Editorial

Perspective — Duane's Retraction Syndrome
— Charuta Bapaye, Sumita Agarkar - SN-ORBIS POLTC

Muscle Puzzle — Uday Naik, Meenakshi S - SN-ORBIS POLTC

Histopathological analysis of 768 enucleated eyes over a period of 7 years in a Tertiary Eye Care Hospital — Jyotirmay Biswas, S Krishnakumar, P Vaijayanthi, K. Vanitha — Department of Ocular Pathology

Chiasmal syndromes — G. Balu, Shikha Bassi — Neuroophthalmology Department

Comparison of Freiburg Visual Acuity and Snellen Visual Acuity tests
— Sheela Evangeline K, Ashwini M, Varadharajan J, Krishna Kumar R
— Elite School of Optometry, Unit of Medical Research Foundation, Chennai

Technology update: — MICROPERIMETER
— Munneswar Gupta Nittala — Sankara Nethralaya Diabetic Retinopathy Project

Left SDOCT image showing area of photoreceptor layer loss. Right microperimeter image showing dense scotoma area where photoreceptor layer loss seen.
EDITORIAL

Greetings.

Enucleation is an end stage surgery performed for varied eye diseases. Article on the histopathological analysis of the enucleated eyes gives a preview about its common indications. Duane’s and Chiasmal syndromes are covered in crisp details to refine our existing clinical knowledge. The comparative study of Freiburg and Snellen visual acuity tests is presented to analyze sensitivity of each as rapid screening tests. Microperimetry- a new diagnostic tool in fundus examination is covered. A muscle puzzle is included to highlight a common strabismus entity.

Dr S Meenakshi - Editor
Dr Shubhra Goel - Associate Editor

ERRATA

This is in reference to the article “Establishment of primary cultures of bovine retinal capillary endothelial cells and pericytes for in-vitro studies” (Vol.XXV No.3 October 2007, page 55).

In the first paragraph of Materials and Methods the dosage of the antibiotics, amphotericin and streptomycin in ‘mg/ml’ is to be read as ‘μg/ml’.

The second paragraph mentioning about the size of filters of the endothelial cells ‘41 m,nylon’ and pericytes ‘60’ is to be read as 41μ and 60μ respectively.
Duane's retraction syndrome (DRS) is an ocular disorder consisting of retraction of the globe with narrowing of the palpebral fissure in attempted adduction, frequent abduction deficiency with variable limitation of adduction, and upshoot and/or downshoot of the affected eye on adduction.\(^1\)

Stilling in 1887 and Turk in 1896 along with many others described this complex syndrome. In 1905 Alexander Duane presented a series of 54 patients sharing these common features. The incidence of Duane's syndrome has been reported in literature as 1-4\% of strabismus patients.\(^1\) There is however no Indian study in this context.

**ETIOLOGY**

A glance through literature reveals the evolution of theories regarding the etiology of DRS. Turk and Wolfe described that it was caused by the lateral rectus being replaced by an inelastic connective tissue band. Gallus hypothesized that this fibrous transformation occurs either as a developmental anomaly secondary to nuclear aplasia, or as a result of hemorrhage in the muscle or damage to the tendon during birth.

Duane felt that the cause was a double insertion of the medial rectus, one component rotating the globe and the other retracting the globe on adduction. He himself later admitted that this was unlikely, as all posterior insertions do not give rise to this phenomenon.

With the advent of electromyography, Breinin described the innervational anomaly behind DRS i.e. absence of electrical potentials of the lateral rectus on abduction, but the presence of action potentials on adduction. This co-contraction of the medial and lateral rectus during adduction was thought to be responsible for retraction of the globe. The observation of slowing of adduction saccades in these patients with limited abduction also supported this theory.

Autopsy studies showed that this innervational anomaly was due to absence of the VI nerve or its nucleus, due to which the part of the mesoderm destined to become the lateral rectus continues to derive innervation from the III nerve nucleus. MRI studies later corroborated this finding. Further observations revealed that DRS is associated with several other anomalies, all of which are of structures developing in the 4\(^{th}\) to 10\(^{th}\) week of gestation. Therefore it was postulated that a teratogenic insult occurring during this period might be responsible for this. Various agents like thalidomide and alcohol were implicated.\(^1,2,3\)

**INHERITANCE**

Most cases of DRS are sporadic. There are reports of familial occurrence in about 10\% cases.\(^1\) However no definite inheritance pattern has been identified. There can be several reasons for this such as incomplete penetrance with variable expressivity of the gene, or environmental modification of the gene. Since DRS occurs in otherwise healthy patients mild cases may be missed. Sometimes co-occurrence of the syndrome in the family may be coincidental.

**DEMOGRAPHICS**

Most studies report a 60\% predominance in females. It is hypothesized that this may be due to the gene being partly sex-linked. The left eye is more commonly involved in a ratio of 3:1. No particular ethnic
group is predisposed to DRS. 18% cases are bilateral. In hereditary cases bilaterality can be as high as 96%.\textsuperscript{1} Reports in literature reveal asymmetry in bilateral cases.

\textbf{CLASSIFICATION}

\textbf{Huber’s classification}\textsuperscript{1}

\textbf{Type I}: Esotropia with marked limitation of abduction with minimally defective or normal adduction, widening of the fissure on abduction.

EMG shows peak electrical impulses of the lateral rectus in adduction and weak impulses on abduction. The impulses of MR are normal.

\textbf{Type II}: Exotropia with marked limitation of adduction with normal or slightly defective abduction, narrowing of the fissure on attempted adduction.

EMG shows peak innervation of lateral rectus in both adduction and abduction and normal behavior of the medial rectus.

\textbf{Type III}: Marked limitation of both adduction and abduction. EMG shows intense innervation of both medial and lateral rectus in all gazes.

\textbf{CLINICAL FEATURES}

\textbf{Globe retraction} - This is seen maximum in adduction in slight down gaze, best seen from the side after retracting the lids. It occurs due to co-contraction of the horizontal recti. It may be absent in mild cases.

\textbf{Narrowing of palpebral fissure on attempted adduction} - This occurs as a passive response to globe retraction due to lowering of the upper lid and raising and straightening of margin of the lower lid. It is the least dependable sign. It may occur physiologically in otherwise normal eyes also.

\textbf{Abduction deficiency} - is usually the most prominent feature with variable limitation of adduction.

\textbf{Upshoot / Downshoot on adduction} - This occurs in more severe cases. There have been several theories to explain its occurrence. These can be differentiated from oblique over action as they are more abrupt and more grotesque.

\textbf{Bridle effect/ Side slip theory}\textsuperscript{1,4}

The up/down shoot occurs when the eye starts moving up/down in adducted position. The original description was that when the eye adducts the tight lateral rectus indents on the globe, and when the eye starts moving up or downwards in adduction the tight muscle slips off the crest of the globe causing the up/down shoot. However imaging studies have shown that it is the eye that slips under the muscle. The lateral rectus maintains its perpendicular position with respect to the lateral orbital wall. When the globe starts moving down/up the relation of the lateral rectus to the center of the globe changes such that it becomes a depressor/ elevator, resulting in a downshoot/upshoot.

