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Delhi Journal of Ophthalmology (DJO), once called Visiscan, is a quarterly journal brought out by the Delhi Ophthalmological Society. The journal aims at providing a platform to its readers for free exchange of ideas and information in accordance with the rules laid out for such publication. The DJO aims to become an easily readable referenced journal which will provide the specialists with up to date data and the residents with articles providing expert opinions supported with references.

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Author/Authors must have made significant contribution in carrying out the work and it should be original. It should be accompanied by a letter of transmittal. The article can be sent by email to the Editor or a hard copy posted. Articles received will be sent to reviewers and their comments will be emailed to the author(s) within 4-6 weeks. The identity of the authors and the reviewers will not be revealed to each other by the editorial team. Detailed instructions to the contributors and for advertisement are included at the end of the journal.

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Editorial Office
Dr Rohit Saxena, Room No. 479, Dr R.P. Centre for Ophthalmic Sciences, AIIMS, New Delhi-110029
Ph +91-011-26593182, Email : editordjo@gmail.com

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59. Instructions to Authors
Dear friends,

We are all smarting under the insult the colonial minded British have heaped upon us. Just when our medical industry was getting set to reverse the flow of patients and break free, these jealous British in their inscrutable way have accused us of producing the superbug: and scaring away our patients.

It took hours of prime time television and front page headings in national dailies to get back our confidence and to reassure all of us that the report is false and the ‘New Delhi’ Metalloproteinase 1 is just jealously and nothing else. After all isn’t it that many countries have reported similar multi drug resistant organisms. Also there is no concrete epidemiological data linking this NDMI to us.

But could there be just a little lesson for us to learn in it. Could it be that our medical practices is helping to create such superbugs. Those that are resistant to increasingly newer antibiotics as we are pushed by the pharmaceutical industry to use the latest available antibiotics for ‘viral URI’s and just about anything else.

Could the easy availability of over the counter cutting edge drugs and their indiscriminate use even by the lay help to create such resistance?

Could we Ophthalmologists be a major contributors to this problem: we give the topical formulation of these latest antibiotics for just about anything: itching, surgical prophylaxis, mild congestion etc. and minute quantities of the antibiotic reaches the gut where the sub-MIC concentrations ensure that the bacteria get enough drug to adapt to them but not kill them.

Should we start working for a rational antibiotic use (especially topical ones) policy for hospitals, for a ban on OTC availability of life saving newer antibiotics, but then I don’t think this is necessary. We know that for all our problems there is always someone to blame: Mr. Kalmadi, the rain gods, the government, insensitive officials etc, we know this time it is the jealous British doctors worried about their loss of livelihood.

That is why I think that the Ostrich should be the National Bird of India.

Dr. Rohit Saxena

With due apologies to the peacock, the national bird of India and our esteemed forefathers who felt that the peacock truly represents the colours and diversity of India. “Hiding their head in the sand, like an ostrich” is an English metaphor which means to be foolishly ignoring their problem, while hoping it will magically vanish.

The Delhi Journal of Ophthalmology is now indexed at Index Copernicus. The editorial board is involved in the task of getting the journal indexed in other sites as well as improving the quality of articles and their presentation. This is only possible with the support of each and every DOS member.

In addition to the present heads, the DJO also publishes original research including thesis work of residents. We also welcome comments to articles and advise on how to improve the DJO. Any DOS member who has not received the previous four issues please contact DOS Secretariat dosrecords@gmail.com, dostimes@airtelmail.in or Editor, DJO editordjo@gmail.com. Some copies have come back due to incorrect addresses, so members are requested to please provide correct addresses and contact details to DOS Secretariat.
Orbit is an anatomical Pandora’s box when it comes to lesions that can occupy its domain. It is a complex cavity containing various compactly arranged tissues; the globe, the extraocular muscles, vessels, nerves, glandular and mesenchymal tissues. A wide spectrum of pediatric orbital tumours can be seen in childhood; these space occupying lesions can be primarily orbital pathology or secondary to some systemic disease. These differ substantially from those in adult patients since in children there is greater incidence of congenital lesions, higher frequency of infection, and unique benign and malignant tumours that involve the orbit. We in this article will primarily describe an approach to a child with proptosis and the commonly seen benign space occupying lesions in children. Malignant space occupying lesions will be the main feature of part II of this review article.

Incidence and differential diagnosis of orbital space occupying lesions in children

Various studies have reported marked variation in incidence of various space occupying lesions in children.[1-7] Bullock et al compared their findings to nine other published series and have reported cystic lesions of the orbit (mainly dermoids) to be the most common orbital masses in children followed by vasculogenic lesions (capillary hemangiomas, lymphangiomas or cavernous hemangiomas).[8] The common malignant lesions of the orbit described in children were rhabdomyosarcoma, secondary malignant tumours/malignancies (including neuroblastoma, Ewing’s sarcoma, and orbital involvement in retinoblastoma), lymphomas and leukemia. In a retrospective analysis of histopathological records from our centre by Bajaj et al, contrary to western reports, secondary orbital involvement of retinoblastoma was the most common cause of proptosis[1-3, 9].

Approach to a child with Orbital space occupying lesion

While evaluation of a child with orbital disease, a meticulous history of the patient’s ocular and systemic systems is vital. The ophthalmic history should include the age of onset, the duration and rate of progression of the proptosis. The informant or if possible the patient should be queried about pain, change in visual acuity or refraction, diplopia, and decreased fields of vision. While eliciting a thorough medical history, the ophthalmologist should also consider orbital involvement secondary to systemic pathology. Past trauma and family history also may aid in the diagnosis.

A thorough general physical examination with review of all systems form the next step in evaluation which should be followed by complete and elaborate evaluation of ocular and periorbital area. Relative protrusion can be observed by simply standing behind a seated patient and gazing downward toward the chin from the forehead to assess the displacement of one globe as compared to the contralateral side. Decreased visual acuity (measured in a child by an age appropriate method), change of refraction, and pupillary abnormalities should be noted. Extraocular motility dysfunction and diplopia should be carefully assessed and documented. One should carefully evaluate status of various cranial nerves in relation to the orbit. Palpation of the anterior orbit can assess associated tenderness, consisteny, and mobility of the mass. In case of proptosis, measurements should be made with Hertel exophthalmometry and also one should note the displacement of the eye in planes other than the anteroposterior dimension (eg, downward, lateral). Slit lamp examination and intraocular pressure measurement, should be done where possible. Dilated funduscopic examination may reveal optic disc edema or pallor, retinal detachment, choroidal folds, vascular engorgement or shunt vessels, or indentation of the posterior pole. Functional evaluation of optic nerve should be done with color vision, visual fields and VER if possible.

Cystic lesions

Dermoid Cysts

Dermoid cysts are benign developmental choristomas, thought to be the most common space–occupying orbital lesion of childhood.[2,4,8] These cysts are congenital arising from nests of primitive dermal elements that have been trapped in bony suture lines at the time of fetal closure. This sequestered tissue forms dermoid cyst which histopathologically is lined with keratinized epithelium and has dermal elements, such as hair follicles, sweat glands and sebaceous glands. Shields and colleagues have classified orbital dermoids into juxtasutural, sutural and soft tissue types[10]. The juxtasutural type are adjacent but not obviously attached to the bony suture line. These are the most common type in children and appear in the superotemporal and superonasal quadrants. A sutural
dermoid cyst extends into the bony suture line, forming a pit or hourglass configuration. Soft tissue or complicated dermoids usually present at an older age. Clinically, the juxtasutural dermoid cyst in children presents as a painless mass in the superotemporal area and is unattached to overlying skin. The mass is smooth, mobile, and nontender with no visual symptoms. The soft tissue dermoid grows slowly over a long period of time and often presents with proptosis. Computed tomography (CT) scan preoperatively is required to confirm the diagnosis and rule out any intracranial extension (Figure 1). Deeper orbital lesions may show complete bony detects.

Management of dermoid cysts is surgical, aiming for complete excision. Intraperatively, rupture of the cysts is to be avoided to limit lipogranulomatous inflammation and scarring. If the cysts is accidentally ruptures, copious irrigation of the site is performed. Excision of sutural and soft tissue cysts is more complicated. Sutural cysts often cannot be removed intact because of their communication into or through the bone. Care is taken to remove all remaining cyst lining, thereby limiting the possibility of recurrence. Soft tissue cysts are removed by way to a transconjunctival or lateral orbitotomy approach.

Teratomas

Teratomas are rare congenital germ-cell tumours that may appear in the orbit. Arising from primordial germ cells, these tumours are characterized by the presence of all three germinal layers i.e ectoderm, mesoderm and endoderm. They are benign growths and do not invade orbital bone, although orbital enlargement is often seen. Presenting at birth as a large orbital mass causing moderate to massive proptosis, the teratomatous lesion may be accompanied by conjunctival keratinization, exposure keratopathy and corneal ulceration. Teratomas may present as primary orbital, combined orbital and intracranial, or sino-orbital masses[11]. Massive teratomas traditionally have been treated by orbital exenteration especially when the eye is severely damaged. Debunking surgery with sparing of globe is possible with even visual preservation in cases which present early for better cosmetic and if possible visual rehabilitation[12-14].

Hydatid cyst

Cystic hydatid disease is a zoonotic infection of humans caused by the larval stage of Echinococcus granulosus (Figure 2). Man is the intermediate host while dog is the definitive host. The prevalence rate varies with endemicity and is 5-10%
The most common site for the development of hydatid cyst is the liver and lungs with act as two natural filters during the process of migration of the larvae from the gut. Orbital hydatid disease is quite rare and represents <1% of the Echinococcus cases. The symptoms include progressive proptosis with or without pain, disturbance in ocular motility, visual deterioration and chemosis. A few cases with acute onset visual loss have been reported due to serous retinal detachment, secondary to local inflammation caused by liberation of toxins from semipermeable cyst wall. In long standing cases visual loss can occur due to secondary optic atrophy or exposure keratitis.

Serological tests: The Casoni’s intradermal test shows an immediate hypersensitivity reaction with high sensitivity and low specificity; the counter immunoelectrophoresis test has been adapted to diagnose hydatid disease with high specificity and sensitivity.

Imaging: USG and MRI are valuable diagnostic aid, for detection of the lesion. The characteristic ‘double wall sign’, as described by Betharia et al. from our centre is often diagnostic of hydatid cyst on ultrasonography. MRI is superior for delineation of its relationship to the adjacent ocular structures.

Complete surgical removal via orbitotomy and cryo-extraction of the endocyst has been described as the gold standard in the management of isolated hydatid cyst. In the present era of anti-heliminthic drugs, preoperative and post operative course of oral albendazole is recommended.

Orbital cysticercosis
Cysticercosis is the infestation of tissue by larval tapeworm Taenia solium. Man is the definitive host while pigs are the intermediate host of T.solium, but at times humans can be infected by taking raw food contaminated with eggs of T.solium or by consuming food or water contaminated with faecal matter containing ova. Human cysticercosis predominantly affects the central nervous system causing neurocysticercosis and the eye and adnexa causing ocular/orbital cysticercosis. Ptosis may be an unusual presentation when the cyst resides in the levator causing mechanical restriction of its action. Intraocular cysticerci are easily diagnosed by ophthalmoscopy, however the diagnosis of orbital cysticercosis was largely speculative until the advent of advanced imaging modalities like ultrasonography, CT and MRI. High resolution ultrasonography displays the characteristic picture of a sonolucent area with well-defined anterior and posterior margins. The presence of a central echodense, curvilinear,
highly reflective structure within the cyst suggestive of scolex, helps to narrow the differential diagnosis to cysticercosis as the aetiological cause. CT and MRI not only confirm the diagnosis but also helps to rule out neurocysticercosis, prior to starting of antihelminthic drugs from an ophthalmologist side. Presence of a cystic lesion without a scolex with positive ELISA for anticysterceral antibodies is also diagnostic. A combination of oral albendazole and corticosteroids is given in confirmed cases for minimum of 4 weeks, after intraocular and intracranial cysticercosis are ruled out by indirect ophthalmoscopy and imaging respectively. Intraocular cysticercosis is associated with a poor prognosis for vision, though sub-retinal cysts can be removed by an intravitreal or transscleral approach, the results are however not very good.

Vasculogenic lesions
In 1999, the Orbital society created a new classification of vascular lesions based on hemodynamic behavior (Table-I). This new system guides management and prevents high risk and unnecessary surgical intervention[17].

Capillary Hemangioma
Capillary hemangiomas are one of the most common benign orbital masses in pediatric patients. One-third of all orbital capillary hemangiomas are diagnosed at birth, and more than 90% are visible by 6 months of age.[18] The lesion can present in one of three ways. Superficial involvement appears as telangiectatic vessels in the skin, with time the lesion becomes more raised and nodular, developing a strawberry like appearance. Deeper lesions form raised, soft, purplish nodules. Deep orbital involvement may have no overlying skin changes and presents solely with proptosis. The child presenting with a typical capillary hemangioma will have a superficial, raised, red or deep blue lesion in the eyelid, conjunctiva or anterior orbit, which may enlarge with valsalva maneuvers or crying (Figure 3)[19]. These lesions enlarge in size until the patient is about 2 to 3 years old, and thereafter stabilize and then start involuting from four years of age and most doing so by the seventh year. Visual complications from capillary hemangiomas are common with amblyopia affecting 43% to 60% of patients having eyelid or orbital involvement.[20] Other major complications of capillary hemangiomas include superinfection, ulceration, hemorrhage and necrosis. Two rare complications are Kasabach-Merritt syndrome, characterized by sequestration of platelets and fibrinogen in the lesion resulting in coagulopathy and high output cardiac failure is seen in patients with large capillary hemangiomas.

For delineating large, diffuse and infiltrating lesions, imaging modalities like CT scan, ultrasound, MR imaging and technetium-99m-labelled red blood cell scintigraphy are useful[21]. The masses are usually well circumscribed and enhance with contrast, although bony remodeling may be seen, no erosion of bone should be evident. Treatment is indicated if there is compromise of the visual axis, high induced astigmatism, optic nerve compression, or exposure keratitis because of severe proptosis. Therapy generally falls into medical, radiotherapeutic, or surgical treatment.

Medical therapy involves the use of intralesional steroid injection, systemic steroids, or interferon. Combining long and short acting steroids (Triamcinolone+ Betamethasone) is common. Complications include eyelid necrosis[22], hypopigmentation of the skin[23], localized fat atrophy and rarely central retinal artery occlusion. Interferon and systemic corticosteroids are very effective, although systemic side effects are common. Surgical excision is reserved for well-circumscribed lesions or those causing severe functional sequelae that are unresponsive to medical therapy.

Lymphangiomas
Lymphangiomas are benign hamartomatous malformations. They may be present in the conjunctiva, eyelids, or deep in the orbit. Classically they are viewed as separate from the vascular system, although some overlap is seen between lymphangiomas and varices, or so called “combined lesions”[24]. Histopathologically, lymphangiomas may or may not be blood filled and characteristically have lymphoid cell aggregates with or without germinal centers. The majority of lymphangiomas appear in the first two decades of life and usually have a gradual course and undergo slow enlargement with increasing proptosis over many years. These may suddenly undergo expansion secondary to intralesional hemorrhage (chocolate cyst) or due to expansion of lymphoid elements during an acute upper respiratory infection and present with sudden proptosis. Hence, a careful history may reveal sudden painful proptosis, facial trauma or that the tumour or proptosis started right after a upper respiratory infection. Physical examination may reveal bluish discoloration of blood vessels within the eyelid skin; however if the vessels extend under the conjunctiva, and are pale straw colored they are called lymphangiectasias. Clinically, these lesions can be differentiated from orbital varices by the lack of enlargement on valsalva maneuver. Additional lesions may be found elsewhere in the body, including on the mucous membranes of the mouth. Non-contiguous intracranial vascular anomalies associated with orbital lymphangiomas have been described[24]. On CT scanning, these lesions can be well-circumscribed or diffusely infiltrative because of a lack of encapsulation. Critical orbital tissues may be encased or difficult to identify. The lesions show either no enhancement or rim enhancement of the large cystic spaces, as in loculated lesions. Phleboliths may be seen[25]. MR imaging may also be useful in delineation of the lesion prior to attempted surgery.
Management requires mainly observation initially for growth (clinical and radiographic studies) prior to considering intervention. Treatment of lymphangioma is typically indicated when it is associated with growth, optic nerve compression, corneal exposure, glaucoma or evidence of vision deterioration. Surgically, most patients require several debulking surgeries to relieve acute optic nerve compression or corneal exposure. In rarest of cases, orbital lymphangioma patients may require exenteration of the orbit.

Orbital varix
The orbital varix is a rare orbital vascular lesion, it is actually a vascular hamartoma typified by plexus of low pressure, low-flow, thin walled and distensible vessels that intermingle with normal orbital vessels. This lesion generally involves the superior ophthalmic vein; however other veins of the orbit can also be affected.

Most patients with an orbital varix develop positional proptosis secondary to its connections to the systemic venous circulation. The proptosis is exacerbated when the patient assumes a prone position, bends over, as performs a valsasla maneuver. Rarely, the lesion can have an acute presentation with painful proptosis and decreased visual acuity, due to the thrombosis or hemorrhage of the affected vein. CT and MRI images show an irregular mass usually located in the posterior orbit. CT is superior to MRI in the demonstration of phleboliths and to rule out orbital wall defects.

The treatment of orbital varix is difficult. Usually the anterior portion of the lesion is excised surgically. Alternatively, drainage of the blood clot and electrocauterization of the vessel wall have been done. It is seldom possible to remove the entire lesion, especially when it is located posteriorly in the orbit. Recently there have been studies indicating the role of percutaneously injected n-butyl cyanoacrylate (NBCA) to embolize orbital varices followed by surgical resection, as an aid in visualization and hemorrhage prevention during surgical resection of symptomatic orbital varices[26].

Orbital abscess
Orbital abscess is a well delineated form of orbital cellulitis or rather a complication there of characterized by collection of pus within the orbital tissues[27]. Adjacent sinus disease accounts for most of orbital cellulitis in children[28,29].

Ethmoidal sinusitis is the most common etiological factor, others being trauma, skin infections, dental infections, otitis media, intraorbital foreign bodies, endophthalmitis, dacryoadenitis, squint surgery, retinal buckling procedures, bacteremia & HIV[30,31].

Orbital cellulitis / abscesses in children commonly present as an acute febrile illness, with painful proptosis, lid swelling, chemosis and impaired ocular motility. These can be associated with visual deterioration, pupillary abnormalities, colour vision defects, and field defects[29,32].

The classification of orbital infections by Chandler et al. (Table-III) does not necessarily imply an order of disease progression; however, it helps explain the physical signs and symptoms of the various infections and helps organize treatment plans. Etiological agents implicated include Staphylococcus aureus, Streptococcus pneumoniae, Hemophilus influenzae, anaerobes, Propionibacterium acnes, Pseudomonas, Pneumocystis, mixed infections and rarely parasites and fungi[33]. Leukocytosis and blood cultures are unreliable. CT scans with coronal sections are useful. It is important to note that computerized scans cannot predict whether the mass represents a hematoma, an exudate or a transudate[34]. MRI is frequently necessary for patients with intracranial infections[34]. Ultrasonography is a useful adjunct.

