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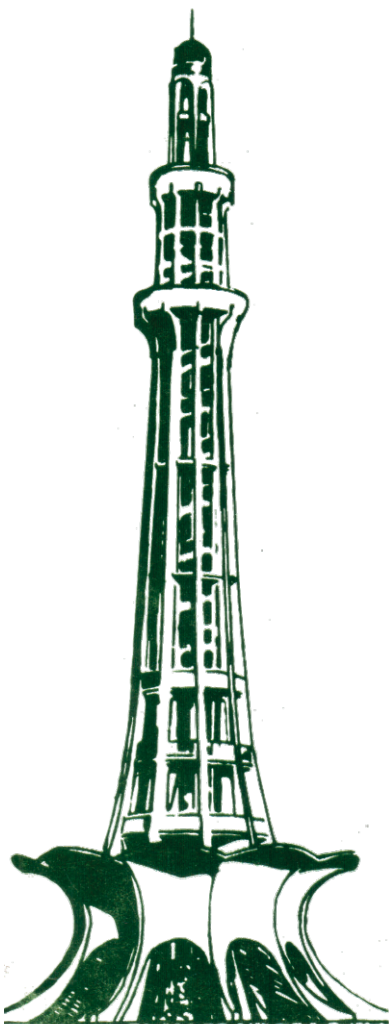
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IN THE NAME OF ALLAH, THE BENEFICENT, THE MERCIFUL

مَجَلَّةُ طِبِّ الْعُيُونِ پَاكِسْتَان
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Vernal Keratoconjunctivitis — Care, Keratoconus and Other Complications

In 1982, during my visit to Faisalabad, Pakistan, a 17-year-old handsome and intelligent boy was brought to me for evaluation of blindness in one eye and ongoing gradual loss of vision in the other. Through the central moderate posterior subcapsular opacities almost total optic disc cupping in the right eye and fairly advanced cupping in the left eye could be seen. The changes of chronic vernal keratoconjunctivitis were present in both eyes. The patient had been using corticosteroid drops to relieve the symptoms of vernal conjunctivitis for such a long time that corticosteroid induced glaucoma and cataracts had robbed him of his sight. To my horror, the boy was still using corticosteroid drops. Unfortunately, he had ignored the warning about the dire complications of unsupervised treatment by the ophthalmologist who initially prescribed these drops. The patient had continued refilling the prescription either by showing the container to the druggist, or by getting renewable prescriptions from a local general practitioner who never bothered to refer the boy to an ophthalmologist for evaluation. The following year, two teenagers who had gone totally blind under similar circumstances were shown to ophthalmologists attending the 1983 Afro-Asian Congress at Lahore. These tragedies are not merely isolated events in Pakistan, as is abundantly clear from the lead article in this issue by Khan, Kundi, Saeed, Gulab, and Nazeer.¹ (See page 111). The sooner more effective regulations on dispensing drugs are formulated and more strictly enforced, the better it will be for the people of Pakistan.

As for the efficacious therapeutic agents for vernal conjunctivitis, the situation remains most unsatisfactory even after a hundred years. A poorly perceived disease cannot have a well-aimed therapy.

Vernal conjunctivitis was first clearly delineated as a clinical entity in 1875 by Saemisch,² who described it as a rare disease of unknown etiology. The disease remained unnoticed for the next twenty years and no mention of it was made in many of the ophthalmic textbooks.³ Another twenty years passed before it found a regular place in various texts on eye disease, when Posey declared, in 1902, that the disease was "in all probability due to a specific germ." Because researchers failed to isolate any organism, the literature soon became replete with alternative theories explaining the etiology of vernal

conjunctivitis. In the 1920s, most popular theories were that it was an expression of some constitutional disorder of ductless glands, a manifestation of vagotonia and diminished suprarenal activity, or an ocular allergy, usually to pollens.^{5, 6} Most recently, increased levels of IgE have been found in the serum and tears of patients with vernal conjunctivitis,⁷ suggesting strongly that it is an (atopic) IgE mediated type 1 hypersensitivity disorder. However, other concurrent immune mechanisms may play a role in pathogenesis, rendering it still incompletely understood. It has also been shown that IgE antibodies to rye grass and ragweed antigens may be produced in the conjunctiva of patients with vernal conjunctivitis.⁸ Now it is also possible to produce and study type 1 hypersensitivity reactions similar to vernal conjunctivitis in laboratory animals.⁹ These developments may be expected to result in new, more effective therapeutic and prophylactic measures.

Recently, encouraging results have been achieved in treating vernal conjunctivitis with non-steroidal agents. Oral aspirin apparently inhibits the synthesis of prostaglandin D₂, a secondary mast cell mediator.¹⁰ Cromolyn sodium in 2% to 4% solution drops q.i.d. has proved another profitable therapeutic and prophylactic agent. Its action depends on the inhibition of the release of histamine from mast cells.^{11, 12} Cyclosporine 2% solution drops caused freedom from symptoms in nine out of 12 patients, but symptoms recurred in most of them at the subsidence of the treatment.¹³ The corticosteroids remain the most effective available therapy, but they also have the most undesirable complications, making it necessary to continue the search for alternatives. Hence, the battle to understand and cure vernal conjunctivitis goes on.

Occurrence of keratoconus in about 9% of the patients reported in the study by Khan et al¹, is most significant. The previous reports have either denied any relationship between keratoconus and vernal conjunctivitis, or found it in less than one percent of the patients.¹³ This study not only establishes that keratoconus is a genuine complication of vernal conjunctivitis, but also suggests that Pakistanis may be more predisposed to it. The overall incidence of corneal complications in this study was nearly 49%. The most important corneal complication, ulceration, has been previously reported in 3.2% of the cases of

vernal conjunctivitis.¹⁴

In Pakistan, the matter of differentiation between vernal conjunctivitis and trachoma must also be considered. They may co-exist in the same patient. However, when this is not the case, following differentiating features may be helpful in making a correct diagnosis.¹⁵ The symptom of itching, characteristic ropy discharge, and seasonal recurrences are more typical of vernal conjunctivitis. Eosinophils are not found in trachoma, and intracellular cytoplasmic inclusions are not found in vernal conjunctivitis. The granulations of the conjunctiva may involve the retrotarsal folds in trachoma, but not in vernal conjunctivitis.¹⁵ Although pannus may be seen in vernal conjunctivitis,¹⁴ it is so rare in it that it may be regarded as an exclusive feature of trachoma.^{6,14,15}

— Khalid J Awan, FPAMS

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Ophthalmic "Pastpourri"

P.I. PEEVES

TODAY:

The "time has come to examine more closely the advantages versus the disadvantages of peripheral iridectomy in cataract microsurgery..." It "may not be necessary at all and may indeed be a potential source of complications..." such as "intraoperative hyphema, disinsertion of the iris, inadvertent cutting of the superior loop of the implanted intraocular lens..." and iridic "irritation."

Schulze, RR, and Copeland, JR: Posterior chamber intraocular lens implantation without peripheral iridectomy - a preliminary report. *Ophth Surg* 13:567, — 1982

A CENTURY AGO:

It "is time to escape from the operative anarchy which has prevailed since (the introduction of iridectomy in cataract operation)." The author strongly recommends "the avoidance of an iridectomy (during cataract extraction), so that the 'resultat esthetique' is infinitely better, while many unpleasant complications and bad results are avoided, such as hemorrhage into the anterior chamber, escape of vitreous, incarceration of capsule, iritis, and iridocyclitis, etc."

Panas, F: Du choix du meilleur proce'de' d' extraction de la cataracte. 1885 (1-15-246-531)



Appreciation Expressed

The following experts in their fields devoted their valuable time to review the manuscripts and provided most meritable help in the improvement of the standard and style of the material that was published in the SECOND VOLUME of the Pakistan Journal of Ophthalmology (January 1986 to October 1986) Without their generous cooperation, the standard of The Journal could not have been maintained. We wish to express our deepest gratitude and indebtedness to them. — Editor.

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A Study of 530 Cases of Vernal Conjunctivitis from the North West Frontier Province of Pakistan*

M. Daud Khan, FRCS,
Niamatullah Kundi, FCPS,
Nasir Saeed, DO,
Arifa Gulab, MBBS,
and Anisa F. Nazeer, MBBS

ABSTRACT: The authors studied 530 patients, 444 men (83.8%) and 86 women (16.2%), with vernal conjunctivitis in the Department of Ophthalmology of Khyber Hospital at Peshawar, from January 1982 to December 1984. Eighty-nine percent of the patients were children and 11% were adults. The disease was active in summer and spring in all of the patients except for 41, in whom it was perennial. The mixed limbal and palpebral variety afflicted 93.6% of the cases. Personal or familial allergies occurred in 39.1% of the patients. Hayfever was the commonest personal and asthma the commonest familial allergy. Topical corticosteroids were required in an overwhelming majority of the patients to control the disease the surgical treatment (cryotherapy and or excision of the papillae tried in 21 cases) was unsuccessful.

Complications of the disease itself or from its treatment developed in 299 (54.4%) of the cases. The cornea was involved in 259 of these. Keratoconus was found in 48 (9%) of the patients. Corticosteroid induced glaucoma (14.2% cases) and cataract (9.1% cases) were the most serious therapeutic complications. (Pak J Ophthalmol 2:111-114, 1986)

Vernal conjunctivitis is a recurrent, bilateral, interstitial inflammation of the conjunctiva, usually of periodic seasonal incidence, self-limited course, characterized by flat-topped "cobble-stone-like" papules on the tarsal area and a discrete or confluent gelatinous hypertrophy of the limbal conjunctiva, and a distinctive type of keratitis, associated with intense itching, redness, lacrimation, photophobia, and a mucinous ropy discharge containing eosinophils.¹ Since the first description of the limbal variety by Arlt,² in 1846, as conjunctivitis lymphatica, the condition has been known in the past as perikeratic hypertrophy, phlyctoena pallida, circumcorneal hypertrophy, recurrent vegetative conjunctivitis, conjunctivitis aestivale, periodic hyperplastic

conjunctivitis, or spring catarrh. The last one having enjoyed the most popularity; notwithstanding that it is a misnomer, because the affliction is neither catarrhal one, nor does it necessarily occur in spring. The disease recurs in early summer rather than in spring.¹ Vernal conjunctivitis is a geographically universal affliction with a widely variable incidence, being rare in England and France and being common in India, Middle East, and the Americas. Because of the apparently high number of patients with vernal conjunctivitis in our clinics, we decided to study the incidence of vernal conjunctivitis and complications associated with it and its therapy in the North West Frontier Province of Pakistan.

MATERIALS AND METHODS

A special proforma was drawn up for this study of the patients with vernal conjunctivitis, indicating the age, sex, and history of the disease in each patient. It also included the age of the patient at the time of onset of the disease, the duration of the disease at the time of presentation, the seasonal variations, other personal or familial atopic disorders in the patient, and the treatment received.

The types of lesions, ocular complications of the

Accepted for publication on September 10, 1986.

From the Postgraduate Medical Institute and the Lady Reading Hospital, University of Peshawar, Peshawar, Pakistan. (The study done at Khyber Hospital Peshawar)

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Reprint requests to M. Daud Khan, FRCS, Department of Ophthalmology, Postgraduate Medical Institute, Lady Reading Hospital, Peshawar, Pakistan.



Figure 1. (Khan, Kundi, Saeed, Gulab, and Nazeer): Because of the extreme severity of symptoms, this child with vernal conjunctivitis had great difficulty in opening her eyes.



Figure 2. (Khan, Kundi, Saeed, Gulab, and Nazeer): Typical "vernal conjunctivitis facies" of an adult patient.

