Toxic / Nutritional Optic Neuropathy

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These disorders are often grouped together as they cause a syndrome characterized by papillomacular bundle damage, central or cecocentral scotoma, and reduction of color vision. The anterior visual pathway is susceptible to damage from toxins or nutritional deficiency (Table 1). Both toxicity and malnutrition, acting either independently or together, have been implicated in the pathogenesis of these disorders.

Ethambutol is one drug that is commonly is associated with toxic optic neuropathy. The optic neuropathy that occurs is dose dependent and duration related. Loss of vision does not tend to occur until the patient has been on the drug for at least 2 months, but symptoms generally appear between 4 months to a year. This onset may be sooner if the patient has concurrent renal disease because this will result in reduced excretion of the drug and, therefore, elevated serum levels. The toxicity of this drug is dose related. The patients on dosages of 25 mg/kg/d or greater are most susceptible to vision loss. However, cases of vision loss, even with much lower doses, have been reported. Isoniazid, another antitubercular drug, also can produce toxic optic neuropathy, and patients with concurrent hepatic or renal disease are at higher risk. Amiodarone, an anti arrhythmic drug has been implicated as a cause of an optic neuropathy. Its most common ocular side effect is verticillate keratopathy. Although the optic neuropathy is typically bilateral and symmetric with visual loss and/or field loss, it also may present unilaterally. With this drug, the toxicity to the optic nerve also appears to be dose related, with dosage varying from 200-1200 mg/d. Visual complaints may start 1-72 months after the initiation of treatment and are slowly progressive. The optic neuropathy from amiodarone, should not be confused with the acute nonarteritic ischemic optic neuropathy-like picture also reported with this drug.

Pathophysiology

The exact mechanisms by which nutritional deficits or toxins damage the optic nerve has not been elucidated. The etiology is most likely to be multifactorial. In most cases, the toxins impair the vascular supply or metabolism. In patients who abuse ethanol and tobacco, malnutrition is the principal cause of the visual loss. The specific deficiencies in vitamin B-12, thiamine, folic acid, proteins with sulfur-containing amino acids, or any combination of these also play a role. Over time, these deficiencies cause accumulations of formic acid. Both formic acid and cyanide inhibit the electron transport chain and mitochondrial function, resulting in disruption of ATP production and ultimately impairing the ATP-dependent axonal transport system.

It has been hypothesized that the chelating properties of ethambutol contribute to its neurotoxicity, but this has yet to be proven. The mechanism of the neurotoxicity that occurs from the antiarrhythmic amiodarone remains unclear. Researchers believe that it may relate to a lipidosis that is induced by the drug.

History: When a patient is suspected of having toxic or nutritional optic neuropathy, a thorough history is invaluable and should cover diet, drug/toxin exposure (eg, heavy metals, fumes, solvents), social history including tobacco and alcohol use; and occupational background, with details on whether similar cases exist among coworkers. Treatment of any chronic disease or illness should always be elucidated. A family history also should be taken. Persons with alcoholism are not always forthcoming with their drinking habits; therefore, obtaining these details, along with diet details, from friends or relatives may be more reliable.

Signs and symptoms: Toxic/nutritional optic neuropathy often presents as a painless, progressive, bilateral, symmetrical visual disturbance with variable optic nerve pallor. Pallor in the temporal quadrant is most common with this condition. The patient may manifest reduction in visual acuity (20/50- 20/200), loss of central visual field (usually relative cecocentral scotoma) and reduced color perception.

If the visual loss is unilateral or significantly symmetric and / or is associated with pain as one of its symptoms, in such cases, other diagnoses should be considered.

In toxic optic neuropathies, the visual loss may be acute as well as chronic, depending on the insult. Ascertaining whether the onset of the visual symptoms was during or very shortly after exposure to a particular toxin is important. Establishing similar illnesses in coworkers or others exposed to the same drug or chemical also may be helpful.

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There is no relative afferent pupillary defect in these cases because the optic neuropathy is virtually always bilateral and symmetric. However, in most patients, the pupils are bilaterally sluggish to light.

In nutritional optic neuropathies, the optic disc may be normal or slightly hyperemic in the early stages. In a small group of patients with hyperemic discs, one could find small splinter hemorrhages on or off the disc. In longstanding cases, one might find papillomacular bundle dropout and temporal disc pallor, followed by optic atrophy.

In the early stages of toxic optic neuropathies, most patients also have normal-appearing optic nerves, but disc edema and hyperemia may be seen in some intoxications, especially in acute poisonings. Papillomacular bundle loss and optic atrophy develop after a variable interval depending on the responsible toxin.

Table 1: Agents that Can Cause Toxic / Nutritional Optic Neuropathy

➢ Ethambutol	➢ Methanol
➢ Isoniazid	➢ Carbon monoxide
➢ Amiodarone	➢ Lead
➢ Ethylene glycol (antifreeze)	➢ Mercury
➢ Chloramphenicol	➢ Thallium
➢ Digitalis	
➢ Chloroquine	
➢ Streptomycin	
➢ Quinine	
➢ Vincristine and methotrexate	
➢ Sulfonamides	
➢ Malnutrition with vitamin B-1 deficiency	
➢ Pernicious anemia (vitamin B-12 malabsorption)	

In ethambutol toxicity, fundus is normal initially; atrophy develops later if the drug is not discontinued. With isoniazid toxicity, optic nerve swelling has been reported.

Patients on amiodarone typically present with bilateral optic disc swelling, which can be quite marked, along with flame-shaped hemorrhages. If appropriate action is not taken, this may be complicated by permanent visual loss and bilateral optic atrophy.

Investigations

Evaluation for toxic optic neuropathy includes a complete ocular examination with color vision testing and threshold visual field testing. Also refer the patient for complete physical and laboratory studies such as a complete blood count with differential, serum B-1, B-12 and folate levels, a heavy metal screening (lead, thallium) and test for the Leber's mitochondrial DNA mutation. Although imaging studies yield normal results in toxic/ nutritional optic neuropathy, they almost always are indicated, unless one is absolutely certain of the diagnosis. The most appropriate imaging study is an MRI of the optic nerves and chiasma with and without gadolinium enhancement.

Visual field testing and color vision testing absolutely essential in the evaluation of any patient suspected of having toxic/nutritional optic neuropathy. Central or cecocentral scotomata with preservation of the peripheral field are characteristic visual field defects. These cases show red green colour defects, but sometimes blue yellow defects may also be seen.

Management: The first step in managing toxic-nutritional optic neuropathy is to remove the offending agent. This may cause some reversal of the process. Other than stopping the drug, no specific treatment is available for the optic neuropathy caused by ethambutol. Once the drug is stopped, recovery may take weeks to months. However, the vision may still decline or fail to recover even when the drug is stopped. High dose vitamin C can be tried with some success in certain cases. For isoniazid, vision also improves when administration of the drug is ceased. Pyroxidine 25-100mg/day may help stabilize or reverse isoniazid-caused toxic neuropathy. Because these drugs may be given concurrently in the treatment of tuberculosis, and both may produce a toxic optic neuropathy, physicians should remember that if stopping one does not result in the improvement of a patient's vision, then the other drug also should be stopped. Prompt discontinuation of amiodarone is essential if evidence exists of toxic optic neuropathy from the drug. The visual symptoms, along with the disk swelling, will improve gradually over the next several months, rather than immediately. However, visual loss or associated field defects reportedly can be permanent despite discontinuation of the drug.

In cases of nutritional optic neuropathy, improved nutrition is the key, as dietary deficiency is the common denominator in these patients. It cannot be overemphasized to patients that stopping, or at least

reducing, their smoking or consumption of alcohol is critical to their recovery. The latter, combined with an improved diet and vitamin supplementation, are the mainstay of therapy in nutritional optic neuropathy. Various authors prefer giving thiamine 100 mg PO bid, folate 1 mg PO qd, a multivitamin tablet daily. Vitamin B-12 injections are usually reserved for patients with pernicious anemia.

Patients with nutritional/toxic optic neuropathy should be observed initially every 4-6 weeks and then, depending on their recovery, every 6-12 months. At each visit, the patient's visual acuity, color vision, visual fields, pupils, and optic nerves should be assessed.

Prevention

Patients in whom ethambutol or isoniazid is indicated need to have a baseline ophthalmologic examination before treatment is instituted and should be monitored by their ophthalmologist periodically as long as they are on the drug to detect any optic nerve toxicity as soon as possible. The more sensitive visual function tests like visual evoked response (VER), Colour vision , Contrast sensitivity etc detect the optic nerve damage early and hence can help to prevent severe visual dysfunction, if the drug is stopped at the initial evidence of optic nerve damage. Any patient for which amiodarone is being considered for treatment requires a baseline ophthalmic examination before the drug is initiated. Furthermore, once on the drug, patients should be evaluated at least every 6 months. Even if a patient presents with corneal changes associated with the drug, their decreased vision should never be attributed to this until any pathology of the optic nerve has been excluded. Chronic smokers and alcohol abusers should seek assistance on methods to stop or reduce their smoking and/ or alcohol intake.

Prognosis

If patients with nutritional optic neuropathy are compliant with the treatment regimen, and unless the loss of vision is already far advanced, the prospect for recovery or at least improvement is excellent, except for the most chronic cases. However, the rate of recovery varies from a few weeks to several months.

For toxic optic neuropathies, when the responsible toxin is discontinued, vision usually recovers to normal over several days to weeks. However, this also depends on the nature of the offending agent and on its total exposure before it was removed. Some cases however fail to improve at all or show minimal improvement.

Suggested Readings

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Impact of Vision Loss on Development, Its Management and Early Intervention

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Unique Needs of Children With Visual Impairments: Early vision loss affects every area of development including cognitive, social, emotional, communication, self-help, and both fine and gross motor skills. The unique educational and developmental needs of children with vision impairments can best be assessed and interpreted by qualified educators. Vision is the primary source of information for most children. No other sense can stimulate curiosity, integrate information or invite exploration in the same way, or as efficiently and fully as vision does.

A baby who is born blind or severely visually impaired experiences the world differently from typical developing children. Since 85% of all early learning is visual, the child who is blind or visually impaired is at great risk for developmental delays. Effective, intensive intervention is imperative in the early years.

When a child is born with, or acquires vision impairment, the family is challenged to understand the visual diagnosis, the impact of vision loss on development, and to begin to bring the world to the child. Without a dependable visual system the child with vision impairment* is called upon to understand the world through incomplete messages from the other senses of touch, sound, taste and smell. The child needs to organize this incomplete information and then respond to what may be a confusing view of the world.

As well, the child cannot learn everyday tasks by observing others. As a result, the ability to understand basic life concepts, and the process of accomplishing most daily activities is seriously compromised. For example, the child who is unable to see family members going through the steps of preparing a meal at home misses valuable understanding of how things happen in daily life.

Family members provide the link to the world for the child with vision impairment. Carrying the baby in a front pack while doing chores around the house, setting aside a drawer with interesting and safe kitchen items for play, taking a sensory walk around the neighborhood, or describing everyday noises to the child are activities that make the connection to people and events in life. The child becomes most familiar with these routines by doing them again and again with family members and friends. At first, the child may be hesitant to touch things, but gentle encouragement over time usually helps the child to place little hands on toys and real objects.

Once pieces of information about the world are understood as a whole (this is a lifelong process!) developing problem solving skills can follow. Only through experience based learning, as described above, does the child gain personal validation of what the world is about in a way that makes sense to him.

Currently, the majority of children with vision impairments have additional disabilities Taking care of general health issues is critical in the early months of life, and appointments with physicians and therapists can often fill the waking hours of family members. Eating, communication and movement skills are the focus of many interventions and therapies.

The Impact of Visual Impairment on Development

There are five primary developmental areas and vision loss has am impact on all or any of these:

- Physical development
- Language and speech development
- Social and emotional development
- Adaptive development and
- Cognitive development

Children may experience mild to severe delays in one or all the areas. Lack of imitation through observation greatly limits their learning.

1 ***Sensory Development*** For the infant born without sight, the other senses have intermittent input and may appear diminished. The child receives inconsistent, discrete, and generally unverified fragments of information. Hearing is the only distance sense available to the blind infant, but the infant has no control over the presence or absence of sound in his environment. Sound without visual verification is only noise coming from nowhere. Only after much tactual, motor, and auditory interaction does sound acquire meaning. Only then can sound provide information about location, cause, or source.

Sound is not the strong motivator that vision is. Not until approximately 12 months - will a blind child reach

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for an object based on sound cue alone. Environmental exploration. is usually delayed until the child reaches this point.

Normally visual dimensions supply the incentive for tactile exploration: color, pattern, shape and location. These dimensions are unavailable to a blind infant; therefore, purposeful tactile activity is minimal because the environment remains unknown and uninviting.

2 Motor Development

Hands: Although the hands are a major perceptual organ, a blind infant has significant developmental delays in his ability to employ his hands functionally. Even at 5 months a blind infant's hands will be fisted and held at shoulder height. There will be no mutual fingering, no engaging at the midline. At this age, a sighted child is practicing coordinated reaching and transference of objects from one hand to another.)

This delay in hand utilization will result in delayed fine motor and gross motor development. Without vision, hand and eye do not work together. Instead, ear- hand coordination must occur. However this takes much experience and is achieved much later than normal eye-hand coordination.

Body: A blind infant usually achieves control of his posture at approximately the same age as sighted infants through the following normal progression:

- sits alone momentarily
- rolls from back to stomach
- sits alone steadily
- takes stepping movements when hands are held
- stands alone
- bridges on hands and knees

However, the achievements that require self- initiated mobility are significantly delayed:

- elevated on arms in prone
- raising to a sitting position
- pulling to a stand
- walking alone

Until a blind child will reach out to grasp a sound cue (12 months), he will not move out in space either on hands and knees or feet. The blind child's difficulty or reluctance in moving around the environment encourages passive behavior such as self- stimulating mannerisms.

Self- Concept: The blind child has an unusual dependence on a sighted person to mediate and help integrate his environment. This notion of dependence must be considered as a major factor in the blind child s development. The blind child has diminished control over his environment and can only control his inner world. As he withdraws into this world, he diminishes the need for social interaction. He may not understand that there is a complex world outside of himself, that he is separate from it, that he can both act on it and be the recipient of action.

3 Cognitive Development

Construct of World: The blind child has limited ability to coordinate and organize elements into higher levels of abstraction, and to verify the information. Therefore, he constructs a reality that is different from the sighted child's. The process of establishing concept-defining attributes and relationships is more problematic for the blind child and less accessible to guidance. The blind child is continually involved in problem solving, but this process, which is essential to future development, is more difficult and less rewarding for him.

Object Permanence: A stable visual field is the basis of object permanence and other conceptual tasks. A blind child cannot obtain object permanence until he has the ability to reach for objects based on sound cue alone. It is acquired nearly a year later than in sighted children.

Causal Relationship: Since the results of actions cannot be seen, the blind child may not be motivated to action. He may not understand his ability to cause things to happen or to retain pleasurable stimuli.

Constancy: Understanding how to align blocks or orient his hands on a page in order to duplicate a pattern will be difficult if he hasn't observed objects in various orientations to know that an object is the same regardless of its position in space.

Classification: Limited opportunities to explore objects and to see similarities are reflected in preschool blind children's classification errors. Concepts of same and different can evolve only if children identify the distinguishing variable on which to focus. A blind child has little difficulty generalizing across size, but numerous experiences with a variety of similar objects were required to expedite generalization and association skills.

Conservation: A blind child exhibits delays in conservation of substance, weight, volume, length and liquids.

4 Social Development

Relationships: In a sighted child the mutual smile between infant and mother is the beginning of attachment, recognition, and communication. The blind child will smile at 2 months in recognition of his mother's voice, but only nuzzling or tickling will regularly elicit a smile.

In later years, the child appears to have ambivalent emotional involvement and appears disinterested, non-communicative, and uninformed about the rudiments of play with his peers. Consequently, he may be avoided by

his peers and rejected or overprotected by strangers and relatives. All in all, his social interactions are more complicated because subtle visual cues are missing and facial expressions are lost.

Self- Help: Many self- help skills that are normally learned by watching are delayed in blind children.

Chewing, scooping, self- feeding skills may be delayed 2 years or more. Brushing teeth is difficult to accomplish since the child may reject the texture and has no opportunity to observe others performing grooming skills. Fear of the unknown and inability to locate the bathroom may contribute to delayed toilet training.

5 Language Development

Imitation: Much of what is learned by the normal child is learned by imitating others.

Total communication; including signing and finger spelling, is rooted in the development of imitation. Imitation signals the beginning of symbolic meaning in a child. The blind child needs planned, systematic instruction directed at the development of deferred imitation.

Use of Language: The blind infant may jabber and imitate sooner than a sighted child, but may show delay when combining words to make his wants known. The blind child primarily uses language to saris- y his immediate needs or to describe currant activities. He initiates few questions and his use of adjectives is sparse. The blind child may take in the sounds which make up the language, but may not grasp the meaning intended by the speaker. His sensory experiences are not readily coded into language. He may store phrases and sentences in his memory and repeat them out of context. The blind child often has a language that is echolalic preservative and meaningless. The early language of the blind child does not seem to mirror his developing knowledge of the world, but rather his knowledge of the language of others.

Personal Pronoun.

To correctly use the personal pronoun "I", a child must have established a sense of himself as separate from the environment. Since the development of self- concept in a blind child is delayed, he tends to confuse the use of personal pronouns, extending the use of the second and third personal pronouns or his own name to refer to himself.

