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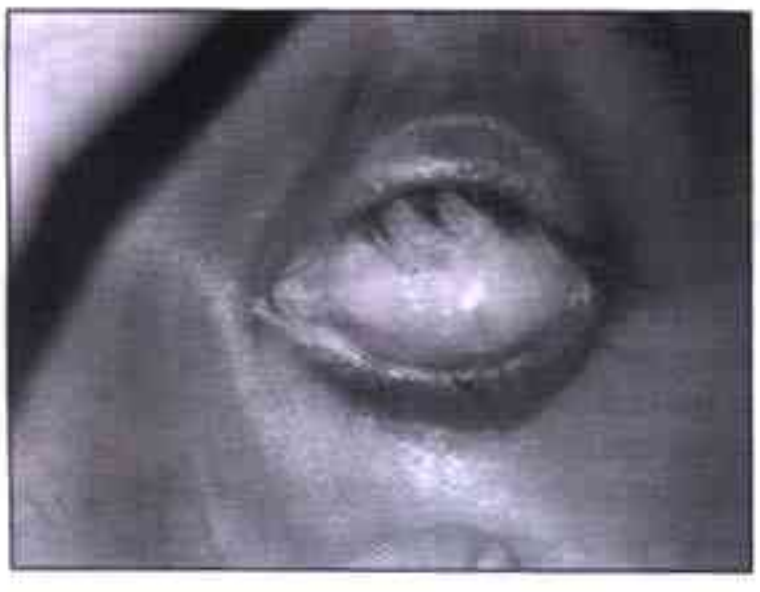
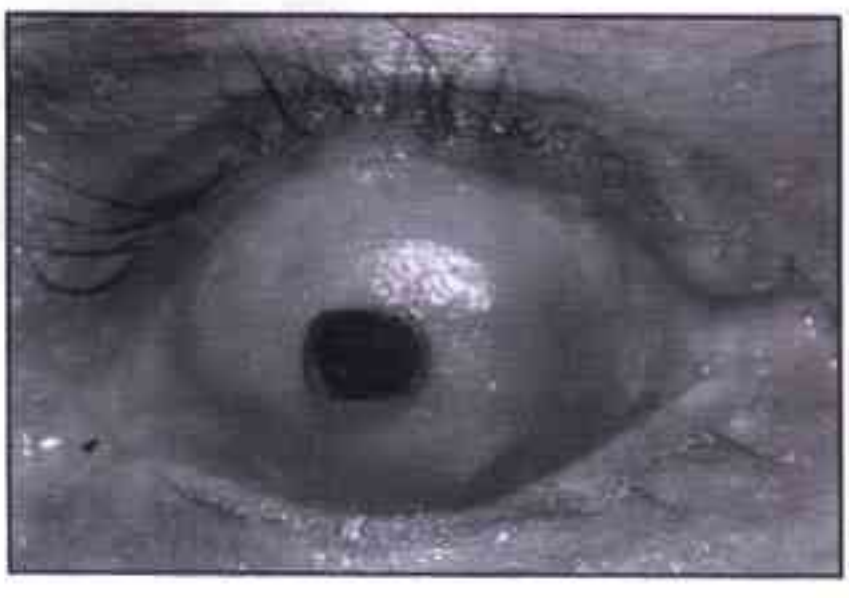


Mucous membrane in situ



Final appearance of the eye

SANKARA NETHRALAYA

EDITORIAL

This issue covers in depth in the perspective section the new technique of OOKP i.e. the osteo odonto keratoprosthesis, a new and exciting treatment offering hope for some forms of corneal blindness. Two case reports of an unusual orbital tumor and secondary infection in a patient with immunodeficiency are presented with detailed clinical features and photographs. High risk pediatric patients can be a challenge both to the anaesthetist and the treating ophthalmologist. Some important aspects of anaesthesia for the high risk infant is covered in the article with a few case illustrations. An intriguing muscle puzzle makes you put on your thinking caps.

Dr S Meenakshi
Editor

Perspective:

Osteo-Odonto Keratoprosthesis (OOKP) - the Indian Experience

Geetha K Iyer, Vinay S and Sitalakshmi G - Department of Cornea Services

HISTORY OF KERATOPROSTHESIS

The need for an alternative to conventional penetrating keratoplasty in patients with corneal disorders compounded by tear deficiency, vascularisation, and cicatrisation cannot be overemphasized. The search for an artificial cornea or keratoprosthesis has yielded several ideas and options, some viable and some not. Starting from glass implants we have moved on to synthetic hydrogels and biological tissues, each with their own advantages and disadvantages.

NEED FOR KERATOPROSTHESIS

Keratoprosthesis surgery is looked upon as the last resort for bilateral corneal conditions that are not amenable to conventional penetrating keratoplasty. These are as follows:

a. Stevens Johnson syndrome
b. Ocular cicatricial pemphigoid
c. Severe chemical injuries
d. Severe trachoma
e. Severe dry eyes
f. Multiple graft failures
g. Vascularised corneas

Being in the process of evolution, precise indications cannot be listed for the keratoprosthesis procedure and the above mentioned only serves as a guideline to the surgeon.

IDEAL ARTIFICIAL CORNEA

An ideal artificial cornea can be described as one that

a. surpasses the natural cornea in terms of:
   - optical quality
   - biointegration
   - freedom from infective agents
b. has unlimited inexpensive availability
c. mimics the natural cornea in terms of:
   - IOP measurement
   - drug penetration

TYPES OF KERATOPROSTHESIS

The keratoprosthesis could be broadly classified as those with

A. Biological skirt	B. Non biological skirt
a. Tooth root	a. PMMA
b. Tibial bone	b. HA (hydroxyapatite)
c. Cartilage	c. PTFE expander (polytetra-fluoroethylene)
	d. Hydrogel
	e. Dacron

OOKP

Among the many types of transcorneal devices developed over the last decades and made up of different materials, Strampelli's OOKP has probably been among the only one to overcome most of the the biointegrability problems conditioning to the performance of ocular mesoprosthesis. OOKP, a surgical intervention conceived by Strampelli in 1963, is a technique designed for patients with severe surface disorders who would be at high risk of graft rejection following penetrating keratoplasty. The prosthesis consists of a

section of one of the recipient's teeth (biological haptics made by osteodental lamina) supporting an acrylic optical cylinder. Because of the isolation of the implant from the recipient's ocular tissues, the rejection reactions against the prosthesis are very low.

The OOKP procedure involves two stages performed over a period of 6-9 months:

Stage IA: Tectonic reconstructive procedures (when applicable)

Stage IB: Mucous membrane graft

Stage IC: Preparation of osteodental alveolar lamina (ODAL)

Stage II: Implantation of ODAL on the bulbar surface

RATIONALE

Success depends upon the implantation of the buccal mucous epithelium covering the eye onto the alveolar dental ligament (periodontal ligament). A condition similar to that in the oral cavity, in which the alveolar dental ligament is in contact with the gingival ligament, is produced.

The alveolar dental ligament measuring 0.15-0.38 mm is present between the dentine and the alveolar bone and prevents the proliferation of the gingival mucosa by contact inhibition. The ligament is highly cellular and is composed of collagen fibres, blood vessels and nerves. In the oral cavity the ligament functions as a support for the tooth, transmits neural input to aid in mastication and has abundant sensory receptors. However, its primary role as a part of the ODAL is to keep the proliferating gingival mucosa in check, preventing it from extending between the lamina and the cylinder into the eye and forming a retroprosthetic membrane which could cause expulsion of the lamina. The protrusion of the ODAL beyond the ocular surface is crucial in that it causes proliferation of the mucous membrane bringing it in contact with the alveolar dental ligament thus causing contact inhibition. This prevents the growth of the membrane over the anterior surface of the cylinder. Thus of all the supports (both biological and non biological) for prosthetic corneal implants, the tooth appears to be a long-lasting solution reducing the rates of extrusion, which happens to be the most common cause for failure among the other prostheses.

CONTRAINDICATIONS

Though there are practically no contraindications to this procedure in a light perceiving eye other than an edentulous patient, the procedure is better deferred in case of the following:

1. retinal detachment
2. advanced glaucoma
3. mentally unstable
4. unavailability for long term follow up
5. the happily blind
6. unreasonable expectations
7. cosmetic reasons

The absolute contraindications being:

1. blind eye
2. edentulous patient

AGE

The procedure is preferably performed in patients over the age of 17 years. This allows for complete development and maturity of the patient's own tooth which can then be harvested for the procedure.

In children, the tooth has to be harvested from a live related donor necessitating the use of immunosuppression. The rate of bone resorption is high in these cases and these patients invariably require a second procedure using the host tooth in adulthood. There is a high risk of amblyopia in patients who have developed the visual loss prior to 4 years of age and might not benefit from the procedure.

PREOPERATIVE ASSESSMENT

A detailed preoperative ophthalmic, dental and psychological assessment is done following which the surgeries are scheduled.

PROCEDURE

STAGE I OOKP

In the first stage, the ocular surface is prepared by performing a superficial keratectomy with fibrovascular pannus removal. (Figs 1 and 2)

The surface is then covered using mucous membrane harvested from the buccal mucosa (Fig 3). This serves as a biological covering for the dental lamina. It provides adequate blood supply to the bone, protects the anterior surface of the lamina (haptic) and acts as a barrier against microbial invasion.

A single rooted tooth, preferably the upper canine is chosen for preparation of the lamina (Fig 4). The tooth with the surrounding alveolar bone is extracted (Fig 5). It is then fashioned into a lamina with bone on one side and dentine on the other (Fig 6 and 7). A central hole is drilled within the area of the dentine, into which the customized PMMA optical cylinder is cemented with acrylic resin (Fig 8 and 9). This is now referred to as the ODAL (Fig 10). This ODAL is then placed in a subcutaneous pouch in the orbitozygomatic area for the next three months to develop vascularisation and to promote the growth of connective tissue. A spiral CT scan is performed prior to stage II to rule out resorption of the lamina and to document lamina measurements.

