We are honored to have Joseph Lang as our ninth Costenbader Lecturer. Professor Lang studied medicine in Zurich under the guidance of Marc Amsler who interested him in the field of ocular motility. He is Professor and in charge of the Orthoptic Department of the University Eye Clinic in Zurich, and for the past 8 years, Secretary of the European Strabological Society.

His book, Strabismus, has two editions in German, one in Spanish and French. His book on Microtropia recently had its second edition. He was awarded the Vogt-Award of the Swiss Ophthalmological Society in 1974 for work on microtropia. His papers are benchmarks of original observations and thought, especially in our understanding of sensory adaptations to strabismus.

The Costenbader Lecture is presented annually at the AAPOS&S meetings in memory of Frank D. Costenbader (1905–1978) of Washington, D.C., the father of pediatric ophthalmology in the USA. Beyond recognizing his many contributions to knowledge of strabismus, we memorialize also his kindness to patients, his helpfulness to colleagues, and his inspiration to a generation of ophthalmologists. The lecture was initiated in 1974 by the Costenbader Alumni Society and adopted by the American Association of Pediatric Ophthalmology and Strabismus at its first meeting in 1975.

Costenbader Lectures

1974 Marshall M. Parks
1975 Robert N. Shaffer
1976 Lorenz E. Zimmerman
1977 T. Keith Lyle
1978 Jules Francois
1979 D. Robison Harley
1980 David G. Cogan
1981 Philip Knapp
SPECIAL FORMS OF COMITANT STRABISMUS

J. Lang

ABSTRACT

This lecture is concerned with some special forms of comitant strabismus. The congenital strabismus syndrome is characterized by early onset, latent nystagmus, dissociated vertical divergence, excyclorotation of the nonfixating eye and anomalous head posture. Microtropia is an inconspicuous strabismus of less than 5 degrees, showing no motor but only sensorial defect with harmonious anomalous correspondence. Normosensorial late convergent strabismus is a motor form of strabismus with acute, intermittent onset and normal retinal correspondence showing excellent surgical prognosis. Theoretical and practical importance and frequency of these forms are discussed.

Three new diagnostic methods are also discussed: the two pencil-test, the fourth image of Purkinje and the new Lang-Stereotest.

INTRODUCTION

First I would like to thank the officers of the American Association for Pediatric Ophthalmology and Strabismus for the kind inviation to give the Costenbader Lecture. If is, of course, a great honor and pleasure for me to speak in front of such a famous audience. It is also a pleasure for me to discuss with you some of my views on strabismus problems, which to you may have some European flavor.

I had the great privilege to meet Frank Costenbader personally twice, for the first time at the First International Strabismus Symposium in Giessen in 1966, and for the second time at the First Congress of the International Strabismological Association in 1970 in Acapulco. I had the good fortune to have a long discussion on strabismus with him on our pleasant two-day bus trip from Mexico City to Acapulco. I was deeply impressed by the wisdom, the modesty and the honesty of this great man. He confessed that he regarded himself not as a scientist, but as an ophthalmological practitioner devoted to the welfare of his patients. Knowing him personally made me gladly accept
the honor and the task of speaking to you. I know that Dr. Costenbader would have appreciated that I prefer to speak to you more on clinical experience than on scientific aspects of strabismus.

I would like to discuss with you three clinical entities, namely the congenital strabismus syndrome, microtropia and normosensorial late strabismus. Together with accommodative strabismus they represent four model forms which can give a better survey on the vast field of convergent strabismus.

I would also like to offer you three small diagnostic pearls, namely the two-pencil test, the forth image of Purkinje and the new Lang-Stereotest.

CONGENITAL STRABISMUS SYNDROME

In 1961, Costenbader(1) published his thesis on infantile esotropia which means an esotropia known to be present before the age of one year. Prior to that(2) he had defined congenital esotropia as "strabismus present by 6 months of age, characterized by a large deviation unresponsive to spectacle treatment of the existing hyperopia". But do congenital cases always show a large deviation? Are there other signs which characterize congenital convergent concomitant strabismus?

At the first International Congress of orthoptists in London in 1967, I(3) discussed a very simple question: Do children who squint from birth or from the first few months of life show a characteristic form of strabismus which may be differentiated from other forms?

