Anterior Segment

8

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8.1 Anterior Segment Examination Sequence

- Introduce yourself to patient
- Set patient up on slit lamp
- Examine lower eyelid — punctum first (ectropion, plug) then eyelashes and eyelid margin for anterior or posterior blepharitis, slide finger across lid to look for lumps
- Ask the patient to look up and pull lower eyelid down and look at conjunctiva for scars, symblepharon
- Ask patient to look down and lift upper eyelid — look for punctum first (ectropion, plug) then eyelashes and eyelid margin for anterior or posterior blepharitis, slide finger across lid to look for lumps
- Look at upper bulbar conjunctiva for blebs/tubes

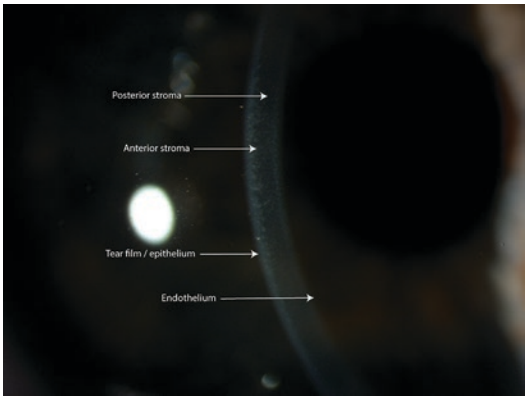

Fig. 8.1 Anterior segment image of a slit beam of the cornea

- Ask the patient to look to the right to look at temporal conjunctiva and then to the left to look at nasal conjunctiva
- Ask the patient to look straight ahead and then examine cornea (epithelium, anterior stroma, posterior stroma, thickness of cornea, endothelium — see Fig. 8.1)
- Examine AC for cells
- Examine iris for PI, areas of atrophy (transillumination), PXF, PS, ectropion uveae, nodules, comment on any corectopia
- Examine lens and comment on position, aphakia, pseudophakia, PCO, previous Yag capsulotomy, cataract and its density, as may apply
- Examine anterior vitreous for cells, degenerations

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8.2 Epithelial Basement Membrane Dystrophy (EBMD) (Fig. 8.2 and Table 8.1)

8.2.1 History

- History of RCES without previous trauma

8.2.2 Examination

- Findings should be present in both eyes
- Grayish-white maplike opacities in the superficial corneal epithelium ("maps") — characteristic and pathophysiologic feature and represent corneal epithelial basement membrane present within the epithelium itself — best seen on retroillumination
- Parallel fingerprint-like lines in the corneal epithelium ("fingerprint") — represent parallel rows of thickened basement membrane within the epithelium — best seen on retroillumination

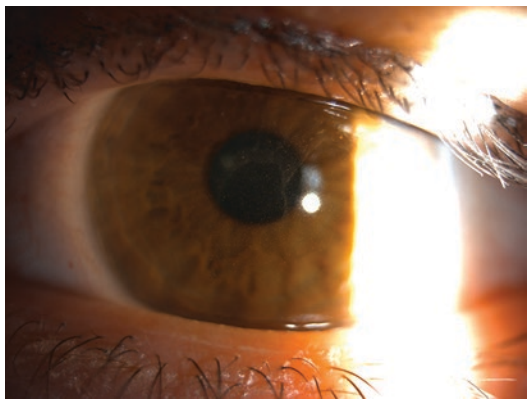

Fig. 8.2 Anterior segment image of a patient with the typical "dot" and "map" changes associated with EBMD

Table 8.1 Theoretical pathogenesis of EBMD

• Epithelial cells produce abnormal multi-laminar basement membrane, both in normal location and intraepithelially, as the intraepithelial basement membrane thickens it blocks normal migration of epithelial cells toward the surface, trapped epithelial cells degenerate to form intraepithelial microcysts, abnormal intraepithelial basement membrane produces map and fingerprint changes

- Intraepithelial gray microcysts (most unusual finding — "dot") — represents individual small mounds of thickened basement membrane beneath the basal layer of epithelium — best viewed by retroillumination

8.2.3 Treatment

- Management of RCES: lubricants, cycloplegia, patching, BCL insertion, mechanical debridement of the loosened epithelium, anterior stromal puncture for recalcitrant recurrent erosions below the visual axis (80% first time success rate), PTK for recurrent erosions in the visual axis — superficial ablation just into Bowman's layer ensuring that all abnormal epithelial basement membrane is removed, alcohol delamination

8.2.4 Other Diagnoses to Consider

- Meesman's dystrophy (see Sect. 8.3)
- Corneal dystrophies of Bowman's layer: Reis Bucklers/Thiel-Behnke dystrophy (see Sect. 8.4)

8.3 Meesman Dystrophy (Fig. 8.3 and Table 8.2)

8.3.1 History

- Positive family history: AD condition
- Symptoms: reduction of vision, photophobia

8.3.2 Examination

- Multiple diffusely distributed tiny vesicles that extend all the way to the limbus
- Vesicles are more numerous in the interpalpebral area and are best visualised with retroillumination
- The epithelium adjacent to the cysts is clear, but there can be whorled and wedge-shaped epithelial patterns
- Findings are bilateral and limited to the corneal epithelium

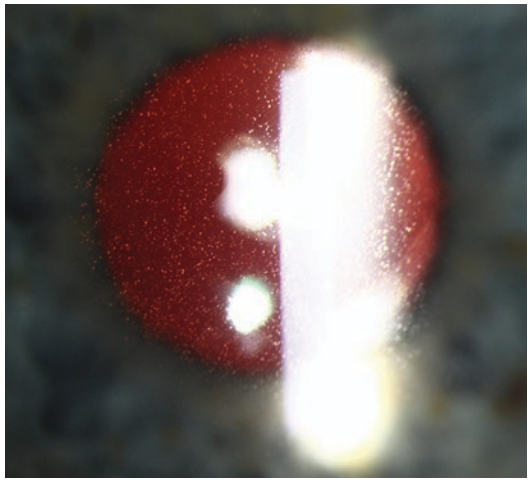

Fig. 8.3 Anterior segment image of a patient with Meesman corneal dystrophy showing the typical tiny vesicles distributed diffusely across the corneal epithelium

Table 8.2 Key facts of Meesman dystrophy

- A type of corneal epithelial dystrophy — defined as a corneal opacity or alteration which is most often bilateral and progressive, occurs after birth, and is not inflammatory
- AD condition caused by mutations in the genes for cytokeratins CK3 and CK12
- Microcysts contain "peculiar substance" — an electron dense accumulation of granular and filamentary material

8.3.3 Treatment

- Rarely required as patients are usually asymptomatic or have minimal symptoms
- Indications for treatment: significant photophobia or visual impairment
- Treatment options
  - Superficial keratectomy
  - Excimer laser phototherapeutic keratectomy
  - Lamellar keratoplasty: DALK

8.3.4 Other Diagnoses to Consider

- "Dot" changes of epithelial basement membrane dystrophy (irregular round, oval, or comma-shaped, non-staining, gray-white intraepithelial opacities of various sizes in close proximity to the maplike patches): hx of recurrent corneal erosions (pain, lacrimation, photophobia), dots alone are never seen — look for maps (geographic circumscribed gray lesions best seen with broad oblique illumination) and fingerprint lines (branching refractile lines with club-shaped terminations — best seen in retroillumination)
- Lisch epithelial corneal dystrophy — x-linked dominant (positive FHx), densely crowded transparent epithelial microcysts in a feathery whorled pattern

8.4 Reis-Buckler Dystrophy (Fig. 8.4 and Table 8.3)

8.4.1 History

- History of RCES since childhood

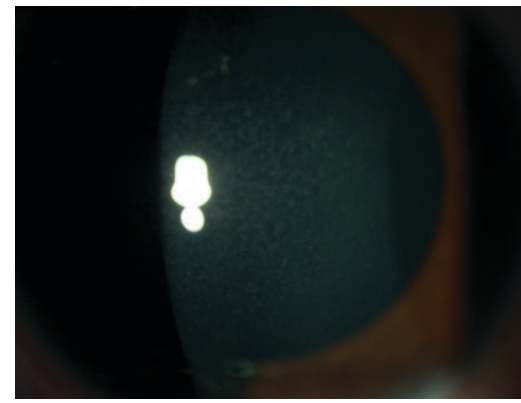

Fig. 8.4 Anterior segment image of a patient with Reis-Buckler dystrophy with typical honeycomb shaped opacities in the Bowman's layer

Table 8.3 Key facts of reis-buckler dystrophy

- A corneal dystrophy of Bowman's layer
- AD condition caused by mutations in the keratoepithelin gene (TGFB1)
- Characterised by replacement of Bowman's layer with a fibrocellular scar tissue (stains blue with Masson trichrome)

8.4.2 Examination

- Honeycomb shaped opacities in the Bowman's layer involving the center of the cornea with/without scarring.

8.4.3 Treatment

- RCES: lubricants, mechanical debridement, BCL with prophylactic topical antibiotics, anterior stromal puncture, PTK, alcohol delamination
- If vision reduced: PTK, lamellar or penetrating keratoplasty

8.4.4 Other Diagnoses to Consider

- Thiel-Behnke corneal dystrophy: distinguished from Reis-Bucklers by electron microscopy looking a fibrocellular scar that is characterised by curly collagen fibers and a saw-tooth configuration.

8.5 Lattice Dystrophy (Fig. 8.5 and Table 8.4)

8.5.1 History

- History of RCES
- Family history of lattice dystrophy

8.5.2 Examination

- Refractile radially orientated branching lines that overlap one another creating a latticework pattern ± a diffuse central anterior stromal haze
- Examine for facial paresis (lagophthalmos and exposure keratopathy), blepharochalasis, and laxity of the facial skin (Type II)

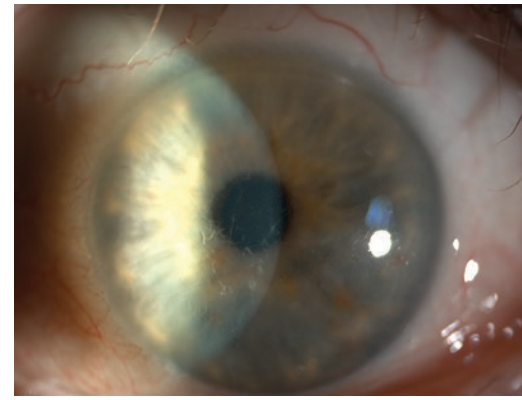

Fig. 8.5 Anterior segment image of a patient with lattice dystrophy with the typical refractile radially orientated branching lines that overlap one another

Table 8.4 Key facts of lattice dystrophy

- Bilateral AD inherited primary localised corneal amyloidosis with progressive deposition of amyloid in the corneal stroma
- Type I: mutation in the TGFβ1 gene on chromosome 5, isolated to the eye
- Type II (Meretoja's syndrome): mutation in the gelsolin gene, ocular and systemic features (cardiac conduction abnormalities, peripheral neuropathy)
- Amyloid stains with Congo red and demonstrate apple green birefringence and dichroism at polarising microscopy

8.5.3 Treatment

- RCES: topical lubricants, BCL, patching, anterior stromal puncture, PTK, alcohol delamination
- VA impaired: lamellar or penetrating keratoplasty (highest rate of recurrence in lattice dystrophy compared to granular and macular dystrophy)
- Refer to medics If type II suspected for assessment of systemic involvement

8.5.4 Other Diagnoses to Consider

- Granular dystrophy (see Sect. 8.6)
- Macular dystrophy (see Sect. 8.7)

- Avellino dystrophy (see Sect. 8.8)
- Schnyder's crystalline dystrophy (see Sect. 8.9)

8.6 Granular Dystrophy (Fig. 8.6 and Table 8.5)

8.6.1 History

- Positive family history
- History of RCES

8.6.2 Examination

- Small discrete sharply demarcated grayish white opacities in the anterior central stroma with intervening clear stroma and sparing of the periphery
- Opacities vary in shape and can be grouped into three basic morphologic types: drop-shaped, crumb-shaped, and ring shaped.

