EXTENDED REPORT

Test characteristics of orthoptic screening examination in 3 year old kindergarten children

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Aim: To analyse the test characteristics of orthoptic screening for amblyopia or amblyogenic risk factors (target conditions) in kindergarten.

Methods: 1180 three year old children were screened by orthoptists in 121 German kindergartens. Orthoptic screening consisted of cover tests, examination of eye motility and head posture, and monocular visual acuity testing with the Lea single optotype test. Children were re-examined in kindergarten by different orthoptists after 3-6 months using a more demanding pass threshold for visual acuity. All children with at least one positive orthoptic test result or an inconclusive re-examination were referred to an ophthalmologist for diagnosis. The gold standard was set positive if a target condition was diagnosed on ophthalmological examination. It was set negative if no target condition was found upon ophthalmological examination, or if a child who screened negative or inconclusive passed the orthoptic re-examination without any positive test result.

Results: The gold standard was ascertained in 1114 children. 26 (2.3%) children had a "positive" gold standard. In 10.8% of the children the initial screening was "inconclusive," mostly due to lack of collaboration. Screening test sensitivity (based on conclusive results only) was 90.9% and specificity was 93.8%.

Conclusions: Orthoptic vision screening of 3 year olds in kindergarten is sensitive and specific. However, owing to a substantial proportion of inconclusive screening results, rescreening of non-cooperative 3 year old children should be considered.

The main purpose of preschool vision screening is to detect amblyopia so early that treatment is likely to be still effective.1,2,3 The fourth year of life is considered best for vision screening as from this age onwards in most children monocular visual acuity can reliably be assessed by simple screening methods.4-6 In Germany, vision assessment is part of the general preventive care examinations conducted by general practitioners and paediatricians, and is paid for by the statutory health insurance. The effectiveness of this programme with respect to the detection of amblyopia is considered insufficient,7,8 since general practitioners and paediatricians lack experience with ophthalmological tests.9 Screening by orthoptists has been studied as an option to improve the sensitivity and specificity of preschool vision screening,10-14 because orthoptists are familiar with visual acuity and cover testing in children. Because in Germany about 50% of children enter kindergarten at age 3, and more that 80% attend kindergarten at age 4,15 kindergarten offers easy access to large numbers of children without efforts from the parents. Vision screening has been conducted successfully in German kindergartens before.16-19 Yet, little is known about the test characteristics of orthoptic vision screening performed in German kindergartens. The purpose of this study was to determine the test characteristics of orthoptic vision screening for untreated amblyopia or amblyogenic risk factors performed on 3 year olds in kindergarten prospectively. Therefore, a community based study was conducted which aimed at recruiting a representative sample of 3 year old children to be screened in real world kindergarten settings. The evaluation of screening tests performed in a community are prone to be affected by examination, or verification, bias.20,21 This bias is introduced if the diagnosis for patients with different screening test results is not equally likely to be confirmed by a gold standard evaluation. Verification bias has been shown to be frequent in paediatric studies.22 In particular, if the gold standard evaluation inconveniences the parents, or if it is associated with possible side effects, children who screened positive will be more likely to have a gold standard evaluation than children who screened negative. This bias results in sensitivity being erroneously inflated, and specificity being falsely reduced.23 Thus, the design of this study aimed at avoiding verification bias by using a practicable gold standard evaluation which was likely to be obtained for all children in the study.

METHODS

The institutional review board approved the study design which followed the tenets of the Declaration of Helsinki.

Study population

In order to recruit a representative study sample, local and regional kindergartens in two counties of south west Germany (Tübingen and Reutlingen) were contacted systematically. The management and parents of 121 kindergartens opted for participation in the study. All 3 year old children (after the third, before the fourth birthday) attending these kindergartens were eligible. Their parents or legal guardians were asked for informed consent in writing. The exact number of eligible children could not be ascertained owing to personal data protection legislation which required that access to the parents had to be obtained through the kindergarten staff. However, the participating kindergarten management and staff welcomed vision screening and were highly committed to recruit all eligible children. According to the kindergarten staff, nearly all parents of eligible children agreed to let their child(ren) participate. To the best of our knowledge, only one mother of a strabismic child who was already being treated refused to let her child participate, and consent could not be obtained from a few parents who did not speak and/or write German well enough to complete the consent form, despite the help offered by the kindergarten staff. The resulting dropouts may be estimated at a maximum of 10 children.
Therefore, we concluded that nearly all eligible children were enrolled and no significant bias resulted.

A succinct history of each child (known ocular diseases, wearing of glasses, current ophthalmological treatment) was collected from the parents on the consent form. Children were recruited regardless of their history; 1184 children recruited for the study received an orthoptic screening in kindergarten between July and December 1999 (phase I). Less than 10% of the children who were enrolled could not be examined on the scheduled day owing to absence (illness, vacation, or other). In four children examined, the birth date was not ascertained until February 2001, and verification was impossible because the families moved. These four children (all without pathology in the orthoptic examinations) were excluded from this analysis, resulting in a sample size of 1180. The mean age was 42.7 months, 50.6% were male.

Children already treated for amblyopia
Among the 1180 children examined and included, there were 21 (21/1, 180 = 1.8%, 95% CI 1.0% to 2.5%) who had already been treated for amblyopia, of whom 11 had large angle strabismic amblyopia and 10 were of refractive/anisometropic origin. This was verified by comparing the ophthalmological records with the information provided by the parents. For these children the gold standard which is explained in the following paragraphs was rated “negative,” since they were already treated. They were excluded from the sample for the calculation of the specificity of screening in order to avoid a bias towards false “positives.”

STUDY DESIGN
Orthoptic screening
Orthoptic screening was performed by five experienced orthoptists and consisted of four items. Results could be rated “positive” (any pathology), “negative” (within normal limits), or “inconclusive” (insufficient cooperation or unclear, borderline result) with the following criteria:

- inspection of the anterior eye segment, “positive”: any potentially vision threatening macroscopic organic anomaly other than ocular misalignment or eye motility disorder
- unilateral and alternate cover, and uncover tests at near and at distance, “positive”: manifest strabismus or unstable re-fusion upon uncovering
- examination of eye motility and head posture, “positive”: any detectable anomaly
- uncorrected monocular visual acuity testing with the Lea single optotype test at 3 metres/10 feet (single symbol book No 2506, Precision Vision, Villa Park, IL, USA) and an eye patch, “positive”: visual acuity £0.4 (10/25), OR if line difference >1 line and visual acuity in the worse eye equal 0.5 (10/20) to 0.63 (10/17) (L Hyvärinen, Lea Test Ltd, Helsinki, Finland, personal communication, 10 October 1998).21

The starting visual acuity was 0.4 (10/25). To pass a line, three out of four symbols (3/4) had to be identified correctly. If the child hesitated or seemingly guessed without concentrating on the symbols, or identified only two symbols correctly—for example, because it was unfamiliar with forced choice testing, it was shown another symbol except for the circle/“ball.” Then three correct symbols out of five (3/5) were accepted to pass the line with the final visual acuity of that eye.

For completeness, we mention that corrected visual acuity was also measured if children had brought their glasses and were cooperative enough to be tested a second time after uncorrected visual acuity testing. However, since all children were to be examined under the same conditions, and regardless of history, the measurement of corrected visual acuity was not used for study purposes but rather to be able to inform parents of their children’s corrected visual acuity—that is, for ethical reasons.

To save time, visual acuity testing of an eye was discontinued when 1.0 (10/10) was reached. Otherwise, visual acuity testing criteria followed the recommendations of the manufacturers’ user instructions.

Visual acuity testing would be performed under varying indoor lighting conditions because of season and weather. Therefore, the orthoptists were instructed to adjust the shades, and to seat the children in a way to avoid direct sunlight and high contrasts if there was bright sunlight; if the light level was low, a portable glass shielded 150 W halogen reflector lamp was used to light up the place from which the visual acuity test was shown. In 26 kindergartens, the ambient light level was monitored with a radiometer (Universal Photometer, Hagner, Solna, Sweden). It was ascertained that this procedure entailed photopic light levels (all measurements above 10 cd/m², average 132 cd/m²).

In order to improve cooperation, the staff of the kindergarten were asked to train the children for the visual acuity testing, with photocopies of the Lea symbols. Parents were asked not to be present in kindergarten during the examination sessions. In the following text, “positive” and “negative” orthoptic examination results were also labelled “conclusive,” as opposed to “inconclusive” results.

Screening outcome
Orthoptic examinations were rated “referral,” if any screening item was “positive”; “no referral” if all screening items were within normal limits—that is, “negative”; or “inconclusive” if cooperation was insufficient for at least one screening item, or if the result was ambiguous.

Gold standard
In this community based study, a mandatory ophthalmological gold standard examination with cycloplegia would most likely not have been acceptable to a considerable proportion of parents. These parents would either not let their child participate in the study at all, resulting in a non-representative study sample, or they would tend not to comply with the ophthalmological examination if the child screened negative, resulting in verification bias as described earlier. In order to avoid these types of selection bias, a practicable gold standard was defined which was likely to be obtained for all children in the study population.

Orthoptic examination
As part of the gold standard procedure, the study population was re-examined in kindergarten after 3–6 months by a different orthoptist (phase II) with a more demanding threshold for uncorrected monocular visual acuity of >0.63 (10/17) in either eye to pass the examination (that is, to be rated “no referral”). Children who only reached 0.5 (10/20) or 0.63 (10/17) in either eye were classified “borderline” which entailed referral for gold standard purposes. The other outcomes of the orthoptic examination were the same as in the screening phase I. The orthoptist in phase II was masked to the results in phase I. However, the orthoptist in phase II had the same history information as in phase I.

If the orthoptic screening (phase I) resulted in “no referral” or “inconclusive,” and the orthoptic examination (phase II) resulted in “no referral,” then the gold standard was set “negative.”

Figure 1 summarises how the gold standard was established starting from the results of the orthoptic screening.

Ophthalmological examination
If any orthoptic screening item in phase I was “positive,” the child was referred for a full ophthalmologic examination;
Figure 1  Flow diagram of gold standard determination. A child could be classified gold standard “negative” in several ways: the result was “negative” or “inconclusive” in the orthoptic screening and “negative” in the orthoptic examination; children who had a visual acuity of >0.63 (10/17) in either eye or better and no anomalies in the orthoptic examination were classified gold standard “negative” without further examination; children with “positive” or “inconclusive” or “borderline” results in the orthoptic examination were referred. Children with a “positive” screening were all referred. To be classified gold standard “positive,” a child had to be referred and had to have target conditions upon ophthalmological examination.

likewise, if upon the orthoptic examination (phase II) collaboration was insufficient so that findings were classified “inconclusive” or if the child was absent, the child was also referred to an ophthalmologist. Application of this procedure meant that any visual anomaly detected in kindergarten had to be confirmed by an ophthalmological examination. If the result of an ophthalmological examination was “positive,” then the gold standard was set “positive;” or if the result was “negative;” then the gold standard was set “negative.” Parents were given the choice to have their child examined in the outpatient clinic of the strabology department of the University Eye Hospital, Tübingen, or in the office of an ophthalmologist in their vicinity to avoid verification bias. About half of the ophthalmological examinations were provided by the strabology department, and the other half by office based ophthalmologists.