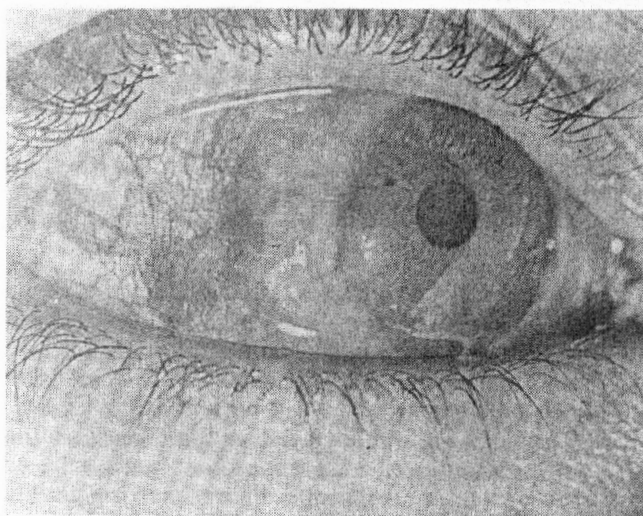


Figure 3. (Khan, Kundi, Saeed, Gulab, and Nazeer): Massive limbal involvement in vernal conjunctivitis.

disease, and of its treatment, and their effects on vision were recorded. The study period began on January 1, 1982 and ended on December 13, 1984. All the patients seen in hospital eye clinic or in private offices of the authors were included in the study. In each case diagnosis was made on the basis of clinical features only (Figures 1-3) and no laboratory tests for blood or conjunctival eosinophils, or serum and tear IgE were done. The proformas were analyzed at the end of the period for the results.

The clinical features used for diagnosis of vernal conjunctivitis were intense itching, typical conjunctival papillary hypertrophy and/or limbal changes, edema, stringy discharge, and seasonal periodicity. Only those patients were included in the study who did not have any other keratoconjunctival disease at the time of first examination. Also the patients who had glaucoma or any cataractous changes at the time of first examination were excluded. However, the patients who were lost to followup and then reappeared for management of the disease were included, despite their having developed these two findings, provided no cause other than the use of corticosteroids was confirmed by history. Glaucoma was diagnosed when definite pathologic cupping and elevation of intraocular pressure over 21 mm Hg on two occasions were present. Biomicroscopic examination was used for evaluation of corneal complications. The diagnosis of keratoconus was made by clinical appearance, retinoscopy, keratoscopy, Placido disk, and slit lamp observation of folds in Descemet's membrane. No keratometry was performed.

RESULTS

Out of a total of 530 cases, 444 (83.7%) were men

Table 1
Age of onset

Age group (in years)	No. of patients	Percent
0-5	123	23.21
6-10	253	47.74
11-15	95	18.11
16-20	43	8.11
21-25	6	1.13
26-30	-	-
31	2	0.38
Unknown	8	1.32
Total	530	100.0

Table 2
Seasonal variations*

Season	No. of patients
Spring	495
Summer	503
Autumn	80
Winter	42
Perennial	41

*The variation in the total is on account of the overlapping of activity periods of disease in many patients.

and 86 cases (16.2%) were women. The age variations at onset are given in Table 1. The disease has been present for five or more years in 342 (64.5%) patients, for six to ten years in 131 (24.7%), and for more than ten years in 57 (10%). The seasonal variation in the activity of disease is given in Table 2. A total of 496 patients (93.6%) had a mixed palpebral and limbal disease, 32 cases (6.00%) had palpebral lesions alone, while only 2 (0.4%) cases had just limbal lesions. A total of 490 cases had received topical steroids alone or in combination with antibiotics at the time of presentation. Both medical and surgical treatment was received by 21 patients. The surgical treatment involved the excision of papillae, cryoapplication to the papillae, or both. Nineteen cases had not used any previous treatment at the time of presentation.

Table 3
Personal and familial allergies.
(207 cases, 39% of 530)

Type of Allergy	No. of patients	Percent
A. Personal allergies	77	14.53 of total
Hayfever	69	89.62
Asthma	5	6.49
Eczema	5	6.49
Drug allergy	1	1.30
Multiple	3	3.90
B. Familial allergies	130	24.53 of total
Asthma	69	53.08
Spring catarrh/Allergic conjunctivitis	59	45.38
Hayfever	8	6.15
Eczema	8	6.15

Table 4
Type of corneal complications - 259 cases (48.9% of 530)*

Type	No. of patients
Superficial Punctate Keratitis	137
Limbal Deposits	83
Keratoconus	48
Ulcers	24
Scarring	3
Edema	2
Ectasia	2
(Multiple Complications)	(39)

*The difference in the total number is due to multiple complications in some patients.

Table 5
Causes of loss of vision to the level of 6/24 (20/80) or less due to complications of vernal conjunctivitis or its treatment (103 eyes)*

1. Steroid induced glaucoma.	22
2. Steroid induced glaucoma with cataract.	15
3. Cataract.	14
4. Keratoconus with cataract.	7
5. Keratoconus.	39
6. Corneal ulcers.	6

*Almost 2/3 of the eyes had less than 6/60 (20/200) visual acuity.

Table 6
Age distribution of complications.

Group in years	Total No. of patients	Number with complications	Percent
0-5	31	7	22.58
6-10	145	73	53.40
11-15	158	92	58.20
16-20	119	74	62.20
21-25	54	36	66.66
26-30	15	9	60.00
31 and above	8	8	100.00
Total	530	299	56.41

History of other atopies (personal or familial) occurred in 207 (39.2%) cases. Personal allergies occurred in 77 (14.5%) cases, in which hayfever was found to be the commonest. Familial allergies occurred in 130 (24.5%) cases, in which asthma was found to be the commonest familial allergy (53.1%). (Table 3)

Corneal complications occurred in 259 cases. (Table 4) steroid induced lenticular opacities and steroid induced glaucoma. Among the corneal complications keratoconus, which occurred in 48 patients, had the most devastating visual results (Table 5). Other causes of senese loss of sight including are shown in Table 5.

The closer analysis of the complications suggested that the longer the duration of the disease, the higher the chances of complications. Maybe this is why the older patients had the higher incidence of complications. (Table 6)

Other diseases not particularly related to spring catarrh noted in this group were 8 cases of high refractive error, 2 cases of corneal ectasia, 2 cases of iritis, 1 case of traumatic cataract and 3 cases of active trachoma.

A total of 116 eyes had poor visual acuity of less than 6/24, (20/80), 79 eyes with less than 6/60 (20/200). In 103 eyes the cause of poor vision was either vernal catarrh itself or the complications arising from its treatment. (Table 5)

More than 90% of the patients in our study were using topical corticosteroids for symptomatic relief.

DISCUSSION

Vernal conjunctivitis is a specific IgE mediated type I hypersensitivity affecting the eyes.³ It is often associated with high levels of eosinophils in blood and conjunctival scrapings.⁴ There is also usually high levels of IgE in serum and tears.⁵ Other atopic disorders like spastic bronchitis, eczema and hayfever are also usual accompaniments of this disease.⁶ The disease usually affects males in the age group 6-20 years.^{1,7}

In the present study the age/sex distribution of the disease was found to follow the generally accepted pattern. Although it is not a universal feature of the

disease, a seasonal variation was prominent in this study. For example, Togby⁸ from Egypt and Neumann et al³ from Israel did not observe any significant seasonal variation in their patients. Similarly Jay⁹ in 1981 reported a perennial character of the disease from England, but his study was confined to 17 cases and all were over 16 years of age.

In the present study, 93.5% of the cases had mixed palpebral and limbal disease. Togby⁸ similarly reported a higher incidence (71.4%) of mixed type of lesions in his study from Egypt, while Posey¹⁰ and Lehrfeld¹¹ respectively reported a higher incidence of 60% to 55% of palpebral disease from the United States. On the other hand some authors have noted an almost exclusive predominance of the limbal variety in black patients and suggested a genetic factor.¹²

An overwhelming majority of our patients were so used to treatment with topical corticosteroids that many stubbornly refused any change in therapy to non-steroidal anti-allergic preparations. This was in spite of obvious ocular complications of steroid therapy. Excision or cryotherapy of the hypertrophic papillae was done in a few cases, but were not found to be of much value.

Personal or familial allergies were found in 39.2% of our patients, which is in agreement with the generally accepted view that concurrent allergies are common in vernal catarrh.⁶ However, in two reports from the Middle East, the incidence of other systemic allergies (6%³ to 11%⁸) was similar to the comparable age-group subjects in the control group.^{3,8}

A number of serious sight threatening complications of the disease or its treatment were noted in the present study. Corneal complications occurred in 259 cases: steroid-induced glaucoma in 48 cases, steroid cataract in 31, and multiple complications in 39. Corneal complications included superficial punctate keratitis, marked limbal deposits, keratoconus, ulcers, plaques, pseudogerontoxon, corneal oedema and corneal ectasia. Except for keratoconus, these corneal complications are similar to those reported by other workers.^{9,13}

We had 48 cases of keratoconus. Keratoconus is reported to be associated with a number of other atopies.^{14,15} Although it has been reported in vernal catarrh by a few workers^{16,17,18} most workers did not find any association between the two conditions.^{14,15,19} Nonetheless, this high incidence of keratoconus in our patients indicates that there is definite relationship between vernal conjunctivitis and keratoconus, at least in population of Pakistan. These complications were responsible for the loss of sight in 103 eyes to the extent of less than 6/24 (20/80) visual acuity. Out of these, almost 2/3 of the eyes were legally blind with visual acuity of 6/60 (20/200) or less. This study thus indicates that vernal catarrh prevalent in the North West Frontier Province of Pakistan is of a severe mixed type variety. It must be emphasized that an unsupervised use of steroids led to severe loss of vision in many young individuals. It is thus suggested

that the disease should be studied in more depth in the community, especially its immunological background. Efforts should be made to identify the allergens responsible, eliminate them if possible, or desensitize the patients.

The efficacy of drugs like sodium cromoglycate^{8,9,20,21,22} and aspirin²² should be studied in these patients and if found effective, their use should be popularized. The corticosteroids must be used with great caution and for a minimum period when necessary. Potent steroids like beta and dexamethasone should be replaced by new synthetic steroidal preparations, such as medrysone and fluorometholone, which are reported to have a lesser effect on intra-ocular pressure. Nonetheless, all patients using steroids must have a careful and regular follow-up of intra-ocular pressure.

Acknowledgment: We thank Khalid J. Awan, FPAMS, for his help.

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Remarks and Replies

Subluxation of the Lens and Ocular Hypertension in Exfoliation Syndrome

I found the article, Subluxation of the Lens and Ocular Hypertension in Exfoliation Syndrome, (Pakistan Journal of Ophthalmology, 2:77-78, 1986), a most stimulating reading. I was rather impressed with the number of patients involved in the study along with the significant prevalence of lens subluxation in them. I feel that at least some degree of lens subluxation is probably more common in this syndrome than what most people appreciate.

To date, there have been few studies examining the prevalence of lens displacement in the exfoliation syndrome. In Gifford's¹ report, three of 62 eyes with the exfoliation syndrome were found to have spontaneous lens dislocation; while a subsequent study by Tarrkanen² of 635 eyes with the exfoliation syndrome demonstrated 9 cases of displaced lenses. Sood, et al³, described 11 cases of phakodonesis, 2 cases of dislocated lenses, and 5 cases of iridodonesis in 17 exfoliation eyes. Bartholomew⁴ evaluated 238 eyes with lens displacement, and found 22 to have the exfoliation syndrome.