Experience: The blind child is often hesitant to explore because of fear of the unknown. He is also often discouraged from exploration by adults who are overprotective. Without concrete experiences, the child will not develop meaningful concepts or the language to describe or think about them.

Early intervention and management of a child with low vision / severe visual impairment

> ***Vision rehabilitation*** services implies a comprehensive approach that focuses on rehabilitating an individual to his / her maximum potential. While vision rehabilitation cannot restore lost sight, it can maximize any existing sight a child has or, it can equip them with techniques to live an independent life.

More children are now surviving infancy with inherited or congenital visual disorders. However, the social and economic consequences of blindness and low vision in children are very important, in terms of the number of years of visual impairment and the emotional and financial demands on their parents and on the whole community. Low vision care is one of the most challenging problems in India. In order to establish rehabilitation protocols and to evaluate the efficiency of established services, the level of benefits and improvement of quality of life of the low vision children population must be determined. Comprehensive medical care includes preventive, curative and rehabilitation medicine. In ophthalmology we usually talk about

- preventive ophthalmology or primary prevention,
- curative ophthalmology or secondary prevention of blindness and
- low vision rehabilitation or tertiary prevention of blindness.

All three aspects of prevention of blindness should be present in the work of each unit. The goal of low vision rehabilitation is to reduce the functional vision loss by *enhancing* the visual function through the use of different optical and non-optical devices and by compensating, *substituting*, the loss by the use of new skills and techniques based on other modalities. In young children there is also an arrow backwards from visual impairment to visual disorder because of the pliability of the visual system. In these cases **therapy functions also as treatment** because it may bring considerable improvement in the visual functioning, more than would be possible through the age related development of vision without supportive therapy. This is an extremely important aspect of paediatric early intervention. The role of vision stimulation and visual efficiency therapy are crucial as paediatric early intervention.

"Visual stimulation" is the exposure of a passive child to some kind of light stimulus in artificial situations. The approach of vision stimulation and active learning was developed over the past 30 years, primarily while working with children who were blind with additional disabilities

such as mental retardation, spasticity, epilepsy, autism and hearing loss. While developing the approach it was discovered that infants and toddlers with vision impairment would also benefit from having optimal opportunity to learn rather than from being trained or taught. From the very outset, seeing is an activity; it means active looking, searching and selecting. Children will seek, fixate, analize, and recognize only those stimuli that are significant to them within their personal life context.

Who require vision stimulation?

Children less than six years of age respond best to vision stimulation based on the concept of plasticity. However if done on older patients for a longer period of time, one could get good results too. Those diagnosed with any residual vision are the best candidates for vision stimulation. Also not to forget those with cortical visual impairment and delayed visual maturation. Amblyopia therapy combined with vision stimulation had also given remarkable increase in functional vision and visual acuity.

***"visual efficiency therapy"* -** How vision is used can be improved with training. Measures of vision do not change after training, that is, visual acuity or visual fields will not change because of the training

Aim of visual efficiency therapy or vision training:

- To encourage and help each person make best use of their vision
- To provide a variety and number of opportunities for the person to learn about and understand their environment.

This is a way of finding out if there are problems in vision and visual development.

Fundus Fluorescein Angiography (FFA)- Chief findings in retinal diseases

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Fundus fluorescein angiography has always remained an important tool in the hands of retina specialists inspite of welcome arrival of another significant tool called Optical Coherence Tomography (OCT) used for in vivo histopathology like retinal evaluation.

In this article, we have tried to very briefly summarize the characteristic angiographic features of various disease entities in its various stages and which particular angiographic phase to look for that characteristic finding. Details of basics of FFA and pathophysiology of disease concerned are beyond the scope of this article.

To describe briefly abnormal fluorescence is either hypo (poor fluorescence) or hyper (more fluorescence). Hypofluorescence is either of choroidal circulation or of retinal circulation and is either due to being blocked by overlying hemorrhage etc. or due to poor filling of choriocapillaris/retinal circulation. Hyperfluorescence is either transmitted i.e. window defects (due to RPE atrophy) or is leakage (this could cause staining of tissues or pooling into potential space). Pre-injection (Pseudo) fluorescence is either due to autofluorescence of certain structures (e.g. optic nerve head drusen) or mismatched camera filters.

1. Mild Non-proliferative Diabetic Retinopathy (NPDR)

i) Microaneurysms- hyperfluorescent dots that may leak in later phases.

ii) Superficial & deep retinal hemorrhages- cause blocked choroidal fluorescence.

2. Severe NPDR (Fig.1)

i) All features as in mild NPDR

ii) Capillary non-perfusion (CNP) areas- seen as areas of hypofluorescence and usually outlined by dilated capillaries unlike hypofluorescence caused by hemorrhages.

iii) Intra-retinal microvascular abnormalities (IRMAs)- are segmental and irregular dilatation of capillary channels lying within CNP areas. As they have immature endothelial tight junctions initially, they may finally slightly leak in later phases at their growing tips.

iv) Venous abnormalities- Such as dilatation, beading, looping & reduplication.

v) Soft exudates cause blockage of choroidal fluorescence like retinal hemorrhages.

FFA characteristics of Severe NPDR are-

MVI 4-2-1 i.e. micros & hges in all 4 quadrants, venous abnormalities in 2 or more quadrants & IRMAs in at least 1 quadrant.

3. Proliferative Diabetic Retinopathy (PDR)

i) Neovascularisation of disc (NVD) or elsewhere (NVE) on retinal surface or elevated into vitreous. These leak the dye profusely which increases in late phase.

ii) Preretinal (Subhyaloid) hemorrhages are well outlined and these block both retinal & choroidal fluorescence.

4. Focal diabetic maculopathy

i) Focal leaks from microaneurysms in macular area.

ii) Hard exudates cause blocked choroidal fluorescence.

5. Diffuse diabetic maculopathy (cystoid)

i) Dilated retinal capillaries are seen leaking diffusely into the macular area.

ii) Typical petalloid or honey comb pattern of CME may be seen in late phases.

iii) Hard exudates typically are not seen.

6. Ischemic diabetic maculopathy (Fig.1)

Foveal avascular zone (FAZ) appears broken i.e. capillary non perfusion (CNP) areas merge into FAZ

7. Mixed variant of Diabetic maculopathy is more common which has any combination of the above 3 variants viz. focal, diffuse & Ischemic.

8. Branch retinal venous occlusion (BRVO) (Fig.2)

FFA advisable only after at least 3 months for the retinal hemorrhages (hges) to absorb sufficiently to allow for proper evaluation.

i) Retinal hges- Block choroidal fluorescence

ii) Delayed filling of involved vein (normally seen in 20-25 seconds of injection) and also delayed emptying (unlike rest of the retinal veins).

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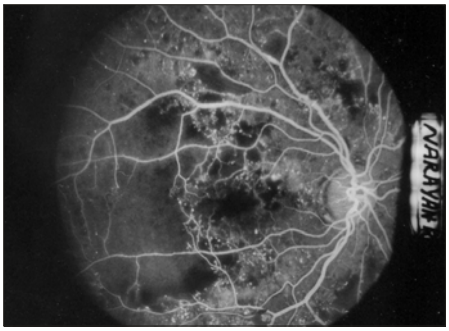

Fig.1: Severe NPDR- with ischemic maculopathy. Extensive CNP areas are seen. Hypofluorescence of non-perfusion areas is outlined by capillaries unlike hypofluorescence because of blocked type. FAZ is grossly broken indicating severe macular ischemia.

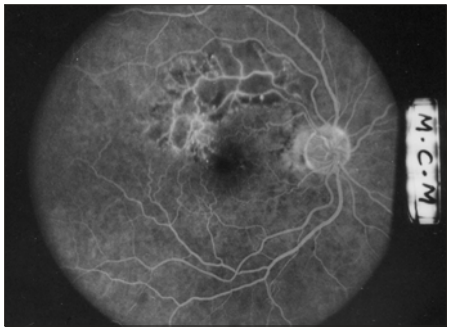

Fig.2: Superotemporal BRVO- Engorged & torturous superotemporal retinal veins with non-perfusion areas. Adjoining FAZ is broken thereby affecting vision greatly.

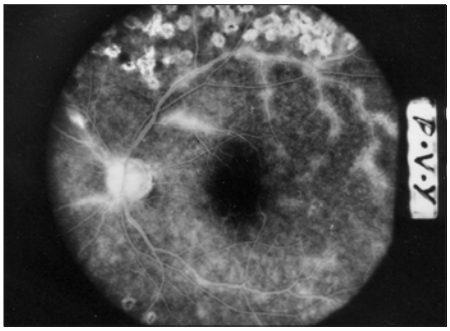

Fig.3: Eales disease- Retinal vasculitis with staining and leak from vessel walls. Laser spots are seen superiorly.

iii) CNP areas (involvement of more than 5 disc diameter areas is critical)

iv) Capillary dilatation and telangiectatic changes.

v) Dye usually stains and leaks from the venous wall in late phases. Macula may show perifoveal capillary leakage (edema) or breakdown (broken FAZ i.e. macular ischemia)

vi) Extensive leakage from NVE may be seen.

9. Non-ischemic Central retinal venous occlusion (CRVO) - FFA advisable only after hemorrhages resolve significantly i.e. after 3-4 months.

i) Delayed central retinal venous filling (normally in 20-25 seconds) and also delayed emptying.

ii) Engorged and tortuous retinal veins

iii) Retinal hges (superficial) block both retinal & choroidal fluorescence.

iv) Dilated and engorged retinal & disc capillaries.

v) Only occasional CNP areas are seen.

vi) Staining and leakage of dye from retinal venous walls.

vii) Macula may show diffuse leaking from perifoveal capillaries (may by cystoid with typically petalloid appearance in late phase)

10. Ischemic CRVO

i) All FFA characteristics as in non-ischemic variant plus there are-

ii) Extensive CNP areas all over the fundus.

iii) CNP areas usually extend into FAZ.

iv) In older cases, extensive leakage from NVD or NVE may be seen.

In Ocular ischemic syndrome (Carotid artery disease) unlike CRVO, veins are usually dilated & irregular in caliber but not tortuous; moreover although NVD is present in 1/3rd cases, disc edema and hemorrhage are not the characteristics. Retinal hemorrhages tend to be in mid periphery

11. Eales' disease (Fig.3)

i) Active vasculitis- Narrowed vascular lumens. Involved inflamed venous walls take staining and may leak dye in late phases.

ii) Picture like that of multiple BRVOs or even CRVO sometimes. Usually peripheral veins involved.

iii) CNP areas in involved segment.

iv) Mid-peripheral NVEs at junction of perfused and non-perfused retina are common. NVEs leak profusely with increasing hyperfluorescence in intensity and size.

12. Hypertensive retinopathy.

FFA picture depends on whether retina, choroid or optic nerve is involved.

i) Focal or generalized arteriolar constrictions or occlusions.

ii) Cotton wool spots are seen as CNP areas (central hypofluorescence bordered by dilated capillaries).

iii) Venous compression is seen at AV crossings.

iv) Choroidal infarcts are seen as Elschnig's spots posteriorly and as Siegrist's tongue shaped streaks peripherally. In acute stage Elschnig's spots appear as hyperfluorescent areas initially which leak dye in later phases. In healed stages, both appear as patches of blocked fluorescence (because of RPE hyper pigmentation) with halo of hyperfluorescence (because of window defect). Lately underlying scleral staining may occur.

v) In disc edema because of malignant hypertension, no looping or corkscrew type tortuosity of peripapillary capillaries and no leak from optic disc vessels is seen unlike that of papilledema.

13. Central Retinal Artery Occlusion (CRAO) (Fig.4)

i) In initial stages, poor choroidal flush (generalized

hypofluorescence) because of blockade by overlying retinal edema and non filling.

ii) Markedly delayed arm retinal circulation time (normally 8-10 sec)

iii) Blood column in arteries is broken (Box-carring)

iv) Poor or no retinal capillary filling.

v) Delayed and incomplete venous return.

vi) After about 2 weeks although dye flow starts in vessels but their calibre remains irregular and capillary filling remains poor.

vii) In combined CRVO & CRAO, filling of arteries by dye is markedly delayed and there are extensive retinal hemorrhages.

viii) 20% population has cilioretinal artery arising out of choroidal circulation which supplies macula. It may fill up in choroidal phase as usual (i.e. spared from CRAO).

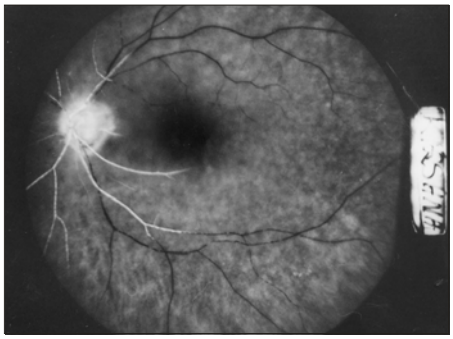

Fig.4: CRAO- Delayed A-R circulation time. Poor filling of retinal vessels. Blood column in arteries is broken (Box-carring).

14. Branch retinal arterial occlusion (BRAO)-

Picture on FFA is similar to CRAO except that it is confined to the affected segment.

15. Exudative vasculopathies-

It includes Coats' disease (multiple telangiectatic and aneurysmal vascular dilatations with massive subretinal exudation and hemorrhages), Leber's miliary aneurysms (localized Coats) and von-Hippel's disease (marked retinal AV malformations with exudation).

i) Telangiectatic and aneurysmal vessels are well made out in early and mid AV phase with leakage from their walls in late phases.

ii) Retinal capillaries show significant dilatation and tortuousities.

iii) Mid-peripheral extensive CNP areas may be seen.

iv) Vascular abnormalities as seen on FFA are much more than that seen on fundus examination.

16. Retinal arterial macroaneurysm-

Occurs in an arteriole before third order bifurcation occurring primarily in old age (usually females) or secondary to other retinal vascular disorders (e.g. Eales', HT, venous occlusions, etc.)

i) On FFA it fills up in early arterial phase as round or fusiform outpouching from arteriole usually at arteriolar bifurcation or AV crossing.

ii) In later phases, its vessel wall may just stain or leak extensively.

iii) Surrounding blocked fluorescence may be seen depending upon site of bleed- subretinal, intraretinal, subhyaloid or into vitreous. Blocked fluorescence may even mask the macroaneurysm.

17. Purtscher's retinopathy-

Results from leucocytic embolization of retinal vessels due to complement activation by head or chest injury, renal failure, pancreatitis or labor.

i) On FFA, CNP areas are seen in involved segments.

ii) Retinal vessels show vascular wall staining and faint leakage.

iii) Hypofluorescent areas also occur due to blocked fluorescence by hemorrhages.

18. Radiation retinopathy-

It most closely mimics the diabetic retinopathy with background, preproliferative or proliferative changes. It can develop months to years after radiation (avg. 2-3 years) with exposure to over 2000 to 3000 rads.

19. Dry ARMD-

i) Hard drusen show as early hyperfluorescent dots increasing in AV phase but fading in later phases.

ii) Soft drusen hyperfluoresce later and may stain in still later phases.

iii) Focal hyperpigmentation and RPE atrophy- Mottled hyperfluorescence i.e. blocked background choroidal fluorescence interspersed with window defects.

iv) Geographic atrophy-

- Early window defects
- Hypofluorescent areas due to choriocapillaris dropouts.
- Border of lesions remain same even in later phases.

Large choroidal vessels and scleral staining are seen in late phases through the atrophic areas.

20. Wet ARMD (Fig.5)

Is characterized by choroidal (subretinal) neovascular membrane (CNVM)

i) Classical CNVM- is well outlined even in early phases as hyperfluorescent patch of lacy pattern with increase in intensity and size in mid and later phases.

ii) Occult CNVM shows 2 patterns on FFA -

Type 1- Fibrovascular PED- well or ill demarcated, mottled hyperfluorescence lesion seen within 1-2 minutes of injection with persistent staining or leakage of dye in late phase.

Type-2- Late leakage of undetermined source-There is

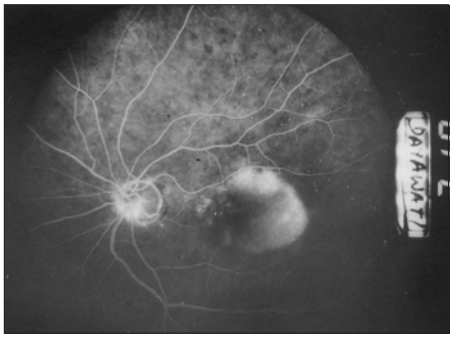

Fig.5: Wet ARMD- Classic & occult CNVM both coexisting with dye slowly filling up neurosensory detachment. Pure classic CNVM in ARMD is uncommon.

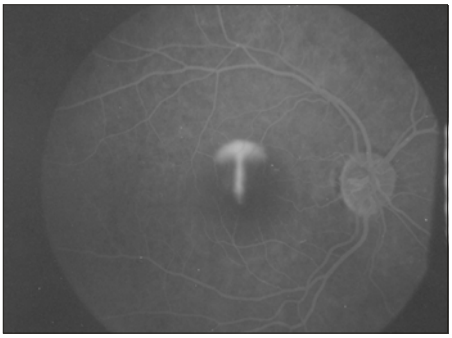

Fig. 6: CSR- Typical smoke stack leak (dye moves up filling neurosensory retinal detachment in an umbrella fashion following osmotic gradient). Patient was 35 years old female (disease much less common in females).

an area of leakage in late phase which doesn't correspond with area of CNVM or fibrovascular PED to account for leakage.