Aggressive parenteral antibiotics with judicious surgical intervention constitute the mainstay of treatment. All children have to be admitted before initiating therapy, even if they lack orbital signs, because children are deficient in IgG2 and are predisposed to bacteremia. For orbital cellulitis, empiric antimicrobial therapy should be chosen to provide activity against S aureus, S pyogenes, and anaerobic bacteria of the upper respiratory tract in addition to the usual pathogens associated with acute sinusitis (ie, S pneumoniae, H influenzae, and M catarrhalis). Intravenous therapy is maintained until the infected eye appears nearly normal. At that time, oral antibiotic therapy can be substituted to complete a 3-week course of treatment. Nasal decongestants can be used to help drain the sinuses. Surgical drainage generally is not necessary for cellulitis; however, any patient of orbital abscess with compromised vision, well-defined abscess, exposure keratopathy or complete ophthalmoplegia and not responding to conservative treatment should receive surgery for drainage and debridement.

Optic nerve Glioma
Gliomas, benign intrinsic tumours of the optic nerve or visual pathway occur primarily in children, with a mean age at presentation of 8.8 years. These tumours occur isolated or may be associated with neurofibromatosis type-I. Histopathologically, optic gliomas arise from the astrocytes of the optic nerve.

Gliomas may occur anywhere along the visual pathways. The signs and symptoms at presentation are related to tumour...
location and patient's age. Optic nerve gliomas in preverbal patients typically present with axial proptosis (Figure 4). Older children may note a decrease in vision or a change in visual field. Other findings on examination include disc swelling, or optic atrophy and decreased motility. Gliomas with intracranial extension may come to the attention of the clinician because of complaints of pain and headache. On CT scan, gliomas appear as fusiform enlargement of the optic nerve. Intracranial gliomas are better imaged with MR imaging. Physiological tests can be used to determine the extent of lesions within the optic pathways. Surgery is the treatment of choice for gliomas of the optic nerve (as opposed to gliomas of the chiasm), particularly when there is profound visual loss and when there is significant proptosis. Radiation therapy for treatment of optic nerve and chiasmal gliomas remains controversial. Others recommend that radiation therapy be given early in the course of the disease to minimize the risk of visual deterioration[35]. Often chemotherapy is administered in an effort to defer the use of radiation therapy. Newer treatments like proton therapy could spare more normal tissue in children with optic pathway gliomas compared with 3D conformal radiation, particularly in larger tumours. As treatment has evolved over time from primary exenteration to
treatment of optic nerve gliomas, survival of these patients has increased dramatically.

**Neurofibroma**

Neurofibroma is a benign peripheral nerve tumour that can affect the orbit. It can be divided into localized, diffuse and plexiform type[36]. The localized type is clinically and radiographically similar to schwannoma and is associated with neurofibromatosis in about 10% of cases. The diffuse type has a variable association with neurofibromatosis and the plexiform type is almost always seen in association with neurofibromatosis. Localized neurofibroma, rare in children, can manifest as proptosis, globe displacement, diplopia and optic nerve compression. Diffuse and plexiform neurofibromatosis are very similar clinically and radiographically, but are classified separately because of subtle histopathologic differences. They usually occur in the first decade of life and show gradual progression, often with involvement of other periorcular and ocular tissues, including the uveal tract, the diffuse poorly defined mass can cause the classic S-shaped curve to the upper eyelid owing to subcutaneous involvement by the tumour. The plexiform type can be very extensive with massive involvement of the orbit, eyelids and intraocular structures. In addition, patients with neurofibromatosis can have congenital defects in the sphenoid bone that can produce a characteristic pulsating proptosis similar to that seen with encephalocele On orbital CT and MRI, it appears as an irregular, ill defined mass often with extensive periocular involvement. While the suspected localized orbital neurofibroma have to be excised completely, the diffuse, unresectable type should be managed more conservatively.

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Peripheral Retinal Degenerations

K Sudhamathi, Umah Venugopal, Ajay Sharma, Deependra V Singh
Eye-Q Superspeciality Eye Hospitals, Gurgaon

The vast array of peripheral retinal degenerations leave an ophthalmologist with numerous possible differentials and prognosticators when an individual presents to him. The following review is an aid in understanding which degenerations make retina susceptible to a possible retinal detachment in near future; and are possible indicators for prophylactic therapy at presentation and which of them do not predispose to any complications and thus can be left untreated.

Peripheral retina defined as an area anterior to equator of the eye is a site for potential pathologic lesions which may predispose to Rhegmatogenous Retinal detachment. This article focuses on differentiation of such lesions from benign peripheral retinal lesions, assessment of the risk of RD associated with them and decision making regarding prophylactic treatment of these lesions.

Relevant Anatomy
A sound understanding of the anatomical location of the peripheral retina structures and their relationship to one another is required. The peripheral retina is three to four disc diameters (3DD-4DD) wide.

The vortex veins and ampullae serve as very good landmarks for an examiner. These vortex ampullae and veins represent the equator of the eye.

Retina ends sharply at ora serrata, where parsplana of ciliary body begins. The border between the two is scalloped, with the scalloped teeth (dentate processes) facing anteriorly alternating with oral bays between the teeth. Pars plana of ciliary body extends 3-4mm anteriorly forwards to the Pars Plicata. Vitreous base spans the ora serrata and extends 1.5mm anteriorly onto the ciliary body and 1.5-3mm posteriorly onto peripheral retina. Vitreous is tightly adherent to retina at optic disc, vitreous base, edges of peripheral degenerations and along the retinal blood vessels. The attachment at vitreous base is considered to be the strongest.

Peripheral retina is best visualized with either 3-mirror Goldman lens or indirect ophthalmoscopy.

The primary reason for performing a dilated examination of the peripheral retina is to detect potential predisposing lesions[1] that can lead to retinal detachment (RD). Retinal detachment constitutes separation of the sensory retina from the underlying retinal pigment epithelium, (RPE) due to

| Table-1 Features of Benign peripheral retinal degenerations (not predisposing to RD) |
|-------------------------------------------------|------------------|--------------------------|-------------------------------------------------|
| Benign Lesions                                | Demography       | Natural Course           | Appearance                                      |
| Paving stone Degeneration                      | 27%              | Rarely lead to breaks    | ○ Atrophy or Absence of Outer Layers of Retina  |
|                                                |                  |                          | ○ Loss of RPE                                    |
|                                                |                  |                          | ○ Absence of Chorio-capillaries                 |
|                                                |                  |                          | ○ Lesions occur as Single or Confluent areas    |
| Congenital RPE hyper trophy                    | Common, no estimate available. Unilateral 1-2% bilateral, in these cases rule out Gardner’s syndrome | Rarely Causes RD                              | ○ Large, Well Demarcated, Usually Black area   |
|                                                |                  |                          | ○ Apex pointing to the Optic Nerve- Bear Tracks |
|                                                |                  |                          | ○ Histologically: Enlarged RPE cells, with Large |
|                                                |                  |                          | ○ Spherical Melanin Granules, or Macromelanosomes |
| RPE hyper plasia                              | Reactive process sec. to infection, trauma or uveitis | Does not cause RD | Posterior to the Ora Serrata along the Vitreous Base. Flat black pigmentation with irregular margins
| Peripheral Cystoid Degeneration               | 18%              | May lead to Retinoschisis| Cavitation of Outer plexiform & nuclear layers   |
| White Without Pressure Areas                  | 5%,in Ged 66% 23% in Myopia | Rarely breaks seen. Considered by some as precursor to GRT | Optical phenomena in which vitreous traction changes color of retina
the accumulation of liquefied vitreous fluid in that potential space. Most RDs are the result of one or more retinal breaks that allow passage of fluid between the sensory retina and the RPE. Since occurrence of RD is frequently associated with moderate to severe loss of vision, it is always imperative to subject all the patients at high risk to periodical peripheral retinal examination. Also it is important to distinguish dangerous from benign peripheral retinal lesions[2].

Peripheral retinal conditions can be classified as –
Benign Peripheral retinal lesions not predisposing to RD.
1) Paving stone/Cobble stone degeneration
2) Peripheral cystoid degeneration
3) RPE Hyperplasia
4) Congenital RPE hypertrophy ‘Bear Tracks’
5) White-without-pressure.

Rhegmatogenous Peripheral retinal lesions
1) Lattice degeneration
2) Retinal Tufts
3) Meridional folds and Complexes
4) Snail Track Degeneration
5) Snow Flake degeneration
6) Pigmentary degeneration and pigment clumps

White-Without-Pressure
W-W/O-P is a fairly common retinal finding and is related to vitreous traction. It causes the retinal surface to have a thin whitish appearance which can even slightly change its location between examinations. It is not uncommon for a narrow zone of optically darker retina to be on the posterior margin of W-W/O-P. Scleral depression can enhance the whitish appearance of the entity and the posterior margins. W-W/O-P is also associated with such retinal degenerations as lattice degeneration and retinoschisis.

White-with or -without-pressure is found to some extent in over 30% of normal eyes, with a strong tendency toward bilateralism. Individuals under 20 years of age have only a 5% occurrence, while those over 70 years of age have approximately a 66% frequency. A study of myopes found a prevalence of 0% in myopic eyes with the shortest axial length and 54% in eyes with axial lengths over 33 mm. In patients of all ages, it is most frequently found in myopic patients at 22.8%. This lesion is rarely associated with retinal break formation.

Lattice Degeneration
Lattice degeneration is a Vitreoretinal pathology that produces pockets of liquefied vitreous and retinal thinning, both of which can result in retinal tear formation and subsequent detachment of neurosensory retina[5]. Lattice lesions are usually hyperpigmented, mottled looking and most often located in the far periphery of the retina. They
Peripheral Retinal Degenerations

Figure 1 Pigmented lattice with holes pre (1A & 1B) and post laser (1C & 1D).

Figure 2 Old Rheg. RD from lattice with holes (2A) and lattice with large atrophic hole in fellow eye. Non pigmented lattice pre (2C) and post laser (2D).

Figure 3 Large Flap tear from PVD with laser marks 3A, Rare retinal tear at the anterior margin of lattice 3B, Flap tear at the posterior margin of lattice pre (3C) and post laser (3D).

Figure 4 Juvenile retinoschisis with cart wheel macula (4A), Large inner wall holes pre (4B,4C) and post laser delimitation (4D).

Figure 5 Low/no risk peripheral degenerations; Fundus photographs showing Paving stone degenerations (5A & 5B), snowflake degeneration (5C) and Cystic tuft (5D).

Figure 6 White without pressure.
are elongated lesions with the long axis parallel with the ora serrata and they can be singular or multiple in number. Lesions which are around blood vessels are known as perivascular lattice degeneration.

Lattice degeneration tends to be both fairly symmetric and bilateral, affecting both eyes 33.0%–48.1% of the time. The number of lesions per eye can vary from 1 to 19 or more and the average number per eye is approximately 2. There is always fairly normal appearing retina between lattice degeneration and the ora serrata and this is important in differentiating prominent vitreous base from lattice. On occasion, only the posterior margin of a pigmented area in the far periphery may be seen during ophthalmoscopy and it may give rise to the presumptive diagnosis of lattice degeneration. The pigmented area should be viewed under scleral depression so as to be able to view anterior to the lesion and if the pigmented area is adjacent to the ora serrata than the diagnosis would most likely be a prominent vitreous base, but if retina is found anterior to the lesion than the most likely diagnosis would be lattice degeneration.

White lines may display a crisscrossing pattern, the appearance that is responsible for the term lattice degeneration[5]. White lines are not commonly seen in young patients, only being found in 3.3% in the 10-19 year old age group, but they do increase in frequency with advancing age, reaching a maximum prevalence of 42.9% after age 50 years. Abnormal pigmentation is the most common finding in lattice degeneration and occurs in 81.7% and 92% of lesions. This pigmentation found in the retina and choroid seems to increase with age. The second most common feature is tiny white or yellow flecks, which are seen in 80% of lesions and are located between the retinal surface and the vitreous cortex.

Lattice degeneration is associated with W-W/O-P, atrophic holes, and linear and flap tears. Atrophic holes may form in a lesion due to the loss of all of the sensory retinal tissue, and occur in 18.2% to 28.7% of cases. The frequency of retinal detachment caused by atrophic holes in lattice degeneration is fairly low and has been reported to be 2.8% and 13.9%. Retinal tears have been reported in 2.4% in 125 autopsied eyes with lattice degeneration and another report found retina tears in 1.5% of 289 patients with lattice lesions followed for 3-10 years. Other reports found retinal tears in 1.0% of eyes with lattice degeneration. Lattice with tractiveal tears or a mixture of breaks was found to be responsible for retinal detachments in 16% to 27% of all primary detachments. Two reports found that 55% to 70% of retinal detachments in eyes with lattice degeneration were caused by tears at the posterior edge of the lattice lesions. Scleral depression enhances the visualization of these lesions and for confirming the existence of retinal breaks.

Lattice degeneration is significant in that it is the most commonly associated retinal finding in rhegmatogenous RDs undergoing surgery. Lattice degeneration and retinal detachment have a significant association; 20% to 41% of patients operated for a rhegmatogenous retinal detachment have lattice degeneration present in the eye. This does not mean that all patients with lattice degeneration are likely to develop a retinal detachment; in fact it has been estimated that this occurs in only 0.3% to 0.5% of patients or 1 in 200-300 patients with the disease.

**Decision Making**

**Natural History of Precursors to Rhegmatogenous Retinal Detachment**

Precursors to retinal detachments are PVD and symptomatic retinal breaks. Nearly all patients with a symptomatic RRD will progressively lose vision unless the detachment is repaired. Prevention or early diagnosis is important because the rate of successful reattachment is higher and the visual results are better if detachment spares the macula. Successful treatment allows patients to maintain their abilities to read, work, drive, care for themselves, and enjoy a better quality of life.

**Posterior Vitreous Detachment**

Posterior vitreous detachment is the cause of many retinal breaks, which can then lead to retinal detachments. The symptoms of PVD include light flashes and floaters, and patients with such symptoms are at significant risk for retinal detachment[3,4]. Approximately 15% of patients with acute symptoms of PVD have a retinal tear at the time of the initial examination. Patients with acute PVD who have no retinal breaks on presentation have a 2% to 5% chance of developing them in the weeks that follow. In patients who present with substantial vitreous hemorrhage, 67% were found to have at least one break, with 31% having more than one break and 88% of the breaks occurring in the superior quadrants.

**Symptomatic Retinal Breaks**

A symptomatic retinal break is defined as one caused by vitreoretinal traction in a patient with a new PVD or a break associated with a significant increase in flashes and floaters. Approximately one half of untreated symptomatic retinal breaks with persistent vitreoretinal traction (horseshoe or flap tears) will cause a clinical retinal detachment unless treatment is applied.

**Risk Factors for Rhegmatogenous RD.**

Aside from retinal breaks, risk factors for RRD include myopia, lattice degeneration, cataract surgery, trauma, and a history of RRD in the other eye. Combinations of these factors in a single eye further increase the risk.

**Myopia**

More than half of nontraumatic RRD occurs in myopic
eyes. As axial length increases, so does the risk of RRD[4]. As compared with that of emmetropes, low myopes (1 to 3 diopters) have a 4-fold risk and higher myopes (>3 diopters) have a 10-fold risk. The tendency to develop atrophic breaks, absence of Gel vitreous to plug holes and occurrence of early and acute PVD are factors responsible for high risk of RD in myopes.

Lattice Degeneration
Lattice degeneration, increases the risk of retinal detachment[5]. Lattice degeneration is present in 6% to 8% of the general population; an individual with this disease has a high risk of RRD.

Cataract Surgery
The overall risk of RRD after cataract surgery is approximately 1%. Two large studies found that the risk of RRD after cataract surgery is 6 to 7 times greater than that of phakic control groups[7,8,9]. The following have been reported to increase the risk of retinal detachment after cataract surgery: vitreous loss; increased axial length; lattice degeneration; Nd:YAG laser capsulotomy; Caucasian race; and younger age. A recent study has found the risk to be higher for as long as 20 years.

Trauma
Patients with blunt or penetrating ocular injuries that have altered the structure of the vitreous or retina are at increased risk of RRD[10]. Although nearly all breaks caused by blunt trauma occur at the time of the injury, the detachment may not be symptomatic for years because the younger age group at risk for trauma has a formed vitreous. Trauma also accelerates the development of PVD.

Rhegmatogenous Retinal Detachment in the Fellow Eye
Patients with a history of non-traumatic detachment in one eye have about a 10% increased risk of developing RRD in the fellow eye[11]. Because pathologic Vitreoretinal changes are frequently bilateral. A pseudophakic RRD is not necessarily caused by cataract surgery alone. The fellow eye in a patient with pseudophakic retinal detachment is also at higher risk for developing a retinal detachment, whether the fellow eye is phakic or pseudophakic. Phakic fellow eyes in patients with pseudophakic retinal detachment have about a 7% risk of RRD, indicating that all the risk for developing RRD cannot be attributed to cataract surgery alone.

Early Detection and Prevention
There are no effective methods to preventing vitreous changes that lead to RRD. If factors associated with an increased risk of retinal detachment are discovered during a routine eye examination in an asymptomatic patient, a peripheral fundus examination is advisable. Patient at high risk should also be educated about the symptoms of PVD and retinal detachment as well as about the values of periodic follow-up examination[12].

Diagnosis
History
- Symptoms of PVD
- Family history of Rhegmatogenous RD
- Prior eye trauma, including surgery
- Myopia
- Examination of the vitreous for PVD, pigmented cells, hemorrhage, and condensation
- Peripheral fundus examination with scleral depression.

There are no symptoms that can reliably distinguish PVD with an associated retinal break from PVD without an associated retinal break; therefore, a peripheral retinal examination is always required. The preferred method of

<table>
<thead>
<tr>
<th>Type of Lesion</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute symptomatic horseshoe tears</td>
<td>Treat promptly</td>
</tr>
<tr>
<td>Acute symptomatic operculated tears</td>
<td>Treatment can be considered</td>
</tr>
<tr>
<td>Traumatic retinal breaks</td>
<td>Usually treated</td>
</tr>
<tr>
<td>Asymptomatic horseshoe tears</td>
<td>Treatment should be considered</td>
</tr>
<tr>
<td>Asymptomatic operculated tears</td>
<td>Treatment is rarely recommended</td>
</tr>
<tr>
<td>Asymptomatic atrophic round holes</td>
<td>Treatment is rarely recommended</td>
</tr>
<tr>
<td>Asymptomatic lattice degeneration without holes</td>
<td>Not treated unless PVD causes a horseshoe tear</td>
</tr>
<tr>
<td>Asymptomatic lattice degeneration with holes</td>
<td>Can be treated if other risk factors are present</td>
</tr>
<tr>
<td>Asymptomatic dialyses</td>
<td>No consensus on treatment due to insufficient evidence to guide management, most surgeons treat</td>
</tr>
<tr>
<td>Fellow eyes with atrophic holes, lattice degeneration, or asymptomatic horseshoe tears</td>
<td>Treatment advocated by most surgeons.</td>
</tr>
</tbody>
</table>
evaluating peripheral vitreoretinal pathology is with indirect ophthalmoscopy combined with scleral depression.