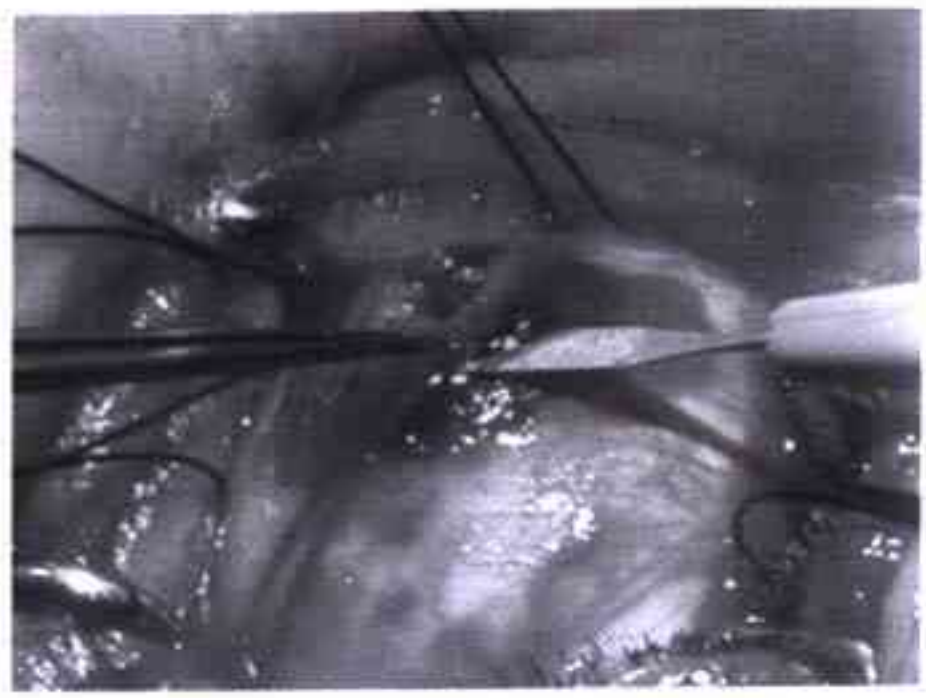


Fig 1. Superficial keratectomy

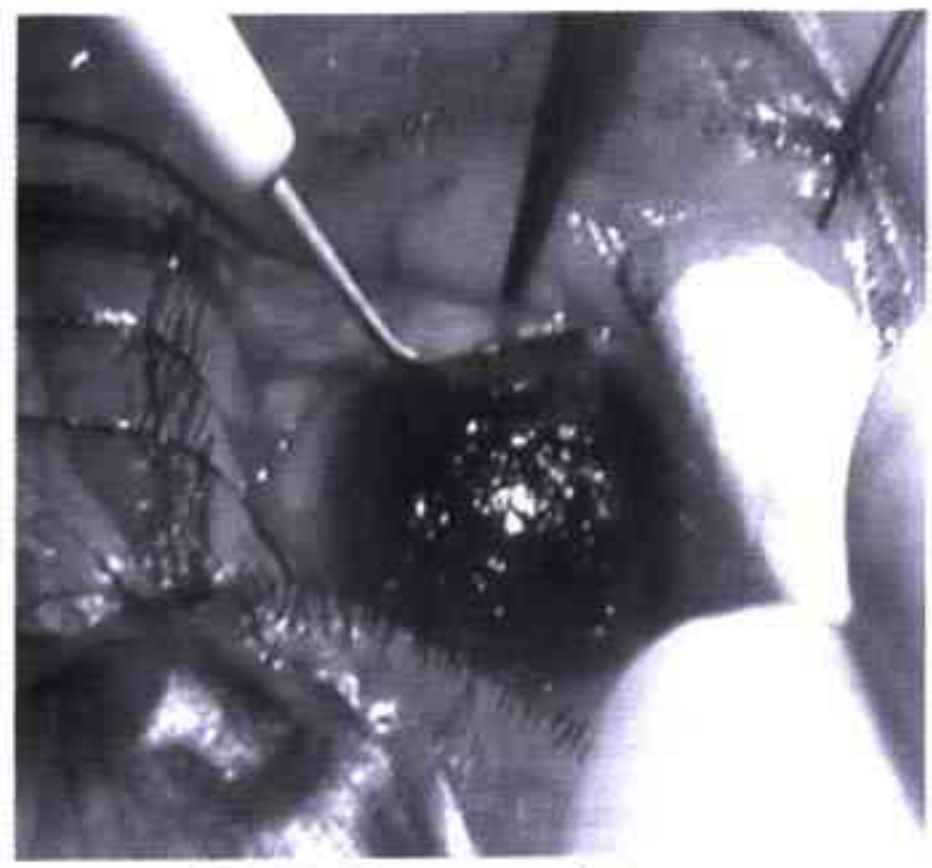


Fig 2 Exposure of stroma

STAGE II OOKP

The ODAL is first dissected off from the subcutaneous pouch and examined for its integrity prior to proceeding with the ocular surgery. Once the intactness of the lamina is confirmed, the ocular surgery is commenced by reflecting the mucous membrane (Fig 11).

The central cornea is trephined according to the dimensions of the optic cylinder. Intraocular procedure done includes total iridodialysis (Fig 12), intracapsular cataract extraction and anterior vitrectomy. Complete removal of these structures reduces the possibility of postoperative glaucoma and formation of retroprosthetic membranes.

