye lids, sparse eyelashes, hairy cheeks, low set ears, deafness and middle ear abnormalities. Plastic surgery may correct some deformities but there is no cure.

TRICHIASIS
Trichiasis is an ingrown eyelash that causes irritation of the eye surface (conjunctiva). It may be caused by an injury to the eyelid, entropion or trachoma.

See also ENTROPION; TRACHOMA

UVEITIS
Inflammation of the iris, the coloured part of the eye, is iritis. When the surrounding tissues of the uveal tract are also involved, it is called uveitis. Both conditions may be due to an infection such as toxoplasmosis, tuberculosis or syphilis (exogenous iritis), or it may be associated with inflammatory diseases in other parts of the body, including psoriasis, ankylosing spondylitis, and some bowel conditions (endogenous iritis). The latter form is more common.

Usually only one eye is involved which will suddenly become red and painful with blurred vision. Bright lights will aggravate the eye pain and the pupil is small. In the exogenous form, there is less pain and the onset is slower, but this form often results in some permanent deterioration in vision.

Any underlying infection or disease must be treated if possible, and the eye is made more comfortable with warm compresses. Steroid eye drops are used to reduce inflammation.
Usually recovery is satisfactorily, but recurrences are common. See also FUCHS UVEITIS SYNDROME; IRITIS; POSNER-SCHLOSSMAN SYNDROME;

VELOCARDIOFACIAL SYNDROME

The velocardiofacial (VCFS or Shprintzen) syndrome is an inherited congenital abnormality of the face and heart occurring in one in 2000 births. It has the same chromosomal pattern as di George syndrome involving chromosome 22, but with different characteristics. It was first described by Dr. Robert Shprintzen of New York in 1978.

Affected children have small stature, abnormal facial features (down turned corners of the mouth, long mid-face and cylindrical nose), nasal speech pattern and heart abnormalities such as a hole between the ventricles or misaligned main vessels (Fallot’s tetralogy). Some children also have a cleft palate, eye abnormalities, feeding difficulty as babies, slender hands, unusual emotional upsets and a learning disability. Numerous other abnormalities may be associated with the syndrome including reflux of urine from the bladder to the kidneys, club foot and an umbilical hernia. Between 10% and 30% develop significant psychiatric disturbances in adult life. A chromosomal gene analysis can confirm the suspected diagnosis.

Treatment involves surgery to correct heart, nasal and facial abnormalities if possible. Speech therapy and physiotherapy may also be beneficial. there is no cure, but patients have a reasonable life expectancy if the heart defects can be corrected.

VISCERAL LARVA MIGRANS

Visceral (and ocular) larva migrans are internal infestations by the larvae of a roundworm (nematode). Dogs infected by the roundworm (Toxocara canis) pass worm eggs out with their faeces to contaminate the soil. Eggs swallowed by humans (often children) hatch into larvae, which penetrate through the gut wall into the bloodstream by which they are carried to a variety of organs, particularly the lungs, liver, brain and eye (ocular larva migrans).

The symptoms are very variable, depending on which organ the larvae are carried to, and the number of larvae present. They usually include fever, tiredness, loss of appetite and
A RATIONALE FOR THE EYES

weight loss. Organ specific symptoms include cough, wheeze, rash, large liver and spleen, visual disturbances, seizures and behavioural disorders. Heart infestation and pneumonia may lead to death.

There is a specific blood test to detect the presence of the larvae, and other blood tests show significant reactive changes. Chest x-rays may show lung inflammation. Masses of larvae in the eye can be seen by looking through the pupil with an ophthalmoscope (magnifying light). Medication (eg. thiabendazole) is available to destroy the larvae, and steroids are used to reduce inflammation.

The larvae cannot develop into worms in humans, and die off naturally after several months, but permanent organ damage may occur. The treatment of eye disease is unsatisfactory.

VITREOUS DETACHMENT
See POSTERIOR VITREOUS DETACHMENT

VOGT-KOYANAGI-HARADA SYNDROME
The Vogt-Koyanagi-Harada syndrome is a form of inflammation of the brain, eyes and ears of unknown cause.

Patients develop recurrent attacks of encephalitis and meningitis, with uveitis (eye inflammation), detachment of the light sensitive retina from the back of the eye (causes patches of blindness), fever, headache and dizziness. It may be associated with a white patch of hair and skin, hair loss (alopecia), cataracts and glaucoma in the eyes, and deafness and ringing in the ears (tinnitus). One or both eyes may be affected.

There is no specific diagnostic test and no treatment is available. Most cases settle spontaneously with time, but permanent eye damage often occurs.