**Diagnostic Tests**
A B-Scan maybe required in case of opaque media like Cataract or Vitreous Hemorrhage secondary to retinal tear or any other cause.

**Treatment**
Table 3 summarizes recommendations for management. Treatment of peripheral horseshoe tears should be extended well into the vitreous base, even to the ora serrata. The surgeon should inform the patient of the relative risks, benefits, and alternatives to surgery. The surgeon has the responsibility for formulating a postoperative care plan and should inform the patient of these arrangements[13].

**Follow-up**
The guidelines in Table 4 are for routine follow-up in the absence of additional symptoms. Patients with no positive findings at the initial examination should be seen at the intervals as recommended in the Table 4. All patients with risk factors should be advised to contact the ophthalmologist promptly if new symptoms such as flashes, floaters, peripheral visual field loss, or decreased visual acuity develop.

**Useful Tips**
**Examination**
- Whenever doubtful indent and depress
- Use both 28d and 20d lens
- Scan anterior to the lesion
- Change posture of the patient, supine is best but upright position gives best view
- Try to take fundus picture if one can capture it, then one can easily barrage it by slitlamp laser photoagulation.

**Treatment**
- Superior lesions more dangerous than inferior ones.
- Patients > 40 years of age without PVD to be treated in view of risk of PVD induced flap tear at the margin of lesion.
- Use more number of smaller spots than large intense burns.
- Always explain to patient the symptoms of PVD and importance of prompt retinal examination after experiencing those symptoms.

**References**
New Insights in Primary Angle Closure Glaucoma

Tanuj Dada, Gaurav Kumar, Lalit Tejwani, Vishal Arora, Meenakshi Wadhwani, Anita Panda
Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi

Primary angle closure glaucoma is one of the major causes for irreversible visual loss in Asia. In addition to pupillary block, lens induced mechanisms, plateau iris and supra-ciliary effusions are important contributing factors elucidated by Ultrasound biomicroscopy. Treatment modalities for management of primary angle closure disease start with a laser iridotomy and include laser iridoplasty, lens extraction, combined procedures with goniosynechiolysis and glaucoma filtering surgery. This article is to give insight into newer concepts in treatment of angle closure based on the improvement in knowledge of pathophysiology and natural history of the disease.

According to the International Society of Geographical and Epidemiological Ophthalmology (ISGEO) Classification, primary angle closure glaucoma is subdivided according to conceptual stages in natural history of angle closure of glaucoma into primary angle-closure suspect (PACS), primary angle closure without optic neuropathy (PAC), and primary angle-closure glaucoma with neuropathy (PACG)[1].

- PACS- Irido trabecular contact (ITC) with normal optic disc and visual field. IOP is normal and PAS is absent.
- PAC- ITC + either raised IOP, PAS or typical symptom
- PACG-ITC + structural glaucomatous changes in optic nerve + Visual field loss.

These categories may clinically overlap or be potentially related, and the natural history of the disease is such that the patient may present to the clinician at any stage of the clinical spectrum. The most important fact to remember about this disease, however, is that a significant reduction in morbidity from this condition is possible as most cases can be detected with available technology; and many cases treated with the timely single application of a simple, safe treatment.

Epidemiology

Of the estimated 67 million people worldwide thought to be affected with primary glaucoma, one-third to one half have primary angle closure glaucoma (PACG)[2]. One of the major factors determining susceptibility to primary angle closure is the ethnic background. In European and African populations primary open-angle glaucoma (POAG) is reported to be approximately five times more common than PACG; while in the Chinese[3,4], Mongolians[5] and Indians[6] the frequency of PACG may equal or be greater than POAG. In Eskimos/Inuit the prevalence of PACG has been found to be higher than any other ethnic group[7].

PACG is 2 to 3 times more likely to cause visual impairment than is primary open-angle glaucoma[3]. The incidence of PAC increases with age, peaking in incidence between 55 and 70 years of age[8] and is greater in females[9]. Population-based studies have shown that most cases of PACG are asymptomatic, whereas chronic PACG may develop after the resolution or precede the occurrence of an acute attack of angle closure[10,11]. Data from India Vellore Eye Study (VES)[12] and Andhra Pradesh Eye Disease Study (APEDS)[13] shows a prevalence of 4.32% and 0.71% for PACG. Hospital based data suggests an equal number of people with POAG and PACG. Prevalence of primary angle closure disease (PACG + PAC) in the rural population according to Chennai Eye Disease Incidence Study[14] is 1.58%. An additional 7% were at risk of developing angle closure glaucoma.

Mechanisms of Angle Closure Glaucoma

Most common mechanism of primary angle closure is pupillary block. The simultaneous activation of dilator and sphincter pupillae muscles produces a resultant force whose vector lies more or less perpendicular to lens surface when the pupil is in mid-dilated position. These pathologic mechanisms exist because of primary anatomic variations in the size, position, and relationship of the anterior segment structures (cornea, iris, ciliary body, lens). In plateau iris[15] there is anterior rotation of ciliary body pushing iris anteriorly crushing peripheral shallow angle and deep central anterior, chamber. A large anteriorly displaced lens may also be an important factor contributing to primary angle closure glaucoma.

Role of Lens in Pathogenesis

Eyes with primary angle closure have significant anatomic differences from normal eyes[16]. The most significant clinical hallmarks of an eye with angle-closure are the shallow AC and narrow angle. The mean anterior chamber depth (ACD) in PAC eyes is approximately 1.8 mm, which is 1 mm shorter than in normal eyes[16,17]. Angle closure becomes a rarity when anterior chamber depth exceeds 2.5 mm[18]. Decreased AC volume[19], small corneal diameter[20], and short axial...
After better understanding the pathophysiology of the disease, factors, and individualized on a case to case basis. Surgical decisions for angle closure should be controlled aggressively with reassessment of the target IOP at each stage. The management of PACG requires repeated ocular examinations with special emphasis on the evaluation of the filtration angle to determine the mechanism of the angle-closure and the clinical stage of the disease. The first treatment of an acute attack is primarily symptomatic, but must be followed by care to prevent the development of chronic angle closure glaucoma. In patients with established synechial closure and optic neuropathy the IOP must be controlled aggressively with reassessment of the target IOP at each stage. Surgical decisions for angle closure should be taken after careful evaluation of anatomic and physiological factors, and individualized on a case to case basis.

After better understanding the pathophysiology of the disease, with the help of newer investigations ultrasound biomicroscopy (UBM) and anterior segment OCT (ASOCT, Visante) more knowledge of role of lens in angle closure glaucoma has been found. Treatment of angle closure and prevention of further progression of the angle anomaly is therefore now based on a better understanding of the pathomechanism of angle closure.

**Laser Peripheral Iridotomy (LPI)**

It eliminates the pressure differential between the anterior and posterior chambers by providing alternative route aqueous trapped in posterior chamber to enter the anterior chamber. It is also a safe and effective prophylaxis in suspect eyes with occludable angles secondary to pupillary block, including fellow eyes of patients at risk for bilateral angle closure. LPI has proven to be an effective treatment for APAC, resulting in widening of the filtration angle and reduction of elevated IOP, it may not protect against chronic angle closure.

A study in Asian eyes with acute PAC that had undergone laser PI showed 58.1% continued to have elevated IOP and 32.7% eventually required trabeculectomy. PI does not always provide long term protection, recurrent attack can cause PAS formation. A study showed 0.65 mm of shallowing, accounting for the total of 1 mm difference in AC depth of the smaller eye compared to the normal eye. Growth of the lens, with an increase in the number of lens fibers continuing throughout adult life, results in an increase in lens thickness and anterior curvature.

Lowe also developed an index of relative lens position calculated as

**Relative lens position**

Anterior chamber depth + ½ (lens thickness) axial length. When the biometry of contralateral eyes of patients having an acute primary angle closure APAC were studied and compared to population-based controls, unfavorable dimensions were found consisting of more shallow anterior chambers and narrow angles, and thicker lenses. These differences were considered to explain in part the estimated 50% risk for APAC in these eyes. Decreased ACD is accelerated in women between the fourth and fifth decades, which may explain their greater propensity for PAC in females.

Relative resistance to flow of aqueous from the posterior chamber (PC) into the anterior chamber (AC) increases greatly when the dimensions of the iris–lens channel are changed in such a manner that flow of aqueous is more impeded. This incremental pressure differential determines the iris contour. As this pressure increment increases, the iris becomes more convex. Clinically significant pupillary block is present when the increased iris convexity brings the iris into apposition with the trabecular meshwork, with extreme anterior iris-bulging, is known as iris bombe.

**Management**

The management of PACG requires repeated ocular examinations with special emphasis on the evaluation of the filtration angle to determine the mechanism of the angle-closure and the clinical stage of the disease. The first treatment of an acute attack is primarily symptomatic, but must be followed by care to prevent the development of chronic angle closure glaucoma. In patients with established synechial closure and optic neuropathy the IOP must be controlled aggressively with reassessment of the target IOP at each stage. Surgical decisions for angle closure should be taken after careful evaluation of anatomic and physiological factors, and individualized on a case to case basis.
control without antiglaucoma medication during the mean follow up of 22 months[38]. Due to low success rate and high rate of complications trabeculectomy in PAC is becoming less popular compared to lens removal. Trabeculectomy should be performed first in eyes with advanced glaucomatous optic neuropathy with central 10 degrees of visual field involvement where IOP is not controlled despite topical medications (atleast 3 topical medications including one prostaglandin ) post laser iridotomy. In such eyes trabeculectomy must be performed with 2 releasable sutures to prevent shallow anterior chamber in the post operative period. The patient must be counselled regarding the requirement for cataract surgery in the near future.

**Role of Lens Removal**

Lens removal seeks to correct persistent pupillary-block and angle crowding after LPI, and therefore is the definitive procedure in both the treatment and prevention of acute and chronic angle closure glaucoma. It relieves all 3 mechanisms of primary angle closure related to pupillary block, plateau iris and the thick anteriorly displaced lens. Hayashi et al[39] demonstrated that anterior camber depth and angle width in ACG eyes approximates that of POAG eye and control normal eyes. In a study done on chronic angle closure patients showed mean increase in ACD and angle width from 2.04 mm to 3.44 mm this was due to exchange of the thickened lens (5mm) for IOL (1mm)[40,41].

**Surgical Tips and Caveats**

With advancement and increased skill of cataract it is possible to do safe and successful cataract surgery in angle closure patient. Surgical challenges in doing cataract surgery in angle

### TABLE 1 Effect of lens removal in primary angle closure disease

<table>
<thead>
<tr>
<th>Study (year)</th>
<th>Lens Procedure</th>
<th>Glaucoma Type (# of eyes)</th>
<th>Preop Gonioscopy</th>
<th>Follow-up (months)</th>
<th>Preop/Postop (mean IOP mmHg)</th>
<th>Success % IOP&lt;22mmHg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Greve42(1988)</td>
<td>ECCE PCIOL</td>
<td>AACG (5)</td>
<td>Near or complete closure</td>
<td>Range 6-42</td>
<td>31 to 16</td>
<td>76%</td>
</tr>
<tr>
<td>Gunning43(1991)</td>
<td>ECCE PCIOL</td>
<td>CACG (41)</td>
<td>PAC</td>
<td>Mean 14.3</td>
<td>22.6 to 15.6</td>
<td>65%</td>
</tr>
<tr>
<td>Gunning44(1998)</td>
<td>ECCE PCIOL vs Trabeculectomy</td>
<td>CACG (22)</td>
<td>PAS 77%eyes</td>
<td>Mean 53</td>
<td>28 to 17</td>
<td>Overall success of 68% reported in both groups</td>
</tr>
<tr>
<td>Roberts45(2000)</td>
<td>PHACO PCIOL</td>
<td>AACG (18)</td>
<td>360°PAS (2)</td>
<td>36, 24 mos</td>
<td>39 to 17</td>
<td>67% (2 eyes)</td>
</tr>
<tr>
<td>Lai46(2001)</td>
<td>PHACO PCIOL GSL DLPI</td>
<td>CACG (7)</td>
<td>360°PAS (7)</td>
<td>Mean 9</td>
<td>33 to 13</td>
<td>100%</td>
</tr>
<tr>
<td>Hayashi47(2001)</td>
<td>PHACO PCIOL</td>
<td>CACG (68)</td>
<td>No data</td>
<td>Mean 25</td>
<td>21 to 15</td>
<td>41%</td>
</tr>
<tr>
<td>Jacoby48(2002)</td>
<td>PHACO PCIOL CSI</td>
<td>AACG (43) AACG (32)</td>
<td>Partial closure (7) Partial closure (9)</td>
<td>Mean 10</td>
<td>41 to 18 40to 20</td>
<td>72% 35%</td>
</tr>
<tr>
<td>Kubota49(2003)</td>
<td>PHACO PCIOL</td>
<td>CACG(13)</td>
<td>PAS &gt;180° (5)</td>
<td>Mean 14</td>
<td>13 to 14</td>
<td>62%</td>
</tr>
<tr>
<td>Nonaka50(2005)</td>
<td>PHACO PCIOL</td>
<td>PAC (13)</td>
<td>2 Q closed by UBM</td>
<td>3</td>
<td>19 to 15</td>
<td>No data</td>
</tr>
<tr>
<td>Lai51(2006)</td>
<td>PHACO PCIOL</td>
<td>CACG (21)</td>
<td>&gt;90 - 270 closed</td>
<td>Mean 21</td>
<td>20 to 15.5</td>
<td>66.7%</td>
</tr>
<tr>
<td>Liu52(2006)</td>
<td>PHACO PCIOL</td>
<td>PAC(29) PAC/suspect(28)</td>
<td>PAS 9 clock hours (17%)</td>
<td>3</td>
<td>15 to12 14 to 12</td>
<td>41% 100%</td>
</tr>
<tr>
<td>Imaiizumi53(2006)</td>
<td>PHACO PCIOL</td>
<td>AACG (18) AACG (2)</td>
<td>No data</td>
<td>6</td>
<td>49 to13</td>
<td>100%</td>
</tr>
<tr>
<td>Pachimkul54(2008)</td>
<td>PHACO PCIOL</td>
<td>PACG(56)</td>
<td>6</td>
<td>6</td>
<td>23.3 to 14.8</td>
<td></td>
</tr>
</tbody>
</table>

**AACG** acute angle-closure glaucoma; **CACG** chronic angle closure glaucoma; **CD** choroidal detachment; **CSI** conventional surgical iridectomy; **ECCE** extracapsular cataract extraction; **GSL** goniosynechialysis; **IOP** intraocular pressure; **mos** months; **#** number; **PAC** primary angle closure; **PACG** primary angle-closure glaucoma; **PAS** peripheral anterior synechia; **PCIOL** posterior chamber intraocular lens; **PHACO** phacoemulsification.
closures patients are that there could be difficulty in access (small palpebral fissure), inflamed eye, high IOP, diminished red reflex, corneal epithelial and stromal edema, reduced working space due to shallow chamber and small pupil, PAS, large size of the lens and an already compromised corneal endothelium. There are increased chances of iris prolapse, problems during capsulorhexis, high capsular bag tension, lens subluxation, posterior capsular rupture, malignant glaucoma and suprachoroidal hemorrhage. To avoid these problems

- IOP must be controlled preoperatively, intravenous mannitol can be used for this purpose.
- Topical anesthesia is best if you are an expert surgeon, if you need to use peri-bulbar block inject only 3-5 ml and give intermittent digital massage.
- Atropine may be used for dilating the pupil.
- If IOP is found to be very high during surgery, a pars plana vitreous tap is an option using a 23/25G vitrectomy probe
- Gradual decompression of the anterior chamber is essential to minimize suprachoroidal hemorrhage.
- Intraoperative preservative free intracameral adrenaline can be used to dilate pupil.
- Use chilled BSS plus and Viscoat to coat and protect the corneal endothelium.
- Meticulous wound construction to avoid iris prolapse. (avoid posterior scleral entry)
- Use iris hooks or pupil ring expanders in small pupil.
- Straight phaco tip should be used as there is less room for manipulation in the crowded AC.
- Goniosynechiolysis may be done if the presence of PAS is confirmed by direct gonioscopy.
- Increased post operative reaction may occur due to excessive iris manipulation, intense topical steroid therapy with cycloplegia may be required in the post operative period.
- Put a drop of timolol at the end of cataract surgery to blunt post operative IOP spikes, may add oral acetazolamide.
- Must check digitally the IOP after you seal the wound and side port with BSS. Never leave a hard eye ball on the table in such eyes and elevated IOP can be very detrimental to the glaucomatous optic nerve head.
- Keep a close check on IOP in the post operative period. Some patients may be steroid responders – switch to bromofenac or nepafenac.

The chronicity and stage of the angle closure process determines the surgical outcome. In cases in which there is early optic disc cupping and mild visual field loss, lens extraction alone may be enough to achieve adequate IOP control; whereas eyes with advanced glaucomatous optic neuropathy are more likely to have poor residual trabecular meshwork function as a result of PAS which affect more than three quadrants or non-synechial damage. In such cases, an additional filtration procedure may be necessary for IOP control.

In recent years many studies are done and data are available showing the effect of lensectomy in PAC patients (Table 1).

### Goniosynechiolysis

It can be effectively done in cases of recent PAS formation (< 1 year), long-standing PAS are likely to be associated with permanent trabecular damage[55]. It is done with the help of direct gonioscope, after entering the chamber viscoelastic injected and synechiae released with the help of blunt tip spatula. Frequent complications include hyphema, fibrinous reaction and synechial reclosure of the angle. Given that the procedure does not address the underlying pathomechanism for synechial angle closure, be it pupillary-block or angle-crowding, its use is recommended as an adjunct to other procedures such as LPI[56], ALPI[57,58], or mainly lens extraction[60,61].

Razeghinejad MR has also shown that combined phacoemulsification and viscogoniosynechialysis seem to be an effective surgical procedure in the treatment of patients with CACG and angle restoration whether controlled or uncontrolled by medication[62].

### Phacoemulsification vs Laser PI for APAC

Lam DS et al[61] compared early phacoemulsification and peripheral iridotomy in acute primary angle closure and found that the prevalence of raised IOP at 18 months was 3.3% of the cases in phaco group vs 46.7% in LPI group. Angle in phaco group was more open compared to LPI group (mean Shaffer gonio grading 2.10 ± 0.76 vs 0.73 ± 0.64). PAS was more in LPI group 228.6 ± 89.2 compared to 101.3 ± 74.6 in phaco group. Requirement for topical antiglaucoma drugs was more in LPI group after 18 month followup. However, there were no statistically significant differences in visual acuity and visual fields between the 2 groups at 18 months. The authors thus concluded that early phacoemulsification is more effective in treatment of APAC as compared to laser iridotomy.

In a prospective nonrandomized trial done in Japan[63], primary phacoemulsification was compared with laser iridotomy in patients with CACG and PAC. In IOL group, IOP decreased from 14.8±4.2mm Hg to 10.8±1.6mm Hg (p <0.05), whereas in PI group 15.5±4.1 mmHg to 14.7±4.7mm Hg (p=0.76). In IOL group postoperative 6 months no glaucoma medication was required, PI group 0.2±0.4 (p <0.05) medication was required. Safety in both procedures was comparable.

