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Chapter 4

The Treatment of Intermittent Exotropia in Childhood: A Long-Term Study

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Abstract

This study presents 124 children aged less than 15 years suffering from a temporary divergent squint when looking into the far distance. These children were treated and observed in an ophthalmologist's office over the past 35 years.

No evidence-based rules for treating intermittent divergent squints are found in the literature. The possibilities are: (1) Prescription of glasses, (2) Short-term occlusion of one eye against suppression, (3) Orhtoptic exercises, (4) Prismatic correction of the squint over some years, (5) Surgery, and (6) Contact lenses in older and myopic children.

The aim of treatment is to establish a steady compensated exophoria with a latent angle as small as possible; in rare cases the result may be orthophoria.

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The patients sample showed some specific characteristics: 44.5% started squinting in the 2nd and 3rd year of life, 76.6% had unilateral strabismus and suppression, but no severe amblyopia and - 67% had approximate emmetropia, which played a role in compliance with wearing glasses.

Most ophthalmologists prefer to wait and observe the child for some time; if the child's squint deteriorates, an operation will be proposed. The success of such operations is uncertain.

I adopted a conservative treatment approach starting with prisms. Unlike older children the younger children – aged less than 10 years – mostly accepted the glasses necessary to apply the press-on-prisms which correct the squint angle for distance. After a short time, the eye position fixing near objects relaxes and the children had almost the same squint deviation for near and distant fixation. Therefore the diagnosis was "pseudodivergence excess" in almost all the children, only two had convergence insufficiency and two others needed bifocals. In periodical controls, the prisms were adapted to the current eye position. In this way, - in spite of squint – fusion was trained throughout the day and suppression eliminated. Step-by-step the squint angle decreased, with a reduction of on average 13Δ, and finally, after average 4.3 years the prisms could be removed. The longer the time of prism-treatment, the more constant was the result, even after years. This method is recommended only for children with a squint deviation of up to 10°. More severe squints require surgery; but the results are better after preparation of the binocular functions by prisms. A small postoperative divergence can be treated again with prisms or with contact lenses in cases of myopia.

The therapy should be started as soon as possible after the onset of the squint, as the recovery time will be less. Parents should be warned that treatment takes a long time. However, treatment is safe and results in good sensorial preconditions for adult life.

Introduction

Children with a periodic divergent squint are a heterogeneous group – and their treatment is not evidence-based. Therefore, I would like to share the experiences I have gained over the past 35 years.

A child closing one eye in bright sunshine is considered the typical first behavioral symptom of a divergent squint. Probably the child experiences double vision and unlike an adult can quickly adjust to normal vision by suppressing the deviating eye. Thus, we cannot differentiate in childhood

between decompensating exophoria and intermittent exotropia; the first results in diplopia, the second in suppression or, later on, other sensorial adaptations.

Clinical features are a child tired, feeling bored or sick, not fixing a certain object, and suddenly turning one eye outwards, which the child is able to correct immediately. One eye drifts outwards only when looking into the far distance while not fixing a near object. With time, the squint phases become longer and more frequent and a stage is reached when the parents seek treatment.

Pathophysiology

The dynamics underlying an intermittent divergent strabismus are unknown. Most ophthalmologists suppose an abnormal "position of rest" of the eye-balls in the divergent orbits. With effort the eyes can be forced to converge to a normal straight position with binocular functions. Thus, the divergence is overcome by "convergence excess". [1] Another theory designates the turning out of one eye as "divergence excess [2], assuming that an active monocular divergence is possible. As yet, no center for divergence functions has been located in the brain; therefore, some ophthalmologists explain a divergent position of the eyes by passive relaxation into the "position of rest" [3]. The study of my patient cohort aimed to give new insights.

The Patients

The study included 124 patients aged up to 15 years who were registered at my office and diagnosed with intermittent exotropia. Not included were patients showing organic defects of the eyes, hypothyreosis, mental disability, cerebral palsy, hydrocephalus, premature birth, eye muscle palsies and consecutive divergence, e.g. after surgical intervention to correct convergence.

