The Ocular Communique

The Journal of the Maharashtra Ophthalmological Society

Editor : Dr. B. K. Nayak

Volume 6, Issue 1. Sep - Dec, 2009

Contents

Vol. 6 No.1, Sep - Dec, 2009

Instructions to Authors

The Journal of Maharashtra Ophthalmological Society publishes three issues in a year. It accepts original articles, rare case reports and short reviews. All the articles are subject to editorial revision.

1. All the papers should be accompanied by a statement that, they have not been published in any other journal or presented in any conference and that, if accepted they will not be offered to any other publisher / conference without the consent of the Editorial Board. All the authors should sign in this statement.

   For the articles already published earlier elsewhere, written permission of the relevant authority should be accompanying the article.

2. Three sets of the articles must be sent with the statement.

3. The matter must be typewritten in double space on bond paper with adequate margin.

4. The title along with the author's name, address of the institution, and reprint request must be typed in a separate page.

5. The manuscript in the case of scientific papers must be in the following format:

   Title, Abstract & Keywords, Introduction, Material and Methods, Results, Discussions and Reference. The abstract must be short, conveying the aim, method of study, result and conclusions.

6. Table should be typed on separate pages and numbered, titled and with suitable column headings.

7. Photographs should be submitted in quarter size (3 ¼ " x 4 ½ ") on glossy paper. Indication of top and figure number should be shown on the back of the photograph. No author's name or institution must be mentioned in the article or on the photograph. Title of the article may be written at the back of the photograph. Legends must be submitted on a separate paper.

8. Colour photographs shall be published at author's cost. (Film Scanning Charges, extra printing charges etc.) The amount should be paid in advance.

9. All contributions will be accepted for publication only after review by two members of the editorial board.

Reference: In accordance with the Vancouver agreement the Journal of Maharashtra Ophthalmological Society has adopted standard bibliographic patterns as follows:

A paper or book cited in the text is referred to by a superscript in numerical order, in which it is first cited in the text, not in alphabetical order by the authors name. For convenience in preparing the typescript the reference number may be typed between parenthesis on the line, not superscript. The titles of the journal will be abbreviated in accordance with the style of Index Medicus. In the typescript they could either be abbreviated in that style or given in full. Authors submitting papers are requested to adopt it in order to facilitate editing.

Examples are given hereunder.

For articles: Nayak B K, Ghose S, Singh J P An evaluation of the NR-1000F Auto Refractometer in high refractive errors. Br J Ophthalmol 1987; 9:682-4.

For books: Mandel Wanger etal,Atlas of corneal diseases, W.B. Sanders, First edition, 1989, 80-2.

Reprints: if required, will be supplied at extra cost. This requirement must be mentioned while submitting the article.

This journal is edited, printed and published by Dr. B. K. Nayak on behalf of Maharashtra Ophthalmological Society.

Cover designing by Mr. Rajesh Dave from Surgicon

Printing by : Balaji Printers, 6 / 4, Khimji Nagji Chawl, S. B. Marg, Opp. Phoenix Tower, Lower Parel (W), Mumbai - 400 013. Tel.: 2431 1963, Mbl.: 98201 48846, E-mail : lokarts@gmail.com

JOURNAL OF MAHARASHTRA OPHTHALMOLOGICAL SOCIETY

Vol. 6 No. 1, Sep - Dec, 2009

Editorial Office :
Dr. B. K. Nayak
P.D. Hinduja National
Hospital, Veer Savarkar
Marg, Mahim, Mumbai - 16.
Tel.: 24447127, 24447176
Fax: 91-22-24440425
Mbl.: 9987315599
E-mail:
drbknayak@gmail.com

Editorial

Dear Colleagues,

Greetings from the Editor's Office of JOMOS.

As per the new guidelines of Medical Council of India, publication in medical journals has been made compulsory for promotion of teachers in various grades. There are two aspects for publishing articles in any peer-reviewed medical journal. To begin with, there has to be something new to say on the basis of a proper and scientific research work. Further, this work has to be written down properly as per the requirements of a peer reviewed journal for getting accepted for publication. Unfortunately, both aspects are deficient in todays' undergraduate and post graduate training programmes in most of the institutions in India. To fill this gap Indian Journal of Ophthalmology (IJO) and P.D.Hinduja National Hospital & Medical Research Centre (PDHNH) who have been conducting regular research methodology and scientific writing workshops have taken the initiative and have come up with another Research Methodology Workshop which is scheduled on 1st and 2nd of May 2010 at Mumbai.

This workshop is the basic programme under the SCIENTIFIC WRITING & SKILL DEVELOPMENT PROGRAMME which will be an ongoing educational series in three phases over the year 2010. The intermediate and advanced understanding of research methodology and medical writing will be organized in due course of time in two subsequent workshops in 2010. The complete programme will be helpful for those aspirants who want to understand the nitty-gritty of scientific research and publication. This will also help in undertaking editor's job subsequently.

It is my sincere request to all those who are interested in the field to take the benefit of the opportunity and contact the editorial office for further details.

Regards,
DR. BARUN KUMARNAYAK
EDITOR – IN – CHIEF
JOMOS, OCULAR COMMUNIQUE

Published by Maharashtra Ophthalmological Society

How to write Good Scientific articles?

Dr. Barun Kumar Nayak

The logical conclusion of any research would be publication of a paper in a scientific Journal. At the same time it is unethical to have completed the research and not to have published the paper subsequently. It is betrayal of trust of the patients who have given their consent to participate in the research for altruistic reason for advancement of science and it also amounts to wastage of resources in the form of time and money. It has been rightly said "every paper is publishable – somewhere". The author should choose the journal judiciously for publication depending upon the subject of the paper and the readership of the journal. If the paper has been written appropriately, the chances of it getting accepted in a good journal becomes very high. It is understandable that unless the study has been done writing cannot be done, but, if the study is sound, then poor writing should not come in the way of publication leading to its rejection. The aim of this communication is to suggest some points to improve the quality of an article.

It is mandatory that for any scientific article written in a peer reviewed journal there has to be something (new) to say. Please remember Scotts Fitzgerald's maxim "Write because you have something to say, not because you want to say something". To justify this statement you have to have a good research. When your research is over you want to say to the world that you "have a problem" and "you have an answer too" If you feel that it is worth saying then you must publish. You should package the message in such a manner that it is appealing to everyone. As a basic principle short, substantive and clear papers are the one that everyone wants to read. You must remember that Scientific writing should be very clear and written in simple language. Earlier Quintilian model (1ce) of writing was followed which says "Clear writing is a writing that can be understood" However, now we follow the Woodford model (1967) which says "Clear writing is writing that cannot be misunderstood".

First of all the person who is writing the article should be familiar with the subject as well as the direction in which research is being done in that field of particular subject. The writer should know the details of the work done in that field. Based on the lacunae in the subject in literature the author should formulate the "new questions" that he / she intends to answer in the paper. The author needs to be at least as knowledgeable as the reviewers are with regards to that particular question.

After choosing a particular journal you must read instructions to authors minutely. The standard format of an article is in the form of IMRAD: where

- I - Introduction (What question was asked?)
- M - Methods (How was it studied?)
- R - Result (What was found?)
- A - And
- D - Discussion (What is the conclusion?)

The other components of the article are title, abstract, keywords, references, and tables and figures.

Title: Will be read by all and reader will decide on the basis of title to read the full text of the article. Therefore, it should be attractive, meaningful, should convey the gist and also include some of the keywords.

Abstract : This forms the first impression about the article. It should be complete and convey the most interesting part of the study. It should be the summary of the study and must include objective and hypothesis, the study designs, methods, main results, and conclusions. The abstract should stand on its own without any reference to any other parts of the paper. Therefore due care should be given while writing the abstract.

Introduction: It should be precise. The reviewer does not appreciate lengthy introduction. Each sentence or statement in the introduction should be supported by appropriate references. First paragraph should state the problem in one sentence with one reference. It should include epidemiology or the clinical significance in one sentence with 2-3 references. It should be followed by uncertainty in literature in one sentence with 2-3 references. Second paragraph should include review of literature to support either the uncertainty in literature or the hypothesis in a maximum of up to five sentences. The summary of literature should be done with specific questions in mind.

- Did the studies answer the scientific questions in mind?
- Are there any conflicting results from different studies?
- Is there an untested population?
- Is there a method that has not been tried?

Dr. Barun Kumar Nayak
P. D. Hinduja Hospital and Medical Research Center, Mahim, Mumbai.

The third paragraph of introduction should mention the purpose of study in one sentence (e.g." The purpose of the study was to find …." Or "In the present study, we compared….")

Problem in real world / literature should lead to the present study logically. Therefore the authors should make clear links between the problem and the solution, question asked and the research design, prior research and this study. The authors should be selective, but not exhaustive, in choosing studies to be cited and amount of detail to be included. In general, the more relevant an article is to the present study, the more space it deserves and the later it appears in the introduction. The common mistakes which are committed by the authors in writing the introduction are:

- Too lengthy
- "Text book" knowledge
- Review of all literature
- End with quick summary of own results.

In a nutshell it should evolve around the research question.

Materials and Methods: It should be elaborate and should include the answer of the following questions:

- What was done (study design) Prospective / retrospective, randomization, masking?
- Who (study population)?
- When (study time frame)?
- Where (Hospital-based, tertiary care, location of study)?
- How (detailed procedure)?

Identify the intervention leading to the main outcome. The Materials and Methods should be so clear that the reader can duplicate it if interested. It should include the following:

- Preparation
- Ethics approval
- Protocol
- What is the purpose of each procedure?
- How does the procedure answer the question?
- Can subtitles signal topics in subsections?
- Methods of measurement
- Independent variable
- Dependent variable
- All controls
- Methods of analysis of data
- Computer programs
- Statistical advice

The usual inadvertent omission in this section by the writer are :

- Ethical approval
- Sample size calculation
- Randomization method
- Blinding
- Questionnaires
- Interventions
- Clinical assessments
- Statistical methods

Result: Present the crude data leading to the specific data. The data should also include the statistical analysis with its significance. This portion is only meant for data presentation. One should never interpret data and draw conclusions in this segment. Never repeat tables and figures in the text. One should not state the statistical techniques, which were not used in the analysis. P- value should be reported up to 3 decimal places and highly significant values should be reported as $p<0.001$ even if the values are much less (e.g. $p <0.0000039$). You should always write central tendency (mean, median), spread (SD, range, inter-quartile range), precision, (standard error, 95% confidence interval) of your data.

Discussion: A particular pattern should be followed in the discussion so that a logical conclusion can be derived at the end. No conclusion can be drawn without a proper discussion. First paragraph should summarize the most important findings. These findings should be interpreted correctly. Based on the findings and interpretations the conclusions should be drawn. One has to decide whether each hypothesis is supported, rejected, or no conclusion can be drawn with confidence (inconclusive). Second paragraph should mention the plausible explanation. The reasons for similarity of results with those in other studies should be discussed and at the same time plausible explanation should be postulated to defend the results that are not in agreement with those of other studies. It is important to see how these results relate to expectations and to the literature cited in introduction (agreement, contradiction and exceptions). Possible reasons for unexpected findings should be discussed. Third paragraph should mention the limitations, and strengths of the present study. Fourth paragraph should include clinical and research implications and future studies. It should not be over generalized. Conclusion should be the last paragraph along with certain suggestions.

- Where do we go next?
- What questions remain? (the best studies open up new avenues of research)
- Did the study lead you to any new questions?
- Briefly suggest new experiments to further address the main questions.

References: References should be given properly and in the order as it appears in the text. Avoid "second hand citation", and try to find the original papers.

Figures and Tables: Figures are preferable to table and tables are preferable to straight text. The figures and tables should be self – explanatory. Figures and tables should be simple, make clear points, style should be similar between tables, show main points in figures and do not repeat the text.

General considerations: Always write in simple language. Someone has said that "The official language of science is not English but it is bad English"

Always write in active voice and use the first person where necessary (e.g. we examined…" or "our current proposal …."). Simple sentences should be used. One should avoid scientific jargons and abbreviations. The nouns and verbs should agree (e.g. "The data are …" or "None is …..")

Present tense should be used to report background that is already published (e.g. "IOP is a risk factor for glaucoma"). The future tense is used for work that is intended to be done (e.g." we will test the hypothesis …."). Past tense should be used to describe the results of a specific study, especially the authors own (e.g." We found that ….")

The title of the article should be adequately descriptive and should make a person want to go through the paper. One should choose and decide the appropriate journal, where it is to be sent for publication. One should procure that journal and read "instructions to authors" minutely. Follow the style of that particular journal meticulously. Do not ignore any of instructions mentioned there.

Finally I would like to deal briefly with the process and reviewing an article. Following points are considered by the reviewer.

Technical aspects:
- Is the writing clear and grammatically correct?
- Is introduction and methodology adequate?
- Whether number of cases and follow-up conclusions can be derived from data?
- Are literature references adequate?
- Is the study ethical?