Patients mostly have mixed & variable components of classical and occult CNVM.

21. Pigment epithelial detachment (PED)-

i) seen as well outlined hyperfluorescent patch with gradual increase in intensity in later phases but *no increase in extent of lesion* (same size throughout)

ii) In long standing cases, pigment migration causes irregular blocked fluorescence within PED.

iii) Slow filling, irregularly filling or notching of PED are signs of underlying CNVM as in ARMD,

22. Idiopathic central serous chorioretinopathy (ICSC) or central serous retinopathy (CSR) (Fig.6)

i) Leakage through RPE- hyperfluorescent areas with increase in intensity and size in later phases. Could be pin-head size, ink-blot, smoke-stack (point leak which spreads superiorly to reach the border of neurosensory detachment), single or multiple, or even diffuse.

ii) No leak could mean resolved leak or associated optic pit (pit shows as early hypofluorescent lesion within the optic nerve head with late staining but no leakage of dye).

iii) In resolved cases, window defects at previous site of neurosensory detachment may form tracts in various shapes.

23. Cystoid macula edema (CME)-

i) Early perifoveal capillary dilatation with late pooling of dye in a typical petalloid pattern around fovea (check at 10-15 min. of injecting dye).

ii) Global blood retinal barrier breakdown may give rise to capillary leakage even from surrounding areas and even from disc.

iii) Leakage outside perifoveal area assumes a honeycomb pattern rather than classical petalloid one.

iv) Post surgical CME unlike diabetic CME is always confined to perifoveal region and also has associated late disc staining.

v) Extent of dye leak in post surgical CME correlates poorly with visual acuity.

24. CNVM (non-ARMD)

Causes include idiopathic, pathological myopia, angioid streaks, post inflammatory, hereditary dystrophies, etc.

i) Look for early lacy hyperfluorescence with increase in size & extent in late phases.

ii) Myopic CNVMs are usually small and close to foveal center.

CNVM may form at edge of old Toxoplasmic scar and also next to disc (juxtapapillary).

25. RPE tears (Rip)-

Occurs in PED, CNVM or following laser.

a. Absence of overlying RPE in affected area gives rise to early large area of window defect and staining of choroid & sclera in late phases. Scrolled RPE at opposite edge remains hypofluorescent.

26. Angioid streaks (cracks in Bruch's membrane)-

a. FFA picture varies with status of underlying choriocapillaris and overlying RPE.

b. Typically seen as hyperfluorescent bands extending radially from the disc in arterial phase without any leak in late phases.

c. Associated CNVM or optic nerve head drusen may be present.

27. Polypoidal choroidal vasculopathy (PCV) –

ICG is essential for its diagnosis.

28. Choroidal rupture-

a. Early hypofluorescent band concentric with optic disc because of absent choriocapillaris with late hyperfluorescence because of staining of underlying sclera or residual choroidal tissues in cases of partial rupture.

b. Associated CNVM is common and is seen as hyperfluorescent lesion with increasing leakage and fuzzy margins in late phase. They usually regress spontaneously.

29. Choroidal folds-

a. FFA shows alternating hyper (crest) and hypofluorescent (trough) bands due to thinning of RPE at crests and crowding of RPE at troughs.

30. Macular hole-

FFA picture depends upon underlying RPE status.

a. If underlying RPE is intact, no abnormal FFA finding is noted.

b. Window defect occurs if RPE is atrophied. This hyperfluorescence fades in late phases.

31. Epiretinal membrane (ERM)

a. Distorted perifoveal arcade and exaggerated retinal vascular tortuosity

b. Leakage from distorted capillaries is common.

c. Associated punctate hemorrhages & cystoid macular edema are possible.

d. In secondary ERM, FFA may reveal the cause e.g. associated BRVO changes.

32. Macular infarction-

Abrupt ending of arterioles approaching fovea because of extensive capillary non-perfusion (absent perifoveal capillary network)

33. Parafoveal Telangiectasia (PFT)-

Focal or saccular telangiectasia (thinning and dilatation) of perifoveal capillaries.

a. Group 1A (U/L, congenital PFT) & group 1B (U/L, idiopathic PFT) - microaneurysmal and saccular dilatation of involved capillaries with staining of surrounding retina in later phases.

b. Group 2 (B/L, acquired PFT) - commonest, bilateral symmetric picture. Angiographic findings are same as above. Associated RPE hyperplasia may cause blocked fluorescence. CNVM may also be associated.

c. Group 3 (B/L, idiopathic PFT) - On FFA in addition to above findings, progressive obliteration of perifoveal capillaries is seen.

34. Best's disease (Vitelliform dystrophy)-

a. FFA picture varies with stage of disease. RPE alteration causes irregular transmitted hyperfluorescence. In yellow areas (vitelliform lesion) blocked choroidal fluorescence is seen.

b. In late stages, areas of hyperfluorescence appear within the lesion with staining in late phases. Large choroidal vessels may become visible in atrophic stages.

c. Associated CNVM may occur (early choroidal hyperfluorescence with increasing leakage in later phases)

35. Stargardt's dystrophy (Fundus flavimaculatus)

a. Blocked choroidal fluorescence (because of lipofuscin like material in RPE) in macula and elsewhere with window defects at places within macula. Retinal capillaries may be made out more prominently because of blocked choroidal fluorescence.

b. Atrophic macular lesion shows up as hyperfluorescent late scleral staining.

36. Cone dystrophy-

FFA picture depends on type of fundal lesion. In classical bull's eye maculopathy central ovoid area of blocked fluorescence is surrounded by rim of transmitted hyperfluorescence.

37. Ocular albinism-

a. In absence of overlying RPE, large choroidal vessels are visible in early phases. While late phases show choroidal & retinal vessels silhouetted against scleral staining.

b. Perifoveal capillary network and hence FAZ is ill formed (foveal hypoplasia).

38. Retinitis Pigmentosa (R.P.)

a. In early stages of RP with damaged RPE but intact choriocapillaris, mottled transmitted hyperfluorescence is seen against normal choroidal flush.

b. In late stages of RP, as choriocapillaris also get destroyed (making prognosis poorer), choroidal flush may even be absent in patches or even generally, hence FFA may help to prognosticate the case.

c. Petalloid hyperfluorescence in macula may be seen because of associated CME.

d. Secondary macular degeneration may give use to Bull's eye like changes with central hypofluorescence surrounded by cuff of transmitted hyperfluorescence.

39. Central areolar choroidal dystrophy-

In macular area, there is complete hypofluorescence because of loss of choriocapillaris and underlying choroidal vessels are prominently visible even in early stages of disease.

40. Choroideremia-

Characteristic FFA feature is diffuse absence of choriocapillaris (no choroidal flush) with prominently visible large choroidal vessels. Even scleral staining is not seen in late phase as atrophic choriocapillaris don't leak.

41. Gyrate atrophy-

FFA picture is like Choroideremia except that there are well defined mid peripheral patches of RPE & Choriocapillaris loss.

42. Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)-

a. In acute stage, multiple discrete or confluent patches of hypofluorescence are seen in posterior pole due to blockade of fluorescein by inflammatory cells and also non-perfusion of choriocapillaris.

b. In late phase, lesions usually stain.

c. In healed stage, mottled hyperfluorescence (window defects) is seen.

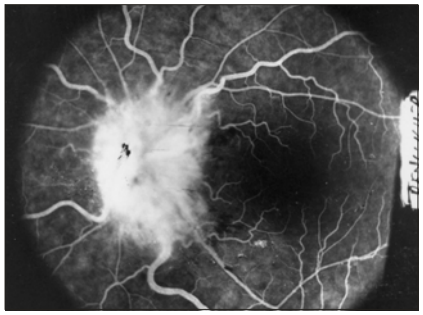

Fig. 7: Papilledema- disc capillaries leaking extensively.

43. Chorioretinitis-

a. In active lesions, FFA shows early hypofluorescence due to blockade by overlying chorioretinal edema and by delayed filling of choroidal vessels. In later phases, hyperfluorescence starts at margins of lesions which then spreads.

b. In healed stages lesions show transmitted fluorescence (RPE atrophy) with visibility of large choroidal vessels. Blocked fluorescence is seen at places due to RPE hyperplasia and hyperfluorescence at margins due to RPE loss and scleral staining. Extensively lasered retina, advanced chorioretinal dystrophies and myopic chorioretinal degeneration also show similar FFA picture.

44. Vogt Koyanagi Harada (VKH) syndrome-

a. Typical FFA picture in acute stage is multiple small hyperfluorescent dots that increase in later phases with extensive pooling of dye in subretinal space (exudative retinal detachment).

b. In healed stages, punctate hyperfluorescent dots are seen but no late pooling. Late staining of disc is common.

45. Intermediate uveitis-

a. Leakage & staining of walls of large peripheral retinal vessels.

b. Perifoveal capillary leakage with typical petalloid appearance in macula indicates associated CME. Leakage is more extensive than in postoperative CME.

46. Optic nerve disorders-

a. Optic nerve head drusen-

i. Autofluorescence in pre-injection (control) photos.

ii. Staining of drusen in late phases.

b. Optic nerve pit-

1. Marked early hypofluorescence in region of pit (absent vessels). Intense hyperfluorescence is seen in and around the pit in late phase due to staining

2. No leakage of dye occurs into associated neurosensory retinal detachment.

c. Papilledema (Fig.7)

FFA role is to diagnose early papilledema or to differentiate it from pseudo- papilledema.

1. Dilatation of disc capillaries with leakage of dye into and around the disc is seen in early phases. Staining of disc is seen in later phases.

2. However normal FFA does not rule out early papilledema.

d. Anterior ischemic optic neuropathy (AION)

Upper & lower halves of disc show differential filling.

e. Optic neuritis and Neuroretinitis have FFA picture somewhat similar to papilledema.

f. Myelinated nerve fibres-

Autofluorescence in control (pre-injection) photos. Fuzzy hyperfluorescence is seen in late phases.

47. Choroidal nevus

i) FFA picture varies with its degree of pigmentation (occasionally amelanotic), location in choroid & secondary RPE changes.

ii) ICG delineates it better

iii) Nevus in choroid with spared choriocapillaris may reveal normal FFA. If choriocapillaris are involved, choroidal hypofluorescence is seen. While hyperfluorescent window defects may occur if overlying RPE has atrophied but choriocapillaries are relatively uninvolved.

48. Choroidal melanoma-

a. Small tumor may show as mottled hyperfluorescence in AV phase and late staining.

b. Large tumor may show double circulation i.e. both normal retinal vessels and tumor vessels fill up together.

c. Late staining of entire tumor mass.

49. Choroidal hemangioma (hamartoma)-

Could be circumscribed or diffuse.

a. In early phase, large choroidal vessels within the hemangioma show the reticular pattern which increasingly leaks the dye causing staining of lesion in late phase.

b. Associated CME (petalloid hyperfluorescence in macula) and overlying RPE atrophy (mottled hyperfluorescence) may occur.

50. Optic disc melanocytoma-

Benign pigmented disc tumor which causes blocked disc fluorescence throughout the angiogram.

Duane's Retraction Syndrome

Gaurav Kakkar, DOMS, MS**, Sunita Lulla,** MS**, Kanak Tyagi, Abhishek Dagar,** MS

Prior to publication of Alexander Duanes description in 1905 this syndrome had been described by Stilling in 1887 and Turk in 1896 .Hence in European literature the retraction syndrome is appropriately referred to as the Stilling-Turk-Duane syndrome after it's early describers. Duane emphasized that the retraction of the retraction of the globe is an essential clinical feature of this syndrome., because of this feature which is so diagnostic the term "Duane's Retraction Syndrome" is deeply entrenched.

Classification

Of the various classifications proposed by various authors Huber classification remains the most popular because he used the clinical descriptions together with electromyographic information to simplify this complex syndrome.

Type I: It is the most common type (78%) characterized by marked limitation of abduction with minimally defective or normal adduction, retraction of globe and narrowing of the palpebral fissure in adduction, widening of the fissure on abduction Electromyography shows absence of electrical activity in the lateral rectus muscle on abduction but paradoxical electrical activity on adduction (fig 1 & 2)

Type II: It is the least common type(<10%) characterized by marked limitation of adduction with exotropia of the affected eye, abduction normal or slightly limited, retraction of the globe and narrowing of the fissure on attempted adduction . On electromyography the lateral rectus showed peak impulses on abduction and a second paradoxical peak on attempted adduction. There was normal behavior of the medial rectus.

Type III: It is present in 15% characterized by a combined limitation or absence of both abduction and adduction, retraction of the globe and narrowing of the palpebral fissure on attempted adduction. The electromyogram demonstrates cocontraction of the horizontal rectus muscles on both adduction and abduction.

The most characteristic clinical presentation of Duane syndrome is an absence of abduction of an eye with some degree of restricted adduction and retraction when an attempt is made to adduct .The retraction is variable: it is conspicuous in some but minimal in others. Additionally, either an upshooting or downshooting, or both, of the adducted eye frequently occurs, particularly as the adducting eye begins to move in the oblique position of up and in or down and in. This overshoot simulates overaction of the inferior and superior oblique muscles. Occasionally, the upshoot or down-shoot is so marked that the cornea is driven completely out of the palpebral fissure, hiding behind the upper or lower lid. Some patients manifest only the upshoot and a few only the downshoot, but most patients have various degrees of both vertical abnormalities.

Pathogenesis

The pathogenesis of DRS is a spectrum of mechanical, anatomical and innervational disorders that has been classified into a single clinical entity.

There is evidence of partial or absent development of the sixth nerve nucleus and /or nerve. Branches from the third nerve are directed to innervate the lateral rectus muscle. This abnormal firing pattern of the lateral rectus muscle results in limitation of abduction and adduction. The cocontraction of the horizontal rectus muscles on adduction causes the retraction of the globe and also may cause slippage of the globe causing upshoot /downshoot depending on the relative position of the globe in reference to the muscles. Further secondary anatomic changes (fibrosis) may occur in the abnormally innervated muscles.

Features

A. Incidence

The frequency of Duane's syndrome in the general population of strabismus patients has been estimated to be 1-4%..

B. Gender Distribution

The most logical way to explain the frequency of the syndrome among females was to suggest that the gene responsible, besides being autosomal dominant with incomplete penetrance, was also partly sex-limited, making females more susceptible to the effects of the gene.

C. Laterality

The predilection for left eye involvement has been cited in practically all the studies of DRS

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Venu Eye institute, New Delhi

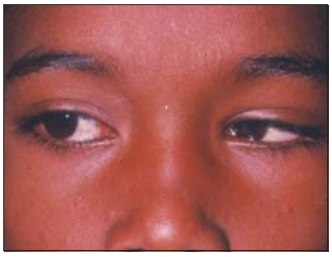

Fig. 1: Type 1 DRS

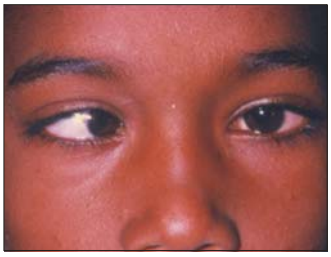

Fig. 2: Type 1 Limitation of abduction

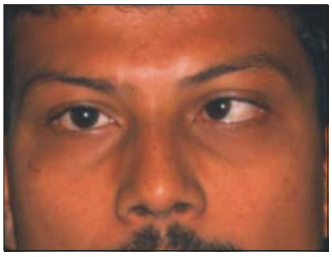

Fig. 3: Esotropia

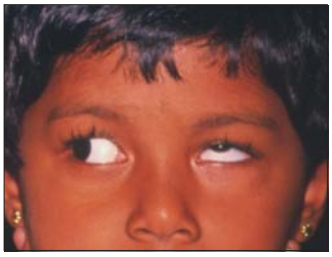

Fig. 4: Upshoot

D. Types of Presentation

The most common form of the syndrome is Type I of the Huber classification system.

E. Ocular Deviation in Primary Position

Most patients with DRS appear to have strabismus in primary position. Patients may adopt small, relatively unnoticed head positions which may obscure a small angle strabismus in primary gaze.. Esotropia was the most common presenting deviation in primary gaze in the majority or reviewed studies and it was more common in Duane's Type I. Orthotropia was the second most common finding in primary position

F. Refractive Errors

Reviews of DRS patients have shown hypermetropia of greater than +1.50 in 71% of the patients

G. Amblyopia

Amblyopia in DRS has been found to be more due to strabismus and not due to anisometropia. The range of Duane's syndrome patients with amblyopia ranged from 3% to 25%, with the weighted average being 14% among studies

Associated Congenital Anomalies and Syndromes

1. Associate Ocular Findings

The most frequently encountered ocular abnormality are nystagmus epibulbar dermoid, anisocoria and ptosis .Many of the epibulbar dermoids were associated with Goldenhar's syndrome.

2. Associated Nonocular Findings

Goldenhar's syndrome (oculo-auriculo-vertebral dysplasia):It is due to an abnormal morphogenesis of the first and second branchial arches and is classically accompanied by vertebral anomalies, epibulbar dermoids, facial hypoplasia and preauricular skin-tags. (fig 6)

Klippel-Fiel anomaly: It consists of a malformation of cervical vertebrae with webneck torticollis and facial asymmetry

Wildervanck syndrome (cervico-oculo-acoustic syndrome) :It consists of klippel –fiel anomaly and congenital sensorineural deafness.