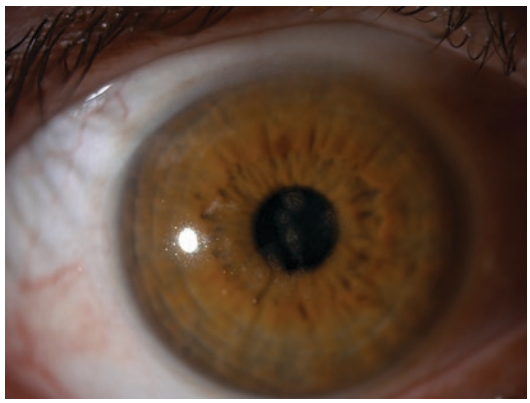

Fig. 8.6 Anterior segment image of a patient with granular dystrophy, with typical multiple white anterior stromal opacities with clear intervening stromal spaces between each opacity

Table 8.5 Key facts of granular dystrophy

- AD condition involving the deposition of hyaline material in the corneal stroma (hyaline material in the stroma stains red with Masson trichrome)
- Caused by a mutation in the keratoepithelin gene (TGFB1)

8.6.3 Treatment

- RCES: lubricants, BCL with prophylactic topical antibiotics, mechanical debridement, anterior stromal puncture, PTK, alcohol delamination
- If vision reduced: keratoplasty (PK or DALK) but high chance of recurrence

8.6.4 Other Diagnoses to Consider

- Macular dystrophy (see Sect. 8.7): recessive family history, decreased CCT, involvement of peripheral stroma, early intervening stromal haze
- Avellino dystrophy (see Sect. 8.8): combination of lattice dystrophy and granular dystrophy
- Schnyder's crystalline dystrophy (see Sect. 8.9)

8.7 Macular Dystrophy (Fig. 8.7 and Table 8.6)

8.7.1 History

- History of RCES

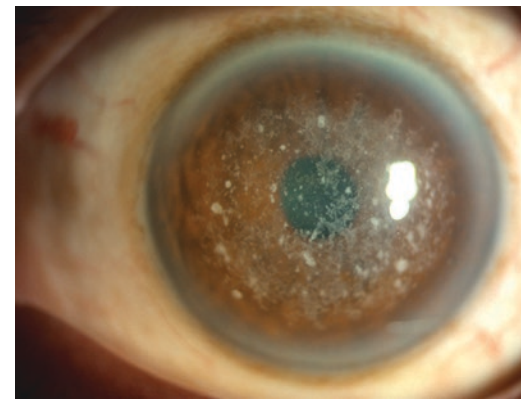

Fig. 8.7 Anterior segment image of a patient with macular dystrophy, showing multiple white opacities in the anterior stroma on a background of diffuse haze of the corneal stroma

Table 8.6 Key facts of macular dystrophy

- AR disorder involving the deposition of a GAG (keratin sulphate) in the stroma (GAG stains with Alcian blue or colloidal iron)
- Arises from mutations in the gene for carbohydrate sulfotransferase (CHST6)

8.7.2 Examination

- Multiple gray-white opacities in the anterior stroma on a background of diffuse clouding in the anterior stroma, extending from limbus to limbus. Guttae are commonly present.
- Associated with reduced CCT

8.7.3 Treatment

- RCES: lubricants, mechanical debridement, BCL with prophylactic topical antibiotics, anterior stromal puncture, PTK, alcohol delamination
- Photophobia: tinted glasses or tinted cosmetic CL
- Reduced vision: keratoplasty (DALK or PK)

8.7.4 Other Diagnoses to Consider

- Granular dystrophy (see Sect. 8.6)
- Avellino dystrophy (see Sect. 8.8)
- Schnyder's crystalline dystrophy (see Sect. 8.9)

8.8 Avellino Dystrophy (Fig. 8.8)

8.8.1 History

- History of RCES

8.8.2 Examination

- Anterior stromal, discrete gray-white granular deposits with anterior stromal haze
- Mid to posterior stromal lattice lesions

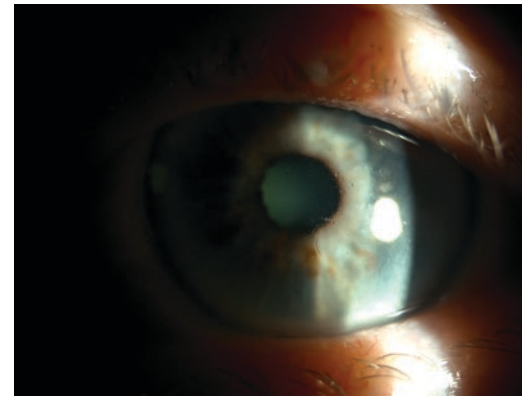

Fig. 8.8 Anterior segment image of a patient with Avellino dystrophy showing signs of both lattice dystrophy and granular dystrophy

8.8.3 Treatment

- RCES: lubricants, mechanical debridement, BCL with prophylactic topical antibiotics, anterior stromal puncture, PTK, alcohol delamination
- Photophobia: tinted glasses or tinted cosmetic CL
- Reduced vision: PK

8.8.4 Other Diagnoses to Consider

- Lattice dystrophy/Granular dystrophy

8.9 Schnyder's Crystalline Dystrophy (SCD) (Fig. 8.9 and Table 8.7)

8.9.1 History

- History of hypercholesterolaemia
- History of paraproteinaemias

8.9.2 Examination

- Bilateral central refractile crystals/opacities in the central anterior stroma with associated arcus lipoides

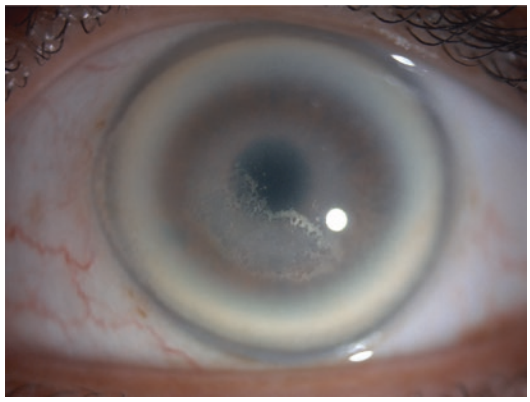

Fig. 8.9 Anterior segment image of a patient with Schnyder's crystalline dystrophy showing subepithelial crystalline deposition

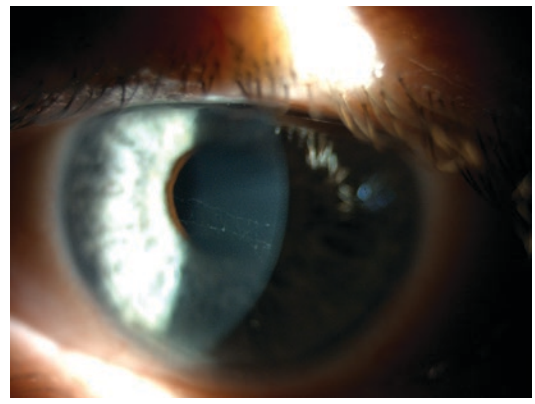

Fig. 8.10 Anterior segment image of a patient with PPCD with typical horizontal band lesions

Table 8.7 Key facts of SCD

• AD stromal dystrophy caused by mutations in the UBIAD1 gene • Characterised by the deposition of cholesterol crystals and phospholipids in the corneal stroma (crystals stain red with Oil Red O)

Table 8.8 Key facts of PPCD

• Bilateral, AD inherited corneal dystrophy • Normally non-progressive although has potential of decreasing vision due to corneal oedema and glaucoma

- Diffuse stromal haze, which may affect all levels of the stroma

8.9.3 Investigations

- Bloods: fasting cholesterol and triglyceride levels (SCD), leucocyte cysteine content (cystinosis), lymphocytosis (leukaemia)

8.9.4 Treatment

- If visual problems: keratoplasty (lamellar or penetrating) although recurrence of cholesterol crystals may occur

8.9.5 Other Diagnoses to Consider

- Cystinosis
- Dysproteinaemia's: multiple myeloma, Waldenstrom's macroglobulinaemia, Hodgkin's disease, leukaemia, benign monoclonal gammopathy, cryoglobulinaemia
- Medications: gold
- Bietti's peripheral crystalline dystrophy — peripheral location of crystals + retinal degeneration

8.10 Posterior Polymorphous Corneal Dystrophy (PPCD) (Fig. 8.10 and Table 8.8)

8.10.1 History

- History of Alport syndrome: 85% XR condition, anterior lenticonus, sensorineural deafness, dot and fleck retinopathy, glomerulonephritis

8.10.2 Examination

- Vesicular-like lesion (hallmark of PPCD): appears as sharply demarcated transparent round cysts surrounded by a grey halo at the

level of DM and endothelium and commonly occur in lines or clusters
- Band lesions: typically horizontal, have parallel scalloped edges, and do not taper towards the ends
- Diffuse deep stromal opacities: small, macular, gray-white lesions at the level of DM. There may be deep stromal haze adjacent to these lesions
- Endothelial guttae
- Corneal oedema occurs infrequently and ranges from minimal stromal thickening to bullous keratopathy
- PAS (characteristic feature of PPCD) seen with or without gonioscopy, corectopia, areas of iris atrophy
- Check IOP — open (compression of TM secondary to high iris insertion) and angle closure (endothelial cell migration across the TM onto the iris forming synechiae) glaucoma
- Look for trabeculectomy or glaucoma drainage implant
- Fundus examination: CDR (glaucoma), dot and fleck retinopathy (Alport syndrome)

8.10.3 Investigations

- Bloods: deranged renal function tests — glomerulonephritis — Alport syndrome
- Urinalysis: haematuria and proteinuria — glomerulonephritis — Alport syndrome
- Hearing test: sensorineural hearing loss — Alport syndrome

8.10.4 Treatment

- Observation: in the great majority of patients, PPCD is stable and asymptomatic
- Treat any glaucoma present: medical/surgical intervention
- PK: if significant reduced vision — presence of PAS visible without gonioscopy and increased IOP must be considered relative contraindications to corneal transplantation

8.10.5 Other Diagnoses to Consider

- ICE syndrome — unilateral, sporadic occurrence, female predilection, always has iridocorneal adhesions, displays iris atrophy, always has glaucoma, progressive and symptomatic
- Posterior corneal vesicle syndrome
- Early-onset CHED
- Congenital glaucoma
- Axenfeld-Rieger syndrome

8.11 Fuchs Endothelial Dystrophy (FED) (Figs. 8.11, 8.12 and Table 8.9)

8.11.1 History

- Symptoms: reduced vision (most predominantly in the morning)

8.11.2 Examination

- Corneal guttae ± melanin pigment pigmentation — appear as dark spots on the posterior corneal surface by direct illumination (beaten

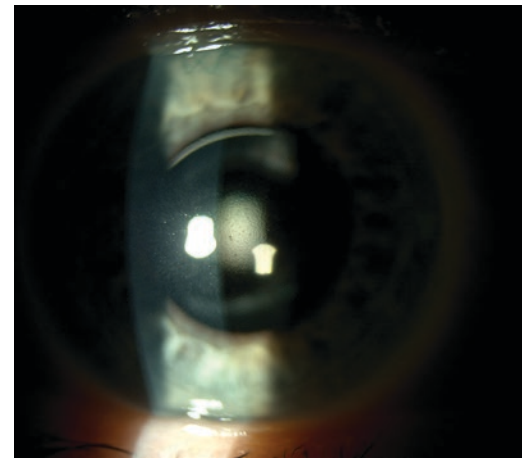

Fig. 8.11 Anterior segment image of a patient with FED showing multiple corneal guttae

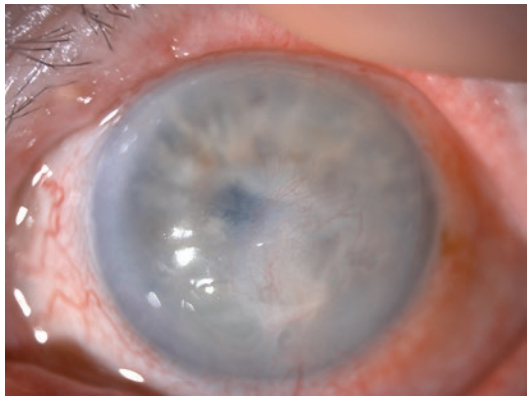

Fig. 8.12 Anterior segment image of a patient with end stage FED with diffuse corneal oedema and vascularised subepithelial fibrosis