Although in my experience, a 16% prevalence of lens displacement in eyes with the exfoliation syndrome seems to be rather high, I think this article has made a significant contribution to a better understanding of and appreciation for exfoliation syndrome.

I would be interested to know if the authors were able to correlate the finding of lens of luxation with any other associated findings in the exfoliation syndrome, i.e., cataract or glaucoma (open-angle or angle-closure), or if it correlated with the degree of exfoliation.

At present, I am in the process of defining a group of patients that I call "exfoliation suspects" on the basis of the presence of secondary clinical signs without evidence of exfoliation material on the pupillary border or anterior lens capsule in either eye. These patients are being assessed for evidence of early involvement by examining conjunctival biopsy specimens for the presence of exfoliation material.

Andrew M. Prince, M.D.
New York N.Y. U.S.A.

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Reply

I am very thankful for Dr. Prince's letter and remarks about our paper, Subluxation of the Lens and Ocular Hypertension in Exfoliation Syndrome.¹ I quite agree with him that the luxation of the lens occurs to some degree in the patients with exfoliation syndrome, but the incidence in our study was appreciably higher. While working in the north of Scotland, I had the chance to conduct ophthalmic outpatient clinics in the Outer Hebrides. I did not collect any data, but the exfoliation syndrome was very prevalent; however, the subluxation of the lens was not. The high incidence of exfoliation syndrome in those patients might be a racial and geographic phenomenon.

As to the incidence of cataract in our patients, it occurred in 49 eyes out of a total of 70 eyes studied which had exfoliation syndrome but had no luxation of the lens. The cataract was mature in 14, posterior subcapsular in five, cortical in 20 and nuclear in four eyes. The remaining six eyes were aphakic.

I would be most interested in learning the results of the research Dr. Prince is doing on exfoliation syndrome.

Shad Mohammad, FRCS
Abbottabad, Pakistan

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The Eye Camps — Risky or Rewarding?

Kindly accept my heartiest congratulations for the excellent production of Pakistan Journal of Ophthalmology. It will help in understanding the ophthalmic problems in the region.

I have gone through your editorial on the eye camps,¹ and I entirely agree with your observations. In fact, we have already laid down certain guidelines for organizing eye camps in Delhi and surrounding areas. The organizer of the camp gives undertaking to abide by the conditions, only then permission for holding the eye camp is granted to him.

We are planning to hold a symposium on the eye camps and I would like "RISKY OR REWARDING" for the title, if you permit me to use these words for your editorial.

S.R.K. Malik, M.D.
New Delhi, India

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Self-Evaluation, Self-Education

Edited by Khalid J. Awan, MD, FPAMS
Muhammad Humayun, FRCS, FPAMS

In this section of the Journal, photographic documentations of interesting and challenging observations will be presented to the readers. They should make their diagnosis from the given information and compare these with the expositions on pages 133-134—Editor

Camera Clinicals



Figure 1



Figure 2



Figure 3

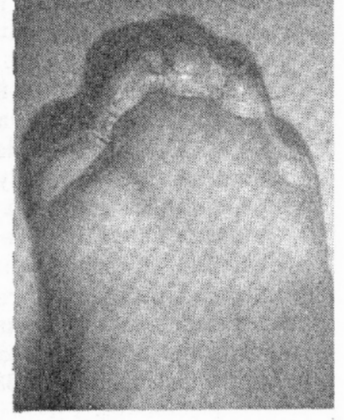


Figure 4

Figures 1, 2, 3, and 4: The patient was a 5-year-old girl with average intelligence. Her skin and hair color were noticeably lighter than the rest of the family. Ocular examination showed visual acuity to be 20/30 (6/9) in each eye with correction. Her prominent eyes also had hypertelorism and exotropia (Figure 1). On biomicroscopy, the irides patchily transilluminated. Ophthalmoscopy showed that the fundi were hypopigmented. The systemic clinical features included interesting facies (Figures 1 and 2) and syndactyly of hands (Figure 3) and feet (Figure 4).

20/20 (6/6) in each eye with correction. The ocular examination was normal with the exception of two things: The ultrasonography showed that the anteroposterior length of the left eye varied from 23.67 mm to 30.16 mm depending on the direction of the gaze. The ophthalmoscopy of left eye showed the changes represented in the composite photograph in Figure 7, with a perfectly normal macula. A small retinal cystic area in the retina at 1 o'clock above the macular area did not affect the overall function of the eye.

Figure 5: A 62-year-old man had suffered for almost 20 years from a disease of the face that completely destroyed his nose. (Figure 5). His refusal to have surgery eventually resulted in so an extensive involvement that radiation remained the only choice for treatment. Although irradiation halted the progress of the disease, it caused ectropion of the lower eyelids, destruction of the medial canthi with direct invasion of the globes on the nasal sides by the proliferating skin, chronic conjunctival irritation and epiphora, and cataracts (Figure 5). The absence of a nose precluded the use of glasses. Eventually modern surgical techniques helped restore useful vision.

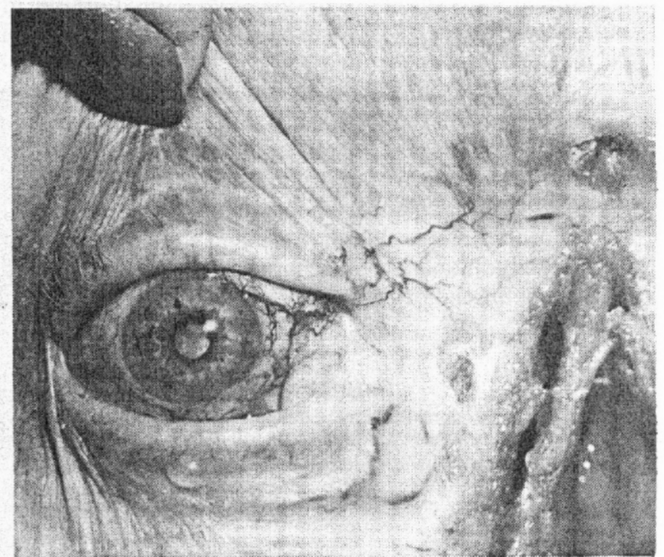


Figure 5

Figures 6 and 7: A 9-year old brilliant girl had totally solid-white leukocoria when examined from a straight ahead position, but not when the observation was made from the sides (Figure 6). Her vision was

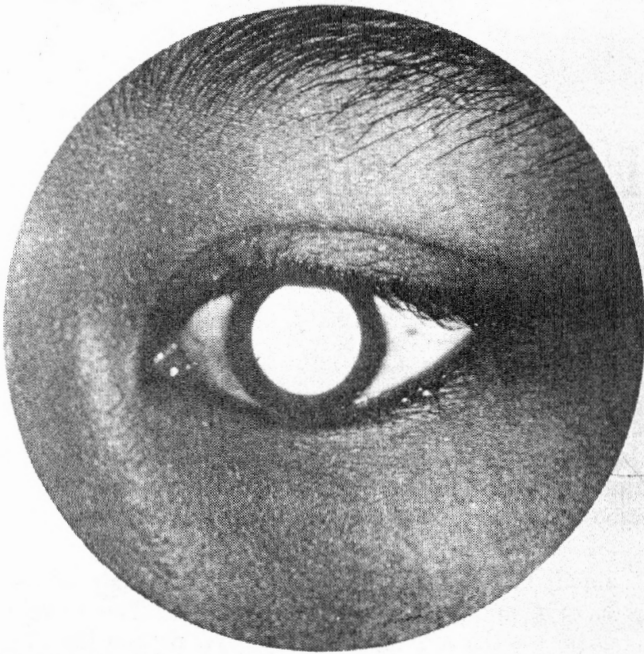


Figure 6

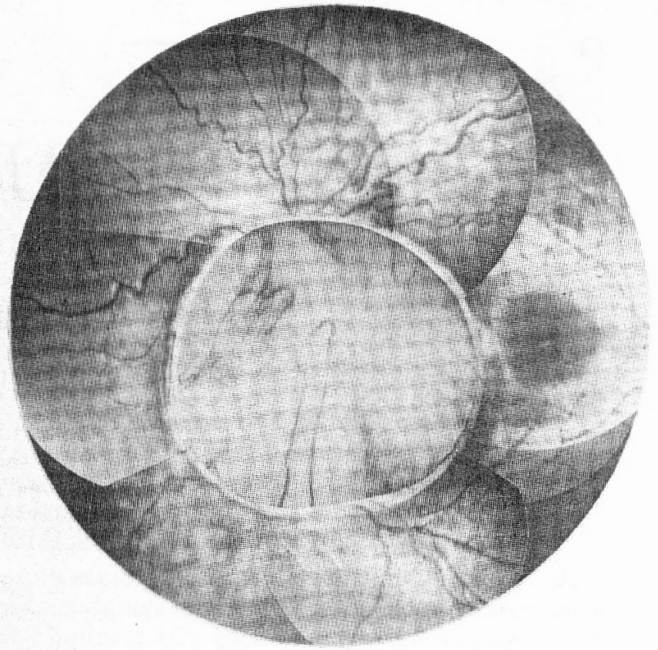


Figure 7

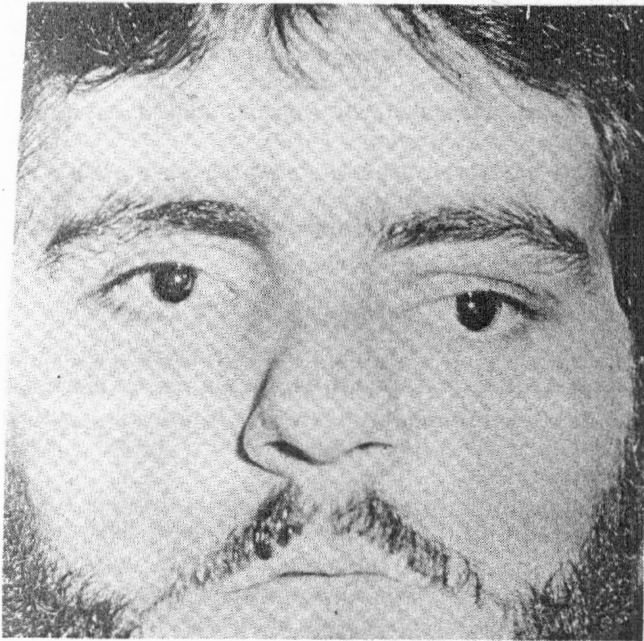


Figure 8

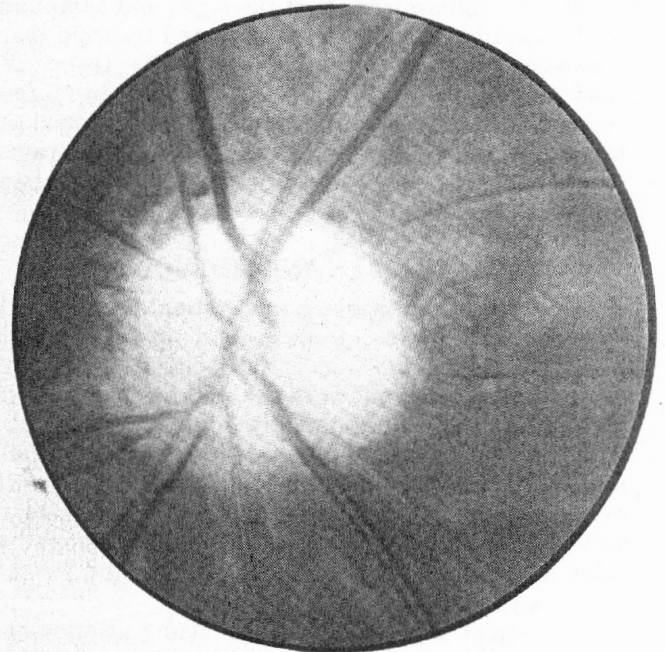
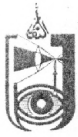


Figure 9

Figures 8 and 9: A 22-year-old man had noticed down and outward turning of his left eye since the age of 5. This was followed by a gradual loss of sight to the present light perception level in that eye only. The right eye has remained normal with 20/20 (6/6) vision and full field of vision.