The combined abnormalities of the vertebral column and limbs, otologic defects and other ocular malformations with Duanes syndrome are called "Duanes syndrome plus".

All structures most frequently altered in association with Duanes syndrome develop during the second month of embryonic life. A common teratogenic factor may be the cause the syndrome during this period.

Variants of Duane's Syndrome

a) Vertical Retraction Syndrome:

It is extremely rare associated with limitation of the affected eye on elevation/depression associated with globe retraction and narrowing of the palprebral fissure.

b) Congenital Adduction Deficit with synergistic divergence: It presents as a unilateral adduction deficit and simultaneous abduction of the eye on attempted lateral gaze into the field of action of the apparently paretic medial rectus muscle.

Clinical Features and Diagnosis

1. Defects in Abduction and Adduction

The most characteristic findings is an absence of abduction of an eye with some degree of restricted adduction. The adduction deficit is best demonstrated by attempting to measure the near point of convergence. All DRS patients seem to have a remote or recessed near point of convergence.

2. Esotropia and Exotropia

Although many patients with DRS are orthophoric in primary position, exotropia in straight ahead position also occurs. Esotropia, however, is the most commonly diagnosed type of strabismus. This reflects the increased frequency of DRS Type I, in which esotropia would be expected to be most common. The head gaze must be stabilized in forced straight ahead position in order to measure the strabismus in primary position. (fig 3 & 9)

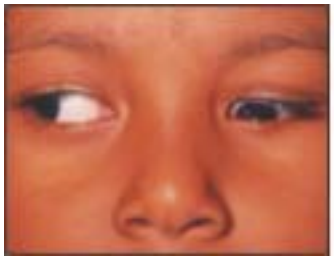

Fig. 5: Downshoot

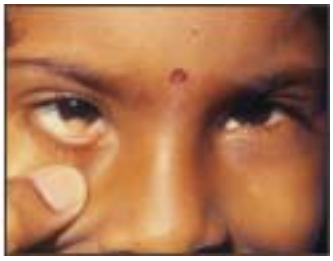

Fig. 6: Epibulbar Dermoid

Fig. 7: Face Turn Pre-op

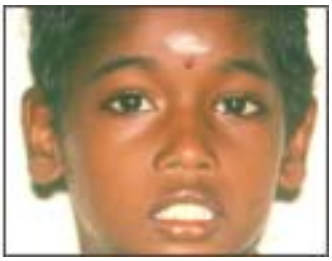

Fig. 8: Post-op

3. *Upshoots and Downshoots*

The characteristic vertical deviations seen in DRS include the over elevation and depression seen in the adducted position. In contrast to typical over action of the superior oblique and inferior oblique muscles, these ductions are often exaggerated and, as von Noorden points out, are of almost grotesque cosmetic proportion, often causing the cornea to disappear from view . The associated lid changes and retraction of the globe help to distinguish these vertical movements from true oblique muscle over action. The eye in DRS does not usually upshoot or downshoot when the eye is adducted in the horizontal position. It occurs when the eye begins to move up or down in the adducted position, and then a "flipping" up or down movement is noted. This abnormal movement had been postulated to be related to a slipping of the lateral rectus muscle over the globe, "the bridle effect" (fig 4 & 5)

4. *A and V Patterns*

Various types of A and V patterns may be found in DRS. The V pattern is more common, but a pattern also occurs. These most frequently manifest as an exotropia in up or down gaze. Some patients will have an X pattern if they have both an upshot and downshoot. Interestingly, patients with bilateral DRS tend to have a A pattern more frequently then V.

5. *Eyelid Changes and Globe Retraction*

The eyelid changes found in DRS in adduction are related to a dropping of the upper lid and elevation of the lower lid.these eye lid changes are more than just passive narrowing from the retraction of the globe which results from cocontraction of the horizontal rectus muscles. Both lid narrowing and globe retraction are often the most pronounced components of DRS. (fig 10)

6. *Torticollis*

Face turn is a major characteristic of DRS. It is a mechanism of maintaining single binocular vision. The face is turned in the direction of the horizontal muscle with the greatest deficit.

Differential Diagnosis

1. Abducens Nerve Palsy (6TH Nerve)-there is a definite onset of new large angle esotropia with diplopia .globe retraction, vertical upshoots/downshoots are not present
2. Moebius Syndrome.-Complete paralysis of bilateral 6 th cranial nerves associated with loss of olfactory and gustatory senses.
3. Congenital or Infantile Esotropia-child may cross fixate and on patching the abduction is full.

Therapeutic Modalities

Goals of Treatment

The cosmetic problems of the affected patient are mainly ocular retraction and narrowing of the palprebral fissure on adduction, torticollis horizontal strabismus anomalous vertical movement and limited abduction.

With the exception of limited abduction other problems may be eliminated totally or partially by means of adequate recession of horizontal rectus muscles.

The most common indication for surgical treatment is an unacceptable face turn. (Fig 7 & 8) The face turn is a secondary manifestation of strabismus in primary position and develops to permit fusion. If it is sufficiently large, the face turn may be a functional handicap in addition to being disfiguring. Patients who have Duane syndrome with exotropia in primary position usually have a face turn away from the affected eye. More commonly, an esodeviation in primary position leads to a face turn toward the side of the affected eye. This face turn usually is more pronounced with distant fixation

Occasionally, fusion is impossible in a patient with bilateral Duane syndrome usually because bilateral

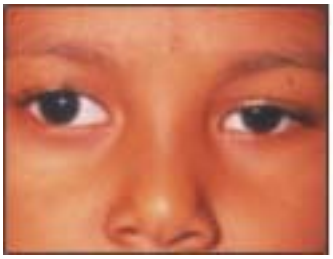

Fig. 9: Exotropia

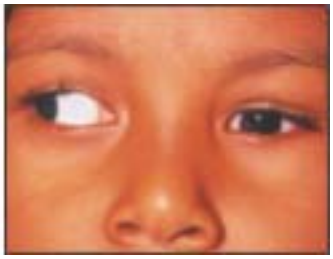

Fig. 10: Globe Retraction

involvement or a vertical deviation precludes fusion with any head posture. In these cases, the strabismus itself rather than the secondary head posture can be the main indication for surgical correction.

When the affected eye is adducted, an upshoot, downshoot, or retraction can be sufficiently disturbing to the patient or the parents to warrant surgical treatment

A reduction of 50% or more of the width of the palpebral fissure during adduction compared with primary position has been suggested as an indication for surgical treatment of the retraction

Contraindications

Many patients with Duane syndrome are orthophoric in primary position or have only an insignificant face turn. Because fusion usually can be obtained by means of a face turn, most children with Duane syndrome have normal binocular function and stereopsis. Therefore, in contradistinction to congenital esotropia, where the goal is to restore ocular alignment at as early an age as possible, the goal in Duane syndrome should be to avoid disrupting normal binocular development. Thus, young age is a relative contraindication for surgery. Severe cases may warrant early treatment, but usually, it is preferable to delay surgery until the patient is at least 4 to 5 years old. At this age, patient cooperation facilitates a detailed examination, and the visual system is relatively mature and less susceptible to damage from a temporary disruption of binocularity, as can occur postoperatively if there is an unfavorable response to surgery.

Surgical Procedures

Horizontal Muscle Surgery

Medial Rectus Muscle Recession

Recession of the ipsilateral medial rectus muscle is the mainstay of surgical treatment for Duane syndrome. In patients with esotropia in primary position, this procedure improves the face turn and the esotropia by weakening the antagonist of an effectively paretic lateral rectus muscle. Medial rectus recession alone also may improve the enophthalmos and vertical overshoots in adduction, in part by limiting adduction of the eye.

Recession of the contralateral in addition to the ipsilateral medial rectus muscle may be performed in cases in which the patient has primary position esotropia greater than 20 Ä and marked co-contraction of the lateral rectus muscle as evidenced by limited adduction, massive retraction.

Lateral Rectus Muscle Recession

An exodeviation in primary position with a face turn away from the side of the affected eye is a less common presentation of Duane syndrome. In these cases, the face turn is treated with recession of the ipsilateral lateral rectus muscle. In patients who have both primary position exotropia and a marked upshoot or downshoot, a lateral rectus recession usually is combined with an additional measure to minimize sideslip of the lateral rectus muscle across the globe: either a Y-splitting procedure or a posterior fixation suture

Large recessions (10 to 12 mm) of both the medial and lateral rectus muscles of the ipsilateral eye are more effective, particularly when enophthalmos in primary position is a major complaint.

Transposition Procedures

Transposition of the vertical rectus muscles to a position adjacent to the lateral rectus, with or without recession of the ipsilateral medial rectus, has been suggested as a means of correcting the primary position esotropia, improving abduction, and enlarging the field of single binocular vision This procedure may provide better abduction than medial rectus recession alone. However, it is more difficult to perform; may exacerbate retraction upshoot, or downshoot; can create new vertical deviations,and particularly in adult patients on whom a medial rectus recession is performed concurrently, runs some risk of anterior segment ischemia. This procedure probably should only be considered as primary treatment for patients with no abduction at all, minimal retraction, and no upshoot or downshoot.

Y-Splitting Procedure

Co-contraction of the medial and lateral rectus muscles on attempted adduction can cause a striking upshoot or downshoot of the eye. This effect is attributed to sideslip of a tight lateral rectus over the globe (mechanical factors) in most cases. The Y-splitting procedure effectively results in a broad lateral rectus insertion that stabilizes its position and prevents it from flipping superiorly or inferiorly over the globe, thus eliminating or greatly reducing the upshoot or downshoot of the affected eye in attempted adduction. The Y-splitting procedure may be combined with a moderate recession of the lateral rectus muscle, particularly if there is associated primary position exotropia.

Posterior Fixation Sutures (Faden)

A posterior fixation suture on the lateral rectus muscle can effectively prevent slippage of the muscle belly over the globe. It may be used as an alternative procedure to treat upshoots and downshoots. As with the Y-splitting procedure, a posterior fixation suture can be combined with a lateral rectus recession when appropriate.

Vertical Rectus muscle Weakening Procedures

Besides the mechanical factors discussed above, innervational factors, presumably aberrant co-contraction of the vertical rectus or inferior oblique muscles, may contribute to an upshoot or downshoot in some patients with Duane syndrome When there is a significant vertical deviation in primary position, horizontal rectus muscle surgery alone generally does not correct the problem adequately, and recession of the appropriate vertical rectus muscle is needed

Surgical Complications

The possible complication of anterior segment ischemia due to the vertical rectus muscle transposition with a recession of the medial rectus of the affected eye has been mentioned. If necessary, the medial rectus recession can be performed as a secondary procedure four months following the transposition.

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Post-Operative Endophthalmitis: Few Practical Tips for its Management

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Endophthalmitis is defined as an inflammation of the inner coats of the eye associated with exudates in the vitreous, which may be infectious (bacterial or fungal) or non-infectious in origin. It is an uncommon entity in clinical ophthalmology but has the potential to cause severe visual loss. Although the incidence of intraocular infections after cataract surgery has sharply declined over the past 3-4 decades since the advent of aseptic techniques and the use of prophylactic antibiotics, endophthalmitis still remains one of the most dreaded complications that an ophthalmic surgeon has to face. Sixty to seventy per cent of cases of endophthalmitis occur after cataract surgery, with an overall incidence of 0.02-0.75% in most large series of operated cataract cases. In the present article we report the practical approach to manage regarding the diagnosis and treatment of postoperative endophthalmitis.

Source of infection

The most common source of infection is probably the patient himself, as it is almost impossible to sterilize the operative field completely[1]. However, there are many other sources which may play a role in the occurrence of endophthalmitis, i.e. the surgeon's hands, airborne pathogens (either from surgeon's nose or the air conditioner), solutions injected into the eye, instruments, antiseptics used preoperatively, and drops & ointments used post-operatively. The most frequent pathogens causing acute bacterial endophthalmitis are gram positive cocci i.e. *Staphylococcus albus* and *Staphylococcus aureus*, followed by the gram-negative organisms, especially *Pseudomonas aerugenosa*, streptococcus and fungi. In recent years, attention has been directed to late onset endophthalmitis caused by *Propionobacterium acnes*.

Clinical features

There are two clinical settings in which postoperative endophthalmitis may be seen. These are *(a) 1st postoperative day:* This is the less common presentation of the disease. Endophthalmitis that presents at this time is severe in intensity and usually associated with increased pain, redness, watering and lid edema. The clinical sign of most utility in suspecting a diagnosis at this time are an increased number of cells in the anterior chamber and the presence of vitreous exudates. A hypopyon may or may not be present at this time. A decreased fundus glow on the first post operative day can be due to many other reasons and is not very specific for the diagnosis of post-operative endophthalmitis. Similarly decreased visual acuity at this time is not of much relevance in making the diagnosis since it can be due to many other causes. *(b) Onset during the postoperative period (usually within 6 weeks of the surgery).* This is the more common mode of presentation. Most of these patients give a definite history of occurrence of sudden deterioration of vision between the second post operative day and the sixth week after initial recovery. The decrease in vision is associated with pain (in three-fourth of patients), redness, watering and clinical signs in the form of increase in anterior chamber reaction with or without a hypopyon and vitreous exudates. Deterioration of visual acuity in a patient after initial recovery is one of the most consistent symptoms of postoperative endoph-thalmitis. The presence of a decreased fundus glow on distant direct ophthalmoscopy at this time is a very important sign of endophthalmitis.

In the situations described above, the cornea may be hazy, and epithelial edema may be present. Inspection of the area of incision may sometimes reveal an area of infiltration, usually along the suture track. If present, this is a bad prognostic sign[2].

If the process manifests soon after the surgery with a fulminating course, *Pseudomonas* infection is a likely cause. However, other gram-negative organisms and *Staphylococcus aureus* could also induce a similar response.

The most definitive clinical signs of endophthalmitis are presence of vitreous exudates along with a hypopyon or significant number of cells in the anterior chamber.

Investigations and Management

Every patient suspected of having postoperative endophthalmitis must be examined thoroughly after taking a brief history. Complete slit-lamp examination should be done to note the findings of the incision site,

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cornea, anterior chamber and anterior vitreous, if visible. Visual acuity should be carefully assessed since it is an important prognostic indicator and is also one of the parameters used for decision making. Ultrasonography should be ordered to assess the posterior segment status. The objective of ultrasonography is to evaluate the eye for the presence and location of posterior vitreous detachment and for the presence of a retinal detachment.

Topical concentrated antibiotics **(Table 1)** should be started immediately along with a cycloplegic-mydriatic (i.e. Homatropine 2% eye drops 2 hourly). As a rule, we prefer to start atleast two topical antibiotics (for gram positive and gram negative organisms respectively) simultaneously. Anti-glaucoma medication is added if required. Systemic steroids (Tab Prednisolone in a dose of 1mg/kg /day after breakfast) are added to reduce the inflammation if the probable cause suspected is not fungal.

Intravitreal injection of antibiotics

The mainstay of treatment for postoperative endophthalmitis is intravitreal injection of antibiotics[3-9]. Usually a combination of two antibiotics is chosen, which are selected based on their activity against coagulase-negative staphylococci (the most common bacterial cause of endophthalmitis), and gram-negative bacilli. The most commonly used and effective combination for this purpose at present is Vancomycin (1mg/0.1ml) and ceftazidime (2.2mg in 0.1ml). ***While amikacin (400 µg/0.1 ml) could also be used in place of ceftazidime, it possesses a higher risk of macular toxicity with the higher as well as the recommended usual doses and should be avoided, if possible*[10-12].** Some people favor intravitreal injection of steroid (Dexamethasone 400µg/0.1ml) to limit post inflammatory consequences. However, we are not at present administering intravitreal steroids routinely to our patients. Great care must be exercised in the preparation of antibiotics for intravitreal injection since they are highly toxic in higher than recommended doses.

Technique

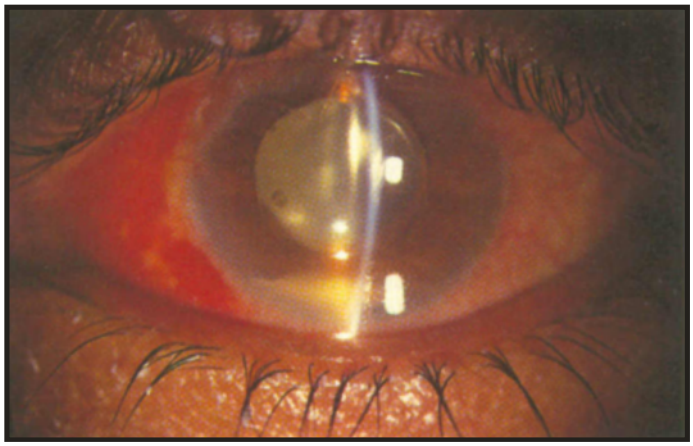
Intravitreal injection of antibiotics should be given in the operation theatre under proper aseptic precautions. It is usually performed under topical anesthesia. However, retrobulbar anesthesia could be administered if the patient is uncooperative. Facial anesthesia prior to the intravitreal procedure simplifies the procedure for the surgeon and improves the patient's comfort significantly and is recommended for all, except the most cooperative patients. The most common site for administering intravitreal injection is inferotemporally 3 mm from the limbus in aphakic and 3.5mm in pseudophakic eyes. Initially around 0.5 cc vitreous should be aspirated with a 21 guage needle for making smears as well as for culture sensitivity; then antibiotics is injected sequentially into the vitreous cavity, with a 26 guage needle. In case of a dry tap, aqueous paracentesis could be done to obtain material for smear and culture sensitivity and even more importantly, for bringing down the IOP prior to the intravitreal injection. It is not necessary to use two different needles for injecting the two antibiotics though different syringes are mandatory. The needle should be kept facing towards the centre of the globe. The bevel of the needle should be dented upwards towards the cornea. The antibiotics should be taken in two separate glass tuberculin syringes. Disposable tuberculin syringes should be avoided. This is so because it is possible to inject the antibiotic slowly drop by drop with a glass tuberculin syringe by rotating the plunger while pushing it forward. This motion is not possible with a disposable syringe. Thus, a jet of fluid is more likely to be injected with a disposable syringe[9].