\textbf{Co-innervation of vertical rectus with lateral rectus}\textsuperscript{1}

In these cases the eye is hypo/hyper tropic in primary position and shoot occurs from the horizontal position. This may be the etiology in certain cases where the shoot can be eliminated by injecting xylocaine into the vertical rectus muscle.

\textbf{Oblique muscle overaction}

This is no longer an accepted cause of up/down shoot.

\textbf{Small angle strabismus} - Horizontal deviations are more common. The deviation is usually less than 30PD. The magnitude of deviation is thus out of proportion to the movement limitation. Esotropia is most common and occurs in Type I DRS. Exotropia occurs in Type II DRS. The patient may even be
orthotropic in primary position. In patients with significant adduction and abduction deficit (Type III) there may be esotropia in abduction, exotropia in adduction and may be orthotropic in primary position.

Vertical deviations can occur in primary position either due to a co-existing congenital superior oblique palsy due to absence of the IV nerve nucleus, a combined horizontal and vertical retraction syndrome, or abnormal superior or inferior insertion of the lateral rectus. Chronic upshoot leading to superior rectus contracture and hypertropia has been described in literature. A corresponding chronic downshoot as a cause of hypotropia has however not been described.

**Face turn** - This may allow preservation of fusion and stereopsis. It is not an essential feature.

**Pattern** - The V pattern is more common than A pattern in unilateral DRS. In bilateral cases A pattern is more common.  

**OCULAR ASSOCIATIONS**

- Anisometropia
- Amblyopia - Anisometric amblyopia is more common than strabismic
- Nystagmus
- Epibulbar dermoid
- Anisocoria
- Ptosis
- Crocodile Tears

**Uncommon**
- Optic Nerve hypoplasia
- Marcus Gunn jaw wink
- Cataract
- Microphthalmos
- Prominent epicanthal fold
- Persistent hyaloid artery
- Ectropion of lower lid
- Situs inversus of optic disc
- Myelinated nerve fibres

A complete ocular examination is therefore essential, though, in majority cases it may be normal.

**SYSTEMIC ASSOCIATIONS**

Some reports in literature show no systemic associations in hereditary cases, while some authors mention genetic links between DRS and certain disorders like deafness. Sporadic cases are associated with congenital malformations of structures that develop in the middle of the first trimester.

Studies show more frequency of congenital malformations in DRS than in the general population. With detailed investigations the incidence increases from 15 to 50%. Therefore systemic examination, at least an audiometry would be worthwhile in patients with DRS. A detailed gestational history should be taken before assuming a genetic problem.

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Developing structure</th>
<th>Gestational age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perceptive deafness</td>
<td>Auditory ossicles</td>
<td>6-8 weeks</td>
</tr>
<tr>
<td>Klippel-Fiel anomaly</td>
<td>Vertebrae, Pinna</td>
<td>4-8 weeks</td>
</tr>
<tr>
<td>Goldenhar syndrome</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thenar hypoplasia</td>
<td>Upper extremity</td>
<td>3-7 weeks</td>
</tr>
<tr>
<td>Cardiac anomalies</td>
<td>Ventricular septum</td>
<td>5-6 weeks</td>
</tr>
<tr>
<td>Cleft palate</td>
<td>Palate, oral fissure</td>
<td>7-10 weeks</td>
</tr>
<tr>
<td>Renal anomalies</td>
<td>Kidneys</td>
<td>4-5 weeks</td>
</tr>
</tbody>
</table>
INVESTIGATIONS

1. Forced duction test - Helps in the surgical plan.

2. Force degeneration test\(^5\) - In this test passive adduction is possible when the patient attempts abduction due to weak action of lateral rectus, but resistance to passive adduction is felt when the patient attempts abduction due to paradoxical firing of lateral rectus on attempted adduction.

3. Saccadic velocity testing \(^1,5\) - DRS has reduced abduction and adduction saccades due to paradoxical innervation.

4. EMG - Not required for diagnosis.

DIFFERENTIAL DIAGNOSIS \(^1\)

1. VI nerve palsy - It is necessary to differentiate this from DRS to avoid unnecessary investigations in a patient with DRS and to formulate the right surgical plan.

The deviation in VI nerve palsy will be in proportion to the abduction limitation, while in DRS the deviation is generally small inspite of a gross abduction limitation. Forced duction test may or may not be positive in either. Force degeneration test for paradoxical innervation will be positive only in DRS. Both conditions will have reduced abduction saccades while reduced adduction saccades will be present only in DRS.

2. Infantile esotropia - An infant with DRS may present with only abduction limitation as other features may develop later. Infantile esotropia also has a large angle of deviation as opposed to DRS.

3. Moebius syndrome - Mask facies and associated limb, chest and tongue anomalies can differentiate this condition from DRS.

4. Congenital Oculomotor apraxia - Impairment of voluntary horizontal gaze may mimic DRS. Jerky horizontal head thrusts are present in the former.

MANAGEMENT

Non-surgical management

1. Correction of refractive error (more commonly hyperopia) and anisometropia
2. Treatment of amblyopia in children.

Indications for surgical treatment

1. Noticeable ocular deviation in primary position.
2. Marked abnormal head posture.
3. Marked globe retraction.

PRINCIPLES OF SURGERY

1. No resection - as it worsens movement limitation, retraction and upshoot/downshoot and does not improve head posture.
2. Recession of overacting muscle is the safest and most effective alternative.
3. Adjustable sutures may be used where possible.

SURGICAL OPTIONS \(^1,9,10,11,12,13\)

1. For esotropia and face turn
   - Recession of ipsilateral or bilateral medial rectus is done depending on the amount of deviation. Recessing the normal medial rectus gives the advantage of fixation and prevents risk of adduction limitation following large unilateral medial rectus recession. It does not cause an adduction deficit in the normal eye. If significant globe retraction is also present then a differential recession of medial recti along with lateral rectus recession is done.
   - Faden operation can be used as a substitute to recessing the normal medial rectus, however experience with it is limited.
   - Transposition of vertical recti to the lateral rectus creates a passive exo-force due to resting tone of the vertical recti being transferred temporally. It may be done as full tendon or partial tendon to spare the ciliary vessels. It is useful in cases where the paradoxical innervation is not severe.
If it is, the recession of the lateral rectus is added to the procedure. It is combined with recession of the medial rectus if it is tight. However this procedure runs the risk of over/undercorrection, of inducing vertical tropias and of anterior segment ischaemia. 10-15 degrees of abduction may be achieved, however this may not improve the binocular field significantly.

2. For exotropia
   - Ipsilateral or bilateral lateral rectus recession is done depending on the amount of deviation.

3. For retraction and upshoot / downshoot
   - Recession of both horizontal recti of affected eye is an option. Posterior fixation suture may be added.
   - Y-splitting with recession of the lateral rectus is effective. The lateral rectus is split as far back as possible and the two halves are splayed 10mm above and below the original insertion. This removes the muscle from the crest of the globe and thus eliminates upshoot / downshoot. As it makes the muscle tight, recession should be done with it.