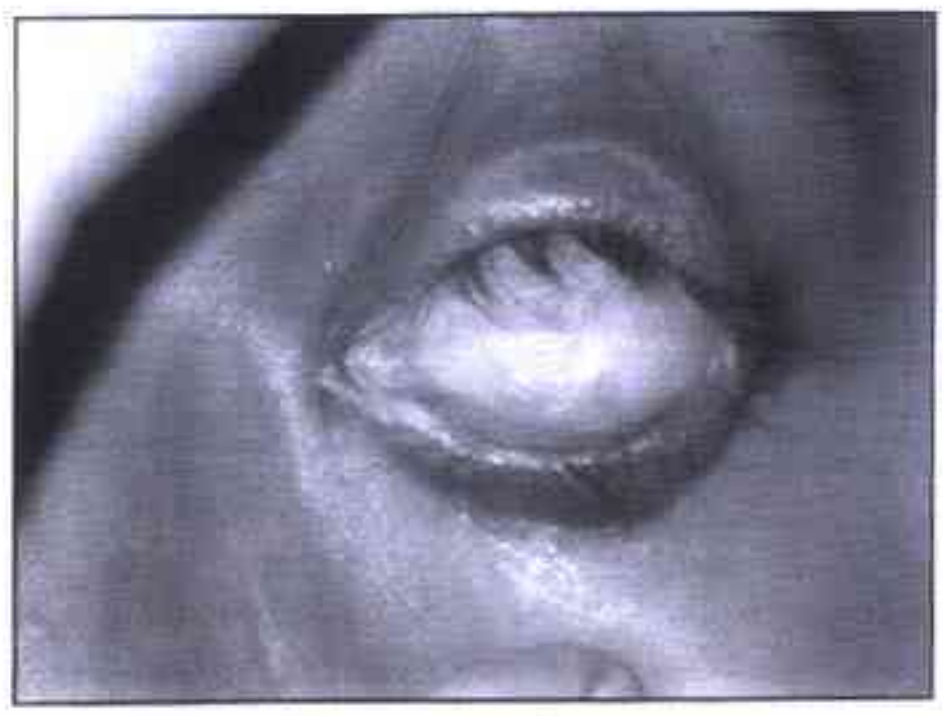


Fig 3. Mucous membrane in situ

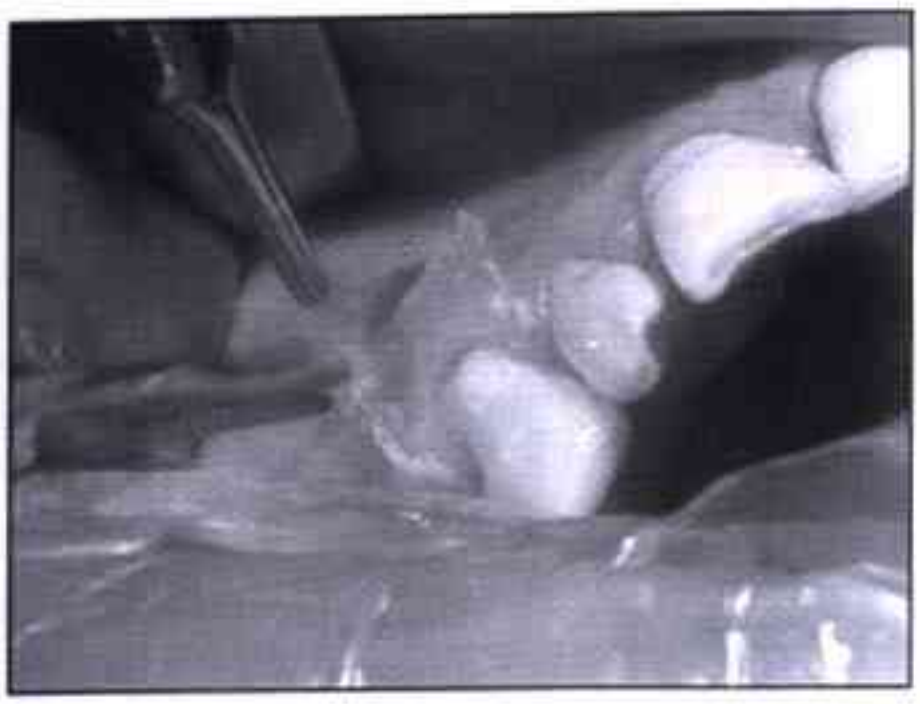


Fig 4. Tooth selected and mucosa reflected

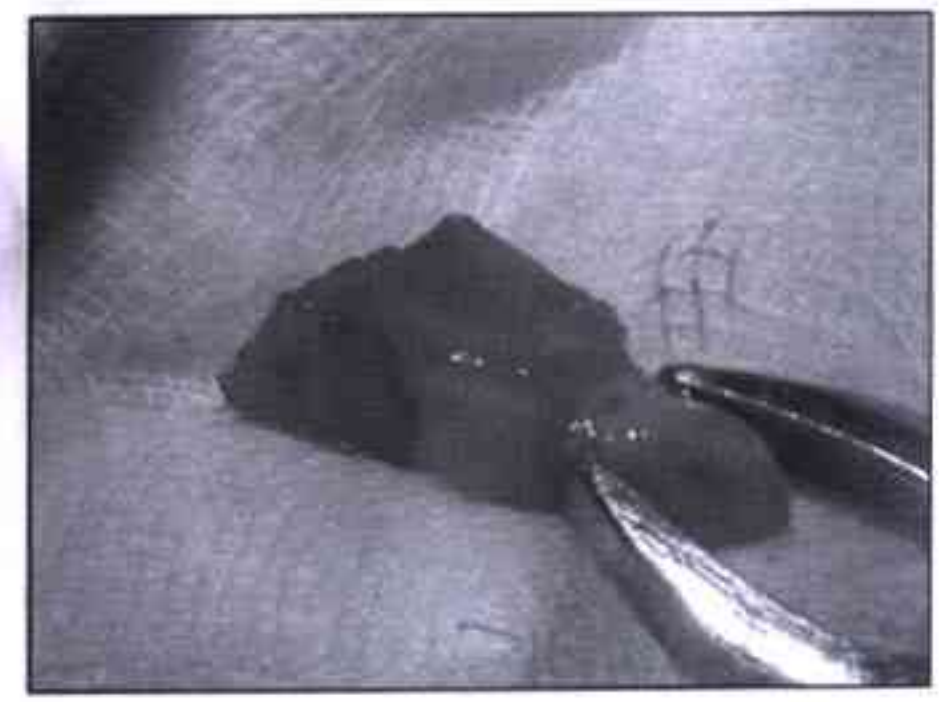


Fig 5. Removal of tooth with bone

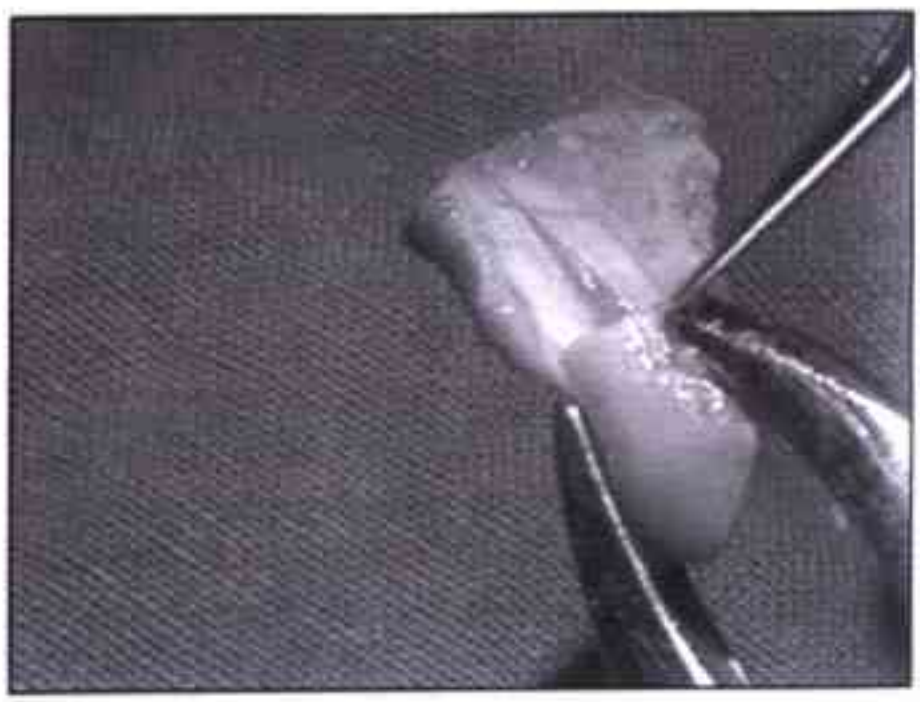


Fig 7. Exposure of root canal

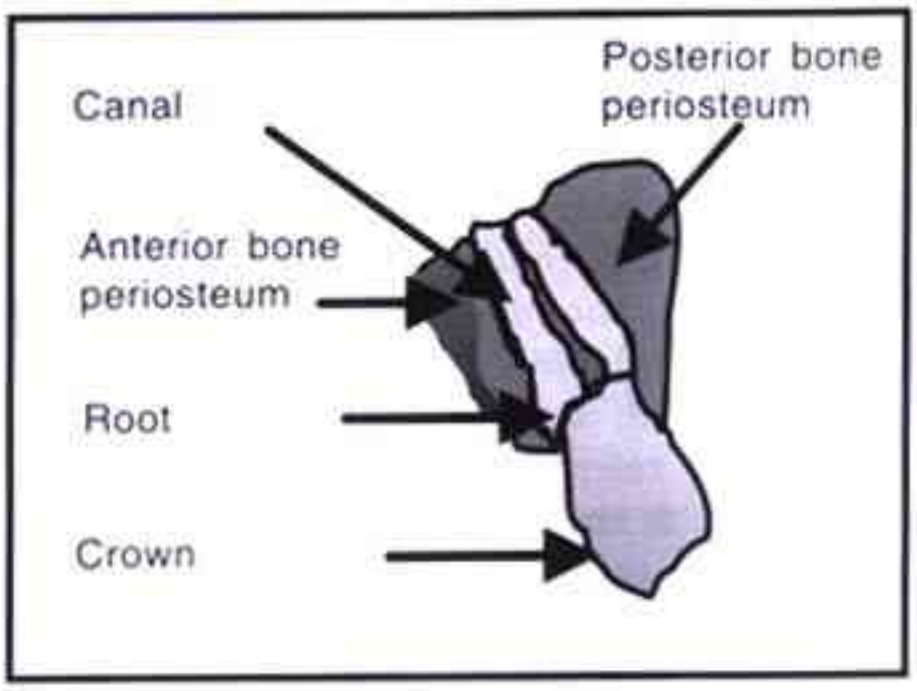
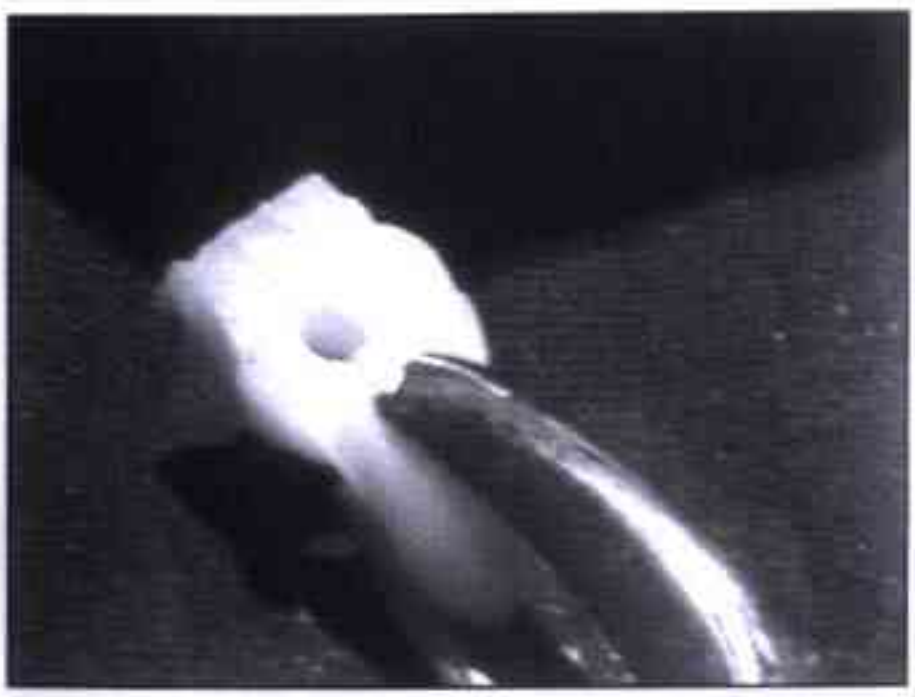


Fig 6. Schematic of adjacent photo



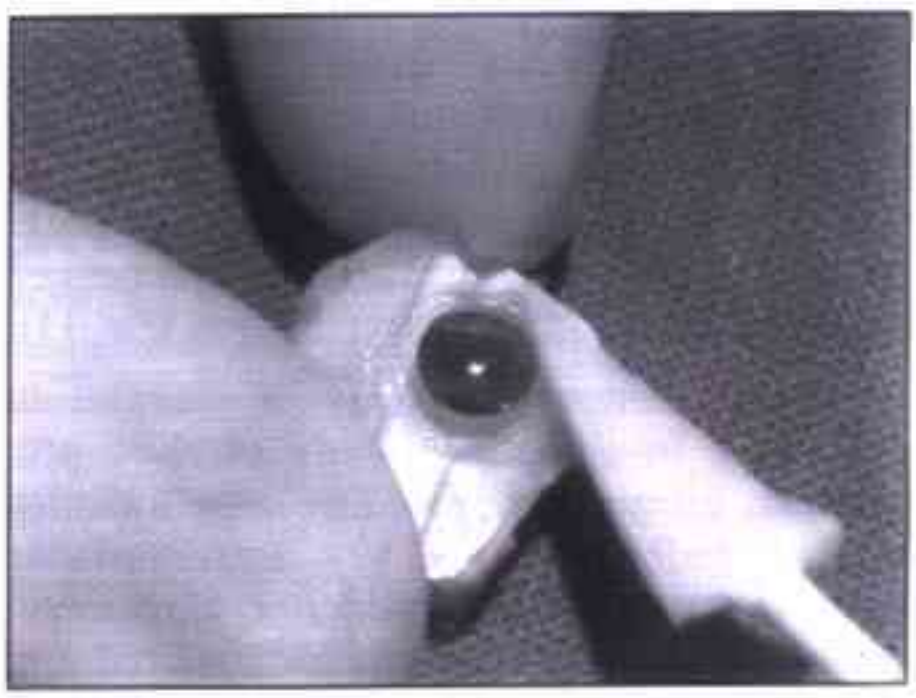


Fig 8 & 9. Drilling of hole with cementing of optic cylinder.