The EVS recommended that systemic antibiotics have no added role in patients being treated by intravitreal injections or vitrectomy[4,8]. As a rule, however, we do administer Tab Ciprofloxacin 500 mg to 750 mg BD to patients of endophthalmitis. The rationale for this decision

Table 1: Concentration and dosage of principal antibiotics used in endophthalmitis

Drug Name	Topical (%)	Intavitreal	Systemic
- Cefazolin Sodium	50 mg/ml (5)	2.25 mg	25-50mg/kg/day QID IV
- Amikacin Sulfate	20 mg/ml (2)	0.4 mg	15mg/kg/day TDS IV
- Vancomycin Hydrochloride	50 mg/ml (5)	1 mg	15-30 mg/kg/day OD-BD IV
- Ceftazidime	—	2.25 mg	1-2 gm/day BD-TDS IV
- Gentamycin Sulfate	13.5 mg/ml (1.35)	0.1-0.2 mg	3-5 mg/kg/day BD-TDS IV
- Amphotericin B	1.5 mg/ml (0.15)	5 mgm	0.4-0.6 mg/kg/day with 5% dextrose over 2-4 hours
- Dexamethasone	—	0.36 mg	6mg/kg/day
- Prednisolone	—	—	1 mg/kg/day Oral
- Tobramycin	13.5 mg/ml (1.35)	—	3-5 mg/kg/day BD-TDS IV

is that oral ciprofloxacin does have a significant penetration into the vitreous cavity as opposed to the systemically administered drugs tested during the EVS in most of their patients.



Vitrectomy in Endophthalmitis

As a rule, all patients seen at our Centre are given an initial intravitreal antibiotic injection on presentation. Vitrectomy is reserved for patients who do not respond adequately to intravitreal antibiotics within 36-48 hours. While we do prefer to carry out an immediate vitrectomy for patients who present with a visual acuity of light perception only, this is often not possible due to logistic constraints. We have shown in a study carried out at our centre that the results of immediate intravitreal antibiotic injection followed by vitrectomy within 48 hours are as good as those of immediate vitrectomy in patients with post operative endophthalmitis. If however, the patient has already received an intravitreal antibiotic injection elsewhere, it is advisable to carry out an immediate vitrectomy at presentation. For all cataract surgeons faced with a patient with endophthalmitis, our advice would be:

Give an intravitreal antibiotic injection immediately. Assess the response after 24 hours. If there is no definitive evidence of improvement, please refer the patient immediately to a vitreo-retinal surgeon. We are not in favour of multiple intravitreal antibiotic injections except in patients with an extremely poor prognosis i.e. those with corneal infiltration or those with a cornea too hazy to perform a successful vitrectomy or those with a retinal detachment on ultrasonography.

Tips for Vitrectomy in Endophthalmitis[9,13]

Today, there is no doubt that the mainstay of management for post-operative endophthalmitis is '***Intravitreal antibiotics***'. However, there are certain situations, in which vitrectomy may result, not only in salvaging the eye, but also in providing a favorable outcome in patients with this devastating disease process. Some of the practical tips regarding successful use of this very effective tool in the ophthalmologist's armamentarium are given below:

Patient Selection

- Inaccurate projection of light is not a contraindication for vitrectomy. In fact, it probably has almost no prognostic role at all.
- Patients with significant corneal infiltration are likely to fare poorly, and may be better managed conservatively.
- Patients who have not responded to their first intravitreal injection of antibiotics given 48 hours earlier are candidates for an immediate vitrectomy. Multiple intravitreal antibiotic injections should be avoided as the means of management for endophthalmitis except in cases which are not fit for vitrectomy.
- Patients with a retinal detachment on USG have a poor prognosis following vitrectomy.

Investigations

- Preoperative ultrasonography (USG) is mandatory prior to vitrectomy for endophthalmitis.
- USG is required to look for presence of a partial/total posterior vitreous detachment and also for the presence of a retinal detachment.
- Patients with dense exudates extending up to the posterior one third of the vitreous cavity on ultrasonography have a poorer visual prognosis.
- Surgery is easier in patients with a partial or total posterior vitreous detachment.
- The decision regarding when to undertake vitrectomy is however not based on ultrasonography. It is based on clinical factors.
- Electrophysiological tests like VER and ERG have no role in evaluating patients for vitrectomy for endophthalmitis.

Surgical Technique

- Surgery under general anesthesia is preferable for uncooperative patients. As a rule, however, peribulbar anesthesia is adequate for surgery in the majority of the patients.
- The corneal incision must be inspected and strengthened by placing additional sutures before starting vitrectomy.
- 3 port pars plana vitrectomy is carried out in all patients.
- A 6 mm infusion cannula must be used.
- This 6 mm cannula is always possible to visualize within the vitreous cavity by depressing the cannula

inwards and forwards towards the pupil (since the patient is aphakic/pseudophakic, there is no risk of lens damage).

- The infusion bottle must not be kept higher than 24 inches from the patients' eye.
- In patients with pseudophakic endophthalmitis, there is almost always a fibrin membrane covering the iris and the pupillary area. This can be removed form the pars plana route by making an iridectomy next to the temporal port with the vitrectomy cutter. An MVR blade is then passed through the iridectomy to engage the fibrin membrane and dislodge it from its adhesions to the iris and the IOL surface[14]. The membrane is then eaten away in the anterior chamber by introducing the vitrectomy probe through the iridectomy. Adequate visualization is possible in all cases by this technique.
- A high cutting rate (> 600 cuts per minute) and low suction (<100 mmHg) must be kept for the vitrectomy cutter. This will ensure that no undue traction is put on the vitreous gel during the vitrectomy.
- In over half the cases, vitreous exudates are not found to extend beyond the anterior or mid vitreous during vitrectomy.
- Only a core vitrectomy is required to ensure a successful outcome in the majority of the cases.
- Vitreous cavity lavage can be carried out by continuing to use the vitreous cutter (in the cutting mode) placed in the central vitreous cavity for at least (5-10 minutes after the infected gel has been removed. This makes it possible to carry out the lavage without causing any turbulence within the vitreous cavity.
- Corneal epithelium debridement can be carried out at any time during the vitrectomy if epithelial edema is preventing adequate visualization, without any risk of causing a corneal ulcer.
- After closure of the sclerotomies, check the corneal wound once more since, the corneal sutures could become loose due to the distortions created.

Today, the results of vitrectomy for endophthalmitis have improved considerably. Approximately 80% of patients can expect a visual acuity better than 6/60 following vitrectomy for endophthalmitis. The technique thus offers a glimmer of hope to the afflicted patients.

Risk factors for poor results

Earlier studies have reported that a positive culture, a more virulent organism, delay in onset of initiation of treatment, presence of concomitant ocular disease such as retinal detachment and rubeosis, and a poor initial visual acuity are the risk factors for worse visual acuity results. The EVS findings showed many similar as well as independent risk factors, i.e., older age, history of diabetes, corneal infiltrate or ring ulcer, abnormal intraocular pressure, rubeosis, absent red reflex, a ruptured posterior capsule, and visual acuity of light perception as predictors of a poor visual outcome[4,8]. However, the most important risk factor for decreased final visual acuity was an initial visual acuity of light perception only. Such patients had twice the risk for a worse acuity outcome compared with patients with better than light perception only. It is our experience that the most important prognostic factor in patient's undergoing treatment for endoph-thalmitis is the presence of significant corneal infiltration. Such patients do extremely poorly even after vitrectomy.

It is our clinical experience that the worst prognosis occurs in patients with corneal infiltration; in these patients vision is usually unlikely to be salvaged. Today however with appropriate management, it is possible not only to salvage many of the eyes with this devastating condition, it is possible to help them retain useful vision also.

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23-Gauge Sutureless Vitrectomy

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Historical Background

Since the introduction of pars plana vitrectomy in the early 1970s by Machemer[1], advances in the field of vitreoretinal surgery have been dramatic. Machemer initially performed pars plana vitrectomy with the use of a 17-gauge (1.5mm diameter) multifunctional instrument capable of cutting and aspirating the vitreous. This instrument utilized a fiberoptic sleeve and required a 2.3mm scleral incision. In 1974, O,Malley designed a smaller vitreous cutter with a diameter of 0.9mm (20-gauge).[2] This less invasive 3-port 20-gauge cannula-entry system is still used today. In 1990, de Juan and colleagues designed a variety of 25-gauge (0.5mm diameter) vitreoretinal instruments for more delicate and precise surgical maneuvers.[3] Recently, Fuji et al designed a 25-gauge microcannular system and an array of 25-gauge instruments referred to as transconjunctival sutureless vitrectomy system (TSV).[4]

Introduction

23-gauge sutureless vitrectomy is adapted from the transconjunctival sutureless vitrectomy system (Fujii et al).[5] One of the most frequent objections to the 25-gauge TSV instruments has been that they are too flexible for many of the complicated tasks performed on the retina and vitreous body.

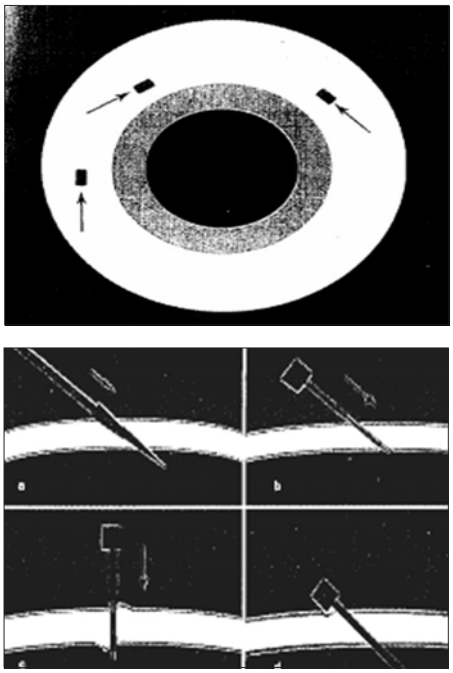

Figure: 1 & 2

Principle

As in transconjunctival sutureless 25-gauge vitrectomy, 3 microcannulas for the instruments and infusion line are inserted transconjuctivally into the area of the planned sclerotomy. The incisions are not made perpendicular to the scleral surface (i.e. towards the posterior pole) but at a 30-40 degrees angle parallel to the corneoscleral limbus (Figure 1 and 2). The tunnel-like nature of these incisions facilitates the self-sealing of the wound after removal of the cannulas.

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Technique

The procedure is started by pushing the conjunctiva 1-2m laterally (i.e. parallel to the coneoscleral limbus) in the inferotemporal, superotemporal, and superonasal quadrants using a special pressure plate (DORC, Zuidland,Holland) with a central opening at 3.5mm to hold it firmly to the sclera. (figure 3) A 23-gauge stiletto blade, 45 degree angled, 0.72mm width is then inserted at a 30-40 degree angle through the conjunctiva, sclera and pars plana 3.5mm from the corneoscleral limbus. (Figure 4). To obtain scleral tunnels parallel to the corneoscleral limbus, the scleral incisions are made radial to the coneoscleral limbus. Constant pressure is applied to the pressure plate while the incision is made and during withdrawal of the stiletto blade to prevent slippage of the conjunctiva against the sclera.

The microcannula is then inserted through the conjunctival incision and into the scleral tunnel using a specially designed blunt inserter, both these are made of steel. The length of cannula (without its head) is 4mm, the internal diameter of the cannula is 0.65mm and the external diameter is 0.75mm. The inserter is not a beveled trocar but a blunt instrument whose spatula-like tip merges with a cylindrical body holding the microcannula. The external openings of two of the three microcannulas are funnel shaped to facilitate insertion of the instruments.

The instruments used for transconjunctival sutureless 23-gauge vitrectomies are a Pneumatic vitreous cutter, Electromagnetic vitreous cutter, wide-angle endoillumination probe, flute needle, endodiathermy probe, endolaser probe, endgripping forceps, vertical and curved scissors, Hem-stopper, RON-knife and illuminated pic. A cutting rate of upto 1200 per minute and suction upto 500mm Hg are used.

At the time of removal of the microcannulas, the IOP should be approximately 20 mmHG. The cannulas are closed with plugs, and removed with a special forceps. Slight pressure is applied to the conjunctiva and sclera with a cotton wool applicator to prevent subconjunctival hemorrhage and any wound leak observed.

Advantages over 20-gauge vitrectomy

1. Minimal trauma of the conjunctiva and sclera. No postoperative scleral thinning in the area of the sclerotomy. High postoperative stability of the sclerotomies.
2. Less postoperative astigmatism.
3. Less postoperative discomfort.

Advantages over 25-gauge vitrectomy

1. Instruments are less flexible and more effective.
2. Shorter operative time.
3. Brighter endoillumination.
4. Better handling of an acute intraoperative hemorrgae due to the more effective use of the fute needle by its larger inner diameter.
5. Longer durability of the instruments.
6. Lesser learning curve.

Fig. 3: Eckradt Pressure Plate

Fig. 4: Eckradt 23G Cannula System

In summary, the sutureless 23-gauge vitrectomy procedure appears to be a viable alternative to 25-gauge vitrectomy. It offers all of the advantages of the minimally invasive transconjunctival vitrectomy system developed by Fujii et al, plus the benefits of a sturdier and larger instrumentarium.

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Principle & Application of Keratometry in Clinical Practice

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Introduction

The development of keratometry or the ophthalmometer has been credited to Helmholtz in 1854, who modified an image doubling and magnifying technique deviced by Ramsdon in 1796. In 1881, Javal & Schiotz modified Helmholtz's instrument for clinical use for measurement of radius of cornea's curvature.

Optical principle of keratometry

The principle of the keratometer is based on the geometry of aspherical reflecting surface. Cornea acts as a convex mirror. An object of known size and distance is reflected off the corneal surface to determine the size of the reflected image with a measuring telescope and calculate the refracting power on the basis of an assumed index of refraction. The index of refraction of the cornea is 1.376, however; the index used in many keratometer is 1.3375.

The object (mires) is the light source in the instrument and the image is viewed through a microscope so that it can be seen clearly.

Within the approximation of paraxial optics is

$$\frac{r/2}{x} = \frac{h'}{h}$$

where

r = radius of curvature

x = distance from object to focal point

h = object height

h' = image height

In the keratometer, the viewing microscope is fixed so that image plane is always the same distance from the objective, and the object light source is fixed to the body of the instrument. As the distance x from object to the focal point is constant, the equation becomes (for the object of fixed size)

r = constant x h

Hence the corneal radius of curvature is directly proportional to image size.

1. In the Javal Schiotz keratometer, the amount of image doubling is constant and the instrument measures the object size needed to produce an image of fixed size.

   Doubling of the image formed by reflection at the cornea is achieved by a Wollaston prism. (Wollaston prism consists of two rectangular quartz prisms cemented together).
2. In the Helmholtz instrument, the object size was fixed and the amount of doubling varied to produce an image of fixed size.

   In the Bausch & Lomb keratometer, in addition to the use of prisms for separation:

   a) Two small holes are used for forming images by Scheiner principle

   b) Two large holes are the part of doubling system

It measures the radius of curvature of a central zone of the cornea approximately 3mm in diameter. The radius of curvature of the axial zone of the emmetropic eye is about 7.8mm.

Instrumentation

Reichert keratometer (Bausch & Lomb)

It is a one position keratometer. This instrument has constant object size and difference in the size of image to determine the different radii of curvature of the cornea. Doubling process produce a vertical & horizontal doubling simultaneously. The light beam reflected from the patient's cornea passes through four apertures so the left and right apertures contain a horizontally and vertically placed prism, whereas the upper and lower apertures contain no prism.

The upper & lower apertures constitute of a Scheiner's disc mechanism, enabling the operator to keep the instrument in sharp focus.

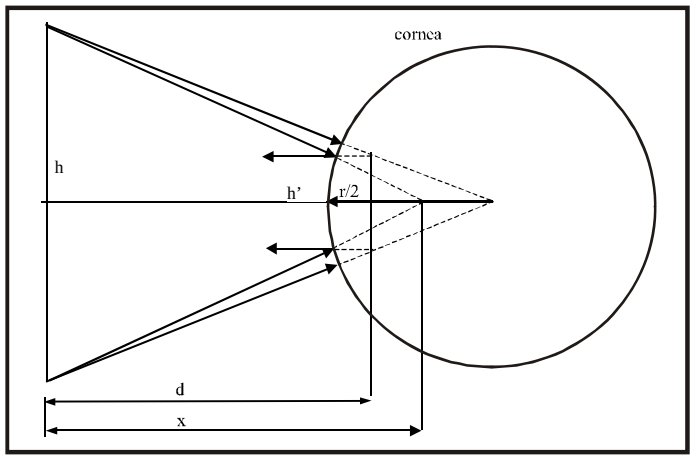

Fig. 1: Principle of keratometry

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All India Institute of Medical Sciences,
New Delhi.

