

Biomechanical Analysis of X-Pattern Exotropia

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- **PURPOSE:** To simulate and check the plausibility of the proposed mechanisms of X-pattern exotropia and to determine the least invasive surgical method that can be used to treat the disorder.
- **DESIGN:** Computational supported analysis and retrospective study.
- **METHODS:** The oculomotor model SEE++ was used to simulate the effects of the different causes that have been proposed for the X-phenomenon. In addition, a retrospective study was conducted using preoperative and postoperative measurements of 10 patients with X-pattern exotropia. Eye movements and surgery of these patients were simulated and analyzed statistically.
- **RESULTS:** Our computer-based simulations showed that only 1 of the 4 proposed theories can account successfully for the observed X-patterns: an overaction of all 4 oblique muscles can induce divergent exotropia in upgaze and downgaze, and an alteration of horizontal muscles can cause the additional divergence in all gaze positions. The simulation of eye muscle surgery confirmed that a sufficient correction of the divergent deviation in all gazes already can be achieved by a recession and resection of 2 horizontal eye muscles.
- **CONCLUSIONS:** In case of X-pattern exotropia, recession and resection of 2 horizontal muscles can be used as a first-line therapy, leading to a simplification of the therapy. (*Am J Ophthalmol* 2011;152:141–146. © 2011 by Elsevier Inc. All rights reserved.)

ALPHABET-PATTERN STRABISMUS, ALSO KNOWN AS A-pattern, V-pattern, X-phenomenon, or diamond-phenomenon strabismus, is characterized by a differing horizontal deviation in upgaze and downgaze. In case of exotropia, A-pattern strabismus shows an increase of the strabismus angle in downgaze, whereas V-pattern strabismus is characterized by an increased angle in upgaze. The combination of the 2, that is, exotropia with an increasing divergent angle in upgaze and downgaze, consequently is referred to as *X-pattern exotropia* or *X-phenomenon*. Because only a few patients

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have X-pattern exotropia, it is difficult to investigate what causes these symptoms and how such patients can be treated most effectively.

Regarding possible causes of X-pattern exotropia, 4 different explanations have been proposed. Jampolsky postulated an ipsilateral overaction of the oblique muscles as a possible reason for the X-phenomenon in unilateral large-degree exotropia. He suggested that overaction of all 4 oblique muscles may produce the increase of exodeviation in upgaze and downgaze.¹ A different cause was suggested by Wright: he proposed that a tightly contracted lateral rectus muscle could produce a leash effect, causing the observed X-pattern exotropia.² The third mechanism was suggested by de Decker: in patients with long-standing exotropia, he observed a slip of all 4 oblique muscles to the polar cap. This dislocation may amplify the abducting effect of the oblique muscles, and therefore may induce an increase of exotropia.³ In addition, in upgaze and downgaze, the lateral rectus muscle may slip above or below the eye and pull the eye up and out or down and out, thereby contributing to the X-shaped exotropia. A fourth possible explanation comes from observations by von Noorden: he reported that patients with X-pattern exotropia show a combination of oblique dysfunction together with horizontal deviations.⁴

The complex interactions of eye positions, pulling directions, and mechanical and neural contributions render purely intuitive interpretations of strabismus patterns highly complex and speculative. For example, the effect of an increase in stiffness of the lateral rectus muscle depends on the gaze position, on the relation of the center of eye rotation to the location of the muscle insertion, on the location and behavior of the corresponding muscle pulleys, and on the insertion width of the lateral rectus. To gain a better understanding of possible biomechanical causes of the X-phenomenon, we therefore decided to look at the predictions of computer-based simulations of the oculomotor plant for the 4 suggested causes of X-pattern exotropia. An overview of these hypotheses and the corresponding suggested treatments, as well as the results of our computer simulation of the mechanisms proposed by these hypotheses, is provided (Table).