### Phacoemulsification vs Phacotrabeculectomy

Two RCTs[64,65] were conducted in Hongkong comparing phacoemulsification with phacotrabeculectomy in medically controlled and medically uncontrolled chronic angle closure glaucoma. They showed lower intraocular pressures with phacotrabeculectomy than with phacoemulsification alone, but the difference was marginal and mostly statistically
New Insights in Primary Angle Closure Glaucoma

insignificant. More complication and more glaucomatous optic neuropathy deterioration was seen with phacotrabecectomy group. Tham CC et al have also shown in another recent study that combined phacotrabecectomy resulted in significantly more surgical complications than phacoemulsification alone in CACG eyes with coexisting cataract. They found that there was no difference in visual acuity or disease progression between the 2 treatment groups[66].

At R.P. Centre we evaluated the effect of phacoemulsification and foldable intraocular lens (IOL) implantation on biometric determinants of the anterior chamber angle in primary angle closure glaucoma (PACG) using Ultrasound Biomicroscopy (UBM). Forty six eyes of 46 patients with chronic PACG and cataract having a patent laser iridotomy were included in this prospective, interventional case series. Angle parameters were measured using UBM, before surgery and 3 months after phacoemulsification with IOL implantation. Intraocular ocular pressure (IOP) was measured by applanation tonometer and records of glaucoma medication administered were maintained. Main outcome measures were IOP, Central Anterior Chamber depth (ACD), trabecular iris angle (TIA), angle opening distance at 250 and 500 microns from scleral spur (AOD250 and AOD500). The mean age of study participants was 56.5 ± 9.9 years (range 44-75yrs). The preoperative mean IOP was 25.0 ± 5.4 mm Hg on maximum antiglaucoma medication which reduced to 15.8 ± 3.8 mm Hg (p=0.0001) at 3 months. Number of antiglaucoma medications also decreased from 2.4 ± 1.1 to 0.4 ± 1.1 (p=0.0001). There was a significant widening of the anterior chamber angle with the TIA increasing significantly after phacoemulsification (p<0.001) with an associated increase in AOD250, AOD500 and ACD (p<0.001). We found that Phacoemulsification in eyes with PACG results in significant widening of the anterior chamber angle. This results in better IOP control after surgery and decreases the need for glaucoma medications. These findings suggest that in eyes with primary angle closure glaucoma – lens extraction should be performed as the first stage procedure as it may control the IOP in majority of eyes with / without medical therapy. If IOP is not controlled medically, a trabeculectomy may be performed as a second stage procedure after 3 months.

So the big questions remains that in an eye with PACG with IOP not controlled medically post laser iridotomy, what is the preferred practice pattern with and without cataract?

Cataract present

Conventionally trabeculectomy has been performed first for IOP control in eyes with PACG. If trabeculectomy is performed first, it leads to development / progression of cataract, phacoemulsification becomes difficult in an eyes with shallow AC with a functional bleb and an already compromised endothelium, there is decrease in visual acuity and all structural and functional tests of glaucoma become difficult to perform and interpret due to presence of lenticular opacification. There is also a surgeon factor as many glaucoma surgeons are excellent when it comes to trabeculectomy but may not be the best of cataract surgeons and hence would not prefer phacoemulsification as the initial procedure. If on the other hand a lens extraction is done first (with IOP being controlled medically) – the patient’s vision improves, all investigations (visual fields, HRT, GDX, OCT etc) become easy to perform an interpret and it is much easier to perform a trabeculectomy in a pseudophakic eye with a deep anterior chamber. The only precaution that needs to be taken is that IOP must be medically controlled at all times during the follow up post cataract surgery.

So a two stage procedure seems to be the best option. Stage 1 – cataract surgery by temporal clear corneal phacoemulsification. IOP monitored and if required managed with topical medical therapy. If IOP not controlled medically:

Stage 2 – trabeculectomy with MMC after 3 months or more. If a patient is not amenable to follow up, has a poor compliance with topical ocular hypotensive therapy, advanced glaucoma with central 10 degrees visual field involvement or 360 degrees synechial closure of the angle confirmed on indentation gonioscopy : a phacotrabecectomy with MMC is the preferred option.

Clear lens

At this moment due to lack of scientific evidence coupled with ethical concerns, clear lens removal cannot be advocated as an option for the management of PACG. A laser iridotomy should be performed first and if there is presence of residual angle closure, ultrasound biomicroscopy should be performed to rule out plateau iris syndrome (treated by ALPI). The patient should be put on medical management with prostaglandin analogues being the first line therapy. If IOP is not controlled on medical therapy, a trabeculectomy should be performed with mitomycin c and use of releasable sutures to prevent early post operative shallowing of the anterior chamber.

Conclusion

The lens has an important role in the pathogenesis of primary and secondary ACG, and clinical studies suggest that lensectomy and PCIOL implantation for ACG patients may offer successful IOP control, with prevention of progression of the glaucomatous optic neuropathy. The mechanism for this is elimination of pupillary block and widening of the angle, thus reducing the iridotrabecular proximity. Medical management and ALPI remain the most common modes of treatment of an acute attack but newer approaches including early lens removal are gaining popularity as a definitive treatment that
addresses the anatomical anomaly. The use of the same during an acute attack and removal of clear lenses for management of PACG however, still remains controversial. Results from future randomized control trials will further elucidate the optimal therapeutic plan for this potentially blinding disease.

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**Major Review**

**Idiopathic Juxtafoveal Retinal Telangiectasis**

Neha Goel, Bhanu Pratap Singh Pangtey, Anisha Seth, Usha Kaul Raina, Basudeb Ghosh
Vitreo Retina Services, Guru Nanak Eye Centre, Maulana Azad Medical College, New Delhi

Idiopathic juxtafoveal retinal telangiectasis (IJRT), also known as idiopathic macular telangiectasia (IMT), refers to a heterogeneous group of well recognized clinical entities characterized by telangiectatic alterations of the juxtafoveal capillary network of one or both eyes, but which differ in appearance, presumed pathogenesis, and management. Classically, three groups of IJRT are identified. Group 1 is unilateral, easily visible telangiectasis occurring predominantly in males, and causing visual loss as a result of macular edema. Group 2, the most common, is bilateral occurring in both middle-aged men and women, and presenting with telangiectasis that is more difficult to detect on biomicroscopy, minimal exudation, superficial retinal crystalline deposits, and right-angle venules along with characteristic and diagnostic angiographic and optical coherence tomography (OCT) features. Vision loss is due to retinal atrophy, not exudation, and subretinal neovascularisation (SRNV) is common. Group 3 is very rare characterized predominantly by progressive obliteration of the perifoveal capillary network, occurring usually in association with a medical or neurologic disease. This article presents a current review of IJRT, including the classification, clinical features, pathogenesis, complications, differential diagnosis, and treatment modalities.

**Introduction**

Idiopathic juxtafoveal retinal telangiectasis (IJRT) is an uncommon cause of vision loss, which may be unilateral or bilateral. It comprises a group of retinal vascular anomalies characterized by retinal vessel dilation and tortuosity, multiple aneurysm formations, varying degrees of vascular leakage, incompetence and lipid exudate deposition. Visual loss is related to intraretinal edema, foveal atrophy and/or the development of subretinal neovascularisation (SRNV) [1,2]. Historically, there has been confusion differentiating Coats’ disease and IJRT. The term Coats’ disease is now reserved for congenital retinal telangiectasis associated with massive exudation, retinal detachment and retinal degenerative changes. This differs from IJRT, whereby exudation or diffusion abnormalities from incompetent capillaries are confined to the juxtafoveal region and are either of congenital or unknown origin[3].

**Classification**

Gass[2], who originally described this entity in 1968 and later coined the term IJRT in 1982, classified the disease into several types on the basis on biomicroscopic and angiographic findings[3]. In 1993, Gass and Blodi revised this classification and defined 3 distinct groups[4]. Group 1 patients have clinically visible retinal telangiectatic blood vessels and retinal exudation; group 2 patients have occult telangiectasis and minimal exudation; and group 3 patients have clinically visible telangiectasis, parafoveal capillary occlusion and minimal exudation. Subclassification of groups (Table 1) include those which are predominantly congenital, exudative and non-familial (groups 1A and IB), and those that are primarily acquired, non-exudative, obstructive and occasionally familial (groups 2A, 2B, 3A and 3B). Group 2A is the most common subtype reported and its development has been summarized into five stages (Table 2)[3]. Group 1A is the second most common.

In recent years, newly recognized manifestations have expanded and refined the clinical spectrum of these macular vasculopathies. Furthermore, the use of high speed angiography and optical coherence tomography (OCT) have provided a better understanding of the nature of the vascular abnormalities. In 2006, Yannuzzi et al proposed a simplified classification termed idiopathic macular telangiectasia (IMT) with 2 distinct types (type I, or aneurysmal telangiectasia, equivalent to group 1A and B and type II, or peripheral telangiectasia, equivalent to group 2A)[5]. The third type, occlusive telangiectasia, was omitted from the classification based on its rarity and presence of capillary nonperfusion rather than macular telangiectasia as the primary abnormality. Perifoveal telangiectasia was further classified in 2 stages: the nonproliferative stage when there are exudative telangiectasia and foveal atrophy, and the proliferative stage with the advent of SRNV.

**Pathogenesis**

Gass and Oyakawa[3] suggested that chronic venous stasis due to obstruction of the retinal veins as they cross retinal arteries on both sides of the horizontal raphe may be a cause of group 2A IJRT. Low-grade nutritional damage induced by specific retinal circulatory disturbances affects the retinal
cells, particularly those at the level of the inner nuclear layer, which includes the Müller cells, leading to degeneration and atrophy of these cells and the connecting photoreceptor cells resulting in growth of vessels and the migration of retinal pigment epithelial (RPE) cells into the retina[4]. Abnormalities of glucose tolerance may be found in cases with Type 2A IJRT[6]. This supports the hypothesis that bilateral disease occurs in relation to a widespread metabolic disturbance in the retina, whereas unilateral cases represent a local and truly vascular defect. Preliminary data using 2-wavelength autofluorescence imaging indicate that macular pigment density (MPD) is significantly reduced in the central retina. These recent findings have provided increasing evidence that group 2A IJT is not a disease limited to the retinal vasculature but that neurons are intrinsically involved as well[7].

Clinical picture and Diagnosis

The diagnosis of IJRT rests on a combination of stereoscopic biomicroscopy, fundus fluorescein angiography (FA) and OCT (Figures 1 – 4). On FA, the telangiectatic vessels are easily visible straddling the horizontal raphe and filling promptly in both the superficial and deep juxtafoveolar capillary plexus. Central cystic or noncystic macular edema is evident angiographically as late intraretinal staining. Diagnostic dilemma commonly exists to differentiate IJRT from occult SRNV or cystoid macular edema (CME) on FA. The merit of OCT is to provide information about the retinal structure and thickness in IJRT, as well as provide diagnostic clues in cases which are equivocal on FA. Following are the OCT features in IJRT[8,9]

1. Foveal cyst in the innermost retinal layers – most common finding
2. Internal limiting membrane (ILM) draping across the foveola related to an underlying loss of tissue
3. Intraretinal hyperreflective lesions – second most common finding. They correspond to ophthalmoscopically visible hyperpigmented lesions.
4. Disruption of the inner segment/outer segment (IS/OS) photoreceptor (PR) junction line
5. Foveal detachment
6. Blunting of the foveal pit/foveal flattening
7. Foveolar thinning
8. SRNV
9. Lamellar or full thickness macular hole[10,11]

Whenever there is absence of macular oedema on OCT in spite of prominent leakage of fluorescein in the fovea, IJRT must be suspected. Presence of foveal thinning despite occurrence of foveal cysts/detachment indicates that there is some degree of retinal atrophy and serves as a distinguishing feature of IJRT. Disruption of the IS/OS line can be visualised even in early cases with good vision, and does not necessarily indicate loss of the photoreceptor cells. Intraretinal RPE proliferation has been explained by the loss of PR cells, which allows the RPE cells to migrate into the overlying retina, especially along the venules. All eyes exhibiting RPE proliferation and migration demonstrate disruption of the IS/OS PR junction[8].

Macular holes (MH) may occur as a sequel to chronic macular oedema. We would then expect macular holes to occur more frequently in association with IJRT. However, the rarity of MH in IJRT as well as the preservation of good visual acuity in patients with MH implies that the holes were the result of lateral separation of the photoreceptors within the fovea and that there could not have been profound atrophy of the photoreceptors. There is a loss of the structural aspects afforded by Müller cells, particularly the Müller cell cone, in the central macula in IJRT[12].

Differential Diagnosis

When IJRT is suspected, it must be differentiated from venous occlusive disease, diabetic retinopathy, radiation retinopathy, Eales’ disease, carotid artery occlusion and sickle cell retinopathy. Group 1 patients, in addition to that noted above, should be distinguished from those with Coats’ disease which is defined by extensive peripheral retinal telangiectasis, exudative retinal detachment, relatively young age of onset and male predilection. Group 2 patients, during the early stage of the disease, may demonstrate foveolar atrophy that simulates lamellar macular hole formation, or may possess a yellow foveal lesion that may be mistaken for adult vitelliform dystrophy or Best’s disease. In the late stage, patients who exhibit macular stellate pigment plaques with SRNV may be misdiagnosed as having age related macular degeneration (ARMD) or focal chorioiditis. In differentiating patients with IJRT associated with SRNV from those with exudative ARMD, IJRT is rarely associated with pigment epithelial detachment and large neovascular complex formation. Group 3 patients who demonstrate atrophy of the juxtafoveolar retina with capillary occlusion and minimal exudation are most similar to those with sickle cell retinopathy[13].

Management

Macular edema and exudation are the main cause of visual loss in group 1 IJRT; the amount of exudation, edema, and subsequent visual acuity loss is variable[3,4]. Treatment options include laser, intravitreal steroids, or anti-vascular endothelial growth factor (VEGF) agents[15,16]. Laser may not always be possible due to the close proximity of the abnormal vessels to the fovea. Also, due to lack of significant improvement in visual outcomes and increased risk of the development of SRNV following treatment, laser photocoagulation for macular edema associated with IJRT is currently not recommended[14]. Intravitreal triamcinolone

Vol. 21, No. 1, July-September, 2010
**Table 1: Classification of IJRT**

<table>
<thead>
<tr>
<th>Group</th>
<th>Mean age of onset of symptoms</th>
<th>Predominance</th>
<th>Typical area of involvement, clinical picture</th>
<th>Visual loss</th>
<th>Systemic association</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>40 years</td>
<td>Male, Unilateral</td>
<td>Telangiectasia and aneurysms temporal to fovea, 2 Disc Diameters</td>
<td>Amount of exudation and visual loss variable</td>
<td>None</td>
</tr>
<tr>
<td>1B</td>
<td>Middle age</td>
<td>Male, Unilateral</td>
<td>Focal, limited to two clock hour</td>
<td>Exudation and edema may or may not occur. 20/25 or better</td>
<td>None</td>
</tr>
<tr>
<td>2A</td>
<td>Middle age and older</td>
<td>Male=Female, Bilateral but asymmetric</td>
<td>Retinal thickening temporal to the fovea, right-angle venules, RPE hyperplastic plaques, superficial crystalline deposits, SRNV</td>
<td>Progressive</td>
<td>Possibly diabetes</td>
</tr>
<tr>
<td>2B</td>
<td>Juvenile</td>
<td>Bilateral</td>
<td>SRNV</td>
<td>Visual loss due to SRNV</td>
<td>None</td>
</tr>
<tr>
<td>3A</td>
<td>Middle age or older</td>
<td>Female, Bilateral</td>
<td>Minimal exudation, capillary obstruction and occlusion</td>
<td>Visual loss due to capillary obstruction</td>
<td>Polycythemia, hypoglycemia, ulcerative colitis, multiple myeloma, chronic lymphatic leukemia</td>
</tr>
<tr>
<td>3B</td>
<td>Middle age or older</td>
<td>Male=Female</td>
<td>Minimal exudation, capillary obstruction and occlusion</td>
<td>and occlusion Visual loss due to capillary obstruction and occlusion</td>
<td>With CNS vasculopathy</td>
</tr>
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**Table 2: Stages of Group 2A IJRT**

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<tr>
<th>Stage</th>
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<tbody>
<tr>
<td>Stage 1</td>
<td>Asymptomatic</td>
<td>Difficult to detect clinically</td>
<td>Abnormal capillaries seen with fluorescein angiography (occult staining)</td>
</tr>
<tr>
<td>Stage 2</td>
<td>Asymptomatic</td>
<td>Mildly dilated perifoveolar capillaries</td>
<td>Slight graying of the retina, mild loss of transparency</td>
</tr>
<tr>
<td>Stage 3</td>
<td>Progressive decreased acuity</td>
<td>Dilated right-angled venules</td>
<td></td>
</tr>
<tr>
<td>Stage 4</td>
<td>Progressive decreased acuity</td>
<td>RPE hyperplasia clumped around the right-angled venules</td>
<td>Pseudovitelliform lesion</td>
</tr>
<tr>
<td>Stage 5</td>
<td>Rapid and severe vision loss</td>
<td>Intraretinal and subretinal neovascularization, exudation and hemorrhage</td>
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acetonide (IVTA) is beneficial in the treatment of macular edema by its anti-inflammatory effect, downregulation of VEGF production, and stabilization of the blood retinal barrier[15]. Intravitreal injections of anti-VEGF agents, namely Bevacizumab, have shown improved visual outcome and significant and sustained decrease in leakage on FA and macular edema on OCT. It is likely that patients with group 1 IJRT with pronounced macular edema from leaky telangiectasis may benefit functionally and morphologically from anti-VEGF injections supposedly even at a lower treatment frequency than in other diseases[16].

When considering treatment for group 2 IJRT, therapeutic attempts for nonproliferative IJRT and those for the SRNV of the proliferative stage must be distinguished. The angiographic late intraretinal staining pattern in nonproliferative IJRT 2A has prompted many ophthalmologists to interpret it as macular edema secondary to retinal vascular leakage. Several treatment modalities have been tried to treat this “macular edema.” To start, laser photocoagulation is not effective in the treatment of nonproliferative IJRT 2A. In addition, treatment might be associated with RPE changes, post-treatment retinal hemorrhages, and increased retinal vascular distortion[17].

Given that OCT shows that the fluorescein leakage seen is not associated with retinal thickening, the angiographic “leakage” is probably due to the staining of the extracellular matrix rather than extracellular leakage, and visual acuity correlates with photoreceptor layer disruption and not the degree of “leakage.” IVTA is likely to have a minor or no therapeutic effect in nonproliferative IJRT 2A[18]. Recent publications on intravitreal anti-VEGF injections, namely Bevacizumab, report on possible short term benefits in some cases of IJRT 2A[19,20]. Inhibition of VEGF may be useful before atrophic changes occur. VEGF plays a pathophysiological role in IJRT 2A, because the structural capillary changes described histopathologically lead to a disturbed exchange of oxygen and substrates between the vascular lumen and neurosensory retina, which in turn may lead to a hypoxia induced increased VEGF release by retinal cells[19]. However, despite increased leakage on FA, underlining the effect of Bevacizumab on vessel stability and permeability, small cystic changes seen on OCT and visual acuity may remain unchanged, emphasizing that visual deterioration is caused by microcystic degeneration and progressive retinal atrophy and not by intraretinal edema, and therefore cannot be halted with intravitreal anti-VEGF injections. Moreover, VEGF plays a role in photoreceptor differentiation and survival, and in maintaining retinal vascular homeostasis. Therefore, blocking VEGF may accelerate apoptosis among ganglion cells and photoreceptors in IJRT 2A[19]. Given the current lack of convincing evidence of efficacy, the concern about the potential deleterious effects of repeated injections and cost of treatment, treatment of nonproliferative IJRT 2A with VEGF antagonists appears questionable.