To ascertain that parents complied, rigorous follow up contacting, by mail and telephone, was performed systematically and, if necessary, repeatedly within a year after the orthoptic examination of phase II. In addition, ophthalmological examination reports from all children who were currently seen by an ophthalmologist (irrespective of this study) were collected. A standardised examination report was requested from the ophthalmologist which contained corrected and uncorrected visual acuity, cycloplegic refraction, ocular motility and ocular alignment assessed by cover testing, stereopsis, fundus examination and retinal fixation behaviour, as well as information on treatment, if started.

Criteria for “positive” gold standard

The gold standard classification of the ophthalmological examination records was done by the study team. The study protocol criteria for a “positive” gold standard were:

- main criterion: any newly administered spectacle therapy if the corrected visual acuity was ≤0.4 (20/50) in either eye, OR difference of visual acuity between right and left eye >2 logarithmic lines, except for myopia; or
- secondary criterion, in the event that the main criterion might not be applicable due to lack of cooperation in the visual acuity testing: any newly administered patching therapy in the presence of risk factors like monolateral strabismus or high refractive error (cycloplegic spherical equivalent difference of ≥1.5D, or ≥3D of astigmatism).

It has to be pointed out that the screening criterion for visual acuity line difference was just >1 logarithmic line. This option was chosen to achieve a high sensitivity of screening (that is, not to miss subthreshold borderline cases) and to avoid a definition of target conditions which would include suprathreshold borderline cases.

In cases of spectacle prescription in which visual acuities were above the thresholds defined above, the gold standard was set “negative.”

The study design aimed at performing a community study compatible with current, not necessarily standardised, diagnostic and treatment practice patterns, at bringing about a high participation rate, and avoid verification bias. Therefore no uniform guidelines were imposed upon ophthalmologists. This helped to obtain the participation of all ophthalmologists in the region (see list of more than 30 participating ophthalmologists in the acknowledgements). Parents were not dissuaded from participation in the study because they did not have to present their child to an ophthalmologist who would be complying with guidelines, while their family ophthalmologist might not.

Validity of the gold standard

The gold standard used in this study required all children who screened positive to have a full ophthalmological examination—that is, a “classic” gold standard evaluation, which by definition is 100% accurate, with sensitivity and specificity both being 100%. Hence all children who screened positive were correctly classified; thus no incorrectly “positive” gold standard result could occur, and consequently the specificity of the gold standard was 100%. However, the gold standard did not require all children who screened “negative” to have a full ophthalmological examination. Although a more demanding pass threshold was used in the orthoptic examination in phase II to make incorrectly “negative” gold standard results unlikely, some of those who screened “negative” may have been incorrectly classified as having a “negative” gold standard. Hence, the estimate of sensitivity may be biased. Yet, this bias is likely to be small, as shown in the following paragraph.

Assuming that the orthoptic screening and the orthoptic examination performed by different orthoptists were independent from each other, the accuracy of the gold standard used in this study can be assessed using conditional probability calculation. Through combining the sensitivities of the orthoptic examinations and those ophthalmological examinations, which were conducted because of “inconclusive” or missing orthoptic examinations in phase II, an overall sensitivity of the gold standard can be computed. Figure 2 shows an example of the calculation of the overall sensitivity of the gold standard used in this study. In this example, the true sensitivities of the orthoptic screening (S1) and the
orthoptic examination (S2) were set at 90.0% each, while the number of examinations corresponds to those conducted in the study. The resulting overall sensitivity of the gold standard would be 98.4%, which means that of 100 children diagnosed “positive” through ophthalmological examinations, 1.6 would be misclassified by the gold standard used in this study. With respect to the observed sensitivity of the orthoptic screening, which was calculated based on “conclusive” results of phase I only, the bias would even be smaller. In the example shown in Figure 2, the sensitivity of the gold standard in children with “conclusive” results in phase I would be 99.2%. Hence, if the true sensitivity of the orthoptic screening was 90% as assumed in this example, it would be estimated at 90.0%/99.1% = 90.7%—that is, overestimated by only 0.8% percentage points.

Table 1 lists other combinations of sensitivities of orthoptic screening and orthoptic examination. Even for unrealistically low sensitivities of orthoptic examinations, the results were still acceptable to achieve a valid gold standard.

Owing to gold standard results incorrectly classified “negative,” the specificity of the orthoptic examination may also be overestimated; however, the magnitude of overestimation would even be smaller than for sensitivity.

RESULTS
Cooperation and attendance in the study population
In phase I, 1047 (88.7%) of all 1180 children showed sufficient cooperation with the orthoptic examination, providing a “conclusive” result (“referral” or “no referral”); 133 (11.3%) children had an “inconclusive” result.

In phase II, 194 (16.4%) of the children examined in phase I were not present in kindergarten on the days of the orthoptic examination, owing to illness, vacation, etc, 957 (97.1%) of the 986 children present were sufficiently cooperative.

Gold standard results
For 1114 children (94.4%), including the 21 children already treated before screening, the gold standard was ascertained as of February 2001. Of those 66 children for whom no gold standard result could be obtained, all had “inconclusive” orthoptic examination results, almost all of them because they did not attend the orthoptic examination in phase II, having mostly “negative,” and to a lesser extent, “inconclusive” findings in the orthoptic screening.

Forty two children were examined by an ophthalmologist although for them the gold standard was already “negative” based on both orthoptic examinations. Some of these children happened to have an ophthalmological examination some time before the kindergarten screening and a few children were presented to an ophthalmologist after the first or second screening because the parents became sensitised. In none of these 42 children was a target disease detected.

“Positive” gold standard results
In 26 children the gold standard was “positive” (26/1, 114=2.3%, 95% CI 1.4% to 3.2%). Of these, three had small angle or accommodative strabismic amblyopia; the rest had refractive/anisometropic amblyopia. Patching and spectacle...
Table 2  Ophthalmological examination findings and orthoptic vision screening results of gold standard “positive” children; all children received spectacle treatment. Refractive errors are those measured under cycloplegia. Subjects 1222, 1221, and 1855 had moderate risk factors, but they nevertheless failed the more important visual acuity testing performed in offices of participating ophthalmologists.

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<tr>
<td>2124</td>
<td>4</td>
<td>-0.5</td>
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<td>1</td>
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<td></td>
<td>yes</td>
<td>“positive”</td>
<td></td>
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</tr>
</tbody>
</table>
Table 3 Results of orthoptic screening (phase I) by test items and cumulated, in 1093 children for whom the gold standard was obtained and who were not treated for amblyopia or amblyogenic risk factors before screening. Inconclusive results of orthoptic screening items were excluded to compute the proportion of gold standard “positive” cases which were detected by each item, and the resulting per item sensitivity and specificity.

<table>
<thead>
<tr>
<th>Screening item</th>
<th>Inconclusive results</th>
<th>Positive results</th>
<th>Negative results</th>
<th>No of conclusive results</th>
<th>Proportion of detected gold standard “positive” cases*</th>
<th>Sensitivity</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No</td>
<td>%</td>
<td>No</td>
<td>%</td>
<td>No</td>
<td>%</td>
<td>No</td>
</tr>
<tr>
<td>Inspection</td>
<td>0</td>
<td>0.0</td>
<td>2</td>
<td>0.2</td>
<td>1091</td>
<td>99.8</td>
<td>1093</td>
</tr>
<tr>
<td>Ocular motility/eyephase posture</td>
<td>47</td>
<td>4.3</td>
<td>12</td>
<td>1.1</td>
<td>1034</td>
<td>94.6</td>
<td>1046</td>
</tr>
<tr>
<td>Cover testing</td>
<td>51</td>
<td>4.7</td>
<td>13</td>
<td>1.2</td>
<td>1029</td>
<td>94.1</td>
<td>1042</td>
</tr>
<tr>
<td>Visual acuity</td>
<td>118</td>
<td>10.8</td>
<td>63</td>
<td>5.8</td>
<td>912</td>
<td>83.4</td>
<td>975</td>
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<tr>
<td>Cumulated</td>
<td>118</td>
<td>10.8</td>
<td>79</td>
<td>7.2</td>
<td>896</td>
<td>82.0</td>
<td>975</td>
</tr>
</tbody>
</table>

*Proportion of detected cases with positive gold standard among all cases with positive gold standard in children with conclusive result for the screening item (equal to sensitivity).

Table 3 illustrates the relative importance of the screening items when “inconclusive” results were excluded to compute the test characteristics of orthoptic vision screening in 3 year old kindergarten children. The most sensitive single test item was visual acuity testing. Two children screened “positive” upon inspection: one child with a “red eye” who was gold standard “negative”, and another child with bilateral ptosis (subject ID 1869, see Table 2) who was gold standard “positive” because of strabismic amblyopia. Therefore, inspection was little helpful for screening in this sample.

DISCUSSION

Study population

Only 3 year olds were included, because these were the youngest children who could be examined in kindergarten in Germany. However, the results are likely to be more favourable in 4 year olds, since these are in general examined more easily. This means that children who enter kindergarten later, and escape screening at age 3, could be screened at age 4.

Two cases of strabismic amblyopia were newly detected in children whose parents erroneously indicated current ophthalmological treatment of their child on the consent form. Therefore, inclusion of children in the screening programme should not be based on the history provided by the parents.

The results of this study suggest that at age 3 most strabismic amblyopias with visible angular deviation have already been detected and that the remaining target conditions are mostly due to refractive errors. Since visual acuity testing—the most important single screening item in detecting refractive errors—depends more on cooperation and age than cover testing does, orthoptic screening in populations with a greater proportion of strabismic amblyopias may be even more sensitive and specific, with less “inconclusive” results.

Choice of screening items

A single optotype test, and without crowding bars, was used because this would make testing of 3 year olds easier: this was backed by the finding in a study which showed that the Lea single optotype test was nearly as sensitive to detect amblyopia as a line test. Similar findings were obtained when using the Sheridan-Gardiner single optotype test of visual acuity. The use of a more difficult test and a higher threshold may have increased the proportion of “inconclusive” or false positive results, without raising the sensitivity. Children with “positive” gold standard who were not detected through screening (phase I) had bilateral balanced refractive errors (see Table 2), and probably a lower risk of severe amblyopia than the unilateral visual deficits of which none escaped the screening.