Computer-based simulations of 3-dimensional eye movements have many adjustable parameters, so results based only on such simulations have to be treated with caution. We therefore combined our computer-based simulations with a retrospective pilot study of all X-pattern exotropia patients who sought treatment at our clinics over

TABLE 1. Comparison of Hypothetical X-Pattern Exotropia Mechanisms with Simulation Results and Suggested Therapies

X-Pattern Exotropia Hypothesis Author	Pathologic Mechanisms	Suggested Therapies	Simulation Results
Jampolsky	Bilateral overaction of all 4 oblique muscles	Surgical treatment of all 4 oblique muscles	Little exotropia in primary position
Wright	Contracted lateral rectus muscle(s)	Recession of lateral rectus muscle(s)	No change in upgaze or downgaze
de Decker	Slip of all 4 oblique muscles to polar cap	No general treatment suggestions	Not simulated, for lack of specific information
van Noorden	Cumulative oblique-dysfunction and horizontal deviations	Surgical treatment of all 4 oblique muscles, or surgical treatment of horizontal rectus muscles	X-pattern can be reproduced, with different options for horizontal deviation

the last 20 years and the effects of the horizontal rectus surgeries that have been used to treat those patients. Our simulations of the oculomotor mechanics allow us for the first time to simulate the effects of the suggested causes of X-pattern exotropia and to analyze the mechanics behind the X-phenomenon.

METHODS

• **SIMULATION:** In the last decade, 2 computer-based models of the oculomotor system have been commercially available. The first one was Orbit (Eidactics, San Francisco, California, USA), which has been developed for the Mac (Apple, Inc, Cupertino, California, USA). An updated, Java-based version is under development, but currently is not available. The second commercial oculomotor model, SEE++, has been developed for PCs (RISC Software GmbH, Hagenberg, Austria). The simulation results of SEE++ have reproduced successfully the effects of surgeries for simple operations, as well as for biomechanically complex pathologic conditions.^{5,6} Our simulations of the suggested biomechanical causes of X-pattern exotropia were carried out with SEE++ release 7.4.

• **SUBJECTS:** Strabismus patients with X-pattern exotropia are very rare. Over a period of 20 years, only 10 patients with X-pattern exotropia qualified for inclusion in our study. The mean age \pm standard deviation of these patients was 31.7 ± 15.2 years. The best-corrected logarithm of the minimal angle of resolution visual acuity on the better eye was 1.0 ± 0.1 , and that of the weaker eye 0.4 ± 0.4 . Five of these patients previously had undergone squint surgery. A statistical comparison of strabismus angles between the 2 groups showed no significant difference between patients with and without squint surgery. We therefore pooled the 10 cases and treated them as 1 homogenous sample.

The strabismus angle was measured using the prism cover test under optimal refractive correction. Measurements were performed before surgery, 2 days after surgery,

and 4 months after surgery. Patients with additional ocular pathologic features were excluded. For achieving the best results when comparing simulations with field data, the preoperative and postoperative measurements were obtained directly as input parameters for the computer simulations.

• **SURGICAL TREATMENT:** The goal of the surgical treatment of these patients was to reduce the horizontal deviations to a level acceptable to the patients, not to eliminate the X-pattern exotropia. To achieve this goal, only the horizontal extraocular muscles were operated on.

In 9 patients, common recession and resection was performed for divergent deviation. In 1 case, a bilateral recession was performed. The dosage of recession was 5.1 ± 1.3 mm, and that of resection was 4.8 ± 3.0 mm. The dosage of bilateral recession of the lateral rectus muscle was 7 mm. The expected effect of muscle displacement by 1 mm was calculated with 2.1 ± 0.4 degrees (4 ± 0.7 prism diopters).

• **STATISTICAL ANALYSIS:** Because of the small sample size of 10 patients, our statistical analysis focused on a comparison between the results of surgery and of the simulation on the mean and median levels of the strabismus in all gaze positions. The statistical tests included the 2-sided *t* test for paired data as well as the Wilcoxon rank-sum test (Mann–Whitney *U* test) for nonparametric analyses.