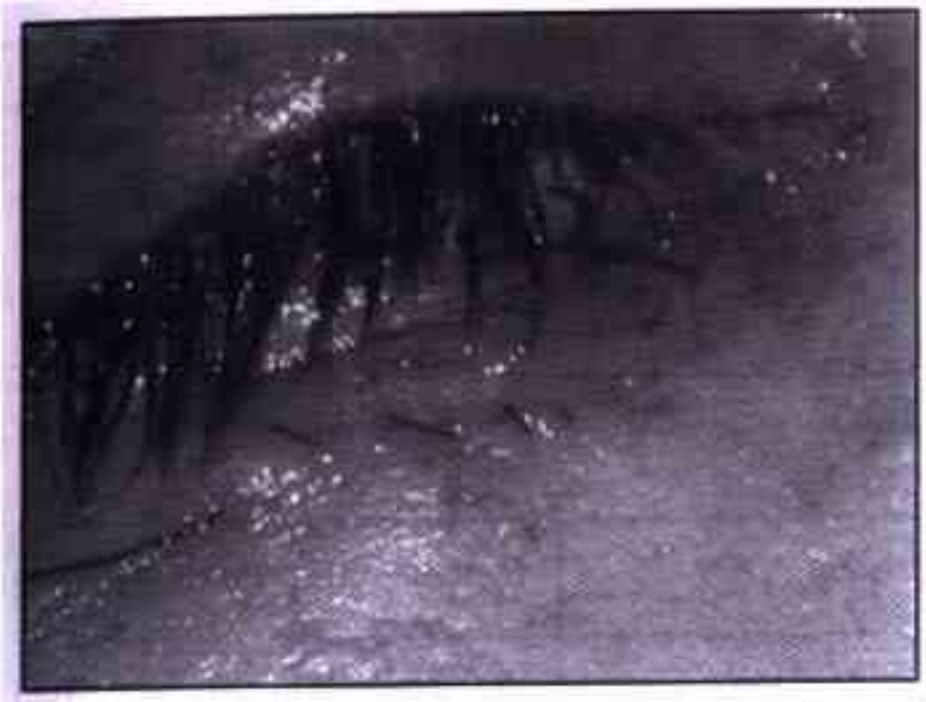


Fig 10.ODAL in subcutaneous pouch

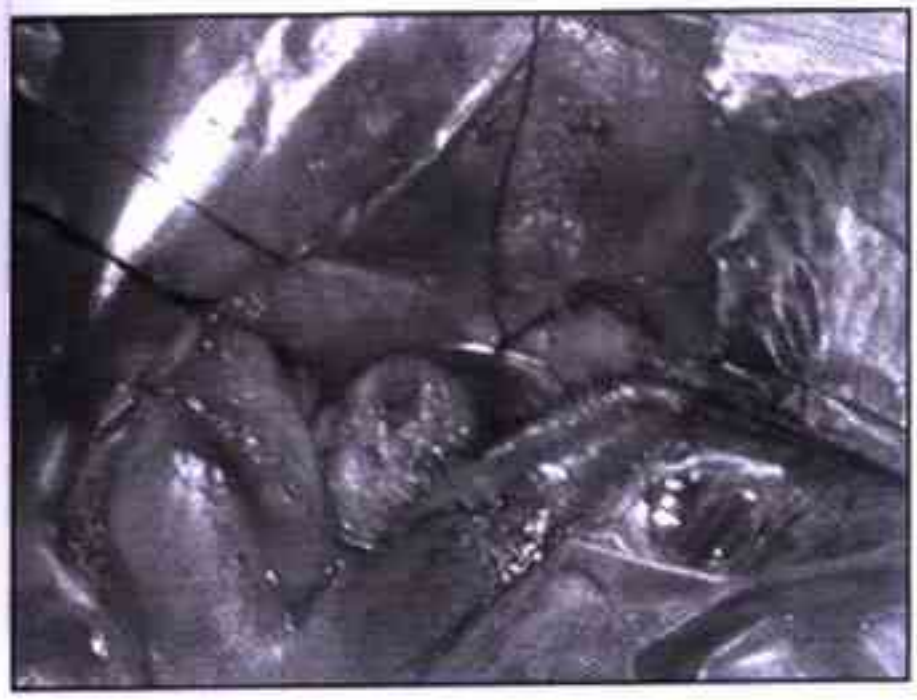


Fig 11. Mucous membrane reflected

Fig 12. Total iridodialysis

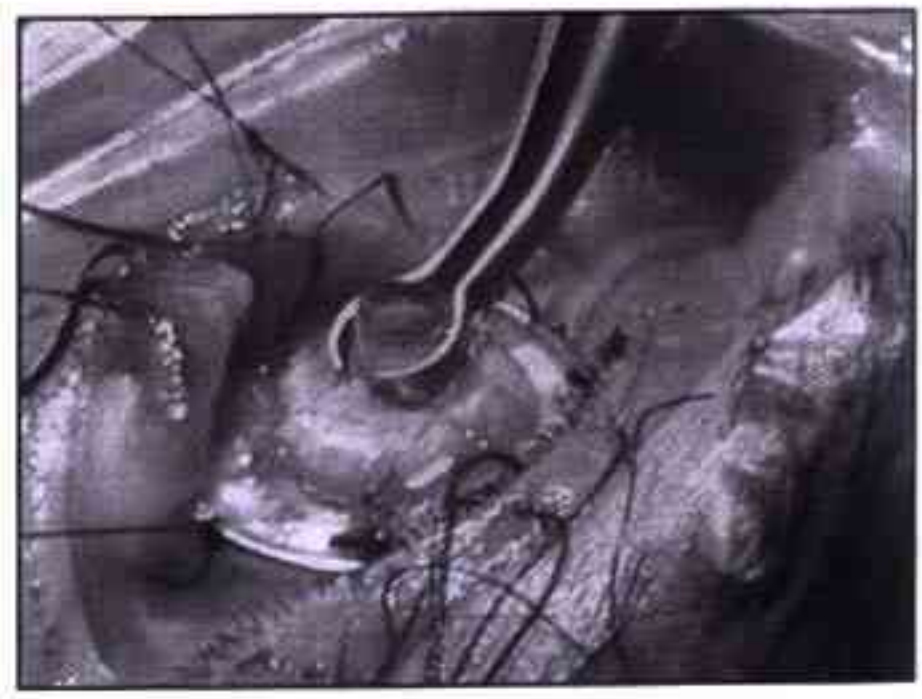


Fig 13. ODAL placed over the eye

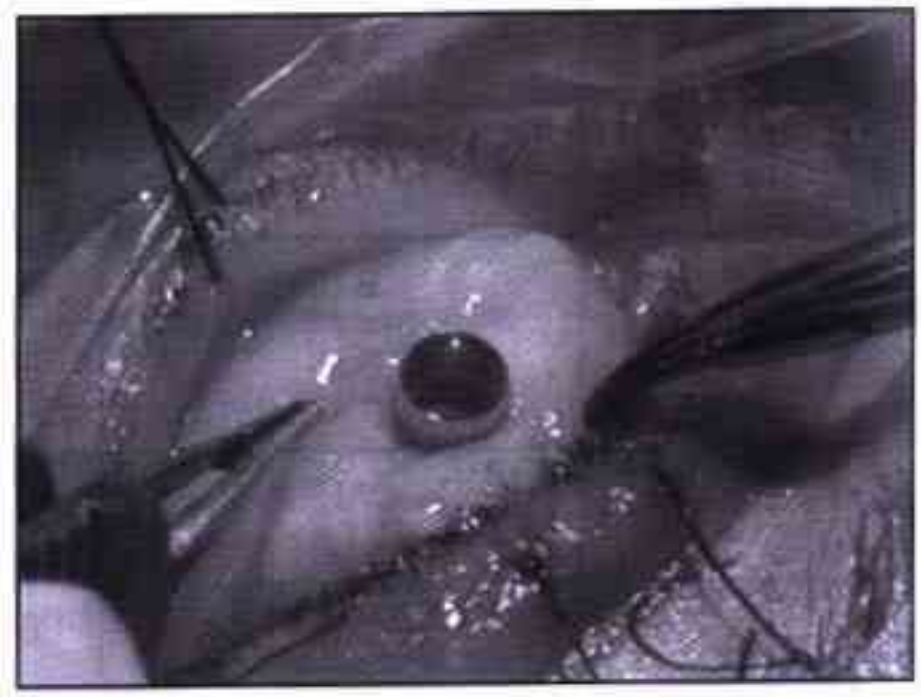


Fig 14. Mucus membrane reflected over ODAL

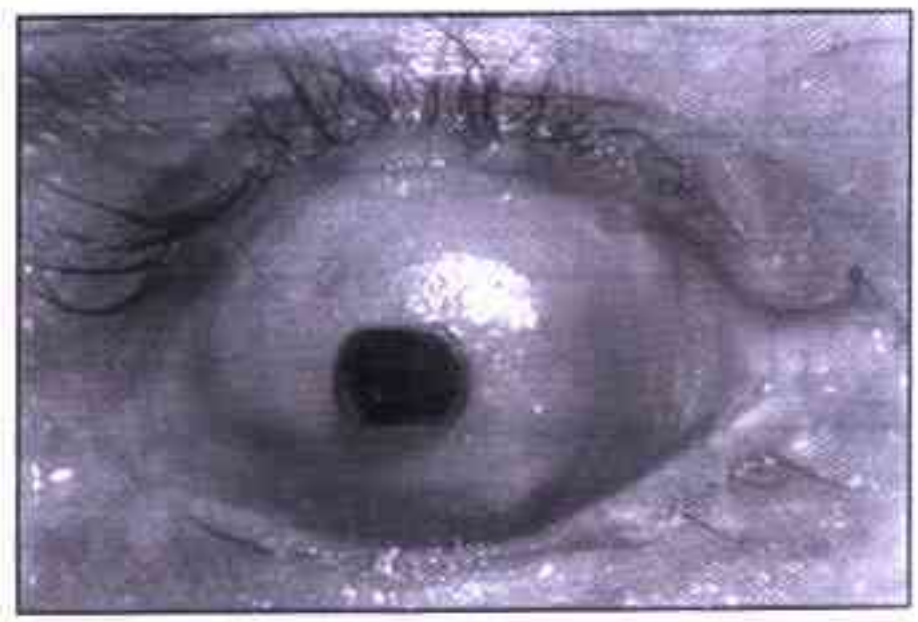


Fig 15. Final appearance of the eye

The lamina is placed with the cylinder centered over the corneal trephination and sutured (Fig 13). The centration is confirmed by indirect ophthalmoscopic finding of a well centered disc and macula and altered by appropriately placed tension sutures if required. The mucous membrane is finally reflected back on the lamina with a

trephination through which the anterior end of the cylinder protrudes out (Figs 14 and 15). A scleral shield with a central opening can be worn for cosmetic reasons.

CONCLUSION

The OOKP has stood the test of time in terms of long term results and complications. Despite the demanding and time consuming procedure that it is, the long term results have been satisfying and worth the effort in terms of helping these otherwise hopeless corneal blind patients in returning back to an almost normal routine.

At Sankara Nethralaya, 10 cases have been successfully completed with good results. The details are as tabulated below.

S.No.	Age (Years)	Primary disorder	Pre op VA	Refraction	Post op VA
1	39	SJS	PL+PR ACCURATE	+5.75DS +4.5DS	6/12. N6
2	50	OCP	PL+PRACCURATE	+3.00DS +3.00DS	6/7.5. N6
3	48	SJS	CFCF	+10.00DS +2.00DS	6/6. N6
4	23	SJS	PL+PR ACCURATE	+13.00DS	6/45. N36
5	20	SJS	PL+PR ACCURATE	+3.50DS +3.00DS	6/6. N6
6	23	CHEMICAL INJURY	CFCF	+0.50DS +3.00DS	6/6. N6
7	19	SJS	PL+PR ACCURATE	+1.00DS +3.00DS	6/9. N6
8	36	CHEMICAL INJURY	PL+PR ACCURATE	-5.50 DS +2.50 DS	6/5. N6
9	40	OCP	PL+PR ACCURATE	-3.50 DS +4.00DS	6/12. N6
10	23	CHEMICAL INJURY	PL+PR ACCURATE	+18.00 DS + 3.00DS	3/60. <N36

OCP- Ocular cicatricial pemphigoid; SJS- Stevens Johnson syndrome

The follow up ranged from 4 months to 32 months.