SURGICAL OUTCOME

Surgical results are not very encouraging and recurrences are common. Ocular motility does not improve and the patient needs to be warned about it pre-operatively. Therefore patients who can maintain fusion and good vision with a slight face turn should not be operated on. Surgery should be done only after a proper assessment only for a positive indication.

Reference


Muscle Puzzle
Uday Naik, Meenakshi S — SN-ORBIS POLTC

16 year old female came with complaints of inward deviation of eyes since birth. Her family history, birth history, treatment history was not contributory. She had Right head tilt. Stereopsis was 500 sec of arc. Her vision with +/- was 6/6, (N6) in both eyes. What is the Diagnosis and treatment?

This patient has infantile esotropia with Bilateral DVD with a Right head tilt.

YOUR DIAGNOSIS ?

(Answer on page 22)

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*Educational Material for optometrists and ophthalmic nursing still in the process of being built up.
Histopathological analysis of 768 enucleated eyes over a period of 7 years in a Tertiary Eye Care Hospital

Jyotirmay Biswas, S Krishnakumar, P Vaijayanthi, K. Vanitha — Department of Ocular Pathology

INTRODUCTION

In our study, we analyzed the main cause for enucleation performed in a tertiary eye care institute. The indications for enucleation were the following: 1) malignant intraocular tumors, 2) painful blind eye, 3) Staphyloma and 4), traumatic ruptured globe. Our objective is to determine the age group distribution; compare pediatric and adult enucleation; and histopathological analysis of eyes enucleated for malignant intraocular tumors.

MATERIALS AND METHODS

A comparison was made between enucleation made from January 2000 – June 2003 and those made from July 2003 to December 2006. The data was retrieved from Snowmed coding system, which was used for coding the specimen in our pathology laboratory.

Enucleated globes were fixed in 10% neutral buffered formalin for 48 hours in 1 in 10 dilutions. A short cylindrical segment including cornea, lens and iris anteriorly and optic nerve posteriorly was used for analysis. Samples of vortex veins were taken, particularly if malignant melanoma was suspected, to look for invasion. Histopathologic slides from all cases were stained with Haematoxylin and Eosin and reviewed for characterization.

RESULTS

Among the total 768 patients, 490 were males (63.8%) and 278 were females (36.2%) with a male to female ratio of 1.76:1. 55.07% of the patients were of pediatric age group (<15 years) whereas only 4.5% were above 60 years. (Table - 1a and 1b)

Histopathological analysis of retinoblastoma, malignant melanoma and other

<table>
<thead>
<tr>
<th>S No.</th>
<th>Age group (in years)</th>
<th>Number of patients</th>
<th>Male patients</th>
<th>Female patients</th>
<th>% Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0-5</td>
<td>335</td>
<td>197</td>
<td>138</td>
<td>43.61</td>
</tr>
<tr>
<td>2</td>
<td>6-10</td>
<td>50</td>
<td>31</td>
<td>19</td>
<td>6.51</td>
</tr>
<tr>
<td>3</td>
<td>11-15</td>
<td>38</td>
<td>26</td>
<td>12</td>
<td>4.95</td>
</tr>
<tr>
<td>4</td>
<td>16-30</td>
<td>123</td>
<td>86</td>
<td>37</td>
<td>16.02</td>
</tr>
<tr>
<td>5</td>
<td>31-40</td>
<td>72</td>
<td>56</td>
<td>16</td>
<td>9.38</td>
</tr>
<tr>
<td>6</td>
<td>41-50</td>
<td>60</td>
<td>36</td>
<td>24</td>
<td>7.81</td>
</tr>
<tr>
<td>7</td>
<td>51-60</td>
<td>55</td>
<td>33</td>
<td>22</td>
<td>7.16</td>
</tr>
<tr>
<td>8</td>
<td>61 and above</td>
<td>35</td>
<td>24</td>
<td>11</td>
<td>4.56</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td></td>
<td><strong>768</strong></td>
<td><strong>490</strong></td>
<td><strong>278</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>

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Table 1b: Enucleation done for pediatric age group & adult age group

<table>
<thead>
<tr>
<th>S No.</th>
<th>Diagnosis</th>
<th>No of cases (0-15 years); N = 423</th>
<th>No of cases (15 and above); N = 345</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Retinoblastoma</td>
<td>326</td>
<td>4</td>
</tr>
<tr>
<td>2</td>
<td>Malignant melanoma</td>
<td>5</td>
<td>80</td>
</tr>
<tr>
<td>3</td>
<td>Ocular inflammation</td>
<td>3</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>Staphyloma</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>5</td>
<td>Painful blind eye Glaucoma</td>
<td>26</td>
<td>111</td>
</tr>
<tr>
<td>6</td>
<td>Phthisis bulbi</td>
<td>13</td>
<td>3</td>
</tr>
<tr>
<td>7</td>
<td>Coats disease</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>9</td>
<td>Hemangioma</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>10</td>
<td>Medulloepithelioma</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>11</td>
<td>Silicone oil globules</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>12</td>
<td>Hemorrhage/trauma</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>13</td>
<td>Peripheral nerve sheath tumor</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>Haematoma</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>15</td>
<td>Leiomyoma</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>16</td>
<td>Squamous cell carcinoma</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>17</td>
<td>Metastatic carcinoma</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>18</td>
<td>Hyphema</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>19</td>
<td>Phakolytic inflammation</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>20</td>
<td>Microphthalmos cyst</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2: Histopathological analysis of Retinoblastoma

<table>
<thead>
<tr>
<th>S No</th>
<th>Retinoblastoma (n=330)</th>
<th>No. of cases</th>
<th>% age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>With invasion</td>
<td>97</td>
<td>29.39</td>
</tr>
<tr>
<td>2</td>
<td>Without invasion</td>
<td>233</td>
<td>70.61</td>
</tr>
<tr>
<td>3</td>
<td>Choroidal invasion</td>
<td>93</td>
<td>24.47</td>
</tr>
<tr>
<td>4</td>
<td>Well differentiated</td>
<td>115</td>
<td>34.84</td>
</tr>
<tr>
<td>5</td>
<td>Poorly differentiated</td>
<td>86</td>
<td>26.06</td>
</tr>
<tr>
<td>6</td>
<td>Undifferentiated</td>
<td>129</td>
<td>39.09</td>
</tr>
</tbody>
</table>

Table 3: Histopathological analysis of Melanoma

<table>
<thead>
<tr>
<th>S No</th>
<th>Malignant melanoma of the choroid (n=85)</th>
<th>No. of cases</th>
<th>% age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Invasion</td>
<td>16</td>
<td>18.82</td>
</tr>
<tr>
<td>2</td>
<td>No invasion</td>
<td>69</td>
<td>81.18</td>
</tr>
<tr>
<td>3</td>
<td>Mixed cell type</td>
<td>45</td>
<td>52.94</td>
</tr>
<tr>
<td>4</td>
<td>Epithelioid cell type</td>
<td>7</td>
<td>8.24</td>
</tr>
<tr>
<td>5</td>
<td>Spindle cell type</td>
<td>5</td>
<td>5.88</td>
</tr>
<tr>
<td>6</td>
<td>Other types</td>
<td>28</td>
<td>32.94</td>
</tr>
</tbody>
</table>

Lesions were done and the statistics are summarized in the tables (2, 3, 4 & 5) respectively.