RESULTS

• **COMPUTER-BASED SIMULATION OF THE BIOMECHANICAL CAUSE OF X-PATTERN EXOTROPIA:** Figure 1 shows the average strabismus angles found in patients (Left), as well as the simulation results for the overaction of the oblique muscles (Middle left and Middle right) and of a tether effect of the horizontal muscles (Right). For the simulations of X-pattern exotropia, muscle forces were changed by increasing or decreasing the active and passive

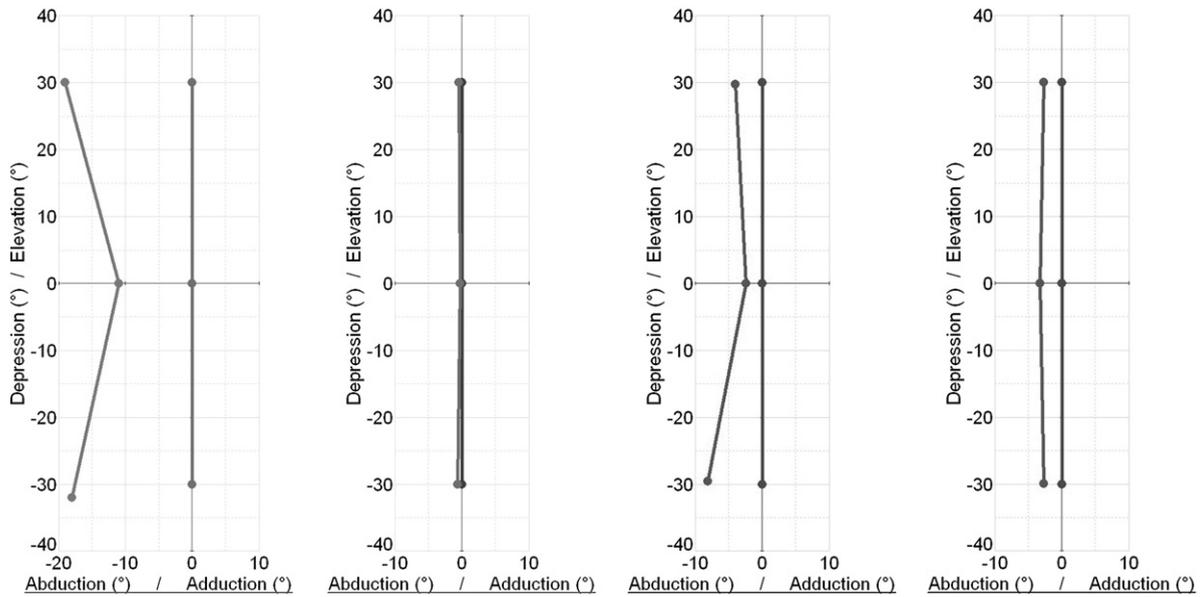


FIGURE 1. Computer-based simulation of X-pattern exotropia in comparison with patient data. (Left) Preoperative measurements of patients with X-phenomenon in primary position, upgaze, and downgaze. (Middle left) Simulated increase of passive strengths of all 4 oblique muscles does not result in a typical X-pattern. (Middle right) Increase of the active muscle strengths of all 4 oblique muscles induces the characteristic X-shaped deviation, but no significant divergent deviation in primary position. (Right) Simulated tether effect with contraction of the lateral rectus muscle showing no typical signs of X-phenomenon.

muscle strengths. The active muscle strength corresponds to the muscle force induced by neural activity; the passive muscle strength describes the contribution of the muscle stiffness and elasticity to the total force exerted. To simulate the hypothesis of Jampolsky of an overaction of all 4 oblique muscles as a possible cause of the X-phenomenon, we first tested the effect of an increase in passive muscle strength of all 4 oblique muscles.¹ However, this did not result in a typical X-phenomenon (Figure 1, Middle left). In contrast, an increase of the active muscle strengths of all 4 oblique muscles, comparable with a muscle overaction, induced typical X-pattern exotropia (Figure 1, Middle right). The second mechanism suggested for the X-pattern is a tether effect, a leash effect induced by a tightly contracted lateral rectus muscle on upgaze and downgaze. We simulated this effect by increasing active and passive forces for the lateral rectus. The simulation results showed no typical signs of the X-phenomenon (Figure 1, Right). None of these hypotheses is able to explain the pronounced divergent deviation, which is typical for the X-phenomenon and which was present in all our patients. The hypothesis of de Decker, a “slip of all four oblique muscles to the polar cap,” could not be simulated because he provided no specific information on this proposed muscle slip.³

The fourth hypothesis for explaining X-patterns, by von Noorden, is a cumulative incidence of oblique dysfunction together with horizontal deviations. The overaction of the oblique muscles is simulated as above (Figure 1, Middle right). Horizontal deviations can be in-

creased in different ways, through modifications of the properties of the horizontal rectus muscles. Figure 2 shows the effect of varying the muscle lengths of the horizontal rectus muscles (Left), their strength (Middle), or their insertion points (Right). Note that these changes are superposed on the increased muscle strengths of the 4 oblique muscles.