At that time, in 82 such cases, I found 92% with dissociated vertical divergence, 57% with latent nystagmus, 65% with excyclorotation of the nonfixing eye and 70% with abnormal head posture. Twenty percent showed A-pattern, 17% showed V-pattern and 15% had, in addition to the strabismus, cerebral damage. The percentage of these characteristics may vary, but I have found the picture as a whole, to have been confirmed ever since that time.

The different characteristics could best be seen in a short film. In this film a girl with congenital strabismus syndrome was shown at the age of 8 months and at the age of 10 years. She was operated on when she was 6 years and 7 months (No. 106 in Malcolm Ing's thesis).(4) At the age of
10, the two pencil test was positive when both eyes were open, but negative when the deviating eye was covered.

Dissociated vertical deviation was first described by Bielschowsky(5) and may also be called occlusion hypertropia. The eyes may seem to be vertically aligned, but the occluded eye slowly drifts upward and after uncovering may drift down again. The darkening phenomenon of Bielschowsky shows the same movements. This phenomenon is of innervational origin and must not be confused with elevation in adduction as seen in V-patterns which sometimes is also called alternating sursumduction. Dissociated vertical divergence is not usually present in the first year of life, but appears in congenital strabismus only at the age of two years.

The second sign of the congenital strabismus syndrome is latent nystagmus. If one eye is occluded jerking nystagmus with a rapid phase to the side of the fixating eye is observed. This jerking nystagmus sometimes is present when both eyes are open, and from it the fixating eye can be recognized in small angles: the rapid phase of the nystagmus is always directed to the fixating eye. In addition, there may be a rotatory component of the nystagmus. Latent nystagmus must be carefully differentiated from congenital pendular nystagmus. Its correlation with strabismus is very high. I once found that in patients with latent nystagmus almost 99% have strabismus, whereas with congenital pendular nystagmus only 30% have a strabismus and between them mainly cases with albinoti fundi. This differentiation has never been made by the advocates of the nystagmus-blockage syndrome - an entity which anyway seems to lose its importance.

Cyclorotational movements are often seen in the congenital strabismus syndrome: the eye that takes up fixation makes an incyclorotation and the eye that relinquishes fixation makes an excyclorotation. Sometimes spontaneous rotational movements are seen.

Abnormal head posture is often seen in the congenital strabismus syndrome. Usually the head is tilted towards the shoulder of the fixating eye and the face is also turned to this side. All these signs must be carefully looked for, be it by slit lamp, by carefully watching rotation movements on the iris, and by funduscopy. Recently, I had the opportunity to examine these cases on the television monitor of an infrared fundus camera. It is impressive how the whole picture of the congenital strabismus syndrome is even more
We call this picture a syndrome because it fulfills the conditions of a syndrome. A syndrome should be a group of apparently unrelated symptoms and signs which have the tendency to appear together and to characterize a clinical entity. This congenital squint syndrome largely corresponds to the entity described in 1954 by Crone(6) as alternating hyperphoria, in 1959 by Anderson(7) as latent nystagmus and alternating hyperphoria and in 1962 by Ciancia(8) as esotropia in infants with limitation of abduction.

What is the pathogenesis of the congenital squint syndrome? Nobody knows for sure. We once thought it had its seat in the brain stem and was due to an imbalance between vestibular and visual influence upon the oculomotor mechanisms. Now we believe that it may be due to an imbalance between the geniculo-striate and the extra-geniculo-striate system and that latent nystagmus is the dominating feature of this syndrome. Clinical observations and neuroanatomical facts support this hypothesis.

Clinical observations show that in almost 99% of cases with latent nystagmus, a strabismus is present. Cases with orthotropia and latent nystagmus are extremely rare. In infants with the congenital strabismus syndrome both eyes are in a convergent position at first. On ophthalmoscopical examination of fixation, the fixation object does not lie in the fovea but on the nasal side of the fovea. Only gradually does fixation move into the foveola. We got the clinical impression that only when fixation has moved to the foveola, latent nystagmus can manifest itself. The fixation object in the nonoccluded eye slowly moves from the foveola to the nasal side and by a saccade returns to the foveola. Imbalance of optokinetic nystagmus may result from this. In some adult cases, one can see that in binocular vision the leading eye does not fixate exactly with the foveola, since when the squint eye is occluded, an adjustment movement of the fixating eye can be observed.