Value:
- Originality of material?
- Importance of articles?What papers are accepted?
- Which covers an important subject
- The message is new and original
- Relevant to the journal's readers
- Well presented and interesting
- Covers topical subject
- Covers neglected area

What papers are rejected?
- Not important
- Not new enough
- Not sufficiently interesting for readers
- Topic is interesting but the methodology is inadequate
- Too super –specialize or narrow
- The findings are difficult to apply clinically

Revision and Resubmission: Whenever you are asked to revise the article by the editor of a peer reviewed journal keep in mind the following points
- Make an effort to incorporate the referee's suggestions
- If a "stupid" referee misunderstood your paper, it is your fault
- Do not attack referee and do not get angry
- Write a detailed response to individual referee
- Resubmit the revision within the required timeframe

Rejection: If you get rejection letter from the editor please follow the following principles:
- Expect a rejection letter or a recommendation to revise extensively
- Do not accept defeat too quickly
- Study reviewers comments thoroughly
- Consider revising and resubmitting the paper
- Listen to what the editor says
- Develop a thick skin
- Be a good loser
- This gives you opportunity to submit this improved paper to a new journal

Hope these suggestions will stimulate many potential writers to produce good useful articles

References:

1. *PeatJ, Elliot E, Baur L, Keena V, Scientific writing Easy when you know how, Byward Viva Publishers Pvt. Ltd., First Indian Edition, 2004.*
2. *Deborah C, Saltman A.M, Writing for Publications: http://www.nswphc.unsw.edu.au/pdf/ShortCourseResMetJun05/Wednesday%20Session%203%20%20Deb%20Saltman%20Writing20and%20Publishing%20DS.*
3. *B.K.Nayak. How to write a good scientific paper? Journal of Maharashtra Ophthalmological Society. Volume 1, issue 2 – Jan- April 2005*

Original Article

"A study of safety and efficacy of Manual Small Incision Cataract Surgery (MSICS) in eyes with white cataract."

Dr. Somen Misra, Dr. Neeta Misra, Dr. Priya Patil

Abstract:

Aim: To assess the safety & efficacy of MSICS in cases of white cataract with the use of trypan blue as an adjunct for performing continuous curvilinear capsulorhexis (CCC).

Methods: A hospital-based, prospective, randomized study was carried out on 40 eyes of 40 Patients with uncomplicated white cataract (intumescent, mature, hypermature) patients at a tertiary level health care centre. Baseline evaluations were done for all patients and were found to be similar in all aspects. 40 eyes with uncomplicated cataracts were randomly allocated into two groups; 20 MSICS with trypan blue(group A) and 20 without trypan blue (group B). The performance of the patients in terms of visual rehabilitation and the incidence of complications in the two groups were studied.

Results: In present study, success rate of CCC was 85% with adjunctive use of trypan blue and remaining 15% were converted to can opener or envelope capsulotomy. All 20 patients (100%) where we had used trypan blue, could put PC IOL in capsular bag. In patients, where we had not used trypan blue, 85% of cases we could put PC IOL in bag, 10% of cases we had put PC IOL in sulcus and only in one case (5%) we had put IOL in anterior chamber. Post operative uncorrected visual acuity of 6/18 or better at end of 6 weeks were seen in 75% (group A 90% and group B 60%). Post operative best corrected visual acuity of 6/18 or better at 6 weeks were seen in 92.5% (group A 100% and group B 85%)

Conclusions: Our study demonstrates that MSICS is safe and effective treatment for patients with white cataract, especially with the adjunctive use of trypan blue dye, even in rural population, where Phacoemulsification may be the unaffordable to majority of population.

Key words: Manual Small Incision Cataract Surgery, trypan blue, continuous curvilinear capsulorhexis

Introduction

Though Phacoemulsification remains the most exciting innovation of cataract surgery in 20th century, a parallel technique of Manual Small Incision Cataract Surgery (MSICS) has evolved as the most popular choice in developing countries. In India, there are about 12.5 million blind & 80% of them are blind due to cataract[1, 2]. Most patients have advanced stages of cataract with intumescent, mature or hypermature cataract. Majority of these patients are socio-economically backward and cannot afford procedures like Phacoemulsification. MSICS is desirable in such advanced stages of cataract as it is cost effective. White cataracts constitute a significant volume of cataract surgical load in India[3, 4, 5]. White cataracts are classified pre-operatively as intumescent, mature or hypermature based on depth of anterior chamber, appearance of anterior capsule and nature of lens matter[2].

It is big challenge to perform MSICS in above three types of cataract because of following reasons: 1] Lack of red reflex[4] 2] Poor contrast between the anterior capsule & the underlying cortex 3] High intra-lenticular pressure in intumescent and hypermature cataract[6] 4] Leaking lens matter from the anterior capsule puncture site 5] In hypermature cataract, zonules are weak, posterior subluxation of lens may occur 6] Occasional presence of capsular fibrosis[2].

Creating a complete, continuous capsulorhexis (CCC) and to prolapse the nucleus from the bag into anterior chamber (especially hypermature cataract where nucleus goes down) are two difficulties encountered during MSICS in eyes with white cataract. Trypan blue staining helps the surgeon to visualize the anterior capsule while performing CCC. Secondly, it makes the prolapse of nucleus using sinskey hook, a very safe maneuver by delineating the CCC margin & the underlying cortex. Thus, safety of MSICS in white cataract is enhanced by adjunctive use of trypan blue dye for CCC[2].

In spite of the fact that MSICS can be cost effective procedure in white cataract, more research on safety, efficacy is warranted. In this study, we propose to evaluate intra operative, post operative findings as well as post operative visual outcome and complications of MSICS in white cataract, especially with adjunctive use of trypan blue.

Materials and Methods

The present study was conducted over a period of two years from August 2006 to August 2008 at a tertiary level health care centre. 40 eyes of patients with uncomplicated senile intumescent, mature, hypermature cataract, attending the Ophthalmology OPD were selected. Patients from either sex were included. Patients with cataract but without any other cause of ocular morbidity which would adversely affect the outcome of the surgery were selected. The criteria for exclusion were: Patients with any cause of ocular morbidity like any active corneal disease, corneal opacities or degenerations, uveitis, glaucoma, posterior segment pathology i.e. retinal detachment, macular disease, hypertensive or diabetic retinopathy or any other retinopathy or neuropathy.

Patients with any major intra or post-operative complications, related to anesthesia, surgeon, or patient related factors which would affect the outcome of surgery were excluded. Patients with any previous ocular surgery like pterygium excision, glaucoma surgery, any other ocular surgery, Patients who did not complete at least 6 weeks of follow-up, Patients with any systemic disease such as uncontrolled diabetes which may complicate

Dr. Somen Misra
Prof., Dept. of Ophth., Pravara institute of medical sciences, Loni 413736, Maharashtra.

intraoperative or postoperative visual recovery were excluded.

Routine investigations like blood pressure, blood sugar (fasting) & urine analysis (for albumin & sugar) was carried out in all patients. All patients were thoroughly worked up for cataract surgery with complete ocular examination including Visual Acuity, meticulous Examination of Anterior Segment with the help of slit lamp, Grading of white cataract into intumescent, mature or hypermature, direct ophthalmoscopic examination, applanation tonometry, syringing of lacrimal sac, Keratometry, IOL power calculation.

40 eyes with uncomplicated cataracts were randomly allocated into two groups: Group A: Included 20 eyes in which MSICS was done with adjunctive use of trypan blue. Group B: Included 20 eyes in which MSICS was done without adjunctive use of trypan blue. A written informed valid consent was obtained in each case preoperatively. Topical antibiotic eye drops (Gatifloxacin 0.3%) were instilled 2 hourly one day prior to surgery. Preoperatively adequate mydriasis was achieved. A peribulbar injection was given. Randomization was done half an hour before the surgery to allot the patient to one of the two groups. (MSICS with adjunctive use of trypan blue and MSICS without trypan blue) .0.1% trypan blue was injected in half the number of cases in anterior chamber to stain the anterior capsule and wash was given after 10secs. Postoperatively all patients were treated with Dexamethasone (0.1%) + (Tobramycin 0.3%) eye drop 6 times a day x 1 week and then in doses tapering weekly to stop at 6 weeks. All patients were followed post-operatively on Days 1, 8 and 42. During each post-operative visit, unaided and best-corrected visual acuity was recorded with the help of illuminated Snellen chart. Post operative slit lamp examination was done on days 1, 8 and 42. Any post operative complications were noted, with the specific reference to wound related problems and any form of inflammation & cellular reaction were noted.

A comparison of all above criteria was done between the two groups of patients. To analyze the various parameters, we applied Chi-square test and 'Z' test, to the data to be analyzed wherever possible. A statistical analysis was done to determine the true results of the two groups using p-values.

Observations

Majority of the patients in both groups belonged to the age group of 60 – 70 years, 50%, and 35% in groups A and B respectively. There was no predominance of either sex. 50% of patients were females and 50% were males. Surgery (MSICS) was done on: 20 (50%) MSC (mature senile cataract), 15 (37.5%) HMSC (hypermature senile cataract), 5 (12.5%) intumescent type of cataract. All patients had visual acuity less than 2meter finger counting and majority of them, 85% in group A and 75% in group B, had vision perception of light (PL) and projection of rays (PR).

Good complete rhexis were achieved in total 21(52.5%) white cataract patients, out of which 14 patients belongs to group A where trypan blue was used, whereas only 7 patients belongs to group B where trypan blue was not used. In those 14 patients in group A, 11 patients were having mature cataract. Small rhexis was seen only in 1 case of (2.5%) of group B. Large rhexis were noted in 3 cases (7.5%) of group B. Extension of rhexis margin to mid periphery was seen in 9 cases (22.5%) out of which only 3 cases were seen in group A, all 3 belonging to HMSC type of white cataract, remaining 6 cases were seen in group B. Capsulorhexis tear was converted to can-opener type of capsulotomy in 2 cases (5%) (1 intumescent cataract in group A and 1 HMSC in group B). Both these patients developed intraoperative miosis. CCC was converted to envelope type of capsulotomy in 4 patients (10%). All these patients were hypermature type of cataract (2 in group A and 2 in group B).

All 20 patients (100%) in group A, where we had used trypan blue, could put PC IOL in capsular bag. In group B, where we had not used trypan blue, 85% of cases we could put PC IOL in bag, 10% of cases we had put PC IOL in sulcus and only in one case (5%) we had put IOL in anterior chamber. Zonular dialysis, Posterior capsular rupture and vitreous loss were seen in 2cases (5%), out of 40 patients in HMSC where trypan blue was not used. Out of these 2 cases, we had put one IOL in sulcus and in other case we had implanted IOL in anterior chamber. Intra operative miosis was noted in total 3 patients (7.5%), 1 in group A and 2 in group B. Total of 17 complications was noted in 10 eyes (3 patients of group A, 7 patients of group B). In 6 cases, 5 (12.5%) patients had corneal oedema with descemet's fold >10 and 1 case (2.5%) in group B had corneal oedema with descemet's fold <10. Mild iritis was seen in total 3 (7.5%) cases, belonging to group B. Moderate iritis with fibrin membrane noted in 1 case (2.5%) in group B. Endothelial touch were noted in center of cornea in 2 (5%) cases of, 1(2.5%) belonging to group A and 1(2.5%) belonging to group B. Secondary glaucoma were noted in 2 (5%) of cases, 1 (2.5%) in group A and 1 (2.5%) case in group B. Trauma to iris is noted in one (2.5%) case of in group B where there was intra operative miosis before prolapsing the nucleus in AC. Macular oedema was noted in 1 case (2.5%) post operatively in group B.

Categorizing postoperative visual acuity as per WHO Guidelines-good outcome (6/6-6/18), borderline outcome (<6/18-6/60) and poor outcome (<6/60). In our study, UCVA (uncorrected visual acuity) of 80% in group A and 60% in group B had good visual outcome on 1st postoperative day, where as BCVA (best corrected visual acuity) of 85% in group A and 65% in group B had good outcome on 1st post operative day.Early and very good visual rehabilitation was achieved in majority of the patients. Considering post operative UCVA of 6/18 or better at end of 6 weeks were seen in 75% (group A 90% and group B 60%). Post operative BCVA of 6/18 or better

at 6 weeks were seen in 92.5% (group A 100% and group B 85%). Significant complications leading to poor visual performances were minimal and seen only in 1 case belonging to group B (where we had put AC-IOL).

Discussion

Cataract is the main cause of curable blindness worldwide, with the developing world accounting for three quarters of blindness[8]. White cataract represents an advanced form of cataract[9]. Millions of people, worldwide are blind from white cataracts. The developing countries cannot afford expensive modern technology to treat these cases[10]. MSICS is not as glamorous as Phacoemulsification. However, it yields the same results and avoids the risk of a dropped nucleus. In addition, it can be done without expensive equipment[11]. Phacoemulsification in dense cataract is associated with intra operative complications. The denser is the nucleus, the greater the possibility of corneal endothelial damage, zonular dialysis and posterior capsular rupture[12].