Some interesting observations were made upon examination of the whole sample:

- Heredity: 20.8% of the children had a family history of intermittent exotropia, assuming the information provided by the children's parents was complete.

- Age at the onset of squint (Figure 1): 44.5% of the children started to squint in their 2nd or 3rd year of life, corresponding to the literature.
- Laterality: 76.6% of the children had an unilateral strabismus, the ratio of right to left eye deviation was 52 : 43. However, none of the patients had a severe amblyopia, only two showed a reduced unilateral visual acuity of 0.5. These patients were treated by short-term occlusion.
- Refraction: The eyes of 120 children were cycloplegically refracted using Lindner's retinoscopy [4]. Surprisingly, 67.5% were found to have approximate emmetropia, i.e., a range between -0.5 D and +0.75 D spherical equivalent (Table 1). No human has exactly 0 D.

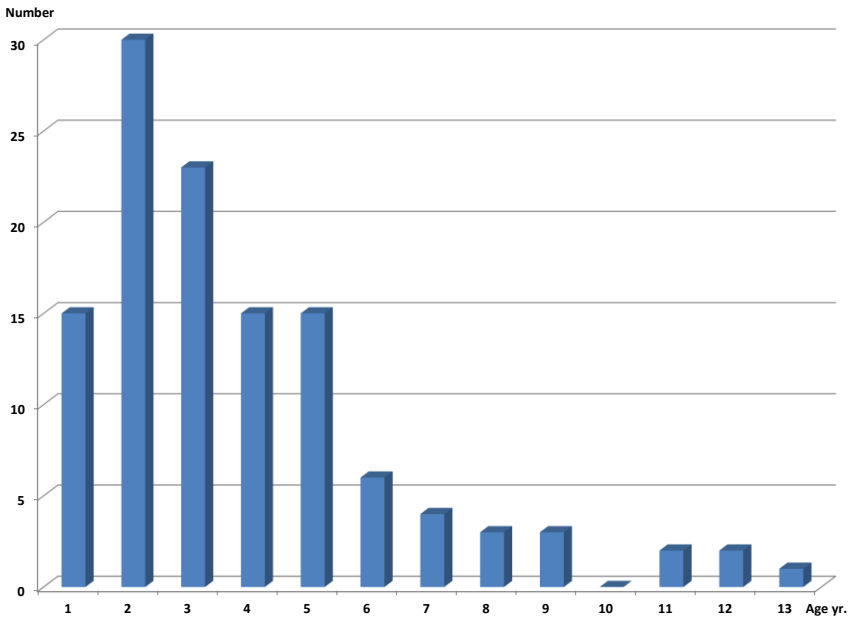

Figure 1. Age at the onset of squint.

A recent study [5] reported that only 25% of the not squinting children aged 2-3 years examined were emmetropic, in contrast to my study, where 71.8% of the children of the same age with intermittent divergent squint were emmetropic. (Table 2). In textbooks [6] the normal refraction of children at this age is assumed to be +2.0 D. The prevailing emmetropia amongst my patient cohort is key to understanding why these children refused to wear

glasses and it was so difficult to use prism therapy with them. 32.5% of my patients needed fully correcting glasses of the refractive error, exceptionally in hyperopic cases 0.5 D were subtracted.

Table 1. Refraction error (120 cases)

	Number	
Emmetropia	81	-
Hyperopia	21	1-2 D
Hyperopia	5	> 2 D
Anisometropia	3	> 1 D
Myopia	5	> 1 D
Astigmatism	5	> 1 D

Table 2. Age and refraction error (120 cases)

	Number	Emmetropia
Age 2-3 yrs	64	46 = 71,8%
Age 4-15 yrs	56	35 = 62,5%
Total	120	81 = **67,5%**

Treatment Method and Results

1. Wait and Observe

Strabismus remains stable for years in some children, but in others the phases of deviation become more frequent and longer. There is an unwritten rule that operation is appropriate if the squinting period exceeds half the time of the child's vigil. The rational is to prevent the development of a steady divergent strabismus and consequent sensorial adaptations. Most ophthalmologists attempt to resolve squints by surgery.