While there is agreement that line tests exhibit a better sensitivity for amblyopia than single optotype tests, and that crowded optotypes could be even more sensitive when tested...
in cooperative subjects, screening with such tests in 3 year old children may not necessarily be more advantageous, since their testability and specificity may be inferior. For instance, a study of the testability of Lea symbols (line chart) versus HOTV line charts found that in three year old children the testability was better for Lea symbols (92%) than for HOTV charts (85%). The authors concluded that Lea symbols should be preferred over HOTV and tumbling E tests. Yet, in the cited study, two screeners were used for each chart. In the study presented here, using the Lea single symbols book administered by a single screener recruited from outside laboratories and universities, the percentage of untestable 3 year old study presented here, using the Lea single symbols book the cited study, two screeners were used for each chart. In the study presented here, using the Lea single symbols book administered by a single screener recruited from outside laboratories and universities, the percentage of untestable 3 year old children was 10.8% (95% CI 9% to 13%) compared to 8% (95% CI 4% to 12%) using Lea line charts in the above cited study, which leaves the results fairly comparable at the testability level. However, the intervention of two trained research screeners to perform visual acuity testing would not seem to be a realistic option to conduct screening cost effectively in a real world setting. Another study reported success (testability) rates of 76% (3 year olds) and 95% (4 year olds) with Lea line symbols.

In the light of the present study's results these data rather point to a lower testability of line symbol tests in 3 year olds.

The rationale of the present study was to foster feasibility at all levels. Therefore, a most simple, fast and easy to use test, the Lea single symbol book, was preferred over the corresponding line or crowded test.

Gold standard

The study protocol was tailored to assess the test properties of orthoptic kindergarten screening in German kindergartens. A practicable gold standard was used, which did not require all children to undergo a full ophthalmological examination. This made it possible to recruit almost all eligible children in the participation kindergartens. The gold standard was ascertained in more than 94% of the study population, thus avoiding verification bias. The effectiveness of screening was based upon practical treatment decisions. The study design helped avoid laboratory conditions which could not necessarily be reproduced in reality.

The validity of the gold standard in children without pathological findings in the orthoptic examinations may be questioned because an ophthalmological examination was not mandatory to rule out the presence of target conditions. This might entail a reduced sensitivity of the gold standard, and an overestimation of the sensibility and specificity of orthoptic screening. However, this error, if it exists, would be small, as demonstrated by the decision analytic model.

Prevalence of amblyopia and amblyogenic risk factors compared to other studies. In a recent review the prevalence of amblyopia (estimated from the yield of screening programs) was found to be between 2.7% and 4.4%. The prevalence of amblyopia or amblyogenic risk factors in the present sample was 47/1114 = 4.2% (95% CI 2.1% to 5.4%) which fits into the cited bracket, and would suggest that the sample prevalence was at the upper end of established prevalence limits.

CONCLUSIONS

In this study, orthoptic screening performed in kindergarten was sensitive and specific for detecting amblyopia and amblyogenic risk factors in 3 year old children. However, in approximately 11% of 3 year olds no “conclusive” screening results were obtained because of insufficient cooperation. To increase effectiveness “inconclusive” results could be rated “referral” together with the “positive” screening results, which would also raise the sensitivity to 92.3% (23/26). In turn, the specificity would decrease considerably. This may require rescreening of these children at a later time, when children will be slightly older and adequate cooperation will be more likely as a result of further developed social behaviour. This and other options were analysed in economic evaluations of different vision screening methods in kindergartens, which demonstrated the impact of test characteristics on the cost effectiveness of screening. While the data show that the screening programme can be conducted effectively, the evaluation of treatment effectiveness was beyond the scope of this study, and remains to be addressed separately.

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Serpiginous choroidopathy presenting as choroidal neovascularisation

Serpiginous choroidopathy is an insidious, relentlessly progressive, idiopathic inflammatory disease affecting the retinal pigment epithelium and inner choroid. Choroidal neovascularisation (CNV) is a well recognised late complication of serpiginous choroidopathy in 10–25% of affected patients. In all previously reported cases CNV was recognised at the time of or after the diagnosis of serpiginous choroidopathy was established. We report a patient presenting with CNV who subsequently developed clinical findings characteristic of serpiginous choroidopathy.

Case report

A 31 year old man presented with decreased vision in his right eye in July 1997. Examination revealed acuities of 20/40 right eye and 20/20 left eye with normal anterior segments. The right fundus showed subretinal fluid and haemorrhage adjacent to the disc (Fig 1A). The left eye showed an irregularity superior to the optic disc (Fig 1B). The vitreous and fundi were otherwise normal bilaterally. Fluorescein angiography (Fig 2A, B) revealed peripapillary choroidal neovascular membranes in both eyes that were treated with argon laser photocoagulation. In April 1998 and February 1999 the left eye required photocoagulation for recurrent peripapillary CNV. Evaluation for floaters in February 2000 revealed 1+ vitreous cells and new lesions in the left eye.

Examination at the National Eye Institute in April 2000 revealed acuities of 20/40 right eye and 20/16 left eye with normal anterior segments. The vitreous contained trace cells without haze bilaterally. The right fundus showed a large peripapillary chorioretinal scar. The left fundus revealed a chorioretinal scar superior to the disc and two yellow, irregularly circumscribed, deep macular lesions (Fig 3A, B). The retinal vessels and discs were normal and no subretinal fluid, haemorrhage, or macular oedema was noted in either eye.

Fluorescein angiography revealed early hypofluorescence and late hyperfluorescence corresponding to the macular lesions in the left eye (Fig 3C, D) with no evidence of CNV in either eye. A diagnosis of serpiginous choroidopathy was made based on the clinical and fluorescein characteristics of the macular lesions in the left eye.

Comment

CNV in serpiginous choroidopathy is associated with a poor visual prognosis. In a small study CNV was reported to develop within 16 months of the serpiginous diagnosis. In a larger retrospective study of 53 serpiginous patients active CNV was found in three patients at the time of initial diagnosis and in three others within 2–17 months. Our patient differs from those previously reported in that he was diagnosed and treated for idiopathic CNV before the recognition of clinical findings.

Figure 1
July 1997. (A) The right eye shows subretinal fluid adjacent to the disc surrounded by subretinal haemorrhage extending to the fovea. (B) An undefined irregularity is noted superior to the optic disc of the left eye.

Figure 2
(A) Angiography of the right eye reveals a wedge shaped peripapillary CNV membrane. (B) A smaller CNV membrane is present angiographically superior to the left optic disc (late phase).

Figure 3
April 2000. (A) The right eye shows a large peripapillary chorioretinal scar resulting from previous laser photocoagulation of the initial CNV lesion. (B) A similar photocoagulation scar is present in the left eye which also shows two non-contiguous active serpiginous lesions. The fluorescein angiogram of the left eye shows early hypofluorescence [C, arrows] corresponding to the locations of the active serpiginous lesions that later become hyperfluorescent [D].
diagnostic of serpiginous choroidopathy. Other causes of posterior uveitis associated with CNV and choriotinal lesions similar to those seen in our patient include acute posterior multifocal placoid placoid epithelio- pathy (APMPE), presumed ocular histoplasmosis (POHS), sarcoidosis, multifocal choroiditis, birdshot chorioreti- nopathy, and toxoplasmosis. As with most cases of serpiginous choroidopathy, the CNV in these entities typically occurs late in the disease course.

The exact pathogenesis of idiopathic CNV is unknown. CNV in eyes with uveitis, however, is believed to develop in direct response to the intraocular inflammation which may alter the balance between vascular growth factors such as vascular endothelial growth factor (VEGF), and inhibitors. In the early stages of development active serpiginous lesions and CNV may appear as poorly defined subretinal lesions difficult to differentiate by ophthalmoscopy. Typically with fluorescein angiography classic CNV and serpiginous lesions are readily distinguished as the former shows early hyperfluorescence while the latter characteristically shows early blockage. Occult CNV, which may show subtle or less pronounced early hyperfluorescence with late leakage, however, may be more difficult to distinguish from an early serpiginous lesion.

This case illustrates that serpiginous choroidopathy may present with CNV. In contrast to idiopathic CNV, optimal treatment of CNV in patients with uveitis may require immunosuppressive treatment that addresses the underlying ocular inflammation with or without adjunctive laser therapy. Further investigation is needed to better define the role of emerging therapies for CNV such as photo- dynamic therapy which may offer promise for the treatment of CNV in uveitis patients.

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References

Optic neuritis in anti-GQ1b positive recurrent Miller Fisher syndrome

Only five cases of optic nerve involvement in Miller Fisher syndrome (MFS) have been documented in the literature. This report fur- ther confirms that optic neuritis may be seen in anti-GQ1b positive MFS.

Case report

This 23 year old woman presented with acute blurry vision, diplopia, and pain with eye movement. Her visual acuity was 20/20 right eye and 20/200 left eye with left relative afferent pupillary defect (RAPD). She had left red colour desaturation. Her visual field on tangent screen revealed an enlarged left blind spot and a left upper quadrant temporal peripheral field constriction. She had bilateral sixth nerve palsies, nystagmus in all gazes, and left optic disc oedema. After 1 week her visual acuity improved to 20/20 in both eyes, but her left disc remained oedematous. She then developed some left lower gait ataxia to such a degree that she was unable to walk. Dysmetria and dysdiadochokinnesia were more marked in her left upper extremity. She had very weak left upper extremity weakness, absent lower extremity deep tendon reflexes, and bilateral Babinski’s. She also had tingling in her hands and feet and decreased lower extremity vibratory sensation. Her mental status was normal throughout her ill- ness. She was not taking any drugs. A magnetic resonance image (MRI) of the brain and entire spine and MR venogram were all normal. Her cerebrospinal fluid (CSF) opening pressure and protein was 170 mmH₂O. Her CSF protein was elevated at 70 mg/dl, but CSF glucose and cell count were normal; CSF VDRL, Gram stain, routine bacterial, viral, and fungal cultures were all negative. Electroencephalogram, EEG, and the visual evoked potential (VEP) were either absent or delayed left P100 latency at 131 ms and her brainstem auditory evoked potential (BAEP) was normal. Electromyogram/nerve conduc- tion study (EMG/NCV) study revealed mildly prolonged median and peroneal F-waves, nor- mal distal motor latencies in her extremities and a reduced left median sensory nerve action potential (SNAP). Anti-GQ1b antibody (162 EIA U (normal = 100) Athena Diagnos- tics, Worcester, MA, USA) and anti-GM1 anti- body (1035 EIA U (normal = 800) Athena Diagnostics, Worcester, MA, USA) were both positive at high titres. Syphilis and Borrelia serology was normal. Antibodies for acetyl- choline receptor, hepatitis A, B, and C, Myocpasma, Campylobacter jejuni, Lyme, Hu, MaTa, Y0, CV-2, and RI were all negative. Sedimentation rate, ANA, and c-ANCA were all normal. Serum and urine toxicological screen were both negative. After 5 days of plas- mapheresis, her anti-GQ1b and anti-GM1 antibodies were negative. Her optic disc oedema, ocular motor palsies, and nystagmus immediately resolved, but she continued to walk with assistance. Two months later she had fully recovered. Six months after her recovery she developed a recurrence of her neurological symptoms and signs with left optic disc oedema. Her visual acuity at that time was 20/20 right eye and 20/100 left eye. She had a mild left lower extremity blind spot again, but no extraocular motility defects. Her VEP showed a delayed left P100 latency at 142 ms and her BAEP was normal. Single fibre EMG of her left frontalis muscle revealed no blocking suggestive of a neuro- muscular transmission defect.HLA-DR2 allele was positive and HLA-Cw3 allele was nega- tive. Her anti-GQ1b antibody (212 EIA U (normal = 100) Athena Diagnostics, Worces- ter, MA, USA) was elevated again. She underwent plasmapheresis with full recovery in about 6 months.