Concerning the neuroanatomical facts, one must bear in mind that beside the retino-geniculo-striate system other pathways exist, the most important being the extra-geniculo-striate pathway to the colliculus superior and to the pulvinar and from there to the cortex, as shown in the diagram by Trevarthen and Sperry.(9) This second visual pathway is claimed to be responsible for ambient vision.
Furthermore Bernheimer(10) in 1899 demonstrated that 20 to 30% of the fibers of the optic nerve do not reach the corpus geniculatum laterale, but pass to the prectectal and tectal area. According to Minkowsky(11) these are probably only uncrossed fibres which stem from the nasal parts of the retina. In addition, it is known that, in the early stages of human fetal development, the primitive phylogenetic arrangement of a complete decussation of the nerve fibers occurs in the chiasm. It is not until the 11th week that uncrossed fibers begin to appear.

Keeping in mind that congenital strabismus is often seen in cases of prematurity or of cerebral damage, we may expect that in these cases the phylogenetic older extra-geniculo-striate system, based on the nasal halves of the retina and on the crossed optical fibers subserving ambient vision, is dominant. This accounts for the convergent position of the eyes and for the tendency to fixate in the nasal part of the retina. As visual development goes on in the first months of life, fixation moves into the fovea to fulfill the task of discriminating forms.

When both eyes are open, no nystagmus is seen since opposite movements of the eyes are counterbalanced. When one eye is occluded counterbalance does not exist any more. Fixation then slowly glides from the foveola to the nasal part of the retina due to the tendency of ambient vision, and then quickly returns to the foveola again (Fig 3), bringing back the object in the area of discrimination.

I am fully aware that this is a hypothesis based only on clinical impressions and on possibly insufficient neuroanatomic facts.(12) But since nowadays everybody is speaking mostly of the work by Hubel and Wiesel on the geniculo-striate system, I find it worthwhile to mention other works and pathways. Maybe this view also explains why patients with congenital esotropia cannot be cured just by aligning them even by the most sophisticated surgical techniques.

MICROTROPIA

But now let us go to another clinical entity which may seem less important, but which is interesting for the whole pathogenesis of strabismus, namely microtrophia.(13)

Microtrophia or microstrabismus may be defined briefly as
fig. 1: Latent nystagmus. a. both eyes open - no nystagmus. b. right eye covered - nystagmus to the left. c. left eye covered - nystagmus to the right.

fig. 2: From Trevarthen and Sperry: extra-geniculo-striate pathway.

fig. 3: Latent nystagmus as seen in the fundi.
an inconspicuous strabismus of less than 5 degrees, usually with harmonious anomalous correspondence. Three forms can be distinguished: primary constant, primary decompensating and consecutive microtropia, which has also been called secondary microtropia.

There are three situations in which the ophthalmologist may be confronted with microtropia:

a. in amblyopia apparently without strabismus (In the past this has usually been diagnosed as congenital amblyopia);

b. in hereditary or familial problems of strabismus;

c. in residual strabismus after treatment, which each of us sees more often than he would like to.

Microtropia does not seem to have been familiar to well-known early strabismologists as Javal, Worth, Duane and Bielschowsky. Let us quote Maddox(14) who in 1898 wrote that "minute squints were exceedingly rare, the natural desire for single vision being too strong to allow minute squints to exist".

Observations of small angles have been made only in the last few decades. There were plenty of theories on the causes of this condition and on the ways of avoiding it, be it by orthoptics, by prisms, by surgery and/or by other sophisticated treatment. Here once again strabismologists have fallen into the same old trap. Before knowing the nature of a condition one already knows where it comes from and how to treat it.

But before dealing with consecutive microtropia, where treatment can produce almost every sensorial perturbation, except, of course, a real cure, one should try to study and to learn as much as possible from primary microtropias. In primary microtropia no treatment has changed the original sensory conditions and more insight may be gained into a fascinating experiment of primary binocular pathology made by nature itself.

As far as I know, cases of primary microtropia were described for the first time by Jampolsky(15) in 1951 under the name of fixation disparity. I will not go further in details about terminology of microtropia, be it retinal slip, cortical slip, fusion disparity, ultrasmall angle, eso flick
or other terms. But since this condition is, in the US, mostly called Monofixation Syndrome, I will say a few words later on about that.