Table 8.9 Key facts of FED

• Age of onset over age 50
• Female: Male ratio of 3:1

metal appearance — coalescence of central guttae)
- Corneal stromal oedema with DM folds, microcystic epithelial oedema (seen as a stippled pattern that stands out in sclerotic scatter, using fluorescein stain the microcystic pattern is highlighted as a disruption in the tear film) that eventually coalesces to form bullae, which can lead to erosions and fingerprint lines.
- In end stage disease, subepithelial fibrous scarring occurs between the epithelium and Bowman's membrane (best seen with tangential illumination)
- Measure IOP and examine the optic disc for glaucoma: increased incidence of primary open angle glaucoma (POAG) in FED

8.11.3 Investigations

- Specular microscopy — reduced endothelial cells counts, increased average cell volume, loss of hexagonality of the cells and increased variation of cell sizes
- Pachymetry — increased CCT

8.11.4 Treatment

- Medical
  - Indication(s): visually significant corneal oedema.
  - Options:
    Topical hypertonic saline solutions
    Dehydration of cornea by a blow dryer in the morning or throughout the day
    Reduction of IOP (avoid topical carbonic anhydrase inhibitors)
    BCL for treatment of recurrent erosions caused by epithelial bullae
- Surgical
  - Indication(s): blurring of vision in the morning, evidence of epithelial oedema on slitlamp examination
  - Options:
    Lamellar keratoplasty (DSEK, DMEK)
    Full thickness PK — preferred if significant corneal scarring present
- FED and cataract (Table 8.10)
  - Staged cataract extraction followed by keratoplasty:
    Advantages: IOL stability in the bag, easier to manage if complications occur, delay of graft procedure
    Disadvantages: two operative procedures on separate occasions required for patient, longer visual rehabilitation procedure for patient
  - Triple procedure (keratoplasty with cataract extraction and IOL implant):
    Indications(s): consider if CCT more than 650, endothelial cell count <1000, dense cataract present

Table 8.10 Management of a patient with FED and a cataract

• If the patient has no cataract and mild corneal oedema — consider conservative medical management
• If the patient has a dense cataract and severe corneal oedema — consider triple procedure
• If the patient has no cataract but has severe corneal oedema — consider keratoplasty first and cataract extraction if a visually significant cataract develops
• If the patient has a dense cataract and no corneal oedema — consider cataract extraction first and keratoplasty if corneal decompensation develops

Advantages: faster visual recovery and reduced number of operations
Disadvantages: may be unable to perform keratoplasty (with subsequent loss of valuable graft tissue) if complications arise from initial cataract surgery

8.11.5 Other Diagnoses to Consider

- Pseudophakic bullous keratopathy
- Chandler's syndrome — unilateral ICE syndrome with a hammered silver appearance of the corneal endothelium, corectopia, iridocorneal adhesions
- Posterior polymorphous dystrophy — corectopia, iridocorneal adhesions
- Central herpetic disciform keratitis — presence of underlying KP's
- Macular dystrophy
- Congenital hereditary endothelial dystrophy — present at birth (CHED 2 — AR)
- Corneal pseudoguttae: post-trauma, post-intraocular inflammation — these apparent guttae are transient and disappear with resolution of underlying condition

8.12 Interstitial Keratitis (IK) (Fig. 8.13 and Table 8.11)

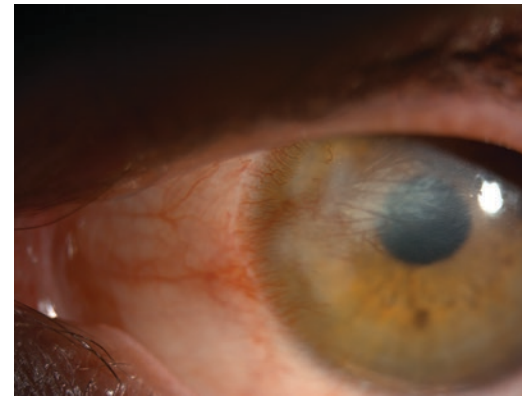

Fig. 8.13 An anterior segment image of a patient with interstitial keratitis. Note the ghost vessels in the corneal stroma arising from the limbus

Table 8.11 Key facts of IK

• Defined as a nonsuppurative inflammation characterised by cellular infiltration and vascularisation of the corneal stroma with minimal primary involvement of the corneal epithelium or endothelium

8.12.1 Causes of IK

- Infectious: bacterial (congenital — if bilateral or acquired — if unilateral syphilis, TB, leprosy, lyme disease, trachoma, brucellosis), viral (HSV, HZV, EBV, mumps, rubeola, HTLV-1), parasitic (acanthamoeba keratitis, microsporidiosis, leishmaniasis, onchocerciasis, trypanosomiasis)
- Non-infectious: cogan's syndrome, sarcoidosis, lymphoma

8.12.2 History

- Syphilis: history of congenital syphilis or hx of parents being treated for STI, symptoms of meningism (headaches, photophobia, neck stiffness), hearing loss
- HSV: History of HSV keratitis, hx of herpetic "cold sores"
- HZV: Risk factors for immunosuppression: immunosuppressive drugs, organ transplant recipients, neoplastic diseases, HIV
- Acanthamoeba: History of CL wear (extended wear CL), poor CL hygiene (rinsing in tap water), swimming with CL in situ (ponds, hot tubs, swimming pools), corneal trauma (rural or agricultural setting)
- TB: fever, night sweats, weight loss, cough, hx of TB, recent travel to endemic areas, contacts of active TB
- Cogan's syndrome: acute tinnitus, vertigo (dizziness), bilateral hearing loss

8.12.3 Examination

- Vascularised corneal opacification is a sign of prior stromal inflammation. Look for ghost vessels (a tracery of intertwined phantom ves-

sels forming a thread like network weaving through a stromal haze) from the limbus
- Syphilis: bilateral in congenital and unilateral in acquired, cellular infiltration of stroma with stromal oedema, stromal neovascularisation (superficial capillaries bud from venules of the limbal arcades, deeper vessels arise as terminal branches of the anterior ciliary vessels), anterior and posterior uveitis, associated findings in congenital syphilis — salt and pepper retinopathy, Hutchinson's teeth (widely spaced teeth with notched incisors), frontal bossing, saddle shaped nose, sabre shins, associated findings in acquired syphilis — maculopapular rash (trunk, palms, soles), lymphadenopathy, painless chancre, painless ulcer of mouth or conjunctiva, light-near dissociation (Argyll Robertson pupil — neurosyphilis)
- HSV: stromal oedema (suggested by increased width of slit beam and DM folds), stromal neovascularisation, stromal cellular infiltration (three hallmarks of IK) ± scleritis or uveitis, pattern of stromal infiltration may be central or peripheral, focal or multifocal, superficial or full-thickness, wessely-type immune rings (if present supports the diagnosis — best seen on sclerotic scatter), look for anterior stromal scars (footprints or ghost dendrites) from previous HSV disease, raised IOP, sectoral iris atrophy, check for reduced corneal sensation
- HZV: stromal infiltration with stromal oedema, pseudodendrites (lack central ulceration, have blunt ends, raised epithelial cells), vesicular eruption of periocular skin and eyelids (look for scabbed lesions), uveitis
- Acanthamoeba: stromal oedema with infiltration, ring infiltrate, perineural infiltrates, pseudodendrites
- TB: unilateral, cellular infiltration of stroma is often peripheral or quadrantic and is followed by localised oedema, and later stromal vascularisation ± scarring or thinning. May have additional uveitis, particularly choroiditis.
- Cogan's syndrome: patchy cellular infiltration of the mid-stroma with stromal oedema, stromal neovascularisation from the limbus, stromal scarring and ghost vessels (late-finding)

8.12.4 Investigations

- Syphilis: non-treponemal tests (VDRL, RPR — better for screening), treponemal tests (FTA-ABS, hemagglutination, enzyme immunoassay — ELISA), HIV test (check for co-infection, HIV antibody detection via ELISA. Positive ELISA results are confirmed with western blot), LP (if active intraocular disease or neurosyphilis — leukocytosis, raised protein levels). Treponemal tests remains reactive for life in patients with congenital or acquired syphilis. Over time, the non-treponemal test titres decrease, ultimately becoming undetectable.
- Acanthamoeba: corneal scrape and stain with calcofluor white (stain cysts visualised under UV light) and culture on non-nutrient agar with E. coli overlay ± PCR, in-vivo confocal microscopy with direct visualisation of cysts
- TB: CXR, tuberculin skin test, Interferon gamma release assay (IGRA — T-spot or QuantiFERON), sputum culture for AFB, early morning urine sample for AFB, corneal scraping for culture and histopathological evaluation
- Cogan's syndrome: bloods for FBC (leukocytosis, eosinophilia), ESR (raised), p-ANCA (PAN), c-ANCA (GPA), hearing test (sensorineural hearing loss), VDRL/RPR (to rule out syphilis)

8.12.5 Treatment

- Syphilis: topical steroids, GU referral with contact tracing, benzylpenicillin, PK for corneal scarring
- HSV: topical steroids with topical antivirals (prevent breakout of epithelial keratitis) — no benefit of adding oral aciclovir (HEDS I), oral acyclovir 400 mg BD reduced the probability of stromal keratitis recurrence by 50% from 28% to 14% at the 1 year time point (HEDS II), cycloplegia (for uveitis), PK for corneal scarring
- HZV: topical steroids, oral aciclovir 800 mg 5×/day (IV aciclovir if patient is immunocompromised to avoid complication of disseminated infection), cycloplegia (for uveitis), PK (high risk of failure) or Boston keratoprosthesis

for corneal scarring. PK only generally performed for tectonic reasons.
- Acanthamoeba: admit, topical biguanide (PHMB 0.02% or chlorhexidine 0.02%) + diamidine (propamidine 0.1% or hexamidine 0.1%), PK for corneal scarring
- TB: topical steroids, systemic anti-TB medication (rifampicin, isoniazid, pyrazinamide, ethambutol), cycloplegia (for uveitis), PK for corneal scarring
- Cogan's syndrome: low dose topical steroids for IK, high dose systemic corticosteroids for inner ear disease

8.12.6 Other Diagnoses to Consider

- Microbial keratitis — bacterial (pseudomonas, syphilis, chlamydia), viral (HSV. HZV), fundal (candida, aspergillus, fusarium), parasitic (onchocerca)
- Traumatic corneal scarring from chemical burns
- LESC failure
- OCP
- SJS
- Terrien's marginal degeneration
- Ocular rosacea
- Atopic keratoconjunctivitis

8.13 Lipid Keratopathy (Figs. 8.14 and 8.15)

8.13.1 Causes

Secondary lipid keratopathy is more common than primary lipid keratopathy and occur in corneal neovascularisation.

- Primary: no history of trauma, corneal vascularisation, or known disorders of lipid metabolism
- Secondary: interstitial keratitis, trauma: surgical — corneal intacs implantation, corneal ulceration, corneal hydrops, mustard gas injuries, disorders of lipid metabolism (fish eye disease, tangier disease, familial LCAT deficiency, apolipoprotein A1 deficiency)

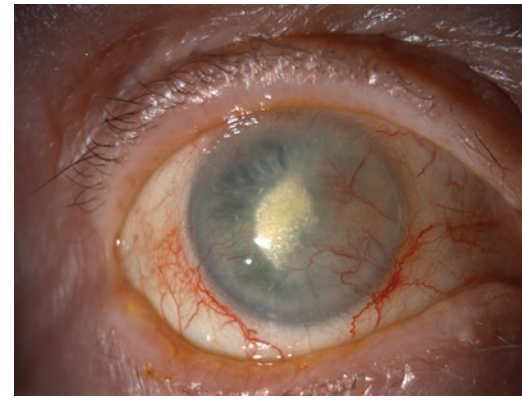

Fig. 8.14 Anterior segment image of a patient with lipid keratopathy in a region of corneal scarring and stromal neovascularisation resulting from previous herpes simplex keratitis

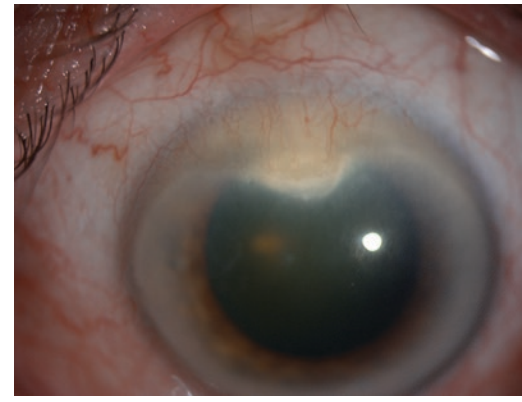

Fig. 8.15 Anterior segment image of a patient with lipid deposition at the leading edge of the superior pannus associated with Terrien marginal degeneration