Comment

In addition to the classic triad of ophthalmoplegia, ataxia, and areflexia, optic nerve involve- ment presenting as optic neuritis may be a feature of anti-GQ1b positive recurrent MFS. Only five cases of optic nerve involvement in MFS have been documented in the literature.2–5 In the two previous cases of visual impairment in MFS, visual evoked potentials were either absent5 or suggestive of pre-chaismatic and post-chaismatic visual pathway dysfunction.2 Demyelinating optic neuropathies confirmed by VEP were reported in one patient with possible MFS.4 Two other cases of presumed optic neuritis were associated with anti-GQ1b positive MFS.6,7 In one patient presented here, markedly decreased visual acuity, pain with eye movement, dyschromatopsia, and optic disc oedema that resulted in good visual recovery are all indicative of the diagnosis of optic neuritis. Since high titres of anti-GQ1b gangliosides are known to be present in the human optic nerve and anti-GQ1b antibodies can cross the blood-brain barrier,3 the optic disc oedema in this patient could repre- sent an anti-GQ1b IgM complement mediated optic disc oedema.9 MFS is considered a variant of Guillain-Barré syndrome (GBS) because some patients who present with MFS progress to GBS.8 High titres of anti-GQ1b IgG antibodies are present in 80% to 100% of patients with MFS.8 MFS may be immunologically different- iated from GBS by the presence of anti-GQ1b and anti-GM1 antibodies. Although both anti-GD1a IgG and anti-GM1 IgG are associ- ated with GBS,7 anti-GM1 IgG is present in patients with typical MFS who have limb weakness,3 as in this patient. As further evidence linking this antibody to MFS,8 the decrease in anti-GQ1b antibody levels after plasmapheresis correlated with the clinical recovery in this patient. Therefore, the ele- vated titres of anti-GQ1b and anti-GM1 antibodies, along with the clinical triad of ophthalmoplegia, areflexia, and ataxia in this patient all support the diagnosis of MFS, and not GBS.

In rare cases, MFS has been known to recur. This patient presented with a relapse of similar clinical features 6 months after her diagnosis from her initial episode. In the study done by Chida et al.,7 patients with recurrent MFS appeared to have similar HLA typing charac- teristics as the non-recurring ones. Both types of patients had HLA-DR2 and Cw3 alleles, but the fre- quency of HLA-DR2 was slightly higher in the patients with recurrent MFS.7 Therefore, this patient’s HLA-DR2-positive status may have been a risk factor for her recurrence of MFS. This case report emphasises that optic neu- ritis may be a central nervous system feature that should be recognised as part of the Miller Fisher syndrome. The presence of both anti- GQ1b IgG and anti-GM1 IgG in this patient provides immunological evidence supportive
of typical MFS. The delayed P100 latency in her VEP also provides electrophysiological evidence that the optic nerve is affected in anti-GQ1b antibody positive MFS. Furthermore, this is the first documented case known to the author of optic neuritis in the recurrent subtype of MFS which is associated with a higher frequency of the HLA-DR2 allele.

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References

Ocular myasthenia gravis and inflammatory bowel disease: a case report and literature review

Myasthenia gravis has been reported to be associated with both ulcerative colitis (UC) and Crohn’s disease (CD). The link between inflammatory bowel disease (IBD) and myasthenia gravis (MG) is thought to be related to the production of autoantibodies. Myasthenia gravis is also associated with other autoimmune diseases including alopecia, lichen planus, vitiligo, and systemic lupus erythematosus.

Similarly, IBD frequently presents with other autoimmune disorders. One study demonstrated a 9.4% prevalence of autoimmune disorders in patients with UC including sclerosing cholangitis, thyroid disorders, vitiligo, insulin dependent diabetes mellitus, thyroid disease, pernicious anaemia, scleroderma, and seropositive rheumatoid arthritis. Despite the association between MG and other autoimmune disorders, there are relatively few reports of ocular findings as the presenting sign of MG in patients with IBD.

Case report
A 21 year old African-American male, with a medical history of biopsy proved ulcerative colitis diagnosed in 1995, focal segmental glomerular sclerosis determined by renal biopsy in 1995, and primary sclerosing cholangitis determined by liver biopsy in 2000 presented to the neuro-ophthalmology service with complaints of binocular diplopia and ptosis of the left upper eyelid. Both the diplopia and the ptosis were better in the morning and worsened during the course of the day. His ulcerative colitis had been in remission for the past 5 years without medication.

Best corrected visual acuity was 20/25 in each eye. The external examination revealed ptosis of the left upper eyelid that worsened in sustained upgaze. He had limited extraocular motility in all fields of gaze (Fig 1). The remainder of the neuro-ophthalmic examination was normal and he had no difficulty with speech or swallowing.

Laboratory evaluation revealed a positive acetylcholine receptor antibody and normal thyroid function studies. There was no evidence of a thymic mass on magnetic resonance imaging of the chest.

The patient returned to the emergency room 1 week later with difficulty swallowing and shortness of breath. He was hospitalised for plasmaphenesis and upon discharge treated with imuran, prednisone, and mestepefate. Review revealed only one other purely ocular presentation of myasthenia associated with ulcerative colitis; however, details of the ocular examination were not included. Another report, of a 21 year old woman with a 3 year history of Crohn’s disease, documented diplopia and unilateral ptosis as the initial findings of MG. She was found to have acetylcholine receptor antibodies and her ocular findings improved with pyridostigmine.

Because of the relatively few reports of ocular myasthenia in patients with IBD we reviewed the English literature and found four additional reports of MG in patients with IBD. Based on these four reports and the three (including the present report) with ulcerative colitis in patients with IBD (Table 1), the mean duration of IBD before the diagnosis of MG was 10 years.

Autoimmune dysregulation is the central defect in both MG and IBD. Both IBD and MG may be associated with an elevated carcinoembryonic antigen (CEA) and decreased peripheral lymphocyte counts that subsequently normalise following thymectomy. Some studies have shown abnormal thymic involution and the presence of an abnormal ratio of T suppressor to T helper cells in both MG and UC, while others have noted a decline in suppressor T cells and an increase in

Table 1 Previous reports of myasthenia gravis occurring in patients with inflammatory bowel disease

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age (years)</th>
<th>Sex</th>
<th>IBD</th>
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<th>AchR antibody reactivity</th>
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<tr>
<td>Miller 1971</td>
<td>35</td>
<td>Male</td>
<td>UC</td>
<td>13</td>
<td>Unknown</td>
</tr>
<tr>
<td>Tan 1974</td>
<td>38</td>
<td>Male</td>
<td>UC</td>
<td>12</td>
<td>Unknown</td>
</tr>
<tr>
<td>Martin et al, 1991</td>
<td>63</td>
<td>Male</td>
<td>CD</td>
<td>15</td>
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</tr>
<tr>
<td>Gower-Rousseau et al, 1993</td>
<td>27</td>
<td>Female</td>
<td>UC</td>
<td>10</td>
<td>Positive</td>
</tr>
<tr>
<td>Finnie et al, 1994</td>
<td>21</td>
<td>Female</td>
<td>CD</td>
<td>3</td>
<td>Positive</td>
</tr>
<tr>
<td>Lossos et al, 1995</td>
<td>11</td>
<td>Male</td>
<td>CD</td>
<td>9</td>
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<tr>
<td>Present report</td>
<td>21</td>
<td>Male</td>
<td>UC</td>
<td>7</td>
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</table>

IBD = inflammatory bowel disease, MG = myasthenia gravis, AchR = acetylcholine receptor, UC = ulcerative colitis, CD = Crohn’s disease.
immature helper T cells suggesting migration without normal maturation.

The immunological link between MG and IBD is highlighted by two reports of patients undergoing surgical treatment. One report of a patient with both MG and CD demonstrated improvement in perineal and perianal disease following thymectomy for severe uncontrolled MG. Another patient with both MG and UC demonstrated regression of the myasthenia following proctectomy.

Although the simultaneous occurrence of these two autoimmune disorders is uncommon, it is important to understand that ocular findings may be the initial manifestation of MG in patients with IBD.

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References

Magnetic resonance imaging findings in malignant melanoma of the lacrimal sac

A case of primary malignant melanoma of the lacrimal sac is presented. This is the first report of the preoperative magnetic resonance imaging (MRI) findings of malignant melanoma of the lacrimal sac.

Case report
A 54 year old Chinese woman was referred to an ophthalmologist complaining of a 6 month history of left sided bloody tears and epistaxis. She had a firm, non-tender left medial canthal swelling, and syringing revealed left nasolacrimal duct (NLD) obstruction. Ocular and periorbital examination was otherwise normal. A dacyrocystogram (DCG) demonstrated a filling defect in the lacrimal sac with NLD obstruction.

Radiological features of lacrimal sac tumours include filling defects on DCG and mass lesions on CT. However, to the authors’ knowledge, this is the first report of the MRI findings of malignant melanoma of the lacrimal sac.

Owing to the paramagnetic properties of melanin, malignant melanoma appears hyperintense on T1 weighted imaging, and hypointense on T2 weighted imaging. A study of six mucosal melanomas of the head and neck found that on T1, five lesions were hyperintense and one was isointense. On T2, five were of mixed intensity and one was iso-intense. They concluded that hyperintensity on T1 of mucosal melanomas was characteristic but not universal.

The majority of malignant lacrimal sac tumours are epithelial in origin. Imaging features suggesting malignancy include invasion of bone, rapid growth, and irregular margins with skin fixation. On MRI, the majority of epithelial tumours have intermediate signal intensity on T1 and high T2 signal intensity. High tumour cellularity is associated with intermediate to low T2 signal intensity.

High signal intensity on T1 is not specific for malignant melanoma. Subacute haemorrhage caused by the presence of methaemoglobin is more likely and although melanoma may undergo intratumoral haemorrhage, other tumours with a tendency to bleed include small cell lung carcinoma, choriocarcinoma, and renal cell carcinoma metastases. Less likely causes include fat containing tumours (lipoma, dermoid, and teratoma).

An ENT opinion was sought, and nasal examination revealed left sided septal deviation, with no obvious cause for the epistaxis. Computed tomography (CT) of the head and orbits demonstrated a left lacrimal sac lesion extending into the NLD with proximal dilation of the duct and no apparent bone erosion (Fig 1A). MRI confirmed the presence of a lacrimal sac lesion with intermediate signal intensity on T1 and T2 weighted images (Fig 2A, B). The lesion enhanced with intravenous gadolinium.

An incisional biopsy of the lacrimal sac (Fig 1B) under frozen section control, and paraffin sections, confirmed malignant melanoma.

A full medical review, including MRI of the chest and abdomen, and liver function tests, excluded tumour elsewhere. However, abdominal MRI and ultrasound revealed a co-incidental polycystic liver.