MSICS promises to be a viable cost effective alternative in this regard. The safety of this procedure in white cataract is enhanced by the adjunctive use of trypan blue dye as in our study the success rate was 85% with adjunctive use of trypan blue and remaining 15% were converted to can opener or envelope capsulotomy. Although we found very good rate of success in completing CCC but still it was very difficult to complete CCC in hypermature and intumescent type of cataract, even with adjunctive use of trypan blue due to high intra-lenticular pressure and liquefied cortex. The challenge of performing CCC in white cataract is well documented. Venkatesh et al[2] made an observation. In their study success rate of CCC was 96% with adjunctive use of trypan blue and all those, cataract converted to can-opener type were intumescent in nature. Jacob et al[13] reported that 3.85% of cases where a CCC had failed, it had to be converted to conventional ECCE. This was not the case with our study, as we could comfortably continue a sutureless procedure, MSICS, with can-opener and envelope capsulotomy.

All 20 patients (100%) in group A where we had used trypan blue, we could put PC IOL in capsular bag. In group B where we had not used trypan blue, 85% of cases we could put PC IOL in bag, 10% of cases we had put PC IOL in sulcus and only in one case (5%) we had put IOL in anterior chamber. Partial Zonular Dialysis was noted in 1 hypermature cataract in group B, while prolapsing the nucleus into anterior chamber and there was posterior capsular rupture superiorly. Vitreous loss was seen in same case. Meticulous anterior vitrectomy was performed in this case and PC-IOL was placed in sulcus. Complete Zonular Dialysis was also noted in 1 hypermature cataract where trypan blue was not used (group B) and rhexis was extended beyond mid periphery after 1st nick on anterior capsule. There was vitreous loss and PC-IOL implantation was not possible. In this case, due to lack of capsular support, AC IOL was implanted, after meticulous anterior vitrectomy. Intra operative miosis was noted in total 3 patients. 2 patients where we had to convert CCC to can- opener type of capsulotomy. 1 case, there was Partial Zonular Dialysis.

In our study, UCVA of 80% in group A and 60% in group B had good visual outcome on 1st postoperative day, where as BCVA of 85% in group A and 65% in group B had good outcome on 1st post operative day. Our results correlate well with those shown by Venkatesh et al[2], in their study, 64% had good uncorrected visual outcome and 94% had good best corrected visual outcome on 1st post operative day.

In present study, on 42nd postoperative day, 39 patients (97.5%) had BCVA of 6/18 or better. No patient had BCVA worse than 6/60, at end of 6 weeks including that 1 patient in whom we had implanted AC IOL.

Our results correlate well with those shown by Dennis et al[14], who did a study to evaluate MSICS in China. They showed that UCVA of 6/18 after MSICS in their study population was 83.4%, and BCVA of 6/18 was 97% at the end of follow up at 6 months. Venkatesh et al[2, 15] got similar results in one study (MSICS in white cataract) with 94% of eyes having BCVA of 6/9 or better on the 40th post operative day. They also conducted one study (MSICS in brunescent cataract) in which BCVA of 6/18 or better were achieved in 97.1% of eyes. Minassian et al[16] reported UCVA of 69% in MSICS at end of 6weeks. Riley et al[17] reported BCVA of 88% in MSICS at end of 6 weeks. Henning et al[18] reported higher rates of final UCVA >6/18 with MSICS. Bayramlar et al[19] in their study found VA of 20/40 or better in 75% at 2months post operatively in MSICS. Kimura et al[20] got final post operative VA 20/30 or better in 80.4% in MSICS.

Total of 17 complications was noted in 10 eyes (3 patients of group A, 7 patients of group B) which is much higher than the 7%, shown in the study by Gogate et al[21]and 8.5%, shown in the study by Dennis et al[14]. In 6 cases, 5 (12.5%) patients had corneal oedema with descemet's fold >10 and 1 case (2.5%) in group B had corneal oedema with descemet's fold <10. These results were comparable to 2 different studies done by Venkatesh et al[2]. One study, found corneal oedema in 13% of cases, where as, in another study done to know safety and efficacy of MSICS for brunescent and black cataract, corneal oedema with descemet's fold >10 were seen in 14.7% of cases and corneal oedema with descemet's fold <10 in 4.9% of cases[15].

So, immediate postoperative complications such as corneal oedema found in 6 cases (15%). It was higher than published results of Phacoemulsification in white cataract[3] raising doubts whether Phacoemulsification is more endothelial friendly. However, all of them were resolved with medical therapy within one week. Mild iritis was seen in total 3 (7.5%) cases, belonging to group B,

out of these 2 cases, in 1 case CCC was converted to envelop type of capsulotomy and 1 case there was complete zonular dialysis. In 1 cataract we noted mild iritis where trypan blue was not used, rhexis was too big and PC-IOL was implanted in sulcus.

Venkatesh et al[2] in their study found, mild iritis in 6%, moderate iritis with fibrin membrane was seen in 3%. Similar results were also found in another study done by them to know safety and efficacy of MSICS for brunescent and black cataract, mild iritis in 5.9%, moderate iritis with fibrin membrane in 2.9%.

Only 3(15%) cases in group A, developed post operative complications, where as 7 (35%) cases in group B developed complications. Complications were less in group A, where we had used trypan blue. The dye stained capsular rim was distinctly visible throughout the surgery and any compromise to capsular bag can be easily detected. So, less surgical complication naturally resulting in good final visual outcome. Complications were more in group B, where we did not use trypan blue before performing CCC. 65% of cases we could not achieve good CCC due to lack of contrast, so only 85% of patients had final BCVA > 6 / 18.

In summary, MSICS is a safe and affordable procedure to perform on intumescent, mature or hypermature cataract, which gives excellent results in terms of visual rehabilitation. Creating a CCC and prolapsing the nucleus from the bag into anterior chamber (especially hypermature cataract, where cortex is liquefied and nucleus sinks down) are two difficulties encountered during MSICS in eyes with white cataract. Trypan blue for anterior capsule staining in mature white cataracts helps the surgeon to visualize the anterior capsule while performing CCC. Secondly, it makes the prolapse of nucleus using sinskey hook, a very safe manoeuvre by delineating the CCC margin and the underlying cortex. Thus safety of MSICS in white cataract is enhanced by adjunctive use of trypan blue dye before performing the CCC. The dye stained capsular rim can be easily detected. So it is also helpful in the bag placement of IOL, leading less post operative complications. From present study, we can also conclude that results of MSICS with adjunctive use of trypan blue are much superior than results of MSICS without adjunctive use of trypan blue.

References

1. *Dandona L, Dandona R, Naduvilath T, McCary CA, Nanda A, Srinivas M, et al. Is current eye policy focus almost exclusively on cataract adequate to deal with blindness in India? The Lancet 1998; 74:341-43.*
2. *Venkatesh R, Das M, Prashanth S, Muralikrishnan R, Manual small incision cataract surgery in eyes with white cataract. Ind J Ophthamol 2005; 53:173-176.*
3. *Chakrabarti A, Singh S. Phacoemulsification in eyes with white cataract. J Cataract Refract Surg 2000; 26:1041-7.*
4. *Kothari K, Jain SS, Shah NJ. Anterior capsular staining with trypan blue for capsulorhexis in mature and hypermature cataracts. A preliminary study. Ind J Ophthamol 2001; 49:177-180.*
5. *Misra S, Misra N, Reddy V. A comparative study of post operative astigmatism between the different external incisions used in manul small incisional cataract surgery. The ocular communiqué 2008; 88-91.*
6. *William JF, Samuelson TW, Masket S. The White Cataract how to manage a cortically mature nucleus. Cataract & refractive surgery today 2005; 19-20.*
7. *Pandey SK, Werner L, Wilson M, Izak AM, Apple DJ. Anterior capsule staining techniques, recommendations and guidelines for surgery. Ind J. Ophthalmol 2002; 50:157-159.*
8. *Natchiar G.Manual Small Incision Cataract Surgery. Arvind Eye Hospital Madurai, 2nd Ed; 2004; chap 3, 5:25-30, 48-57.*
9. *Lee YC Great white cataracts. EyeWorld Asia-Pacific 2008; 03:25-26.*
10. *Nowak R. SICS-a cost effective alternative to Phacoemulsification for developing countries in Nepal. Curr opin opthalmol.2008; 19:55-9.*
11. *James PG, Andrea C. Small incision manual extracapsular cataract surgery in Ghana, West Africa. J Cataract Refract Surg 2003; 29:57-64.*
12. *Junsuke A, Shuzo K, Shiro H. Manual sutureless cataract surgery using a claw vectis. J Cataract Refract Surg 2000; 26491-496.*
13. *Jacob S, Agarwal A, Agarwal S, Chowdhary S, Chowdhary R. Trypan blue as adjunct for safe Phacoemulsification in eyes with white cataract. J Cataract Refract Surg 2002; 28: 1819-25.*
14. *Dennis Lam SC, Congdon NG, Rao SK. Visual outcomes and astigmatism after sutureless, manual cataract extraction in rural China. Arch Ophthalmol. 2007:125(11):1539-1544.*
15. *Venkatesh R, Tan CSH, Singh GP, Veena K, Krishnan KT, Ravindran RD. Safety and efficacy of manual small incision cataract for brunescent and black cataracts. Eye 2008; 00:1-3.*
16. *Minassian DC, Rosen P, Dart JKC, Reidy A, Desai P, Sidhu M. Extracapsular cataract extraction compared with small incision surgery by Phacoemulsification: a randomized trial. Br. J Ophthamol 2001; 85:822-829.*
17. *Ridley et al. The Auckland Cataract Study: co-morbidity, surgical techniques, and clinical outcomes in a public hospital service. Br. J. Ophthalmology. 2002; 86: 185-190.*
18. *Hennig A, Kumar J, Yorston D. Sutureless cataract surgery with nucleus extraction: outcome of prospective study in Nepal. Br. J. Ophthalmol 2003; 87: 266-70.*
19. *Bayrmmlar H, Cekic O, Totan Y. Manual tunnel incision extracapsular Cataract extraction using the sandwich technique. J Cataract Refract Surg 1999; 25: 312-15.*
20. *Hideya K, Shin-ichiro K, Naonori M, Hiroo T, Miyo M, Makoto N. Extracapsular cataract extraction with a sutureless incision for dense cataracts. J Cataract Refract Surg 1999; 25:1275-1279.*
21. *Gogate PM, Deshpande M, Wormald RP, Kulkarni SR. Extracapsular cataract surgery compared with manual small incision cataract surgery in community eye care setting in western India: a randomised trial. Br J Ophthalmol 2003; 87: 667-672.*

Target Intraocular Pressure in Management of Primary open Angle Glaucoma

Dr. Barun Kumar Nayak

The various steps in the medical management of glaucoma involves the following considerations:

- Establish a diagnosis
- Establish a good baseline intraocular pressure (IOP) and other parameters
- Set a target IOP
- Initial therapy and attempt to lower IOP to target
- Follow up with baseline parameters
- Modify target IOP
- Consider quality of life of patients

In this communication I would be discussing the step of "Set a target IOP".

Normal IOP is defined for populations as the IOP that is safe for each individual in health and disease. As we notice here that this definition defies the statistical definitions, hence it is very difficult to calculate target pressure in all cases accurately. However, we also understand that IOP is the only parameter that can be influenced directly by glaucoma treatment and treating glaucoma makes sense only if it preserves the well being, the health status and quality of life of patients.

Target IOP is the mean IOP obtained with treatment that prevents further glaucomatous damage in the eye of the individual under considerations. Target IOP is often treated as a single figure, but it should be considered as a range of IOPs as depicted in figure 1.

Before we proceed we must understand some of the characteristics of IOP

- As the IOP increases the risk of glaucomatous damage increases and if crosses the threshold level the chances of developing glaucoma becomes high (Fig 2)
- Figure 3 depicts the rationale for IOP reduction which decreases the rate of ganglion cell loss.
- The fundamental goal of glaucoma treatment is to slow the rate of retinal ganglion cell loss to the age dependent rate (Fig 4)
- Individual vary in susceptibility to pressure dependent damage (Fig 5)
- There is a change in the susceptibility of patients during the course of glaucoma (Fig 6)

Main problems of target IOP assessment:

1. As we have noticed so much of variation in the behavior of IOP, the target IOP must be individualized to each patient and to each eye.
2. Target IOP should be accurate estimate to prevent further damage.
3. Target IOP needs to be determined in advance
4. The glaucomatous damage must increase to verify that the target IOP estimated earlier was inadequate
5. Many glaucoma patient do not have elevated IOP (normal tension glaucoma)

In deciding target IOP, earlier ophthalmologist used to consider the IOP less than 20 mm of Hg adequate but now there are enough evidence in literature that this may not be sufficient for all glaucoma patients.