8.13.2 Examination

- Primary lipid keratopathy: lipid deposition centrally or peripherally
- Secondary lipid keratopathy: gray to yellow-white infiltrates (often at edge of scars/lesions) associated with the presence of an adjacent corneal blood vessel

8.13.3 Treatment

- Indications: vision, cosmesis
- Options

- Argon laser to limbal feeder vessel or needle point cautery to induce absorption of the lipids through destruction of the feeder vessels
- Intrastromal anti-VEGF agents with off license avastin
- Keratoplasty: PK or DALK

8.14 Radial Keratotomy (RK) (Fig. 8.16 and Tables 8.12 and 8.13)

8.14.1 Examination

- Radial incision lines (from center to peripherally)
- Star shaped iron deposit forming centrally (tear star — no effect on vision)

8.14.2 Indications of RK

- Refractive surgery procedure to correct myopia (−1.00 to −4.00 D)

8.14.3 Complications of RK

- Intraoperative: penetration into AC causing iris/lens damage

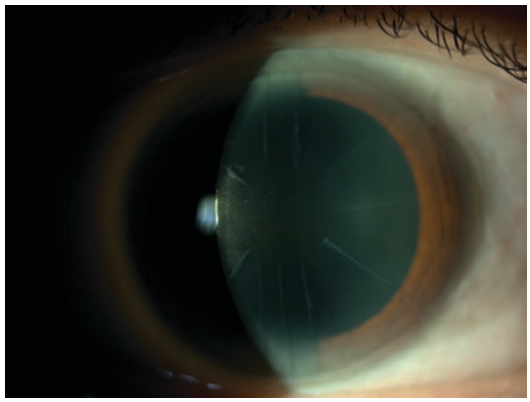

Fig. 8.16 Anterior segment image of a patient who previously underwent a radial keratotomy refractive procedure to correct an underlying myopic refractive error

Table 8.12 Cataract surgery considerations in patients who have undergone a previous radial keratotomy refractive procedure

- IOL power calculation may be problematic and may result in undercorrection and hyperopia
- Calculation of implant power for cataract surgery after RK should be done using a third-generation formula (Haigis, Hoffer Q, Holladay 2, or SRK/T) rather than a regression formula (SRK I or II)
- Keratometric power is determined in one of three ways: direct measurement using corneal topography; application of pre-RK keratometry minus the refractive change; or adjustment of the base curve of a plano contact lens by the overrefraction.
- Scleral tunnel incisions preferred (clear corneal incisions increase the risk of the blade transecting the RK incision, which can induce irregular astigmatism)
- Place incision in the steep meridian of the cornea or use of toric IOLs to reduce preoperative astigmatism
- Prevent overhydrating the cataract incision in order to avoid rupture of the RK incision

Table 8.13 Measuring intraocular pressures in patients who have undergone a previous radial keratotomy refractive procedure

- Changes in the corneal shape without corneal thinning can lead to falsely low IOP values with central applanation tonometry
- In eyes that have undergone RK, non-Goldmann measurement of IOP and continued examination of the optic nerve and possibly VF is recommended.

- Postoperative: irregular astigmatism (visual glare + distortion) due to wound gape and epithelial plugs, epithelial downgrowth, endophthalmitis, traumatic cataract, starburst effects (starburst patterns around lights at night — light scattering off radial incisions), perforation of the cornea, progressive hyperopic shift (can be corrected with LASIK or PRK)

8.15 Vortex Keratopathy (Fig. 8.17)

8.15.1 History

- Use of medications (dose and duration): amiodarone, hydroxychloroquine, chloroquine, phenothiazines (chlorpromazine), tamoxifen, indomethacin

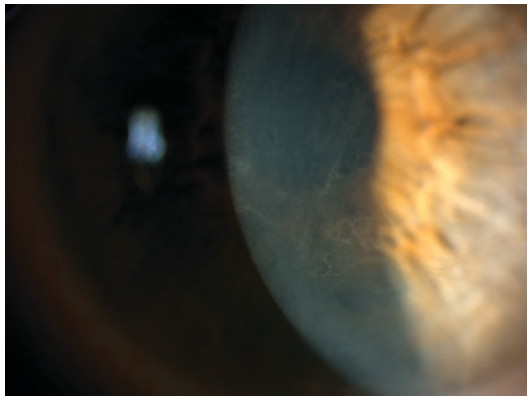

Fig. 8.17 Anterior segment image of a patient with vortex keratopathy who was on long term amiodarone treatment

- History of Fabry's disease: x-linked recessive disorder, deficiency of alpha-galactosidase A enzyme, renal failure, cardiovascular disease, neurologic changes, cutaneous angiokeratomas

8.15.2 Examination

- Pattern of non-elevated white to brown whorl-shaped opacities within the basal corneal epithelium, consisting of fine lines emanating from a central nodal point (most commonly located in the inferior paracentral region — typically bilateral and symmetric)
- Look for aneurysmal dilatations and tortuosity of the conjunctival and retinal vessels and a spoke-like posterior subcapsular cataract: Fabry's disease
- Look for bulls eye maculopathy (ring of depigmentation surrounded by an area of hyperpigmentation seen centered on the fovea): hydroxychloroquine toxicity, chloroquine toxicity
- Look for optic neuropathy (VA, RAPD, colour vision, VF), retinopathy, and anterior subcapsular lens opacities: amiodarone

8.15.3 Investigations

- Screening for greatly deficient or absent alpha-galactosidase A activity in plasma or peripheral leukocytes or gene sequencing: Fabry's disease

8.15.4 Treatment

- Cornea verticillata is visually insignificant and is not an indication for stopping any associated medications.
- Discontinuation or reduction of the dose of amiodarone should be considered if optic neuropathy is observed.

8.15.5 Other Diagnoses to Consider

- Corneal iron lines (faint yellow to dark brown discolouration in the corneal epithelium): tear star after RK, Hudson-Stahli line (horizontal line located in the lower third of the cornea), Ferry line (appears on cornea anterior to filtering bleb), Stocker's line (advancing edge of pterygium), Fleischer ring (base of the cone of KC)
- Corneal stromal deposits: gold (chrysiasis — gold to violet like fine deposits scattered from the corneal epithelium to the deep stroma ± bulbar conjunctiva), silver (argyrosis — lids and conjunctiva have a light to dark slate-grey appearance, deposits of blue-gray material in the peripheral deep stroma of the cornea), antacid, retinoid deposition

8.16 Calcific Band Keratopathy (BK) (Fig. 8.18 and Table 8.14)

8.16.1 Causes of BK

- Ocular: chronic anterior uveitis, JIA, herpetic keratouveitis, long standing interstitial keratitis, phthisis bulbi, prolonged corneal oedema, silicone oil in AC
- Systemic: hypercalcaemic states (CKD, hyperparathyroidism, sarcoidosis), hyperphosphataemia (CKD), hyperuricaemia (CKD), x-linked ichthyosis

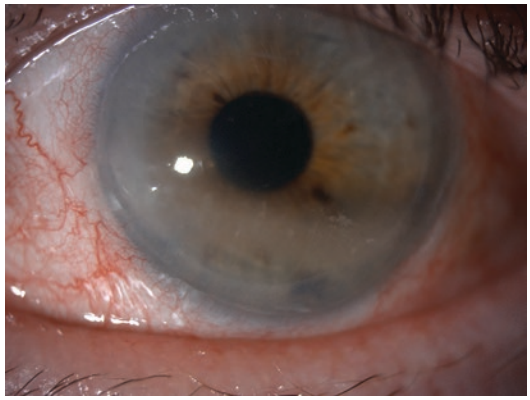

Fig. 8.18 Anterior segment image of a patient with calcific band keratopathy

Table 8.14 Key facts about BK

• Calcific BK is a deposition of calcium hydroxyapatite across the cornea at the level of Bowman's layer

8.16.2 History

- History of uveitis
- History of RD repair requiring the use of silicone oil
- History of chronic kidney disease (CKD)
- History of gout
- Symptoms suggestive of hypercalcaemia: abdominal pain, depression, renal stones
- Use of vitamin D and calcium supplements

8.16.3 Examination

- White and chalky band of opacity beginning peripherally at the 3 and 9 o'clock positions, with a sharply demarcated peripheral edge separated from the limbus by a lucent zone
- Lucent holes are scattered throughout the opacity and represent penetrating corneal nerves through Bowman's layer
- Exclude the presence of uveitis and the presence of silicone oil in AC.
- Exclude phthisis bulbi: hypotony, small eye

8.16.4 Investigations

- Bloods: U&E's, calcium, phosphorus, uric acid, ACE (if sarcoidosis suspected), PTH levels (if hyperparathyroidism suspected)
- CXR: sarcoidosis

8.16.5 Treatment

- Indication: patient becomes symptomatic with reduced VA, FB sensation, tearing or photophobia
- Options:
  - Lubricants and BCL for comfort (temporary measure)
  - Removal of epithelium followed by application of ethylenediaminetetraacetic acid (ETDA) to calcific areas. No. 15 blade used to remove any residual calcium
  - Excimer laser phototherapeutic keratectomy (PTK)

8.16.6 Other Diagnoses to Consider

- Spheroidal degeneration (climatic droplet keratopathy): bilateral golden-yellow globular deposits within the interpalpebral area, deposits located within Bowman's layer and the anterior stroma
- Urate keratopathy: brown scintillating crystal deposits in the superficial cornea

8.17 Infectious Crystalline Keratopathy (ICK) (Fig. 8.19 and Table 8.15)

8.17.1 History

- ICK: history of keratoplasty, history of chronic use of topical steroids
- Multiple myeloma: backache
- Waldenstrom's macroglobulinaemia: epistaxis, bleeding gums

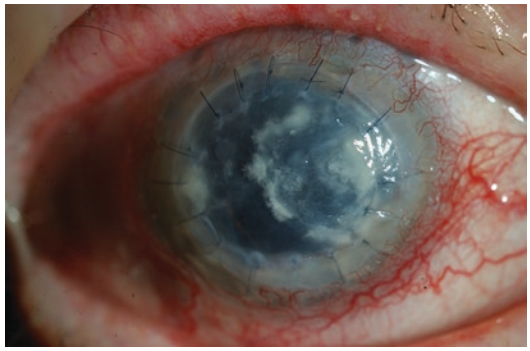

Fig. 8.19 Anterior segment image of a patient with infectious crystalline keratopathy secondary to Candida

Table 8.15 Key facts about ICK

- A non-inflammatory intrastromal microbial colonization of a graft: bacterial (*S. viridans*, *S. pneumoniae*, coagulase negative *streptococcus, P. aeruginosa*), fungal (*Candida*)
- Diagnosis of ICK can be made clinically based upon a history of keratoplasty, a history of topical steroid use, and the clinical appearance of the lesion

- Cryoglobulinamia: raynaud's phenomenon, urticaria, ulceration of the skin
- Drug history: gold

8.17.2 Examination

- ICK: crystalline fine needle like branching opacities in the anterior or mid-stroma (resembling a snowflake) occurring beneath an intact epithelium in the absence of clinically evident stromal inflammation
- Cystinosis: myriad of fine needle shaped, highly refractile opacities present in the conjunctiva and corneal epithelium, stroma, and endothelium (see Fig. 8.20)
- Multiple myeloma: prominent corneal nerves, presence of numerous scintillating polychromatic crystals scattered throughout the corneal stroma and conjunctiva, BRVO/CRVO
- Waldenstrom's macroglobulinaemia: needle-like crystals scattered throughout the corneal stromal and conjunctiva, BRVO/CRVO
- Cryoglobulinaemia: superficial scattered crystalline corneal deposits

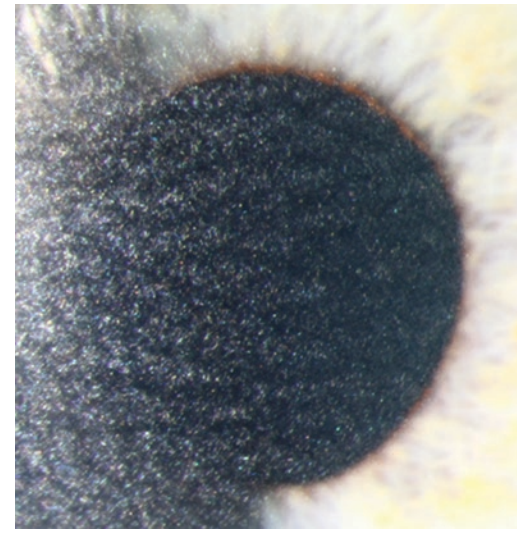

Fig. 8.20 Anterior segment image of a patient with cystinosis showing the diffuse presence of needle-shaped crystals throughout the corneal surface