The X-rays of the skull at age 6 had shown some thickening of the roof of the left orbit and also hypertrophy of the lymphoid tissue in the nasopharynx. However, the optic foramina and canals

were normal on repeat radiologic studies. These findings were recently confirmed by a CT scan. The examination with Hertel's exophthalmometer has revealed no proptosis of the left eye. It is, however, exotropic with hypotropia (Figure 8), and has distinct fundoscopic changes shown in Figure 9. The patient has no similar skeletal changes in the rest of the body. The laboratory workup showed an elevation of alkaline phosphatase, 24 a.k.u. (normal, 17 a.k.u.) and serum phosphorus, 5.3 mg (normal, 4.5 mg).



Elicited Lid Lag Sign in Early Graves' Ophthalmopathy

Amin M. Nasr, M.D., Eduardo P. Penna, M.D.
and
Pamela S. Chavis, M.D.

ABSTRACT: The authors describe a unilateral lid lag sign in 10 out of 12 patients with early Graves' disease. These patients did not show lid lag when the examination was conducted with both eyes open. This early sign of Graves' ophthalmopathy is elicited only after visual occlusion by covering of and the eyelid movement suppression by application of pressure over the fellow eye. (*Pak J Ophthalmol* 2: 118-119, 1986)

The diagnosis, of Graves' disease becomes difficult or impossible in patients with subtle symptoms and a few nonspecific signs that overlap with manifestations of other diseases with unilateral eye involvement in a clinically and chemically euthyroid patient.¹ A physician's full awareness of the signs and symptoms of thyroid ophthalmopathy is essential to avoid more sophisticated and expensive diagnostic tests. We report a new sign of lid lag induced by forced suppression of visual contact and eyelid function of the fellow eye in patients with early Graves' ophthalmopathy, which to our knowledge has not been previously described.

MATERIALS AND METHODS

Twelve apparently euthyroid patients with subtle evidence of Graves' disease and unilateral mild proptosis (2-3 mm) had full clinical and eye examinations. They all were checked for unilateral lid lag with one eye intentionally suppressed and also with both eyes open while following a vertically moving target. All patients had further diagnostic evaluation with ultrasonography and CT scanning to establish the diagnosis of Graves' ophthalmopathy. Also, several normal individuals were tested for this unilateral lid lag.

The patient is examined in the sitting position and allowed to fixate with both eyes at a target placed 50 cm away in the primary direction of gaze. A sufficient distance is required between the patient and the target to minimize convergence, to maintain a full range of motion of the upper eyelid, and to permit the examiner an adequate observation. The target is elevated to a point where the patient has to maximally elevate his eyelids to see the target; then the target is

slowly lowered with the patient continuously following it to a maximal downward position. The examiner observes the movement of the eyelids and the globes for any jerky or asynchronous motility. The examiner observes the following during the test: 1. any lid lag in either eye; 2. scleral show and the position of the upper eyelid margin in relation to the pupillary border and the limbus. 3. the synchronous movement of both the eyeballs and upper eyelids. 4. any jerky or abnormal movement of eyelids; and 5. asymmetry between the two eyes.

The examiner repeats the procedure with visual suppression of one eye by covering it and preventing the movement of its upper lid by applying pressure over it with the palm of his hand.

RESULTS

There was no evidence of lid lag in either eye in any patient when tested with both eyes open. There also was no asynchronous movement of the eyes and the eyelids, and no innervational "jerky" eyelid movements.

In 10 out of 12 patients, a unilateral upper lid lag, manifested by 1 - 2 mm of scleral show between the superior limbus and the upper eyelid margin, was clearly noticed when the opposite eye was covered and its upper eyelid function suppressed by pressure application during testing by the method outlined above (Figures 1 - 3). The normal subjects tested during the study had no lid lag, or it was insignificant in degree.

COMMENTS

In 1969, Werner² classified the eye changes in Graves' disease into Class 0, no signs or symptoms; Class 1, no symptoms but signs limited to upper eyelid retraction and stare, with or without lid lag and proptosis; Class 2, soft tissue involvement with signs and symptoms; Class 3, proptosis; Class 4, extraocular muscle involvement; Class 5, corneal involvement; and Class 6, visual loss due to optic nerve

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Figure 1.
(Nasr, Penna,
and Charis):

The patient observing and following the target, from a maximal elevated position to a maximum downgaze position with both eyes open. Notice no evidence of lid lag in either eye.



Figure 2.
(Nasr, Penna,
and Charis):

Left eye occluded. Patient is following the target with right eye from upgaze to downgaze. Not the definite lid lag in the uncovered right eye.



Figure 3.
(Nasr, Penna,
and Charis):

Right eye occluded. Patient is following the target with left eye. Note the prominent lid lag with scleral show.

involvement. This classification proved to be unsatisfactory, and several years later had to be modified.³ Nonetheless, the clinical usefulness of this classification remains, at least in ophthalmology, seriously limited,^{4,5} and a new clinical classification of Graves' ophthalmopathy based on computed tomographic (CT) scans has been proposed.⁶ It is obvious that an early diagnosis of Graves' ophthalmopathy is important. The unilateral lid lag sign we have reported appears to be a helpful corroborative clinical feature. Although it is not present or is present in minimal degree in normal individuals, the sign of unilateral lid lag by suppression of the fellow eye must not be regraded as an absolute but only as a supportive evidence of euthyroid Graves' disease.

Von Graefe's lid lag sign, elicited by having the patient look down from up gaze and noting the follow movement of the upper eyelid with the eyeball in its descent, may not be clinically evident in early stages of Graves' disease. We have noted the absence of this sign in several patients with mild unilateral proptosis of Graves' disease when examination is conducted with both eyes open. However, the unilateral lid lag sign reported here could be elicited in these patients.

In addition to the lid lag of von Graefe, several other signs have been reported in Graves' disease. They include Dalrymple's sign (lid retraction) which may also be seen in other conditions, such as contralateral ptosis with pseudoretraction of the fellow eyelid, hydrocephalus, aberrant regeneration of the third nerve, hypokalemic periodic paralysis, cirrhosis of the liver, etc.; Ballet's sign (palsy of one or more extraocular muscles); Cowen's sign (jerky pupillary contraction to consensual light); Gifford's sign (difficulty in everting the upper eyelid); Jellinek's sign (brownish pigmentation of the eyelids); Joffroy's sign

(absence of forehead wrinkling on up gaze); Lowy's sign (large dilation of the pupil on instillation of weak epinephrine solution drops), which may also be seen in pancreatic insufficiency; Mobius's sign (weakness of convergence); Rosenbach's sign (fine rapid tremors of the closed eyelids), should not be confused with Rosenbach's sign in neurasthenia in which patient is unable to close eyelids immediately upon command; Stellwag's sign (infrequent blinking); and Suker's sign (a poor fixation on lateral gaze).

The physiology of eyelid movement is multifactorial. It depends on visual, sensory, anatomic, and neuronal factors that contribute homogeneously to the total functioning potential of the eyelid. Visual occlusion without forceful suppression of eyelid function did not elicit a lid lag. The dual symmetrical innervational pattern to both upper lids appears to be disrupted by the visual and mechanical interference in one eye. However, the pathophysiology of the sign we have described remains to be explained. It may be that disruption of binocularity by suppression of one eye affects the bilateral synchronous pattern of eye movements which enhances the early pathologic change.

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Transition From Intracapsular Cataract Extraction to Extracapsular Cataract Extraction Facilitated by Viscosurgery

Emanuel Rosen MD, FRCS

ABSTRACT: The transition from intracapsular cataract extraction (ICCE) to extracapsular cataract extraction (ECCE) may be facilitated by two approaches: (a) utilization of viscosurgery in order to provide the surgical control that generates safety and surgical confidence and (b) adoption of the intercapsular (endocapsular) technique of extracapsular cataract extraction which ensures in the bag implantation and the flexibility to aid implantation in complicated cases. (Pak J Ophthalmol 2:120-123, 1986)

Ophthalmic surgeons generally are conservative in their operating habits and accordingly feel more comfortable and secure in the practice of familiar methods. Thus intracapsular cataract surgery with or without lens implantation is likely to be more successful in the hands of an experienced intracapsular trained surgeon than would be less familiar extracapsular methods.

Voices of progress may encourage ophthalmic surgeons to adopt new methods but it is the eyes of patients which are at risk during a transitional period. Adoption of new and improved techniques are required in order to achieve higher standards of care with long term patient visual benefits. The need is to achieve evolution of practical systems which are able to minimize the trauma of a transitional surgical period.

Intracapsular Cataract Surgery

Intracapsular surgical technique has benefitted since the introduction of viscosurgical methods in 1980 with Healon® and subsequently with other viscoelastic agents.

Viscosurgery immediately offered: a. better surgical control of the intraocular procedures, b. better protection of vulnerable tissues during surgery and, therefore, c. increased confidence in surgical performance with consequential improvement in overall results.^{1,2,†}

Specifically in the intracapsular procedure, Healon® allows: a. protection of the anterior hyaloid membrane after cataract extraction, b. recreation of

the posterior chamber space for implantation of a posterior chamber IOL or recreation of the anterior chamber for placement of an angle supported anterior chamber IOL, c. protection of the corneal endothelium from the hydrophobic polymethylmethacrylate IOL, d. control of bleeding from an iridotomy or iridectomy when performed, and e. other uses in complicated cases e.g. separation of synechiae.

Problems of Extracapsular Cataract Surgery

The problems facing the neophyte extracapsular surgeon are: a. performance of a safe and sound anterior capsulotomy, b. successful removal of the lens nucleus, c. complete evacuation of cortical remnants after nucleus expression, d. maintenance of the integrity of the posterior lens capsule, e. incorporation of the IOL into the capsular bag, and f. avoidance of damage to the corneal endothelium which is particularly at risk from mechanical and fluid trauma especially during a prolonged procedure.

Some of these problems can be overcome by the design of the surgical technique e.g. intercapsular methods i.e. working within the capsular bag until the IOL is implanted. Other potential problems can be minimized by utilizing viscosurgery.

Viscosurgery

"Viscosurgery" is defined as the use of clinically proven, physiologically compatible, viscoelastic substances to facilitate a surgical procedure. This takes three principle forms: a. creation or recreation of space between ocular tissues to allow safe intraocular surgical maneuvers, b. the maintenance of these spaces for the duration of surgical maneuvers by the elastic and viscous forces which counteract collapse or deformation, and c. the use of viscoelastic

Accepted for publication July 18, 1986.

From Manchester Royal Eye Hospital, Manchester England.

Reprint requests to Emanuel Rosen, MD, FRCS.

materials to protect vulnerable tissues from mechanical or chemical trauma by coating and/or separating those tissues from potential noxious agents.

Substances or solutions which are merely viscous but without significant elastic properties would fail to fulfil the requirements of a surgical 'liquid' instrument which has to be delivered to required sites within the eye through a narrow calibre cannula. Such deformation and reconstitution of a substance that this method of delivery requires, dictates the physical characteristics of a medium which allows a surgeon freedom of maneuver that is implicit in 'viscosurgery'.

With the availability of Healon® in particular (or other visco-elastic agents when clinically proven) the intracapsular surgeon is able to undergo the transition from intracapsular extraction to extracapsular extraction and IOL implantation with a degree of confidence bred from improved control that is lacking if viscosurgical methods are not utilized.