We are extremely grateful to Professor Giancarlo Falcinelli and Dr Giovanni Falcinelli for their initiation and continued guidance of the OOKP procedure at Sankara Nethralaya.

REFERENCES

1. *Ricci R, Pecorella I, Ciardi A, et al. Strampelli's osteo-odonto-kerato-prosthesis. Clinical and histological long term features of three prostheses. Br J Ophthalmol. 1995:79:129-134.*

2. *Caiazza S, Falcinelli G, Pintucci S. Exceptional case of bone resorption in an osteo-odonto keratoprosthesis. A scanning electron microscopy and x-ray*

microanalysis study. Cornea.1990:9:23-27.

3. Marchi V, Ricci R, Pecorella I, et al. Osteoodonto keratoprosthesis. Description of surgical technique with results in 85 patients. Cornea. 1994; 13:125-130.

4. Hille K, Grabner G, Liu C, Colliardo P, Falcinelli G, Taloni M, Falcinelli G. Standards for Modified Osteoodonto-keratoprosthesis (OOKP) Surgery According To Strampelli and Falcinelli. Cornea.2005:24:8:895-908.

Ocular syphilis presenting as vitritis in a patient with HIV infection

Amitabh Kumar, Chekitaan, Jyotirmay Biswas — Department of Uvea services

Introduction

The incidence of syphilis has been on the rise[1]. Co-infection with human immunodefiency virus (HIV) has serious implications for the pathophysiology and treatment of syphilis. Syphilis is an uncommon cause of uveitis in HIV infected patients accounting for less than 1% of this population[2] which is comparable to its incidence of 1.1% in the pre HIV era[3]. There are various manifestations of ocular syphilis in HIV infected hosts including iridocyclitis, papillitis, optic neuritis, branch retinal vein occlusion (BRVO), chorioretinitis, neuro-retinitis, intermediate uveitis, periphlebitis and serous retinal detachment[2-8]. Dense vitritis as the primary manifestation of ocular syphilis in HIV infected patients is rare. Syphilitic uveitis should be considered in all patients with rash and/or head ache where there is retinitis and/or retinal vasculitis or in any uveitis of uncertain origin. We report a case of a 33 year old HIV infected male who presented to our clinic with vitritis.

Case report:

A 33 year old gentleman presented to us in January 2006 with complaints of reduced vision in his left eye for the last three months. He complained of seeing floaters in front of his left eye since November 2005. His visual acuity in the left eye, then, was 6/18, N/36. He was treated with posterior subtenon injection of triamcinolone acetonide and was on oral steroids. Subsequently his vision improved to 6/9. He had another episode of reduced vision in his left eye along with anterior chamber reaction, vitritis, sclerosed vessels and discrete yellow deposits in the retina. Polymerase chain reaction (PCR) of the aqueous tap was negative for viruses (herpes simplex, herpes zoster and cytomegalovirus) and Mycobacterium tuberculosis. At presentation best corrected visual acuity (BCVA) in the right eye was 6/6, N/6 and the left eye was hand movements with accurate projection of rays. Slit lamp examination of the left eye revealed old pigmented keratic precipitates on the endothelium, and a reaction of 1+ flare and 1+ cells and a grade 3 relative afferent pupillary defect (RAPD). Intra ocular pressures (IOP) by applanation tonometry were 10 and 8 mm Hg in the right and left eyes respectively. A clinical diagnosis of intermediate uveitis (? Sarcoid) [figure-1] was made and the patient was sent for blood investigations which included routine blood, serum angiotensin converting enzyme and peripheral blood smear. All the blood reports were normal except a raised ESR (30mm at the end of one hour) and a negative mantoux test. He was put on tablet prednisolone 40mg / day. The patient was reviewed after a week when his vision had become worse to

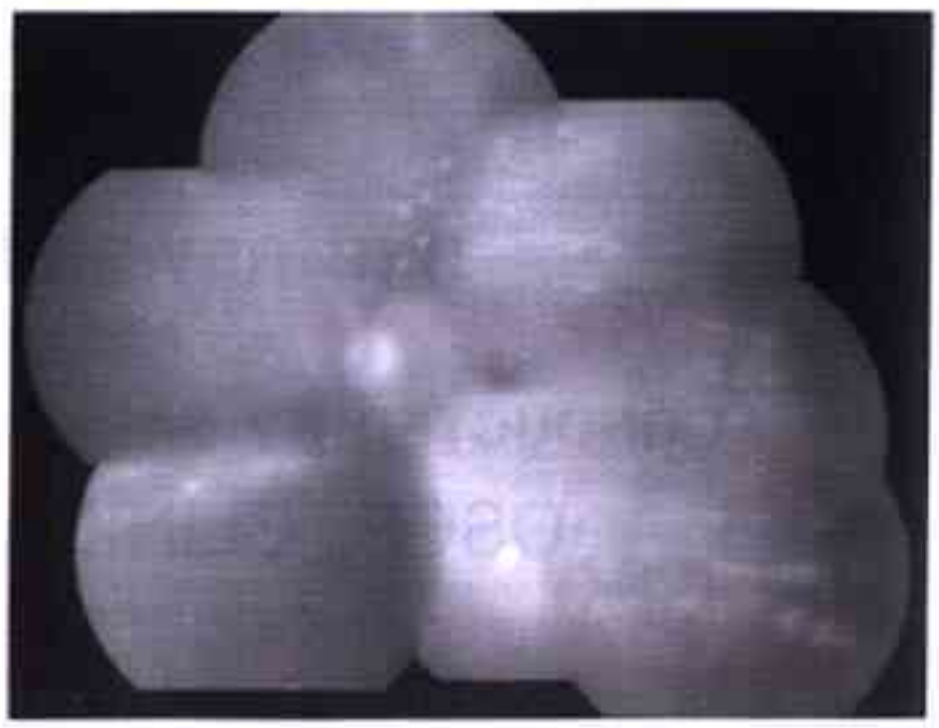


Figure 1: Montage photograph of the left eye at presentation showing dense vitritis and cotton ball vitreous exudates along with vascular sheathing.

inaccurate PR. The IOP were within normal limits. He had 1+ flare and cells along with vitritis. A differential diagnosis of endogenous endophthalmitis / masquerade syndrome was made. Enzyme linked immunosorbent assay (ELISA) for HIV was performed which detected anti HIV 1 antibody. It was confirmed by Western blot. The patient was referred to an AIDS care centre where he was found to be having positive reactive rapid plasma reagin (RPR) and Treponema pallidum heme agglutination test (TPHA). His CD4 counts were 187 cells/ micro litre. A vitreous biopsy was done and a microbiological examination grew Acinetobacter calcoaceticus. PCR of the vitreous sample for viral and fungal studies were negative. In view of positive tests for syphilis he was treated with crystalline penicillin G. 4 million units IV every 4 hours for 14 days followed by injection benzathine penicillin 2.4 million units IM weekly for three weeks as per the treatment guidelines for neurosyphilis along with HAART therapy. Two weeks later his vision improved to counting fingers. The same treatment was continued and at a subsequent visit a month later, vision in the left eye had improved to 6/60, N/18 [figure-2]. At the last visit (May 1, 2006) his vision was maintained. His latest CD4 counts were 800 cells/micro litre.

Figure 2: Photograph of the left eye post treatment showing a significant reduction in vitritis, a pale disc and sclerosed vessels.

Discussion:

HIV positive patients with syphilis may present with atypically dense vitritis. Moreover in some patients syphilitic vitritis may be the initial manifestation of HIV disease.

Ocular inflammation is often the only clinical finding in syphilis. In a recent retrospective study none of the patients with syphilis and HIV exhibited a vitritis on initial examination[1]. The optic nerve, retina and other neuroepithelial structures are embryologically derived from the CNS and the involvement of these structures constitutes neurosyphilis. Contrary to common belief involvement of the CNS can occur in all stages of syphilis[1]. CSF pleocytosis, one of the earliest and most sensitive findings in neurosyphilis may not be useful in HIV positive patients[9,10]. It is encountered much earlier in HIV infected patients. Given their findings and the difficulty in diagnosing neurosyphilis in HIV positive patients on the basis of serum and CSF some clinicians have advocated aggressive treatment of syphilis in HIV co-infected patients. Schaegel et al have reported 3 patients presenting with vitritis in a review of 28 patients with syphilis. The important differential diagnoses include toxoplasmosis, tuberculosis, sarcoidosis, leukaemia and ARN syndrome[11]. One must consider HIV infection in patients who exhibit ocular syphilis on initial examination[6]. Moreover atypical presentations of ocular syphilis such as vitritis in our case may arise because of co infection with HIV. Syphilitic uveitis in HIV has an altered clinical profile and a more severe course[5]. Panuveitis is commoner than isolated anterior uveitis[2]. Diagnosis of ocular syphilis should be considered in any HIV positive patient who presents with visual symptoms irrespective of the CD4 counts.

References:

1. *Margo CE, Hamed LM. Ocular syphilis. Surv Ophthalmol 1992;37:203-220.*
2. *Shalaby IA, Dunn JP, Semba RD, Jabs*

D. Syphilitic uveitis in human immunodeficiency virus infected patients. Arch Ophthalmol 1997:115:469-473.

3. Schlaegel TF Jr, Kao SF. A review(197-1980) of 28 presumptive cases of syphilitic uveitis. Am J Ophthalmol 1982;93:412-414.

4. Mc Leish WM, Pulido JS, Holland S, Culbertson WW, Winward K. The ocular manifestation of syphilis in the human immunodeficiency virus type 1 infected host. Ophthalmology 1990;97:196-203.

5. Becerra LI, Ksiazek SM, Savino PJ et al. Syphilitic uveitis in human immunodeficiency virus infected and non infected patients. Ophthalmology 1989;96:1727-1730.

6. Levy JH, Liss RA, Maguire AM. Neurosyphilis and ocular syphilis in patients with concurrent human immunodeficiency virus infection. Retina 1989;9:175-180.

7. Kleiner RF, Najarian L, Levenson J. AIDS complicated by syphilis can mimic uveitis and Crohns disease. Arch Ophthalmol 1987;105:1486-1487.

8. Radolf JD, Kaplan RP. Unusual manifestations of secondary syphilis and abnormal humoral immune response to Treponema pallidum antigens in a homosexual man with asymptomatic human immunodeficiency virus infection. J Am Acad Dermatol 1988;18:423-428.