- Benign monoclonal gammopathy: diffuse deposition of gray-white, yellow, gray-brown, or polychromatic iridescent dot-like opacities with a diffuse, fine stippled appearance

8.17.3 Investigations

- ICK: corneal scrapings for cultures and smears (help select appropriate antibiotics)
- Corneal biopsy: histopathologic analysis of the crystalline deposition
- Cystinosis: measure the leucocyte cysteine content, U&E's, conjunctival biopsy to measure the extracted free cysteine, electron microscopy to detect the characteristic crystals
- Multiple myeloma: bloods — hypercalcaemia and pancytopenia, XR spine — vertebral fractures and lytic punched out lesions, serum and urine electrophoresis, bone marrow biopsy

8.17.4 Treatment

- ICK: fortified intensive topical antibiotic drops — coverage for S.viridans includes top-

ical vancomycin, stop topical corticosteroids, repeat PK in order to eradicate the infection or to treat scar formation that may have caused decreased VA
- Cystinosis: oral and topical cysteamine

8.17.5 Other Diagnoses to Consider

- Lipid keratopathies: Schnyder's crystalline dystrophy, Tangier disease, familial lipoprotein disorders, lecithin-cholesterol acyltransferase (LCAT) deficiency
- Errors of protein metabolism: cystinosis, tyrosinaemia, hyperuricaemia
- Acquired immunoprotein keratopathies: multiple myeloma, benign monoclonal gammopathy, cryoglobulinaemia, Waldenstrom's macroglobulinaemia
- Drug deposition: chrysiasis

8.18 Bullous Keratopathy (Fig. 8.21 and Table 8.16)

8.18.1 Causes (Table 8.17)

- FED
- Trauma from cataract surgery
- Angle-closure glaucoma (ACG)
- PXF keratopathy
- HSV endotheliitis
- Chandler's syndrome
- PPCD

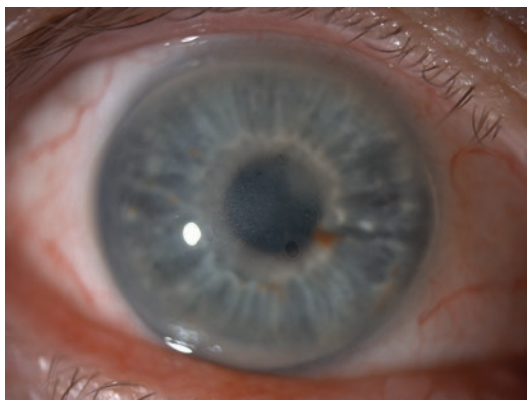

Fig. 8.21 Anterior segment image of a patient with pseudophakic bullous keratopathy, showing diffuse epithelial microcysts with several larger epithelial bullae

8.18.2 Examination

- Diffuse epithelial microcysts with the presence of adjacent epithelial bullae
- Examine for aphakia or pseudophakia: ABK/PBK
- Examine for guttae: FED
- Examine for a fixed and dilated pupil, iris atrophy (transillumination defect), presence of patent PIs (transillumination), shallow AC (using gonioscopy or Van Herrick's method — narrow slit beam at angle of 60° onto cornea just anterior to the limbus — narrow if distance between iris and posterior cornea is <¼ of the corneal thickness), glaukomflecken, increased CDR, defects on HVF testing: ACG
- Examine for whitish dandruff-like material on pupillary margin and anterior lens capsule centrally and peripherally with clear intermediate zone (if phakic), peripupillary transillumination defects, iridodonesis/phaco- or pseudo-phacodonesis: PXF
- Examine for KP's overlying stromal and epithelial oedema (pattern of KP's and distribution of oedema — diffuse, disciform, linear) and anterior uveitis: HSV endotheliitis (stromal infiltrate and neovascularisation, which

Table 8.16 Characteristics of bullous keratopathy

• Bullous keratopathy is characterised by corneal oedema with formation of epithelial bullae. It is secondary to loss or dysfunction of the corneal endothelial cells

Table 8.17 Causes of pseudophakic bullous keratopathy

• Pre-operative factors: pre-existing endothelial disease — FED, PXF, trauma, ACG, HSK
• Intraoperative factors: phacoemulsification in the AC, IOL-to-cornea touch, vitreous loss ± nuclear loss, instrumentation, ultrasound damage
• Postoperative factors: retained lens fragment in the AC, IOL dislocation — touch, vitreous-to-endothelial touch, flat AC (immediate with wound leakage or delayed with pupil block) with IOL touch to cornea, chronic post-op uveitis, ACIOL (chronic uveitis, IOL-to-cornea — touch)

are signs of stromal inflammation, are absent in endotheliitis)

8.18.3 Treatment

- Medical management: topical corticosteroids for inflammation, reduction of IOP if raised, hypertonic agents (5% NaCl), BCL, anterior stromal puncture (form adhesions between epithelium and stroma, decreasing formation of bullae) — reserved for eyes that have poor visual potential or are poor surgical candidates
- Surgical management: repositioning or exchange of IOL, if IOL subluxed or dislocated, removal of retained lens fragment if present, endothelial keratoplasty when visual loss and discomfort becomes medically untreatable, conjunctival flap — reserved for eyes with poor visual potential or patients who are not candidates for corneal transplantation

8.19 Salzmann Nodular Degeneration (Fig. 8.22 and Table 8.18)

Table 8.18 Key facts about Salzmann nodular degeneration

- Dense collagen plaques with hyalinisation are located between the epithelium and Bowman's layer
- Female predilection

8.19.1 Causes

- Vernal keratoconjunctivitis
- Phlyctenular keratitis
- IK
- Post-corneal surgery
- MGD/Ocular Rosacea
- Trachoma

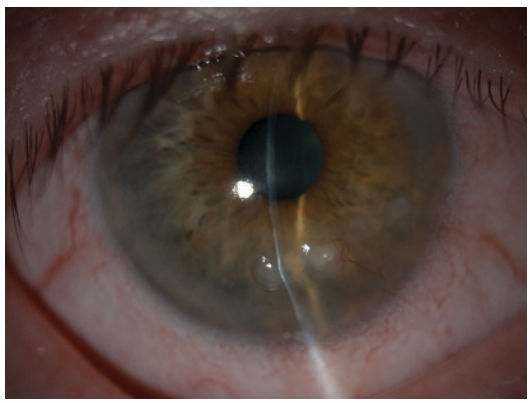

Fig. 8.22 Anterior segment image of a patient with Salzmann nodular degeneration with multiple elevated nodules inferiorly

8.19.2 History

- History of allergic eye disease, dry eye, rosacea, or trachoma
- History of previous corneal surgeries

8.19.3 Examination

- Single or multiple white or blue elevated nodular lesions adjacent to areas of corneal scarring or a corneal pannus
- An iron line may be seen at the edge of the nodules (indicate chronicity)
- Examine for rosacea
- Examine for ghost vessels
- Examine everted upper eyelid for papillary changes, Horner-trantas dots (VKC)
- Examine everted upper eyelid for Herbert pits or Arlt line

8.19.4 Treatment

- Indications: comfort and vision
- Options:
  - Lubricants and BCL for comfort
  - Superficial keratectomy by manual dissection or PTK for nodules involving visual axis and causing decreased vision

8.19.5 Other Diagnoses to Consider

- Corneal keloids: more frequently in men, younger age group, occur post-trauma or in

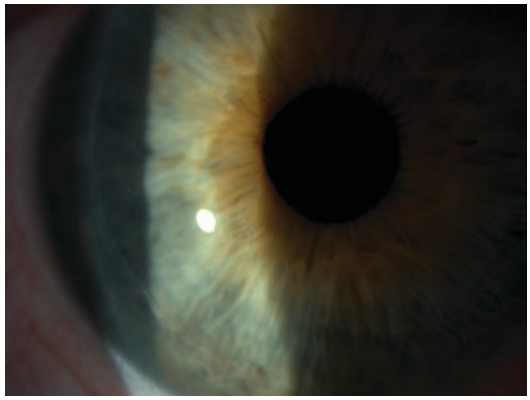

Fig. 8.23 Anterior segment image of a patient who previously had a LASIK refractive procedure to correct myopia. The LASIK flap is seen as a fine white line (concentric with limbus) in the anterior corneal stroma

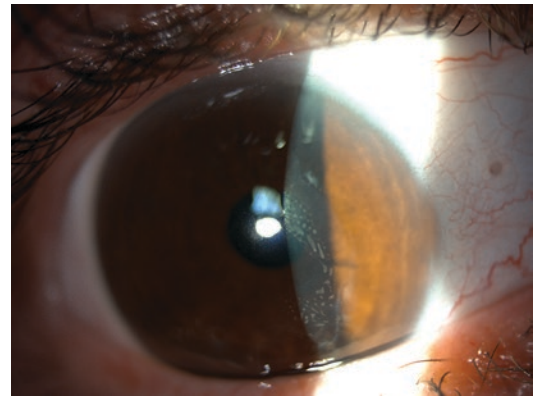

Fig. 8.24 Anterior segment image of a patient who previously had a LASIK refractive procedure, which has been complicated with epithelial downgrowth

association with chronic ocular surface inflammation
- Spheroidal degeneration

8.20 Laser In-Situ Keratomileusis (LASIK) (Fig. 8.23)

8.20.1 LASIK Surgical Technique

- Two stage procedure:
  - Creation of a thin flap (consists of epithelium, Bowman's layer, and superficial stroma) on the surface of the cornea using a microkeratome blade or using a FemtoSecond laser
  - Flap is lifted and the stromal bed is then reshaped with the excimer laser

8.20.2 Complications of LASIK

- Intraoperative:
  - Creation of the flap with the microkeratome related problems:
    Incomplete or irregular cut flap
    Button hole
    Corneal perforation
  - Photoablation-related problems:
    Treatment decentration
    Interface debris
    Wrinkles
- Postoperative
  - Early:
    Dislodged flaps:
    - First 24 h
    - Emergency repositioning ± sutures + BCL to prevent fixed folds and epithelial ingrowth

    Diffuse lamellar keratitis (shifting sands of Sahara):
    - First 24 h
    - Diffuse inflammation at flap interface without microbial cause
    - White sand-like deposits in lamellar cut plane in absence of both epithelial defect and AC activity
    - Aggressive topical steroid leads to rapid resolution

    Infectious keratitis:
    - ≤10d — gram +ve organisms, >10/7d — mycobacteria
    - Flap lift, scrape, frequent topical antibiotics

    Epithelial downgrowth (see Fig. 8.24):
    - Flap lift and scrape
  - Late
    Iatrogenic keratectasia
    Dry eye

8.21 Keratoconus (KC) (Fig. 8.25 and Table 8.19)

8.21.1 Associations of KC

- Ocular: retinitis pigmentosa (RP), vernal keratoconjunctivitis (VKC), leber's congenital amarousis (LCA), floppy eyelid syndrome
- Systemic: *atopic disease* (asthma, eczema, hayfever), *Down's syndrome* (habitual ocular massage), *craniosynostosis* (Crouzon's syndrome, Apert syndrome), *connective tissue disorders* (Marfan's syndrome, Ehlers-Danlos syndrome, Osteogenesis Imperfecta)

8.21.2 History

- Atopic disease — asthma, eczema, hayfever
- History of connective tissue disorders — Marfan syndrome, Ehlers-Danlos syndrome, Osteogenesis Imperfecta
- History of Down's syndrome
- History of craniosynostosis
- Previous laser refractive surgery

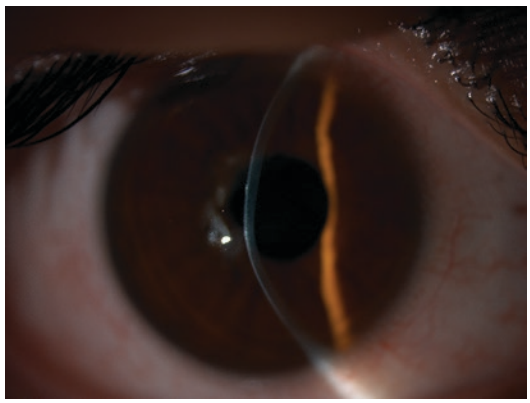

Fig. 8.25 Anterior segment image of a patient with KC showing thinning and scarring of the corneal stroma