Economics of Viscosurgery

Financial considerations in the way ophthalmic surgery is practiced are ever with us but cost effectiveness in ocular microsurgery is a complex issue. Some components are easy to identify, others require a philosophical approach. A procedure that succeeds in its aims the first time will be much cheaper than one which has to be repeated or modified to achieve its effects. Thus, we all should be aiming for the highest standards that help us and our patients to achieve lasting success from minimum intervention. However, operations are perfected by experience and experience teaches slowly and always at the cost of some mistakes. From time to time we all face difficult moral issues with individual patients and specific problems, knowing that someone somewhere has greater experience or that more expensive equipment and materials will allow us to perform a better procedure.

This situation is common to all and is resolved by reacting to practicalities and necessities through our rationalizations may not stand up to the closest scrutiny.

Viscosurgery is a case in point. Ophthalmic surgeons differ in their attitude to the availability of viscosurgical agents. Reactions include: "I can manage perfectly well without these materials," "far too expensive," "air will do just as well," "only poor surgeons require these substances" and so on. Surely the point is that any adjunct to surgery, instrument or material that makes the operation safer and more reliable is not only worthwhile but should be utilized in our hands as we undoubtedly would have it utilized in our own eyes. The control that viscosurgery adds to an intraocular microsurgical procedure is unquestionable, provided that the materials used have passed the acid tests which must apply to all substances introduced into the intraocular environment.³

Looking beyond the immediate costs of disposable items used in the operating theaters, the repayment is to be found in the results. Proper development of viscosurgical materials should be encouraged, not only that they may be improved upon, but so that market forces will ensure that they reach the patients at a fair price.

Transition from Intracapsular to Extracapsular Extraction

The transition from intracapsular to extracapsular extraction is enhanced by the adoption of a form of extracapsular extraction, intERCapsular cataract extraction, especially as a prelude to IOL implantation, which offers intrinsic safeguards to which may be added the facility of "viscosurgery."

Viscosurgical Recommendations in IntERCapsular Cataract Extraction and IOL Implantation

Specific recommendations for the application of viscosurgical methods in intERCapsular (endocapsular) extracapsular extraction:

1. Reform the anterior chamber with Healon® (and occasionally enhance dilatation of a sluggish pupil) after completion of the incision to allow a safe anterior capsulotomy (Figures 1 & 2).
2. Utilize the viscosurgical agent to dissect recalcitrant cortical material from the fornices of the lens capsule.
3. Reform the capsular bag with Healon® for IOL insertion and protection of the posterior lens capsule (Figures 3 & 4).

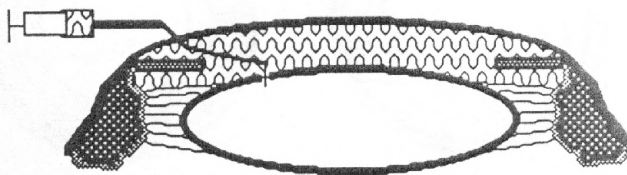


Figure 1. (Rosen): After a full limbal incision the anterior chamber is reformed and maintained by Healon® to allow a safe anterior capsulotomy by a cystotome attached to a Healon® syringe.

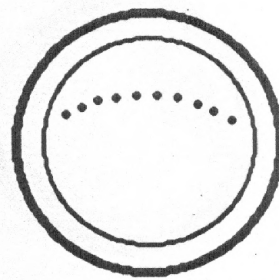


Figure 2. (Rosen): Linear capsulotomy under Healon®.

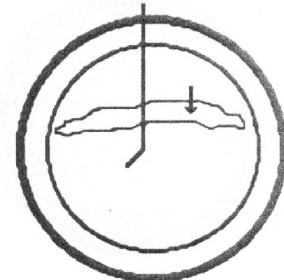


Figure 3. (Rosen): Healon® injected through a 30 gauge cannula into the 'empty' capsular bag to create the space for IOL 'in the bag' implantation.

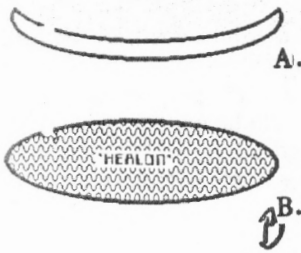


Figure 4. (Rosen): The capsular bag before (a) and after (b) Healon® injection, an illustration of a 'viscosurgical' principle.

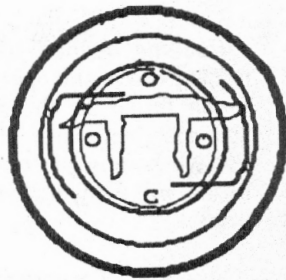


Figure 5. (Rosen): After IOL insertion into the capsular bag, an anterior capsulotomy extension is performed by Vannas scissors under Healon® protection of the corneal endothelium by recreation and maintenance of the anterior chamber, allowing room for maneuver.

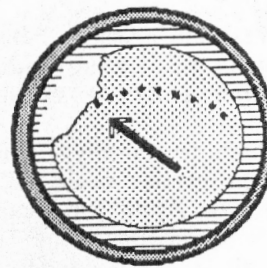


Figure 12. (Rosen): Axis for alignment of IOL.



Figure 13. (Rosen): IOL in position, haptic providing equatorial bag support where needed at site of dehiscence.

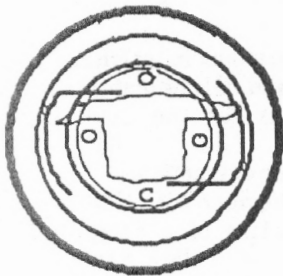


Figure 6. (Rosen): Anterior capsulotomy by tearing of the central capsular flap, once again performed under Healon® protection of the vulnerable tissues.

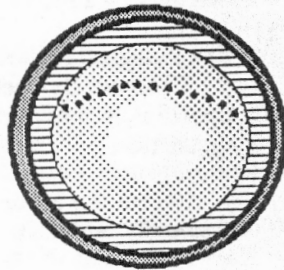


Figure 7. (Rosen): Posterior lens capsule tear.

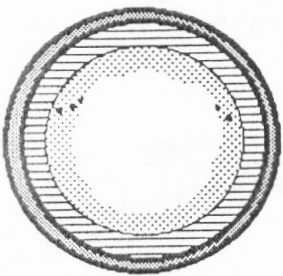


Figure 8. (Rosen): Controlled by careful capsulotomy and automated anterior vitrectomy followed by Healon® tamponade of vitreous to allow subsequent IOL implantation 'in the bag.'

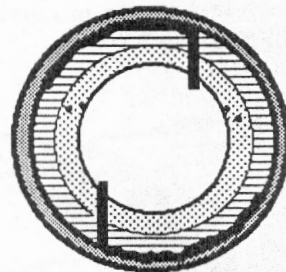


Figure 9. (Rosen): IOL implantation 'in the bag' or utilizing the anterior lens capsule minus its central area, in the sulcus fixation, under Healon® control.



Figure 10. (Rosen): Dehiscence of the zonule in the 10-11 o'clock meridians, during cortical lens matter aspiration.

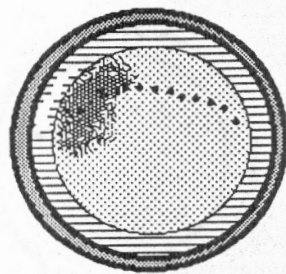


Figure 11. (Rosen): Reformation of the capsular bag contours by Healon® manipulation of the capsule, providing intra-operative support until IOL implantation and location of the lens haptic at this site for continuing support.

4. Reform the anterior chamber with Healon® to allow anterior capsulotomy (Figures 5 & 6).

5. In the face of a problem e.g. a small dehiscence appearing in the posterior capsule during the irrigation/aspiration procedure, inject Healon® to protect and retard the anterior hyaloid face from prolapse. (Figures 7, 8 & 9).

6. In the presence of a localized dehiscence of the zonule, reform the contours of an empty capsular bag with Healon® to allow IOL insertion in the bag safely and position the IOL haptic in the meridian of the dehiscence for maximum bag support. (Figures 10, 11, 12 & 13).

By adherence to these methods a surgeon making the transition from intracapsular to extracapsular extraction will find fewer problems and their better control when they do occur.

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EDITORIAL DISCUSSION

After a perusal of this useful and practical article Dr. Awan, the Editor, felt that the following discussion between him and Mr. Rosen on a few specific aspects of this paper's topic will be helpful to the readers of The Journal.

Editor: What are the limitations and the advantages of an "in the bag" approach?

Mr. Rosen: First the advantages of "in the bag." After such implantation, post operative experience shows that the capsular bag "shrink wraps" or encapsulates the IOL haptics and some of the optic providing "biological" insulation from uveal tissue which should reduce the release of the mediators of inflammation and breakdown of the blood ocular

barrier both by reduction of chemical and mechanical stimulation. I see little theoretical difference between haptics placed in the angle of the anterior chamber or iris supported IOLs each with uveal contact and a sulcus supported IOL with its haptics in potential contact with iris pigment epithelium. Indeed the latter is perhaps the least desirable because of the additional problems of pigment dispersion and the worry of later pigmentary glaucoma.

Secondly an "in the bag" IOL should be automatically centered and therefore optically satisfactory.

My third and equally important point is that most of the surgical procedure can be conducted in the capsular bag rather than in the chamber between posterior capsule and corneal endothelium. The logic of this procedure is obvious and to my mind obviates the value of phakoemulsification for this technique cannot be performed within the capsular bag in a satisfactory manner.

Finally, when I decided to change generally from intracapsular cataract extraction and IOL implantation to extracapsular cataract extraction and IOL implantation I wanted to take advantage of the full logic of extracapsular cataract extraction as I saw it. Namely, to maintain an intact posterior capsule, to avoid uveal contact during and after the procedure (no iridectomy or iridotomy, no instrument or IOL contact with uveal tissue), to encapsulate the IOL, i.e. make full use of the capsular bag and to find and utilize an extracapsular cataract extraction method that guaranteed the IOL would be "in the bag."

Coupled with the viscosurgical element, which is very important, I found in intercapsular surgery surgical satisfaction and control not only in my own surgery but that adopted consequentially by my trainees.

Therefore I feel there are no limitations for an "in the bag" approach if the intercapsular method is utilized and, as emphasized in the text of my paper, the flexibility it offers in complicated cases is an added advantage.

Editor: How does an eye with "in the bag" IOL behave after blunt trauma?

Mr. Rosen: Much as a phakic eye would behave, i.e. in a patient with a weak zonule or one which has been weakened by less satisfactory surgery, subluxation or dislocation of the IOL plus capsula may occur. Depending on its extent, an 'intracapsular' extraction of the IOL could be necessary and at least is possible.

Editor: What precautions are necessary, and what are the risks of "in the bag" IOL in an eye with zonular dialysis? What is the chance of future subluxation or dislocation of the IOL?

Mr. Rosen: Provided that the zonular dialysis is limited e.g. 30 degrees of arc, I would not hesitate to use the technique described above for IOL placement. The precaution is to use Healon® to reposition the

equatorial capsule and tamponade vitreous herniation. If vitreous herniation is deemed significant then a careful cleaning up operation using an efficient vitrector is indicated and "in the bag" IOL placement is unlikely to be possible with safety. However, if the dialysis of the zonule is limited, then the chance of later dislocation occurring should not be high unless trauma is the initiating factor.

Editor: How many degrees of zonular dialysis are safe for "in the bag" implantation? If the zonular dialysis is too great, what alternatives do you advise?