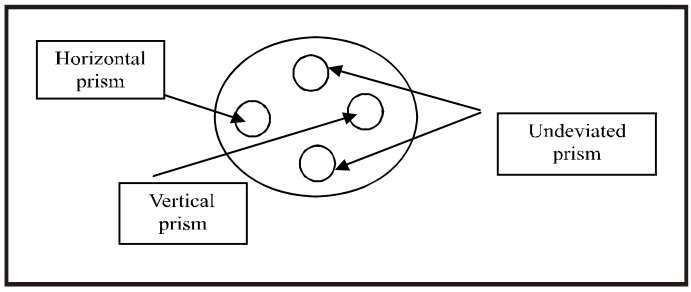


Fig. 2: Apertures in the Keratometer Optical System

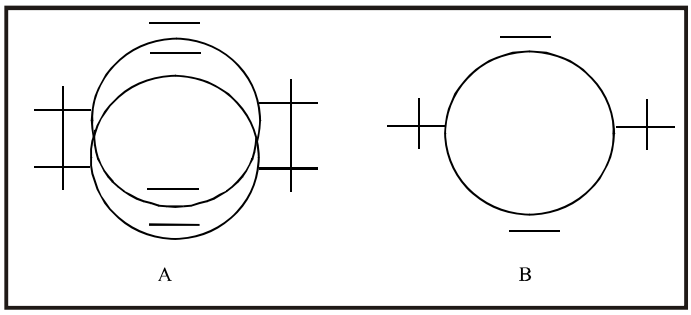


Fig. 3: The Scheiner's disk focusing mechanism Out of focus (A), in focus (B).

The Bausch & Lomb keratometer measures radius of curvature from 36.00D to 52.00D. By placing -1.00 D or +1.25D lens in front of the objective the range can be extended from 30.00 to 61.00D respectively.

Steps for adjustment while performing keratometery

(A) Movement of instrument as a whole to keep the mires focus.

(B) Adjustment of mires size and separation by knobs.

(C) Adjustment of axes by tilting the instrument along the anterio posterior axes to accurately overlap mires.

Clinical procedure for the keratometry

- Focusing the eyepiece
- Instrument alignment
- Focusing the mires
- Position the patient
- Instruct the patient
- Axis measurement
- Curvature measurement
- Record the measurements
- Expand the range of measurement

While performing kerotometery, it is important to keep in mind following limitations and assumptions

1) Calculation based on reflection from a spherical surface
2) Sample only four central points of the cornea
3) Para-axial optics to calculate surface power
4) Assume symmetry about instrument optics axes
5) Distance to focal point approximated by distance to image
6) Assumed index of refraction in radius to diopter conversion
7) One position instruments assume regular astigmatism

Interpreting the findings

The difference in power between the two principle meridians indicates power of the cylindrical lens, which is necessary to correct patient's astigmatism. The meridian of least refracting power indicates the position of minus axis of correcting cylinder.

In With The Rule (WTR) corneal astigmatism - mires will be horizontally oval.

In Against The Rule (ATR) corneal astigmatism- Mires look like a vertical oval.

In Oblique astigmatism, the principle meridians are between 30 - 60 degree and 120 -150 degree.

A. If mires in Bausch & Lomb are a perfect sphere, it indicates a spherical cornea.

B. If mires are irregular or doubled, it indicates an anterior surface irregularity of the cornea.

C. Pulsating mires is indicative of keratoconus.

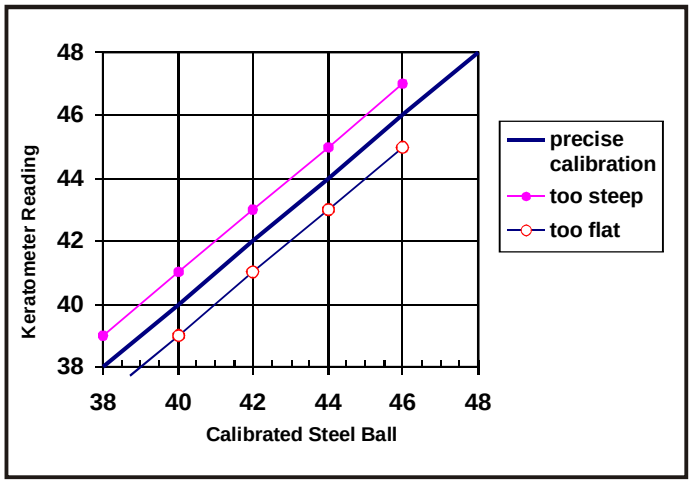
D. Following are Keratometric signs of keratoconus:

i) Early signs:

- Inclination and Jumping of mires (while attempting to adjust the mires, the mires jump. If an attempt is made to superimpose, the plus mires, they will jump above and below each other).

ii) Other signs:

- Minification of mire: In advance keratoconus (K> 52 D), the mires begin to get smaller, due to increased amount of myopia.



Keratometer calibration chart

- Oval mires: Occur due to large amount of astigmatism, mires are normal size and distinct borders.

iii) Distortion of mires: The mire image is "Irregular", "Wavy" and "Distorted".

Clinical uses of keratometry

1. Objective method for determining curvature of the cornea.
2. To estimate the amount and direction of corneal astigmatism
3. The ocular biometery for the IOL power calculation
4. To monitor pre and post surgical astigmatism.
5. Differential diagnosis of axial versus refractive anisometropia.
6. To diagnose and monitor keratoconus and other corneal diseases.
7. For contact lens fitting by base curve selection
8. To detect rigid gas permeable lens flexure.

Calibration of keratometry

A steel ball of known radius of curvature is placed before the keratometer and its value is set on the scale or dial. The mires are focused by clockwise and anti clockwise movement of the eyepiece through trail and error. When mires are in focus, calibration is complete.

Two examples of calibration against standard steel ball bearings. Filled circles show readings that are approximately 0.60D too steep across the measured range. Open circles show readings that are increasingly too flat.

(I) Expanded range of keratometer by direct method:-
   (a) With +1.25D add+9.00D to the scale reading
   (b) With -1.00D, 6.00 D is deducted from the scale reading

(II) Expanding the range of measurement by Mandell

A graph is plotted between the scale reading along ordinate and the actual value of the steel ball along abscissa by placing +1.25 and -1.00 in front of the objective.

For increasing the accuracy

a) Focus the hair cross of Bausch & Lomb and black line of Haag-striet by taking out of the eyepiece anti-clockwise and bring it in clockwise till the lines are first focused.

b) Always occlude the eye which is not examined, otherwise it becomes the fixing eye and reading will come from eccentric point on the cornea.

c) Bausch & Lomb keratometer was made extremely accurate by employing vernier type of alignment by Shick.

Source of errors in keratometry

a) Improper calibration

b) Faulty positioning of the patient

c) Improper fixation by patient

d) Accommodative fluctuation by examiner

e) Localised corneal distortion, excessive tearing, abnormal lid position

f) Improper focusing of the corneal image.

Surgical/ Operating Keratometer

The operating Keratometer is attached to the operating microscope. The accuracy of operating keratometer is limited due to

- Difficulty in aligning the patient's visual axis and the keratometer's optical axis
- Keratometers are calibrated for a fixed distance from the anterior cornea. The different microscope objective lenses result in different focal lengths and therefore working distance.
- Air in the anterior chamber result in a second target reflection.
- External pressure on the globe results in change in corneal curvature.

Automated Keratometer

Automated kertometer is similar to manual keratometer. In this keratometer, the reflected image of target is focused onto a photodetector, which measures image size, and radius of curvature is computed. The target mires are illuminated with infrared light and infrared photodetector is used. The advantage of this instrument is that it is compact and very short time.

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Antiviral Preparations used in Ophthalmic Practice

Sanjay Teotia, DOMS

Currently available antiviral agents are virostatic, They are active against D.N.A. viruses, especially herpes simplex virsus and varicella- zoster virus chemotherapeutic intervention can occur before or at the time of viral particle attachment to host cell membranes, during uncoating of viral nucleic acids, by inhibiting a cellular receptor or factor required for viral replication, or by blocking specific virus coded enzymes and proteins produced in the host cells that are essential for viral replication but not for normal host cell metabolism.

Commonly used antiviral preparations are:

Idoxuridine (IDU, 5-iodo-2 deoxyuridine)

Description

This compound is chemically closely related to thymidine. It inhibits viral metabolism by substituting for thymidine in D.N.A. synthesis and thus prevents replication of virus. The drug acts on virus and host cell D.N.A. and is highly toxic to host cells. Because of its high systemic toxicity IDU has been limited to topical therapy of herpes simplex keratoconjunctivitis.

Indications

Commonly used in acute epithelial herpetic keratitis, herpetic ulcerations of cornea, therapy resistance to acyclovir.

Adverse reactions

Adverse reactions are irritation, pain, pruritus and inflammation or edema of eyelids, follicular conjunctivitis, photophobia.

Dosage and administration

One drop of 0.1% solution is instilled one hourly during day and 2 hourly during night and treatment should be continued for 5 to 7 day after complete healing to lesson the chance of recurrence.

Acyclovir (acycloguanosine)

Description

Acyclovir is a purine nucleoside analogue with activity against herpes viruses (in order of potency) HSV-1, HSV-2, VZV, Epstein Barr Virsus (EBV). It has minimal activity against cytomegalovirus (CMV). It has proved to be an extremely safe and effective agent. It inhibits viral D.N.A. synthesis. It differs in that the virus replication is inhibited without damage to the host cell. It is thus more selective in action. Immunocompromised patients who require prolonged treatment may develop resistance via-mutation in the viral thymidine kinase.

Indications

Topical – Commonly used in epithelial as well as stromal herpes simplex keratitis,

Oral- After penetrating keratoplasty in patients suffering with herpes simplex keratitis, recalcitrant stromal or uveal disease caused by HSV, and to reduce Ocular complications of Keratitis and Uveitis in Herpes zoster ophthalmicus.

Adverse Reactions

A few cases show slight punctate epithelial keratopathy which cease once the drug is stopped. On systemic use nausea, vomiting, diarrhea, headache and rashes may occur

Dosage and adminisration

Topical – 3% ointment is used 5 times a day

Oral – 800 mg 5 times a day for 5 to 7 days

Adenine Arabinoside (Ara-A, Vidarabine)

Description

It is a purine nucleoside, interferes with viral D.N.A. and is effecitve in treatment of H.S.V. infections Adenine Arabinoside appears less susceptible to the development of drug resistant viral strains than I.D.U., and I.D.U. resistant infections often respond to vidarabine.

Indications

Acute keratoconjunctivitis, Recurrent Superficial Keratitis caused by HSV-1 and HSV-2

Adverse Reactions

Possible adverse effects include tearing, irritation, pain, photophobia and superficial punctate keratitis.

Dosage and administration

Available as 3% ophthalmic ointment used 5 times a day till epithelization occurs and than reduced to once or twice daily for 4 to 5 days to prevent recurrences.

123, Laxmanpuri, Near Classic Hotel
Delhi Road, Bulandshahr (U.P.)

Trifluorothymidine (TF-3)

Description

It is a pyrimidine nucleoside. It has advantage over IDU of higher solubility, greater potency, lack of toxicity and allergic reactions. It is also effective in IDH resistant cases. It is a D.N.A. inhibitor with same mechanism as IDU.

Indication

Use in treating primary keratoconjunctivits and recurrent keratitis caused by HSV and HSV-2, Herpetic ulcers, prevention of complications produced by corticosteroids.

Adverse Reactions

More toxic than acyclovir, may cause mild superficial punctate keratitis on prolonged use.

Dosage and Administration

1% eye drop one drop instilled 4 hourly results in healing of 90% of herpetic ulcers in a period of two weeks.

Cidofovir

Description

Cidofovir (cytosine; HPMPC) is a nucleotide analog and, in contrast to nucleoside analog contains a phosphonate group that does not require virus- dependent phosphorylation Celluar enzymes convert cidofovir to the active diphosphate form, which has a long intracellular half life. Cidofovir has inhibitory in vitro activity against a broad spectrum viruses including HSV-1, HSV-2, VZV, CMV, EBV, adenovirus, human papillomavirus and human polyomavirus.

Indication

Used to treat CMV Retinitis in patients with HIV

Adverse Reactions

Renal failure, rash, headache, fever

Dosage and administration

5 mg / Kg I.V once weekly for 2 weeks with maintenance therapy every other week.

Ganciclovir

Description

Ganciclovir is a nucleoside analog of 2 deoxyguanosine, Ganciclovir inhibits viral D.N.A. synthesis by competitive inhibition of viral D.N.A. Polymerase and is incorporated into viral D.N.A. as D.N.A. chain terminator.

Indication

CMV Retinitis

Adverse Reactions

Bone marrow suppression perticularly neutropenia, anemia, rash, fever, liver function abnormalities.

Dosage and administration

Parentral – 10 mg / kg/ day IV in 2 divided doses for 2 to 3 weeks and require suppressive thereapy to prolong time to relapse at 5 mg/ kg/ day.

Intravitreal – 400mg administered through parsplana are useful for patients with CMV retinitis who are resistant to IV ganciclovir.

Foscarnet

Description

Foscarnet in an organic analog of inorganic pyrophosphate. It selectively inhibits virus specific D.N.A. polymerase and reverse transcriptase. It inhibits HSV-1, HSV-2, human herpes virus 6, EBV and VZV. Foscarnet's efficacy is similar to that of ganciclovir for treating and delaying progression of CMV retinitis with a different adverse effect profile

Indication

Used in CMV Retinitis

Adverse Reactions

Intravitreal use- direct retinal toxicity, intravitreal bleeding and endophthalmitis.

IV use- nephrotoxicity, hypokalemia, hypocalcemia

Dosage and Administration

Parentral – 60 mg / kg TID for induction or 90 mg IV BID for 2 week is followed by maintenance of 90 to120 mg/ kg IV daily.

Intravitreal

Doses between 1200 and 2400 mg used with an induction period of twice a week for 3 weeks, followed by weekly injections.

Immune Globulins

Hyperimmune CMV immunoglobulin has attenuated CMV disease associated with kidney transplantation but it has not proved useful in preventing CMV disease in HIV infected persons. A human monoclonal anticyto-megalovirus antibody may be useful as adjunctive therapy with foscarnet or ganciclovir for the treatment of CMV retinitis.

Zidovudine (azidothymidine)

Zidovudin is recommended for selected AIDS patients with non vision threatening retinitis.

Surgical Eye Camps: Can we make them Safe?

Harsh Goel DO, MS, **Subodh Sinha** MS, **Abhas Sinha** MS, **Anil Tara** MS

Camps are an effective modality for medical service delivery on a mass scale and in a cost effective manner. Ophthalmology is probably the only discipline which has tapped the potential of camps as a very effective strategy in combating illness at grassroots level. Other medical disciplines have also adopted this approach, but, in terms of sheer numbers, ophthalmology has been miles ahead of other disciplines.

Over the years, ophthalmic camps have become cataract centered as cataract remains the major cause of blindness globally, responsible for nearly 60% of blindness worldwide. One day screening camps and refraction camps as a part of school screening activities are also common, but handle far lesser volumes of patients, and are not involved in any surgical intervention. Cataract surgical camps are sometimes the only modality available in inaccessible areas which lack eye care services. These camps have remained popular because they are not only cost effective, but also give dramatic results. The major reason for the latter is that cataract surgery is the most successful of all surgical interventions across all the medical disciplines. The participation of various governmental and non-governmental/ voluntary agencies has added to the effort.

The flip side to this scenario is the attempt to exceed the numerical results of the competitor (?) leading to a number race and a game of one-upmanship between different camp organizers in a region. The worst case scenario is when these camps are organized by dubious individuals to fulfill their personal or political agenda. This kind of a number game has led to a series of disasters at eye camps which have cast a shadow of doubt on the safety and efficacy of camps. The year 1986 was a watershed year in the checquered history of eye camps because of the disasters at Khurja and Moradabad in UP in quick succession. These incidents led to the institution of the norms and guidelines for surgical camps at the insistence of the Supreme Court of India which are still in force. The tendency of throwing caution to the winds, taking shortcuts by ignoring standard norms and protocols and servings vested interests has made *CAMP* the proverbial *'four letter word'*. Any organization which supports the concept of surgical camp is often treated like a pariah by other, '*holier than thou'* organizations.

The authors have been actively involved in surgical eye camps over the last two decades, and having worked in varying environments, have also been actively participating in improvisations and innovations aimed at making camp surgeries safe and reliable. The authors are of the opinion that through stringent internal quality audits and constant innovations and up-gradation in service delivery systems, an organization will be able to institute standards in surgical eye camps which will make camp surgery as safe and reliable as surgery at a tertiary centre of excellence.

Venu Eye Institute & Research Centre
New Delhi.

So, How Do We Make Eye Camps Safe?

What follows is a guided tour of such a model surgical eye camp. No doubt, this model entails higher expenditure in terms of technology, human resources and consumables, but, cost cutting at the expense of protocols and safety has led to the current crisis. In a service oriented profession, health restoration is of supreme importance, and, no expense should be spared to ensure this.