Table 8.19 Key facts about KC

- Clinical term used to describe a bilateral condition in which the cornea assumes a conical shape because of thinning and protrusion
- Process is non-inflammatory with no cellular infiltration and vascularisation
- Onset occurs at about the age of puberty, where the cornea begins to thin and protrude, resulting in irregular astigmatism

8.21.3 Examination

- Corneal thinning from one-half to one-fifth of normal thickness is observed at the cone apex, the point of maximal protrusion
- Prominent corneal nerves
- Munson's sign: angulation of the lower lid in downgaze
- Vogt's striae (ruptures in Bowman's layer): fine vertical posterior stromal folds, which disappear when external pressure is applied to the globe
- Fleischer ring (provides a landmark for the peripheral edge of the cone): partial or complete annular iron line commonly seen at the base of the cone — cobalt blue illumination in the widest possible slit beam can enhance the appearance of a subtle iron ring
- Rizutti's sign: conical reflection on nasal cornea when pentorch is shone temporally
- Charleaux oil droplet sign: dark reflex in area of cone on observation of the cornea with the pupil dilated using a direct ophthalmoscope set on plano
- Corneal hydrops (ruptures in DM): stromal imbibition of aqueous and result in marked corneal oedema
- Deep stromal scarring of cornea: suggestive of previous hydrops
- Examine lids for upslanting of palpebral aperture (Down's syndrome) and floppy eyelid syndrome (easily everted upper eyelid tarsal plates + upper palpebral conjunctival papillary reaction)
- Examine for conjunctival papillae, Horner-Trantas dots (raised white accumulations of eosinophils at the limbus), shield ulcer: VKC
- Examine for blue sclera (results from visualisation of the underlying choroid through a thin sclera): Osteogenesis Imperfecta, Ehlers-Danlos syndrome
- Examine for ectopia lentis: Marfan's syndrome (superotemporal lens subluxation), Ehlers-Danlos syndrome

- Dilated fundus examination for mid-peripheral bone spicule retinal pigmentation, waxy pallor of the optic disc, arteriolar attenuation (RP), retinal detachment (Marfan's syndrome, Ehlers-Danlos syndrome)
- Examine for nystagmus: LCA

8.21.4 Investigations

- Refraction: scissoring on retinoscopy (see Table 8.20), irregular astigmatism, high myopia (Marfan Syndrome)
- Keratometer: inability to superimpose the central keratometric rings suggest irregular astigmatism, look for inferior corneal steepening (early sign of KC) by performing central keratometry followed by keratometry in upward gaze
- Placido disc: a focal area of increased corneal curvature appears as an isolated area of smaller ring spacing and distortion — look for decreased ring spacing inferiorly in early KC. As the condition progresses, the ring spacing decreases overall and becomes increasingly irregular
- Corneal topography (see Sect. 2.9.4)

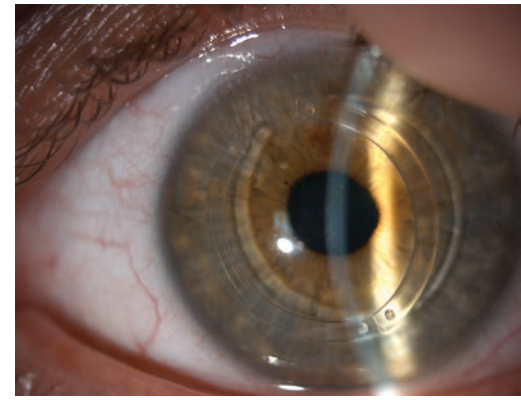

Fig. 8.26 Anterior segment image of a patient with KC who had corneal intacs implanted into the mid-peripheral corneal stroma to improve vision

Table 8.21 Dresden protocol for CXL

- Gold standard protocol for CXL
- Technique:
  - Topical anaesthesia
  - Central 9 mm of corneal epithelium removed
  - 0.1% riboflavin every 1–5 min for 30 min
  - Exposure to UVA light (3 mW/cm^2, 365 nm wavelength) for 30 min (total energy 5.4 J/cm^2)
  - Topical antibiotic
  - BCL insertion
- Contraindications:
  - CCT <400
  - Age >35 years old

8.21.5 Treatment

- Spectacle correction
- Rigid gas permeable (RGP)/scleral/miniscleral CL fitting
- Intacs (see Fig. 8.26): improves VA by flattening the central cornea, reducing astigmatism and centering the cornea, indicated for mild to moderate KC without corneal scarring — goal of procedure is to improve CL fit and comfort ± improve BCVA, ideal candidates have low spherical equivalent and average K readings of <53 D, ring segments do not prevent progression of the underlying disease, CCT needs to be ≥450 µm
- CXL (Table 8.21): slows or halts progression of the underlying disease + improves CL fit, improves VA by decreasing both corneal curvature and astigmatism, CCT needs to be >400 µm thick. Failure of CL is not an indication for CXL
- Keratoplasty (PK or DALK — if normal DM): Indications — intolerance to CL wear or severe irregular astigmatism with poor vision despite gls or CL or significant stromal scarring involving the visual axis
- Corneal hydrops: hypertonic NaCL drops and/or ointment, BCL, topical steroids, cycloplegia, intracameral gas — 20% SF6 (acts as a mechanical barrier, reducing aqueous flow into the stroma through the rupture in DM)

Table 8.20 Causes of a scissoring reflex on retinoscopy

- Corneal ectasias: KC, pellucid marginal degeneration
- Corneal scars
- Tilted lens
- Irregular retina (e.g. from a RD)

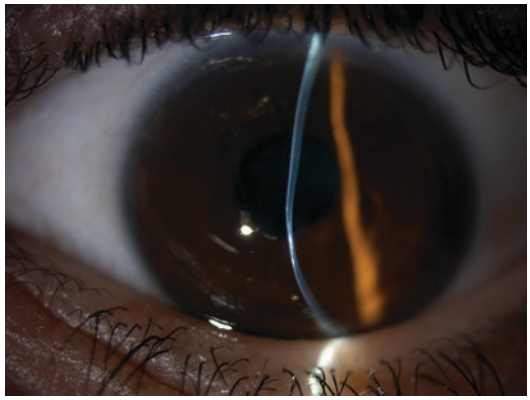

Fig. 8.27 Anterior segment image of a patient with pellucid marginal degeneration with inferior corneal thinning and corneal protrusion above the area of inferior thinning

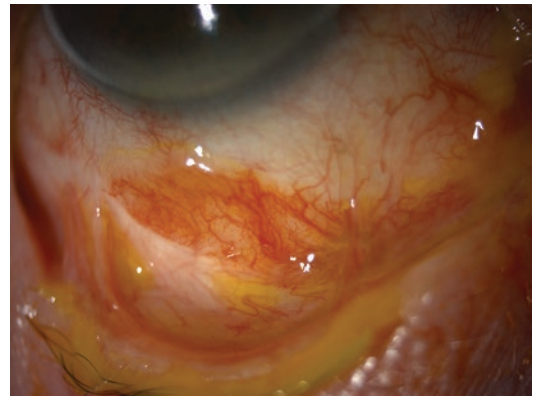

Fig. 8.28 Anterior segment image of a patient with OCP with symblepharon formation

8.21.6 Other Diagnoses to Consider

- Pellucid marginal degeneration: bilateral inferior corneal thinning (typically from 4 to 8 o'clock position) with maximal protrusion occurs just superior to the area of thinning (see Fig. 8.27), shift in axis of astigmatism from against-the-rule superiorly to with-the-rule near the point of maximal protrusion, crab-claw appearance on power map of corneal topography (see Fig. 2.63)
- Keratoglobus: bilateral globular protrusion of the cornea results from generalised thinning most marked in the periphery, prone to corneal rupture after minimal trauma, corneal topography reveals diffuse steepening on the power map and diffuse thinning on the pachymetric map
- Post-traumatic ectasia e.g. post-LASIK ectasia
- Protrusion of cornea subsequent to corneal thinning from ulceration

8.22 Ocular Cicatricial Pemphigoid (OCP) (Fig. 8.28)

8.22.1 Causes of Symblepharon Formation

- Steven-Johnson syndrome (SJS)/Toxic epidermal necrolysis (TEN)
- Chemical burns
- Trachoma
- Avitaminosis A
- Severe chronic keratoconjunctivitis caused by bacteria or viruses
- Medications (causing cicatricial conjunctivitis): systemic — penicillamine, topical — pilocarpine, timolol, gentamicin 1.5%)

8.22.2 History

- Exclude SJS/TEN: drug history (sulfonamides, anti-convulsants, allopurinol) and previous or current infections (mycobacteria, HSV)
- Exclude avitaminosis A: nyctalopia, history of cystic fibrosis or hx of jejunoileal bypass (fat malabsorption)
- Ask about previous chemical injury to eye
- Ask about history of trachoma

8.22.3 Examination

- OCP
  - Subepithelial conjunctival fibrosis (stage I)
  - Fornix foreshortening (stage II)
  - Symblepharon formation (stage III)
  - Ankyloblepharon and surface keratinisation of ocular surface (stage IV)
  - Progressive sicca syndrome with unstable tear film (obstruction of lacrimal

ductules and meibomian gland ducts), trichiasis and entropion occur from subepithelial fibrosis with eventual corneal neovascularisation, corneal ulceration and scarring

- SJS/TEN
  - Acute eye findings (bilateral): pseudomembranous conjunctivitis ± secondary purulent bacterial conjunctivitis, corneal ulceration, anterior uveitis
  - Chronic eye findings (bilateral): lid margin keratinisation, scarring (subconjunctival fibrosis), symblepharon formation, and cicatrization of the conjunctiva — leads to entropion formation, trichiasis, and instability of tear film (goblet cell dysfunction), keratoconjunctivitis sicca (KCS), persistent ED, corneal scarring and stromal neovascularisation, LESC failure
  - Non-ocular findings:

    SJS: erythematous macules that develop central necrosis to form vesicles, bullae, and areas of denudation on the face, trunk, and extremities, two or more mucosal surfaces are involved (conjunctiva, oral cavity, upper airway or oesophagus, GI tract, anogenital mucosa), less than 20% of body surface area involved, *TEN*: involvement of more than 30% of the body surface area, positive Nikolsky's sign (friction applied to healthy areas of skin, causing epidermis is wrinkle and separate), mucous membranes are usually involved with severe erosions of the lips, oral surface, conjunctiva, and genital areas.
- Trachoma (*C. trachomatis* Serovars A–C): follicular conjunctivitis, Herbert's pits (breakdown of follicles around limbus with subsequent tissue necrosis), Artl's line (linear scarring on the upper tarsus), cicatricial lid changes with tarsal plate contraction with accompanying trichiasis — lead to superior pannus, scarring or opacification of cornea (Table 8.22).
- Avitaminosis A: conjunctival xerosis (metaplastic keratinised surface) and Bitot spots (foamy), corneal xerosis (localised or generalised oedema with a typical dry, lackluster appearance, later the cornea may develop a "peau d'orange" appearance from keratinization), corneal ulceration/keratomalacia (frank necrosis or sloughing of the corneal stroma)

Table 8.22 WHO grading system for Trachoma

TF grade: 5 or more 0.5 mm or more in diameter follicles in upper tarsal conjunctiva
TI grade: inflammatory thickening of tarsal conjunctiva that obscures more than half of normal deep tarsal vessels
TS grade: scarring in tarsal conjunctiva
TT grade: trichiasis
CO grade: corneal opacity over pupil

8.22.4 Investigations

- OCP: conjunctival biopsy to look for linear immune deposits (must be demonstrated before treatment initiated) along the epithelial basement membrane zone (immunofluorescence microscopy — Michel's transport medium)
- SJS/TEN: bloods for raised inflammatory markers, conjunctival biopsy to look for absence of goblet cells in SJS, referral to dermatologists/plastic surgery for skin biopsy in TEN
- Avitaminosis A: vitamin A levels
- Trachoma: swabs for immunofluorescent staining, cell culture, PCR, ELISA