Mr. Rosen: About 30 degrees as answered above when there is more than this, bearing in mind the usual importance of an implant to the patient, from the optical point of view, I revert to an anterior chamber angle supported lens, usually the Symflex or Surefit type. At this stage you have to accept that the chances of later complications has risen, particularly cystoid macular edema and retinal detachment, but in my opinion these risks are acceptable in respect of the level of benefit usually achieved.

A further point I would make here, though not implied in your question, is the route to follow after a posterior capsule dehiscence. Here the intercapsular procedure provides the flexibility afforded by the continued presence at this stage of the anterior capsule. I usually use this to support an angulated all polymethylmethacrylate posterior chamber IOL in the sulcus, having cut a small notch into optical axis of the anterior capsule prior to implantation and after tamponade of the vitreous face by Healon® and anterior capsule, in layers.

Editor: Most ophthalmologists in Pakistan will be unable to afford Healon®, are there any alternatives?

Mr. Rosen: Effective viscosurgical agents are perhaps expensive in one sense but cost effectiveness is the point I would labour. A 'cheap' operation is one which succeeds first time and does not therefore involve the patient and surgeon in the untold and unquantified expense of later complications. Live now, pay later may be the way that some surgeons wish to work. The immediate joy of an apparently successful surgical procedure may be followed by delayed problems. Viscosurgery cannot but help improve the standards of every surgeon and as a community we should try and obtain viscosurgical agents at an immediately affordable price. The competition of market forces will produce this result eventually but meanwhile the temptation will be to use agents which are neither viscous, elastic or definably pure. Methylcellulose in various forms is widely used but this is not a single substance, available in consistent proprietary form. Only general comments can therefore be made about these substances which, in our analyses,² leave much to be desired from an ophthalmic surgical point of view. I would offer the dictum "do unto others as you would have done unto yourself" as a reliable surgical guide.



A Clinical Approach To Uveitis Diagnosis

Carmen Santos MD and Robert A Nozik MD

ABSTRACT: The great majority of uveitis cases fall into a small list of approximately 30 uveitic entities. The clinical characteristics of each of these "most likely uveitic entities" are different enough so that based on the detailed history, complete ocular examination, and the familiarity of the ophthalmologist with terminology, it is possible to construct a specific profile of each one. The arrangement of these profiles in a "naming-meshing system," proposed by Drs. Smith and Nozik, results in a very limited differential diagnosis and in the correct diagnosis in the great majority of patients, reducing considerably the amount of laboratory and diagnostic workup. (Pak J Ophthalmol 2: 124-132, 1986)

The diagnosis and management of uveitis can often be very frustrating for practicing ophthalmologists. This stems from the fact that uveitis is not a single disease but can be a manifestation of systemic disease as well as a primary ocular condition. Although a non-specific treatment of uveitis is available, different treatments are indicated for different types of uveitic syndromes, so as accurate and specific a diagnosis as possible is desirable.

The more common ways of classifying uveitic syndromes have been based on location (anterior vs posterior vs generalized) and on the type of inflammation (granulomatous vs non-granulomatous). Unfortunately this information alone is not enough to establish an etiological diagnosis. Frequently a battery of laboratory tests is ordered to rule in or out systemic diseases which may be associated with uveitis. This laboratory work-up is often costly and in many instances non-rewarding.

While there are many possible causes of uveitis, the majority of cases are limited to a selectively small group. Furthermore, there are some particular characteristics to each uveitic entity that frequently make it possible to differentiate one from another. Drs. Smith and Nozik have used these characteristics to construct a profile for each of the most common uveitic entities and have proposed a new system of classification which allows an accurate primary diagnosis in most cases. This system is called the "naming-meshing" system.¹

Several things are necessary in order to be able to

use this system to its fullest advantage. The ophthalmologist should become familiar with the important diagnostic features of the common uveitis entities. For example, it is very helpful to know that a red, painful and photophobic eye is characteristic of the acute anterior uveitis associated with the HLA-B27 antigen. The fact that posterior synechiae are extremely rare in pars planitis and Fuchs' heterochromic iridocyclitis and that increased intraocular pressure is characteristically seen in only a few uveitic syndromes (toxoplasmosis, herpetic uveitis, Fuchs', and Posner-Schlossman syndrome) is of great diagnostic value.

Besides being familiar with the most prominent characteristics of this group of diseases, a knowledge of uveitis terminology is important. The "naming-meshing" system consists of exactly what its name implies: an accurate "naming" of the patient's disease using as many descriptive characteristics as possible, and a matching of this profile with the profile of the uveitic entities included in the list of "most likely uveitic entities" (See Table).

When this is done correctly and accurately, the result is a small differential diagnostic list usually consisting of only two to four entities. Once this is achieved, the appropriate laboratory tests, special procedures or consultations can be performed to confirm the diagnosis. This "guided" laboratory work-up is significantly more limited and has a much higher positive yield than the battery of tests which is usually ordered for uveitis patients. Furthermore, some types of uveitis have such characteristic findings that no laboratory work-up is necessary (eg: snowbanks in pars planitis, heterochromia, cataract and absence of synechias in Fuchs' heterochromic iridocyclitis, and a focal necrotizing area of retinitis adjacent to an old scar in recurrent ocular toxoplasmosis.)

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UVEITIS TERMINOLOGY

The use of precise uveitic terminology is necessary to carry out the first part of this diagnostic system. Some of the most important ones are reviewed below.

Acute uveitis. Inflammatory attack lasts a few weeks or months and then disappears.

Chronic uveitis. Uveitis lasts months or years without completely clearing up between attacks.

Anterior uveitis. Inflammation of the iris and/or the ciliary body (iritis, iridocyclitis or cyclitis).

Pars Planitis. Also known as chronic cyclitis, and peripheral or intermediate uveitis. Refers to inflammation of the peripheral retina and/or is characterized by the presence of large inflammatory aggregates ("snowbanks") over the inferior pars plana.

Posterior uveitis. Inflammation of the choroid. Retinitis which secondarily involves the choroid (retinochoroiditis) is also included in this category.

Diffuse uveitis. Inflammation of the iris, ciliary body, and choroid.

Diffuse choroiditis. Generalized inflammation of the choroid.

Disseminated choroiditis. Many scattered foci of inflammation in the choroid.

Multifocal choroiditis. A few scattered foci of inflammation in the choroid with fairly normal tissue in between.

Granulomatous uveitis. Although this is a pathological term, some clinical characteristics are indicative of granulomatous inflammation. These are a tendency to form iris (Busacca) and pupillary (Koeppe) nodules or mutton fat keratic precipitates.

Keratic precipitates (K.P.). Inflammatory cell aggregates on the back of the cornea. They are large in granulomatous uveitis ("mutton-fat" or "mashed-potato") and small or medium-sized in non-granulomatous uveitis.

Posterior (K.P.s). Round inflammatory cell aggregates in the vitreous of patients with severe retinitis or retinochoroiditis.

Retinovasculitis. Inflammation of the retinal blood vessels. It may involve the veins or arteries or both. It may be generalized or limited to an area of retinitis.

Vitritis. Inflammatory cells in the vitreous usually secondary to a primary inflammation in the retina or uveal tract.

UVEITIS HISTORY

A detailed uveitis-directed history is the first step of the "naming-meshing" technique; the construction of a "working name" of the particular uveitis in a particular patient.

Geographic history is important in some types of uveitis. Histoplasmic choroiditis in the United States is seen in patients who have lived in the "histoplasmosis" belt which includes states bordering the Ohio-Missouri-Mississippi rivers; Behcet's syndrome is more common in Japan and the Mediterranean countries; VKH is also more common

in Japan and the middle east, while lepromatous uveitis, yaws, pinta and microfilaria-induced uveitis are seen almost exclusively in certain areas of Africa, Central and South America.

Family history is often relevant in uveitis caused by infectious agents. Toxoplasmosis, syphilis and cytomegalovirus may be transmitted congenitally, while tuberculosis may be acquired from exposure to an infected family member. Family history may also be important in uveitis associated with the HLA-B27 antigen. A spectrum of diseases including arthritis (ankylosing spondylitis), uveitis, urethritis (Reiter's syndrome), and colitis may be seen in family members who are positive for this antigen.

Demographic history (age, sex, and race) will often provide information of diagnostic value. Uveitis in children, for example, is frequently associated with juvenile rheumatoid arthritis (JRA), toxocara or sarcoidosis. Pars planitis and Fuchs' heterochromic iridocyclitis are seen more commonly in young adults. HLA-B27 associated iridocyclitis, acute multifocal posterior placoid pigment epitheliopathy (AMPPPE), birdshot choroidopathy, Vogt Koyanagi Harada (VKH), and Behcet's syndrome usually appear first in middle age, while large cell lymphoma (reticulum cell sarcoma) is characteristically seen in old age.

Some uveitic syndromes are more commonly seen in one sex than the other. The uveitis associated with JRA is seen predominantly in girls while that associated with ankylosing spondylitis and Reiter's syndrome is more common in males. Behcet's syndrome and sympathetic ophthalmia (SO) are also more commonly seen in men.

Race is also important in the diagnosis of some types of uveitis. HLA-B27 associated iritis is more common in Caucasians while sarcoidosis is seen more frequently in Blacks. Behcet's syndrome is found more frequently in Orientals and in Mediterranean races, VKH is seen in Orientals and American Indians, and coccidioidomycosis is more common in Filipinos.

Some aspects of personal history should also be noted at the time of initial examination. Pets and pet handling are important. Toxoplasma oocytes are excreted in cat feces, so that a history of handling cat feces by a pregnant mother would be significant in a patient with suspected congenital toxoplasmosis. In cases of suspected ocular toxocara infections, contact with unwormed puppies or kittens is of value, since it is caused by the larvae of *Toxocara canis* or *Toxocara cati*. A history of eating raw meat is important in patients suspected of having acquired ocular toxoplasmosis or during pregnancy in the mothers of patients with the possibility of congenital toxoplasmosis. A history of sexual practices may be of value in patients suspected of having uveitis secondary to a venereally transmitted disease.

The presence of systemic disease should be determined since many systemic diseases may be associated with uveitis. Previous ocular history should also be investigated in order to determine the possible

cause of the present uveitis (trauma in SO, previous dendritic or herpetic disease in herpes simplex virus or herpes zoster ophthalmicus uveitis), and the pattern of the uveitis (acute vs chronic, unilateral vs bilateral, initial attack vs recurrence, age at first attack). All of these factors may be very helpful in the differentiation of different uveitis entities.

OCULAR EXAMINATION

A detailed and precise ocular examination is of great importance in patients with clinical uveitis. Many clues that will point to the correct diagnosis can be discovered during the exam if the ophthalmologist is alert and observant. The presence or absence of certain features may be important depending on the particular situation. The most important of these signs will be briefly reviewed.