See also UVEITIS

von HIPPEL-LINDAU SYNDROME
The von Hippel-Lindau syndrome is an inherited developmental abnormality of the blood vessels in the eye and brain that affects young adults. Patients develop visual disturbances from the growth of tumours made of overgrown capillaries on the light sensitive retina at the back of the eye. Tumours may also develop in the cerebellum (lower back part of brain) and cause varied brain disturbances. Ophthalmoscopy (examining the eye with a magnifying light) shows the abnormal growths and a CT or MRI scan of the brain may show tumours.

Treatment involves laser photocoagulation or surgical resection of retinal tumours, and brain surgery to remove any tumours that develop. There is no permanent cure, and the outcome depends on the number, severity and location of the tumours.

WAARDENBURG-KLEIN SYNDROME
The Waardenburg-Klein syndrome is a familial (passes through families) genetic developmental abnormality. It causes deafness in one or both ears, a broad root of nose with sideways displacement of the corner of the eyes, the eyebrows join together, there is a white hair forelock and eye irises of different colours. There is no specific diagnostic test and the only treatment is plastic surgery for the facial abnormalities.
A RATIONALE FOR THE EYES

WAGR SYNDROME
The WAGR syndrome is a genetic developmental abnormality caused by damage to chromosome 11. Its name is an acronym for the principal features that are a Wilms tumour of the kidney, no iris in the eyes (Aniridia), ovarian or testicular tumours (Gonadoblastomas) and intellectual disability (Retarded). The diagnosis can be confirmed by a chromosome analysis. The Wilms' and gonad tumours can be treated but the prognosis is very poor.

WILLIAMS SYNDROME
Patients with the genetic developmental abnormality Williams syndrome (named after a New Zealand physician) have a bright blue lacy iris in each eye, prominent folds of skin at the inside corner of the eyes, prominent lips, droopy cheeks, slight mental retardation and a friendly outgoing personality. Unfortunately they may also have damaged heart valves and problems with calcium deposits in various tissues. There is no treatment available, and it sometimes can be a family trait but patients have a reasonable life expectancy.

WILSON DISEASE
Wilson disease (hepatolenticular degeneration) is a rare inherited disorder of copper metabolism with symptoms relating to the brain, the liver or both, that occurs in both sexes and is usually diagnosed between 10 and 30 years of age. It results in the excessive deposition of copper in the liver and brain.

Excess copper in the brain may cause psychiatric disorders, rigid muscles and a tremor. Liver disease symptoms include jaundice (yellow skin), an enlarged liver and/or spleen, anaemia and hepatitis. A brown/green ring (Kayser-Fleischer ring) around the iris (coloured part) in the eye is easily visible. The diagnosis confirmed by blood tests that detect the excessive copper.

Copper can be removed by a number of drugs (eg. penicillamine), and a diet low in copper (eg. avoiding shellfish, beans and offal) must be followed. Lifelong treatment is necessary to keep copper levels low. Any damage to the brain or liver caused before the treatment is started cannot usually be reversed, but the long-term outlook is normally good.

Samuel Wilson (1878-1937) was an English neurologist.

WOLFRAM SYNDROME
Wolfram syndrome is a rare inherited condition characterised by the presence in a child of both diabetes mellitus and diabetes insipidus, and progressive deafness and eye wasting that leads to blindness.

WYBURN-MASON SYNDROME
The rare Wyburn-Mason syndrome is caused by congenital dilated connections between arteries and veins (arteriovenous aneurysms) in the brain and head that cause abnormalities of the retina in the eye, angiomias (dilated blood vessels) on the skin of the face and intellectual disability.

ZELLWEGER SYNDROME
The Zellweger syndrome or cerebro-hepato-renal syndrome is a genetic abnormality of the brain, liver and kidneys. It is a familial (runs through families) condition, but both parents must be carriers.
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Sufferers have a flat long face, large fontanelle (soft spot) in skull, poor muscle tone, difficulty in breathing from birth, a large liver that fails to function and seizures. The eyes may be affected by glaucoma and cataracts, joints may be contracted, bony abnormalities occur and the testes do not descend in boys. Numerous blood, urine and skeletal x-ray abnormalities can be detected. No treatment is available and death within two months of birth is usual.

It is named after the American paediatrician Ulrich Zellweger (1909-1990).
EYE ANATOMY

Light enters the eyes and stimulates nerves, which in turn transmit impulses to the brain where the information is interpreted as visual images. Light travels in straight lines, but it can be bent if it passes through a lens, be that in a camera or the human eye. The shape of the lens can precisely control the degree of bending, and tiny muscles attached to it can change the shape of the lens in the human eye.

Each eye is a slightly flattened sphere consisting of three layers. The outer layer is called the sclera and forms the white of the eye, except for the very front section, which is transparent to allow light in, and is called the cornea. The second layer is the choroid and contains the blood vessels that service the eye. The front of the choroid forms the iris, which is the part that gives the eyes their colour depending on the inherited genes. In the centre of the iris is a small gap called the pupil. Muscles in the iris enlarge or reduce the size of the pupil according to the amount of light - the more light the smaller the pupil. Just behind the iris is the lens.