There are certain guidelines for target IOP assessment (The European glaucoma society: treatment, principles and options) which must be understood clearly before setting a target IOP for any glaucoma patient:

1. IOP level before treatment: Higher the initial IOP at which the damage has occurred or at the time of first diagnosis, higher target IOP can be set at and if the initial IOP was lower the target IOP also should be set at lower level. While setting the target IOP the maximum and minimum IOP as well as diurnal variation of IOP should be kept in mind. Higher diurnal variation or maximum IOP at higher level than target IOP should be considered as inadequate and bad control of IOP.

2. Stage of glaucomatous damage: If at the time of diagnosis the glaucomatous damage is greater then it calls for a lower target IOP. One should keep in mind that with severe pre-existing damage, the safety margin is very narrow and any further damage may make the patient functionally handicapped,

3. Rate of progression of glaucomatous damage: If the glaucomatous damage is not progressing the treatment becomes questionable. Faster rate of progression of glaucomatous damage calls for a lower target IOP. If patient has come with advanced damage then history may provide some clue regarding the progression, which can be helpful in deciding the target IOP. If a patient is on certain medication and glaucomatous damage is stationary then even raising the target IOP can be considered by withdrawing some of the medications. Repeated assessment of field of vision and other parameters can be helpful in this regard. Sometimes it may need 5-6 visual field examination to establish a progression of glaucomatous damage.

4. Age of the patient: A long life expectancy requires a lower target IOP and vice versa. With this fact in mind

Dr. Barun Kumar Nayak
P. D. Hinduja Hospital and Medical Research Center, Mahim, Mumbai.

if mild glaucomatous damage is detected at the age of 70 yrs, an aggressive treatment is not called for but the similar situation in a 30yrs old patient calls for management with lower target IOP. One should also keep in mind the fact that the nerve fibre can withstand longer the damaging effect of IOP in the younger age group.

5.Likelihood of future glaucomatous damage: This factor is again dependent on IOP and the risk increases with higher IOP. Severe pre-existing damage in the same or fellow eye is a possible risk factor. Other risk factor may be a possible family history of visual handicap caused by primary open angle glaucoma. Fig -7 represent the effect of some of the factors in deciding the target IOP.

Target IOP is a dynamic process and it should be assessed frequently and adjusted at higher and lower level if the follow up tests suggest so.
After describing the basic principles of establishing the target IOP, I will describe two different, easy and logical methods for the same to actually calculate the target IOP.

I. There are many formulae available in literature, but the one which I follow is given below:
TP = (1 - IP / 100) -FD
TP - Target pressure
I P - Initial pressure
FD - Field defect factor, which should be 0 for no, 1 for mild, 2 for moderate and 3 for severe glaucomatous field defect.

This formula looks complicated but still it is simple. Basically it implies that reduce the IOP by the same numerical percentage as that of the initial IOP and substract the field defect factor from the value to get target IOP.

Explanation: If initial IOP was 50 mm of Hg reduce IOP by 50% and substract the field defect factor to get target IOP. If initial IOP was 40mm of Hg, reduce IOP by 40% and substract the field defect factor to get target IOP. If initial IOP was 30mm of Hg, reduce IOP by 30% and substract the field defect factor to get target IOP.

Examples :
a. Initial IOP was 50 mm of Hg.
50 % reduction will make it 25mm of Hg
After considering the field defect factor

No damage	25-0	= 25mm of Hg
Mild damage	25–1	= 24mm of Hg
Moderate damage	25-2	= 23mm of Hg
Severe damage	25-2	= 22 mm of Hg

These are mean target IOPs to be aimed at initially with a range on either side and to be adjusted periodically in follow ups.

b.Initial IOP was 40 mm of Hg

40% reduction	=	24 mm of Hg
After field defect factor		
No damage	=	24mm of Hg
Mild damage	=	23 mm of Hg
Moderate damage	=	22 mm of Hg
Severe damage	=	21 mm of Hg

c. Initial IOP was 30 mm of Hg
30% reduction = 21mm of Hg
After field defect factor consideration it can be 21 to 18 mm of Hg

d. Initial IOP was 20 mm of Hg
20 % reduction = 16mm of Hg
After field defect factor consideration it can be 16 -13 mm of Hg

We notice that this method of calculation does take care of higher IOP – higher target IOP and lower target IOP in greater damage as per our guidelines described earlier. But other guidelines are not taken into account, which the clinician should adjust as per their judgment and experience.
I. Other method to follow is based on American society recommendations. The basic principles are:
i. For unquestionable damage as a result of glaucoma in early stage reduce IOP by 20%, in moderate damage reduce by 30% and in advanced damage reduce IOP by 40%.
ii.Additional 3% for each risk factors and for each decade of life expectancy upto the maximum of 4 factors.

Examples:
a. Initial IOP of 30mm of Hg with mild damage.
I. Reduce IOP by 20% = 30-6 =24mm of Hg
ii.With 2 risk factors
Reduce IOP by (20 +3x2) = 26% = 30-8 = 22mm of Hg approx
iii.With 2 risk factors and 2 decades of life expectancy
Reduce IOP by (20+3x4) = 32% = 30-10 = 20mm of Hg approx

b.Initial IOP of 30 mm of Hg with moderate damage
I. Reduce IOP by 30% = 30-9 = 21mm of Hg approx
ii.With 2 risk factors
Reduce IOP by (30+ 3x2) = 36% = 30-11 =19 mm of Hg approx

iii. With 2 risk factors and 2 decades of life expectancy
Reduce IOP by (30+ 3x4) = 42% =30-13 = 17 mm of Hg approx

c. Initial IOP of 30 mm of Hg with advanced damage
I. Reduce IOP by 40% = 30-12 = 18 mm of Hg
ii. With 2 risk factors
Reduce by (40+ 3x2) =46% = 30-14 =16 mm of Hg approx

iii. With 2 risk factors and 2 decades of life expectancy
Reduce IOP by (40 +3X4) =52% =30-16 =14 mm of Hg approx

I am sure these guidelines will definitely be helpful in deciding the target IOP.

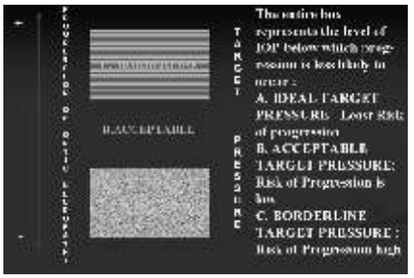


figure 1.

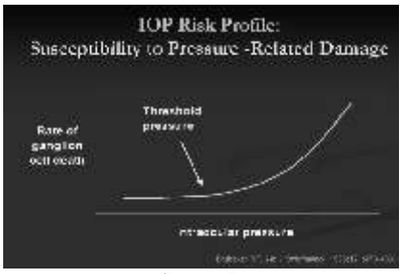


figure 2.

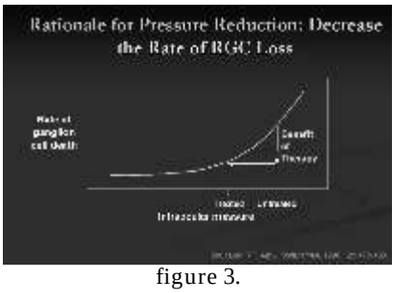


figure 3.

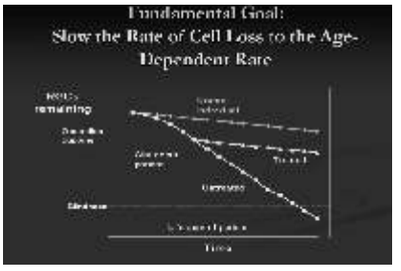


figure 4.

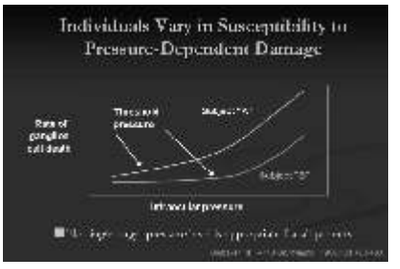


figure 5.

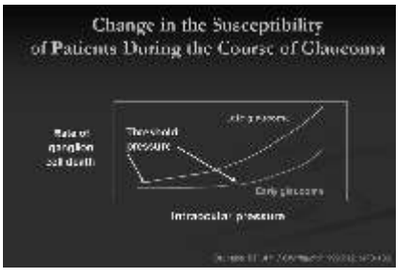


figure 6.

Figure 7.
(from the European glaucoma society treatment, principles and options)

Current Management Options for Uncomplicated Congenital Nasolacrimal Duct Obstruction

Dr. Moushmi Chaudhari, Dr. Ashwin Sainani

Introduction

Congenital Nasolacrimal Duct Obstruction (CNLDO) is a common condition seen in the first year of life with above 20% of all newborns suffering from epiphora.

The nasolacrimal duct (NLD) is about 12mm long and is the continuation of the lacrimal sac. It descends and angles slightly laterally and posteriorly to open into the inferior nasal meatus, lateral to and below the inferior turbinate. The opening of the duct is partially covered by a mucosal fold (valve of Hasner), the most common site for CNLDO.

The lacrimal drainage system begins at about 6 weeks of gestation. It develops embyologically from the naso-optic fissure which becomes buried as a solid epithelial cord because of the growing together of the lateral nasal process from above and maxillary process from below. The epithelial cord thickens and divides to form the NLD, and the canaliculi develop as outbuddings from the upper end. Canalization of the passageways starts from the upper end and progresses downwards. A thin membrane separating the NLD from the lacrimal mucosa usually persists up to the time of birth.

Causes

It is commonly caused by the presence of membrane at the valve of Hasner. Other rare conditions are congenital absence of valves, congenital atresia of NLD, congenital lacrimal sac mucocele, absence or atresia of canaliculi or puncta, craniofacial anomalies or adjacent tumour mass such as hemangiomas, lymphangioma. The incidence of this disorder is higher in children with craniofacial disorders & Downs syndrome.

Presentation

The parents would usually give a history of the child having continuous or intermittent epiphora which may be unilateral or bilateral. Associated features include stickiness of the eyes, recurrent conjunctivitis, crusting of eyelids with or without boggy swelling over inner canthal region. The severity of the symptoms should always be taken into account by taking a detailed history from the parents.

Evaluation

External examination

The eyelids should be examined for swelling, crusting, ectropion and entropion.

Punctal stenosis should be looked for.

Regurgitation Test is done by exerting pressure over the lacrimal sac area which may result in punctual reflux of mucopurulent material.

Fluorescein Dye Disappearance test should be performed by instillation of 2% fluorescein into both conjunctival fornices and observing for the retention of dye after 5 minutes. Prolonged retention of dye is indicative of inadequate lacrimal drainage.

Although the above three evaluation techniques are adequate to make the diagnosis in most cases one can perform the following invasive tests in special cases

Contrast dacryocystography is done to confirm the site of obstruction especially prior to lacrimal surgery by injecting radio-opaque dye into the canaliculi and taking magnified X-ray images.

Nuclear lacrimal scintigraphy using radionuclide technetium-99 is more sensitive in assessing incomplete blocks especially in the upper part of the lacrimal system.

Differential diagnosis

The differential diagnoses include congenital dacryocystocele, congenital conjunctivitis, corneal abrasions and congenital glaucoma.

Treatment

Treatment of nasolacrimal duct obstructions including the method and timing varies among the ophthalmologists. Despite the controversy most cases result in success.

Conservative Methods:

Massage over the lacrimal sac area is first described by Crigler: It helps to increase the hydrostatic pressure in the sac and opens up the membranous occlusions. It also empties the sac of stagnant tears which can be a source of infection. This can be followed by instillation of antibiotic drops only if there are signs of infection like mucopurulent discharge or redness and swelling of eyes. Massage is almost universally prescribed as the first line of therapy for a CNLDO and there are number of studies that have shown that the incidence of spontaneous recovery of the condition by conservative methods is very high (93%) in the first year of life . A short course of anti-biotic drops like ciprofloxacin or tobramycin can be prescribed only when there is increased mucopurulent discharge.

Surgical techniques:

Probing with Bowman's probes is best done under sedation or general anesthesia. After dilating the punctum the upper canaliculus is probed upto the nasal wall of the lacrimal sac until one can feel a “hard stop”

Dr. Moushmi Chaudhari
P. D. Hinduja Hospital and Medical Research Centre,Mahim, Mumbai.

after which the probe is rotated inferiorly, slightly posteriorly and laterally into the nose. Once the membranous obstruction is bypassed, one feels a sudden give-way. The location of the probe can be confirmed by direct visualization with the nasal endoscope or direct metal-to-metal contact using another probe through the nose. This procedure can be done in very young babies (below 6 months) under topical anesthesia although it usually results in a poorer success rate.

Balloon catheter dilatation:

This technique was described by Becker in 1991.The balloon catheter is introduced through the upper punctum until it is in the NLD.The balloon is then inflated to a pressure of 8 atmospheres for 90 seconds and deflated for 10 seconds. A repeat inflation up to 8 atmospheres is done for 60 seconds. The balloon is then retracted 5 mm, to lie at the junction of the sac and the NLD and two more inflations are performed. This technique is gaining importance again in the recent years.