The question arises whether a spontaneous improvement with a change to exophoria is possible. Hiles [7] found that 31 of 48 patients decreased their deviation by an average of 18Δ after a mean of 11.7 years, but normalization was not achieved.

Results of the Study

Three children with an intermittent squint in their 2nd year of life were orthophoric six months later without having received any treatment. Similar spontaneous remissions were never observed in older patients.

2. Glasses

The prescription of glasses depends on the kind of refraction error and, whether or not the child squints. Children who squint – convergent or divergent – require fully correcting glasses, even with low diopters. This is because convergence, accommodation and fusion form a sensitive control system of binocularity; these components influence each other. Even a slight accommodative relaxation with glasses of only +0.5 D may increase the fusion range [8] and relieve exophoric patients of asthenopic complaints. The recommendation in the literature [9] is to prescribe minus-lenses to overcome the divergence by activating the accommodative convergence. This method does not seem to be physiologic and may overstrain the accommodative power. In a recent study [10] children with intermittent exotropia were found to have a reduced accommodative amplitude compared with controls.

Results of the Study

Eight patients, aged between 4 and 13 years were treated with glasses alone; one had myopia of -4 D, the others a refraction error with a maximum of +1.5 D and glasses up to +1.0 D. Only one child refused to wear glasses. After 0.5 to 6 years' observation five patients reduced their squint angle from -25Δ to orthophoria, and the others had at least well compensated exophoria.

3. Occlusion

Occlusion seems to work against suppression of a deviating eye, but at the same time interrupts the fusion present at near fixation. One should consider that suppression of the squinting eye is produced binocularly and only occurs in those moments when the eye is in an anatomical outward-position; [11] therefore, elimination is only possible in this state by prismatic correction of the squint angle.

Occlusion of the master eye is indicated to treat amblyopia or deep-rooted laterality. The occlusion film, fixed on the glass of the dominant eye, should

be translucent and reduce the vision to an acuity of 0.1. Light impermeable foils could also be used, but only for a few hours a day.

Results of the Study

Two patients of pre-school age eliminated their squint within a few months.

4. Orthoptic Exercises

Orthoptists are obligated to give patients orthoptic exercises with the aim of improving the fusion depth, activating the convergence and eliminating suppression. Unfortunately, the exercises are time consuming.

Results of the Study

The control checkup of 13 patients after training showed more power to compensate the deviation, but the squint angle in general was the same as before.

5. Contact Lenses

In my experience, myopic patients who wear hard contact lenses (CL) reduce their squint angle. This is because more effort is needed for accommodation in eyes with CL. Another factor may be that the optical imaging in the periphery of the visual field is better with CL than with glasses. The peripheral visual perception is dominant over the central one when looking into the distance.

Result of the Study

The benefit of hard CL seen in two of my myopic patients:

Case 1: Girl, father squinting divergent
Age 14 yrs: Refraction: RE -5.75 +0.75x LE -6.5 +0.75x
Decompensating exophoria, sometimes diplopia
Squint angle distance (F) -35Δ, near (N) -40Δ
Age 18 yrs: Fitting of hard CL resulting in → exophoria
Age 41 yrs: After wearing the CL since 23 yrs: F = N exophoria,
Latent angle F = N -14Δ

Case 2: Girl
Age 12 yrs: Refraction: RE -2.5s, LE -1.25s
Intermittent divergent squint, angle F -30Δ, N -4Δ
Age 17 yrs: wearing CL, squint angle F -4Δ, N -2Δ

6. Long-Term Wearing of Prisms without Operation (19 Patients)

6.1. Background

In 1967, French researchers [12, 13] found that continuously wearing prisms corrected squint deviations; the prisms were progressively reduced and no operations were needed if the angle did not exceed 8° or 10°.

Press-on prisms have been available in Austria since 1970 and have been used at the 1. University Eye Clinic in Vienna with success one year before a squint operation [14].