Although rare, development of SRNV generally results in poor visual acuity if left untreated, with 80% (21 of 26) of eyes in 1 study having a final acuity of 20/200 or worse[14]. Histopathologic studies show that while neovascular membranes in ARMD originate from the choroidal vasculature, vessels in proliferative IJRT originate from the retinal vasculature and contain more vessels and less fibrous tissue. Before the advent of VEGF antagonists, therapeutic options for SRNV in IJRT 2A included laser photocoagulation, photodynamic therapy (PDT) with or without IVTA, transscleral thermotherapy (TTT), and surgical removal of the CNV[18]. VEGF has been implicated as the major angiogenic stimulus responsible for neovascularization in IJRT 2A. Given the risk of permanent RPE damage with PDT, coupled with the huge evidence of efficacy of VEGF antagonists in the treatment of SRNV in various entities, the anti-VEGF approach is a reasonable treatment alternative for proliferative IJRT 2A. Anatomical peculiarities related to neovascular lesions in the setting of IJRT, such as the location above the RPE and the presence of anastomotic retinal vascular connections may facilitate inflow and concentration of the drug in the neovascular complex. Both intravitreal Bevacizumab (1.25 mg)[20,21] and Ranibizumab (0.5 mg)[22] have been used successfully in proliferative group 2A IJRT. Recently, primary treatment with combined intravitreal Bevacizumab or Ranibizumab and PDT have been reported anecdotally for proliferative IJRT 2A[23,24]. In both cases, the PDT was performed with a laser spot of the same size as the SRNV and followed by the intravitreal injection. Thus anti-VEGF therapy combined with or without PDT appears efficacious and should be considered as a treatment option for proliferative IJRT 2A.

Conclusion

IJRT comprises essentially three groups that differ in their appearance, presumed pathogenesis and management. In group 1, the unilateral telangiectasis is easily visible and vision loss is a result of exudation in the macula. Intravitreal steroids or Bevacizumab is generally effective in controlling the macular edema. In group 2, the most common, the bilateral capillary telangiectasis is more difficult to detect biomicroscopically, but the angiographic and OCT findings are characteristic and diagnostic. Vision loss is progressive and primarily due to retinal atrophy, not exudation or development of SRNV. Treatment options for this group are still limited, and have shown effectiveness only for the neovascular component. This is primarily because the pathogenesis of this telangiectasis remains an enigma and is possibly secondary to a retinal neuronal dysfunction. New imaging modalities and functional
Figure 1 – (a) Colour fundus photograph of a patient with IJRT 2A showing a greyish ring around the foveal centre with numerous superficial retinal crystals and a blunted, right angled draining venule (arrow). (b) Corresponding fluorescein angiogram shows in the early phase clearly visible dilatation and telangiectasis of the perifoveal capillary network with confirmation of the right angled venule (arrow). These capillaries show late intraretinal staining (c). Horizontal (d) and vertical (e) OCT scans show foveal detachment (asterisk), subfoveal cysts (arrowhead) with partial loss of the highly reflective line considered as the boundary between photoreceptor inner segments and outer segments (arrows).

Figure 2 - (a) Colour fundus photograph of a patient with IJRT 2A showing a greyish ring around the foveal centre with early intraretinal pigment deposition in the vicinity of a right angled venule (black arrow). (b) Corresponding fluorescein angiogram shows late intraretinal staining. Vertical (c) OCT scan shows blunting of the foveal pit (arrowhead) with foveal thinning. Horizontal OCT scan shows the characteristic ILM drape (dashed arrow) with partial loss of the highly reflective line considered as the boundary between photoreceptor inner segments and outer segments (arrows).
Figure 3 - (a, e) Colour fundus photograph of a patient with IJRT 2A showing stellate intraretinal pigment epithelial plaques, right angled draining venules and refractile retinal crystals (Stage 4). The disease is asymmetric, being more advanced in the left eye. (b, f) Corresponding fluorescein angiograms show blocked fluorescence due to the RPE hyperplasia with some intraretinal staining. Vertical (c) and horizontal (d) OCT scans of the right eye show blunting of the foveal pit, foveal thinning and a hyperreflective intraretinal lesion corresponding to the pigment (arrow). In the left eye, the flat pigmentary proliferation on the foveal surface masks the underlying retinal structure on OCT (g, h).

Figure 4 - (a) Colour fundus photograph of the right eye of the patient in Figure 2 showing temporal parafoveal retinal elevation with few crystalline retinal deposits, a right angled venule (arrow), nasal RPE alterations and subretinal blood characteristic of SRNV (IJRT 2A, Stage 5). (b) Corresponding fluorescein angiogram shows early hyperfluorescence with intense late leakage (c). Horizontal (d) and vertical (e) OCT scans show elevation of the juxtafoveal retina secondary to the presence of a hyperreflective fusiform complex lying at the outer retina/retinal pigment epithelium (RPE) level (extent marked by arrow) and minimal intraretinal fluid.
tests will hopefully improve the understanding and treatment capabilities of this condition. Group 3 is a perifoveolar capillary occlusive condition and is poorly understood because of the scarcity of cases reported.

References

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<td>— Transmits healthy blue light¹</td>
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Contact Lens Fitting in Keratoconus

Pooja Singh, Raghav Gupta, Jeewan S Titiyal, Rajesh Sinha

Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi

Keratoconus is a progressive, non-inflammatory, asymmetric ectatic corneal disorder that results in high amount of myopia and astigmatic refractive error. Early cases may be managed by spectacles; however, the definitive management is use of contact lens. In the mild stage, soft toric contact lens may be used. In the moderate and advanced cases, rigid gas permeable contact lens is the mainstay of treatment of these cases. Rose-k contact lenses have now become the contact lens of choice in these cases; however, the SOPER design is also very useful for management in these cases. Some cases may require piggyback and scleral lenses for better stability of the lens, and improved visual acuity and comfort to the patient.

Clinical Manifestations of Keratoconus

Symptoms of keratoconus

People with early keratoconus typically notice a minor blurring of their vision and come to their clinician seeking corrective lenses for reading or driving. At early stages, the symptoms of keratoconus may be no different from those of any other refractive defect of the eye. As the disease progresses, vision deteriorates, sometimes rapidly and is usually not correctable with spectacles. Visual acuity becomes impaired at all distances, and night vision is often quite poor. Some individuals have vision in one eye that is markedly worse than that in the other eye. Some develop photophobia, eye strain from squinting in order to read, or itching in the eye, but there is normally little or no sensation of pain. The classic symptom of keratoconus is the perception of multiple ‘ghost’ images, known as monocular polyopia. This effect is most clearly seen with a high contrast field, such as a point of light on a dark background. Instead of seeing just one point, a person with keratoconus sees many images of the point, spread out in a chaotic pattern. This pattern does not typically change from day to day, but over time it often takes on new forms. Patients also commonly notice streaking and flaring distortion around light sources.

Signs of keratoconus

External signs

Munson’s sign
Rizzuti phenomenon

Slit Lamp Findings:

Stromal thinning
Posterior stress lines (Vogt’s striae) (Figure 1)

Iron ring (Fleischer ring)
Scarring: Epithelial or Subepithelial

Retroillumination Signs:

Scissoring on retinoscopy

Figure 1 Vogt’s striae

Figure 2 Oil droplet sign

Oil droplet sign (“Charleaux”) (Figure 2)

Photokeratoscopy signs:
Compression of mires infer temporally (egg-shaped mires)
Compression of mires inferiorly or centrally

Videokeratography signs:
Localised increased surface power
Inferior superior dioptric asymmetry
Relative skewing of the steepest radial axes above and below the Horizontal meridian
Classification of keratoconus:
On the basis of Radius of Curvature of anterior segment of cornea:
1. **Mild/early** – 8.00-7.00(mm)/42D-47D
2. **Moderate** – 6.90-6.50(mm)/47D-52D
3. **Moderate/advanced** – 6.40-6.00 (mm)/53D-56D
4. **Advanced/severe** – <6.00(mm)/>57D

Keratoconus cones can be classified as:
1. **Nipple**: Small diameter (5mm); cone lies in the lower nasal quadrant
2. **Oval**: Larger (> 5mm); lies more commonly in the inferno- temporal quadrant
3. **Globus**: Largest diameter (>6mm); 75% of the cornea is affected

**Related conditions:**
Keratoconus has been found related with atopic conditions asthma and eye rubbing.
It has also being linked to Leber’s congenital amaurosis, Down’s syndrome and mitral valve prolapse cases.

**Posterior keratoconus:**
It is the rare form of condition reported in some cases. Uniform corneal steepening was observed in generalized posterior keratoconus and corneal steepening was observed in localized central and posterior keratoconus and corneal flattening was observed in localized peripheral posterior keratoconus.

**Investigations**
**Slit lamp examination:**
**In early kc:**
- thinning of epithelium due to cone progressing
- extra cellular and intercellular accumulation in epithelium at the periphery of the cone

**In later stages:**
- ruptures or folds in descememt’s membrane
- eventually some form of corneal scarring is seen
- small stromal scars are due to idiopathic breaks in bowman membrane
- hydrops: marked stromal oedema occurs as a result of descememt membrane rupturing allowing aqueous humor to enter the stroma. (Figure 3)

**If scarring in visual axis area it can affect the vision.**

**Corneal Topography**
**Videokeratography(3,4) Orbscan Pentacam**
1. In basic orbscan height map, a mean radius of curvature of corneal map is calculated and relative height above or below this ideal best fitting spherical surface is known.
2. Warm colors show areas higher than best fitting sphere and cooler color which are lower than best fitting sphere

**Pachymetry**
It provides additional information about corneal thickness in diagnosing keratoconus. Ultrasound pachymetry is best accepted.

**Refraction**
Retinoscopy is difficult /scissoring reflex. Subjective refraction is needed. In early keratoconus and mild cases can be corrected with glasses. Monocular mild keratoconus is best dealt with glasses.

**Dry Eye Assessment**
Dry Eye is ruled out by asking for relevant symptoms and by doing schirmer’s test and tear breakup time.

**Contact Lens Options For Keratoconus**
In cases of high irregular astigmatism and moderate/advanced keratoconus cases rigid contact lenses will be required effectively to provide new anterior surface to the eye giving good, clear vision. They are considered when good vision is not attained by glasses less than 6/9 and shadowing and diplopia persists with glasses and patient becomes symptomatic

**Rigid Gas Permeable Lenses**
- First choice of correcting irregular astigmatism
- A mild to high dk/t material is preferred providing stability required for high powered lenses.
- A balance is required between material which is deposit resistance especially in patients who are atopic and providing sufficient oxygen flux. (Figure 4)

**Fitting Philosopies**
**a. Apical bearing:**
Lens bears heavily on corneal apex. It provides good vision but may result in corneal abrasions scarring. (Figure 5)
b. Apical clearance:
The back surface of the lens vaults apex of the cone. There is lesser risk of corneal scarring. It causes variable vision due to uncorrected astigmatism. (Figure 6)

c. Three Point Touch:
Lens rests lightly against the cone apex. Also supported on nasal and temporal zones by mid periphery of the back surface of the lens. It provides stable fitting and good vision. (Figure 7)

Type of RGP Lens Design:
**Early keratoconus**
- Aspheric or multicurve lenses. This is the Preferred lens fitting technique.
- Kera I and II (No.7)Acuity K
- Rose K (David Thomas)

**Moderate keratoconus**
- Rose K 2(David Thomas)
- Woodward KC3/SOPER LENSES
- Kera II
- Quasar KNO7

**Moderate/Advanced keratoconus**
- Rose K 2/IC(David Thomas)
- Kera II/III
- Profile K (J Allen)

**Advanced keratoconus**
- Small diameter lenses
- S-Lim (J Allen)
- Dyna-intra limbal (No.7)
- Scleral lenses/JUPITOR lenses
- Gas permeable (innovative sclerals)

**Preliminary examination**
Many patients with keratoconus will be in their late teens or early twenties. They will need information and reassurance. They may present with concerns about the speed with which their vision has deteriorated.
- It is important to explain the reason why the spectacle prescription has been changing rapidly over the past 12-24 months.
- The nature of the corneal thinning disorder and the reasons for corneal distortion should be explained. The advantages of contact lenses over spectacles should be emphasized.
- The progression of the condition and the prognosis should be discussed.
- An information leaflet explaining the condition, and information about the local keratoconus support group, should be provided

**Fitting Considerations**
- cone position shape and size
- corneal radius (central and steepest)
- corneal toricity
- degree of myopia and corneal astigmatism

**Fitting Algorithm**
1) After taking full history and symptoms, the preliminary examination should include age, occupation, and motivation. Any history of previous contact lens intolerance or allergies should be noted.
2) Full slit lamp biomicroscopy is vital.
3) Examine the keratometer readings. The mires may be much distorted; however they provide useful information at the initial stages.
4) Choose the correct base curve using corneal topography; start with the base curve equivalent to the steeper of the two keratometer readings. Many variations on this philosophy exist as already discussed.
5) Allow the lens to settle for about 20minutes before evaluating the fluoresce in pattern.
6) Examine the central area, the mid peripheral area and the periphery.
7) Evaluate the lens in the central position. Once you have judged the fit, alter the fit as necessary (for example flattens, if pooling) until you obtain gentle apical touch and the three-point-touch. Use the Guillon grading scale
for assessing the fluoresce in picture. There should be minimal bearing (touch) at the apex of the cone, as well as an area of bearing between the periphery of the lens and the intermediate zone of the cornea.

8) The lens should be ordered in mid-high Dk material after an over-refraction has been undertaken. A collection appointment should be arranged. An aftercare appointment should follow four weeks after the collection appointment, when slight modifications may be necessary.

**Soper Contact Lens**

The Soper lens system uses bicurve lenses based on sagittal depth. (Figure 8) A steeper lens is selected by maintaining the same base curve but increasing the diameter. Three fitting sets are available in these contact lenses. Mild (7.4mm diameter, 6.0mm OZ), moderate (8.5mm diameter, 7.0mm OZ) and advanced (9.5mm diameter, 8.0mm OZ). The smaller lenses are used for smaller, centrally located cones. The large diameters are used for oval cones. The peripheral curve of this system is constant. Peripheral alignment and apical clearance typify the soper contact lens design[5].

**Rose-K Contact Lenses**

Unique keratoconus lens design with computer generated peripheral curves based on data collected by DR. Paul Rose of Hamilton, New Zealand. The system incorporates triple peripheral curve system standard flat and steep in order to achieve ideal edge lift of 0.8mm. Available in base curves: 4.75-8.00mm, Diameter: 7.92-10.00mm. Toric surfaces are available on front and back and periphery too. (Table 1)

<table>
<thead>
<tr>
<th>Primary indication</th>
<th>Rose K2</th>
<th>Rose K2 IC</th>
<th>Rose K2 Post graft</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nipple/Oval Keratoconus</td>
<td>Pellucid Marginal Degeneration, Keratoglobus(PMD), Lasik Induced Ectasia</td>
<td>For patients who have undergone penetrating keratoplasty</td>
<td></td>
</tr>
<tr>
<td>Early PMD</td>
<td>Oval/Nipple Keratoconus</td>
<td>Oval/Nipple Keratoconus</td>
<td></td>
</tr>
<tr>
<td>BC:4.3- 8.6mm OD: 7.9-10.4mm</td>
<td>BC: 5.7-9.3 mm OD: 9.4 – 12.0 mm</td>
<td>BC: 5.7- 9.3mm OD: 9.4- 12.0mm</td>
<td></td>
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</table>

*Table 1 Type of Rose- K Contact Lens*

Edge lift can be varied with standard steep/flat optimum lift. It is a multi-spherical posterior design with aberration control aspheric optics across the back and front optic zone diameters. The design provides an easy-to-fit systematic approach with a flexible edge lift for the practitioner enabling them to reduce their chair time and dispense the optimum fitting lens to the patient. The system (26-lens set) allows the practitioner to choose lens options based on a systematic fitting approach. The design starts with a standard 8.7 mm. diameter that incorporates a decreasing optic zone as the base curve steepens coupled with an intrinsic, computer designed peripheral curve system. The lens is provided through Paragon Optics and is manufactured on a DAC lathe. The standard lift lenses should work approximately 70% of the time. Additional lens diameters are available when needed (8.3, 9.0). Peripheral curves and even base curves can be configured in a toric design.[6-8]. (Figure 9)

**Soft Lens/hydrogels/silicon Hydrogels**

**Why Soft Contact Lenses?**

Clinical experience has demonstrated that small subset keratoconus patients may experience a corneal hypersensitivity in certain stages of their condition. This hypersensitivity may be due, in part, to a dilatation or stretching of the corneal nerve fibers; in addition, minute ruptures in Bowman’s layer may be a contributing factor. This subset of patients may find it difficult, or even impossible, to tolerate well-fitted GP lenses.

**Soft Designs for Keratoconus**

A viable option is to use one of the many custom toric soft contact lenses we currently have at our disposal. These have proven successful in the early stages of the condition or in form...
fruste keratoconus.2 However, custom soft spherical lenses may also prove greatly successful. These lenses incorporate a tricurve posterior lens design with increased central thickness to mask much of the regular and irregular astigmatism. You can custom design these lenses using a broad range of base curve radii, powers, diameters and center thickness values.

Advantages
1) They afford higher levels of comfort and longer wearing times, especially in patients intolerant of RGP corneal lenses or in monocular keratoconus.
2) They are useful where the cone apex may be displaced, especially if it is very low.
3) They are useful for certain groups of patients, for example airline pilots.
4) They are relatively simple to fit.

Disadvantages
1) Visual acuity may be variable in cases of very high minus lenses.
2) Low-powered diagnostic lenses may not provide an accurate guide to the fit of the final lens, which may be extremely high powered.
3) There may be reduced oxygen transmissibility and the risk of neovascularisation if the lenses are over worn.
4) If the condition has progressed, it may be difficult to change to RGP’s at a later stage.

Piggyback lenses
With the advent of silicone hydrogel oxygen concern with piggybacking have greatly lessened. Weiss man and ye (2006) have shown that the oxygen transmission of a system using both high-Dk GP lenses and silicone hydrogel lenses is only slightly less than that of silicone hydrogels alone. It’s an improvement over the GP lenses alone of a decade ago. The system using both high-Dk GP lenses and silicone hydrogel lenses also offers significantly more oxygen transmission than custom hydrogel toric lenses or available hybrid lenses other options you might consider to provide a similar comfort and fitting benefit. While slightly inconvenient with two lenses and care systems per eye, patients have responded very positively to piggyback contact lenses due to the comfort and wearing time benefits.