Probing with Intubation was described by Quickert and Dryden in 1970. A normal anatomical pathway is created with a probe and then a silicon tube is passed as a stent. Tubes can be left in place for 3-6 months although most studies suggest 6 weeks as adequate. There are a number of tubes available in the market. Olive tipped probes with silicon tubes (Crawford tubes) are retrieved with a help of a specially designed (Crawford) hook which can lock onto the olive tip. A nasal endoscope is useful to retrieve the probe under direct vision in which case a simple mosquito forceps can be used to pull the probe out. Although probing with silicon intubation has a high success rate but it may require a longer general anesthesia, more post operative office visits, more expenditure and increased risk of punctal stretching, infection, corneal abrasion ,tube dislodgement, tube breakage, canalicular trauma and subsequent stenosis. This method also requires a second procedure for removal of the tube.

Turbinate Infracture:

Occasionally, the inferior turbinate lies against the lateral wall of the nose and may contribute to the obstruction to the nasal end of the NLD. The turbinate may easily be fractured if gasped by a sturdy surgical clamp and twisted. This is often done even in uncomplicated cases of CNLDO.

Dacryocystorhinostomy:

It can be performed either traditionally by making a small incision on the side of the nose and removing the anterior lacrimal crest and bones forming the lacrimal fossa. The ostium should be at least 1 cm in diameter. Silicon stents can be used in case of inflammation. The success rate is reported to be 80 to 95%. It can be done endoscopically as well to avoid an external scar. A success rate of 76 to 87% has been reported.

Timing and method of intervention:

Treatment is initiated because of simple tearing or because of recurrent or persistent mucopurulent drainage from one or both eyes. Epiphora and mucopurulent discharge may decrease or disappear after administration of topical antibiotics and massage but recurrences are common. Controversy remains regarding the optimal timing and method of treatment. Studies have shown that most CNLDO's clear spontaneously by about one year of life. Therefore conservative management such as observation, periodic topical antibiotics and massage will be helpful.

Some have suggested that the age of the patient at treatment may be inversely related to success while the recent PEDIG study concludes probing as a primary procedure of choice with an overall success rate of 78% and no age related decline in success until atleast 3 years of age. Although the initial report on balloon dilatation suggested that it produced a greater rate of success than probing these greater success rates have not been found in subsequent well-controlled studies . Also its added cost makes it non-acceptable as a primary procedure. The three recent PEDIG studies on probing , balloon dilatation and probing with intubation as a primary procedure are probably the most well controlled studies on the topic. Although the success rates are similar (probing-78%, balloon catheter dilatation-80% and probing with intubation-90%) they cannot be directly compared due to substantial selection bias and lack of randomization. All the 3 approaches have a fairly good success rate and choosing between them is a matter of picking the least invasive to begin with and working upwards from there.

A number of authors recommend a stepwise treatment paradigm for CNLDO in children where simple probing is the primary procedure of choice failing which a second probing can be considered or taking into account the cost factor a balloon dilatation can be done. If the latter fails, probing with NLD intubation is recommended.

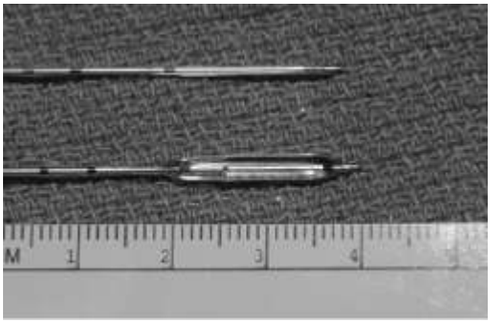
Surgical treatment should be considered only after allowing time (1 year) for spontaneous recovery during which conservative therapy in the form of antibiotic eyedrops and nasolacrimal massage can be advised. Simple probing should be considered as the primary surgical treatment of choice for uncomplicated CNLDO outweighing its benefits over the other interventions. Infracture of the turbinate can be attempted if the turbinate has been impacted against

the lateral nasal wall. If probing fails either a second probing or balloon dilatation should be attempted. Probing with intubation can also be a second line of treatment especially in difficult probings, anatomical abnormalities and canalicular stenosis.

Dacryocystorhinostomy should be reserved for all older children in whom all efforts to establish function of the physiological system have failed especially those with bony obstruction or craniofacial anomalies.

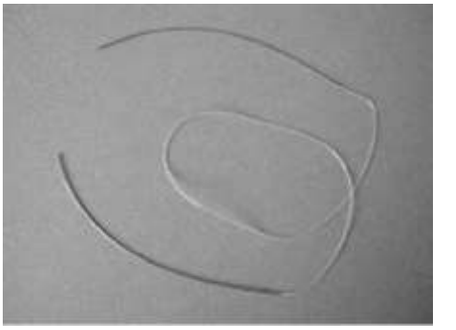
References

1. *Macewen CJ. Congenital nasolacrimal duct obstruction. Compr Ophthalmol Update. 2006 Mar-Apr;7(2):79-87*
2. *Kapadia MK et al .Evaluation and management of congenital nasolacrimal duct obstruction.Otolaryngol Clin North Am. 2006 Oct; 39(5):959-77.*
3. *Crigler LW. The treatment of congenital dacryocystitis.JAMA 1923;81:21-24.*
4. *Paul TO. Medical management of congenital nasolacrimal duct obstruction. J Pediatr Ophthalmol Strabismus 1985;22:68-70.*
5. *Becker BB et al. Balloon catheter dilatation in pediatric patients. Ophthalmic Surg.1991; 12(22):750-752.*
6. *Quickert MH et al. Probes for intubation in lacrimal drainage.Trans AM Acad Ophthalmol Otolaryngol 1970;74:431-433.*
7. *Welsh MG et al.Timing of silastic tubing removal after intubation for congenital nasolacrimal duct obstruction.Ophthal Plast Reconstr Surg.1989; 5(1):43-48.*
8. *Migliori ME et al.Silicone intubation for the treatment of congenital lacrimal duct obstruction: successful results removing the tubes after 6 weeks. Ophthalmology 1988 Jun;95(6);792-795.*
9. *Pediatric Eye Disease Investigator Group. Primary surgical treatment of nasolacrimal duct obstruction in children younger than 4 years of age.J AAPOS 2008;12:427-28*
10. *Hakin KN, Sullivan TJ, Sharma A, Welham RAN. Pediatric dacryocystorhinostomy. Aus NZ J Ophthalmol 1994;22(4):231-235.*
11. *Jones DT, Fajardo NF, Petersen RA, VanderVeen DK. Pediatric endoscopic dacryocystorhinostomy failures: who and why? Laryngoscope 2007 Feb;1117(2):323-7*
12. *Nelson LR, Calhoun JH, Menduke H. Medical management of congenital nasolacrimal duct obstruction. Ophthalmology. 1985 Sep;92(9):1187-90*
13. *Katowitz JA et al Timing of initial probing and irrigation in congenital nasolacrimal duct obstruction. Ophthalmology 1987; 94:698-705.*
14. *Tao S et al Success of balloon catheter dilatation as primary or secondary procedure for congenital nasolacrimal duct obstruction. Ophthalmology 2002; 109:2108-11.*
15. *Pediatric Eye Disease Investigator Group. Primary treatment of nasolacrimal duct obstruction with probing in children younger than 4 years. Ophthalmology 2008; 115:577-84.*
16. *Becker BB et al .Balloon catheter dilatation for treatment of congenital nasolacrimal duct obstruction. Am J Ophthalmol 1996;121:304-9.*
17. *Pediatric Eye Disease Investigator Group. Primary treatment of nasolacrimal duct obstruction with balloon catheter dilation in children less than 4 years old. J AAPOS 2008; 12:451-5.*
18. *Gunton KB et al. Comparison of balloon dacryocystoplasty to probing as the primary treatment of congenital nasolacrimal duct obstruction. J AAPOS 2001;5:139-42.*
19. *Pediatric Eye Disease Investigator Group. Primary treatment of nasolacrimal duct obstruction with nasolacrimal duct intubation in children younger than 4 years of age. J AAPOS 2008;12:445-50.*
20. *Casady DR et al.Stepwise treatment paradigm for congenital nasolacrimal duct obstruction.Ophthal Plast Reconstr Surg 2006;22:243-7.*



Balloon Catheter

Bowman Probes



Olive tipped probe with silicon tube for nasolacrimal duct intubation.

Spectrum of Phakomatoses

Dr Asif virani, Dr Nayana Potdar, Dr Chhaya Shinde, Dr Roshni Shetty , Dr Vishnu Ghonsikar

Group of disorders with hamartomas(abnormal rest of mature cells) of skin,CNS,eye and visceral organs

Includes

- Neurofibromatosis 1
- Sturge Weber Syndrome
- Neurofibromatosis 2
- Klippel Trenaunay syndrome
- Tuberous sclerosis
- Ataxia Telangectasia
- Von Hippel Lindau Disease
- Wyburn Mason Syndrome

All are autosomal dominant except Ataxia Telangectasia which is AR

Neurofibromatosis Type 1 / Von Reckling Hausen Disease / Peripheral Neurofibromatosis.

AD Inheritance. Mutation of NF1 gene on chromosome 17q12-22 results in loss of neurofibromin leaving ras oncogene unopposed for cell growth.

Cutaneous	Ocular
1. Café –au –lait spots(fig 1)– 99% with NF1flat hyperpigmented all over body (spares scalp, palms and soles) since birth ,increases in size and number with time 2. Freckles(fig 2) – 81% Hyperpigmented intertriginous areas (groin,axilla, inframammary) Presents at 6 yrs of age. 3. Neurofibromas (benign peripheral nerve tumor composed of axon,schwann cells and fibroblast) 90% are localized .seen after 6 yrs increases in size and number with age . Plexiform(fig3,4,5) – 10-20% of lesions. • Congenital • Diffuse With soft tissue hypertrophy • Rare Malignant change.	1.Lisch Nodule Brown / yellow dome shaped nodule (melanocytic Hamartoma) on iris surface .seen after 6 yrs of age ,increases with age. 2. Lid – Café- au-Lait spots, localized and plexiform neurofibromas. Plexiform of lid associated with congenital glaucoma and sphenoid bone defects.(pulsating proptosis) 3 .Orbit – Neurofibroma ,Proptosis 4. Optic nerve Glioma (Gr 1 pilocytic astrocytoma) Predominant Intracranial neoplasm seen in 15% NF1 patients. T2 weighed MRI – fusiform enlargement of optic nerve chiasmal may involve Hypothalamus with endocrine abnormality. 5. Enlarged corneal nerve 6. Hamartomas of retina ,RPE – uncommon.

CNS	Visceral
1. Tumours common are Optic pathway Gliomas ,Brainstem gliomas andSchwannoma,Meningioma 2. Aqueductal stenosis 3. Macrocephaly 4. Headache, seizure, impaired intellect	Increased risk of malignancy like Neurofibrosarcoma,Malignant myeloid disorders,RhabdomyosarcomaWilms tumour And Pheochromocytoma(Secondary Hypertension). Skeletal abnormalities Short stature , scoliosis,pseudoarthrosis

Dr Asif virani
LTMMC & LTMGH Sion, Mumbai

Diagnostic Criteria : at least two of below

- 6 or > café –au-lait spots prepubertal >5mm / postpubertal >15mm.
- 2 or > Neurofibromas (any type) / one plexiform
- Axillary / inguinal freckle
- Optic nerve glioma
- 2or > Lisch Nodules
- Osseous lesions like sphenoid dysplasia , cortical thinning.
- 1 st degree relative with above criteria.

Neurofibromatosis type 2 / Bilateral acoustic neurofibromatosis / Central NF

1 in 50,000.

AD inheritance NF 2 (tumour suppressor gene) on chr 22 q 12 encoding for protein schwannominor merlin.

- Cutaneous lesion uncommon.
- Ocular – Posterior subcapsular cataract, epiretinal membrane,combined hamartomas of RPE and retina.
- CNS : B / L Vestibular schwannomas – hallmark Progressive hearing loss, tinnitus, vertigo.

Other includes : Schwannomas of other cranial , spinal , peripheral nerves.

Meningiomas , Optic nerve sheath ,intracranial , intraspinal.

Gliomas , Ependymomas.

Diagnostic Criteria One of below

- B / L VIII nerve schwannomas
- Ist degree relative with NF2 and U / L VIII nerve Schwannoma.
- Ist degree relative with NF2 and two of lesions (Neurofibromas, meningioma .glioma. schwannoma, posterior subcapsular caract)

Sturge weber syndrome / encephalotrigeminal angiomatosis

Fascial angioma / port wine stain / naevus flammeus

- Reddish purple usually unilateral along V1,V2and less oftenV3(fig7).
- May be bilateral ,associated with hemi hypertrophy or extensive limbs and trunk involvement.