The innermost layer of the eye, curving around the back of the sphere, is the retina. This is a light-sensitive structure containing nerve cells, commonly called rods and cones because of their shapes. The rods are sensitive to light and will function in dim light but do not produce a very sharp image. The cones are sensitive to colour. When you go into a darkened room such as a theatre, it is difficult to see for the first few moments. This is the time it takes the rods to adjust to the change in light. The nerves in the retina all meet together to form the optic nerve, which connects to the brain.
The fovea is a small depression in the retina at the back of the eye where the light sensitive cells, which detect vision are very highly concentrated. Light focused on this point has the greatest degree of definition. It is directly opposite the pupil.

Between the lens and the cornea is a chamber filled with a watery fluid, called the aqueous humour. The ball of the eye behind the lens and in front of the retina is filled with a jelly-like substance called vitreous humour. It is this that gives the eyeball its firmness and maintains its spherical shape.

Light passes through all the transparent layers starting with the cornea, then through the pupil and aqueous humour, the lens, followed by the vitreous humour to finally impinge on the retina. All these layers refract (bend) the light so that light from the large area outside is focused in the small area of the retina. The most important refractive body is the cornea, which is responsible for about 70% of the process. The lens focuses the light according to whether it is for near or distance vision. When the light reaches the retina, the nerve cells convert it into electrical impulses and send these along the optic nerve to the brain, which records visually the objects we are looking at. Just like the lens of a camera, the lens of the eye produces an upside down image. The brain is responsible for the right-side-up, three-dimensional view that we eventually get. If the optic nerve is damaged, it can cause blindness even though the eyes themselves are still functioning. The point where the optic nerve leaves the retina has no rods and cones and so forms a blind spot.

The lacrimal glands, behind the outer end of the upper eyelids, produce tears that constantly refresh the eye surface. About a dozen tiny ducts lead from the gland to the surface of the eye behind the upper lid.

The lacrimal duct leads from the inner corner of each eye, through a dilation known as the lacrimal sac, to the back of the nose. It drains excess tears from the eye surface.

The eye is one of the most mobile organs in the body. Each eye has a set of six external muscles so that it can move in all directions.

Because the eyes are so sensitive, the body provides a great deal of protection for them. They are set in two bony sockets in the skull. Externally they are guarded by the eyebrows and the eyelids. The eyebrows help to ward off blows, shield the eyes from sunlight and deflect sweat so that it does not run into the eyes. The eyelids form a protective covering, while the fringe of eyelashes stops dust and dirt getting in. When an eyelid blinks, it wipes a film of antiseptic tears over the eye. The inner surface of both upper and lower eyelids, as well as the eyes themselves, are covered by a transparent membrane called the conjunctiva. This helps to keep the eyes moist so that they can move freely.
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AMSLER GRID

An Amsler grid is an image that is used to detect visual field defects. The patient looks at a spot in the centre of a grid of lines on a wall chart and the examiner moves a small ball on the tip of a thin pointer around the edge of the grid, and slowly towards its centre, as the patient tells the examiner if the ball can be seen. Patches of vision loss within the visual field may also be detected. The examiner plots the visual field on a smaller paper version of the grid during the examination. Some patients are able to plot their visual fields themselves while looking at the grid.

With amblyopia the grid may be distorted in some areas of the visual field.

BLIND SPOT

When we see something, light reaching the retina at the back of the eye stimulates nerve cells, which convert it into electrical impulses and send these along the optic nerve to the brain. The point where the optic nerve leaves the retina has no light sensitive cells and so forms a blind spot. You can find your blind spot by the following simple test.

Hold this page at arms length and close your left eye. Look at the cross with the right eye, and move the page slowly towards you. When the dot disappears, its image has fallen on the blind spot of the right eye.

+  

COLOUR BLINDNESS TEST

Colour blindness is tested by means of specially designed plates with patterns made up of different coloured dots. This is known as the Ishihara test, after Shinobu Ishihara, the Japanese ophthalmologist who first devised this sophisticated system of coloured dots in 1917.

To test whether a patient can distinguish between red and green, they will be shown a page covered with dots in various shades of green with a numeral in the middle composed of red dots. People who are able to distinguish between red and green will be able to pick out
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the figure, whereas the plate will appear to be all the one colour to those who can't tell the
difference between red and green. Other plates with different patterns and colour
combinations test other types of colour blindness.

EYE COLOUR

The colour of the eye is really the colour of the iris, which can contract and relax to change
the size of the pupil and therefore the amount of light entering the eye. The genetics of eye
colour is very complex as there are at least three genes that determine eye colour.