Long-term observations of numerous patients have revealed that – in some cases-- there is a spontaneous reduction of the deviation or a consecutive divergence some years after an operation. Furthermore many patients, operated in early childhood, squinting again when adult. This raises the question of whether the ophthalmologists are performing too many operations on children of too young an age. Is there another therapeutic way? My first series of cases where patients were treated only with prismatic glasses was published in 1998 [15]. Intermittent divergent cases were not included in that study.

6.2. Guidelines for Fitting Prisms

The strength of the prisms base in (-Δ) is determined by the alternating cover-test at both distances, far and near.[16] Once the prisms fit, no movement should occur at any distance. At this point the Bagolini test is positive in many cases. Both eyes should be controlled by the unilateral cover-test; if a manifest deviation is evident, the prism power is insufficient. Press-ons should be used at the start of treatment, 20Δ maximum for each eye. The prisms should be applied to both eyes with the stronger one on the dominant eye against suppression. The difference between the right and left eye should not exceed 10Δ.

Shortly after the first application of the prisms the eye position for near fixation relaxes from parallel to divergent; now the same prismatic power fits for both distances, the “pseudo-divergence-excess” type of squint has changed

into a basic type. Check the prisms every 2nd month and be careful not to overlook the development of an amblyopia behind the prisms, which may be caused by a microtropia, not by the prisms per se. For cosmetic reasons school children are given total or partially correcting prismatic glasses without or with thin press-ons.

6.3. Guidelines for Reducing the Prisms

If the alternating cover-test shows a movement from the nasal to temporal side, the prisms are overcorrecting and need to be reduced. If the prisms seem to be correct, a trial is possible: Apply a press-on of low power base out to one prismatic glass and let the patient wait for half an hour; possibly, in this time the child will stabilize a new eye position and reduced prisms can be ordered.

After different intervals of time the squint deviation changes slowly to a straight direction. The best possible result is orthophoria with good stereo-acuity. Most of my patients showed at least a slight and well compensated exophoria without wearing prismatic glasses.

6.4. Results of the Study (19 Patients)

Prismatic glasses were unsuccessful in two of the patients who wore (?) them for 5 years without any improvement. However, 17 patients had the staying power to finish the treatment and showed improvement.

For example (figure 2):

Case 3: Boy, onset of squint at age 3 yrs, deteriorating, Parents refused an operation

Age 7 yrs: Intermittent exotropia LE, angle F = N -12Δ

Refraction: RE +0.75s LE +0.75 +0.25x, VA: RE = LE 1.0

Prisms: -12Δ distributed to both eyes

Age 8 yrs: Prisms: -18Δ

Age 9-10 yrs: Slow decrease to -10Δ

Age 11-15 yrs: Decrease to -4Δ, prismatic glasses stop

Age 22: Well compensated exophoria -6Δ, no complaints

To give a clearer overview, the sample of patients was divided into two groups according to their visual impairment estimated:

Group 1: Onset of squint at an early age with subsequent deterioration.

Group 2: Onset of squint at 4 years or older, decompensating only when tired, after a cover-test, occlusion or when excited.