Ocular

- Glaucoma (fig8)ipsilateral to fascial angioma mc with upper lid involvement due to angle anomaly or elevated episcleral pressure
- Choroidal haemangioma seen in 40% ipsilateral to fascial angioma
- Ipsilateral heterochromia iridis
- Conjunctival and scleral angiomas.

CNS Ipsilateral leptomeningeal haemangioma between pia and arachnoid over parieto – occipital cortex . Calcium deposition in blood vessels and superficial cortical layers produces “tram track ”appearance on CT scan.

NO visceral lesions

Diagnostic : 2 of these

- Fascial angioma with ipsilateral intracranial haemangioma
- Ipsilateral Choroidal haemangioma
- Congenital glaucoma

Management of congenital glaucoma include trabeculotomy and trabeculectomy with or without mitomycin (fig9,10,11)

Tuberous sclerosis / Bournevilles Disease

AD . 2 tumour suppressor gene TS C1 (chr 9q 34- protein – hamartin)

And TSC 2 (chr 16 p13- protein – tuberin).

Cutaneous	Ocular
Adenoma Sebaceum 75% of patients. Angiofibromas , reddish brown popular rash on malar region. Ashleaf hypomelanotic macules seen in 90% patients. Subungual/ periungual fibromas of toe > finger nails . seen in 25% patients. Shagreen patches (connective tissue hamartomas) thickened skin over lumbosacral region.	Retinal astrocytic hamartoma . 75% cases. Multiple astrocytic hamartoma , either flat smooth or modular and calcified. RPE depigmentation Angiofibroma of eyelid and conjunctiva

CNS	Visceral
Cortical Tubers – MR, Seizure Subependymal Nodules – obstructive hydrocephalus.	Lymphangiomyomatosis of lung Angiomyolipomas of kidney seen in 80% cases of TS. Renal cell cancer rare 2% Rhabdomyomas of heart Sclerosis of calvarium and spine. Pitting of tooth enamel.

Von Hippel Lindau disease / Angiomatosis of retina and cerebellum

AD . 3p26.

No cutaneous lesion

Ocular : Retinal haemangioblastomas first manifestation can be missed due to peripheral location.

Stages

1. Preclassical small lesion
↓.
2. Classical globular with feeder vessel
↓
3. lipid and plasma extravasation.
↓
4. RD may occur
↓
5. Blindness secondary to RD< glaucoma or persistent uveitis.

CNS :

Haemangioblastoma - cerebellum(most common)Spinal cord and brainstem

Majority asymptomatic

Syrinx : in spinal cord or brainstem.

Endolymphatic sac tumours

Visceral

RCC, Pheochromocytomas, benign cysts of kidney, liver, pancreas Polycythemia

Klippel trenaunay weber syndrome

Sporadic disease

Triad of

Cutaneous port wine stain present in 98% patients

Darkens with age.

Ocular fascial port wine stain ,orbital varix, choroidal angiomas and retinal varicosities.

Visceral

Varicose veins

Bone and soft tissue hypertrophy with increase in length and girth.

Diagnostic 2of the triad

Cutaneous vascular abnormality , Bone and soft tissue hypertrophy , Varicose veins

Ataxia telengiectasia / Louis-Bar syndrome

AR inheritance . AT gene (chr 11q22-23) encodes ATM protein for cell cycle control and dna repair.

Cutaneous : Telengectasias of ears, nose ,neck.

Ocular : B / L bulbar conjunctival telengectasia.

Cerebellar atrophy

Visceral hypoplastic thymus,tonsil,adenoid.

Increased risk of leukemia and lymphoma.(defective dna repair , increased cancer risk following uv exposure.

Wyburn Mason syndrome / Retinocephalic vascular malformation

Cutaneous lesions are rare

Ocular :

Retinal A-V malformations / Racemose angioma : U / L over posterior pole. Other like orbital or optic nerve AVM.

CNS : A-V malformations I / L to retinal AVM.

Visceral : AVM I / L maxilla, mandible, pterygoid fossa and spine.

Intracranial and retinal AVM are diagnostic

Fig 1

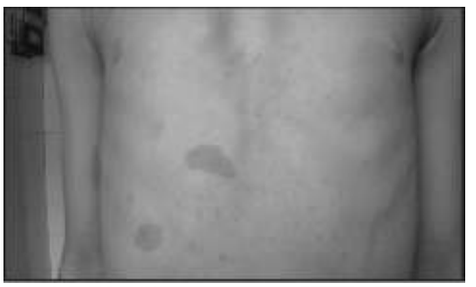


Fig 2

Fig 2

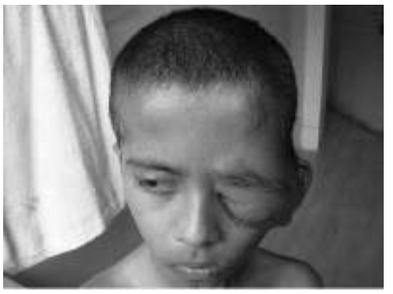
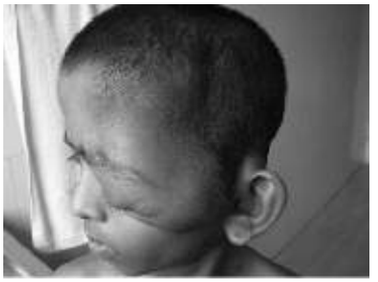


Fig 4

Fig 5



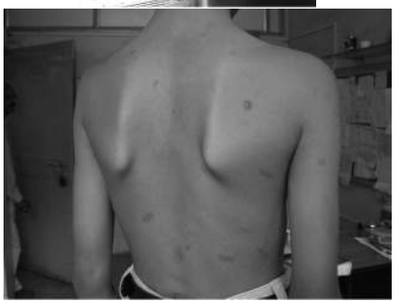


Fig 6

Fig 7

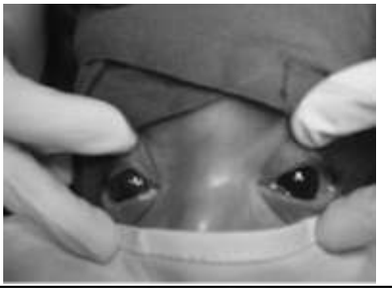
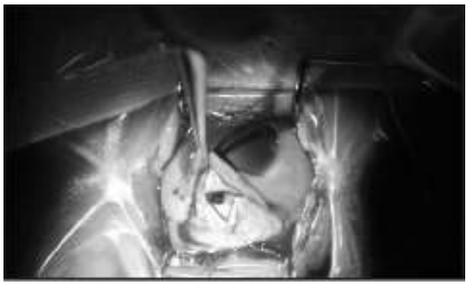


Fig 8

Fig 9

Fig 10

Fig 11



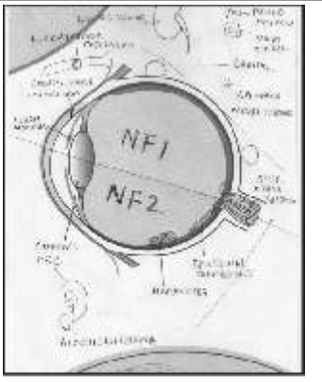


Fig 12

Case report

Tolosa Hunt Syndrome Presenting with Visual loss but Pupillary Reaction to light Remains Normal

Dr. Kedar Dhige

Keywords:
orbital apex syndrome, pupillary light reaction , medial rectus thickening

Case Report:
A fifty seven years old Muslim lady came to the OPD with complaint of swelling of left eye, sudden dimness of vision in left eye associated with severe pain in left side of head for two weeks duration.
Patient had taken treatment elsewhere with partial relief of symptoms. however severe pain recurred again for last five days.
On examination ,she was obviously very distressed and not even able to sit properly. She reported that the pain increased in intensity at night and in early morning it was most severe.
Visual acuity was two feet counting fingers in left eye not improving with pinhole. Right eye visual acuity was 6/6
She had 2mm proptosis with near total ptosis in left eye with mild lid edema. Lateral movement of eyeball was absent All other movements were normal .Anterior segment exam of left eye revealed minimal congestion with clear cornea. Pupils were 3 mm size in both eyes.
IOP was 14 mm Hg in both eyes.

Interestingly and rather inexplicably she had brisk reaction to light in both eyes .No relative afferent pupillary defect was noted. Fundus exam was normal.

Right eye was With in Normal Limits

She was advised CT scan orbits which showed thickening of the left medial rectus muscle not involving the tendinous insertions and Left side proptosis. WBC Count, ESR & thyroid function tests were within normal limits.

MRI left orbit however showed thickening of medial rectus , superior rectus and superior oblique muscles with minimal adjoining fat stranding Suggestive of orbital pseudotumour.

She was prescribed Tab prednisolone 60 mg daily .After two days,the patient improved remarkably. Ptosis and lid edema and proptosis subsided, visual acuity LE improved to 6/12.Extra ocular movements were normal in all directions. Restriction of lateral movement in left eye improved completely.

Discussion:
Considering the sudden onset of visual loss with orbital pseudotumour , optic nerve compression leading to visual loss was suspected. However Pupil reaction to light remained normal. Traditionally, lot of emphasis is placed on the pupil reaction to light while considering optic nerve lesions such as compression. In the given clinical setting of proptosis causing orbital apex syndrome ,one tends to remain complacent as pupillary urgently. Response to steroids in initial adequate high doses promptly reversed the damage inflicted by orbital apex pseudotumour. The limitations of this case study is that VEP was not done, corneal sensations also could not be noted due to poor patient co-operation and ptosis.
The second interesting feature of this case is that though medial rectus thickening was present it had led to restriction of lateral movement of the eyeball which generally occurs due to fibrotic contracture of medial rectus. However adduction of left eye was preserved. Abduction of Left eye also returned to normal following steroids. As biopsy of the extraocular muscle was not done ,the pathological entity causing medial rectus thickening could not be established.

Conclusion:
This case illustrates that Tolosa Hunt syndrome can present with visual loss and normal pupillary reaction to light.

References:

1. *Barbur JL, Moro S, Harlow JA, Lam BL, Liu M. Comparison of pupil responses to luminance and colour in severe optic neuritis. Clin Neurophysiol. 2004 Nov;115(11):2650-8. PMID: 15465455 [PubMed - indexed for MEDLINE]*

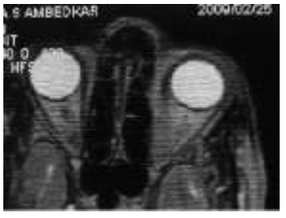
2. *Lau KC, So KF, Campbell G, Lieberman AR. Pupillary constriction in response to light in rodents, which does not depend on central neural pathways. J Neurol Sci. 1992 Nov;113(1):70-9. PMID: 1469457 [PubMed - indexed for MEDLINE]*

3. *Milton JG, Longtin A, Kirkham TH, Francis GS. Irregular pupil cycling as a characteristic abnormality in patients with demyelinative optic neuropathy. Am J Ophthalmol. 1988 Apr 15;105(4):402-7. PMID: 3358432 [PubMed - indexed for MEDLINE]*

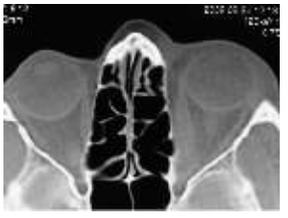
4. *Hamilton W, Drewry RD Jr. Edge-light pupil cycle time and optic nerve disease. Ann Ophthalmol. 1983 Aug;15(8):714-21. PMID: 6660715 [PubMed - indexed for MEDLINE]*

5. *Visual acuity and pupillary reactions after peribulbar anaesthesia. S J Talks, N H Chong, J M Gibson and I R Francis . British Journal of Ophthalmology 1994;78:41-43; doi:10.1136/bjo.78.1.41. Copyright © 1994 by the BMJ Publishing Group Ltd*

6. *Ramsay, A. S, Ray-Chaudhuri, N., Dayan, M., Walshaw, D. (2001). Quantification of relative afferent pupillary defects induced by posterior sub-Tenon's, peribulbar, and retrobulbar anaesthetics. Br. J. Ophthalmol. 85: 1445-1446*



MRI orbit LE medial rectus thickening



CT Scan orbit LE medial rectus thickening

Dr. Kedar Dhige
MGMS New Bombay Hospital, Vashi,

Contact Lens Induced Acanthamoeba Keratitis

Dr. Neha Gadaria, Dr.C. A. Shinde, Dr. Vishnukant Ghonsikar, Dr.Nutan Darda

A 26 year young male presented to an ophthalmologist with foreign body sensation, irritation, photophobia, and watering in right eye with history of using friend's contact lens two days back. For these symptoms he was taking treatment in the form of local antibiotic and lubricating eye drops .The condition worsened over a period of next two days with increased photophobia, redness, watering, blurring of vision, and pain. So he was referred to our institute.

On Examination:

The right eye anterior and posterior segment were within normal limits.The left eye vision was finger counting close to face.The lids were edematous with matted lashes with no vesicles.On Slit lamp examination tarsal Conjunctiva showed congestion and presence of follicles.Cornea showed Grey white ring infiltrates, edema,epithelial irregularity with small area of thinning. Rest details not visualised.The fundus details were not visualised.