A systematic approach should be used and each ocular tissue examined sequentially so that no steps are omitted. The degree of inflammation should be noted; a red photophobic eye is characteristic of acute anterior uveitis such as that seen associated with the HLA-B27 antigen, while a quiet eye is the rule in posterior or intermediate uveitis. The conjunctiva should be examined for isolated nodules which may be biopsied for a definite diagnosis in cases of suspected sarcoidosis. Any corneal pathology should be noted. Scars indicative of previous trauma may be significant if SO is a diagnostic possibility. Dendritic or disciform scars would be indicative of previous herpetic ocular disease. Corneal sensation should be tested since it is usually absent or markedly diminished in cases of herpetic kerato-uveitis. The inferior cornea should be particularly examined for localized thickening and opacification of the posterior area in the area of thickening. This localized inferior corneal thickening has been termed by some authors as "prismatic effect" and is suggestive of long standing granulomatous iridocyclitis. It is believed to be the result of endothelial damage from the chronic presence of granulomatous keratic precipitates in this area and is very frequently seen in cases of ocular sarcoidosis.²

The type and distribution of keratic precipitates on the corneal endothelium may have diagnostic significance. In most uveitis the KP's are distributed in the inferior third of the cornea in a triangular shape with the base towards the limbus (Arlt's triangle). There are two principal exceptions to this rule; in both herpetic uveitis and in Fuchs' heterochromic iridocyclitis the KP's may be randomly distributed on the whole cornea. In Fuchs, furthermore, the keratic precipitates have a characteristic stellate appearance which is not usually seen in other entities. The size of the KP's helps to differentiate granulomatous from non-granulomatous conditions. Large keratic precipitates may have a greasy appearance (mutton-fat KP's) or may be granular, tending to get larger and coalescent towards the bottom of the cornea (mashed-potato type KP's). Differentiating between granulomatous and non-granulomatous inflammation

is important because granulomatous inflammation has diagnostic significance; the more common entities include sarcoid, tuberculosis, syphilis, VKH, sympathetic ophthalmia, herpetic uveitis and toxoplasmosis. It is important to remember that granulomatous uveitis may masquerade clinically as non-granulomatous disease but the reverse is almost never true.

The anterior chamber is next examined for the presence of flare and cells. A standard grading system which goes from 0 to 4+ has been described which is very useful in determining the severity of iritis as well as measuring the effect of therapy.³

The iris is another structure in which signs of granulomatous disease may be detected. This would be the presence of iris nodules which may be of three kinds: Koeppe nodules which are found at the pupillary margin, Busacca nodules which are seen in the iris stroma, and large iris granulomas which are most characteristic of sarcoid. Koeppe and Busacca nodules are usually clear or cream colored, while iris granulomas are pink, opaque and vascularized. It should be mentioned that small Koeppe nodules may sometimes be seen in non-granulomatous uveitis, particularly Fuchs' heterochromic iridocyclitis; the small Koeppe nodule therefore, has less diagnostic value. The iris should also be examined for any areas of stromal or pigment epithelial atrophy which may be seen in Fuchs' or herpetic uveitis. The ophthalmologist should also be familiar with the normal architecture of the iris which may be highly variable. An absence of the normal iris "crypts" and "valleys" may be suggestive for the chronic granulomatous inflammation of sarcoidosis.

Posterior synechias may form in almost any kind of anterior uveitis. Their absence, however, is characteristic of three entities: pars planitis, Fuchs' heterochromic iridocyclitis and glaucoma-cyclitic crisis (Posner-Schlossman syndrome).

Cataracts are frequently associated with uveitis and are characteristically of the posterior subcapsular type. They may be secondary to the inflammatory process itself or the consequence of steroid use. Cataract is a frequent early finding in Fuchs' heterochromic iridocyclitis.

The vitreous should be evaluated for cells and other inflammatory debris. The amount and location of cells should be noted. In pars planitis, vitreous cells are most numerous in the retrolental area while in retinochoroiditis the cells are concentrated in the posterior vitreous, usually over the active retinal lesion. The diagnosis of large cell lymphoma should be considered in older patients who develop vitritis later in life.

Indirect ophthalmoscopy should be performed in all patients with uveitis. Choroiditis, chorioretinitis, retinitis and retinochoroiditis are terms used to describe posterior lesions depending on the primary tissue involved and whether or not there is extension of inflammation to adjacent tissue. Presumed ocular

histoplasmic lesions are located in the choroid while toxoplasma causes a primary retinitis which, often, eventually involves the choroid, leading to the characteristic retinochoroiditic scar. Toxoplasmic scars typically have a white center corresponding to scleral tissue, surrounded by hyperplastic pigment epithelium. This is the result of the severe necrosis of retinal and choroidal tissue which characterizes these lesions. The ophthalmoscopic picture of an active white, fluffy, retinal lesion adjacent to an old pigmented scar is almost pathognomonic for recurrent ocular toxoplasmosis. A marked vitreous reaction is usually present over the active lesion. The vitreous reaction may be severe enough in some cases to prevent adequate visualization of the retinal lesion. In these cases a "light in the fog" effect has been used to describe the ophthalmoscopic picture.

The acute retinal necrosis syndrome is a recently described entity in which there is also an acute retinitis which may be associated with severe vitritis. It may be unilateral or bilateral and some evidence suggests that a virus from the Herpes group is the etiologic agent.⁵

The intraocular pressure may provide data of diagnostic value in some types of uveitides. In most uveitis, at least in the more acute stages, the intraocular pressure is decreased due to inflammation of the ciliary body, which reduces aqueous secretion. The pressure, however, is characteristically elevated in the iridocyclitis associated with toxoplasmosis, herpetic uveitis, and glaucomatocyclitic crisis. In chronic types of uveitis the intraocular pressure may be elevated as a result of secondary uveitic glaucoma. This may result from peripheral anterior synechia, seclusio pupillae from posterior synechia or trabecular scarring or clogging by inflammatory products. Gonioscopy may be helpful in detecting any of these conditions and may also reveal the presence of filiform vessels crossing the anterior chamber angle in cases of Fuchs' heterochromic iridocyclitis.

NAMING AND MESHING

Once the ophthalmologist has obtained a pertinent history and done a uveitis-oriented ocular examination, he has the essential tools for constructing a working "name" for the patient's uveitic condition. In essence, a profile of the patient's uveitis is constructed from the pertinent information obtained from the history and physical examination. This profile is then matched to the "profile" for the uveitic entities listed in the table. This latter profile is based upon these characteristic clinical features. Several examples will follow.

Example 1. A 31-year old caucasian female has a history of floaters in her left eye and "foggy vision" for about a year. Her eye has never been red, painful or photophobic. Her symptoms are only slightly improved with topical steroids. There is no history of

arthritis or any systemic illness. On examination, her vision in the affected eye is 20/30. Fine keratic precipitates with stellate projections are scattered over the entire corneal endothelium. Mild iris stromal atrophy is observed in the affected eye. There is +1 flare and cells in the anterior chamber and there is no evidence of posterior synechia. Early posterior subcapsular changes are seen in the lens and a significant amount of cells are noticed in the retrolental area. Intraocular pressure is 14mmHg OD, and 22mmHg OS. No fundus abnormalities are seen on indirect ophthalmoscopy.

This clinical picture would be named as follows: Chronic, nongranulomatous, unilateral, iridocyclitis in a 31 year old female associated with cataract and increased intraocular pressure. When this profile is matched to the ones in table 1 it is obvious that the best match is Fuchs' heterochromic iridocyclitis. Other diagnostic possibilities based on this meshing system would be an immunologic uveitis, or less likely, glaucomatocyclitic crisis. Herpes simplex or zoster uveitis would be unlikely in the absence of corneal lesions or scarring and the lack of skin lesions in the distribution of the first division of the trigeminal nerve. Laboratory tests, if indicated in a similar but less clear-cut case, would consist of sedimentation rate and a search for systemic condition.

Example 2. A 37-year-old male of middle eastern background is seen because of pain and redness of both eyes. He has a history of previous similar attacks within the past year which have resolved with topical steroids. He also gives a history of aphthous ulcerations of the mouth and occasional pain in his knees. Ocular examination is remarkable for conjunctival and episcleral vessel injection, medium-sized KP's and +4 flare and cells in both eyes. In addition, a small hypopyon is noticed in the right eye. Indirect ophthalmoscopy reveals perivascular sheathing of vessels in both eyes and occlusive vasculitis, especially of the arterioles. Optic nerve pallor is observed, more prominent in the right eye.

This second clinical syndrome would be named as an acute, recurrent, bilateral, non-granulomatous uveitis and retino-vasculitis in a 37-year-old male associated with aphthous ulcerations of the mouth and arthralgias. When we match this clinical picture with those in the table, we see that Behcet's syndrome would be our most likely diagnosis. Other possibilities which should be included in the differential diagnosis are the other causes of retinovasculitis, also obtained from the table, such as tuberculosis, syphilis, sarcoidosis and Eale's disease. In this particular case a laboratory work-up is extremely important due to the morbidity and mortality associated with some of the diagnostic possibilities.

Example 3. A seven-year-old caucasian girl is examined because of diminished visual acuity during a school test in both eyes. There is no history of any

systemic illnesses. Her visual acuity is 20/30 in the right eye and 20/60 in the left eye. Early band keratopathy is observed in both corneas. Fine KP's are seen on the corneal endothelium and the anterior chambers show +2 flare and cells. Posterior synechias are seen in both eyes as well as posterior subcapsular changes which are more advanced in the left eye. Anterior vitreous cells are observed and what can be seen of the fundus appears normal. Intraocular pressure by applanation is 23 and 20 in the right and left eyes respectively.

An appropriate name for this patient's uveitis is a chronic, nongranulomatous, bilateral iridocyclitis in a seven-year-old female associated with band keratopathy, posterior synechias and cataract. It better fits the diagnosis of uveitis in young girls which may produce uveitis identical to that seen associated with juvenile rheumatoid arthritis. An antinuclear antibody test is often positive in these patients after repeated testing, while the rheumatoid factor is negative. Other causes of bilateral iridocyclitis with synechiae in children are rare and would consist mainly of sarcoidosis.

Example 4. A 40-year-old caucasian male is examined due to pain, redness and photophobia of the right eye. He has had previous similar episodes in the past five years which have been usually in the right eye but occasionally have involved the left eye, but never at the same time. They usually respond to treatment with intensive topical corticosteroids. The patient also gives a history of chronic low back pain. Examination discloses a red, photophobic right eye with small and medium-sized KP's, +3 flare and cells and posterior synechias. Retrolental cells are also present in this eye. The left eye looks quiet but some pigment deposits are seen over the anterior lens surface which seem to be evidence of previous posterior synechias.

The clinical picture described above would be named as an acute, recurrent, bilateral, nongranulomatous iridocyclitis in a young male associated with low back pain. When this profile is matched with those in the table we see that it is most compatible with ankylosing spondylitis or uveitis associated with the HLA-B27 antigen which is positive in over 90% of these patients. An x-ray of the sacroiliac joints would also help in the diagnosis. Other conditions which could present as bilateral nongranulomatous uveitis, such as syphilis and sarcoid should be considered but are less likely.

Example 5. A 45-year-old black female is seen because of decreased vision and floaters in both eyes since two years previously. She responds poorly to topical medications but has significant improvement when systemic steroids are used. Flare-ups occur shortly after the systemic steroids are discontinued and the patient is never completely free of symptoms. Her visual acuity is 20/50 in both eyes. There is no

history of systemic illness. Large mutton-fat KP's are present on the inferior 1/3 of the cornea with localized thickening of the cornea in this area. Minimal flare and occasional cells are seen in the anterior chamber. Koepe nodules and posterior synechiae are observed in the pupillary area. A +3 vitreal cell reaction is seen in both eyes. Indirect ophthalmoscopy reveals the presence of small yellow choroidal lesions scattered throughout the peripheral fundus in both eyes.

The working name for this patient's uveitis would be a chronic bilateal granulomatous diffuse uveitis in a 47-year-old female and the differential diagnosis would include all causes of granulomatous uveitis. In the United States, the most common cause is probably sarcoidosis. Others to be considered would include tuberculosis, syphilis, VKH and SO. A diagnostic work-up is particularly important in a patient like this since the treatment for the various diagnostic possibilities is different.