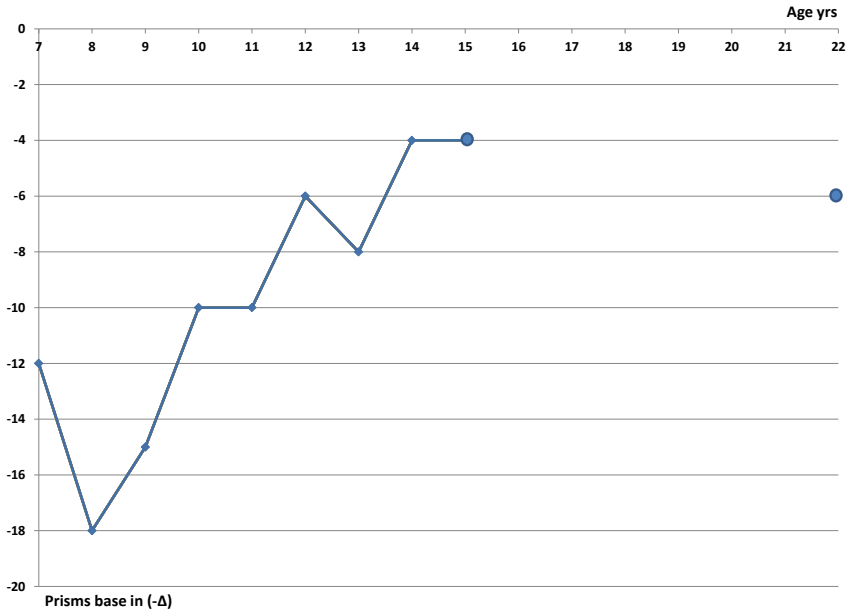

Figure 2. The fluctuations of the squint angle of case 3 during the prism treatment.

Table 3. Prism therapy alone (Group 1)

Name	C.L.	M.K.	W.M.	H.A.	K.M.	M-H.F.	W.C.	T.J.	K.J.	***av.***
Onset of squint										
age yr.	1	1	1	2	3	3	3	4	5	***2.6***
Start prisms										
age yr.	4	6	8	4	5	7	8	5	7	***6.0***
Δ	-12	-10	-20	-25	-22	-12	-18	-17	-20	***-17.3***
Final result										
age yr.	8	7	12	11	10	14.5	15	12	13	***11.4***
phoria Δ, no tropia	0	-6	-4	0	-16	-4	0	-5	-2	***-4.1***
Later findings										
age yr.	13				13	22	19		21	***17.6***
phoria Δ	-2				-4	-6	-10		0	***-4.4***
Duration of prism treatment yr.	4	1	4	7	5	7.5	7	7	6	***5.4***

Comment on group 1:

Prismatic treatment was initiated in most cases with a mean delay of 3.4 years; deviation was between 10 and 25 Δ base in, average 17.3Δ ; both, the time of onset and the degree of squint seemed to influence the time required to correct the squint (1-7.5 yrs, average 4.5 yrs). Every patient showed a

remarkable reduction of the squint angle, average 13.2Δ, which remained constant after some years.

Comment on group 2:

The results for group 2 were more optimistic. These older children were treated with less delay, mean 1.8 yr; wearing of prisms lasted 0.5 - 8 yrs, average 3.1 yrs. The reduction of the angle was - similar to group 1 –an average of 13Δ. This seems to be the limit for improvement with the help of prisms. The two children who wore the prisms for the longest time had the worst starting positions -the largest squint angle or the longest delay of treatment.

Table 4. Prism therapy alone (Group 2)

Name	P.F.	M.S.	L.C.	B.T.	W.C.	G.M.	St.D.	St.L.	***av.***
Onset of squint									
age yr.	4	4	4	4.5	5	6	7	8	***5.3***
Start prisms									
age yr.	6	6	8	5	7	7	8	9	***7.0***
Δ	-15	-30	-20	-16	-20	-12	-20	-20	***-19.1***
Final result									
age yr.	6.5	11	16	6	9	10	10.5	12	***10.1***
phoria Δ, no tropia	-3	-14	-6	-2	-6	-6	-12	0	***-6.1***
Later findings									
age yr.		11	16			13			***13.3***
phoria Δ		-14	-12			0			***-8.7***
Duration of prism treatment yr.	0.5	5	8	1	2	3	2.5	3	***3.1***

7. Operation and Prisms Combined (20 Patients)

The sample was divided into two groups using the same criteria as above.

Example from group 1, the more complicated group (figure 3).

Case 4: Boy, squint onset 1st year of life, convergence was trained at home.

Age 4 yrs: Squint angle F -24Δ, N -12Δ, refraction: RE +0.75s LE +0.5+0.25x

VA: RE = LE 1.0, first prisms: -12Δ each eye

Age 5 yrs: Prisms -35Δ distributed to both eyes

Age 6 yrs: Operation: LE Recession rect.ext. 5.5mm, resection rect. int. 4 mm

After the operation: angle F +8Δ, N 0Δ, prismatic correction and alternating occlusion were done, yet the angle increased up to +20Δ (diplopiaphobia). After 4 weeks the eye position was stable with +6Δ, no more occlusion was done, 2 months later the prisms were removed.