For many years I have carefully looked for primary microtropias in my practice. Out of a total of 41,678 there were 7,751 motility patients who were listed on special punch cards. In 3,338 cases of convergent strabismus there were 1,176 total microtropias and 626 cases of primary constant microtropia, which means 18.7%. Whereas in convergent strabismus, I found 35.2% microtropia, in divergent strabismus there were only 3.3% of microtropias. In order to understand these figures I should add that I worked as a general ophthalmologist at that time and only later became a full-time strabismologist. From these numbers I estimate that 1% of the general population in Switzerland has a microtropia.

The most important characteristic of microtropia is amblyopia. What are the clinical findings in microtropic amblyopia? In 113 children with untreated primary microtropia we found 69% with isometropia whereas 31% had an anisometropia. Again we found 51% with central fixation whereas 49% had eccentric fixation. In isometropia central fixation prevailed with 58%, whereas in anisometropia central fixation was found in only 37%. Vision was best in cases with isometropia and central fixation, the mean value being 0.58. Vision was worst, as can be expected in anisometropia with eccentric fixation, the mean value being 0.08. Isometropia with eccentric fixation and anisometropia with central fixation range in between.

We have done a lot of interesting studies in adult microtropias on the nature of fixation behaviour, on the nature of the scotomas and on reading capacity, but time does not allow us to go into details. More details can be found in the second edition of my monograph on microtropia.

Let us come to hereditary problems of microtropia. Since the times of Hippocrates one knows that squinters stem from squinters and it has recently become apparent, that microtropia here plays a role. Ogge and Rochels(16) recently published a pedigree of a family with three generations and 23 members, where 10 members had a primary microtropia and 4 a large angle strabismus. I would also like to report on three sisters (Fig 4). The youngest and the eldest sister started to squint at the age of 3 years. Both had a hyperopia of about 3.5 diopters and an amblyopia
of one eye. After correction of hyperopia and treatment of amblyopia a consecutive microtropia resulted. The second girl never squinted, but at the age of seven years a microtropia of the left eye was found with amblyopia of 0.2.

I am sure that all of you have seen similar situations and agree with me that microtropia plays an important role in hereditary factors in strabismus, although we do not yet know the exact hereditary mode.

This example also shows the importance of microtropia in the pathogenesis of large angle strabismus.

We all assume that there are some factors which cause an aligned patient to develop strabismus. When this is true for parallel cases, this must even more be probable in cases with microtropia, since there exists a primary disturbance of binocularity. Factors which predispose to decompensation may be a hyperopia, a convergent position of rest, convergence excess and an amblyopia. Such factors may also be combined. From there results a large convergent deviation. Cases with this mechanism may be suspected when shortly after the onset of strabismus a deep amblyopia already exists, or when in intermittent cases in the apparent parallel position, there is microtropia or eccentric fixation. Since anomalous correspondence exists already, it can easily adapt to the large angle. After successful treatment, be it by glasses, orthoptics or surgery, not orthotropia, but the preexisting microtropia shows up again. This supplies a simple answer to the question, why even after careful treatment not an orthotropia but a consecutive microtropia usually results.

Now I may be allowed to explain differences which exist between microtropia and the monofixation syndrome of Marshall Parks. I have, of course, studied carefully Marshall Parks fine thesis on the monofixating syndrome.(17) In analysing the 100 cases in his thesis I interpreted 12 cases as primary microtropias, 59 cases as consecutive microtropias, 5 cases as amblyopia ex anisometropia, 6 as stereoamblyopias, 6 as fully accommodative cases, 8 as cases of normosensorial late strabismus, 3 as cured intermittent divergence and 1 as a macular lesion. All these different conditions have one symptom in common, namely subnormal stereopsis with supression of one eye. By definition microtropia describes only cases with manifest deviation, discovered by the cover-test or by eccentric fixation, whereas the monofixation syndrome includes also cases with orthotropia.
fig. 4: Three sisters, two with large angle strabismus, one with primary microtropia.

fig. 5: Decompensation of primary microtropia into a large angle and return to microtropia after treatment.
This may be explained by the example of anisometropic amblyopia, where there is no deviation, whereas in microtropia there exists a deviation of less than 5 degrees. In anisometropic amblyopia a normal retinal correspondence exists, whereas in microtropia usually there is a anomalous correspondence. Amblyopia ex anisometropia is essentially a refractive condition and must be treated mainly by optical correction whereas microtropia is primarily a sensorial defect and its amblyopia should be treated by occlusion. Of course, both conditions may be combined, as a cataract can be combined with a maculopathy – but they should all the same be differentiated as carefully as possible.