9. Carey LA, Glesby MJ, Mundy LM, Janis EM, Hook EW. Lumbar puncture for evaluation of latent syphilis in hospitalised patients. Arch Intern Med 1995;155:1657-1662.

10. Gordon SM, Eaton ME, George R et al. The response of symptomatic neurosyphilis to high dose intravenous penicillin G in patients with human immunodeficiency virus infection. N Engl J Med 1994;331:1469-1473.

11. Stoumbos VD, Klein ML. Syphilitic retinitis in a patient with acquired immunodeficiency syndrome-related complex. Am J Ophthalmol 1987;103:103-104.

Muscle Puzzle

Mayee Rishikesh Charudatta and Sumita Agarkar
Sankara Nethralaya - ORBIS Pediatric Ophthalmology Learning and Training Centre

Twenty two year old male presented with history of drooping of upper lid and decreased vision in the left eye since childhood. No history of trauma, diplopia or previous ocular surgery.

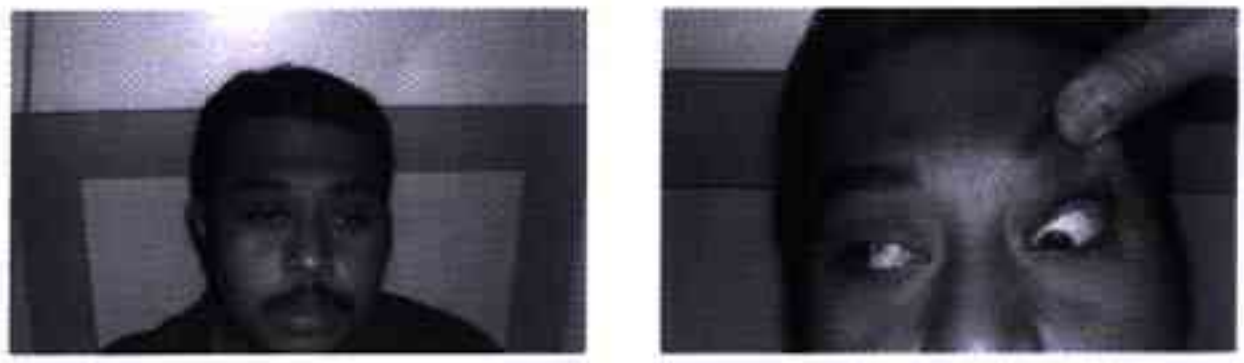
On examination, visual acuity was 6/6 (N6) in right eye and 6/24 (N6) in the left eye.

Pupillary examination was within normal limits. Anterior and posterior segment examination was also normal.

Ocular motility is shown in the photos below.

WHAT IS YOUR DIAGNOSIS AND PLAN OF MANAGEMENT?

(Answer on page 55)



Orbital Schwannoma - Clinical, Histopathological and Radiological Study of a Case

Priti Udhay, E Ravindra Mohan and J Biswas, Department of Oculoplasty and ORBIT

Schwannoma of the orbit is a rare tumour accounting for 1% of all orbital tumours. This is a slow growing tumour in adult occurring in 2nd to 5th decade.[2] We report a case of orbital schwannoma in a 63 year old female who had lower lid swelling for 5 years. The tumour mass was removed in toto through inferior orbitotomy. Histopathology confirmed orbital schwannoma.

Case Report:

A 63 year old female presented with slowly progressive right eye proptosis and swelling of lower lid, of 5 years duration and pain of 2 months duration (Fig-1). Best corrected visual acuity in the affected right eye was 6/6, N/6. She had a palpable, firm orbital mass in the inferior orbit. The eyeball was displaced upwards. Ocular movements were full in all gazes. Pupil examination, slit lamp examination, intraocular pressure and fundus examination were essentially normal. Ultrasonography B scan of the right eye showed an extraconal orbital mass in the inferotemporal orbit with high surface reflectivity and heterogenous internal echoes. MRI scan of the orbit showed a well defined oval soft tissue lesion in the inferior orbit between the inferior and lateral rectus displacing the globe superiorly. It displayed homogenous isointense signal on T1 weighted images and hyperintense signal on T2 weighted images with moderate contrast enhancement (Fig-2). The possibility of cavernous haemangioma was considered. She underwent complete excision of the mass through inferior orbitotomy using a swinging eyelid approach (Fig-3). Gross examination revealed greyish white encapsulated mass measuring 23mm X 17mm X 14mm. Specimen was cut into two halves.Cut surface was smooth (Fig-4). Histopathology revealed a tumour mass comprising of fibrous capsule and composed of solid cellular areas (Antoni A pattern) (Fig-5) and loose myxoid tissue (Antoni B pattern) (Fig-6). There were some areas of lipidisation. These findings were consistent with a diagnosis of orbital schwannoma. Post operatively, the patient had complete resolution of proptosis with excellent cosmesis.

Discussion:

Schwannoma of the orbit is a rare tumour. Within the orbit sensory and motor nerves can be affected. The most common site is the superonasal quadrant of the orbit There is no pathognomic feature of this tumour. The tumour can have a wide range of presentation. The disease occurs primarily in the adults. The tumour is slow growing and non invasive. There is minimal effect on adjacent structures. If the lesion is close to the optic nerve there can be pressure effect on the optic nerve causing optic atrophy, visual field defect, vascular engorgement and optic nerve head oedema. Schwannomas may be noted in association with von Recklinghausen's disease. Of various investigations, computerized tomography is quite useful. It helps in localization of the lesion. The tumour can be within the orbit or can extend from periorbital soft tissues or paranasal sinuses[2]. The configuration varies from round to oval. On CT scan, orbital schwannoma appears as a homogenous well-defined, minimally enhancing mass. Bony erosion of adjacent walls is seen in long-standing lesions. Heterogenicity is a feature

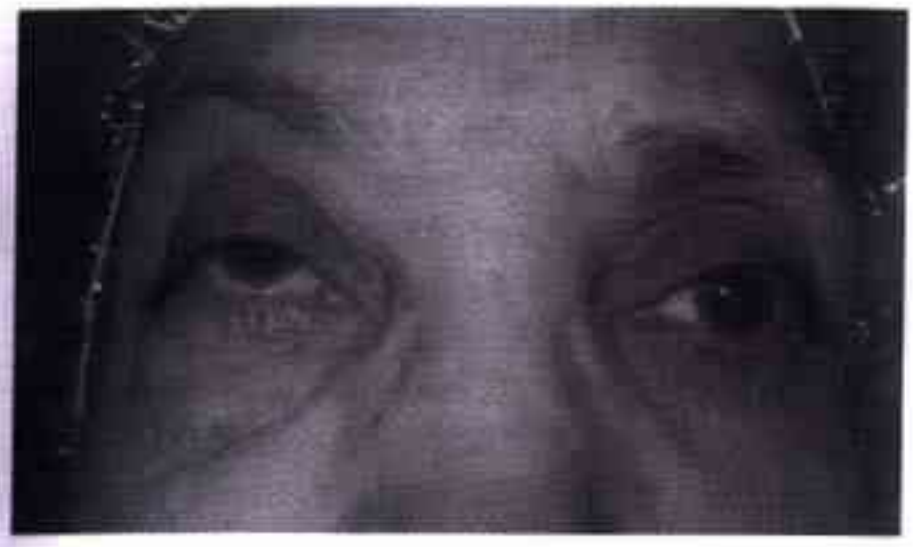


Figure 1: Photograph of the patient showing lower lid mass and superior displacement of the globe

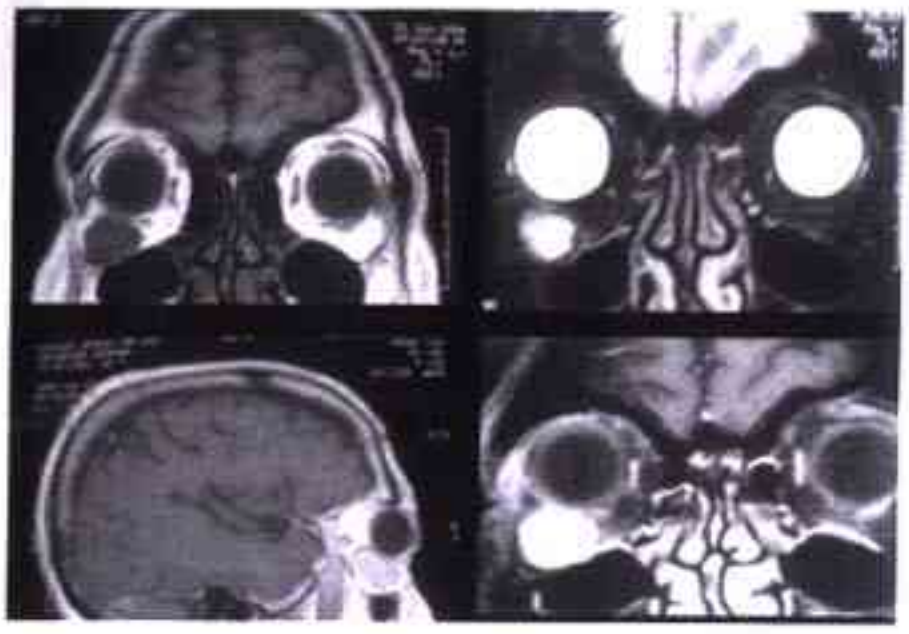


Figure 2: MRI showing isointense signal on T1 weighted images and hyperintense signal on T2 weighted images with moderate contrast enhancement

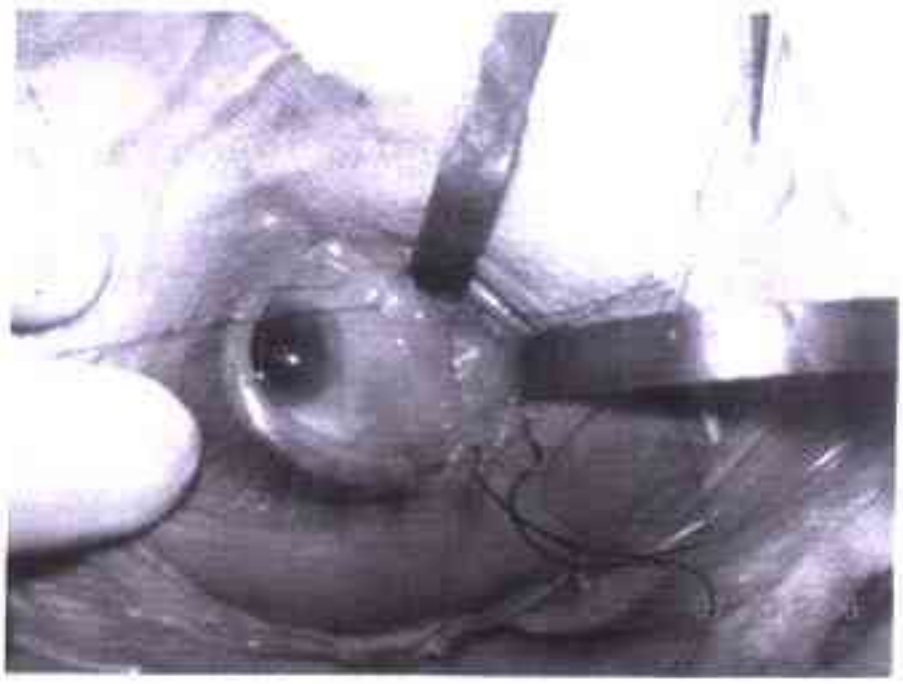


Figure 3: Intraoperative picture showing inferior orbitotomy using a swinging eyelid approach