The system consists of a rigid lens fitted on top of a soft lens. The aim is to maintain the same level of visual acuity as with a single lens.

The RGP lens should be fitted first. Good centration is important and a slightly larger area of apical touch is usually acceptable as the RGP lens will be cushioned by a soft lens. A silicone hydro gel soft lens should be used where possible, with good movement and coverage/ cent ration as in a normal soft lens fitting. Caring for the two lenses can be difficult long-term. Ideally, try to have the patient use the same care regime for the two lenses as this will make cleaning easier, or alternatively consider a disposable soft lens. The cornea should be observed carefully for dryness and neovascularisation. Piggyback contact lenses can play a beneficial role for patients who experience comfort or wetting problems, yet need the optical properties of a GP lens. At the least, the ability to use high-oxygen transmissible lens materials makes piggyback lens fitting a technique certainly worthy of a second look[9].

Hybrid lens system
Hybrids refer to contact lenses that have a rigid GP central portion surrounded by a soft lens material skirt. The earliest lenses in this category were introduced as Saturn contact lenses by Preci-sion-Cosmet in the early to mid-1980s. This initial lens concept went through several generations of development and several changes of ownership before arriving at what is offered today as the Soft Perm lens from CIBA Vision. Among the changes in the Soft Perm lens from the original Saturn design was the addition of peripheral curves onto the GP portion and changes to the peripheral curves of the soft skirt. Activity in the area of hybrid contact lenses increased with the formation of SynergEyes, Inc. The company introduced the SynergEyes A hybrid lens, which became the first in a family of lenses that include SynergEyes KC (for keratoconus), SynergEyes Multifocal (an annular, center-near design), and SynergEyes PS (for post-surgical fittings). One advancement that these entire lenses offer is the option of choosing among different peripheral curves (either two or three options depending on the design) for the soft lens skirt portion. This adds another fitting parameter apart from just the base curve of the GP central portion to allow better refinement of the fit. Of recent note are a number of new developments from SynergEyes. These include:

1. Increased center thickness of the GP lens portion in the company’s Enhanced Profile (EP) version, available in each of the different SynergEyes lens designs. An EP lens provides a center thickness that is 0.09mm thicker than that of the standard lens for each design. This is helpful in reducing some flexure in highly astigmatic corneas. Due to the effect of the soft lens skirt, the increased GP thickness does not significantly alter the fit, centration or comfort of the EP lenses.

2. Introduction of the Clear-Kone design for keratoconus, which is designed to better vault advanced keratoconic corneas without bearing. This new option may allow better success in some of the most challenging cases.

3 FDA approvals to begin clinical trials of hybrid contact lenses utilizing a soft silicone hydrogel material for the skirt portion of the lens. SynergEyes hopes to launch products
with this new skirt material in the first half of 2010. Limiting limbal encroachment or neovascularization is very important in post-surgical cases or in eyes that may eventually need corneal surgery.

The main disadvantages of hybrid lenses are frequent breakage of the lens, giant papillary conjunctivitis and peripheral corneal neovascularisation. It should be noted that the Soft perm lens was not designed for keratoconus, but for a normal cornea. As it provides the comfort of a soft lens and visual acuity of a rigid lens it has been adopted by keratoconic patients who inevitably over-wear these lenses and end up with complications.

Scleral lenses
Scleral lenses play a very significant role in cases of advanced keratoconus where corneal lenses do not work and corneal surgery is contra-indicated. Scleral lenses completely neutralize any corneal irregularity and can help patients maintain a normal quality of life. A PMMA lens can be used in cases of Scleral toxicity.

PMMA scleral lenses are made by the impression method. This practice is confined to the HES. An impression is taken of the cornea, generally with alginate material (ortho print) and a clear shell is made from poly-methyl methacrylate material. Optic curves are ground on to the shell. This can be done in-house or the shell can be sent to Cantor & Nissel. The shell is fenestrated, adjusted, and ground until a desirable fit is obtained. Once an acceptable fit is obtained the lens can be sent for working to the required power. (Figure 10)

![Figure 10 Scleral Contact Lens](image)

**Advantages**
- Easy to use and remove
- Any type of irregularity is removed
- Easy to store and dry
- Long life

**Disadvantages**
- Much chair time is needed
- A very specialized fitting technique

**References:**
Fuch’s endothelial corneal dystrophy is a progressive disorder of the endothelium wherein a gradual decline of functional endothelial cells over the years, manifesting with visual disturbance in late adulthood. The visual loss in these patients can be either due to cataract or due to corneal dystrophy itself. Determining which patient requires cataract surgery is imperative for an ophthalmologist to visually rehabilitate the patient. However cataract surgery could potentially decompense an already diseased cornea with borderline endothelial cell reserve. This review will help the surgeons in proper evaluation and practice during surgery to provide the patient with maximal visual gain.

**Fuch’s dystrophy and cataract surgery**

Fuch’s corneal dystrophy (FCD) is bilateral progressive endothelial dystrophy usually presenting with visual disturbance at an age of 50-60 years. Abnormal production of collagenous material by the affected endothelial cells causes marked thickening of Descemet’s membrane, which becomes studded with excrescences (guttae). Eventually there is a gradual decline in the number of functional ATP dependent Na K pumps resulting in corneal edema[1]. FCD is associated with a low endothelial cell count as well as abnormal cell morphology (pleomorphism and polymegathism) (Figure 1), where in an unplanned cataract surgery has the potential to cause early decomposition of cornea incurred due to mechanical, hydrodynamic, thermal, chemical, or possibly free radical damage secondary to high-frequency ultrasound used during phacoemulsification[3]. Patients with decreased vision due to FCD and cataract can present with a number of challenges to determine the best surgical option for restoring sight because intraocular surgery may accelerate corneal endothelial cell loss. Finding an unresolving corneal edema in an eye with an uneventful phacoemulsification surgery is every surgeon’s nightmare. Therefore proper preoperative assessment and thorough planning is a prerequisite before actual surgery in these patients and one must evaluate, whether cataract surgery alone or cataract surgery combined with full thickness or lamellar keratoplasty (triple procedure) is required for an individual.

**Symptomatology**

**Glare**

The debilitating symptom of glare due to confluent guttae with pigment can occur even when there is little to no stromal or epithelial edema and pachymetry is relatively normal (stage 1) or may also occur secondary to stromal edema (stage 2)[4].

**Visual loss**

This can result from the cataract, the dystrophy, or both. Vision which is worse in the morning is likely attributable to the FCD as corneal edema increases at night while sleeping. Several hours may elapse into the morning before vision improves and documenting what time the vision clears in a particular patient over time can become a measure of how the corneal disease is progressing (stages 2 and 3). In the fourth stage, vision reduces to hand motions due to growth of avascular subepithelial connective tissue and peripheral corneal vascularisation[1].

**Pain**

It is usually seen during the third stage which is characterised by epithelial edema. As epithelial cysts coalesce to form large bullae, their rupture results in severe pain and predisposition to ocular infection[1].

**Preoperative considerations:**

These include

a) Patients with increasing age, history of previous ocular surgery, history of ocular trauma, ocular infection, angle closure glaucoma, and pseudoexfoliation syndrome, diabetes of more than 10 year duration and long-term contact lens use possess a poor endothelial cell density reserve or abnormal cell morphology[5].

b) If the patient has glaucoma and is on topical carbonic anhydrase inhibitors, it is reasonable to stop their use as they interfere with fluid pumping ability of the endothelium by blocking the active sodium potassium pump thus worsening the edema in such patients[5].

c) Consider for endothelial keratoplasty with phacoemulsification if preoperative corneal edema or central corneal thickness (CCT) is more than 650µ. These are the cases which are most likely to decompensate after an uneventful cataract surgery[3].

d) Werblin had found 9% endothelial cell loss at 1 year after phacoemulsification with posterior chamber lens insertion, with 11.5% loss at 3 years[2]. However, in the presence of a hard nucleus (LOCS grade 4 and above) or a low endothelial cell count (ECC) of less than 1500/
mm2, conventional cataract surgery like extracapsular cataract extraction (ECCE) or small incision cataract surgery (SICS) is preferable [6]. No significant difference in postoperative ECC exists when comparing ECCE with intracapsular cataract extraction (ICCE) [2].

**Operative considerations**

**Anesthesia**

It is preferable to give peribulbar block. Avoid intracameral anesthesia as these drugs can cause endothelial toxicity. If required, preservative free 1% lignocaine can be used instead of 5-10% lignocaine, 0.5% bupivacaine or 0.5% proparacaine all of which are endothelio-toxic[2].

**Drugs**

Povidone iodine 5-10% when used for sterilisation should be thoroughly washed after use as it leads to significant corneal toxicity if it accidentally penetrates the anterior chamber. Acetylcysteine 1% is toxic; instead use of 0.001% carbachol is recommended if intraoperative miosis is desired. Vancomycin in a concentration of >1 mg/ml is not considered safe for the endothelium[2].

**Sterilization**

Avoid use of instruments dipped in 2 % glutaraldehyde. Aqueous substitutes: Among the aqueous substitutes used intraoperatively, 2.3% or 1.4% sodium hyaluronate and a combination of sodium hyaluronate and chondroitin sulfate should be preferred. Avoid air injection and use of trypan blue for capsular staining. Amongst the irrigating fluids, balanced salt solution (BSS) Plus (containing reduced glutathione, bicarbonate and dextrose) is better than BSS alone followed by Ringer Lactate and least protection being offered by balanced salt solution (BSS) Plus (containing reduced glutathione, bicarbonate and dextrose) is better than BSS alone followed by Ringer Lactate and least protection being offered by balanced salt solution (BSS) Plus (containing reduced glutathione, bicarbonate and dextrose). Role of preservatives: Epinephrine containing sodium bisulphate as a preservative leads to increased cell loss, hence use of preservative free epinephrine is recommended. Hydroxypropylmethylcellulose (HPMC) containing benzalkonium chloride is known to cause striate keratopathy[2].

**Incisions**

Limbal or clear corneal incision leads to an increased endothelial cell loss. Amount of cell loss is directly proportional to the length of incision. Therefore ECCE or ICCE incision leads to a greater amount of cell loss as compared to phacoemulsification wound. Superior incision has been found to be better than temporal incisions in terms of cell loss. Scleral tunnel incisions in comparison to clear corneal incisions cause less endothelial damage[2]. Sleeve should be appropriate size and should not fit too tight in the wound such that fluid should pass out easily, thus minimising the thermal damage to the cornea[5].

**Viscoelastic considerations**

‘Arshinoff soft shell’ technique should be considered whenever possible wherein a viscodispersive agent preferably HPMC or chondroitin sulfate is injected intracameral followed by the use of a viscocohesive agent like 1.4% sodium hyaluronate which thereby pushes the viscodispersive against the endothelium providing protection during the surgery. Sodium hyaluronate 2.3% used alone, being a viscoadaptive agent serves the purpose of both the agents, and hence can also be used[5].

**Phaco-technique**

During phacoemulsification, phaco parameters in cases of compromised endothelium should include low phaco power along with high vacuum, or restricted use of energy by using fractions of second pulses or bursts, and millisecond-level microburst. The OZil Torsional system (Infiniti, Alcon, Fort Worth, TX) is a hardware and software upgrade which includes a dedicated handpiece that produces side-to-side rotary oscillations of the phaco tip[10,11]. Comparing with the jackhammer motion in conventional longitudinal phaco, the improved OZil Torsional oscillation sheers the lens material with virtually no repulsion, thereby dramatically reduces phaco energy required for lens removal without compromising efficiency. While in Torsional phaco, although the tip moves at a lower frequency of 32 kHz than the 40 kHz in traditional phaco, the side-to-side tip movement sheers the lens material with no repellent force, and cuts with both direction of the tip movement, thus significantly improving emulsity efficiency. Other modifications include include non ultrasonic energy such as sonic frequencies, NeoSoniX-generated tip rotation, and pulse water-jet technology[10,11]. Use of specific chopping techniques with endocapsular emulsification of nucleus is recommended. Entering the anterior chamber repeatedly with phaco tip or second instruments damages the endothelium; therefore minimum manipulation is advisable. Entry and exit should always be made in a formed anterior chamber to avoid chamber fluctuations during the surgery; therefore, it is advisable to inject viscoelastic through side port before taking out the phaco probe. In a recent study by Liu et al, microincision cataract surgery was not superior to a standard phacoemulsification in terms of endothelial cell loss and produced statically insignificant greater cell loss but allowed excellent visual results in his series of patients[12].

**Intraocular lens (IOL)**

During IOL insertion, coat the anterior surface of the lens with a viscodispersive agent. Hydrogel / acrylic/ silicon IOLs have been found to be safer for endothelium than PMMA IOLs. Furthermore, posterior chamber (PC) IOLs lead to lesser cell loss when compared to anterior chamber (AC) IOLs. IOLs with surface passivisation e.g. Teflon coated or heparin surface...
modified are strongly recommended. Corneal endothelial cell loss over time has been found to be more in aphakes as compared to pseudophakes[2].

Postoperative considerations:
Certain postoperative factors like shallow anterior chamber, peripheral anterior synechiae, postoperative inflammation, raised intraocular pressure, vitreous corneal touch, pseudophacodonesis, IOL corneal touch all predispose to increased cell loss over time and thus should be carefully looked for in all such patients[5].

Cataract surgery with endothelial keratoplasty
DSAEK when done together with phacoemulsification and IOL implantation is known as triple procedure. In patients with FCD the view through the cornea may be acceptable to allow for a closed cataract extraction by phacoemulsification followed by DSAEK. When the view is obscured by significant stromal and epithelial edema, the DSAEK may be performed first, waiting for the edema and view to improve followed by cataract surgery. The use of viscoelastic material during cataract surgery has not been found to adversely affect graft adherence after DSAEK. Performing a smaller capsulorhexis, using a larger lens diameter, and constricting the pupil with miotics after intraocular lens implantation allows for the insertion of the graft without increasing endothelial loss. In a prospective study on triple procedure in 89 eyes by Terry et al[7], 32% cell loss was found in donor corneas at the end of 12 months representing a mean endothelial cell density of 1979 cells/mm[2].

Conclusion
Therefore patients presenting with FCD and cataract pose a special challenge to the ophthalmologist and should be dealt with a proper preoperative assessment and postoperative follow up to give the possible & desired visual outcomes without causing any inadvertent complications.

References
Morning Glory Syndrome

Rashida Shabbir Tankiwala, Memuna Bahadur
Dr Babasaheb Ambedkar Memorial Hospital, Central Railway, Mumbai

Morning glory syndrome is a congenital optic disc anomaly described by Kindler1 in 1970. It is a congenital funnel-shaped excavation of the posterior fundus that incorporates the optic disc, resembling the morning glory flower. Ocular complications may include strabismus, reduced visual acuity and retinal detachment and it may have systemic associations as in Aicardi’s syndrome. A patient with monocular morning glory syndrome and reduced visual acuity is reported. The pattern reversal visually evoked potential was reduced.

Introduction
We report this case of unilateral Morning Glory Syndrome with no associated systemic abnormalities which is rare in occurrence, although, the exact incidence of its occurrence is unknown.

Case Report
A 14 yr old girl was brought by her mother with complaints of headache. On inquiry, her birth history and family history were unremarkable. She was found on examination to have less vision in her right eye.

Detailed examination: Her facial features appeared normal. Ocular adnexa including bony orbital rim and soft tissue were normal. She was orthophoric for distance and near. Best corrected visual acuity with -2.75 DS was 6/18. Slit lamp examination of the anterior segment was within normal limits. Pupils were round and regular and the reflexes were normal to light and accommodation in both eyes with no relative afferent pupil defect. Intraocular pressure (IOP) checked digitally was normal in both eyes.

Fundus examination revealed the presence of a funnel-shaped excavation containing an enlarged, somewhat indistinct, optic disc surrounded by a wide annulus of chorioretinal pigmentary disturbance. A white tuft of glial tissue was seen over the central portion of the disc and blood vessels seen emanating radially from the disc margin [Figure 1].

Macula appeared normal, no evidence of retinal detachment, holes or degenerative changes seen in the peripheral retina. Left eye showed vision of 6/6, unaided with normal ocular examination.

Special Investigations
- Colour vision on Ishihara charts was normal.
- Ocular ultrasonography confirmed the presence of marked irregular deepening of the optic nerve head on the right side with the presence of a density over the disc. No evidence of retinal detachment and normal retrobulbar region was noted. Left eye revealed normal ultrasonic examination [Figure 2].

<table>
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<th>ISCEV STANDARD PATTERN - REVERSAL VEP</th>
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<tr>
<td><strong>P 100</strong></td>
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<tr>
<td>Implicit Time</td>
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<td>Amplitude</td>
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The P 100 difference between both the eyes is 3 ms (normal > 10 ms)
There is normal latency of conduction in both the eyes eye.
The amplitude of P 100 is normal on the left side.
Using the Humphrey’s Field Analyser, 30-2 threshold showed [Figure 3].

- Right eye: Enlargement of the blind spot
- Left eye: Fields to be within the normal limits
- There was no evidence of multi-organ involvement, which had been investigated.

**Discussion:**
Vision is usually poor in patients with morning glory syndrome[8]. This vision loss may be due to the presence of retinal abnormalities in the macula[12], or amblyopia secondary to anisometropia or strabismus[1], or due to the developmental defect of the optic nerve head. Usually unilateral[11], but a rare case of bilateral MGS have been reported[8].

Other ocular findings commonly observed in the affected eye with MGS include: strabismus[1], an afferent pupillary defect[10], visual field defects consisting of blind spot enlargement and/or dense central scotomata[13], mild to moderate myopia[1]. Ocular associations found in the affected eye with morning glory disc anomaly may include: non-rhegmatogenous retinal detachment[14], the presence of marked persistent hyperplastic primary vitreous[3], lens coloboma[3], ciliary body cyst, congenital cataract, lid haemangiomia, vitreous cyst and preretinal gliosis[12].

Morning glory disc may be mistaken for disc swelling in the affected eye. It needs to be carefully evaluated to prevent misdiagnosis[5].

Ocular abnormalities have also been reported in the fellow eye. These include: microphthalmos, anterior chamber cleavage syndrome, microcornea and Duane’s retraction syndrome[12]. Systemic associations in morning glory syndrome have been well-documented.

**References**
An Unusual Case of Metastatic Tubercular Endophthalmitis

Meenakshi Kabra, Sarita Beri, Rajiv Garg, Pamela d’Souza, Rajesh Jain, Anita Nangia, Sanjay Kumar Mishra
Lady Hardinge Medical College, Shahid Bhagat Singh Marg, New Delhi -110001

Tuberculosis is prevalent in developing countries like India, however ocular tuberculosis is a diagnosis of exclusion. This unusual case is of an 18 year old patient of pulmonary Kochs who presented to the out patient department of our hospital with unilateral acute pain in right eye with the marked diminution of vision of 3 days duration. On Examination his vision in Right eye was PL-negative. This was preceded by gradual painful loss of vision in the right eye for the past two months duration for which he was already taking medication. He was diagnosed as a case of endophthalmitis with secondary glaucoma and ciliary and intercalary staphyloma in R/E. His eye was eviscerated. On the basis of clinical presentation, radiological and histopathological findings diagnosis of Tuberculous endophthalmitis was therefore confirmed.