I know the opinion that in microtropia the presence of an anomalous correspondence is not shared by all strabismologists. This difference is most probably due to different examinations. I, myself, have relied mostly on ophthalmoscopical examination of correspondence and I can not just neglect my findings. I have also developed a photographic method to show anomalous correspondence.(18) I admit that this is, of course, easier done in adults than in children. But I think that results from these studies are also valid for children. Curiously enough nowadays results from animals, such as cats and monkeys, who naturally do not squint – are readily accepted for humans, whereas results from strabismic adults are not regarded as valid for strabismic children.

THE FOURTH IMAGE OF PURKINJE

Now I would like to speak about two diagnostic methods which I have developed in the last years(19) and which to me are real pearls and which may help to further clarify some points of discussion. Hirschberg's method of assessing strabismus by using the first image of Purkinje is well known, but so is the limitation of this test for small angles. By chance I discovered that by using the fourth image of Purkinje, the one resulting from the posterior surface of the lens, our discrimination threshold for small angles can be sharpened. The fourth image of Purkinje is very weak but can be seen in photographs when these are centered exactly on the plane of the iris. The patient should fixate in the objective and the electronic flash must be positioned exactly vertical above the objective. The fourth Purkinje image can then be seen below and temporal from the first reflex. In cases of microtropia the fourth image is less temporal, as can be seen in a case of microtropia. Once you are familiar with this method, you
automatically check every photograph for the fourth image of Purkinje.

LANG STEREOTEST

The other test I would like to acquaint you with is the new Lang-Stereotest (19) which is a combination of the random-dots of Julesz (20) with cylinder screens. Most of you know cylindrical screens or panography from postcards where the attraction does not exactly correspond to their ethical value. But nobody knows that these cylindrical gratings were invented by the same Walter Robert Hess (21) who had described coordimetry for examination of eye muscles. Beneath each fine cylindre there are two fine strips of pictures (Fig 7).

I have combined these two methods which have the advantage that no glasses are necessary for the examination. Children do not always easily accept glasses or time must be spent to make them accept them. Since my test works without glasses, examination in young children is made easier and quicker. Three objects are seen binocularly: a cat, a star and a car. Children may name these objects, or they may point to them or by observation of fixating movements of the eyes a positive results may be found. Interestingly enough, children recognize the figures usually quicker than adults (Fig 8).

In examining about 1000 cases, I have found that patients with anisometropic amblyopia may pass this test whereas children with microtropia usually fail this test. This is, of course, no hard and fast rule.

This leads us to the question if the patients with constant strabismus have binocular stereopsis. The answer again depends on the methods we use.

TWO PENCIL TEST

When using the two pencil test (22) (Fig 9), which is based on large disparity of gross objects (Fig 10), we can even find in the congenital strabismus syndrome a binocularity. The examiner holds a pencil vertically in front of the patient. The patient is asked to hold a second pencil, bottom down, vertically above the examiner's pencil and to bring his pencil slowly down in an attempt to touch the examiners pencil. This test should be done first with both eyes open, then with the weaker of deviating eye occluded. The binocular and monocular performance are then
fig. 6: The fourth image of Purkinje in a case of microtropia. a: both eyes open - microtropia of the right eye. The fourth image of Purkinje is on the nasal side of the first in the right eye and on the temporal side in the left eye. b: Left eye covered - adjustment movement of the right eye. The fourth image is now temporal below the first image.

fig. 7: Principle of cylinder screens.
fig. 8: The three pictures of the Lang Stereotest.

fig. 9: The two pencil test. a: with both eyes open, b: with one eye covered.

fig. 10: The two pencil test represented in the fundi. Adjustment of large vertical lines is possible with ARC.
fig. 11: Onset of squint in 70 cured patients with normosensorial late convergent squint (average age 3.5 years).

fig. 12: Refraction in 70 cases of normosensorial late convergent squint.
compared.