Three weeks after biopsy, a wide local excision including the medial upper and lower eyelids, dacryocystectomy and medial maxillectomy was performed. A tumour, confined to the sac, and invasion through the medial wall of the upper NLD, into the lateral wall of the nose, and apposing nasal septal mucosa, was seen peroperatively and confirmed historically.

She underwent postoperative adjuvant radiotherapy (55 grays) and to date, 4 months later, remains well.

Comment
Malignant melanoma of the lacrimal sac is rare accounting for 5% of lacrimal sac tumours. It has an unfavourable prognosis compared with other causes of lacrimal sac tumour, and is considered more aggressive than cutaneous malignant melanoma. Response to treatment is generally poor, with up to 80% of cases recurring within 2 years.

Figure 1 (A) Coronal CT scan demonstrating a solid mass of the left lacrimal sac with proximal dilation of the nasolacrimal duct (arrow). (B) Incisional biopsy with lacrimal sac opened and melanoma visible.

Figure 2 (A) T1 weighted sagittal MRI demonstrating intermediate signal intensity mass lesion of the lacrimal sac and proximal nasolacrinal duct (arrow). (B) T2 weighted axial MRI demonstrating intermediate signal intensity mass lesion of the left lacrimal sac (arrow).
require MRI fat suppression methods, paramagnetic material (manganese, iron, and copper), and very high (non-paramagnetic) intratumoral protein concentration.

MRI has been reported as a useful investigative tool in the assessment of lacrimal disease owing to its ability to delineate soft tissues. Intravenous and intracanalicular gadolinium adds useful information on lesional enhancement and lacrimal apparatus structure and function. The predictive value of MRI for lacrimal sac melanoma, however, appears to be variable. Hyperintensity on T1 relies on the paramagnetic properties of melanin, the presence of which is variable in amelanotic melanoma. This is supported by our case, where only moderate T1 hyperintensity with contrast enhancement was demonstrated.

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Photodynamic therapy for recurrent myopic choroidal neovascularisation after limited macular translocation surgery

Limited macular translocation (LMT) is one of the treatment options for subfoveal choroidal neovascularisation (CNV) resulting from pathological myopia.1 The fundamental surgical principle involves the transposition of the foveal neurosensory retina to a new site with more healthy underlying retinal pigment epithelium.1,2 Direct laser photoacoagulation is usually employed as an adjunct measure in eradicating the original CNV after the surgery. It has been observed that geometrically sizeable translocation is a prerequisite for a long term surgical success.3,4 The degree of translocation is, however, not often predictable and any ineffective displacement may render the subsequent laser photoacoagulation extremely difficult or even impossible to perform.5,6 As a result, the recurrent or persistent CNV intruding the newly relocated fovea may jeopardise the final visual outcomes.7,8 Photodynamic therapy (PDT) may be considered as a viable adjunct treatment option in such circumstance.

Case report
A 41 year old woman with pathological myopia of −11.0 dioptres in both eyes presented with a subfoveal CNV and subretinal haemorrhage in her right eye in July 2000. The best corrected visual acuity (BCVA) was 20/200 in her right eye and 20/30 in her left eye. LMT with superotemporal 6 mm scleral imbrication was performed in July 2000. The operation was uneventful and an inferior displacement of the fovea by 600 µm was achieved. The CNV however, was still located in the vicinity of the juxtapfoveal area and therefore laser photoacoagulation, bearing the potential risk of late creeping scar, was not suggested. At the 4 months postoperative visit, her left BCVA was 20/200 and the original CNV became more fibrotic with minimal leakage upon fluorescein angiogram. Nevertheless, she came back at 5 months with a return of metamorphopsia and a drop in her right vision to 20/200, and later to 10/200. Dilated fundus examination showed a tiny patch of submacular haemorrhage in direct continuity with the old fibrotic scar (Fig 1A). Fluorescein angiogram of the early phase demonstrated a fresh recurrent CNV budding out from the original CNV and extending to the centre of the foveolar avascular zone (Fig 1B). Moderate fluorescein leakage could be seen in the late phase (Fig 1C). Treatments comprising revision macular translocation surgery, submacular surgery, photodynamic therapy, and observation had been thoroughly discussed with the patient. In view of minimal invasiveness and comparatively better preservation of surrounding neurosensory retinal tissue, PDT was adopted in treating the CNV recurrence. PDT with verteporfin infusion and laser delivery was performed in accordance with the standard protocol. After the treatment, the blood clot in the fovea was gradually reabsorbed and the vision improved to 20/200 at 3 months of follow up. Complete regression of the recurrent CNV at the fovea without angiographic leakage was documented over the follow up angiogram at 3 months and subsequently (Fig 1D). The vision remained stable at 20/200 in the latest visit at 24 months after the PDT.

Comment
It has been shown that significant visual improvement may be achieved by LMT for the treatment of subfoveal CNV associated with age related macular degeneration (AMD) or pathological myopia.23,4 However, the surgical techniques are demanding and the potential complications are not unusual. One of the late postoperative visually important complications is recurrence of the CNV and this is partially caused by an ineffective translocation of the fovea or a large lesion size of CNV.5,6 The incidence of persistent or recurrent CNV after limited LMT has been reported to be 40% and 35% respectively in age related macular translocation and being 21% and 14% respectively in pathological myopia.7,8 Not many treatment options are available once the fovea is involved. Viable surgical options including repeated LMT, full 360 degree retinotomy MT, or submacular surgery may be considered but the surgical risk may be inadvertently higher in the redetachment of the neurosensory retina. PDT induces a selective thrombosis of the abnormal CNV and has been proved to be an effective treatment in preventing a significant loss of vision in patients with CNV secondary to AMD or pathological myopia.9,10

Figure 1 Right eye with recurrent myopic CNV after LMT. (A) Fundus photograph of the patient showing the recurrent part of CNV budding from the original one with haemorrhage involving the subfoveal area. (B) Early phase fluorescein (FA), demonstrating the filling of choroidal vascular complex with early hyperfluorescence. (C) Late phase FA showing moderate fluorescence leakage from the CNV. Photodynamic therapy (PDT) with the size of the laser spot as marked was delivered. (D) Late phase FA at 12 months revealing a complete regression of the recurrent CNV and late scar staining of the original CNV.
Its clinical indications and applications are expanding. Its minimal invasiveness and clinical efficacy make it a safer and visually desirable supplementary treatment in recurrent CNV after LMT. In our patient, the complete closure of CNV was achieved with concurrent vision improvement after a single session of PDT without evidence of recurrence at 24 months.

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Acquired Glanzmann’s thrombasthenia causing prolonged bleeding following phacoemulsification

Phacoemulsification under topical anaesthesia using clear corneal incision is not a challenging procedure for the haemostatic system. In patients with known bleeding dia-
tasis, this may be the procedure of choice to remove cataract. We report a patient who bled continuously for 36 hours following phaco-
emulsification under topical anaesthesia through a clear corneal incision. This was managed by using a topical haemostatic agent that has not been used in ophthalmic surgery before. Extensive haematological evaluation revealed the underlying cause to be an acquired form of Glanzmann’s thrombasthes-
tia, a very rare condition.6

Case report
A 79 year old woman underwent left phaco-
emulsification with intraocular lens implanta-
tion through a clear corneal temporal incision. The procedure was uneventful but she was seen to bleed from the operated eye in the recovery room. The eye was patched but the bleeding continued soaking the pads. When re-examined 2 hours later, as there was continuous bleeding, the eye was patched with gentle pressure. Examination the next day showed that the bleeding was persistent. Pressure bandage was reapplied. Examination in the operating theatre con-

firmed the conjunctival origin of the bleeding from the site where the left handed surgeon held the conjunctiva during surgery. Cauter-
sation and an attempt to suture the conjunc-
tiva were unsuccessful. It was decided that the safest option was to use a small piece of oxidised regenerated cellulose (Surgicel, Ethicon) on the bleeding site and patch the eye.

The piece of Surgicel with clotted blood that was lying loose on the conjunctiva was removed at review 24 hours later. The conjunctival site had stopped bleeding with evidence of altered blood on the surface where Surgicel had been applied (Fig 1A). At her last review 8 weeks later, she was found to have a corrected visual acuity of 6/18 due to pre-
existent macular changes secondary to retinal detachment that was reattatched in 1976. The conjunctiva had healed well (Fig 1B). The patient had previously undergone an un-

eventful phacoemulsification and intraocular lens implantation in her right eye under sub-

Tenon’s anaesthesia.

The patient’s recent medical history was significant for recurrent admissions else-

where for investigation of severe anaemia fol-

lowing gastrointestinal bleeding. Platelet count and clotting screen had been normal. Angiodysplasia of stomach and duodenum were treated with laser and angiodyplasia of colon was treated by hemicolecotomy. Three episodes of epistaxis and an episode of vaginal bleeding were managed conservatively. She

had received 60 units of blood transfusion over a period of 1 year. Interestingly, she had appendicectomy and multiple dental extrac-
tions elsewhere many years previously with-

out any significant bleeding. She has not been on any antplatelet agents or anticoagulants. There was no family history of bleeding disor-

ers.

A defect in the platelet function was suspected, as her coagulation screen including the platelet count was normal. Platelet aggreg-
tion tests showed no aggregation against any agonists other than ristocetin, which is dependent on platelet glycoprotein Ib. The platelets showed normal normalisation of glycoprotein antigens IbIIa and Ib. The patient’s serum showed presence of inhibitory antibody against glycoprotein IbIIa. This led to a diagnosis of acquired Glanzmann’s syndrome, an extremely rare condition of autoimun thrombasthenia. No underlying malignant, autoimmune, or lymphoprolifera-
tive disorder had been identified as a cause for this patient’s acquired Glanzmann’s throm-

basthenia.

Comment
The patient described had uncontrollable bleeding for 36 hours following a procedure, which is generally considered safe even in patients with a bleeding disorder. She developed bleeding from the conjunctival site where the surgeon grasped the conjunctiva during cer-

tain stages of the procedure. One would usually not expect any significant bleeding from this site; however, in a patient with com-

promised haemostasis the bleeding may be prolonged. Although the bleeding was no more than a gentle ooze at any point in time it was persistent enough for 36 hours before the topical haemostatic material Surgicel had been put to use. The consequences of an intraocular bleed may have seriously threat-

ened her sight.1

We are not aware of any reports of the use of Surgicel in ophthalmic surgery. All reports of its use are in other fields of surgery.6 This material is supposed to swell up with blood and form a gelatinous mass that aids in the formation of clot. It acts as a haemostatic adjunct. The exact mode of its action in this patient with antplatelet antibodies is unclear. Our experience shows that oxidised regenerated cellulose (Surgicel) may have a role in ophthalmic surgery especially in lacrimal and orbital surgery, when faced with bleeding that is difficult to stop. Various cautionary tales associated with use of Surgicel have been reported.8

Our report suggests that in the presence of a severe bleeding disorder, clear corneal phacoemulsification under topical anaesthe-

sia may not be totally safe. When performing such a procedure in a patient with known bleeding disorder it may be safe to take all the necessary precautions in consultation with a haematologist to avoid a serious bleed that may be sight and life threatening. There may be a role for haemostatic agents like Surgicel.