Age 9 yrs: Decompensating exophoria was again corrected with prisms up to -10Δ, decreasing to -3Δ.

Age 16 yrs: Glasses no longer required.

Age 18 yrs: orthophoria

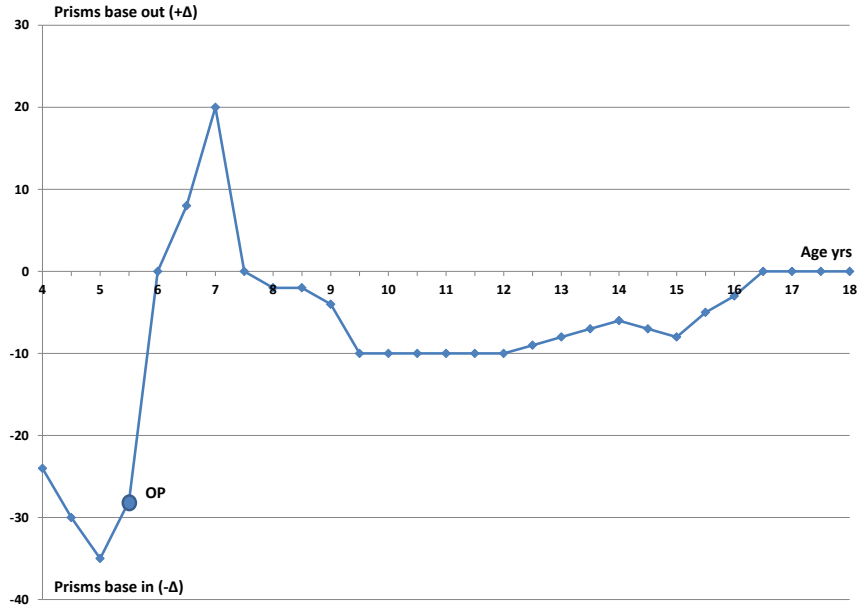

Figure 3. The fluctuations of the squint angle of case 4 during the prism treatment.

The Outcome of the Operated Group 1 (Table 5)

The initial squint angle was an average of -28.8Δ. In spite of the good operation results, in most cases the divergent position recurred within a few months of the operation and prisms were prescribed again. The patients who had the second prismatic correction seemed to have more stable long-term results. Only the one patient with the largest deviation needed a second operation. Example from group 2 (Figure 4):

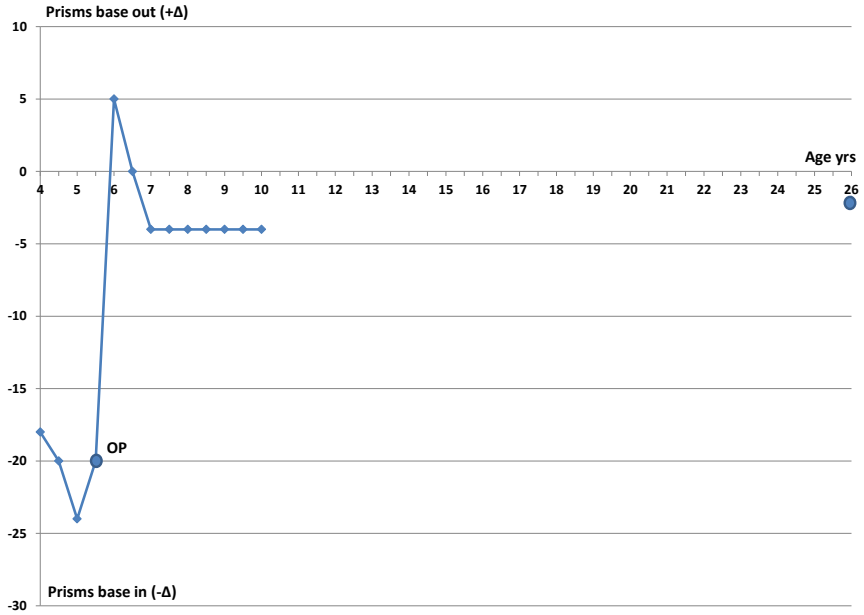

Figure 4. The fluctuations of the squint angle of case 5 during the prism treatment.