• **SURGICAL RESULTS:** All our patients showed X-pattern exotropia, with an increase of divergent deviation in upgaze and downgaze. The mean \pm standard deviation strabismus angle in primary position was -19.0 ± 10.3 degrees (-33.16 ± 18.0 prism diopters [PD]). The strabismus angle in upgaze and downgaze was -26.6 ± 12.8 degrees (-46.4 ± 22.3 PD) and -27.5 ± 12.4 degrees (-48 ± 21.6 PD). The changes in strabismus angles in upgaze and downgaze were significantly different from looking straight ahead ($P = .027$ and $P = .012$, respectively).

The surgery on the horizontal oculomotor muscles resulted in a statistically significant reduction of the strabismus angle in primary position as well as in upgaze and downgaze ($P < .05$). The postoperative measurements of the strabismus angle showed an angle of -5.3 ± 6.8 degrees (-9.3 ± 11.9 PD) in primary position and of -9.3 ± 9.2 degrees (-16.2 ± 16.1 PD) and -9.0 ± 10.5 degrees (-15.7 ± 18.3 PD) in upgaze and downgaze, respectively.

The measurement results of the strabismus angle before and after surgery are given as box plots in Figure 3. Before

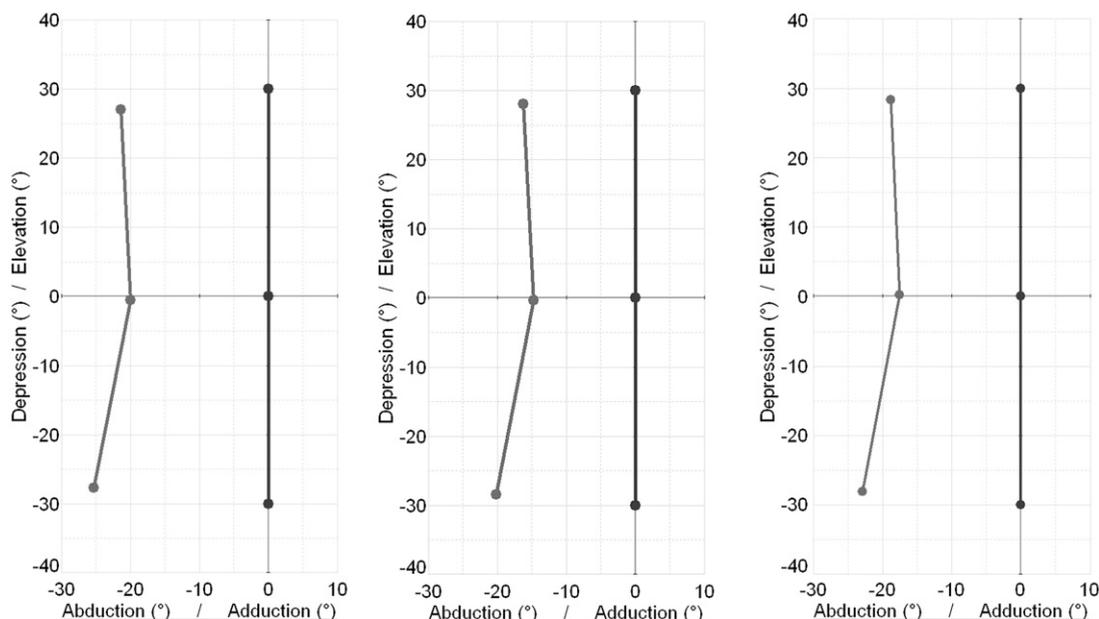


FIGURE 2. Different effects can cause a horizontal deviation. In each figure, the line on the right side stands for the fixing eye, and the line on the left side represents the deviating eye. (Left) Alteration of muscle lengths (here an elongation of the medial rectus muscle by 6 mm and a shortening of the lateral rectus muscle by 5 mm) can reproduce the observed horizontal deviation and maintains the X-pattern. (Middle) Alteration of muscle strengths (here an increase of the medial rectus strength by 10% and of the lateral rectus strength by 300%) has a similar effect. (Right) The same deviation also can be produced through a displacement of the muscle insertions (here a recession of the medial rectus by 5 mm and an advancement of the lateral rectus by 5 mm).