Figure 4: Gross specimen showing encapsulated mass

Figure 5: Histopathology showing Antony A pattern of schwannoma (haematoxylin and eosin, x200)

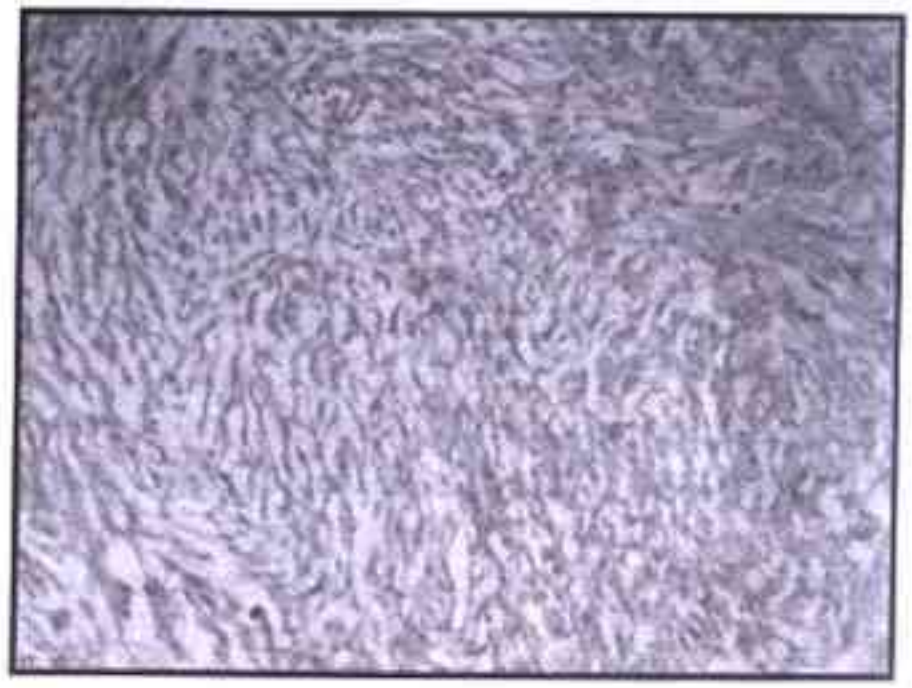


Figure 6: Histopathology showing Antony B pattern of schwannoma (haematoxylin and eosin, x200)

of cystic schwannomas. MRI tends to depict schwannomas as homogeneous masses. Schwannomas are typically isointense or slightly hypointense relative to gray matter on T1-weighted images and slightly hypointense to CSF on T2-weighted images. Gadolinium enhancement is typically homogeneous, although larger schwannomas can show areas of cystic degeneration and heterogeneous signal intensity. CT Scan and MRI findings are indistinguishable from neurofibroma[1]. Other lesions which it can mimic are cavernous haemangioma, meningioma and haemangiopericytoma.[2] In fact, our case was diagnosed by radiologist as cavernous haemangioma. The tumour can be differentiated from thyroid ophthalmopathy as it tends not to follow a muscle or mimic muscle enlargement as seen in thyroid ophthalmopathy. It can be distinguished from orbital pseudotumour as it does not usually extend into and obscure the apex of the orbit.[2]

Schwannoma is a benign proliferation of Schwann cells that envelope peripheral nerves. Histopathologically they are encapsulated by the perineurium of the nerve of origin. Such perineurium can be seen in the capsule sometimes eccentrically. Classic feature is the alteration in the same tumour. Solid cellular areas which is called Antoni A pattern with areas of looser myxoid tissue having stellate or oval cells within a mucinous background which is known as Antoni B pattern. Such Antoni A and Antoni B pattern was seen in our case. In addition, one may also see Verocay bodies which comprises of highly regimented organization of picket-fence nuclei with imbricated polar cytoplasmic processes. Reticulin stain can nicely demonstrate abundant fibres present in schwannoma. Immunohistochemical study with S-100 stain is also positive in this tumour.[1] However, light microscopic features are often so typical that immuno-histochemical study is rarely required. Cystic changes can be seen in schwannomas.

We have reported earlier a series of 4 cases of cystic schwannomas.[3] Schwannomas are less often multiple than neurofibromas and can be easily removed. If removed incompletely chances are high that it will recur. Recurrent lesions can be malignant. Schwannomas are benign tumours but there are atleast two reports of malignant tranformation of orbital schwannomas. This makes long-term follow-up of these patients necessary.

Surgical removal is the treatment of choice in orbital schwannomas. Location of the lesion indicates what type of surgical approach is required anterior, lateral, or combined lateral orbitotomy and frontal craniotomy. Complete removal ensures prevention of recurrences. Exenteration is recommended for malignant schwannomas.

We feel, though rare, primary orbital schwannoma should be kept in the differential diagnosis of all slow-growing soft tissue orbital or periorbital masses. CT scan or MRI is most helpful in identification and investigation of the lesion. Although no pathognomic feature exists, surgical removal is the treatment of choice.

References:

1. *Ophthalmic Pathology:An Atlas and text book 4th edition.Edited by Spener W H . W . B . S a u n d e r s Company,Philadelphia.1996.PP2645-2650.*
2. *Rootman J, Goldberg C, Robertson W. Primary Orbital Schwannoma.B. Br J Ophthalmol. 1982; 66:194-204.*
3. *Subramanian N, Rambhatla S, Mahesh L, Menon SV, Krishnakumar S, Biswas J, Noranha OV. Cystic schwannoma of the orbit-a case series .Orbit 2005 :24:125-129.*

Muscle Puzzle

(Answer to Muzzle Puzzle on page 51)

The ocular motility shows left exotropia and left hypertropia with ptosis in primary position. Exotropia increases in right gaze and reduces in left gaze. However, there is left upshoot in left gaze and downshoot in right gaze with narrowing of palpebral fissure on attempted adduction. The motility is suggestive of Duane's retraction syndrome- type 3 in the left eye.

Duane's (Stilling-Turk-Duane) syndrome constitutes 1-4% of all strabismus entities, more common in females (60%) than males and on left side (60%) than right side. Various etiopathogenetic mechanisms have been proposed like anomalous co-contraction of horizontal recti, structural anomaly in rectus insertion, hypoplasia of the 6th nerve nucleus and teratogenic malformation of rectus muscles.

Huber classified the syndrome into 3 types: type 1 (limitation in abduction), type 2 (limitation in adduction) and type 3 (limitation in both) in addition to narrowing of palpebral fissure on attempted adduction. Type 1 is seen in 78%, type 2 in 7% and type 3 in 15% cases. Bilaterality is seen in 18%.

Associated upshoots and downshoots (Lisch phenomenon) are believed to be due to slippage of globe under contracting rectus tendon.

Diagnosis is clinical, but electromyopic and saccadic velocity testing show reduced amplitudes.

Surgery is indicated for noticeable horizontal squint, abnormal head posture, marked globe retraction and cosmetically unacceptable up/down shoot. Various muscle weakening procedures have been tried: recession for horizontal deviations, transposition of vertical recti for abduction deficit and Faden and Y-split of lateral rectus for up/down shoot.

Anaesthesia for High Risk Pediatric Patients

Ian Sundarraj, Department of Anaesthesiology

Risk is defined as the possibility of incurring misfortunes or "losses "during the administration of anaesthesia. In general surgery in most instances of high risks takes a risk against their life for a life saving surgery. If they do not undergo the corrective surgery it may endanger their life. But in ophthalmic surgery it is the other way. Even if they don't under go the surgery still they may live a long life, however a life of poor quality due to poor vision.

Hence an ophthalmic anesthetist has more responsibility in taking any decision in accepting a high risk case. He has to gather all information about possible risks and must take preventive steps to manage them. The steps should include the following.

1. Risk identification: What will go wrong? How it could happen?
2. Risk analysis: How likely is an adverse reaction? How serious it could be?
3. Risk elimination: How can risk be eliminated or its effect reduced?

Taking a clinical decision balancing the risk and its benefits and deciding if it will be life or vision is not easy.

Anesthetist and the surgeon have to consider the following:

1. How necessary is the surgery in this case?
2. If all goes well is the patient likely to gain the real benefit ?
3. What will be the impact of surgery on the quality of life ?
4. What are the usual risks ?
5. Does the medical condition of the patient increase the degree of risk ?

He should consider whether it would be helpful to involve other specialties like the Cardiologist, Nephrologist, Endocrinologist, Neurologist, and Neonatologist & Pediatrician in the assessment of patient to decide on risks and benefit.

All counseling must be done in a way the childs parents or guardian can grasp in the language they can understand well.

Critical attention has to be paid to cardiovascular and respiratory functions in the preoperative period. Patients may benefit from preoperative evaluation and treatment of cardiovascular and respiratory system and electrolyte imbalance. Monitoring in the perioperative period with monitoring of vital functions can detect early any perioperative problem for its timely correction.

In High Risk Patients special consent has to be signed and obtained with witness after explaining to the patients, parents or guardian the following.

a. What the procedure involves.
b. Type of anesthesia and its risks
c. Specific risks due to the co-existing medical problems
d. Choice of alternative treatment and anesthesia if available.
e. If any medical complication occurs are the parents prepared for transferring the child to any other hospital where facilities are available

Four High Risk cases are presented below

Case No. 1

History of Congestive cardiac failure 34 days after birth due to hypocalcaemia. Seizures with last episode 1 month back, history of sepsis, thrombocytopenia, myocarditis secondary to hypocalcaemia, hypomagnesemia, hypoparathyroidism.

Investigations included Birth Wt.2.7 kg. (mother was on Eltroxine for hypothyroidism). Hb.10gm/%. Blood smear-hypochromic, anisocytosis. Platelet count 3.10.000.