CONCLUSION

A system has been presented for diagnosis and management of clinical uveitis which, we believe, will result in the correct diagnosis of the primary systemic or ocular condition in the vast majority of cases seen in an ophthalmologist's office or in a general ophthalmology clinic. The originators of this system have called it the "naming-meshing" system.¹ The system is based on history and physical examination rather than on laboratory testing. It entails the construction of a detailed "working name" which includes as many descriptive terms as possible which characterize the patient's particular disease. This profile is then matched to the profile of the most common uveitis entities which are summarized in the table. It is possible then to establish a small list of differential diagnosis and a limited laboratory work-up should be enough to arrive at the correct diagnosis. It should be remembered, however, that this system will not work for certain uveitides since there are hundreds of uveitic entities and we are only dealing here with a few of them. They are, nevertheless, by far the most common ones, and this system has proven to be extremely valuable in the diagnosis and management of uveitis patients for those of us who have learned it and use it.

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Likely Uveitic Entities (part 1)									
(Arranged anatomically Anterior uveitis first, then Posterior and finally Diffuse)									
Uveitic Entity	Ana-tomical	Chron-ology	G/NG ¹	M/F ²	U/B ³	Lab Work-up*	Therapy	Complications	Prognosis
I. Viral/ Non-specific/ Trauma	I	A	NG	—	U	Diagnosis based on antecedent history	Depending on severity, simple observation, mydriatic cycloplegic only, or rarely local steroids.	Usually none.	Good. Rare recurrence.
II. Rheumatoid, Spondylitis (Ankylosing Psoriatic)	I-Cy	A-R	NG	M	B	ESR, HLA-B27, SIJt x-ray, Rheumatological consultation.	Intensive local steroids (at least every 2 hours) in attacks. May need periocular and even short course high dose systemic steroids in severe cases for acute attacks. No Rx between attacks unless case is converted to chronic iridocyclitis due to inadequate treatment of acute attacks — then treat as chronic nongranulomatous iridocyclitis — See V if chronic.	May be none if acute attacks are treated promptly and vigorously. Cataracts, posterior synechiae glaucoma, rarely phthisis.	Good, if attacks are treated vigorously and promptly. Poor if case is allowed to become chronic (see V).
III. Reiter's Syndrome	I-Cy	A-R	NG	M	B	ESR, HLA-B27, S-I SIJt x-ray. Medical consultation, (chlamydial CF, culture). ⁵	Same as II.	Same as II.	Same as II.
IV. Immunologic (altered iris vascular permeability, focus of infection, immunogenic focus)	I-Cy	A-R,C	NG	—	U	Search for systemic focus, ESR, Medical Consultation.	Same as II but there is a great danger of converting to a chronic nongranulomatous iridocyclitis (if chronic see V).	Same as II.	Same as II. See V if chronic.
V. Juvenile Rheumatoid Arthritis, Uveitis in young girls	I-Cy	C	NG	F	B	ESR, ANA, Pediatric consultation.	Use medium strength mydriatic/cycloplegic agent at least night-time even in remissions. Intensive (at least every 2 hr.) steroids and more frequent mydriatic cycloplegic during exacerbations. May need periocular or systemic steroids during exacerbations. Some patients do well on low dose chronic systemic steroids.	Posterior synechiae, secluded pupil, pupillary block glaucoma, secondary glaucoma, cataract, band keratopathy, severe danger of ultimate phthisis.	Very poor, even with close follow-up, especially for particular cases. Patients handle intraocular surgery very poorly. Due to extreme danger of ultimate phthisis, avoid glaucoma surgery if at all possible.
VI. Glaucomato Cyclitic Crisis (Posner Schlossman) Syndrome	I, I-Cy	A-R	NG	M/f*	U	HLA-Bw54 © in 40%.	Topical steroids, mild mydriatics, anti-glaucoma therapy during acute attack only.	Very rarely cupping and glaucomatous field loss.	Good
VII. Heterochromic Cyclitis	Cy, I-Cy	C	NG	—	U	—	No treatment needed in vast majority of cases. Night-time dilatation with short acting mydriatic cycloplegic agents only if Koepe nodules are present. Periocular steroids for posterior polar edema (very rare).	Cataract. Glaucoma.	Good, with no treatment. Cases do well with cataract surgery.
VIII. Herpetic Uveitis (Simplex or Zoster)	I, I-Cy KU	A-R,C	G/NG	—	U	Diagnosis based on previous history of herpetic corneal disease. Test for corneal sensation. Skin lesions in zoster. Look for evidence of previous keratitis.	Topical antivirals. Systemic acyclovir? Topical steroids may be necessary in some cases and antiviral coverage should be given. Glaucoma therapy if necessary.	Glaucoma cataract. Corneal scarring and vascularization.	Fair, in absence of glaucoma or scarring. Poor if there is glaucoma, scarring, frequent recurrences and poor response to treatment.
IX. Chronic Cyclitis/Peripheral Uveitis/Pars Planitis	Cy	C	NG	—	B	Fluorescein angiography	Periocular (posterior sub tenons) repository steroids for secondary macular or disc edema only. Occasional short course systemic steroids for exacerbations or immunosuppressive agents or cyclotherapy may be considered for severe recalcitrant cases.	Macular cystoid and cyst formation. Cataract, secondary steroid glaucoma.	Good, if cases are treated vigorously for cystoid macular edema. Cases do well with cataract surgery.

*See last page for key to abbreviations

• A = acute; A-R = acute-recurrent; C = chronic.

* See tabulation of lab abbreviations on last page

¹Granulomatous (G), Nongranulomatous (NG)

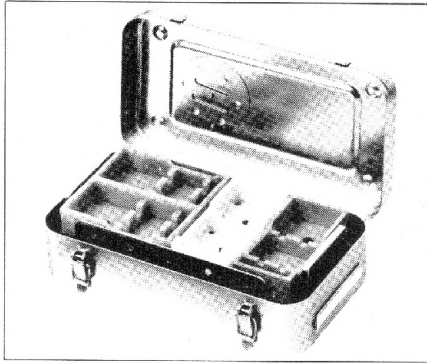
²Male (M), Female (F)

³Unilateral (U), Bilateral (B)

*If lower case, this is of lesser importance.

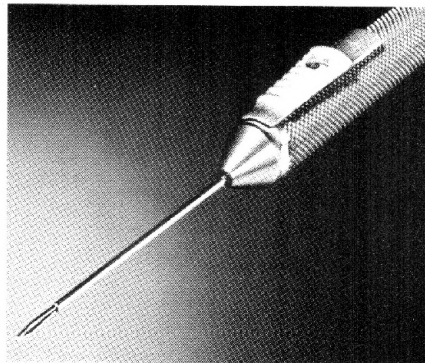
⁵Laboratory tests in parenthesis are those of lesser importance.

Likely Uveitic Entities (part 2)									
(Arranged anatomically Anterior uveitis first, then Posterior and finally Diffuse)									
Uveitic Entity	Ana-tomical	Chron-ology	G/NG ¹	M/F ²	U/B ³	Lab Work-up*	Therapy	Complications	Prognosis
X. Toxocara	Cy Macular End.	C	G/ng ¹	-	U	ELISA test for toxocara (vitreous aspiration). ESR, CBC, EOS (AC tap for calcification) (Ocular x-ray dental film - for calcification).	Periocular steroids and/or systemic steroids during active period.	Cataract, macular scar, glaucoma, retinal detachment.	Poor. If the macula is involved. Poor if endophthalmitis is present. If the involvement is peripheral, the prognosis is good with treatment of the severe phases of inflammation. Once the case is quiet for an extended period, intraocular surgery is tolerated well.
XI. Toxoplasma	R.R.Ch	A-R	G	-	-	Toxoplasma dye or FA test or ELISA test for toxoplasmosis.	Vigorous treatment for lesion of 1) the macula, 2) maculopapillary bundle, 3) optic nerve, 4) severe endophthalmitis, and 5) macular edema secondary to lesions superior to the macula. Standard treatment: Daraprim and sulfa, can use sulfa alone, can use tetracycline, and can use systemic or periocular clindamycin. Concurrent systemic steroids may be a benefit. Avoid periocular steroids. Avoid systemic steroids without anti-microbial coverage.	Secondary anterior uveitis with posterior synechiae secondary glaucoma, secondary cataract. Retinal scarring. Detached retina.	Good, if the active lesion(s) is away from the macula or optic nerve. Fair to poor if the lesions in or near the macula. Recurrences are common and posterior, macular threatening lesions may recur over and over and eventually affect the foveal region.
XII. Acute Retinal Necrosis (ARN)	R.R.V.A	A	C/ng ¹	-	U/B B < 33%		Systemic acyclovir. Intravitreal acyclovir? Prophylactic photocoagulation posterior to area of necrosis if visibility allows. Retinopexy with broad posterior scleral buckle if RD develops.	Retinal detachment in 75%. Optic atrophy. Macular pucker.	Poor
XIII. Large Cell Lymphoma (Reticulum Cell Sarcoma)	R.Ch.V, I-Cy	C	NG/g ¹	-	B	Vitreous Biopsy. Systemic evaluation for lymphoma. LP for cells.	Radiation therapy to orbit and CNS.	CNS disease.	Poor. 90% of patients will die within 1 year of CNS disease if untreated.
XIV. Cytomegalic Inclusion	R.R.-Ch	A-R	G	-	B	Virus studies of urine, serum (tears), Pediatric Consultation, CF test.	q-(1,3 Dihydroxy-2-Propoxy-methyl) Guanine (DHPH) is a new effective antiviral agent. Recurrences may occur after the drug is stopped or tapered. High doses of systemic steroids and immunosuppressive drugs are contraindicated and may in fact cause exacerbation. Transfer factor may be helpful.	Same as XI.	Same as XI. Represents a poor prognostic sign for patients with the acquired immune deficiency syndrome (AIDS).
XV. Retinal Periphlebitis (Eales's Disease)	RV	C	NG	M	B	PPD, Chest x-ray, Fluorescein Angiography	Retinal photocoagulation, cryoablation?	Vitreous hemorrhage. Retinal hemorrhages. Retinal detachment.	Fair
XVI. Birdshot	R.Ch, RPE, RV, (I-Cy)	C	NG	-	B	Flourescein HLA-A29	Systemic and/or periocular steroids (often poor results).	Cystoid/cystic macular edema and scarring.	Fair to poor.
XVII. Acute Multi-focal Posterior Placoid Epitheliopathy	RPE, I-Cy	A	NG	-	B	Flourescein (R/o systemic virus infection)	-0-	Macular pigment alteration and scarring.	Fair to poor.
XVIII. Geographic Choroiditis, Serpiginous Choroidopathy, Helicoid Choroidopathy	Ch,RPE	A-R	NG	-	B	Flourescein	-0-	Macular scarring.	Fair to poor. (Good if macula spared.)



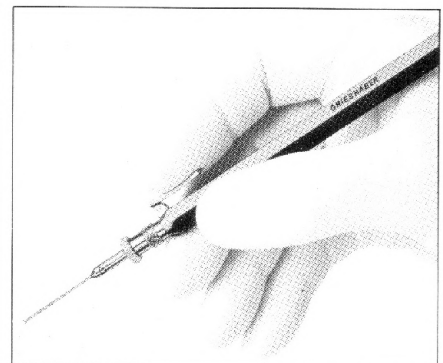
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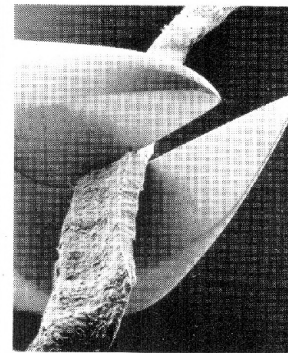
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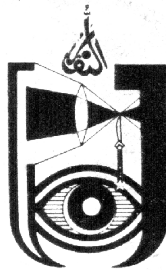


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