surgery, the strabismus angle was much more variable and showed a pronounced left-skewed distribution (Figure 3, Left). The X-pattern is clearly visible by the increase in the median strabismus angle in upgaze and downgaze. After surgery, the patients still show the characteristic X-pattern in upgaze and downgaze. However, the median strabismus angle was reduced strongly, as was the variability (Figure 3, Right). The 95% quantile shifted from approximately -8 degrees (-14 PD) to approximately $+5$ degrees ($+8.7$ PD).

DISCUSSION

THE MAIN GOAL OF OUR RETROSPECTIVE PILOT STUDY was the investigation of possible biomechanical causes of the X-pattern exotropia and the best way to treat patients presenting with such symptoms. Our simulation results indicate that 3 of the 4 suggested causes of the X-pattern cannot sufficiently account for the observed symptoms. In 1965, Jampolsky postulated an overaction of the oblique muscles as a possible cause of the X-phenomenon.¹ Our simulations indicate that an increase in the contractile forces of all 4 oblique muscles in fact can cause the typical X-deviation, with an increase of exotropia in upgaze and downgaze. However, the considerable exotropia observed in patients also in the primary position could not be elicited in this way.

The second hypothesis that we tested was formulated by Wright, who speculated that tight lateral rectus muscles may lead to a leash effect, causing exotropia in upgaze and downgaze.² To simulate this theory, the active and passive strengths of the lateral rectus muscle were increased. The outcome of the simulations showed no typical X-pattern deviations. To reproduce the observed significant divergent deviations in primary position as well as in upgaze and downgaze, we had to alter the activity of the horizontal rectus muscles. This way, different parameters could be modified to obtain the horizontal deviations. However, changes of the horizontal recti alone produced only minimal X-shaped deviation in upgaze and downgaze, and not the observed pronounced X-pattern.

Hence, the observed X-shaped deviations in our patients could be generated only through the fourth hypothesis, an overaction of the oblique muscles combined with a modification of the activity of the rectus muscles. The finding correlates nicely with a report of von Noorden, who found a cumulative incidence of oblique dysfunction together with horizontal deviations.⁴

The second part of our investigation was the retrospective analysis of patients and the results of surgical interventions in patients who had X-pattern exotropia. We performed the study as a retrospective study, because X-pattern exotropia is a rare clinical occurrence, with only 10 patients over 20 years qualifying for inclusion in our