8.22.5 Treatment

- OCP
  - Treat sicca syndrome (artificial tears, ointments, punctal occlusion if Schirmer values less than 5 mm/5 min)
  - Treat chronic blepharitis and meibomitis (warm compresses, lid hygiene, oral doxycycline 100 mg OD for 3 months then 50 mg OD)
  - Immunomodulatory therapy if cases that are active and rapidly progressive (mild: dapsone + oral prednisolone, moderate:

MTX, AZT or MMF + oral prednisolone, severe: IV/PO cyclophosphamide + oral prednisolone)
  - Keratoprosthesis for stage IV disease
- SJS/TEN
  - Systemic disease: refer to intensive burn care unit for monitoring of fluid balance, respiratory function, nutritional requirements, and wound care
  - Ophthalmic disease — acute stage: frequent conjunctival irrigation + prophylactic topical antibiotics to prevent secondary infection, frequent PF topical lubricants (lubricate conjunctival and corneal epithelium), cycloplegics (anterior uveitis), topical steroids but monitor for secondary bacterial keratitis (anterior uveitis), lamellar keratoplasty or PK if perforation impending or occurs, conjunctival flap (Gunderson flap) for an impending perforation, daily lysis of the symblepharon or placement of a symblepharon ring
  - Ophthalmic disease — chronic stage: repair cicatricial entropion and treat trichiasis (epilation, electrolysis, cryotherapy), frequent PF topical lubricants to treat KCS, fornix reconstruction through mucous membrane grafts, BCLs, AMG or tarsorrhaphy for persistent ED, limbal stem cell transplant for LECS failure, keratoprosthesis for end stage corneal scarring and neovascularisation
- Avitaminosis A
  - Vitamin A supplementation
- Trachoma
  - TF grade — azithromycin 1 g PO, TT grade — bilamellar tarsal rotation, TI grade — azithromycin 1 g PO, TS grade — lubricants (infection no longer present at TS stage and so antimicrobial therapy is not useful), CO grade — PK

8.23 Penetrating Keratoplasty (PK)

8.23.1 Definition

- Transplant procedure in which full-thickness host corneal tissue is replaced with donor tissue

8.23.2 Examination

- Examine for a full thickness graft with no graft-host interface present to identity patients who have had a PK surgical procedure

8.23.3 Indications

- Visual (restoration of corneal clarity): keratoconus, pseudophakic/aphakic corneal oedema, FED, deep stromal corneal dystrophies, corneal scarring secondary to herpes simplex keratopathy, infections, trauma
- Tectonic (restoration of structural integrity): corneal thinning, actual or threatened perforation
- Therapeutic: infective keratitis

8.23.4 Contraindications

- Relative
  - Epithelial dysfunction secondary to limbal stem cell deficiency (aniridia, chemical injuries), severe neurotrophic, and dry eye states
  - Stromal vascularisation, especially when involving more than two quadrants
  - Multiple (two or more) graft failures

8.23.5 Advantages of PK

- Familiarity

8.23.6 Disadvantages of PK

- Higher risk of graft rejection
- Risk of suture-related complication
- Risk of traumatic wound dehiscence and globe rupture

8.23.7 Complications

- Intraoperative
  - Scleral perforation from bridal suture placement or scleral ring placement

  - Damage to donor cornea
  - Trephine reversal: causes unwanted hyperopia and difficulty in closing wound
  - Retained Descemet's membrane: suggested by presence of double AC on post-op day 1 (patient will need re-grafting)
  - Iris-lens damage during trephination
  - AC haemorrhage from iris damage
  - Suprachoroidal expulsive haemorrhage
- Early postoperative
  - Wound leaks (shallow AC, seidel positive): if leak controlled with BCL resuturing is not necessary
  - Epithelial defect: treat with lubricants, topical antibiotics ± BCL ± tarsorrhaphy, decrease application of topical steroids
  - Filamentary keratitis: treat with lubricants and removal
  - Suture-related complications:
    Infection
    Immune infiltrates: multiple on recipient side of cornea and treated with intensive topical steroids and removal of suture
    Endophthalmitis
    Kaye dots: small epithelial dots a short distance from sutures on the donor side — not pathologic and usually disappear after suture removal
  - Glaucoma
  - Anterior synechiae: increases graft oedema and risk of rejection — postop dilatation increases AC depth
  - Pupillary block: shallow AC, raised IOP, intact wound — aqueous misdirection, choroidal detachment
  - Urrets-Zavalia pupil: irregular pupil a few days post PK for KC — iris stromal atrophy, ectropion uvea, multiple posterior synechiae, discrete subcapsular opacities
- Late postoperative
  - Graft failure (see Fig. 8.29): causes include immunologic rejection, poor donor quality, intraoperative trauma to donor endothelium, prolonged flat AC in the immediate postoperative period, HSV reactivation

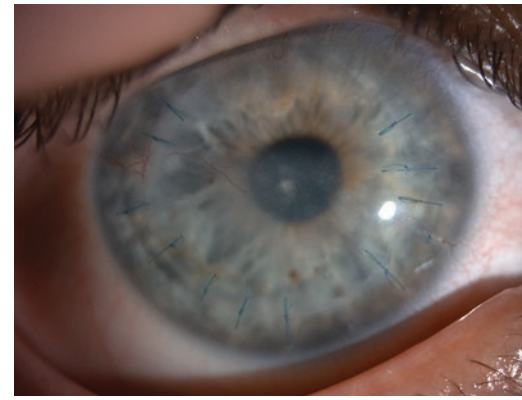

Fig. 8.29 Anterior segment image of a patient with a failed penetrating keratoplasty showing diffuse corneal haze with stromal neovascularisation

  - Immunologic rejection (risk of an immunologic rejection episode is constant over the first 3 years after transplantation):
    Epithelial rejection: elevated epithelial rejection line that stains with fluorescein — responds to topical steroids
    Subepithelial infiltrates: immune reaction seen only in the donor tissue without associated conjunctivitis — responds to topical steroids
    Endothelial rejection line (Khodadoust line): does not extend beyond graft-host junction with KP's at the leading edge with an overlying hazy and oedematous stroma ± DM folds — if left untreated this line will proceed across the donor endothelium from the point of origin at the graft wound leaving damaged endothelium behind it — treat with intensive topical steroids or IV methylprednisolone
    Diffuse endothelial rejection: isolated or diffuse KPs are scattered across the endothelium and are limited to the donor endothelium and often associated with an AC reaction — treat with intensive topical steroids or IV methylprednisolone)
    Post-operative astigmatism

8.23.8 Risk Factors for Immunologic Rejection (Poor Prognostic Factors)

- Corneal stromal vascularisation
- Reduced corneal sensation
- Ocular surface disease (Dry eye, OCP, SJS)
- Previous graft failure
- Large graft size (8–8.5 mm) and eccentric grafts
- Uncontrolled glaucoma
- Active or recurrent herpetic inflammation

8.23.9 Penetrating Keratoplasty in Children

- Preoperative complications
  - Need for frequent topical drop applications
  - Requirement of frequent postoperative visits
  - Commitment to amblyopia therapy
- Intraoperative complications
  - Higher posterior pressure: forward displacement of lens and iris with increased risk of iris prolapse, lens extrusion, suprachoroidal haemorrhage
  - Smaller donor grafts and more pliable tissue with more difficulty with suturing
- Postoperative complications
  - Stronger inflammatory response to surgery than adults: increased risk of graft rejection and failure, contraction of tissue at the 360° interface between host and donor tissue can lead to loosening of the sutures which is a risk factor for suture abscesses and neovascularisation of the corneal tissue
  - Frequent application of topical steroids to reduce high risk of rejection
  - High risk of infection: loose sutures, wound dehiscence, high dose of topical steroids — longer use of prophylactic topical antibiotics
  - Amblyopia: high rate of vision loss despite clear graft
- Prognosis
  - Only about 33% of the grafts performed for Peter's anomaly, Sclerocornea, or intrauterine infection were clear at 1 year
  - In infants less than 1 year old, only 25% of grafts were clear at 1 year
  - In children between 1–4 years old, about 50% were clear at 1 year
  - In children older than 4 years, about 65% were clear at 1 year

8.24 Deep Anterior Lamellar Keratoplasty (DALK) (Fig. 8.30)

8.24.1 Definition

- Surgical technique whereby the majority of the corneal stroma down to the Descemet's membrane is replaced by donor tissue, leaving the host endothelium intact

8.24.2 Examination

- Examine for a graft-host interface to identify patients who have had a DALK surgical procedure

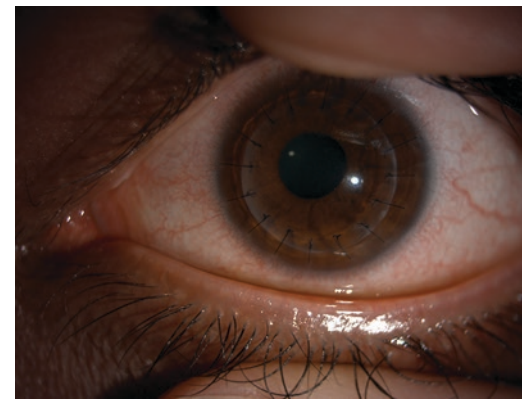

Fig. 8.30 Anterior segment image of a patient who had a DALK procedure

8.24.3 Indications

- Visual: corneal ectasias, stromal dystrophies sparing Descemet's membrane and endothelium, corneal stromal scars from previous infections or chemical injuries
- Tectonic: descemetocoele, pellucid marginal degeneration, advanced Terrien's marginal degeneration, peripheral corneal melts from autoimmune disorders (e.g. rheumatoid arthritis)

8.24.4 Contraindications

- Absolute: endothelial dysfunction
- Relative: epithelial dysfunction

8.24.5 Advantages of DALK

- Preserve host endothelium
- Lower incidence of graft rejection

8.24.6 Disadvantages of DALK

- Working near or at Descemet's membrane with potential risk for perforation

8.24.7 Complications

- Intraoperative
  - Perforations and ruptures of Descemet's membrane during trephination, stromal dissection or with a suture needle
- Postoperative
  - Urrets-Zavalia syndrome (fixed dilated pupil from irreversible damage to the iris sphincter): associated with air/gas injection into the AC at the end of surgery
  - Suture-related complications:
    - Loosening of sutures
    - Suture abscesses
  - Stromal rejection

8.25 Descemet Stripping Automated Endothelial Keratoplasty (DSAEK) (Fig. 8.31)

8.25.1 Definition

- Partial thickness cornea transplant procedure that involves selective removal of recipient Descemet's membrane and endothelium, followed by transplantation of donor corneal endothelium in addition to donor corneal stroma

8.25.2 Examination

- DSAEK grafts can be visualised as a subtle ring in the diffuse illumination and can be seen protruding posteriorly from the cornea in the inferior aspect of the slit beam.