Indications for enucleation during each year are given in Table 6. There were 305 eviscerations during the period of 2000-2006 (Table 7). An increasing trend for evisceration for indications other than intraocular tumors was seen in recent years.

In the present series, malignant tumors (54.03%) were the most frequent indication for enucleation followed by painful blind eye (16.7%), intraocular inflammation (1.6%), trauma (1.17%) and staphyloma (0.6%). Retinoblastoma was the most common ophthalmic tumor (42.96%) requiring enucleation followed by malignant melanoma of the choroid (11.06%).

DISCUSSION

Enucleation of the eye is a major organ-removal surgery performed for end-stage eye disease. Enucleated eyes contributed to 10% of the total volume of the specimens received in the laboratory during 7 year period.
Table 4: Various types of enucleated specimens in different periods

<table>
<thead>
<tr>
<th>S No.</th>
<th>Different types of enucleated specimens</th>
<th>No. of cases (Jan-2000 to Jun-2003)</th>
<th>No. of cases (Jul-2003 to Dec-2006)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Ocular Inflammation (Endophthalmitis and Panophthalmitis)</td>
<td>7</td>
<td>6</td>
</tr>
<tr>
<td>2.</td>
<td>Staphyloma</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>3.</td>
<td>Painful blind eye / Glaucoma</td>
<td>72</td>
<td>57</td>
</tr>
<tr>
<td>4.</td>
<td>Phthisis bulbi</td>
<td>29</td>
<td>49</td>
</tr>
<tr>
<td>5.</td>
<td>Coat’s Disease</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td>6.</td>
<td>Sympathetic ophthalmia</td>
<td>-</td>
<td>3</td>
</tr>
<tr>
<td>7.</td>
<td>Hemangioma</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>8.</td>
<td>Medulloepithelioma</td>
<td>-</td>
<td>3</td>
</tr>
<tr>
<td>9.</td>
<td>Silicone oil globules</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>10.</td>
<td>Hemorrhage/Trauma</td>
<td>27</td>
<td>31</td>
</tr>
<tr>
<td>11.</td>
<td>Intraocular tumors</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>i) Peripheral nerve sheath tumor</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>ii) Haematoma</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>iii) Leiomyoma</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td></td>
<td>iv) Squamous cell carcinoma</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td></td>
<td>v) Metastatic carcinoma</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

Table 5: Comparative table between our study and Vemuganti et al.

<table>
<thead>
<tr>
<th>S No.</th>
<th>No. of Intraocular tumors</th>
<th>Our study n=768</th>
<th>Vemuganti G K\textsuperscript{14} n=151</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Retinoblastoma</td>
<td>330 42.96%</td>
<td>48 31.78%</td>
</tr>
<tr>
<td>2</td>
<td>Malignant Melanoma of uveal tissue</td>
<td>85 11.06%</td>
<td>4 2.64%</td>
</tr>
</tbody>
</table>

Table 6: Indications for enucleation changed during the year

<table>
<thead>
<tr>
<th>S No</th>
<th>Indications for enucleation</th>
<th>2000</th>
<th>2001</th>
<th>2002</th>
<th>2003</th>
<th>2004</th>
<th>2005</th>
<th>2006</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Retinoblastoma</td>
<td>35</td>
<td>50</td>
<td>37</td>
<td>59</td>
<td>51</td>
<td>48</td>
<td>50</td>
<td>330</td>
</tr>
<tr>
<td>2</td>
<td>Malignant melanoma</td>
<td>12</td>
<td>14</td>
<td>11</td>
<td>11</td>
<td>17</td>
<td>10</td>
<td>10</td>
<td>85</td>
</tr>
<tr>
<td>3</td>
<td>Other causes</td>
<td>74</td>
<td>66</td>
<td>55</td>
<td>42</td>
<td>59</td>
<td>42</td>
<td>19</td>
<td>357</td>
</tr>
<tr>
<td></td>
<td>Total no. of cases</td>
<td>121</td>
<td>130</td>
<td>103</td>
<td>112</td>
<td>127</td>
<td>100</td>
<td>79</td>
<td>772</td>
</tr>
</tbody>
</table>
Table 7: Evisceration done during 7 years period (2000 - 2006); N = 305

<table>
<thead>
<tr>
<th>Sl. No.</th>
<th>Year</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>2000</td>
<td>20</td>
</tr>
<tr>
<td>2.</td>
<td>2001</td>
<td>21</td>
</tr>
<tr>
<td>3.</td>
<td>2002</td>
<td>32</td>
</tr>
<tr>
<td>4.</td>
<td>2003</td>
<td>43</td>
</tr>
<tr>
<td>5.</td>
<td>2004</td>
<td>29</td>
</tr>
<tr>
<td>6.</td>
<td>2005</td>
<td>46</td>
</tr>
<tr>
<td>7.</td>
<td>2006</td>
<td>114</td>
</tr>
</tbody>
</table>

Population based studies have estimated the incidence of enucleation to be 2.6-5.0 per 100,000 population.\(^2\,^3\) Indications of enucleation vary across different centers in the world and also denote a changing trend.\(^4\,^6\)

While some workers report an increase in the relative proportion of enucleation for tumors,\(^5\,^7\) others report a decline.\(^8\) Though no population based study has been reported from India, the indications for enucleation and evisceration have been studied in adults and children.\(^9\,^{11}\)

In the non-tumor group, anterior staphyloma was the most common indication for enucleation, contributing 0.65% of all cases. The numbers in our study compare with those reported from India (12-33%),\(^9\,^{11}\) Ethiopia\(^12\) and West Africa.\(^13\) The large number of this condition in our series could possibly be due to childhood trauma, infections and inflammations, which predispose the patient to staphyloma. It could also be due to other factors such as ignorance on the part of the patient, increased tolerance to glaucoma symptoms.

A history of trauma was seen in 1.17% cases, as compared to the findings of other series which reported 33-41% (Spraul et al.,\(^7\) Haile and Alemayehu.\(^12\)) The mode of injury included fire cracker injury, blunt injury, penetrating and gun shot injury. Evisceration was the procedure performed for infective etiology at our centre.

At the time of enucleation, the clinical diagnoses of most of the cases were confirmed except in few cases where clinical diagnosis was not available. There were 14 false positive diagnoses and 24 false negative diagnoses in our study.