A presumptive diagnosis of pseudomonas keratitis was made, and he was started on eye drops gatifloxacin and fortified tobramycin, both one hourly and eye ointment neosporin 3/day and eye ointment atropine 2/day. Initially his symptoms improved slightly, but two days later he developed worsening corneal edema and increased ring infiltrate over cornea.

In the mean time Corneal scrape and the contact lens was sent for the microbiological studies. Gram stain showed neutrophils, epithelial cells,necrotic debris with plenty of acanthamoeba cysts. Calcofluor white stain was suggestive of acanthamoeba. Bacterial and fungal cultures were negative.The culture on non nutrient E.coli agar showed growth of Acanthamoeba.

Hence patient was started on eye ointment Neosporin tds,eyedrop Fluconazole 6/day.As PHMB drops were not available we got it couriered from Arvind Eye Hospital,Madurai.It is available as 20% stock solution which has to be reconstituted to 0.02%PHMB drops with distilled water,which was given half hourly to the patient.However the patient developed corneal perforation with iris prolapse so an urgent therapeutic penetrating keratoplasty was done.

Acanthamoeba keratitis

Risk factors for developing acanthamoeba keratitis include contact lens wear, use of homemade saline, exposure to contaminated water, and corneal trauma.Over 80% of cases are related to contact lens use. All lens types have been implicated, including soft, hard, gas-permeable, disposable, extended wear.

Inoculation occurs through corneal epithelial breaks. Early infection involves the epithelium only followed by spread to all layers of cornea as well as nerves.The trophozoite produces cytolytic enzymes that aids in tissue invasion, loss of keratocytes, and stromal necrosis.

Innate immunity plays an important role in fighting acanthamoeba infection. In an animal model of Acanthamoeba keratitis, exacerbation of keratitis occurred when neutrophil migration was inhibited, and resolution of keratitis occurred when neutrophils were recruited.

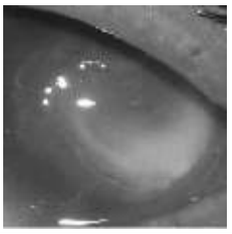
Adaptive immunity may also play a role, as anti-Acanthamoeba IgA antibodies have been demonstrated in tears.

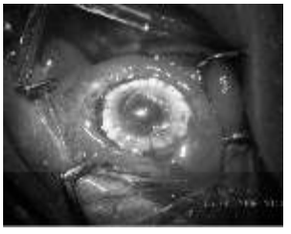
Symptoms are usually unilateral, and often include ocular pain out of proportion to signs, redness, decreased visual acuity, foreign body sensation, photophobia , and protracted progressive course with no therapeutic response to topical antimicrobial agents.

Many corneal manifestations may occur, including elevated epithelial lines that may form dendritic lesions, epithelial erosions, decreased corneal sensation, a gray-white ring infiltrate, nummular infiltrates, radial keratoneuritis, satellite lesions, disciform edema, thinning, and lack of neovascularization.Other signs may include follicular conjunctivitis, tender preauricular node, iritis, and lid abnormalities (edema, pseudoptosis).

Diagnostic modalities include culture, Giemsa Stain, corneal biopsy stained with Calcofluor White, confocal microscopy to detect organisms in vivo, and PCR (which is more sensitive than culture).

Early initiation of treatment is most effective and typically requires long-term therapy. Treatment modalities include epithelial debridement, diamidine derivatives (brolene, pentamidine), imidazole derivatives (miconazole, clotrimazole, ketoconazole), aminoglycosides (neosporin, paromycin) and cationic antiseptics (PHMB, chlorhexidine diacetate). Other therapies include penetrating keratoplasty, steroids, and cryotherapy.





Dr. Neha Gadaria
LTMMC & LTMGH Sion, Mumbai

Case Report

Alstrom Hallgren Syndrome – A Case Report

Dr. Vaishali Une, Dr.Prasad Gurav, Dr. Vilas Wangikar

Abstract :

Oculedoto diabetic syndrome is characterized by (i) Juvenile onset DM, (ii) Blindness, (iii) Deafness, (iv) Skin lesion and (v) Normal mental capacity. Alstrom Hallgren Syndrome has mutated gene ALMSI which was recently identified. Cases are commonly seen in scandinarian countries. We report a rare case of 18 year old diabetic patient fulfilling all the characteristic features of Alstrom Hallgren Syndrome.

Introduction :

Definition : It is an inherited disease characterized by progressive blindness, juvenile onset diabetes mellitus, obesity, deafness and normal mental capacity.

Alternative Names :

Alstrom syndrome, Alstrome Hallgran Syndrome, Alstromes retino-otodiabetic syndrome.

Case report :

An 18 years old patient diagnosed with juvenile DM on Inj. Insulin, resident of Jalna came with chief complaints of dark stools. He was admitted in Medicine Department of Government Medical College and Hospital, Aurangabad. On investigations, he was earlier diagnosed as diabetes mellitus and was on Inj.Insulin. Ocular examination revealed Vision PL+, best corrected vision RE 6 / 60 LE 6 / 18 on Snellen's chart,

Slitlamp examination revealed in both eyes –

- Lid : Normal,
- Conjuctiva : Normal
- Cornea : Clear
- Anterior Chamber : Normal epth
- Iris / Pupil : Circular normally reacting to light
- Lens : Clear
- Pendular nystagmus present
- Intra ocular pressure in both eyes was normal.

Fundus - Media clear, disc pallor++, arteriolas attenuation and pigmentary changes (bone corpuscular pigment along vessels. He was diagnosed as R.P.

Systemic Examination :

- Ear – Audiometry s / o bilateral severe sensorineural hearing loss more at high frequency.
- Mental Capacity – studied till X std. in Braille School.
- Skin
- USG
- Echocardiography
- Family History
- Treatment : Inj. Insulin.

Characteristic Features :

1. Absence of polydactyly / syndactyly.
2. Males and females inherited with equal probability.
3. Cases common in Holland and Sweden.

Discussion :

Pigmentary degenerations of the retina has been associated with multiple systemic associations. Variants are seen and some manifestations are absent. Uptill now very few cases have been reported. In Indian reference, we found 4 references.

Summary :

1. Pigmentary retinopathy leading to blindness.
2. Sensorineural deafness.
3. IDDM
4. Acanthosis migrans
5. Short stature
6. Normal mental capacity.

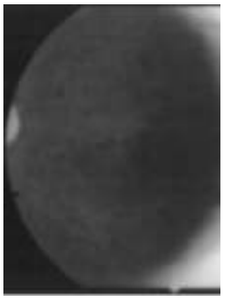
Conclusion :

A diagnosis should be considered in congenital sigmentary retinal dystrophy particularly when association of DM and cardiomyopathy and deafness. As patient have normal mental capacity, rehabilitation of the patient must be done to reduce economic burden. As there are very few cases reported all over and specially in India, support groups are not established.

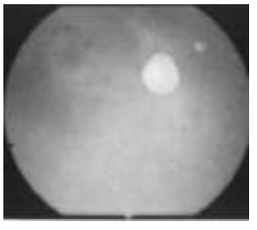
References :

1. Raja LN, Deodhar JN, Magotra ML : Journal of Indian Pediatrics, 1982.
2. RA Garg, BB Mathur, J Singh : Journal of Indian Pediatrics, July 1991, Vol.28; No.7 : pg.799-801.
3. Lt.Col.GP Singh, Maj.VS Gurunath, Lt.Col.s.Chaudhary : Medical Journal Armed Forces India, Jan.1995.
4. Rajan SK, Ameen KHN, Hariharan LM, Natarajan VS : Journal of Associate Physician of India 2002.

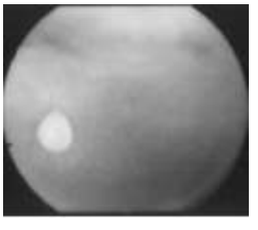
Pic 1



Pic 2



Pic 3



Pic 4

Dr.Vaishali Une
Government Medical College, Aurangabad

Goldenhar Syndrome: A case report & review of literature

Dr Pooja Jain, Dr Asif Virani, Dr Chhaya Shinde , Dr Nayana Potdar

Goldenhar syndrome also called oculo auriculo vertebral dysplasia or first & second branchial arch syndrome, is mostly unilateral, occurs most commonly in men and involves anomalies of face, ears, eyes and vertebrae in most cases. When malformations primarily involves the jaw, mouth and ear of one side, the disorder is referred as Hemifacial microsomia, but if abnormalities of vertebrae and the eyes are also present, the disorder is often called Goldenhar syndrome.

Case History :
A seven year female patient with negative family history presented to the outpatient department of sion hospital with dimness of vision in right eye and painless swelling in right eye since birth. On ophthalmic examination, Best Corrected Visual Acuity in right eye was 6/36 and in left eye it was 6/6 not improving with refraction. There was against the rule astigmatism of 3 dioptre due to inferolateral limbal dermoid of about 5X5 mm size. She had 15 degrees concomitant divergent squint with full eyeball movement. There was no other ocular abnormality. Fundus examination was normal in both eyes.

On ENT examination, patient had pre auricular skin tags which were lying in line joining tragus & angle of mouth. Patient had no complaint of deafness in both the ears. X-ray PNS & Schullers views were also normal with normal external auditory meatus.
CVS & CNS examination was normal.
X-ray cervical spine was not showing occipitalisation of atlas or hemi vertebrae which is usually associated with Limabal Dermoid in Goldenhar syndrome.
CT scan was not showing any abnormality.
Thus, the patient was having following findings:
Limbal dermoid(figure 1) Abnormalities of ear (pre auricular skin tag) (figure 2 & 3)

Discussion :
The incidence of this syndrome is 1:3000 to 1:26,500 with onset at birth. It occurs in family & / or as sporadic cases. Familial cases are consistent with autosomal dominant, autosomal recessive and multifactoral patterns of inheritance. Chromosomal abnormality or defective gene disorder is common in men (70%). It can be unilateral or bilateral but mostly it is unilateral.

Pathogenesis :
Genetic defect: abnormal neural crest morphology-malformation of the derivatives of the first & second branchial arches-facial, auricular, ocular, vertebrae & other anomalies.(figure 4,5 & 6) Variable phenotypic expression is characteristic with this syndrome & the spectrum of phenotypic anomalies can range from mild to severe even within the same affected family. It may be present in monozygotic twins with only one twin being affected.

Review of Literature :
First identifies by M.Goldenhar in 1952 First called oculo auriculo vertebral dysplasia by R.J. Gorlin.

Clinical Features Facial asymmetry :
65-70% cases have some facial asymmetry & in 20% of cases the asymmetry is marked.
The right side of the face is more severely affected in 60% of cases.
10-33% has bilateral facial involvement and / or mandibular regions on the affected side.
There also may be unilateral hypoplasia of the facial musculature with facial muscle weakness and depressor anguli oris hypoplasia on the affected side.

Macrostomia :
Greatly exaggerated width of the mouth due to lateral cleft like extension of the corner of the mouth.

External Ear :
May range from mildly dystrophic ear to microtia to anotia. Pre-auricular skin tags, pits and / or sinuses and occur between the tragus and the angle of the mouth.
Usually unilateral but can be bilateral in 33% of cases.

Middle Ear :
Narrow or atretic(figure 7)

Inner Ear :
Conductive & / or sensorineural hearing loss.
Ocular Manifestations
Epibulbar Tumors
- Dermoid or lipodermoids.
- Found in 35% of cases.
- Yellow or pink, solid ovoid masses up to 10 mm in diameter.
- Unilateral or bilateral.
- May impair vision.

Others :
- Strabismus
- Microphthalmia or anophthalmia.

Dr Pooja Jain
LTMMC & LTMGH Sion, Mumbai

- Blepharo-phimosis in 10% of cases.
- Narrowing of palpebral fissure
- Retinal anomalies.
- Upper lid colobomas.

Other manifestations Neurological :
- Microcephaly or hydrocephaly.
- Occipital encephalocele.
- Arnold chiari malformation.
- Mental retardation in 5-15% of patients.
- Spina bifida.

Congenital Heart disease :
- VSD, PDA, Tetralogy of Fallot, Coarctation of Aorta.
- Transposition of Great arteries.

Renal :
- Ectopia & / or fused kidneys.
- Renal agenesis.
- Multicystic dysplastic kidneys.
- Hydronephrosis, hydro-ureter, ureteral duplication.
- Uretero-pelvic junction obstruction.
- Vesico-ureteral reflux.

Musculoskeletal :
- Scoliosis
- Talipes equino varus (club feet)
- Bifid or digitalized thumb
- Hypoplastic Thumb

Investigations
Skeletal X-Rays(fig 7) Skull :
- Hypoplasia of the maxillary, temporal & malar bones.
- Hypoplasia or absent mandibular ramus & condyles.
- Reduced pneumatization of mastoid region.