ECG and Echocardiogram showed HR.150/min. normal sinus rythm. PR interval 0.12 sec. No ST-T abnormalities. Normal RV & L:V contraction & functions. Stroke Volume 5 cc. LVEDV 70 cc. LVESV 2cc. LVEF 75 %.

Medical Management included Syrup shelcal 5ml tds. Rocaltral (dissolved in water). Syrup Domstal 0.5 ml tds. T. Gardinal 30 - ½ dissolved in shelcal. Syrup Fevovit 3 drops od.

First Surgery

Anaesthesia was induced with Sevoflurane 4%. IV line was secured. IV - Ketamine 5mg and Norcuron 0.5 mg were administered. Direct vision intubation with 3.5 mm red rubber tube;maintained with Isoflurane 0.4%, and manual ventilation. Recovery was achieved with glycopyrrolate. 0.04 mg and neostigmine. 0.2mg IV. Patient recovered well

Second Surgery was a week later Anaesthesia induction with 4% Sevoflurane. IV line secured. Isolyte P was administered.

IV 0.5 mg Ketamine and Norcurone 0.5 mg were administered. Intubation and maintanence and recovery was as before.

Case No. 2

STORMY BIRTH HISTORY

History of galactosemia, proximal renal tubular dysfunction, hyperkalaemia, mild pulmonary valvular stenosis, acquired haemolytic anaemia, dehydration (post diarrhea)

Normal full term - B.Wt. 3.5 kg. H/O jaundice after birth, poor weight gain and delayed development, history of anaemia; received blood transfusion, treated for loose motion.

On examination, found to be marasmic, pallor, roving eye movements, anterior fontanelle was depressed.

Cardiovascular system examination revealed a soft systolic murmur. Respiratory system was normal. Abdominal exam showed a distended liver of 4 cm.

Investigations included urine albumin, sugar - Nil, Hb.11.1 gms % PCV 33 % platelets adequate, Smear - Hypochromic, anisocytosis, RBS 48 mg % Blood urea 27 mg/dl, SGPT - 34 SGOT - 29, Electroyte Na+ 14.6, K+ 6.4 Cl - 102 HCO 316 meq /l, ECG - mild pulmonary valular stenosis.

First Surgery

Lensectomy OD. Theoped 3 drops started 2 days prior to surgery. Patient was premedicated with Atropine 1 /5th cc IM; mask induction achieved with Sevoflurane 0.5 to 2 %; IV line was secured. Atracurium 2.0 mg and 0.5 mg.later administered. Patient was intubated with 3.5 mm endotracheal tube and manually ventilated.

Oxygen desaturation occurred. Endotracheal tube was adjusted, stomach decompressed and Deriphylline 0.125 mg, Decadron 0.75 mg. IV administerd. Anaesthesia was maintained with 0.5% Sevoflurane. Reversal was achieved with glycopyrrolate. 0.04 mg and neostigmine. 0.2mg IV. During surgery nitrous oxide was discontinued to prevent distension of abdomen. Child recovered well, observed for 2 hours in ICU was fed Soya milk; had no vomiting, and was transferred to ward.

Second Surgery was a week later and lensectomy of the other eye.

Premedication and induction was similar to the first surgery. Atracurium 2.0 mg followed by 0.5 mg, Deriphylline 0.125 mg and Decadron 0.75 mg was given. Oxygen desaturation occurred which resolved with adjustment of the endotracheal tube. Reversal was achieved uneventfully in the usual manner.

Case No. 3

Past medical history was significant for the presence of a patent ductus arteriosus, acute gastroenteritis, hypernatremia (Na 178

mEq / l)), metabolic acidosis (bicarbonate 6 mEq / l) Dehydration. Hyperglycaemic -Blood Glucose 204 mg /dl. Hypocalcaemica- S.Cal 6.5 mg/dl. Anaemia and thrombocytopenia. Blood culture had shown no growth. USG Brain showed mildly dilated LV with Gr III Intraventricular Hge with old clot. - Gasping respiration and severe retraction with respiratory rate of 70 / min.

Electrolyte correction with IV fluid 5% Dextrose & Sodium bicarbonate had been given and baby was ventilated on IMV mode for 24 hours.

Birth History include Low birth weight - (1.75 kg). Premature by 2 weeks. Neonatal jaundice. Intracranial haemorrage - Gr. III. Had been diagnosed as Congenital Rubella Syndrome.

Investigations include Haemoglobin 16.5 gms%. PCV 49 %. Platelet Count - 1.25.000 mm^3.

Total Bilirubin -3.5 mg / dl. Direct Bilirubin - 0.5 mg / dl. Indirect Bilirubin - 3.0 mg / dl. Random Blood Sugar - 106 mg / dl.

Anaesthesia

First surgery was an examination under anaesthesia followed by external trabeculotomy of the right eye for congenital glaucoma. The child was started on Theoped drops - 2 twice daily for 2 days. The child was premedicated with 0.1mg intramuscular Pedichloride 1/4 tsp.

Inhalational induction was with sevoflurane. The child was paralysed and Intubated on third attempt. The child was then manually ventilated with sevoflurane, Norcuron - 0.3 mg (wt. 3.5 kg). At the conclusion of the surgery reversal was achieved with glycopyrrolate 0.03 and neostigmine 0.15 mg. Baby cried well after extubation. Baby was observed in ICU, breast fed and shifted to ambulatory ward after he sucked well during feeds.

The second surgery was two weeks later which was a lensectomy for a congenital cataract. This time anaesthesia was induced with Nitrous, Halothane and Oxygen. Intubation was achieved on third attempt. Baby was administered Norcuron 0.4 mg and was manually ventilated. After completion of surgery during fundus exam the child went into bradycardia leading to cardiac arrest and could not be revived.

Case No. 4

Child had congenital acyanotic heart disease i.e. ASD, VSD, and moderate valvular pulmonic stenosis, failure to thrive, bilateral small kidneys, electrolyte imbalance, cardiac failure, persistent tachycardia and hurried respiration.

On examination the baby was afebrile, there was hurried respiration and subcostal in- drawing of chest and systolic murmur over the chest

Investigations included Hb. 12.4gm . PCV 37 %. RBS 105 mg / dl. TC 5600 /mm^2. Blood Urea 20 mg / dl. Serum Creatinine 0.4 mg/ dl. Antibodies for Rubella Virus - ELISA Ig G - High Positive - IgM Low Positive

Electrolytes

	21-01-2004	30-01-2004
Sodium	125 meq / L 1	128 meq / L
Potassium	2.8 meq / L ?	4.1 meq / L
Chlorides	92 meq /L	93 meq /L
Bicarb	25 meq / L	25 meq / L

Anaesthesia

The first surgery was lensectomy for congenital cataract. Patient was induced with sevoflurane, nitrous oxide and oxygen and maintained with sevoflurane. Intubation was possible at first attempt. Surgery lasted one hour. Recovery uneventful.

The other eye also underwent a lensectomy one week later. Anaesthesia induced with sevofluorane. Intubation was possible in the second attempt. Patient developed bradycardia. Sevofuorane was changed to Isoflurane. Patient was manually

ventilated without muscle relaxant. Recovery was good.

Discussion :

1. Preoperative care may include
   A) Prematurity (Apnea Neonatorum)- advise Theophylline drops for two days prior to surgery.
   b) Low Birth weight (Hypothermia) - keep the child warm.
   c) Congenital Defects (Airway abnormalities may be present) - anticipate Intubations difficulties.
   d) Congenital valvular Heart Disease - administer antibiotic prophylaxis.
2. Preoperative treatment and stabilization by pediatrician, management of electrolyte imbalance due to vomiting, diarrhea, by fluids and antibiotics.
3. Management of metabolic disorders: Hypocalcaemia, hypokalaemia and hypoglycaemia.
4. Difficult Venous Access - All veins punctured for IV infusions and blood samples; trans illumination may help identify the veins.

CONCLUSION

The choice of the anesthetic technique and the choice of the anesthetic drugs and analgesic will depend on the individual child's physical status and the experience of the anesthetist.

We feel it will be wiser to avoid respiratory depressants, longer acting muscle relaxants and hepatotoxic anesthetics. Drugs like Fentany for pain relief, relaxants like Atracurium and Anesthetics Isoflurane and Sevoflurane are our choice.We prefer manual ventilation rather than mechanical ventilators with which we will be able to feel the lung compliance.

Inspite of ECG, HR, SpO2 monitoring we still prefer continous auscultation of the heart and lungs during surgery (displacement of the endotracheal tube, obstruction of the tube due to kinking of the tube or obstruction due to blood or secretions in the endotracheal tube can be found out earlier by continuous auscultation by a stethoscope placed over the precordium)

Intravenous line is a must for all cases for not only fluid maintenance but also for administration of drugs and most of all for an emergency.

The inherent danger in ophthalmic anesthesia is the habit of freely using eye drops like "DROSYN". (T' Plus which has phenylephrine) before or during anesthesia cousing sudden rise in heart rate, blood pressure and ventricular arrhythmias. Bradycardia due to Oculocardiac reflex and cardiac arrhythmias may also require early treatment.

Airway problems with displacement of endotracheal tube, blocking with secretions or blood resulting in oxygen desaturation are not uncommon. Hypothermia of the risky child in cool operation theatre has to be taken care of with electric warmers and thermal drapes.

We prefer extubation after reversal and recovery with stable vital signs and good oxygen saturation.

Maintenance of the anesthetic level should not be decreased at nearing the end of surgery especially if the surgeon is going to do a fundus examination after the surgery. Lighter plane of anaesthesia can lead to bradycardia when the eye is handled during the exam which may lead to even a fatal cardiac arrest in a high risk child.

Before extubation all equipments must be ready for reintubation with emergency drugs loaded in syringe.

The child is disconnected from the OT monitors & shifted to the Post Operative ICU inside the OT and administered oxygen by mask and kept warm. A trained nurse takes care of the child.

The child is kept in the ICU for two hours and transferred ton the room with no laryngeal spasm, an unobstructed good breathing, good suctioning and cleared orophryngeal airway, gastric deflation, with good movement of all four limbs, and normal SpO2 & ECG tracing, and with a good cry.

If the child doesn't make a good recovery the child is transferred to ICU of any well-equipped pediatric institute for further management.

INSPITE OF OUR BEST EFFORTS & CARE MISFORTUNES OCCUR

All we have to do is to obtain senior advice at the earliest opportunity.

Inform relatives in detail immediately after any procedure from which a patient may not make a straight forward recovery. This is appreciated as courtesy if all goes well and helpful to avoid complaints if problem do occur.

When adverse events happen early and thorough counseling of the patients family is important. This guards against later feeling of a 'cover up' which is often the stimulus for complaint and litigation.

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