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Propionibacterium acnes endophthalmitis diagnosed by microdissection and PCR

Although Propionibacterium acnes, a Gram positive anaerobic bacillus, is the most commonly identified cause of delayed onset postoperative endophthalmitis, routine vitreous cultures are frequently inadequate for its diagnosis. This case describes the utility of the histopathological technique of microdissection and polymerase chain reaction (PCR) for the diagnosis of delayed postoperative endophthalmitis.

Case report

A 78 year old man with a history of vitreous floaters, a coronary bypass, and aortic valve replacement underwent an uncomplicated cataract extraction with intraocular lens (IOL) implantation in the right eye. Three months later, he developed increasing floaters in the right eye and was diagnosed with vitritis unresponsive to corticosteroid treatment. Examination revealed acuities of 20/25 in the right eye and 20/20 in the left with normal intraocular pressures. The right eye was significant for no anterior chamber cells or flare, dilated iris vessels, an IOL without deposits, 3+ vitreous cells with trace haze, and peripheral pigmentary degeneration. The left eye was normal with the exception of trace vitreous cells and a choroidal naevus. A diagnostic vitrectomy was performed in the right eye. A portion of the vitreous specimen was cultured for fungi, aerobic and anaerobic bacteria, and the remainder was processed for histopathologic examination, and PCR analysis. The engulfed bacilli were microdissected under a microscope with a 30 gauge needle and submitted for PCR.

Nested PCR with P acnes specific oligodeoxynucleotide primers complementary to regions of 16S rDNA was used. The primers were Pa1, AAG GCC CTG CTT TGT TGG; rPa2, TCC ATC CGC AAC CGC CGA A; and rPa3, ACT CAC GCT TGG TCA CAG. Nested-PCR analysis revealed P acnes (Fig 2). A diagnosis of delayed postoperative endophthalmitis was made.

Comment

The most common causes of vitritis in elderly patients are acquired or postoperative infections, sarcoidosis, and intraocular malignancies masquerading as uveitis. An early diagnostic procedure is indicated if postoperative endophthalmitis is suspected. In this case, although the chronic inflammation and intracytoplasmic Gram positive bacilli in a few macrophages suggested an infectious process, the negative cultures precluded the diagnosis of an infectious endophthalmitis. To further investigate the possibility of a bacterial infection nested PCR was performed on the microdissected bacilli. Molecular analysis verified the presence of P acnes and a diagnosis of delayed postoperative endophthalmitis was confirmed.

Vitreous cultures are positive in less than 50% of postoperative endophthalmitis cases. In a study of 23 patients with delayed onset endophthalmitis aqueous culture and microscopy were diagnostic in 0% of cases, vitreous culture was positive in 24% and PCR from the aqueous and vitreous yielded a positive diagnosis in 84% and 92%, respectively. Treatment of P acnes endophthalmitis includes intravitreal vancomycin plus consideration of pars plana vitrectomy with or without capsulectomy with or without IOL removal. Although aggressive surgical intervention eradicates the infection similar visual outcomes are reported with more limited surgical treatment.

In our case the intracytoplasmic bacteria in the macrophages were the only evidence of a bacterial infection. To detect the presence of P acnes we referenced the PCR method described by Hykin that used 150 μl of the vitreous for culture and 100 μl for PCR. Using the technique of microdissection and PCR with a similar volume of vitreous we additionally performed cytology and cytokine analysis which are helpful in the diagnosis of other causes of vitritis.

This case further illustrates the benefits of molecular analysis for the diagnosis of culture.
negative delayed onset endophthalmitis. It also describes for the first time microdissection and PCR for the evaluation of endophthalmitis. Advantages of this technique are that it allows for a more comprehensive pathological examination on a limited specimen and provides the option of having the molecular studies being performed elsewhere.

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Interferon treatment of childhood conjunctival lymphoma

Mucosa associated lymphoid tissue (MALT) lymphoma is the most common ocular adnexal lymphoproliferative disease in children. MALT lymphomas have a more indolent course than non-MALT lymphomas, are usually found in the older age groups (50–70 years), are usually limited to localised (stage I) disease at presentation, and radiotherapy and chemotherapy have been the mainstay of treatment.

Case report
A 15 year old male was referred by an ophthalmologist after an 8 month history of unilateral painless follicles at both nasal fornice (Fig 1A). There were no visual symptoms and, based on a working diagnosis of an atypical vernal reaction, topical steroid treatment was prescribed. Incisional biopsy was performed at the lesions (36%), observation (9%), and palliative therapy (15%). Radiotherapy has been used with some success but persistent ocular morbidity in the form of corneal ulceration, radiation induced conjunctivitis, and ocular irritation disorders have been reported.4 Intranasal IFN-α is a relatively new therapy which has been shown to be both effective and safe in a small number of cases.1, 4 Non-sight threatening ocular complications such as subconjunctival haemorrhage and local chemosis have been reported, as well as minor transient systemic effects including headaches, nausea, fevers, chills, and myalgia.1 Administration of intranasal IFN-α is also a relatively simple and quick procedure. It shows great promise as a first line agent to treat conjunctival lymphoma, but long term follow up is needed.

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Unilateral corneal anaesthesia and ulceration following squint surgery in a child with Pendred syndrome and bilateral sixth nerve palsy

We present a 4 year old child with Pendred syndrome and bilateral sixth nerve palsy. To our knowledge this association has not been previously reported. In addition, this patient developed unilateral corneal ulceration with associated corneal anaesthesia following squint surgery. We will discuss the pathophysiology of this unusual complication following squint surgery.

Case report
This patient presented when he was 6 months old with right convergent squint. He was diagnosed with Pendred syndrome (sensoneural hearing loss and thyroid dysfunction) by the paediatricians and the otolaryngologists following abnormal thyroid function tests and a computed tomograph (CT) scan of the temporal bones showing Mondini malformations of both cochleas. At presentation his visual acuities were 6/60 right eye and 6/36 left eye using the Cardiff acuity cards. He had bilateral alternating esotropia with an inability to abduct either eye. There was no globe retraction or abnormal lid movements and a magnetic resonance imaging (MRI) scan had shown congenital absence of the auditory nerves but no other abnormality. A diagnosis of bilateral sixth nerve palsy was made. The squint was cosmetically poor and measured at 45 prism diptres in the distance and near. He had low hypermetropia with no significant anisometropia. Funduscopy was normal. He was reviewed regularly in the paediatric eye clinic over the next 3 years during which time his visual acuities were within normal limits, the best recorded acuity being 6/9 right eye and 6/9 left eye using singles.

When he was 4 years old, he underwent bilateral superior rectus and inferior rectus lateral transpositions under general anaesthesia, which was uneventful with no immediate postoperative complications, and a cosmetic improvement. He also underwent an alternating convergent squint of 15 prism diptres for distance and near. Two months later he developed a left inferior corneal ulcer (Fig 1) with surrounding punctate epitheliopathy which surprisingly did not seem to cause him as much distress as expected. The left corneal sensation was definitely reduced compared to the right eye. There was no previous history of herpes simplex keratitis, postoperative anterior segment ischaemia, surgical trauma to the long posterior ciliary nerves or ciliary ganglion, congenital absence of sensation, and surgery reducing Bell's phenomenon.

The clinical course was not typical of herpes simplex and there was no previous history of corneal pathology. Postoperative anterior ischaemic syndrome was unlikely as only two recti muscles were operated on and no previous uveitis was observed. To our knowledge there are no reported cases of corneal anaesthesia after squint surgery. There was no evidence of previous involvement, which one may expect with trauma to the long posterior ciliary nerves or ciliary ganglion.

Congenital absence of corneal sensation was the most likely cause, especially in view of his unusual cranial nerve anomalies, and we believe he had pre-existing corneal anaesthesia before squint surgery despite the absence of any other fifth cranial nerve signs. Following the lateral transposition of the superior rectus his Bell's phenomenon was noted to be absent thereby compromising his corneal protection. In addition, he was observed to have significant lagophthalmos while asleep.

We believe that the combination of corneal anaesthesia, abolished Bell's phenomenon, and lagophthalmos compromised his corneal integrity resulting in corneal ulceration.

This case highlights the importance of determining corneal sensation before transposition surgery on the superior rectus as Bell's phenomenon may be abolished thereby compromising corneal protection. This is especially relevant in patients with unusual cranial neuropathy and lagophthalmos.

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Gemella haemolysans acute postoperative endophthalmitis

Endophthalmitis is perhaps the most feared complication of cataract surgery, with a reported incidence between 0.07% and 0.13%. The most common organisms reported in previous studies are Gram positive staphylococci and streptococci. We report a case of severe endophthalmitis with an unusual Gram positive organism, after uncomplicated phacoemulsification, with foldable intraocular lens implantation.

Case report
A 66 year old white man underwent routine phacoemulsification cataract extraction with posterior chamber lens implantation (AcrySof, Alcon, and Hydrosview H60M, Bausch & Lomb) to the right eye in January 2002.

The left eye had previously undergone similar surgery in September 2001. He was generally in good health, and on no medication. There was a past medical history of sarcoidosis treated with oral prednisolone in 1970, which has since been in remission, and an episode of staphylococcal septicaemia in 1987, without sequelae.

On the first postoperative day, visual acuity measured 6/9 unaided and ocular examination was unremarkable. That same afternoon the patient developed ocular pain, initially relieved by paracetamol (acetaminophen), which however, worsened during the night with progressive deterioration of vision. He presented to the ophthalmic emergency department the following morning with the aforementioned symptoms. Visual acuity was reduced to hand movements right eye and 6/9 left eye. Slit lamp examination revealed an oedematous cornea with Descemet’s folds. The anterior chamber was hazy, with 1 mm hyperopia and the intraocular pressure measured 38 mm Hg.

There was no red reflex. B-scan ultrasound examination showed extensive vitreous debris with attached retina. The left eye was pseudoephelitic with no abnormalities of note. A diagnosis of acute postoperative endophthalmitis was made. Anterior chamber and vitreous samples were obtained for aerobic and anaerobic culture/sensitivity and Gram staining. Intravitreal vancomycin 2 mg and amikacin 300 µg, each in 0.1 ml of balanced salt solution and subconjunctival cefuroxime 125 mg were administered. Oral ciprofloxacin 500 mg twice daily, prednisolone 1 mg once a day, topical gentamicin hourly, ofloxacin hourly, and atropine 1% twice a day were commenced.

Preliminary Gram staining suggested a Gram positive coccus sensitive to ciprofloxacin—oral and topical antibiotics were therefore continued. Owing to difficulty in identifying the nature of the organism, the samples were sent to a regional reference laboratory, which identified Gemella haemolysans from both anterior chamber and vitreous aspirates. The organism was reported to be sensitive to gentamicin, ciprofloxacin, levofloxacin, amoxicillin/clavulanate, chloramphenicol, and resistant to trimethoprim.

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The patient continued to make steady progress; 2 months later vision had improved to 6/9 unaided. The patient at that time was troubled by floaters secondary to considerable vitreous debris. At last review in September 2002, visual acuity had further improved to 6/4 with −0.75 D sph correction.