Vertebral :
- Cervical fusion in 20-25% of cases.
- Platybasia & occipitalisation of the atlas in 30% of cases.
- Klippel-Feil anomaly.
- Hemi vertebrae
- Hypoplasia of vertebrae

CT/MRI :
- Agenesis of or lipoma in corpus callosum
- Calcification of falx cerebri
- Hypoplastic septum pellucidum.
- Intra-cranial dermoid cyst.

Management Supportive Multidisciplinary approach
- Pediatrics, plastic, orthopaedic, ophthalmic and ENT surgeons
- Early and regular hearing tests.
- Genetic counselling.

Prognosis :
Normal life span and intelligence in a majority of those patients without significant complications.

Goldenhar Syndrome Axial :: CT w / contrast (IV):
Contrast enhanced axial CT images of the neck demonstrate agenesis of the right pinna, external auditory canal and middle ear. The right inner ear appears intact with slight increased sclerosis about its osseous components. The right mastoid air cells are essentially absent. The right zygomatic arch is hypoplastic and the right mandibular ramus and condyle are markedly hypoplastic and malformed. The right mandible does not articulate at the TMJ. The globes appear symmetric bilaterally.

References :

1. Baraitser M. & R. M. Winter, color atlas of congenital malformation syndrome (1st edition P : 40-41 (1996).
2. Baum J. L. & Feingold M., ocular aspects o Goldenhar syndrome. AJO, 75:250, 1973.
3. Bergsma D. (Ed) Birth defects, atlas and compendium. Baltimore: Williams & Wilkins.
4. Benjamin S. N. & Allen H. F. classification for limbal dermoids, choriostomas & branchial arch anomalies: presentation of an unusual case. Arch Ophthalmology, 87:305, 1972.
5. Geeraets W. J. ocular syndromes (3rdedi) Philadelphia: Lea & Febiger, 1976. Jones K. L., Smith's Recognizable patterns of human malformations (5th edi), P: 642-645 (1997).
6. Journal of Genetics, Human 1:243 (l952).
7. Journal of Pediatrics 63:991-999 (1963).
8. Mansour A. M. et. al, ocular findings in facio auriculo vertebral sequence (Goldenhar Gorlin syndrome) AJO 100:555, 1985.
9. Gorlin RJ, Cohen MM Jr, Levin LS.Oculoauriculovertebral Spectrum.In: Syndromes of Head and Neck.3rd ed.oxford University Press: New York; 1990.p.641-9
10. Das A, Mazumdar S, Chatterjee SS.Atypical case of Oculo-Facioauriculo - vertebral Dysplasia (Goldenhar Gorlin Syndrome).IJO 1999; 47:131-3
11. A. Das, B.Ray, D.Das, S.Das.A Case of Goldenhar Gorlin syndrome with unusual association of Hypoplastic thumb.IJO 2008; 56:150-2

Fig 1

Fig 2

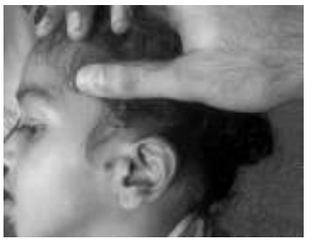


Fig 3

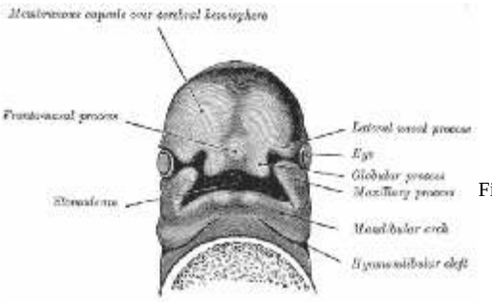


Fig 4

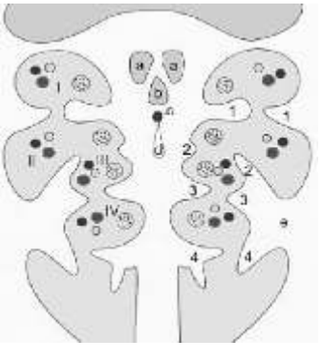


Fig 5

Arch / nerve	Sreleial	Ligamenis	Muscles	Pouch
Fist (V)	1) Malleus 2) Incus	1) Ant Ligament of Malleus 2) Sphenomandibular Ligament	1) Muscles of Mastication 2) Tensor tympani 3) Tensor Paiati 4) Mylohyoid 5) Ant belly of Drastic	1) Auditory tube 2) Tympanic Cavity
Second (VII)	1) Stapes 2) Styloid process 3) Hyold bonelesser hom upper half of body	Stylohyold Ligament	1) Muscles of Facial Express on 2) Stapedus 3) Stylohyold 4) Post belly of Didactic	Linng (crypts) of Palatine Onsils
Third (IX)	Hyold bone greater hom, lower half of body	----------	Stylophatryngeus	1) Interior parathyroid gland 2) Thymus
Fourth (X)	Certilages of Larynx	----------	1) All muscies of Larynx 2) All Muscies of Pharynx (except Stylophanyngeus) 3) All muscies of Soft Palate (except Tensor Palat)	1) Supernor parathyroid gland 2) C-cells of Thyroid
Sisth (XI)	----------	----------	1) Sternocleidomastocd 2) Trapezes	----------

Fig 6

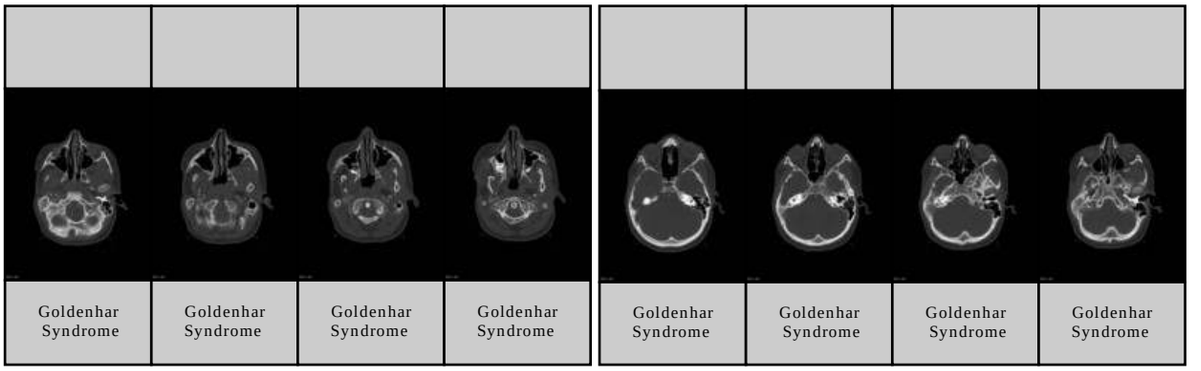


Fig 7

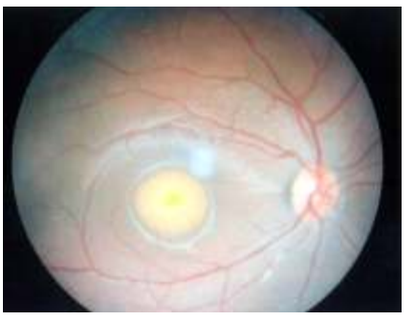
Juvenile Best Macular Dystrophy

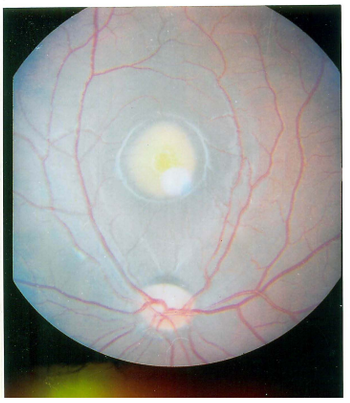
Dr Indrajotkaur Ahuja, Dr Rajat Gupta , Dr Megha Arora, Dr Amit Pandey , Dr Karishma Sardeshpande , Dr Mahananda Galgali, Dr Suresh Ramchandani

Best's vitelliform dystrophy is an autosomal dominant disease[1]. It affects patients of age group 3-15yrs even though adult cases have been reported[2,3].It pathologically involves the RPE and symmetrically affects the macula[1]. Visual acuity tends to remain quiet good for long periods of time. In later stages atrophic changes of RPE or scarring secondary to subretinal neovascularization with haemorrhage may cause a central visual loss[1,4].

Case report:

Our patient is a 6yrs boy with no systemic illness. Chief complaint as per mother was dimunition of vision in both eyes with nystagmus. There was no other significant history. On examination visual acuity was 6/60 OD and 6/36 OS. Both eyes fundus examination revealed symmetrical involvement of macula. The lesion was 3 DD in size ,egg yellow colored , round and elevated structure. Patient was not cooperative for Intraocular tension and EOG. So the diagnosis of bilateral best's macular dystrophy was made on the basis of fundus examination. The condition was explained and the patient's mother counselled. No active treatment was advised.





Discussion:

Juvenile Best's macular dystrophy is autosomal dominant with variable penetrance and expressivity with gene locus on 11q13[5]. Dr Franz Best, a German ophthalmologist, described the first pedigree in 1905 [6,7] .The presentation varies from a small round yellow dot at fovea to a chorioretinal scar resembling toxoplasmosis . Various stages are :

Normal fovea (pathologic EOG)
Previtelliform
Vitelliform
Scrambled egg stage
Cystic stage
Pseudohypopyon
Round chorioretinal atrophy

In Previtelliform stage lesion is a small,round yellowish dot at the site of foveola or tiny honeycomb structure centrally. Classic vitelliform lesion is egg yellow or orange round elevated structure surrounded by a darker border , 0.5 – 3 disc diameters in size. Peripherally vitelliform structures may be seen with a normal looking macula. The contents of disc disintegrate forming a scrambled egg appeareance and later a cyst or a cyst with fluid level resembling a pseudohypopyon. Complications like subretinal neovascularization , haemorrhage and consequently macular hole or a chorioretinal atrophic scar is left.Visual acuity not much affected.slightly subnormal in vitelliform stage. And sudden decrease in later stages due to complications . the loss is reversible

Visual fields show a decreased sensitivity. Scotoma for red and later for green. A relative scotoma for white and a central absolute scotoma in later stages. Color vision shows mild red-green dyschromatopsia [1,8]

Dark adaptation and ERG is normal[1]. EOG is hallmark of the disease[9]

EOG is subnormal with light-dark ratio rarely higher than 1.5. even subnormal in carriers [1,6]. FFA shows hypofluorescence when the disc is intact and on rupture atrophic areas of RPE show hyperfluorescence. Histologically , there is predominant atrophy of RPE and accumulation of lipofuschin granules within RPE,in subretinal space.

No other significant ocular abnormalities or systemic problems have been associated with this genetic disorder.

No therapy exists for halting the progression of disease. Laser photocoagulation along with intravitreal bevacizumab for SRNV [8] .Accurate diagnosis and pedigree analysis is important for family and genetic counselling.

Dr Suresh Ramchandani
MGM Hospital And Research Centre- Kamothe . Navi Mumbai

References:

1. *Stephen J Ryan . Vitelliform macular dystrophy . Retina : Elsevier:4(1) : 673-4.*

2. *Zhuk SA, Edwards AO. Peripherin/RDS and VMD2 mutations in macular dystrophies with adult-onset vitelliform lesion. Mol Vis. 2006; 12: 811–5.*

3. *Renner AB, Tillack H, Kraus H, Kohl S, Wissinger B, Mohr N, Weber BH, Kellner U, Foerster MH. Morphology and functional characteristics in adult vitelliform macular dystrophy. Retina. 2004; 24: 929–39.*

4. *Fishman GA, Baca W, Alexander KR, Derlacki DJ, Glenn AM, Viana M. Visual acuity in patients with best vitelliform macular dystrophy. Ophthalmology. 1993; 100: 1665–70.*

5. *Jack J Kanski.Fundus dystrophies.Clinical Ophthalmology A Systematic Approach :Elsevier:2007:6:672*

6. *Best F. Ubereine hereditare Maculaaffektion. Beitrage zur Vererbungslehre. Augenheilkd. 1905;13:199.*

7. *Falls HF. The polymorphous manifestations of Best's disease (vitelliform eruptive disease of the retina). Trans Am Ophthalmol Soc. 1969;67:265 82. [Medline]*

8. *Graefe's Arch Clin Exp Ophthalmol (2007) 245:1723–1725*

9. *Testa F, Rossi S, Passerini I, Sodi A, Di Iorio V, Interlandi E, Della Corte M, Menchini U, Rinaldi E, Torricelli F, Simonelli F. A normal electro oculography in a family affected by best disease with a novel spontaneous mutation of the BEST1 gene. Br J Ophthalmol. 2008; 92: 1467–70.*

10. *Blodi CF, Stone EM. Best's vitelliform dystrophy. Ophthalmic Paediatr Genet. Mar 1990;11(1):49-59. [Medline].*