False positive diagnosis was defined as a clinical diagnosis that is suggested malignancy when on enucleation there was no evidence. False negative diagnosis was defined as a clinical diagnosis which did not favour malignancy but at enucleation, malignancy was confirmed.

Very few studies are available in India regarding the histopathology findings of enucleated eyes. In the present study, the commonest indication for enucleation were malignant intraocular tumor like retinoblastoma or melanoma, ocular inflammation (1.69%), followed by staphyloma (0.65%), trauma (1.17%), phthisis bulbi (10.15%) and painful blind eye (16.79%). Other studies however showed some differences\(^4\,^5\) e.g. enucleation due to trauma was commonest (40%) in the series published by Tahri H et al.,\(^14\) Gassler and Lommatsch\(^15\) reported 46.4% intraocular tumors in enucleated specimens followed by glaucoma (31.6%), atrophic/Phthisis bulbi (11.9%), inflammation (6.5%) and trauma (0.9%).

Retinoblastoma was the commonest intraocular tumor (42.96%) in the present study followed by melanoma (11.06%). In contrast, Kitzmann\(^16\) reported 48.2% intraocular tumors, among which 82.5% were malignant melanomas. In India, Vemuganti et al.\(^17\) showed 49% intraocular tumors, among enucleated eyes of which 74% were retinoblastomas. The higher incidence of retinoblastoma in our study in comparison to the western countries might be due to the fact that retinoblastoma cases were detected earlier in western world and other modalities of treatment were offered before enucleation. The ratio of retinoblastoma to melanoma in our series was 3.8:1, as compared with 1:2\(^4\) for tumours of the oculo-adnexal region and 1:13\(^18\) in intraocular tumors. This ratio reflects the low incidence of melanoma in
the pigmented races and the relatively high incidence of retinoblastoma.19

The incidence of children requiring enucleation was high in our study which is contrast to the declining trends reported from the western world. This could be due to early diagnosis and availability of other forms of treatment like brachytherapy in the western countries. Timely intervention may negate the need for enucleation. It requires awareness and early diagnosis of the disease by primary eye care centers, pediatricians and general practitioners. Tumors were the commonest cause for enucleation in children.

To conclude, malignant intraocular tumors were the commonest indication of enucleation in our study. Prognosis of Retinoblastoma patients was determined by optic nerve invasion and tumor cells found at nerve resection margin. Most of the melanomas were of mixed cell type with a mixture of spindle and epithelioid cells while epithelioid cell melanomas were less common.

The survival rate of malignant melanoma patients depended on cell type; epithelioid cell variant has the worst prognosis. There was no significant correlation between year-wise enucleation rates of malignant intraocular tumors (Table 6).

In non-tumor group, staphyloma, ocular inflammation, phthisis bulbi, Coat's disease and sympathetic ophthalmia were the indications for enucleation.

CONCLUSION

We found that in children below 15 years, retinoblastoma was the most common intraocular tumor, whereas in adults, malignant melanoma of the choroid was the most common intraocular tumor. Painful blind eye was more common in adults than in children. During the period 2000-2006, the percentage of enucleation performed to remove intraocular tumors tremendously increased from 39% to 77%. At the same time, number of evisceration is significantly increased for trauma and phthisis bulbi. Our data suggest a decrease in number of enucleation of an eyeball from 15.7 % to 10% during the year 2000 to 2006, for non-tumor cases.

REFERENCES


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**COME, GIVE THE GIFT OF SIGHT**

Vol. XXVI No. 1 February 08 14
Chiasmal syndromes

G Balu, Shikha Bassi - Neurophthalmology Department

The chiasmal syndromes are distinct clinical entities characterized by visual field changes (usually bitemporal hemianopia), loss of visual acuity, diplopia, nystagmus, optic atrophy and oculomotor nerve palsies. It can be due to various causes both intrinsic and extrinsic to the optic chiasm. Pituitary tumors are the most common cause of chiasmal syndromes.

CLINICAL ANATOMY

The **Optic Chiasm** is quadrilateral overlying the sphenoid body. It is separated from it by a variable distance of 10 mm. It is embedded in the anterior wall of the third ventricle, between the two thalami and projects into the chiasmal cistern. Is obliquely situated continuing the inclination of optic nerves at an angle of 45°.

The Chiasm has a transverse diameter of 13 mm, a height of 4 mm and antero posterior width of 8 mm. It is in relation, *anteriorly* with anterior cerebral and anterior communicating arteries, *superiorly*, with the lamina terminalis, *inferiorly*, with the tuber cinereum and on *lateral sides*, with the anterior perforated substance. The optic chiasm is separated from structures beneath it by the basal cistern of subarachnoid.

Within the chiasm, the optic nerves undergo a partial decussation. The principal part of the chiasm consists of two sets of fibers, crossed and uncrossed. The **crossed fibers**, which are more numerous, occupy the central part of the chiasm, and pass from the optic nerve of one side to the optic tract of the other, decussating in the chiasm with similar fibers of the opposite optic nerve. The **uncrossed fibers** occupy the lateral part of the chiasm, and pass from the nerve of one side into the tract of the same side. The crossed fibers of the optic nerve tend to occupy the medial side of the nerve and the uncrossed fibers the lateral side. The crossing of the nasal half of macular fibers of central vision occurs posteriorly in the chiasm. The inferior and superior fibers remain inferior or superior, respectively. However, the inferonasal fibers pass more anteriorly in the chiasm, these fibers mingle with the optic nerve fibers still parallel to nerve axis forming interlacing basketwork the **Knee of Wilbrand**, while the superonasal fibers pass more posteriorly. In the posterior most part of the chiasm the macular axons and those from the central retina occupy most of the central part of the chiasm.

The relative position of the optic chiasm over the sella turcica is variable. In 79% of the cases it lies above the diaphragma sellae, (the dural covering of sella turcica). In 12% of the case the chiasm lies above the tuberculum sellae (the anterior elevation of the sella turcica) known as **prefixed chiasm**. And in 4% of the cases the chiasm lies above the dorum sellae known as **postfixed chiasm**.

CLASSIFICATION

1. **Anatomical classification by Lee**

Lee has divided optic chiasmal syndromes into anterior, middle and posterior. Anterior chiasmal syndrome affects the junction of the optic nerve and chiasm. Middle chiasmal syndrome relates to the decussating fibers in the body of the optic chiasm while posterior chiasmal syndrome involves the caudal fibers.

The classic anterior chiasmal lesion affects the optic nerve fibers and the contralateral inferonasal fibers located in Wilbrand’s knee. This will produce an ipsilateral optic neuropathy, often manifested as a central scotoma, and a defect involving the contralateral superotemporal field. This is also known as a **junctional scotoma** (fig. 1).
body of the chiasm most commonly disrupt the crossing nasal retinal fibers. This leads to a bitemporal hemianopia. The field of vision may still be full when both eyes are open but stereovision will not be possible. However, if fusion of the images is lost, perhaps due to a preexisting phoria, binocular diplopia may result.