Case report
The prevalence of tuberculosis in India is 30% and the annual incidence of infection is 1-2%. India forms the highest TB burden country in the world, with 1.8 million cases occurring annually[1]. An 18 year old male patient presented to the out patient department with unilateral acute pain in right eye with the marked diminution of vision of 3 days duration. There was history of dull ocular pain with gradual diminution of vision in the right eye for the past two months. He was diagnosis as uveitis at another centre and was started on topical steroid antibiotic combination, atropine and timolol. He was on regular treatment for his eye condition, without any improvement, for the past two months.

He presented to us with severe ocular pain of 3 days duration. He gave a past history of pulmonary kochs for which he was already on regular four drug antitubercular treatment for the past four months. On systemic examination of the patient, chest examination revealed bilateral crepts. On ophthalmic examination his vision in the Right eye was PL-negative and in the left eye was 6/6. Right eye had ciliary congestion, with a superior ciliary and intercalary staphyloma was present, corneal haze, hyphema and exudates in the anterior chamber Digital tension was high. The details of iris and pupil were not clear. The left eye on examination did not show any other abnormality except absent consensual pupillary reaction. USG B scan of the right eye showed low to moderate amplitude spikes suggestive of vitreous exudates. The intra ocular pressure was 29 in the right eye and 11 in left eye. A provisional diagnosis of endophthalmitis with secondary glaucoma and ciliary and intercalary staphyloma of right eye was made and the treatment in the form of intravenous Omnatix and Amikacin, oral acetazolamide, analgesics was started. Topically he was put on gatifloxacin-dexamethasone combination, antiglaucoma medication, lubricating agent and hot fomentation in right eye. His systemic antitubercular treatment was continued.

Blood profile was Hb 12gm, TLC -5800/mm3, ESR was 83 in 1st hour by Westergens method while random blood sugar, liver and kidney function tests were with in normal limits. Chest x-ray showed, resolving right upper lobe and left upper and lower lobe parenchymal infiltration suggestive of pulmonary Kochs responding to antitubercular treatment. Evisceration of right eye was done and the Ocular tissue sent for histopathological examination. The histopathological report showed focal areas of caseous necrosis (Figure-1A) with multiple epitheloid cell granulomas and Langhans giant cells. Ziehl Neilsen stain showed acid fast bacilli (Figure-1B). Diagnosis of Tuberculous panuveitis leading on to endophthalmitis was therefore confirmed.

Discussion
Mycobacterium tuberculosis, the etiologic agent of tuberculosis can cause infection in many organs including the eye. Ocular tuberculosis can involve any part of the eye and can occur with or without evidence of a systemic focus of tuberculosis[2]. The patient probably developed tuberculous panuveitis leading to endophthalmitis. Only sporadic cases of metastatic tubercular endophthalmitis have been reported. Metastatic endophthalmitis is twice as common in the right eye as in the left, probably because of the shorter, more direct route of arterial blood flow from the internal carotid artery to the eye on the right side[2,3]. The visual prognosis following diffuse posterior metastatic bacterial endophthalmitis is very poor. Removal of the affected painful blind is the treatment of choice. There is no case in the literature where visual recovery is seen after this type of infection regardless of type of therapy[2].
There are two possible contributory factors responsible for endophthalmitis in this patient. Firstly, from the pulmonary infection, a septic embolus occludes the central retinal artery and embolic fragments are then disseminated peripherally, so ischemia as well as infection may add to the poor prognosis. [2]. Secondly, In patients with bacteremia the blood-borne organisms permeate the blood-ocular barrier either by direct invasion or by changes in vascular endothelium caused by substrates released during infection. Destruction of intraocular tissues may be due to direct invasion by the organism[2-4].

**Figure 1A** section shows epitheloid cell granuloma with Langhans giant cell. Part of pigment layer is seen in periphery of granuloma. Haematoxylin and Eosin stain (400x magnification)

**Figure 1B** Ziehl Neelson stain (1000x magnification) arrow showing Beaded long M.tuberculosis identified.

**Conclusion**

All patients of systemic tuberculosis presenting with ocular involvement, metastasis as an etiology should be considered as one of the differential diagnosis. Prompt diagnosis and appropriate management can prevent blindness.

**References**

Cases Reports

Atypical Presentation of Conjunctival Neoplasia

Vandana Jain, S. Gupta, R. Matai, R. K. Srivastava
ESI Hospital, Indore

This communication delineates variable presentations in two cases of conjunctival neoplasia, histopathology of the same was suggestive of squamous neoplasia. Though incidence of conjunctival neoplasia is low, any case presenting as non resolving chronic conjunctivitis or recurrent pterigium should undergo cytological examination to rule out malignancy.

Case 1
A 46 year old male presented with redness, photophobia, blurred vision and membranous growth in the right eye since 1 year. Subsequently he had progressive diminution of vision. He gave history of blunt trauma to affected eye preceding the redness (by a wire) for which he was treated conservatively elsewhere. On local examination of the right eye invasion of conjunctival tissue over cornea was present for 360 degree with papillary process extending up to optical zone. Grossly it had gelatinous appearance with superficial vascularisation. (Figure 1)

His best corrected visual acuity was 2/60. Fundus examination revealed faint red glow. Systemic examination was non contributory. Serology for HIV was negative. A provisional diagnosis of keratitis (D/d : Chronic conjunctivitis or Limbal stem cell deficiency) was made and he was initiated on topical antibiotic-steroid combination. As he did not respond, conjunctival scraping was done. Histopathology of the same revealed plenty of atypical cells and dysplasia consistent with moderate grade of squamous intra epithelial neoplasia. (Figure 2)

This patient was treated with superficial keratectomy with Amniotic membrane transplant & topical Cyclosporin A drops.

Case 2
A 60 year old female presented with painless conjunctival growth of right eye of 2 1/2 months duration. Local examination revealed a sessile firm growth which had no fluctuation, pulsation and was nontender. Dimensions of the growth were 6mm X 4 mm. It was located at temporal limbus with a 2 mm corneal invasion. (Figure 3)

Visual acuity was 6/12 with normal fundus examination bilaterally. Systemic examination was non contributory. A provisional diagnosis of conjunctival melanoma with differential diagnosis of sessile papilloma and squamous cell carcinoma were considered. Subsequently total excisional biopsy of the mass was done with 2mm clear margins of conjunctiva & cornea.

Histopathology was suggestive of well differentiated squamous cell carcinoma with stromal invasion. (Figure 4)
Discussion:
We present atypical presentation of conjunctival neoplasia observed in 2 cases. In case1 360 degree corneal involvement was noted as opposed to a single quadrant location which is more commonly described in literature. In addition the patient was young male which is extremely uncommon. The second case was an elderly female which is the more common age of presentation. In both the cases there were no predisposing factors. Both the cases continue to be on regular follow up as recurrences are known and need to be picked up early. Few studies have given trial of 5-Flurouracil (5 FU) and/or 0.4% Mitomycin-C (MMC) and/or Cyclosporin A and/or Interferon with non conclusive results.

Conclusion
Though conjunctival neoplasias are uncommon, in cases of recurrent pterygium or chronic conjunctivitis (> 3 months) cytological examination is must for early diagnosis. A high index of suspicion is must to pick up these cases early and prompt and complete excision with a 2mm disease free margin may help in complete resolution of the disease. Additional topical treatment has been tried in with unclear benefit. Close follow up of all cases is necessary for early identification of recurrences.

References
Isolated Strabismus as a Presenting Feature of Large Pituitary Macroadenoma

Kamaljeet Singh, Prateek Gujar, Nida Usmani, Santosh Suman

Department of Ophthalmology, Moti Lal Nehru Medical College, Allahabad

Pituitary adenomas usually present with visual field defects and headache. Large tumors may cause diplopia and ophthalmoparesis by cranial nerve compression in cavernous sinus. However, all such cases almost always have poor visual acuity also. We present a rare case of a 35-year-old male with a large pituitary adenoma who presented with headache, ptosis and diplopia but had normal vision in both eyes. He had supero-temporal quadrant visual field defect in the right eye and an atypical visual field defect involving the temporal quadrants in the left eye. The relevant literature is reviewed in the discussion.

Introduction

Pituitary tumours comprise 12 - 15 % of all intracranial tumours. A majority of these are histologically benign [1]. Pituitary adenoma is a relatively common intracranial tumor that presents to ophthalmologists with vision loss and field cuts due to its location below the optic chiasma [2, 3]. It can also present with hormonal disturbances- hyperpituitarism (functioning adenomas) or hypopituitarism (from compression of normal hypothalamic pituitary axis). Clinically, pituitary adenomas present as secretory or non-secretory tumours; visual manifestations are more common amongst non-functional adenomas [4]. Large tumor also compresses cranial nerves in the cavernous sinus. When it does so, some degree of vision loss/temporal field cut is almost always present. The prevalence of field defects in pituitary adenomas in general has been reported in various studies as 37 to 96 % [5]. It is distinctly uncommon for pituitary adenomas to present only with features of extraocular palsy in absence of vision loss [6,7,8]. We report a 34-year-old man with non-functional pituitary macroadenoma who presented to us only with complaints of strabismus in absence of vision loss.

Case report

A 35 year old male came with the complains of headache for the past six months. Two months back he woke up one morning and found he could not open his left eye. He also suffered from double vision when he forced the left eye to open. On admission, visual acuity was 6/6 right eye and 6/6 left eye. Vision was assessed using Snellen’s optotype on self illuminated vision drum for distance. On testing the near visual acuity patient had a vision of N/6. Neurological examination revealed complete left oculomotor nerve paresis and right abducens nerve paresis. Left eye on examination revealed fixed abductant ocular position and absence of light reflex in the left eye. On testing the unocular movements (in the four directions), in the left eye patient had restriction of movement in adduction, elevation and depression. On testing in the right eye, he had restriction of abduction. On testing for binocular movements (Figure 1), our findings revealed paresis of right lateral rectus and left superior, inferior rectus and inferior oblique muscles. Fundus examination was within normal limits. Physical examination revealed no definite clinical signs of endocrinological dysfunction.

Computerized tomography showed a large expansile intrasellar mass with small suprasellar and left parasellar extension, with extension of mass in peripontine cistern. On the left side it was indenting on pons and compressing optic chiasma with possible invasion of left cavernous sinus and internal carotid artery. We suspected it to be a pituitary macro adenoma. On magnetic resonance imaging (figure 2), a large (41× 40× 38 mm) defined soft tissue lesion was seen expanding in sella turcica extending superiorly into suprascellar cistern and compressing the optic chiasma , laterally having left cavernous sinus invasion and posteriorly invading the prepontine cistern. Levels of prolactin, cortisol and TSH measured are normal. The visual field examination revealed a supero- temporal quadrant defect in the right eye and an atypical defect involving the temporal quadrants in the left eye.

We referred the patient to the department of neurosurgery for gamma knife surgery for further management. The patient is in our follow up.

Figure 1- Figure A& B demonstrates the position of the eye in primary gaze. Notable is the out and down looking left eye with ptosis Figure C-H demonstrates the extraocular motility in the different diagnostic position of gazes. Notable is the lack of adduction and dextroelevation in the left eye (C,D,E). There is limitation range of abduction in the right eye (D).
Isolated Strabismus as a Presenting Feature of Large Pituitary Macroadenoma

Discussion
Pituitary adenomas constitute 10-15% of intracranial tumors [2]. Hormone secreting adenomas present early due to characteristic adenomas. Pituitary macroadenomas classically presents with asymmetrical bitemporal hemianopia, although other patterns of visual dysfunction commonly occur depending upon the size of tumor, direction of growth, anatomic configuration of chiasma (prefixed, normal or postfixed) and chronicity of the process. Nonfunctioning adenomas are usually detected after they attain large sizes, and present with vision loss (70-90%), headache (40%), hormonal deficiency (15-40%), extraocular nerve involvement (1-4%), involvement of other cranial nerves in the middle fossa skull base (2-4%) and seizures (4%) [2,9,10].

Diplopia or extraocular nerve weakness is rare, and should raise the possibility of metastatic tumor/pituitary apoplexy/other diagnosis rather than routine non-secreting pituitary macroadenoma no matter how so ever large it may be [2,10]. Of patients presenting with extraocular nerve palsy, characteristic vision changes (bitemporal hemianopia/blindness) is almost always present, which help in clinical localization of the lesion [2].

Non-functioning pituitary adenomas constitute 25 - 30% of all pituitary tumours and present with predominantly ophthalmic features; field defects being the most common [1]. Patients with pituitary macroadenomas may not have symptoms of visual disturbance, yet may have field defects consistent with compression of visual pathways. It is therefore important to perform field testing on patients with pituitary adenomas even if they have no visual complaints. Automated perimetry is a sensitive method for detecting visual field damage and quantifying treatment results.

Review of relevant literature reveals that it is distinctly rare for such tumors to attain large sizes, cause extraocular nerve palsy and yet have no symptom/sign of optic nerve compression (not even on fundus examination). Visual evoked response (VEP) may reveal subtle optic nerve compression features in these patients.

Pituitary adenomas are responsible for a few cases of strabismus in adults, but these patients have other ocular/ neurological findings as well. Pituitary adenoma as a cause of isolated strabismus is very rare and ophthalmologists must be aware of this unusual presentation [6,7,8]. The other similar cases that have been reported are pituitary macroadenoma with isolated partial oculomotor nerve palsy in the setting of apoplexy [6], a case of haemorrhagic non-functioning pituitary adenoma presenting with abducens nerve palsy [8] and another case of acute third nerve palsy as the sole presenting sign [7]. Though these tumors are usually treated by neurosurgeons with micro neurosurgery or gamma knife, they frequently present initially to ophthalmologists with visual/ocular complaints. Ophthalmologists must therefore be aware of all presentations of these tumors so that diagnosis is made early and timely intervention/referral done.

References
Amblyopia A Historical Consideration

Shibal Bhartiya, Sumita Sethi

Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi

“To an unbiased observer the amblyopia treatment domain could appear to be a sort of privileged enclosure exempt from the obligation to apply the methodological rules universally adopted in clinical research concerning treatment of other diseases.”

Paliaga

A historical consideration of amblyopia treatment would undoubtedly lead the reader to conclude that this topic is fraught with empirical innovation, yet lacking in critical evaluation, and appearing to be insufficiently evidence based. If the history of amblyopia were to be classified on this basis of particular study designs within its hierarchy, defining the relative weight given to research findings when treatment decisions come to be made, then pictorially this would resemble a somewhat bottom-heavy pyramid with perhaps just a few systematic review at its apex. This article does not attempt to elucidate the extraordinary complexity of amblyopia, nor does it attempt to define the current scope of our knowledge. It just endeavours to summarize certain major milestone as regard to development and treatment of amblyopia in animal models and in humans. Our understanding of amblyopia is based on studies of experimentally produced strabismic, anisometropic and stimulus deprivation amblyopia in animal models. Extensive studies of clinical amblyopia have been made using psychological, electrophysiologic and behavioral methods. The results of these investigations have revealed a complex syndrome of sensory and motor anomalies, of which reduced visual activity is the most prominent part of the overall disturbance clinically. Inherent limitations in clinical research methods, however, have precluded precise analysis of the amblyogenic mechanisms and identification of its seat within the retino-cortical pathway. Von Graefe defined it as the condition in which the observer sees nothing, and the patients very little. It is the functional counterpart of the neurophysiologic and neuroanatomic aberrations that result from abnormal visual experiences in early childhood. Chavasses concept of amblyopia of arrest states that the visual acuity remains at the level of development present at the time of onset of strabismus, which if persistent would result in superimposition of suppression amblyopia called amblyopia of extinction[1].

Experimental Amblyopia

A major breakthrough in this field occurred in the early 1960’s when Wiesel and Hubel published their now classic papers on the effect of visual deprivation induced by lid suture in visually immature kittens, on the physiology of the visual cortex, and on the histology of the lateral geniculate nucleus (LGN). They established that unilateral lid closure during the first 12 weeks of life severely reduces the number of cortical neurons which can be stimulated through the deprived eye as well as the number connected to both eyes (binocular neurons) and functional anomalies are accompanied by histologic changes in the LGN layers receiving input from the deprived eye. Following the pioneering work of Hubel and Wiesel, and of Ikeda and Wright[2-9], there has been a virtual cascade of information on changes in the visual cortex and the lateral geniculate nucleus (LGN) related to amblyopia, in both animal[10-14] and human models[15,16]. Due to the anatomic and functional differences in the organization of the visual system in the various species used, the data from later studies on several different animals cannot be employed to explain the mechanism of amblyopia in humans, strabismic or...
and treatment have lower prevalence of amblyopia than those finding that those populations that undergo early intervention better understanding of the natural history of amblyopia. The studies of early treatment intervention regimes allow clinical evidence to suggest that the function of an amblyopic eye is subject to inhibitory factors elicited by stimulation- of the normal eye. Thus a dual concept of amblyopiogenic factors or in the visual cortex. When visual experience is abnormal takes place either in the LGN, via translaminar connections, or in the visual cortex. When visual experience is abnormal early in life, the deprived cells are at a disadvantage in the competition and their growth is inhibited. Inhibiting interocular visual processes are known to occur even in normal binocular vision, as exemplified by retinal rivalry or the extinction phenomenon of Aulhorn. Binocular interaction has an even more profound effect in amblyopia, and there is ample clinical evidence to suggest that the function of an amblyopic eye is subject to inhibitory factors elicited by stimulation- of the normal eye. Thus a dual concept of amblyopiogenic factors has evolved from all these investigations.

Treatment of Amblyopia

The studies of early treatment intervention regimes allow better understanding of the natural history of amblyopia. The finding that those populations that undergo early intervention and treatment have lower prevalence of amblyopia than those that do not implies that amblyopia does not improve of its own accord.

Hubel and Wiesel coined the term ‘critical period’: a period of time in early life, during which the visual system shows lability of deprivation and ability for reversal of the effect of deprivation. Hardman Lea et al defined the sensitive period as that passage of time during which the development of the immature visual system may be altered by change in the quality, quantity or balance of the visual input via the 2 eyes.

Jastrzebsik et al devised a model of amblyopia, which describes sensitivity, plasticity and elasticity (SPE) in relation to its response to occlusion. Sensitivity indicating a propensity for the patched eye to worsen and for the amblyopic eye to improve and, among the sensitive eyes, some are distinguished in the model as elastic and some as plastic. Elasticity implying reversibility, after occlusion is discontinued, of improvement in the amblyopic eye, and of damage in the patched eye and plasticity implying a permanent change in both the eyes. A study done by Oster et al suggests that younger age may be associated with both greater sensitivity and elasticity. Their study indicates that stability i.e. loss of elasticity and presence of sensitivity and plasticity becomes evident between the third and fourth birthdays.

Recent randomised, controlled treatment trials, together with reviews of patients who have not been compliant with treatment, indicate that the natural history of amblyopia is not that of spontaneous recovery. Intervention is required to maximize potential visual acuity in the affected eye. The age at which that intervention will still be effective has not been confirmed and is the subject of on-going studies.