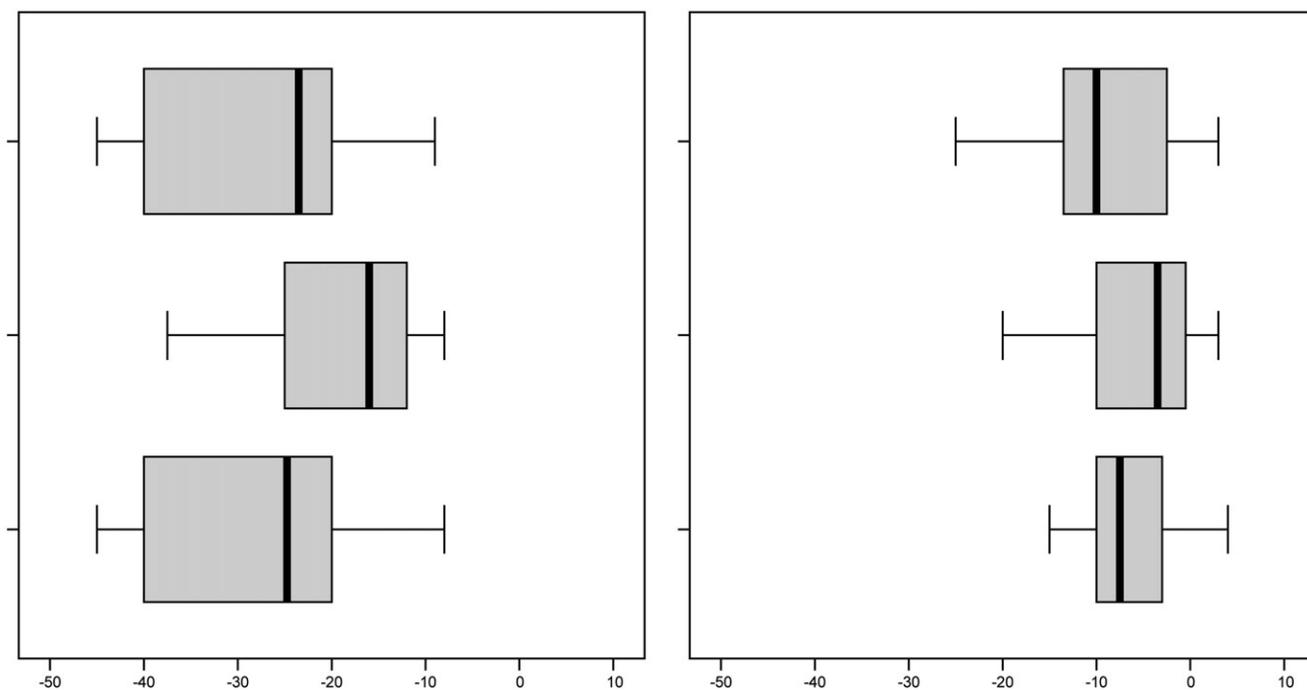


FIGURE 3. Box-and-whisker plots comparing preoperative and postoperative strabismus angles in upgaze, primary position, and downgaze (top to bottom). The whiskers represent the 5% and 95% quantiles. (Left) Before surgery, the strabismus angle is much more variable and shows a pronounced left-skewed distribution. (Right) After surgery, we still find the characteristic X-pattern in upgaze and downgaze, but the strabismus angle is reduced considerably.

study. Unfortunately, because this type of patient is seen only rarely, we had to include patients with and without previous squint surgery to obtain the already small number of 10 patients.

In patients with X-phenomenon caused by an overaction of the oblique muscles, Jampolsky and Limón de Brown postulated good results through surgery of the horizontal eye muscles.^{1,7} After correction of exotropia, dysfunction of the oblique muscles was no longer apparent. This also was our preferred way to treat patients with X-pattern exotropia. The results of our retrospective study show that surgery on the horizontal oculomotor muscles with recession and resection resulted in a statistically significant reduction of the divergent deviations in primary position as well as in upgaze and downgaze. Consistent with our biomechanical simulations, a significant but reduced X-phenomenon re-

mained. The postoperative measurements of the strabismus angle resulted in a reduction of approximately two thirds of the preoperative angle in primary position, upgaze, and downgaze. Because all patients were happy with the results of the operation, no further surgery was performed.

Our results agree with the previous observations by von Noorden.⁴ In case of an abnormal overaction of all 4 oblique muscles, he suggested a combined surgery of the horizontal and oblique muscles. Our results indicate that in patients with high exodeviation in primary position and with a low increase of strabismus angle in upgaze and downgaze, surgery on the horizontal muscles alone may be sufficient and may simplify the treatment. Given the low incidence of X-pattern exotropia, a multicenter study may be the best way to confirm our hypothesis on a larger number of patients.

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Gray's *Anatomy* is unique in medicine in that it has been continuously in print and read (although much changed) in over 40 editions since its initial publication in 1858. It was the work of two young men not long out of medical school – Henry Gray, who dissected and wrote the text, and Henry Vandyke Carver, who also dissected, and drew the illustrations. The full title of the work is *Anatomy, Descriptive and Surgical* but the spine was imprinted as *Gray's Anatomy*, thus shutting Carver out of equal memory.

In the highly competitive culture at St. George's Hospital, Gray had started to make a name for himself even before embarking on the *Anatomy* by entering the competition for

the 1848 Triennial prize of the Royal College of Surgeons, which had set the research topic to be the nerve origins of the human eye as illustrated by comparative vertebrate anatomy. Gray won the award and then had the further good fortune to have his paper read before the Royal Society by the famed ophthalmologist William Bowman.

Gray's promising career was cut short at age 34 when he died of smallpox in 1861.

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Richardson R. *The Making of Mr. Gray's Anatomy*. Oxford: Oxford University Press, 2008.

Submitted by Ron Fishman of the Cogan Ophthalmic History Society.