When we use random dots, then we find a much worse stereoaucity in microtropia than by using tests with lines and contours, such as the Titmus test. There seems to be a fundamental difference between using small random-dots and tests with lines and contours.

NORMOSENSORY LATE CONVERGENT STRABISMUS

I would like to close my discussion with another strabismus form, the so-called normosensory late convergent strabismus. Here in the manifest stage, there is no stereopsis at all, even by the two pencil test. But after surgery full stereopsis usually is reached, even with random dots.

For many years, I have been interested by the simple question, what cases of strabismus could really be cured by surgery. Let us look at 70 cases which I reported on some years ago. In studying this strabismus form the importance of case history is revealed and the old saying that in medicine the case history already means half of the findings proves to be true again.

The most striking feature in our 70 patients is the late onset of strabismus: on an average at the age of 3 years and 5 months. In almost all of these cases the onset was not gradual but acute or sudden. At the beginning the strabismus was usually intermittent and then became constant. Often the strabismus did not start in the evening or with fatigue but appeared immediately on waking in the morning, or after a nap. Those children seem to wake up as squinters, but after one or two hours the eyes may return to parallelism.

Recently, the daughter of one of our residents, at the age of 3-1/2 years, suddenly showed a constant convergent strabismus one morning. But the parents had noticed some months before that when woken up at nights the eyes were convergent. Of course, Barbara work up in the morning much earlier than her parents so that an intermittent phase could easily have escaped observation. She was unhappy. Before the onset she loved to draw and to walk on low narrow walls. She stopped doing this immediately. She complained of double vision. She grossly missed the two pencil test. After operation she switched to orthoorthopia, was happy again and passed the two pencil tests and other more sophisticated stereotests gloriously.
Not all children, of course, complain about diplopia, but closing one eye in a convergent strabismus may be regarded as an objective sign of diplopia. Seventeen out of our seventy children closed one eye. But again it must be taken into account that not all children can close one eye, but rubbing one eye or closing one eye with a hand has the same significance.

You must always ask specifically for this information because it tends to get forgotten quickly by the parents.

With this fresh anamnesis one would believe that the parents could give exact information about trigger factors. In two cases, a short occlusion of one eye was indicated as cause, in a few cases minor children's illnesses were connected with the onset, but even the parents doubted if that was really the cause.

Refraction shows almost normal distribution for this age. In all cases full correction was given and in most cases prismatic correction was tried. But not in all cases a prismatic orthotropia was reached. Surgery was the treatment of choice. In most cases, I performed a standard operation with 4 mm recession and 7 mm resection of the nondominant eye.

Besides those 70 successful cases, I had, of course, failures. Time does not allow me to go into details about them and most differential diagnoses.

I hope you agree with me that there is a clinical entity of normosensorial late convergent strabismus, which can be cured by simple surgery, if surgery is not delayed too long after the onset of strabismus. All of the 70 cases reached orthotropia, some of them even orthophoria. Those cases usually are not described as a separate entity, but are reported to show the usefulness of some sophisticated treatment methods and greatly help to improve statistical results on those methods.

With these three clinical entities and with accommodative convergent strabismus now let us come back to our model forms of strabismus and let us try to localise their seat.

Accommodative strabismus without high AC/A is the result of hypermetropia and has its seat in the refraction of the
eyes.

The congenital strabismus syndrome may be due to an imbalance in development between the extra-geniculo-striate and the geniculo-striate visual system and has its seat in the midbrain.

Microtropia may be due to a statistical variation in the interplay between the feedback mechanisms of monocular fixation and binocular fusion, and may have its seat in the visual cortex.

Normosensorial late convergent strabismus may simply be due to convergent position of rest with its seat in the orbit.

Be that as it may. I would like to end with a quotation from the great physiologist Hering, whom you know from his law of equal innervation of yoke muscles and from his law on identical visual direction. In 1899, Hering wrote that, concerning strabismus, it is not the judgement of the physiologist that is crucial, but that of the experienced ophthalmologist. I think this quotation would have pleased Frank Costenbader.

REFERENCES