Comment

Gemella haemolysans is an aerobic or facultative anaerobic, Gram positive coccus, a normal commensal of the oral cavity and upper respiratory tract of low virulence.1' Systemic infection may lead to septic shock, meningitis, arthritis, or pneumonia, all of which are rare. Identification is difficult. Though Gram positive, the cocci are easily decolorised and hence may appear Gram variable or even negative. Identification is difficult. Though Gram positive, the cocci are easily decolorised and hence may appear Gram variable or even negative. Initially Gemella was included under the genus Neisseria but is now classified as a separate genus within the family Streptococcaceae.2 No studies on susceptibility to antiseptics have been published, though there is no reason to believe that it may be resistant to povidone-iodine preparations. The organism is still viable in vitro to penicillin, streptomycin, vancomycin, chloramphenicol, and tetracyphathiazole. A literature search revealed only one previously reported case of infection by Gemella haemolysans, with ketarthritis and consecutive endophthalmitis.3 Interestingly this patient was reported to have active sarcoidosis on systemic steroid therapy, whereas our patient had a past history of sarcoidosis. This possible association between sarcoidosis and infection by Gemella may be purely coincidental, as no such association has been reported with systemic infection.

Gemella haemolysans is difficult to identify, because of its close resemblance to viridans streptococcus and Neisseria. As diagnostic technology improves, Gemella haemolysans endophthalmitis may be described more often in the future. This report highlights the importance of infection with rare commensal organisms in healthy, immunocompetent individuals after uneventful phacoemulsification cataract surgery.

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Does topical brimonidine tartrate help NAION?

There is no proved treatment for non-arteritic anterior ischaemic optic neuropathy (NAION). Topical brimonidine tartrate has been reported to have a neuroprotective effect for retinal ganglion cells following experimental elevation of intraocular pressure and optic nerve injury in the rat, which is blocked with coadministration of the α-2 antagonist, rauwolscine.4 Increased retinal ganglion cell survival has also been shown to occur following oral administration of brimonidine in monkeys with experimental glaucoma.5 These results were the basis of the recently aborted clinical trial of topical brimonidine purify for acute NAION and our retrospective study of 31 patients with NAION, who were evaluated within 3 weeks of the onset of visual loss, and followed up for a minimum of 8 weeks. During 2001–2, we treated all 14 patients with brimonidine tartrate within 14 days (mean 5.3, SD 5.32) of the onset of visual loss. Five patients were treated after 1 day of symptoms. Three were taken four times a day in 11, three times a day in one, and twice a day in two patients. All 17 untreated patients were evaluated at the year before and were matched to the treated group for age, sex, cardiovascular risk factors, previous aspirin use, and previous first eye NAION.

Snellen visual acuity and colour vision, using the Ishihara colour plates, were documented and expressed as a decimal equivalent (for acuity: 20/60 = 0.33 and light perception = 0.001; for colour vision: the number of correctly identified plates/the total number of plates). The visual fields (Humphrey or tangent perimeter) were analysed and defects were graded according to the following scale: 0 = normal, 1 = accurate nerve fibre bundle defects, 2 = retinal defects (6–12 degrees), 3 = altitudinal defect plus additional loss, 4 = no light perception. A third examiner, who was unaware of the dates of the visual fields and the patients’ treatment status, also evaluated all visual fields and determined, in each patient, whether the field was better or worse than or equivalent to the other field. The intraocular pressure was measured four times in all patients. The pressure was 25 mm Hg in one patient in the treated group and 24 mm Hg in one patient in the untreated group.

Statistical analysis of the data involving comparisons of the treated and untreated groups at baseline and 8–12 weeks was performed using the two tailed t test.3 The Wilcoxon signed rank test was used to compare the individual vision performance from baseline to the 8–12 week examination.2 For visual acuity and colour vision, a positive rank indicated improvement and a negative rank indicated a worse visual outcome. For the visual field grade value was better for the treated group than for the untreated group (p=0.007). At the 8–12 week examination, the mean visual acuity was 0.29 (SD 0.30) for treated and 0.49 (SD 0.39; p=0.12) for untreated patients. The mean visual field grade was 2.2 (SD 0.81) for treated and 1.0 (SD 0.70; p=0.04) for untreated patients. The mean colour vision was 0.42 (SD 0.41) for treated and 0.55 (SD 0.46; p=0.43) for untreated patients.

For the masked examiner’s evaluation, the mean baseline visual field (2.0, SD 0.91) was similar to the field (1.93, SD 0.96; p=0.85) for controls. At the 8–12 week examination, the mean visual field grade was 2.15 (SD 0.99) for treated and 1.87 (SD 0.92; p=0.43) for untreated patients. This examiner further found that the output visual fields for the treated group were improved in two patients, worse in six patients (50%), and unchanged in four patients. The outcome visual fields for the control group were improved in five patients, worse in two patients (13%), and unchanged in eight patients.

The Wilcoxon signed rank analysis demonstrated that for visual acuity, two patients in the control group and 10 patients in the treated group had negative values or a worse outcome at 8–12 weeks (p=0.003). For colour vision, one patient in the control group and eight patients in the treated group had negative values or a worse outcome (p=0.013). For visual fields, one patient in the control group and four patients in the treated group had worse and positive values or a worse outcome at 8–12 weeks (p=0.046).

The average time to start the drops was 3.5 days from the onset of vision loss. In the treated group, the mean baseline visual field (2.0, SD 0.91) was similar to the field (1.93, SD 0.96; p=0.85) for controls. At the 8–12 week examination, the mean visual field grade was 2.15 (SD 0.99) for treated and 1.87 (SD 0.92; p=0.43) for untreated patients. This examiner further found that the output visual fields for the treated group were improved in two patients, worse in six patients (50%), and unchanged in four patients. The outcome visual fields for the control group were improved in five patients, worse in two patients (13%), and unchanged in eight patients.

The Wilcoxon signed rank analysis demonstrated that for visual acuity, two patients in the control group and 10 patients in the treated group had negative values or a worse outcome at 8–12 weeks (p=0.003). For colour vision, one patient in the control group and eight patients in the treated group had negative values or a worse outcome (p=0.013). For visual fields, one patient in the control group and four patients in the treated group had worse and positive values or a worse outcome at 8–12 weeks (p=0.046).

The average time to start the drops was 3.5 days from the onset of vision loss in those who worsened. There was no correlation with a worse outcome and time to initiate therapy.

For all parameters of vision testing, there was a trend for worse visual performance at 8–12 weeks in the group treated with topical brimonidine. Although there was no significant difference for the colour vision outcome, this might reflect that the baseline colour vision value was better for the treated group. The outcome visual field grade was significantly worse in the treated group. The masked examiner’s visual field evaluations demonstrated that more treated patients worsened than in the untreated group. When the baseline and outcome of all visual parameters for each individual were compared, the treated group had a significantly worse outcome at 8–12 weeks.

Our results are not the first description of worse outcome in patients treated with α-2 agonists for central nervous system ischaemic disease. Studies in animal models and clinical studies in humans suggest the potential of drugs, including α-2 receptor agonists, may impede recovery following stroke. Clonidine administration caused recurrence of the neurological deficit in animals who had initially recovered. In a retrospective study, the level of motor recovery of stroke patients was worse in those treated with α-2 agonists than in patients not receiving these agents.8

Although in experimental optic nerve injury in animal models, brimonidine appears to offer neuroprotection, our results demonstrate that brimonidine tartrate, applied topically up to four times daily, does not appear to be a beneficial treatment for acute NAION. It
is possible earlier treatment might have been more effective, although patients who worsened received treatment sooner than those who did not worsen. Increased dosing frequency or using a different preparation of botulinum toxin might be more effective. Additionally, the number of subjects in the study was small and a negative trend could appear more profound.

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**References**


**Chronic eye movement induced pain and a possible role for its treatment with botulinum toxin**

Chronic ocular pain may have many causes and can be a frustrating problem for both patient and doctor alike. We describe two patients who had similar symptoms and eye findings who had been unable to relieve their pain with conventional analgesia. We postulate a cause for their pain and describe our experience of a treatment strategy using a standard dose of botulinum toxin injection into an extraocular muscle.

**Case 1**

A 56 year old white woman presented with right sided facial weakness, nausea, and right sided ptosis. She had a 9 month course of oral steroids and despite this needed tramadol, paracetamol, and flurbiprofen to control her pain. Her symptoms and examination findings slowly stabilised until she was left with marked limitation of upgaze in her right eye. Her symptoms did not change over the next 3 years, at which point she was referred to our care. When she attempted to look up she described a juddering sensation and severe pain just above the eye. She rarely had pain at night but was still using regular oral ibuprofen for pain relief. Her pain was exacerbated by reading or looking at the computer and she complained of vertical diplopia.

On examination she had limitation of abduction and elevation of her right eye and prisms did not improve her symptoms. A tentative diagnosis of inflammatory spasm was made. She was treated with botulinum toxin injection to her right inferior rectus. Two weeks later there was much less tightness and discomfort in the orbit but she still had diplopia in all positions of gaze and was forced to occlude one eye. Three months later the pain was much improved and she could no longer find the diplopia intolerable and declined further treatment.

**Case 2**

A 46 year old white man presented complaining of chronic constant ocular discomfort which followed strabismus surgery 8 years earlier for an A-pattern exotropia with diplopia on downgaze. The pain was worsened by prolonged television watching and prisms in his glasses did not help. Pain was much worse on upgaze and right gaze, which were limited. Ocular non-steroidal anti-inflammatory agents (NSAIDs) did reduce the pain a little but only when taken in high doses (100 mg three times daily flurbiprofen).

On examination he had a right hyperphoria, with an A-pattern exotropia and an abnormal head posture for distance. He still had diplopia. Botulinum toxin was injected into his left medial rectus muscle, which resulted in a profound reduction in his symptoms, leaving him with a small exophoria. His diplopia resolved completely after 10 weeks. The “pressure sensation” and pain in the right eye recurred after about 6 months, this time with no diplopia. He had a further injection of toxin 8 months after the first injection which again significantly improved his pain but gave him diplopia for 3 weeks. He continues to take flurbiprofen 50 mg three times daily orally.

**Comment**

The pain demonstrated by these two patients is typically much worse in certain directions of gaze and particularly during prolonged gaze holding such as when reading or watching television. It had a clear precipitating event and the most remarkable feature is that it had persisted for over 2 years in each case without significant progression or regression. No active disease process could be found to account for the continued pain. The pain is severe and responds only to high doses of analgesics, particularly NSAIDs. None of our patients felt that their pain was satisfactorily controlled by their analgesics.

We believe that there may be a process of chronic low grade inflammation affecting the extraocular muscles or the tissues around them which is exacerbated by continued contraction and relaxation of the same muscles. Muscular spasm perhaps triggered by this inflammatory process may be the cause of the most severe pain and this could account for the exacerbations of pain in certain directions of gaze and on prolonged gaze holding activities. Ocular muscle ischaemia, perhaps caused by constricting scar tissue, remains a possibility but the onset of the pain is very fast making this less likely.