Because macular fibers cross more posteriorly in the chiasm, they are damaged in posterior chiasmal syndrome. This leads to a smaller, paracentral bitemporal field loss. Because the temporal macular fibers haven't been damaged, it is possible to preserve color vision and visual acuity. Posterior lesions may also involve the optic tract and cause a contralateral homonymous hemianopia.

2. Etiological classification by Foroozan

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Traumatic</th>
<th>Iatrogenic</th>
<th>Intrinsic lesions</th>
<th>Extrinsic lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Achiromatism</td>
<td>Skull fracture</td>
<td>Radiation</td>
<td>Glioma</td>
<td>Pituitary adenoma</td>
</tr>
<tr>
<td>Albinism</td>
<td></td>
<td>Surgical</td>
<td>Demyelination</td>
<td>Craniopharyngioma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fat packing</td>
<td>Inflammation</td>
<td>Meningioma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Empty sella</td>
<td>Ganglioma</td>
<td>Aneurysm</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dopamine</td>
<td>Cavernoma</td>
<td>Mucocele</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Agonists</td>
<td>Histiocytosis</td>
<td>Hydrocephalus</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Arachnoid cyst</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Metastasis</td>
</tr>
</tbody>
</table>

- **Congenital**: The lesions include achiasmatism and albinism. MRI of patients with albinism shows optic chiasm of smaller size and they have misrouting of nerve fibers.6

- **Traumatic**: The mechanism of injury is due to direct tearing, contusion hemorrhage and necrosis or a combination of these. Most patients have bitemporal hemianopia, traumatic optic neuropathy and anosmia.7

- **Iatrogenic**: Most commonly occurs due to radiotherapy. MRI shows thickening and contrast enhancement. Chiasm can herniate into an empty sella after surgical intervention and is responsible for visual loss.8

- **Intrinsic lesions**: Gliomas are most commonly associated with neurofibromatosis I. Patients have slow and insidious visual loss, except in glioblastoma multiforme.9 Gliomas usually present with decreased central visual acuity and complex visual field abnormalities not limited to the crossing fibers. Demyelination can be the first sign on MRI in patients with multiple sclerosis. Sarcoidosis presents as diffuse infiltration of chiasm and patients have abnormal chest X-ray and elevated serum ACE levels.10

- **Extrinsic lesions**: Most common of the extrinsic lesions are of the pituitary etiology of which common are the micro adenomas. Pituitary tumors can present as
acromegaly, amenorrhea, galactorrhea, and infertility depending on the tumor etiology. In a case series 20% of acromegalic patients having visual symptoms are due to pituitary tumors. Visual field change is shown to improve within the first few days after transsphenoidal decompression. One study highlights that chiasmal syndrome develops in any pituitary lesion larger than 10 mm.

Clinical features such as younger age, visual field defect that is more inferiorly, unilateral disc pallor, RAPD is highly suggestive of an etiology other than pituitary neoplasm. Craniopharyngioma can present with intermittent visual symptoms due to periodic absorption of the cysts, patients are commonly misdiagnosed to have optic neuritis.

Suprasellar meningiomas can present with visual loss. Radiotherapy is advocated in presence of optic nerve involvement, and some patients may need surgical therapy. Long-term follow up has shown good visual prognosis.

Aneurysms can present with visual field loss. The internal carotid artery aneurysm can present with bi nasal visual loss due to compression of temporal fibers of both, the ipsilateral and contralateral side.

### CLINICAL FEATURES

Visual loss can be the earliest manifestation of chiasmal syndromes. The commonest visual field defect being bitemporal Hemianopia, (Fig-2) which respects the vertical meridian due to involvement of crossing of nasal fibers. Lesions posterior to the optic chiasm lead to Homonymous Hemianopia. Patient can also present with central visual loss due to involvement of macular fibers, which constitute the majority of the chiasm.

Loss of depth perception, loss of stereo acuity and post fixation blindness can lead to inability in performing fine tasks.

Diplopia can occur due to involvement of the cavernous sinus, third nerve being the commonest affected. Diplopia can also be non-paretic due to loss of overlap of corresponding temporal visual fields. Loss of hemi field contrast sensitivity has been shown to occur in chiasmal syndromes.

Chiasmal lesions can produce optic atrophy known as band atrophy due to damage of ganglion cells nasal to fovea and the optic disc with preservation of superior and inferior nerve fibers.

Nystagmus rarely can occur, described as see saw nystagmus.

Presenting features such as amenorrhea, galactorrhea, gigantism, loss of libido, infertility can also occur depending on the type of the secreting pituitary adenoma.

### INVESTIGATIONS

Visual field testing by Humphrey’s visual field analyzer using 30-2 program is the primary investigation. It is always important to test the visual field of the contra lateral eye in presence of an acute monocular visual loss.

Testing of color vision and contrast sensitivity shows hemifield contrast loss in early compression.

Titmus stereo test is used to detect the loss of stereopsis.

MRI is the gold standard for pituitary lesions. It demonstrates soft tissue delineation between pituitary and cavernous sinus. (Fig-3, 4). Intrinsc lesions enhance with intravenous contrast by use of gadolinium.
contrast.\textsuperscript{25} CT is better for delineation of calcification, bone erosion and bone remodeling.

Progressive deterioration of visual field is often the principle criteria on which surgical management decisions are made.

In an intrinsic lesion, finding of dural enhancement on MRI suggests inflammation and appropriate systemic investigations to be considered like PPD, Lyme titer, Chest X ray, serum ACE levels, RPR, HIV and CSF analysis. Treatment options include oral or intravenous steroids.

In patients with isolated chiasmatic thickening, a thorough medical history, the progression of visual loss, and evaluation of inflammatory and infiltrative causes should help in revealing the cause.

A finding of blood products in the optic chiasm on MRI suggests vascular malformations. Treatment options include laser resection.\textsuperscript{26}

Presence of white matter lesions is suggestive of multiple sclerosis. A finding of abnormalities on MRI can also be due to Neurofibromatosis or radiation necrosis.

**FOLLOW UP**

The patients of chiasmal syndromes are followed with serial visual field testing and MRI radiographs every half-year.

**REFERENCES**


Comparison of Freiburg Visual Acuity and Snellen Visual Acuity tests

Sheela Evangeline K, Ashwini M, Varadharajan J, Krishna Kumar R
— Elite School of Optometry, Unit of Medical Research Foundation, Chennai

INTRODUCTION

Testing visual acuity which is the resolving power of the eye is important to screen and treat people with visual impairment. Snellen chart is a simple chart that is sensitive to most common causes of visual impairment and is commonly used world-wide for visual acuity measurement.