8.25.3 Indications

- Endothelial dystrophy: FED, posterior polymorphous corneal dystrophy
- Bullous keratopathy: aphakia, pseudophakia
- Endothelial failure: trauma, previous surgery, angle closure, tubes

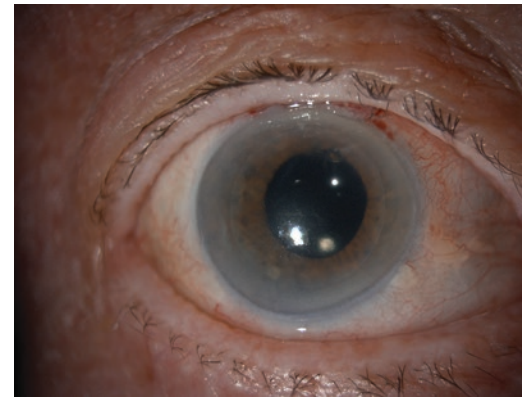

Fig. 8.31 Anterior segment image of a patient who had a DSAEK procedure

- ICE syndrome
- Failed PK — if acceptable refractive result was achieved

8.25.4 Contraindications

- Stromal opacity or scarring that limits visual potential
- Keratoconus, ectasia
- Hypotony/pre-phthisical eye

8.25.5 Advantages of DSAEK

- No suture-related complications
- Reduced incidence of graft rejection
- Rapid visual rehabilitation

8.25.6 Disadvantages of DSAEK

- Possible interface haze and deposits
- Potential significant loss of donor endothelium during surgical procedure
- Risk of graft dislocation

8.25.7 Complications

- Intraoperative
  - Complications with donor tissue preparation: damaged donor tissue, eccentric trephination, thin donor tissue (increase chance of graft striae)
  - Retained Descemet's membrane: prevents proper attachment of the DSAEK graft
  - Air management: donor tissue attachment appears to be dependent on the maintenance of air in the AC (challenge in aphakic eyes or pseudophakic eyes with an open posterior capsule and in patients with previous trabeculectomy or glaucoma drainage implants)
- Postoperative
  - Donor dislocation (methods to avoid: cornea surface massage with a flap roller, vents are made in the midperipheral recipient cornea down to the graft interface)
  - Primary graft failure (graft failing to clear within the first 2 weeks after the op):
    Primary failure: unhealthy donor endothelial, inadequate preservation of the corn, or traumatic pre- or intraoperative technique
    Secondary failure: graft rejection, detached donor from residual fluid or viscoelastic in interface, retained Descemet's membrane
  - Graft rejection: presence of AC cells ± KP's and concomitant corneal oedema
  - Pupillary block glaucoma: due to placement of excessively large gas bubble in the AC or migration of the air bubble behind the iris — tx by partial evacuation of the air bubble and concurrent replacement with BSS to maintain the AC
  - Endothelial cell loss
  - Refractive change: hyperopic refractive shift post DSAEK
  - Interface deposits and epithelial ingrowth: epithelial cells may be dragged onto stromal surface during insertion of the donor tissue — if epithelial ingrowth does not progress to the visual axis and if the donor is well attached to the recipient surface, the patient can be followed closely without surgical intervention

8.26 Anterior Scleritis (Fig. 8.32 and Table 8.23)

8.26.1 Associations of Scleritis

- Non-infectious: systemic diseases (RA, GPA, relapsing polychondritis, PAN, IBD, SLE, Churg-Strauss syndrome), surgically induced (strabismus surgery, trabeculectomy, cataract extraction, RD repairs, pterygium excisions)
- Infectious: traumatic ± FB, post-surgical, extension from other structures (keratitis, choroiditis, endophthalmitis, conjunctivitis, orbital cellulitis, sinusitis, dacryocystitis)

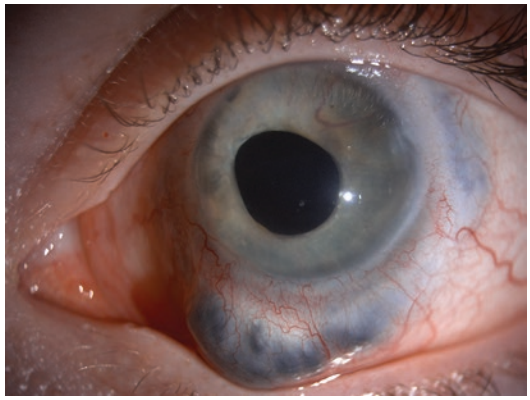

Fig. 8.32 Anterior segment image of a patient with multiple previous episodes of scleritis showing severe scleromalacia with diffuse purple or blackish hue from increased uveal visibility. There is a large area of uveal protrusion with only a thin layer of conjunctiva overlying the uvea

Table 8.23 Causes of a blue sclera

- Scleritis
- Osteogenesis Imperfecta: hx of fractures (brittle bones), hyperextensibility of the joints, sensorineural hearing loss, hyperopia
- Connective tissue disorders: Ehlers-Danlos syndrome (ectopia lentis, RD, hyperelasticity and fragility of the skin), Marfan's syndrome (ectopia lentis, RD, myopia, arachnodactyly, high arched palate), Pseudoxanthoma elasticum — PXE (angioid streaks ± CNV, optic disc drusen, yellowish lumps on the skin of the neck, axillae, and antecubital fossa)
- Keratoconus/Keratoglobus

8.26.2 History

- Scleritis: severe pain enough to awaken patient at night
- Episcleritis: mild discomfort or FB sensation
- POHx of previous ocular surgeries
- PMHx of any systemic diseases (GPA — sinusitis, bloody nasal discharge, saddle nose, relapsing polychondritis — degenerative pinna cartilage)

8.26.3 Examination

- Anterior scleritis (no blanching with topical phenylephrine): diffuse (distortion and tortuosity of both episcleral and scleral vascular networks, post resolution the sclera appears translucent or bluish-gray suggesting thinning of the sclera), nodular (firm, immobile, and tender yellow to deep red nodule to palpation), necrotising with inflammation (white avascular areas of sclera and conjunctiva, with surrounding scleral oedema and congestion, blue-black uveal tissue may appear as the overlying sclera becomes thin and translucent), necrotising without inflammation (scleromalacia perforans — complete lack of symptoms with thinned and avascular areas of episclera and sclera with no surrounding inflammation)
- Posterior scleritis: choroidal folds, choroidal effusions, exudative RD, CMO, optic disc oedema
- Examine hands for RA: swollen MCP, PIP, wrist, or MTP joints, Z-deformity of thumb, swan-neck and Boutonniere deformity of fingers
- Examine for skin rashes: SLE (discoid rash, malar rash), PAN (livedo reticularis)

8.26.4 Investigations

- B-scan: T-sign — posterior scleritis
- Bloods: c-ANCA (GPA, PAN), p-ANCA (Churg-Strauss syndrome), ANA (SLE), rheumatoid factor (RA), anti-CCP (RA), HLA-B27 (IBD)
- CXR: GPA (pulmonary haemorrhage), Churg-Strauss syndrome
- Urinalysis: proteinuria and haematuria (GPA)

8.26.5 Treatment

- Oral NSAIDS: reduce pain and inflammation in non-necrotising scleritis
- Systemic corticosteroids: indicated in severe non-necrotising scleritis unresponsive to oral NSAIDs and in necrotising scleritis — oral prednisolone at a starting dose of 1 mg/kg/day followed by a slow taper according to response
- Systemic immunosuppressants: indicated in cases of necrotising scleritis and scleritis of any type in the setting of GPA (cyclophospha-

mide) or PAN, or when maintenance dose of oral prednisolone ≤7.5 mg OD is not possible without recurrence of disease

8.26.6 Other Diagnoses to Consider

- Episcleritis: simple: sectoral or diffuse redness which blanches with topical phenylephrine, globe non-tender, no investigations required unless history suggestive of systemic disease, treatment with topical lubricants, oral NSAIDS, topical corticosteroids / nodular: red nodule arising from episclera that can be moved separately from the sclera and conjunctiva, globe non-tender, blanches with topical phenylephrine, no investigations required unless history suggestive of systemic disease, treat as for simple episcleritis
- Peripheral ulcerative keratitis (PUK): unilateral crescent-shaped peripheral corneal ulceration with overlying epithelial defect and stromal thinning, sectoral or diffuse scleritis, treatment involves ensuring adequate tear film (lubricants, punctal plugs, punctal cautery), prophylactic topical antibiotics, oral doxycycline and oral vitamin C (inhibit proteases and free radicals, respectively), systemic immunosuppression (liaise with rheumatologist, corticosteroids — IV methylprednisolone or oral prednisolone, MTX, AZT, MMF, ciclosporin, cyclophosphamide), BCL + cyanoacrylate glue for pending/actual perforation, conjunctival recession, tectonic freehand lamellar keratoplasty, conjunctival flaps
- Mooren's ulcer: diagnosis of exclusion (rule out Hepatitis C), unilateral peripheral ulceration with stromal melt with leading edge undermining epithelium, ulcer advances centrally and circumferentially, no associated scleritis (but conjunctiva and episcleral inflammation), treatment similar to PUK — nay require interferon if coexistent hepatitis C (as directed by hepatologist)

8.27 Limbal Epithelial Stem Cell (LESC) Deficiency (Fig. 8.33)

8.27.1 Causes

- LESC deficiency occurs when there is sufficient disturbance or destruction of the limbal stem cells of the corneal epithelium, resulting in disruption of the normal physiologic regenerative process and repopulation of corneal epithelium
- Typical causes include
  - Aniridia
  - Chemical (alkali injury) or thermal injury
  - Chronic contact lens wear
  - Preservative drop toxicity
  - OCP
  - SJS/TEN
  - Ocular surgery: multiple previous pterygium excisions

8.27.2 History

- History of ocular chemical injuries
- History of contact lens wear
- History of ocular surgeries e.g. pterygium surgery
- History of aniridia

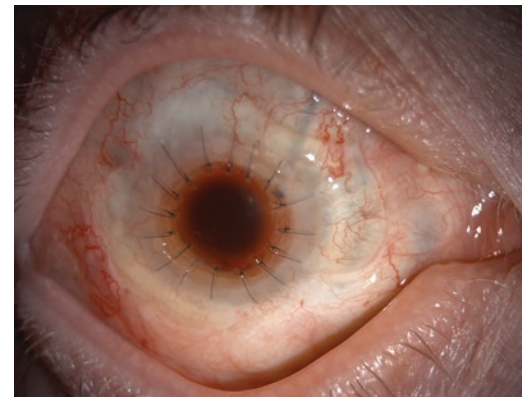

Fig. 8.33 Anterior segment image of a patient with congenital aniridia who had a limbal stem cell transplantation

8.27.3 Examination

- Examine for loss of palisades of Vogt, late staining of the corneal epithelium with fluorescein, persistent epithelial defects, corneal neovascularisation, corneal scarring, and the development of a peripheral pannus
- A "whorled" appearance of the corneal epithelium (results from conjunctivalisation of the corneal epithelium — invasion of conjunctival epithelium onto the corneal surface — occurs when <25–33% of LESC remain)
- Examine for a limbal stem cell transplant graft

8.27.4 Investigations

- Impression cytology (nitrocellulose paper is pressed onto cornea and examined under the microscope): presence of mucin-containing goblet cells on the corneal epithelium and the absence of normal differentiation markers of corneal epithelium (CK 3 and 12)

8.27.5 Treatment

- Any chronic inflammation of the conjunctiva, conjunctival scarring, symblepharon formation, trichiasis, and MGD must be identified and treated prior to limbal stem cell grafting
- Partial: conjunctivalised metaplastic epithelium on cornea — if visual axis affected: sequential sector conjunctival epitheliectomy + AMG, if visual axis is not affected: sequential sector conjunctival epitheliectomy, with fibrovascular pannus — sector limbal transplant + AMG
- Total: unilateral — conjunctival limbal autograft (CLAU) from contralateral better eye, bilateral — living-related keratolimbal allograft (CLAL), cadaveric CLAL

8.28 Gundersen Flap (Fig. 8.34)

8.28.1 Definition

- A Gundersen flap is a surgical technique that uses a thin, bipedicle, total conjunctival bridge flap to achieve a number of purposes including
  - To restore the integrity of a compromised ocular surface
  - To provide a metabolic and mechanical support for corneal healing
  - To relieve pain
  - To provide an alternative to invasive surgery or enucleation

8.28.2 Indications

- Persistent corneal epithelial defect
- Unresponsive ulcerative microbial keratitis
- Corneal thinning and perforation

8.28.3 Advantages

- Can improve quality of life in patients with an eye with poor visual potential and a chronically irritated anterior segment

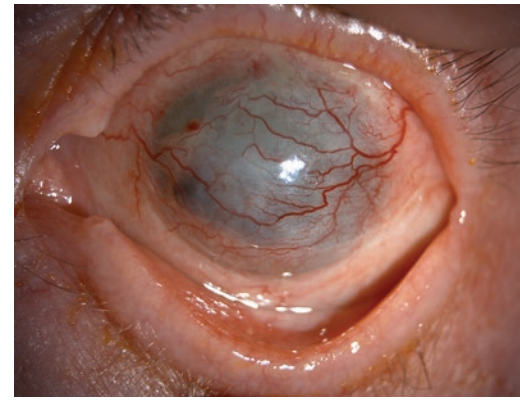

Fig. 8.34 Anterior segment image of a patient with a Gundersen flap

- Can eliminate the need for chronic medication and bandage lenses

8.28.4 Disadvantages

- Vision significantly decreased by the conjunctival flap that covers the visual axis
- Prevents monitoring of disease progression by obstructing any view of the cornea and anterior chamber
- Poor cosmetic appearance

8.28.5 Complications

- Intraoperative
  - Buttonhole formation
  - Dissection of an inadequate flap
  - Excessive haemorrhage
- Postoperative
  - Ptosis
  - Retraction of the flap
  - Vascularisation and opacification of the underlying cornea