Planning

For a particular camp, the planning starts off with the local organizer's approach letter. The local organizer is sent the organization's Camp methodology information, which is followed by a camp site feasibility study. This study includes the infra-structure for OPD, IPD and Operation theatre, along with the willingness of the organizer to provide support facilities like generators as electrical Back-up, air-conditioner for the operation theatre and other logistic support. The level of any existing medical services in the area is also assessed, in case any assistance from other medical service provider may be needed during the camp. Once the Camp site is found feasible, and the organizer accepts the terms and conditions for conducting the camp (including the assistance of a General physician and anesthetist for the duration of the camp), the camp dates and targets are finalized and formalities for obtaining requisite permissions from local authorities are instituted by the local organizer. At the same time, human resource planning for the team according to the target, and logistic planning regarding transport, equipment and consumables required for the camp is initiated by the eye care provider organization.

A Typical Surgical Camp Team

For an average surgical camp with a target of 200 surgeries, the team would be as follows

- Surgeons (3-4) including one Consultant and 2-3 Senior Resident Medical officers
- Optometrists (4-5)
- OT Technicians (2)

- Camp Coordinator, especially in those camps which are being held for the first time in a particular region. In camps which are being held year after year at the same site with the same organizer, the camp coordinator's role would be limited.

Duration and Programme

A typical camp would be of 9 days' duration; OPD is conducted on 1-2 days according to the need. The second OPD day is also dedicated to a review of all admitted patients. The next three days are for surgery, at approximately 75 surgeries per day between 9.00 am and 7.00 pm. The last four days are spent in post-operative patient care, as well as for refraction in patients advised the same at the time of OPD screening. Emergency eye care services are also provided.

At the end of the camp, the operated patients are discharged with advice to report for first follow-up at the same site after six weeks, when sutures are removed after preliminary evaluation. The patients are then advised to return again after two weeks for assessment of visual outcome by recording best corrected Visual acuity after refraction. At every stage, patients with complications are referred to the base hospital for treatment for which the organizer provides logistic support.

OT Installation Protocol

The team reaches the camp site a day in advance, the installation and fumigation of the Operation Theater being the first priority.

The selection of an appropriate room to be used as the Operation Theater is the most crucial task in any camp, as even a small mistake or slip up in the Operation Theater preparation can lead to unimaginable disasters. The recent trend of disallowing camps where a makeshift Operation Theater is to be prepared in places like schools and dharamshalas, has emerged out of instances of improper OT planning and compromises on standards. It is advisable to get even a running Operation Theater in a Government or private hospital freshly whitewashed or painted as it takes care of basic cleanliness once the final washing is done.

OT Installation Procedure

The freshly whitewashed/ painted room to be used as the Operation Theater is first cleaned thoroughly with detergent and disinfectant. All openings and cracks are sealed properly, and the air-conditioner and other electrical appliances are installed. Next, the furniture and equipment like operating microscopes *etc* are cleaned and installed. After a final inspection by the senior most surgeon, the operation theater is fumigated and sealed, to be reopened only after 36-48 hours, on the first day of surgery. Subsequently, the Operation Theater is fumigated again everyday after the surgeries are over.

OPD Procedure

The equipment available in the OPD are Torches, Direct and Indirect ophthalmoscopes, Schiotz Tonometers, complete refraction units, keratometer, A-scan biometer and a Hand-held slit-lamp Biomicroscope, besides BP instruments , Urinalysis strips and handy Glucometers.

Pre-Operative Assessment Protocol

The primary OPD activity in a surgical eye camp is screening for operable cataract.

- Patients thus identified are initially screened for any systemic ailment like hypertension, diabetes, Coronary artery disease, prostate enlargement, asthma and any skin lesion or open wound that may be a cause for concern during surgery.
- Once cleared for a systemic ailment, the patients have their presenting and pin-hole assisted Visual Acuity recorded.
- Patients Aphakic in the other eye are assessed for best corrected visual acuity in that eye.
- The patients then undergo Keratometry and A-scan biometry for axial length and IOL power calculation.
- The patients next have their IOP and Xylocaine sensitivity checked, lashes trimmed, eye to be operated marked with a marker pen, and pupils of both eyes dilated for slit lamp and fundus examination.

The above sequence in which the various investigations are carried out is strictly followed, after which each patient's case sheet complete with consent for surgery is prepared. At the end of the OPD, exhaustive information about all patients is ready. Once the list of all admitted patients is available, the following among them are identified and listed separately for the senior most surgeon to operate:

- One eyed and practically one eyed patients
- Patients aphakic in other eye with best corrected aphakic visual acuity of less than 6/18 (treated as practically one eyed patients)
- Young patients(less than 45 years)
- Patients requiring complicated surgeries including combined cataract+glaucoma surgeries
- Patients pseudophakic in the other eye

Pre-Operative Treatment Protocol

All admitted patients are frequently administered a broad spectrum antimicrobial topically. In between, at regular intervals, Povidone Iodine is also administered topically in the eye to be operated. The skin of the eyelids and the surrounding area is painted with Povidone Iodine solution. Patients already on any systemic medication are advised to continue the same. Patients are also advised on personal hygiene and cleanliness, especially during their stay in the camp.

OT Protocol

The Operation theater technicians spend the OPD days in sterilizing the surgical instruments sets and the linen including the surgical gowns and eye drapes by autoclaving. Once this job is completed, they move over to the OPD work-up area to assist the team working there. The sealed operation theater is opened early on the day of commencement of surgeries, and the air-conditioner is switched on. Nobody is allowed to enter the operation theater in their street clothes; in fact, the camp team carries freshly laundered OT dresses in excess of the total number of team members. After administration of the local anesthetic block, patients are escorted into the Operation Theater after covering their dresses under specifically earmarked gowns, and making them wear a surgeon's disposable cap to cover their hair. Special care is taken to prevent the entry of woolen garments like shawls, sweaters, jackets, socks *etc* into the Operation Theater, as these garments tend to attract and trap a lot of dust particles and germs.

Surgical Protocol

A standard surgical protocol developed for any surgical intervention based on internationally accepted principles is adopted. The surgery is performed according to a standard technique which clearly defines the various steps in a surgical procedure. Major points of emphasis are:

- Changing disposable surgical gloves after every surgery
- Strictly following the 'one needle-one prick' principle
- Using one set of surgical instruments, including various cannulae, only once; the same set is used only after all instruments have been thoroughly cleaned and re-sterilized by autoclaving
- Discouraging use of chemical sterilization for surgical instruments
- At least one of the surgeons in the Operation Theater should be a senior and experienced person, capable of guiding a lesser experienced colleague in case of any difficulty during surgery.
- The area at the entrance of the Operation Theater, and the OT floor are regularly mopped with antiseptic/ disinfectant at frequent intervals while the surgeries are being performed.
- Physician, anesthetist, emergency kit, Oxygen Cylinder and transport facility, preferably an ambulance should be available at the camp site for the duration of the surgeries to assist in case of any medical emergency.
- At the end of the day's surgeries, the Operation Theater floor, furniture *etc* are thoroughly cleaned and the operation theater is again fumigated and sealed till the next morning.
- The instruments sets and linen for the next day are also autoclaved at the end of the day's surgeries.
- Attention to Bio-Medical Waste management at the camp site, especially to handling of used sharps is important.

A word of caution here; the temptation to utilize camps as a training (? hunting) ground for freshly qualified ophthalmic surgeons or trainee residents is quite strong. However, one should realize that surgical camps are an area of extraordinary stress for the entire team, not to mention the team leader and the organization, as it is an unfamiliar, uncontrolled environment; part of the stress emanates from being prominently in the limelight, thus exposing oneself to public scrutiny and unscrupulous elements. In such a setting, the camp should not become a training ground. It is advisable to tread with caution, and lay down strict guidelines for the surgical cases to be allowed to the least experienced surgeons/ trainee residents, and their close monitoring during surgery in a camp.

Post Operative Care Protocol

The post-operative care is provided for a minimum of four days after the surgery. This includes a daily examination with the hand-held Slit lamp Biomicroscope. Appropriate topical and systemic medication as required is started, and review rounds in the evening are a must. Patients with any surgical complications are immediately examined by the consultant at the camp site, and are managed as warranted.

At the time of discharge, presenting and pin-hole assisted visual acuity in the operated eye is recorded, the fundus examined and the patients are advised about the date and location of their first follow-up visit, which is usually at 6 weeks from the last surgical date in a particular camp. This information is properly documented and shared with the local organizer; the same information is then passed on to the camp coordinator's office.

First Follow-up (Sixth Week)

The first follow-up is done after six weeks of surgery. The team consists of one Senior Resident Medical officer and two optometrists, and carries all the case records of the patients operated in the camp, along with a list of these patients, and instruments for suture removal. After case record retrieval, the presenting and pin-hole assisted visual acuity is recorded. After a preliminary examination to ensure that there no late post-operative complications, the sutures are removed, a topical antimicrobial preparation advised for use over the ensuing two weeks, and the patients advised to report again for a second follow-up after two weeks. All clinical findings including visual acuity are properly documented in the case records and the additional list. The emphasis in this visit is on safe suture removal, which is again ensured by adhering to strict sterilization norms and protocols.

Second Follow-up (Eighth Week)

The team strength and composition for this visit is the

same as for the first follow-up visit, the team this time is equipped with a complete refraction unit and an ophthalmoscope. The patients' presenting and pin-hole assisted visual acuity is recorded, after which refraction is done. The patient is then prescribed appropriate spectacles. The clinical and refraction findings are again properly documented in the case records and the list, along with the best corrected visual acuity. Patients with poor visual recovery have their fundus examined to determine the cause of poor visual outcome.

The rationale behind a second follow-up is that it then becomes feasible to assess the visual outcome in a particular camp. It also provides an opportunity for the drop-outs from the first follow-up to get follow-up attention and care.

Data Entry, Collation and Analysis

At completion of various stages in a camp, the data generated from a camp are computerized; such data from a series of camps is collated at regular intervals and analyzed for visual outcomes on the basis of internationally accepted criteria. The WHO guidelines on classifying visual recovery into *good, satisfactory/fair and poor* on basis of best corrected visual acuity are quite acceptable. Regular analysis of visual outcome helps in identifying shortcomings in the system and rectifying them.

Technical Up Gradation

It is quite evident that the authors have described an eye camp where the choice of surgical modality is a conventional Extra-capsular cataract Extraction with IOL implantation where sutures are applied as a routine; this should not be misconstrued as an attempt to discourage or disparage Small Incision Cataract Surgery (Manual SICS) or Phacoemulsification camps. Both of these techniques have proved their efficacy and safety in all settings. An additional advantage is that there are no sutures that need to be removed. However, the tendency among SICS/Phaco camp surgeons of mocking at ECCE+IOL camp surgeons should be contained, as it would not be prudent for an inept or inexperienced SICS/Phaco surgeon to stubbornly insist on adopting these techniques just for egoistic reasons, and in the process leave a trail of destruction behind him. It would be more prudent on his part to instead stick to ECCE+IOL with sutures and give better results. Proper training in these techniques under supervision of experts at a tertiary level centre can prepare more confident and competent surgeons.

Camps, as already pointed out, have become cataract centric due to the magnitude of cataract blindness and the dramatic results of surgery. However, as the facilities are taken to the doorsteps, other major causes of blindness can also be tackled. Add on services can be provided for conditions like glaucoma, diabetic retinopathy screening, refractive errors, low vision *etc* thus widening the scope of services delivered through surgical camps.

In Conclusion

Despite the recent progress in eye care service delivery in the developing and under-developed countries, huge pockets of high cataract blindness exist in these areas as their cataract surgical rates are dismal. The main reason for this is the stark paucity of eye care service delivery infra-structure in these areas. Till such permanent service systems are developed in these areas, surgical camps would remain the major source of succour to millions of cataract blind. Instead of condemning surgical eye camps for their poor results due to safety and reliability issues, it would be more prudent for eye care providers to do a bit of introspection, analyze their mistakes and shortcomings and develop and adopt standard clinical, management and administrative protocols to improve upon their surgical eye camp delivery systems. The model described in this presentation has been implemented and replicated repeatedly with outstanding success. Once an organization decides to develop and stick strictly to such protocols, which may involve a bit more expenditure, eye camps would be able to shed their unsafe and unreliable image, and again become an effective service delivery modality in areas of dire need. This would definitely justify the extra expenditure in the long run.

Digital Fundus Camera: Mechanics and Applications in photography and angiography

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Although fundus photography has a long history, with the first published report of human retinal photography dating to 1886, commercial systems are now undergoing a notable evolution in the new millennium. Ophthalmologists have relied on fluorescein angiography as an important tool in the understanding, diagnosis and treatment of retinal disorders, for nearly four decades. This diagnostic procedure utilizes a specialized fundus camera to capture not only the retina, but also rapid-sequence photographs of the retinal vasculature following an intravenous injection of an orange-colored, fluorescent dye (fluorescein). Fluorescein angiography facilitates the in-vivo study of the retinal circulation.

Equipment

Digital Fundus Photography

Fundus photography documents the retina, the neurosensory tissue in our eyes which translates the optical images we see into the electrical impulses our brain understands. The retina can be photographed directly as the pupil is used as both an entrance and exit for the fundus camera's illuminating and imaging light rays. The patient sits at the fundus camera with their chin in a chin rest and their forehead against the bar. An ophthalmic photographer focuses and aligns the fundus camera (Figure 1 a-e). A flash fires as the photographer presses the shutter release, creating a fundus photograph like the picture.

Improved Resolution: While film-based photography has a resolution of about 4,500 x 3,000 pixels (when color film is exposed and developed correctly), most color digital fundus images have had 640 x 480 pixels-and 1,024 x 1,024 pixels for black-and-white digital angiography-until recently. The resolution of the newest generation of color digital photographic systems (at 2,000 x 3,000 pixels) now more closely approaches that of film, although these higher-resolution units are just beginning to make their way into clinical settings.

Fundus Camera

A fundus camera is a specialized low power microscope with an attached camera. Angiography requires the use of a specialized fundus camera (Figure 2) equipped with a matched pair of exciter and barrier filters along with a fast recycling electronic flash tube that allows a capture rate of up to 1 frame per second. Its optical design is based on the indirect ophthalmoscope. Fundus cameras are described by the angle of view ie the optical angle of acceptance of the lens.

An angle of 30° (Figure 3), considered the normal angle of view, creates a film image 2.5 times larger than life, specially used for documenting macular details. Wide angle fundus cameras capture images between 45° and 140° and provide proportionately less retinal magnification, and are useful in documenting larger areas of the retina. A narrow angle fundus camera has an angle of view of 20° or less.

Narrow band-pass interference filters are utilized to allow maximum transmission of peak wavelengths, while minimizing any crossover of transmission curves. The exciter filter transmits blue-green light at 465-490nm, the peak excitation range of fluorescein. The barrier filter transmits a narrow band of yellow at fluorescein's peak emission range of 520-530nm (Figure 4). The barrier filter effectively blocks all visible wavelengths but the specific color of fluorescein while doing angiography.

Images are captured either with high-speed black-and-white 35mm panchromatic film or electronically, with a charge-coupled device (CCD) and computerized system for digital imaging. Film-based angiography requires either the use of a processing service, or access to a darkroom for processing films on-site.

A. Stereo imaging

Stereo photographs (Figure 5) can be achieved by shifting the fundus camera laterally between sequential photographs. The lateral shift causes the illuminating beam of the fundus camera to fall on opposite slopes of the cornea. The resulting cornea-induced parallax creates a hyperstereoscopic effect that is evident when the sequential pair of photographs is viewed together. This is particularly useful in identifying the histopathologic location of angiographic findings within the retina. The simultaneous stereo fundus cameras use one exposure to place two images side by side on a single 35mm frame.

B. Digital imaging

Digital angiography imaging systems have been available for over fifteen years and continue to improve in quality each year. Although photographic film is still

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Table 1: Characteristics of Fluorescein	
Chemical compound	A highly fluorescent chemical compound synthesized from the petroleum derivatives resorcinol and phthalic anhydride
Absorption of light	Absorbs blue light, with peak absorption and excitation occurring at wavelengths between 465-490nm. Fluorescence occurs at the yellow-green wavelengths of 520 to 530nm.
Dose	Adult dosage is 500mg Injected intravenously
% of Dye	5ml of 10%, 2ml of 25%

capable of capturing greater detail than current digital systems, digital imaging offers some distinct advantages over the more traditional film-based angiogram. Instant access to the electronic images increases efficiency and promotes better patient education by reviewing images on a monitor with the patient. Images can be stored on magnetic media like CD-ROMs and transmitted electronically to remote sites equipped with a computer for viewing. Digital systems also offer the additional advantage of shortening the learning curve for novice angiographers. Having instant feedback allows the angiographer adjust exposure settings and camera alignment to correct any flaws in technique.

Optics of the Fundus Camera

The light is generated from either the viewing lamp or the electronic flash which is projected through a set of filters and onto a round mirror. This mirror reflects the light up into a series of lenses which focus the light. A mask on the uppermost lens shapes the light into a doughnut. The doughnut shaped light is then reflected onto a round mirror with a central aperture, exits the camera through the objective lens, and proceeds into the eye through the cornea (Figure 6). Assuming that both the illumination system and the image are correctly aligned and focused, the resulting retinal image exits the cornea through the central, un-illuminated portion of the doughnut. The light continues through the central aperture of the previously described mirror, through the astigmatic correction device and the diopter compensation lenses, and then back to the single lens reflex camera system (Figure 7).