A rapid and examiner-independent test of visual acuity was desirable and hence the FrACT was developed. The “Freiburg Visual Acuity Test” is an automated procedure for self-administered measurement of visual acuity. Landolt-C is presented on a monitor in one of eight orientations. The subject presses one of eight buttons, which are spatially arranged on a response box according to the eight possible positions of the Landolt-C’s gap. To estimate the acuity threshold, a best PEST procedure is employed, in which a psychometric function having a constant slope on a logarithmic acuity scale is assumed. Measurement terminates after a fixed number of trials.1

FrACT gave lower visual acuity when compared with Landolt ring chart2 and decimal progression chart.3 The FrACT coincided closely for visual acuity greater than or equal to 0.02 when compared with ETDRS.3

However there was no study done comparing FrACT and the commonly used SVAT.

A prospective observational pilot study was carried out to compare Freiburg Visual Acuity Test (FrACT) and Snellen Visual Acuity Test (SVAT).

METHODOLOGY

Thirty subjects with basic knowledge of English alphabets and of any age and sex who were the students and staff of Elite School of Optometry, irrespective of their refractive status and pathology were enrolled in the study. The eye tested and the test to be performed first was randomized by tossing a coin. Each subject’s visual acuity was measured using FrACT and SVAT by two different investigators, who were blinded of each other’s results.

The subjects were instructed not to tilt their head or squeeze their eyes and were allowed to guess in both the tests. The optotype used in FrACT was Landolt C and English alphabets were used in SVAT. The optotype for FrACT was presented in a 14inch CRT monitor. The SVAT was done in a normal clinical set up and the visual acuity was noted as Snellen fraction. The FrACT was done in a 5 meter room with dim background illumination. Care was taken so that there was no reflection on the monitor. The examiner pressed the corresponding button of the patient’s response (8 alternative forced choice method). The size of the next optotype was calculated using the best PEST procedure incorporated in the Freiburg software.1 Eighteen trials (standard- determines visual acuity as rapidly as possible without losing accuracy) were run to estimate the acuity threshold1. The visual acuity displayed on the monitor as Snellen fraction, decimal acuity and log MAR were noted. Paired t-test was used for analysis. A maximum difference of 0.05 log MAR which is tolerable as per DIN EN ISO 8597 was considered.2 The time taken for the completion of FrACT and SVAT was noted down using a watch.
RESULTS

The age of the subjects varied from 17 to 42 years with a mean of 23.67 years ± 7.6.

23 were females and 7 were males. The mean log MAR visual acuity was 0.007 ± 0.335 with SVAT and 0.1483 ± 0.409 with FrACT. The average visual acuity value found with FrACT was 0.14 larger than SVAT value. The difference between FrACT and SVAT obtained was 0.14 (intolerable by DIN EN ISO 8597 standards) and hence both tests were not comparable.

Analysis using the paired t-test showed a significant difference (p<0.0001) between the visual acuity measured by FrACT and SVAT. The average time to complete the test was 2.47 min for FrACT and 1.03 min for SVAT.

DISCUSSION

Our study showed that visual acuity estimated using FrACT was less than the visual acuity estimated using commonly used SVAT. This result was similar to previous studies. However previous studies did not document statistically significant variations in estimation. The difference could be attributed to the difference in optotypes (Landolt's ring in FrACT and English optotypes in SVAT) used in our study. Also the room illumination was not standardized while measuring the visual acuity tests.

Our study showed a higher average testing time per run for FrACT (2.47 min) compared to previous study done by Michael Bach (1.7 min). This difference can be attributed the variation difference in the sample subjects.

CONCLUSION

This study shows that FrACT was not rapid testing method of visual acuity and does not give an accurate self administered visual acuity when compared with SVAT.

Acknowledgement: We sincerely thank Dr L S Varadarajan for reviewing the article and giving his valuable comments.

REFERENCES

Muscle Puzzle
(Answer to Muzzle Puzzle on page 8)

This patient has infantile esotropia with Bilateral DVD with a Right head tilt.

**Dissociated Vertical Deviation**

Most commonly known as DVD it is among the most intriguing and least understood of all forms of strabismus. Though it has unique clinical features, the diagnosis may be difficult when associated with other forms of strabismus especially with cyclovertical deviations. Although, Bielschowsky credited others for the first reports of this entity, it was he who first provided the first comprehensive description and minute clinical analysis of DVD. The lack of precise etiologic information about DVD is reflected by the plethora of terms in use at one time or another: anatopia, alternating hyperphoria or hypertropia, double hypertropia, occlusion hypertropia, alternating sursum-duction, dissociated double hypertropia, dissociated alternating hyperphoria, and dissociated vertical divergence.

DVD is characterized by the spontaneous drifting of either eye upward when the patient is fatigued or daydreaming or when fusion is artificially interrupted by covering one eye. The amount of elevation when the eye is covered is variable, tending to increase after prolonged occlusion, and often differing between two eyes. Latent nystagmus occurs in approximately half the patients with DVD and, in fact, is seldom encountered in the absence of this anomaly. Anomalous head postures in the form of head tilt away from the eye with larger vertical deviation is reported in the range between 23% and 35% of the patients. DVD is usually bilateral phenomenon, often asymmetric, and commonly associated with congenital esotropia and other entities that disrupt binocular vision early in life. It can be latent or manifest, comitant or incomitant. Comitant form is more common. DVD occurs in patients with and without overaction of inferior oblique muscles and may also be associated with overaction of superior oblique muscles and an A pattern exodeviation in downward gaze. Bielschowsky phenomenon can be elicited by putting photometric neutral filters of increasing intensity in front of the fixating eye. Variants of DVD in the form of dissociated horizontal deviation and dissociated torsional deviation can also be seen in some patients.

Various theories have been proposed regarding the etiology of this condition like maldeveloped supranuclear centers, hypofunctioning superior oblique, interruption of neuromuscular transmission of superior oblique muscle, compensatory mechanism to dampen cyclovertical component of latent nystagmus but none has been substantially proven. The role of visual stimulation relative to other eye in the manifestation of DVD is however, proven beyond doubt. Spielmann has shown that DVD does not occur (or gets neutralized) when both eyes are covered by translucent occluders. Imbalance of binocular stimulation is thus important but it is known to occur in individuals with normal binocular functions, which needs explanation. Bielschowsky’s original explanation of this form of strabismus, confirmed by modern eye movement recording techniques, has established indisputably that DVD is a vertical vergence eye movement. However, the stimulus for this movement and its relationship to various forms of strabismus, especially to essential infantile esotropia, have yet to be convincingly identified.

Patients with DVD are usually asymptomatic. Indications for treatment are mostly cosmetic, presence of anomalous head posture; manifest DVD in-patient with peripheral fusion. Although in most patients with conspicuous DVD, surgery is the recommended treatment, the possible
effectiveness of a conservative approach should not be ignored, which includes optical blur of fixating eye to switch fixation preference and spectacle correction to strengthen fusion. Surgical procedures preferred by various authors are recession of the superior combined with resection of the inferior rectus muscles, resection of the inferior recti, retroequatorial myopexy (posterior fixation) of the superior recti combined with or without a recession of these muscles, unconventionally large recessions (7 to 10 mm) of the superior recti, and anterior displacement of the inferior oblique insertion, which may be combined with superior rectus recession. Recently many surgeons have proven effectiveness of anterior transposition of the Inferior oblique muscle. Inferior oblique muscle is inserted just anterior to or at the level of insertion of inferior rectus muscle.