Most babies are born with blue eyes, but in most of these babies the eye colour changes
after about six months. This transformation has to do with the brown coloured protein
melanin, which also adds colour to the hair and skin. At the time most babies are born,
melanin has not been deposited in the various layers of the iris of the eye and they appear
blue. With age the iris changes colour depending on the amount of melanin. A lot of melanin
will turn the iris brown or black, a medium amount causes green eyes, and if only a small
amount is deposited the eyes stay blue.

Brown colour tends to dominate over green and both dominate blue. The inheritance of
grey and the multiple shades of other iris colours is a complex interaction.

As a guide, the following pattern of inheritance is usual, but certainly not absolute, and it is
not possible to be definite about parentage by comparing eye colours.

<table>
<thead>
<tr>
<th>EYE COLOUR</th>
<th>MOTHER</th>
<th>FATHER</th>
<th>CHILD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blue</td>
<td>Blue</td>
<td>Blue</td>
<td>Blue or brown</td>
</tr>
<tr>
<td>Brown</td>
<td>Blue</td>
<td>Blue</td>
<td>Brown</td>
</tr>
<tr>
<td>Green</td>
<td>Blue</td>
<td>Blue</td>
<td>Green</td>
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<tr>
<td>Blue</td>
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Red or pink eyes are due to albinism in which there is a complete lack of any melanin or
colouration in the iris.

Heterochromia is the condition in which the eyes are different colours. This may be due to
an injury or disease (eg. Fuchs uveitis syndrome of the iris, or due to an inherited trait.

A blue cornea may be a sign of brittle bones (osteogenesis imperfecta) and a bright blue
lacy iris occurs with Williams syndrome.

HORIZONTAL LIGHT TEST

The horizontal light test is a test for glaucoma and involves a doctor shining a bright narrow
beam of light horizontally onto the eye from its outer aspect, and observing the shadow cast
on the iris (coloured part of the eye).

If the iris is evenly illuminated or the shadow is on the outer side only, glaucoma is unlikely.
If the shadow falls on the nose side of the iris, there is a significant potential for acute
glaucoma. The iris is normally concave, but when convex due to increased pressure within
the eye it casts a shadow on the nose side of the eye.
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OPTIC DISC
The optic disc is the slightly off-centre point in the retina at the back of the eye where the optic nerve enters the eye. Because of all the nerve fibres congregating at this point, there is no room for any light sensitive cells, so this area has no vision and forms the blind spot. There is a depression in the centre of the optic disc known as the optic cup.

When the eye is examined through an ophthalmoscope or slit lamp, abnormalities of the optic disc may be noted. The presence of extra blood vessels (increased vascularity) may be due to optic neuritis. Bulging of the disc (papilloedema) may be due to increased pressure within the skull from a brain injury, infection or tumour. Atrophy (reduced number of arteries) may be due to optic nerve disease. A pale disc can be caused by a central retinal artery occlusion (blood clot in the artery supplying the optic nerve), while haemorrhages (bleeding areas) on the optic disc may be due to a central retinal vein occlusion.

OPTIC CUP/DISC RATIO
The optic cup is a depression within the pale optic disc on the retina at the point where the optic nerve enters the eye. It is the site of the blind spot. The optic cup/disc ratio is the ratio of the vertical diameter of the optic cup to that of the optic disc, measured on ophthalmoscopic or slit lamp examination of the retina.

The normal optic cup/disc ratio is 0.2. A result of less than 0.5 is probably normal, but a ratio of more than 0.5 is suspicious of glaucoma, and a ratio of more than 0.8 is usually diagnostic of glaucoma. Rising pressure within the eye with glaucoma flattens and enlarges the optic cup.

SNELLEN CHART
A Snellen chart is one on which letters and symbols of varying sizes are printed. It is used to test vision at a standard distance (either six or three metres) and the results are written as 6/6 (perfect vision), 5/6 (better than average vision), 12/6 (minimal vision acceptable for activities such as driving) and 60/6 (legally blind). In the USA imperial measures (feet) are used and 6 metres becomes 20 feet, and perfect vision becomes 20/20.
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It is named after the Dutch ophthalmologist Hermann Snellen (1834-1908).

SWINGING TORCH SIGN

The swinging torch sign can be an indication of glaucoma (more advanced in affected eye), optic nerve demyelination (damage to nerve insulation layer), an eye tumour or other eye disease.

In a darkened room, shining a torch in a normal eye results in an immediate constriction of the pupils in both eyes. Swinging the torch to the affected eye causes initial dilatation of both eye pupils with subsequent constriction of the pupils. Swinging the torch back to normal eye results in immediate pupil constriction again.

As the light shifts from the less to the more diseased eye, the direct stimulus passes along the more damaged optic nerve but this stimulus is now no longer of sufficient in intensity to keep the pupils as small as they had been when the better eye was illuminated, so both pupils dilate.

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