Focussing of the Camera

The relationship between sharpness in the reticle, the fundus view, and the sharpness in the final fundus photograph is given in (Figure 8 a&b). When the image is focused without regard to the sharpness of the focusing reticle, the fundus image perceived would be as sharp while the reticle image appears blurry. The image seen is focused by the observer above the focusing screen, closer than infinity. When viewed through a correctly adjusted eyepiece before the fundus has been focused on, then the fundus appears blurry while the reticle is distinctly sharp. After correctly adjusting the eyepiece and the fundus camera's focusing mechanism, both the fundus image and the reticle appear sharp and clear, and this is the end point. The observer's eyes are focused on the reticle, and the image from the fundus camera corresponds with the film plane. Only this final combination will yield a sharply focused fundus photograph. To brief the whole procedure, a step by step approach is proposed.

Visualization Modes

The fundus camera allows operating modes fluorescein angiography (FLUO), red free (GREEN) and true color photography (COLOR) as well as monochromatic photography with blue and red filters. The ICG mode is not provided as the technical requirements for the use of infrared film are extremely high.

In this article we would discuss about the fundus camera application in angiography.

How angiography is performed?

1. Mydriatic eye drops are administered to dilate the pupil.

Table 2: Adverse Reactions

- Extravasation of dye
- Transient nausea
- Vomiting
- Pruritis
- Urticaria
- Bronchospasm
- Laryngeal edema
- Anaphylaxis
- Hypotension
- Syncope
- Seizures
- Myocardial infarction
- Cardiac arrest

Table 3: Common Indications
Diabetic retinopathy
Age related macular degeneration
Subretinal neovascular membrane
Central retinal vein occlusion
Branch retinal vein occlusion
Central serous chorioretinopathy
Cystoid macular edema
Hypertensive retinopathy
Central retinal artery occlusion
Branch retinal artery occlusion
Retinal arterial macroaneurysms
Pattern dystrophies of the retinal pigment epithelium
Choroidal tumors
Chorioretinal inflammatory conditions
Hereditary retinal dystrophies
Post treatment angiograms also check the efficacy of laser treatment.

2. The chin is placed on a chin rest, and the forehead against a support bar to keep the head still during the test.
3. Fundus photographs of the inside of the eye are taken.
4. Then, the dye is injected into a vein, usually at the bend of the elbow.
5. As the dye is injected, a series of photographs are taken.
6. The needle is removed and pressure is applied to the injection site for several minutes.
7. If indicated late frame photographs are taken and the test is over.

Characteristics of Fluorescein Sodium

The fluorescent properties have made fluorescein useful in a variety of industrial, scientific and medical applications. The dye was first synthesized by Von Baeyer in 1871.

Mechanism of action

The dye when injected intravenously enters the circulation, where approximately 80% of the dye molecules bind to serum protein. The remaining unbound or free fluorescein molecules fluoresce when excited with light of the appropriate wavelength. The dye is metabolized by the kidneys and is eliminated through the urine within 24 to 36 hours of administration. The side effects of intravenous fluorescein include discoloration of the urine for 24 to 36 hours and a slight yellow skin discoloration that fades within a few hours. Nursing mothers should be cautioned that fluorescein is also excreted in human milk.

Complications and adverse reactions

Usually, fluorescein is well tolerated by most patients, but angiography is an invasive procedure with an associated risk of complication or adverse reaction. Adverse reactions occur in 5 to 10 percent of patients and can range from mild to severe.

Indications and uses

The most common uses of fluorescein angiography are in retinal or choroidal vascular diseases. The angiogram is used to determine the extent of damage, to develop a treatment plan or to monitor the results of treatment. The common diagnostic use of fundus fluorescein angiography is summarized in the table below.

Phases of the normal angiogram

The normal angiogram is divided into three phases: the early, mid and the late phase. The normal arm to retinal circulation time is approximately 8-10 seconds.

Early phase: On the contrary the early phase of angiogram is further subdivided into the following phases:

- *Choroidal flush:* In a normal patient, the dye appears first in the choroid in 10-12 seconds. The major choroidal vessels are impermeable to fluorescein, but

Table 4: Abnormal Angiogram

Hypofluorescence:
- Filling defect
- Blocking defect

Hyperfluoresence:
- Autofluorescence
- Pseudofluorescence
- Transmission or "window" defect
- Leakage
- Pooling
- Staining

Table 5: Ultra Widefield
Ret Cam
➢ The Ret Cam was developed as a pediatric instrument to image infants under anesthesia.
➢ Light enters and exits through the front optics of a handheld probe. It has a 130° field of view.
➢ This camera requires pupillary dilation and contact with the patient's cornea. Thus, patient comfort and compliance are still issues.
Panoret 1000
➢ This camera is mounted on an arm that can hold the probe just above the eye.
➢ It still requires an external light source; however, the more sensitive detector systems and computer-controlled illumination allow for a lower power light source.
➢ The image quality is good. It has a 100° field of view, and dilation is not required.
Optomap Exam
➢ This system uses an ellipsoidal mirror to miniaturize the scanning laser raster of light, producing a virtual scan point of the real scan point just behind the pupil.
➢ The virtual scan point allows for an ultra widefield image with the light entering and exiting through the pupil.
➢ There's no contact with the eye, and dilation is not required, resulting in better patient cooperation and ease of use.

the choriocapillaris leaks fluorescein dye freely into the extravascular space. There is usually little detail in the choroidal flush as the retinal pigment epithelium (RPE) acts as an irregular filter that partially obscures the view of the choroid.

- *Arterial phase:* The retinal arterioles typically fill a second or two after the choroid.
- *Arteriovenous phase:* Complete filling of the retinal capillary bed follows the arterial phase and the retinal veins begin to exhibit laminar filling.
- *Venous phase:* Complete filling of the veins occurs over the next ten seconds with maximum vessel fluorescence occurring within 30-35 seconds after injection.

Mid phase: This phase is also known as the recirculation phase, and occurs about 2 to 4 minutes after injection. The veins and arteries remain roughly equal in brightness. The intensity of fluorescence diminishes slowly during this phase as much of the fluorescein is removed from the bloodstream on the first pass through the kidneys.

Late phase: The late or elimination phase demonstrates the gradual elimination of dye from the retinal and choroidal vasculature. Staining of the optic disc is a normal finding. Any other areas of late hyperfluorescence suggest the presence of an abnormality.

The abnormal angiogram

Interpretation of the abnormal angiogram relies on the identification of areas that exhibit hypofluorescence or hyperfluorescence. These are descriptive terms that refer to the time specific, relative brightness of fluorescence in comparison with a normal study.

Advances in digital fundus photography

- Live-motion, fluorescein angiography and color fundus imaging.
- Sharper optics
- Early, real-time angiography
- Instant feedback on focus and alignment
- Dynamic parallax stereo imaging
- No-flash procedures for increased patient comfort

Newer modalities available for fundus photography and angiography

View with a fundus camera was limited to between 30° and 60° of the retina. To view the periphery, patients

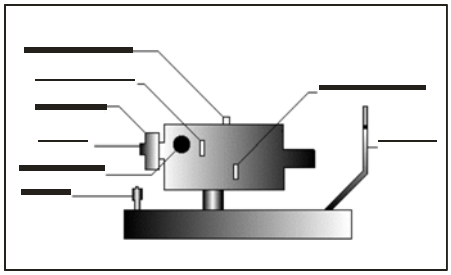


Fig. 1a: Parts of the fundus camera

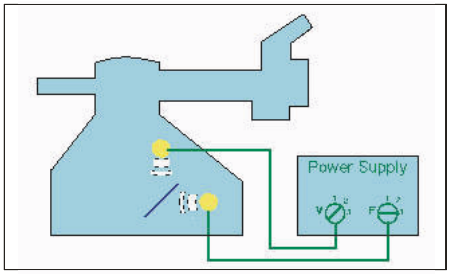
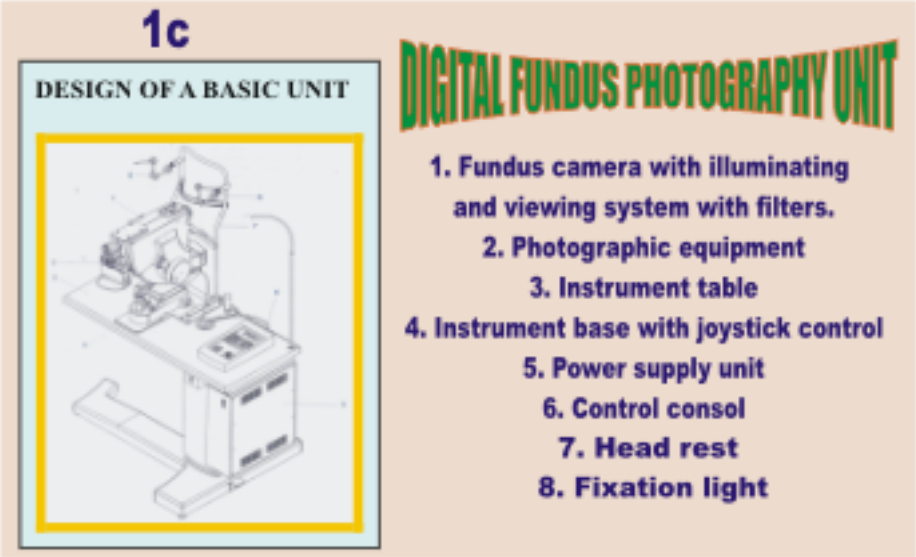
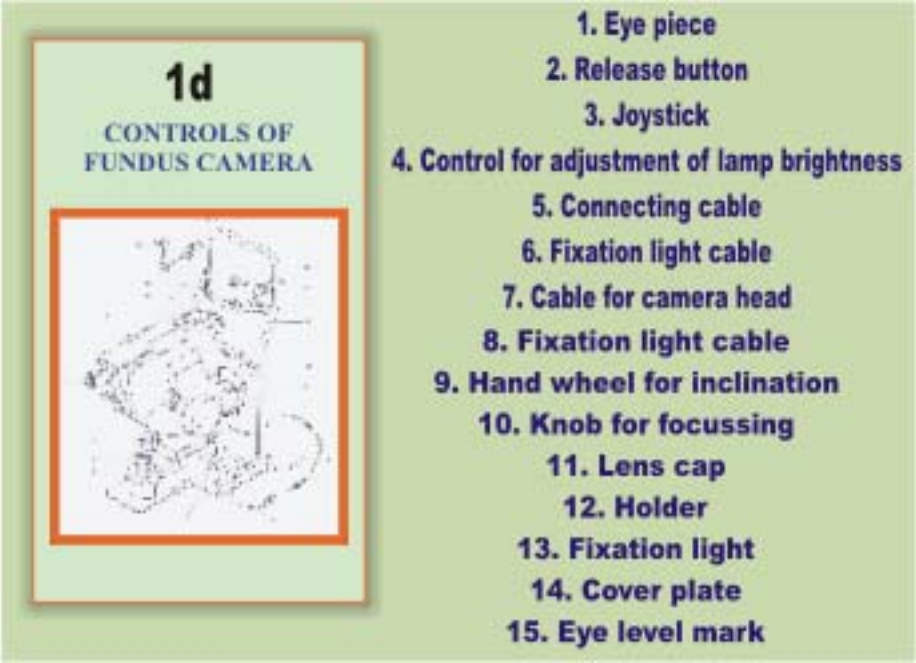
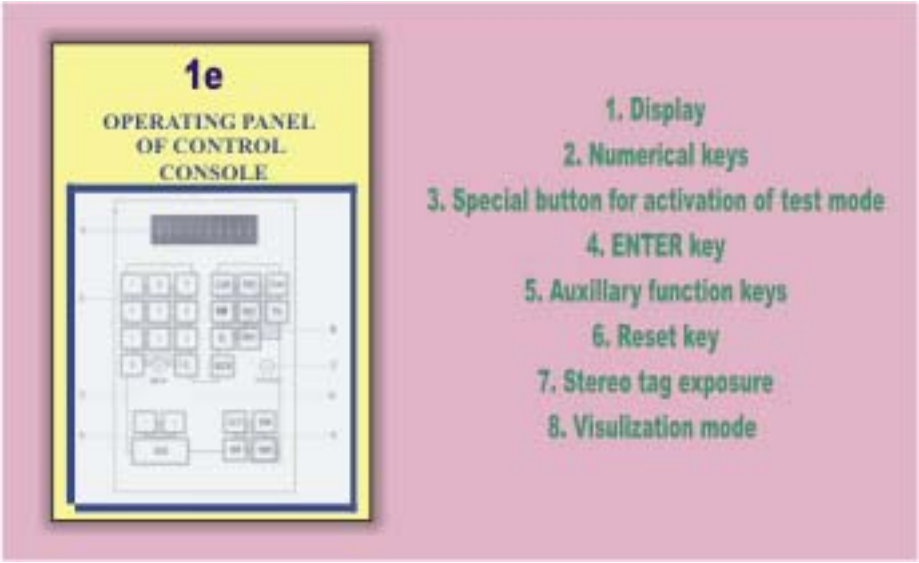


Fig. 1b: Fundus camera: specialized low power microscope with an attached camera, showing the power supply







had to be asked to move their eyes into the direction of the fundus area in question. Tilting the camera along the horizontal plane could increase the peripheral field of view in the 3 o'clock and 9 o'clock positions. And some fundus cameras could be tilted along the vertical plane to capture the 12 o'clock to 6 o'clock positions. Though it was possible to view the retina mid peripherally, the extreme periphery was difficult to image. With this in mind, in 1980's introduction of the first widefield camera, the Equator Plus, so named because it could obtain images beyond the equator, was introduced. Although a significant advance, the Equator Plus still had drawbacks.

1. The camera had to touch the patient's cornea, a procedure that's difficult for most patients to tolerate.
2. The light could not pass through the camera's front lens but had to be delivered to the eye externally through the bulbar conjunctiva, sclera, choroid and retina.
3. The light passing through the fiber optic system had to be very bright, which created heat on the external eye.
4. The area of the fundus that was being illuminated externally was a very bright and saturated (washed out) region of the image, making it impossible to see lesions in that area of the image.
5. The greatest drawback to the Equator Plus was that light passing through the choroid with its rich blood vessel supply added considerable red color to the white light, resulted in a fuzzy image with a red hue.

Even though it produced truly widefield images, the Equator Plus camera never became popular. Then came in the Ultra widefield technology.

Ultra widefield technology

It wasn't until the late 1990s and early 2000s that the imaging systems could obtain good quality, ultra

Fig 2: Fundus Camera with its accessories

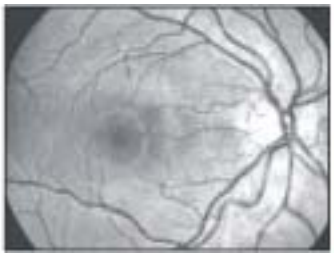


Fig. 3: An angle of 30°: Normal angle of view

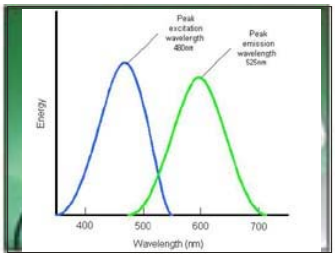
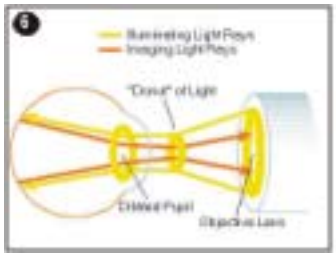


Fig 4: Excitation and Emission Curves of Fluorescein Sodium

Fig. 5: 3-Dx Stereo Fundus Camera



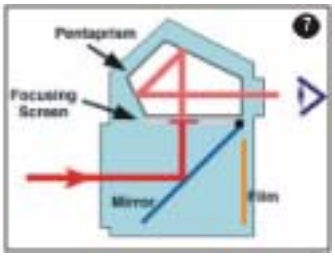
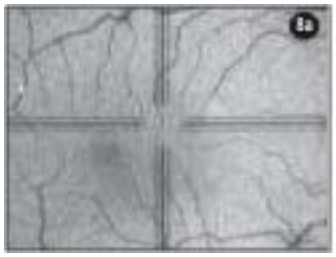


Fig. 6: Illumination system and image correctly aligned and focused: the resulting retinal image exits the cornea through the central, un-illuminated portion of the doughnut.

Fig. 7: Light rays passes through the central aperture of the mirror - through the diopter compensation lenses - back to the single lens reflex camera system



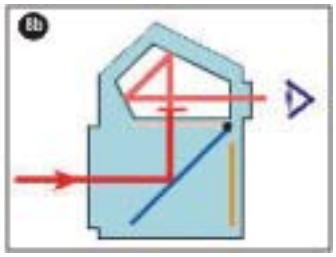


Fig. 8a&b: Showing the reticle formation for clear viewing and photography of the fundus.

widefield technology was introduced. Among the most successful are:

1. Ret Cam
2. Panoret 1000
3. Optomap Retinal Exam.

Each of these instruments produces good digital images that show the peripheral fundus anterior to the equator, but there are some differences.

Conclusion

Hence, with the advent of the highly specialized digital fundus camera, angiography aswell as documenting fundal findings have become of greater ease and less time consuming, than the earlier systems. Fluorescein angiography is a test which helps in the differentiation of retinal disease and is used to determine if laser treatment of the retina is warranted. Practical interpretation of fluorescein angiography images requires diligence and continual training of the clinician's eye. Sequential assessment of angiograms based on an understanding of retinal physiology proves to be the clinician's best aid for utilizing fluorescein angiography as a diagnostic tool.

Suggested readings

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