Priestly observed that after occlusion therapy some patients may show a change in fixation preferences without an improvement in visual acuity, where as others may improve in vision without a change in fixation. The possibility of angle of deviation being influenced by occlusion has drawn little attention. Swan in 1947 noted a significant increase in the angle of deviation in four out of one thousand patients following occlusion therapy. Pine and Shipman observed that occlusion therapy as a treatment of amblyopia whether full-time or part-time carries a very small risk (4%) of increasing a preexisting esodeviation by five prism diopters or greater so as to become cosmetically unacceptable.

Treatment modalities

Oclusion therapy

Since 1722, where Saint Yves first described occlusion of the dominant eye to promote use of the squinting eye, it has remained the mainstay of amblyopia therapy. Worth noted that the age at which a squint developed and the age at which treatment began were important in establishing prognosis. From his results developed Worth’s fraction:
age in months when permanent turn become apparent / age in months at which training began and it was used for prognosis. Occlusion provides the amblyopic eye a preferential chance of development as the dominant eye is withheld from binocular participation. The success rate of occlusion recedes with age, good compliance and age less than 6-7 years ensures a success rate of almost 100%[26-28]. Rutstein et al showed that patients with strabismic or anisometropic amblyopia show a better and faster gain in visual acuity when less than 7 years or less[28]. Most of the visual acuity improvement occurs within the first three months of treatment. Epelbaum et al studied 407 patients of strabismic amblyopia and noted that recovery of acuity of the amblyopic eye was maximum when the occlusion was initiated before 3 years of age, decreased as a function of age and was almost nil by the time the patient was 12 years of age[29]. Assaf, in a retrospective study involving 1904 patients of strabismic amblyopia noted that the period of maximum sensitivity to short periods of occlusion extended to 18 months, declining to about 30 months of age in terms of transfer of fixation[30]. Bangerter recommend the occlusion of the amblyopic eye to treat eccentric fixation[31].This inverse occlusion was supposed to interrupt the subnormal fixation behavior. Its use has now been discontinued as its much less effective than conventional occlusion[32].

Penalisation
Penalization has been described as an unpleasant neologism used for defining methods of treating amblyopia that selectively fogs the image of the sound eye[33,34]. In spite of better acceptance due to binocular stimulation, its only useful in unilateral amblyopia, requires prior solution of squint, anisometropia, and aniseikonia. It is also known to carry the risk of occlusion amblyopia[35] and therefore, its use is limited to non-strabismic, mild amblyopia and for maintenance therapy.

Pleoptics Therapy
In 1936, Comberg started active stimulation of the macula to treat eccentric fixation. Bangerter, who coined the term, propagated its use with inverse occlusion[36,37]. This method involves dazzling of the eccentrically fixating area with bright lights while protecting the fovea with a disc projected on to the fundus, followed by intermittent stimulation of the macula. Treatment is given under direct observation using a modified Gullstrained ophthalmoscope (Pleoptophor). Cuppers used a modified ophthalmoscope (Euthryoscope), which has discs of various sizes to create a central after image, apart from dazzling the eccentric point[38]. He also used the attenuate flashing of room illumination (alternoscope) to perpetuate after images, and devised the visuooscope. The afterimage is projected on the space coordinator where the hand-eye coordinator is releases. This is then followed by exercises with Haidinger brushes on Cuppers coordinator. The latter device uses the property of the fovea to polarize light. Inverse occlusion is continued till central fixation is achieved, after which direct occlusion is started. Both these methods, however, are time consuming, requires, elaborate instrumentation and a regular follow up. It requires the cooperation of an intelligent patient (therefore useful for older than 5 years old) and the duration of therapy required is longer.

Medical Treatment
There is evidence that plasticity of the visual system during the sensitive period is dependent on inputs from noradrenergic neurons, and is subject to pharmacological manipulations. In 1871, Nagel[39] used strychinine for the treatment of amblyopia, while Bieth attempted its use with oxygen. Barany and Hallden used alcohol as a inhibition mechanisms involved in amblyopia are known to involve synaptic neurotransmitters[40]. Pettigrew and Kasamatsu[41,42] used neither activation of neither central nor epinephrine system for enhancing neuronal plasticity, while Kasamatsu used beta-blockers like propranolol[41-44]. Duffy proved that bicuculline, a GABA receptor blocker, which causes catecholamine depletion, reversed visual deprivation[45,46]. Kasamatsu used beta-blockers like propranolol, and together with Pettigrew, also used central norepinephrine system to enhance neuronal plasticity[41-43]. Exogenous NGF (nerve growth factor) prevents the effects of deprivation in rats; Maffei et al predicted that loss of competition for deprived eye is due to lack of neurotropic factor, and replenishing it may prevent amblyopia[47]. Visual deprivation is known to decrease retinal dopamine concentration in children and monkeys. Catecholamines and other neurotransmitter like GABA, glutamate and acetylcholine are involved in neuronal plasticity in deprivation amblyopia and can restore partial visual acuity[48]. Bodis Wallner and Yahr reported that the cortical visual patterns evoked by a stimulus pattern might be altered in latency and waveform in Parkinson’s disease, which characterized by a pathological deficiency of the dopaminergic system[49,50]. Demenic L et al showed that dopaminergic drugs affect the visual performance of normal subjects, producing an improvement in contrast sensitivity[51]. Gottlob observed that the administration of dopamine in normal subjects increases the ERG b wave, selectively changes the amplitude of oscillatory potentials and reduces implicit time of the pattern VEP and pattern ERG[52].

Gottlob et al investigated the short-term effect of a large single dose (200µg) of levodopa on contrast sensitivity and binocular suppression in adult amblyopes, in a cross over, double masked study[53]. They reported an increase in the contrast sensitivity and a decrease in the fixation point scotoma, and
an increase in visual acuity by a half line in 2 of 9 patients tested (22%). Leguire et al confirmed these results in 8-12 years old amblyopes using 400mg of levodopa[54]. In order to determine the tolerance and efficacy of levodopa-carbidopa with part time occlusion therapy for childhood amblyopia, Leguire and coworkers carried out a double masked placebo controlled randomized longitudinal study in 10 amblyopic children between 6 and 14 years of age[55]. Subjects received on average, 0.48/0.12mg/kg body weight levodopa/carbidopa three times per day combined with part-time occlusion of the dominant eye (3 hrs/day) over a 3 weeks period. At the end of the dosing regime the levodopa/carbidopa group significantly improved in visual acuity by 2.7 lines and in mean contrast sensitivity by 70% in the amblyopic eye. The placebo group improved in visual acuity by 1.6 lines in the amblyopic eye. Tolerance and occlusion compliance was similar between groups. One month after termination of the treatment the levodopa/carbidopa group maintained a significant 1.2 lines improvement in visual acuity and 70% improvement in contrast sensitivity in the amblyopic eyes. The placebo group did not maintain an improvement in visual acuity between the eyes. They concluded that levodopa/carbidopa, at an average of 0.48/0.12 mg/kg. Body weight is well tolerated, and when combined with part time occlusion, is efficacious in improving visual functions in amblyopic children.

Citicoline (Cytidine–5 diphosphocholine) has been used clinically for head injury and Parkinsonism. In a dose of 1000 mg I.M. for 15 days, without any amblyopia therapy to patients aged 9-37 years (mean 16.6 years), it caused a temporary increase in visual acuity by a half line in 2 of 9 patients with moderate amblyopia. About 1 in 5 achieved visual acuity of 20/25 or better in the amblyopic eye.

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Multifocal Electroretinography

Lalit Aalok¹, Swati Phuljhele²

Vitreoretina Consultant, Perfect Vision Eye Hospital Faridabad¹, Senior Research Associate, Dr RP Centre²

Visual electrophysiological tests provide information about the functional integrity of the pigment epithelium (Electrooculogram or EOG), the photoreceptors and inner layers of the retina (electroretinogram or ERG), and the optic nerve and occipital cortex (Visual Evoked Potential or VEP). The information is objective and independent from the voluntary response of the patient and thus is especially useful for non-cooperative patients and infants. The multifocal ERG technique provides reliable parameters for the accurate evaluations of diseases and their different treatment modalities existing and emerging.

### Introduction

The full-field or Ganzfeld ERG (ffERG) provides a mass response from the retina, and thus will not be altered substantially by localised lesions of the macula, which contributes only 10% to the ERG response. Therefore its greatest value lies in disorders that result in diffuse functional impairment of the retina. On the other hand, a diffuse retinal disease with macular sparing will have abnormal ERGs but normal central visual acuity. The multifocal ERG technique (mfERG), developed by Sutter and Tran in 1992, provides spatial maps of ERG responses from multiple areas of the macula and would be a very useful technique to assess macular status in such situations. Multiple local ERG responses, typically 61, are recorded from the cone-driven retina under light-adapted conditions. Although multifocal ERG recording of rod function is possible, maintaining dark adaptation during testing is difficult in an actual clinical setting. Focal ERG uses a focal light stimulus to elicit a local ERG response, a process which would be excessively time consuming if multiple areas were to be tested in succession as in mfERG. While mfERG provides useful information about the photoreceptor status, it requires preparation of the patient and the output is complex, requiring expertise in interpretation; its use is therefore mainly limited to specialist centres where research is carried out. Wide-field mfERG permits assessment of retinal function from the central 90 degrees of the retina.

### Methodology

#### Patient preparation

The pupils must be fully dilated prior to mfERG testing as changes in pupil size alter the mfERG responses significantly. Patients must be light adapted for at least 15 minutes in room light for obtaining the cone driven responses, and recording is done with the room light on. Patients need to have a visual acuity permitting fixation upon the target. To help patients having poor vision maintain an accurate fixation upon the presented target, as well as to promote results of the best quality, an optical correction appropriate for viewing distance must be used at all times. The near infra red image sensor incorporated in the optoelectronic stimulator enables checking for stable central fixation during mfERG testing. “Large field” eye glasses should be preferred to avoid masking of the peripheral parts of the stimulation display. Recording may be done monocularly (preferable), or binocularly to aid in fixation at the target in instances of low subject visual acuity.

#### Recording electrodes

These may be of contact lens or non-contact lens types.

### Table 1: Comparison of different ERG techniques

<table>
<thead>
<tr>
<th></th>
<th>Focal ERG</th>
<th>Full-field/Ganzfeld ERG (ffERG)</th>
<th>Multifocal ERG</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Test duration</strong></td>
<td>Time consuming</td>
<td>Time for dilatation and dark adaptation-30 minutes</td>
<td>Time for dilatation and light adaptation is around 20 minutes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Time for actual test is around 30 minutes</td>
<td>Time for actual test is less than 10 minutes</td>
</tr>
<tr>
<td><strong>Identification of disease</strong></td>
<td>Localised disease can be better detected</td>
<td>Unable to detect localised retinal disease; examines entire retina</td>
<td>Localised disease can be better detected; more sensitive; examines the central retina</td>
</tr>
<tr>
<td><strong>Interpretation</strong></td>
<td>Requires familiarity</td>
<td>Requires experience</td>
<td>Output requires expertise in interpretation</td>
</tr>
</tbody>
</table>
Multifocal Electrotretinography

Contact lens type electrodes are optically clear and lie in contact with the cornea, contact being aided by use of either topical anaesthetic drops or artificial eye drops. Repeated use should be avoided to prevent the risk of disease transmission or disturbance of their transparency. Corneal electrodes provide responses of larger amplitude and with fewer artefacts. Examples of contact lens electrodes are Jet electrode (unipolar), Dorian Gold lens (bipolar) and Burian-Allen electrode (unipolar or bipolar). Burian-Allen electrodes make use of an incorporated speculum to hold the lids apart and prevent blinking.

Non-contact lens electrodes may lie in contact with the cornea or the bulbar conjunctiva. Examples of non-contact lens type electrodes are gold-foil electrodes, H-K loop and DTL fiber. The active electrode lies in the inferior fornix or on the cornea, the reference electrode lies lateral to the lateral canthus (if using a unipolar electrode). The ground electrode is placed in the centre of the forehead. (Figure 1: Placement of electrodes)

Recording technique

The stimulus source may be any one of Cathode Ray Tube (CRT), Light emitting diode (LED), Liquid Crystal Display (LCD) screen or Scanning Laser Ophthalmoscope (SLO). The advantage of the scanning laser ophthalmoscope (SLO) is that it shows the exact anatomical location of the stimulus relative to the fundus of eye.

The recording technique describes the one utilised in the MetroVision system (Perenchies, France). A field of ±30° horizontally and ±24° vertically centred on the fovea is stimulated at a viewing distance of 30 cm with a stimulus consisting of an array of usually 61 squares or hexagons (scaled or of uniform size) on a high resolution colour monitor. (Figure 2) Increasing the number of stimulated hexagons to 103 or 241 or higher improves the spatial resolution but progressively increases the testing time. The sizes of the hexagons are scaled (increased in size) with retinal eccentricity from the fovea to the periphery to elicit approximately equal amplitude responses from all locations. This is because the concentration of the cones decreases with increasing distance from the fovea.

A red fixation target is presented within the central hexagon. The luminance of each hexagon is independently alternated between black and white (93% contrast) according to a pseudorandom binary ‘m-sequence’ at a frame rate of 75 Hz i.e every 13.33 seconds. The mean luminance of the stimulus is 100 cd/m² with the stimulus screen being surrounded by an uniformly illuminated background cover with the luminance set at 30 cd/m² to eliminate the rod responses. At a stimulus frequency of 18 Hz, 5000 responses are acquired over a period of 5 minutes for each eye.

Understanding the mfERG response

The human mfERG is dominated by the cells of the outer retina, namely the photoreceptors and the bipolar cells. The standard mfERG mainly measures the cone function and the response components are technically known as kernels. The most commonly analysed responses are the first-order and the second-order kernel components. First order kernels are obtained by adding all the records following the presentation of a flash in that hexagon and subtracting all the records following a dark frame. (Figure 3) shows the typical waveforms in the first-order kernel mfERG electrical response. The waveform is biphasic with an initial negative deflection followed by a positive peak. There may be a second negative deflection after the positive peak. These peaks are labelled as N1, P1, and N2, respectively. The N1 response amplitude is measured from the starting baseline to the base of the N1 trough; the P1 response amplitude is measured from the N1 trough to the P1 peak. The peak implicit times are measured from the stimulus onset.

The second order kernel represents the temporal non linearity of the local responses of the retina or in other words is a measure
of how the mfERG response is influenced by the adaptation to successive flashes. Many authors claim that temporal non linearities arise from the inner retina and therefore abnormal second order kernel may indicate abnormal processing or adaptation at the level of the inner retina. It has an initial

Table 2: P1 wave characteristics according to damage to different retinal layers

<table>
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<tr>
<th>Damage to</th>
<th>P1 wave of mfERG</th>
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<tbody>
<tr>
<td>Cone receptor</td>
<td>Amplitude</td>
</tr>
<tr>
<td>Outer plexiform layer</td>
<td>Normal or larger</td>
</tr>
<tr>
<td>Inner plexiform layer</td>
<td>Normal</td>
</tr>
<tr>
<td>On-bipolar cells</td>
<td>Smaller</td>
</tr>
<tr>
<td>Off-bipolar cells</td>
<td>Larger</td>
</tr>
<tr>
<td>Ganglion cells</td>
<td>Normal</td>
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Figure 3: The multifocal ERG waveforms

Figure 4: Trace array display

Figure 5: Three dimensional topographic display showing foveal peak and the blind spot

Figure 6: Group average by rings

Figure 7: Display output of results

Figure 8: Macular sparing with a foveal peak on multifocal ERG in a patient with retinitis pigmentosa.
positive peak followed by a negative trough, called P1 and N1, respectively.

The mfERG response output can be in several forms.

1. Trace array: The individual electrical trace response from each stimulated zone is displayed in an array from the entire tested area. (Figure 4)

2. Topographic color map displays of amplitude and implicit times: These may be 2 or 3 dimensional displays. (Figure 5)

3. Group averages: Grouping of the electrical responses may be done by concentric ring zones, quadrants or by user defined zones. (Figure 6) The most commonly reported result is response density, in which the responses from the elements in each ring are summed and then divided by the area of these elements.

The average response per zone is displayed in nV/deg² in graphic form, or as a histogram. (Figure 7) The amplitudes and implicit times for the different waveforms are also displayed for each zone. The RMS analysis (root mean square) identifies the presence of a response when noise is present. The RMS measurement of the bar in color, which represents the signal column, is compared to the RMS measurement of the black bar, which represents the noise column. The multifocal map can also be superimposed on the image of the patient’s eye fundus. Each laboratory must develop its own normative data which preferably should be age-adjusted.

Test guidelines for mfERG testing have been published by the International Society for Clinical Electrophysiology of Vision (ISCEV: http://www.iscev.org), to allow for further research before standards are set. As is expected, the stimulus and recording parameters, and the adaptive state of the eye strongly affect the electrophysiological responses, necessitating standardisation for purposeful scientific exchange of test results.

Indications

1. The mfERG can be used to differentiate diseases that affect the outer retina from those that affect the ganglion cells or optic nerve. (Table 2)

2. Age-related macular degeneration (AMD): A significant reduction in the foveal P1 amplitude and delay in N1 implicit time is seen in early AMD. More severe alterations are seen in more severe disease. It has been suggested that rod-mediated mfERG may be more sensitive in detecting retinal dysfunction in early AMD.

3. Diabetic retinopathy (DR): The implicit time measures are more sensitive as compared to the amplitude changes, which are reduced, in detecting retinal dysfunction in DR. The foveal thickness measurement on Optical Coherence Tomography (OCT) has been found to correlate significantly with the mfERG amplitudes and implicit times.

4. Retinitis pigmentosa: The amplitude is decreased typically along with delays in implicit times. Implicit time alterations have been found at times when changes in response amplitudes had not occurred, laying stress on the measurement of implicit times over amplitude parameters. These changes are more prominent in the peripheral regions and wide-field mfERG may be more useful in detecting affectation of the more peripheral retinal areas. (Figure 8) A study by the author in 87 eyes with retinitis pigmentosa revealed highly significant correlation between the visual acuity and the P1 and N1 response amplitudes in patients with retinitis pigmentosa.

5. Patients with Stargardt’s disease have decreased amplitude in the region of central macula with minimal implicit time delays.

6. Retinal vascular occlusions (RVO): There is reduction in the amplitude and increase in the latency of mfERG responses in vascular occlusions. The alterations on mfERG are found to correlate with the areas of visual field changes in both branch arterial and venous occlusions. Branch arterial occlusion causes damage to the inner 2/3rd of the retinal layers and thus the mfERG changes are more prominent in the second-order kernels than the first-order kernels. Laser treatment is followed by implicit time delays and amplitude reduction in DR and vascular occlusions after 8-12 weeks, with the implicit time alterations being more prominent.

7. Glaucoma: Though significant reductions in both first-order and second-order kernel response amplitudes are seen in glaucoma, correlation with the visual field changes are inconsistent and lacking and mfERG may not be a reliable indicator of functional loss in glaucoma.

8. The mfERG technique can be used to follow the effects of clinical intervention, for example after PDT therapy for AMD, before and after retinal detachment surgery or macular hole surgery, after treatment of macular edema in diabetic retinopathy using different treatment modalities, after retinal transplant procedures or in patients with retinitis pigmentosa after stem cell therapy.

References


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