In our case the elevation in adduction was due to disruption of fusion due to presence of nose as an obstacle, which resulted in manifestation of DVD. This was recognized and bimedial recession with differential bilateral Superior rectus recession was done. This resulted in correction of head tilt and correction of DVD.

Reference
TECHNOLOGY UPDATE:

Microperimeter

Munuver Gupta Nittala — Sankara Nethralaya Diabetic Retinopathy Project

The Microperimeter combines fundus-tracking microperimetry with color fundus photography in a single instrument. It was developed by a group of European SLO users who wanted a richer feature set as well as color photography.

The end product of perimetry and microperimetry exams is a sensitivity map of the examined retina. This is obtained by measuring patient’s ability or inability to perceive light of varying intensities projected on different areas of the retina. In conventional perimetry the stimulated fundus areas are identified by their geometric position with respect to the patient fixation area. The sensitivity map is generated by observing a live picture of the examined retina and allows therefore referring stimuli location to precise anatomical references. Microperimetry is then correlated with the fundus of the patient, reason for which it is also called fundus related microperimetry.

The main examinations that can be performed with Microperimeter MP-1 are:

- **Fixation exam**: Allows determining the site of fixation and evaluate its stability in time.

- **Microperimetry**: Allows performing perimetry exams by projecting a stable fixation target and a sequence of Goldman type stimuli. Patient answers at each stimulus (seen/not seen) are recorded and stimuli are displayed on the same, fixed, retinal picture at the beginning of the exam.

- **Non-mydriatic fundus photography**: Allows acquiring fundus images at 45° without use of any mydriatic agents.

**INDICATIONS FOR MICROPERIMETER**

- **Advanced age related macular degeneration (atrophic and neovascular AMD)**
  Detecting the location and fixation (foveal and extra foveal), quantification of scotoma characteristics, quantifying longitudinally the functional impact of any treatment (medical, laser or surgical) at specified retinal location.

- **Early AMD**
  Evaluate the functional deterioration of discrete macular lesions along with the natural history or following treatment.

- **Diabetic macular edema**
  Evaluating the functional impact of different degrees of macular edema, comparison of functional values with OCT data and evaluating the effect of different laser treatment modalities on macular function.

- **Vitreo retinal interface disorder**
  Prognostic value of microperimetric data vs. Vitreo retinal surgery.

- **Low vision patients**
  Quantification of fixation, location and stability, planning of visual rehabilitation program and evaluation of results.

- **Macular hole/ macular cyst**
  Quantification of central scotoma and evaluation of central fixation.

- **Hereditary retinal dystrophies**
  Quantification of retinal sensitivity in hereditary retinal dystrophies like Retinitis Pigmentosa, Stargart’s disease, Best’s disease etc.
METHODOLOGY

Microperimetry can be performed in undilated pupil under dim illumination. Explain the test procedure to the subject and assure the subject that it’s non-invasive and that it takes less time to finish. Answer any of the queries the subject may have about the procedure. The Microperimeter initially takes an infrared photograph (1392-1038 pixel resolution, 45° field of view). The software package allows the operator to select a biological landmark of high reflective under infrared i.e. the branch of a retinal vessel. This image is then digitally registered and matched with the corresponding area on the real-time fundus image of the patient. All stimuli are projected directly onto the retina in relation to this landmark, using a liquid crystal diode. The operator views the retinal image projected on the monitor, with the stimulus as part of the image. Background illumination is set at 4 apostilbs (asb) (1.27 cd/m²; 1 asb = 0.31831 cd/m²). Stimulus intensity may be varied on 1 (0.1 log) step scale from 0 to 20 dB, where 0 dB represents the brightest luminance of 400 asb (127 cd/m²). Stimulus size may be varied by the examiner from the Goldmann I to V standard size. The fixation target set at 100 asb, may be varied in size and shape according to the patient’s visual acuity and/or macular scotoma. Adjustments for eye movements are made at 25 times per second. This active tracking allows the Microperimeter to get reliable perimetry data even when the patient is unable to fixate. If the Microperimeter loses tracking, automatically it will stop projecting stimuli until active tracking is reestablished. A false positive test stimulus is projected every 60 seconds onto the optic nerve head area to check for a false positive answer. When the exam is complete, color photography is taken (1392 X 1038 pixels, xenon flash) and a similar registration technique is used to correlate the visual field data over the fundus photo. This makes it easy to correlate the pathology with scotoma. The microperimeter allows the reporting to be numerically in decibels, schematically or a color scheme. This same technology allows actively tracking and mapping the patient’s fixation. There is also a separate fixation exam available that requires less than one minute to perform.
ANALYSIS OF RESULTS

Microperimeter allows the reporting the results to be numerically in decibels, schematically or in a color scheme and it also calculates the mean retinal sensitivity in particular retinal location as selected by the operator. The stimuli intensity is well represented by an intuitive color scale. A red color stimulus represents a high intensity stimulus corresponding to a low sensitivity instead of a green color stimulus that represents a low intensity stimulus corresponding to a high sensitivity. A seen stimulus is represented by a filled square, while a not seen one is represented by an empty square.

CASE ILLUSTRATION

This is a case of 65-year-old patient complaining of gradually decreasing vision for distance in left eye for last one year. He was previously diagnosed to have choroidal neovascularization in the left eye and underwent intravitreal gas \( \text{C}_{2}\text{F}_{6} \) and Avastin injection 2 months back. On examination his BCVA for distance was 20/20 with refractive correction of \(-1.00/-0.50\times90^\circ\) in right eye and 20/26 with refractive correction of \(-1.50\times90^\circ\) in left eye. With the addition of +3.00DS was able to read binocularly N6. Slit lamp examination showed early posterior sub capsular cataract in both eyes and intraocular pressure was 18 mm of Hg in both the eyes. Dilated fundus examination with indirect biomicroscopy showed few retinal pigment epithelial alterations in macular area of the right eye, the left eye showed mostly scarred choroidal neovascular membrane surrounded by retinal pigment epithelial defect and cystoid macular edema. He was advised for optical coherence tomography and microperimetry. SDOCT showed loss of photoreceptor layer superonasal to the macula in right eye and scarred choroidal neovascular membrane with cystoid macular edema in the left eye. Patient underwent Microperimetry (MP1, Nidek, Italy) for both the eyes and the mean retinal sensitivity was 8.9 dB in the right eye and 10.4 dB in the left eye. Microperimeter showed a dense scotoma in the area where SDOCT showed loss of photoreceptor layer in right eye and dense scotoma in infero-temporal foveal area where pigment epithelial detachment (PED) was noted in SDOCT at macular area (fig-1)

BIBLIOGRAPHY:


Fig 1: Left SDOCT image showing area of photoreceptor layer loss.
Right microperimeter image showing dense scotoma area where photoreceptor layer loss seen.