The pain relief seen in our patients may simply be the result of paralysing an inflamed muscle but there is growing evidence for a separate antinociceptive effect of botulinum toxin.2 No direct peripheral cutaneous antinociceptive effect could be shown by Biersch et al.,6 however inhibition of release of substance P has been demonstrated in vitro and it can be hypothesised that botulinum toxin treatment may reduce the local release of nociceptive neuropeptides from either cholinergic neurons from C or A delta fibres in vivo.7 The mechanisms by which botulinum toxin may relieve pain, including a possible analgesic effect of botulinum toxin metabolites, are reviewed by Guyer.8 There is a growing literature on the use of botulinum for painful conditions,9 particularly those in which muscle spasm plays a part. These include writer’s cramp,9 postoperative pain in spastic cerebral palsys,10 and perhaps more surprisingly, canaliculitis and painful tic convulsivus.9 Many of the reported uses are single case studies and not all controlled trials have shown a positive effect of treatment.

It is not possible to rule out a powerful placebo effect in our patients but, whatever the mechanism of action, their pain was vastly improved and botulinum toxin treatment is very safe in competent hands.

In these cases described botulinum toxin served a dual purpose in that it had the potential to improve their ocular deviation for which it is well known and it also reduced the severe ocular discomfort. Unfortunately, the resulting diplopia limited its usefulness in one case but we feel that this treatment should be considered in this unusual group of patients who present a difficult management problem even to the most experienced ophthalmologists.

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**References**


www.bjophthalmol.com
Intrastral lamellar femtosecond laser keratoplasty with superficial flap

Lamellar keratoplasty has usually been performed taking a trephine to delineate the extent of the tissue to be excised, and a knife or similar instrument to remove the lamellar corneal tissue from the underlying deep corneal bed. In a similar way, the lamellar donor tissue was prepared and inserted into the recipient bed. The depth of the lamellar excision was chosen according to the thickness of the superficial stromal level was removed starting from the peripheral edge of the lamellar graft and the recipient corneal bed caused by irregularities of both surfaces. The purpose of the present report was to describe the new femtosecond laser technology, which may allow us to perform a new type of intrastromal lamellar keratoplasty with preservation of an intact Bowman’s layer and regular corneal epithelium.

Case report

Using a corneal contact lens and a femtosecond laser (20/10 Perfect Vision, Am Taubenfeld, D-69123 Heidelberg, Germany) with a wavelength of 1060 nm, a spot size of about 10 μm, and a laser pulse duration of several hundred femtoseconds, a pre-descemetal incision running parallel to the corneal surface was created in five postmortem eyes of slaughterhouse pigs. The diameter of the deep stromal incision was 7 mm. In a second step, a circular sagittal incision was performed starting from the peripheral edge of the already existing incision in the pre-descemetal level to the superficial layer of the corneal stroma. In continuation of the latter surgical step, the corneal flap was prepared with a diameter of 7 mm, a thickness of about 100 μm, a hinge, and three positional pikes. The pikes in the flap with the corresponding numbered hole of the flap were formed to increase the rotational stability of the flap after repositioning. The height of the peaks was about 0.40 mm. After opening of the flap the superficial segment situated between the pre-descemetal incision and the incision in the superficial stromal level was removed and exchanged against a similar formed segment obtained from another (donor) pig eye; the flap was repositioned.

For all eyes included in the study, the intrastromal corneal button and the superficial flap with the three positional pikes could be prepared without major difficulties. The corneal buttons could easily be repositioned into their original beds as well as into the recipient beds of other eyes in which the recipient beds were created with the same diameter as the donor button. The time taken for preparation of the intrastromal corneal button and the corneal flap, and for the exchange of the corneal buttons was less than 10 minutes in all cases.

Comment

Femtosecond laser technology allows a new type of intrastromal lamellar keratoplasty with removal of a mid-stromal segment and preservation of an intact Bowman’s membrane. Considering the decreased amount of allogenic corneal tissue transplanted, and regarding the preservation of the original corneal surface, lamellar intrastromal femtosecond laser keratoplasty may be associated with a smaller rate of immunological graft reaction and with a lower postoperative corneal astigmatism in some eyes. Future clinical studies may show whether positional edges in the superficial flap increase its postoperative rotational stability.

Proprietary interest: none

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References


Demographic study of paediatric allergic conjunctivitis within a multiethnic patient population

From October 1999, all patients referred to the paediatric ophthalmology service in Bradford have been added to a computerised database. This is the only paediatric ophthalmology service within the city of Bradford and receives all GP referrals of this type. Patients referred from both groups with severe disease, there was a relative prevalence of Asian children and white children. This difference in sex was not significant between both groups (Fisher’s test, p=0.71).

The overall age distribution for all males was 9.18 years and for all females was 10.01 years. For Asians, the mean age for males was 19.8 years and for females was 10.57 years. For white children, the mean age for males was 7.00 years and for females was 9.25 years.

Corneal complications

There were 14 with punctate epithelial erosions (10 Asians and four white children). Comparing patients from both groups with severe disease, there was a relative prevalence of Asians by 6.75 to 1 (Fisher’s test, p=0.001). In two cases, visual loss occurred after the onset of chronic allergic conjunctivitis from epithelial plaque and corneal pannus. Both were Asian.

Comment

Various studies have reported allergic eye disease to be more common among Asian and black patients. This may be due to genetic and/or environmental factors.

We found allergic eye disease to be more common in Asians than white children. It is possible that ocular allergy is multifactorial but perhaps with a greater genetic predisposition in certain ethnic communities. We could not comment on the prevalence of chronic allergic conjunctivitis in the community because of referral bias since we only see patients referred by GPs. The extent to which milder cases are treated in the community is not known but we feel that the more severe cases are the ones referred to our department.

Our findings highlight that allergic eye disease appears to be more common and complicated in Asian patients in the Bradford population. This potential risk of sight threatening disease means that they are more likely to require topical steroid treatment. This has led us to recognise that appropriately aggressive treatment is essential in these patients.

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CORRECTION

We wish to apologise for an error in the extended report by Barry and König (Br J Ophthalmol 2003; 87:909–16). On p 910 under the heading Orthoptic screening, point four of the bulleted list, line four should have read: “positive”: visual acuity $0.4 (10/25).

NOTICES

Helping the blind and visually impaired

The latest issue of Community Eye Health (No 45) discusses help for the blind, with an editorial by Sir John Wall of the Royal National Institute for the Blind on the rights of blind people. For further information please contact: Journal of Community Eye Health, International Resource Centre, International Centre for Eye Health, Department of Infectious and Tropical Diseases, London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK (tel: +44 (0)20 7612 7964; email: Anita.Shah@lshtm.ac.uk; website: www.jche.ac.uk). Annual subscription (4 issues) UK£22/US$45. Free developing country applicants.

Second Sight

Second Sight, a UK based charity whose aims are to eliminate the backlog of cataract blind in India by the year 2020 and to establish strong links between Indian and British ophthalmologists, is regularly sending volunteer surgeons to India. Details can be found at the charity’s website (www.secondsight.org) or by contacting Dr Lucy Mathen (lucymathen@yahoo.com).

SPEcific Eye ConditionS (SPECS)

SPEcific Eye Conditions (SPECS) is a not for profit organisation which acts as an umbrella organisation for support groups of any conditions or syndrome with an integral eye disorder. SPECS represents over 50 different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. The website acts as a portal giving direct access to support groups own sites. The SPECS web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECS contact: Kay Parkinson, SPECS Development Officer (tel: +44 (0)1803 524238; email: k@eyeconditions.org.uk; website: www.eyeconditions.org.uk).

The British Retinitis Pigmentosa Society

The British Retinitis Pigmentosa Society (BRPS) was formed in 1975 to bring together people with retinitis pigmentosa and their families. The principle aims of BRPS are to raise funds to support the programme of medical research into retinitis pigmentosa for this hereditary disease, and through the BRPS welfare service, help members and their families cope with the everyday concerns caused by retinitis pigmentosa. Part of the welfare service is the telephone help line (+44 (0)1280 860 363), which is a useful resource for any queries or worries relating to the problems retinitis pigmentosa can bring. This service is especially valuable for those recently diagnosed with retinitis pigmentosa, and all calls are taken in the strictest confidence.

Many people with retinitis pigmentosa have found the Society helpful, providing encouragement, and support. There is a Help line, the welfare network and the BRPS branches throughout the UK (tel: +44 (0)1280 821 334; email: lynda@brps.demon.co.uk; website: www.brps.demon.co.uk).

Surgical Eye Expeditions International

Volunteer ophthalmologists in active surgical practice are needed to participate in short term, sight restoring eye surgery clinics around the world. Contact: Harry S Brown, Surgical Eye Expeditions International, 27 East De La Guerra, C-2, Santa Barbara, CA 93101–9858, USA (tel: +805 963 3303; fax: +805 965 3564; email: hsbrown.md@cox.net or seeinl@seeinl.org; website: www.seeinl.org).

Rise in organ transplant numbers

According to UK Transplant, the UK has seen the highest number of organ transplants in 10 years. The number of organ transplants from 8:30 am to 5:00 pm at The Royal Glaucoma Society 24th Annual Meeting and Dinner

The Glaucoma Society 24th Annual Meeting and Dinner will take place on 20 November 2003 from 8:30 am to 5:00 pm at the College of Physicians, London, UK. Further details: Ms Janet Flowers (email: glauoc@uakeire.freeserve.co.uk).

Detachment Course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding the Annual Meeting of Iranian Society of Ophthalmology

The detachment course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding Annual Meeting of Iranian Society of Ophthalmology will be held...
on 29–30 November 2003 and 1–4 December
2003 respectively, at the Razi Conference
Center, Hemmat Hyw, Tehran, Iran. Further
details: Scientific programme: Prof Ingrid Kre-
issig, University of Tuebingen, Schleichstr. 12,
Breuningerbau, 72076 Tuebingen, Germany
(tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de). Local
organisation: Dr Arman Masheyekeyhi, Dr Sia-
mak Moradian, Dept of Ophthalmology, Lab-
banfinejad Medical Center, Pasdaran Ave,
Booostan 9, Tehran, 16666, Iran (fax: +98 21 254
9039; email: labbafl@hotmail.com).

5th International Symposium on
Ocular Pharmacology and
Therapeutics (ISOPT)

The 5th International Symposium on Ocular
Pharmacology and Therapeutics (ISOPT) will
take place 11–14 March 2004, in Monte Carlo,
Monaco. Please visit our website for details of
the scientific programme, registration, and
accommodation. To receive a copy of the Call
for Abstracts and registration brochure please
submit your full mailing details to http://

Further details: ISOPT Secretariat (website:

XVth Meeting of the
International
Neuro-Ophthalmology Society

The XVth Meeting of the International Neuro-
Ophthalmology Society will take place 18–22
July 2004, in Geneva, Switzerland.
Further details: Prof. A Safran, University
Hospital Geneva, c/o SYMPORG SA,
Geneva (fax: +4122 839 8484; email:
info@symporg.ch; website: www.symporg.
ch).