Table 5. Prism therapy + operation (Group1)

Name	B.C.	H.P.	Sch.C.	R.P.	E.M.	H.M.	K.C.	W.K.	Qu.H	H.I.	***av.***
Onset of squint											
age yr.	1	1	1	1	1.5	1.5	2	2	2	3	***1.6***
Start prisms											
age yr.	4	4	5	8	5	3	3	4	2	10	***4.8***
Δ	-40	-24	-25 Bifo	-35	-15	-25	-45	-30	-25	-20	***-28.8***
Operation											
age yr.	5	5.5	6	8.5	7	7	4	7	6	11	***6.7***
Result angle Δ	-6	0/+8	-4	-6	0	0	0	0	0	0	***-1.8***
Again prims											
age yr.		6/7	6.5			8	4.5	8	8		***7.0***
Δ		+20/-10	-15			-12	-35	-16	-15		***-18.6***
2. Operation											
age yr.							9				
Result angle Δ							0				
Final result											
age yr.	11	16	11	9	7	8	9	8	10	12	***10.1***
phoria Δ, no tropia	-8	-3	-6 Bifo	-18	0	-12	0	-16	-8	-4	***-7.7***
Later findings											
age yr.	30	18		19	9	17	16		21	18	***18.5***
phoria Δ	-3 CL	0		-16	0	-10	-8		-8 CL	-12	***-7.7***
Duration of prism treatment yr.	1	12	6	1	2	5	6	4	8	1	***4.6***

Case 5: Male

Age 4 years: divergence of right eye recently noticed. Refraction: RE -0.5+0.75x, LE -0.25+0.5x, VA RE = LE 1.0, RE suppression tendency

Age 4.5 yrs: First prisms: RE -5Δ, LE -12Δ, increasing to RE -8Δ, LE -15Δ

Age 6 yrs : Operation: RE Recession rect. ext. 4mm, resection rect. int. 3.5 mm

Result: 0Δ, stereopsis. 2 weeks later convergence +5Δ, prisms for 1 month

Age 10 yrs: Myopia is developing, exophoria -4Δ

Age 26 yrs: Exophoria -2Δ, contact lenses, no complaints

Table 6. Prism therapy + operation (Group 2)

Name	H.M.	L.O.	St.V.	G.T.	K.T.	Y.Y.	K.Th.	M.F.	St.M.	W.K.	***av.***
Onset of squint											
age yr.	2	2	2	4	4	4	5	5	5	8	***4.1***
Start prisms											
age yr.	5	4	4	4	5.5	4.5	6	5	5	8.5	***5.2***
Δ	-30	-22	-25	-20	-25	-35	-20	-15	-25	-22	***-23.9***
Operation											
age yr.	5.5	5	6	5.5	6	5.5	9	8	6	10	***6.7***
Result angle Δ	0	0	0	0	-10	0	-2	2	0	0	***-1.0***
Again prims											
age yr.	6	5.5	7	5.6							***6.0***
Δ	-6	-18	-16	5							***-8.8***
Final result											
age yr.	8	8	8	6	7	6	9.5	11	7	10.5	***8.1***
phoria Δ, no tropia	-4	-16	-10	-4	-10	0 Bifo	-12	-8	-2	-2	***-7.6***
Later findings											
age yr.		10	27	26	11	15	22	16	10	12	***16.6***
phoria Δ		-16	-8 CL	-2	-14	0 Bifo	-16	-10	-16	-4	***-11.1***
Duration of prism treatment yr.	3	4	4	1.6	0.5	1	3	3	1	1.5	***2.3***

The Outcome of the Operated Group 2 (Table 6)

The initial squint angle was a mean of 23.9Δ (smaller than in group 1), prisms were worn for an average 2.3 years (shorter than for group 1). Only 4 patients needed prisms again after surgery and, -no one needed a second operation. But the degree of the remaining exophoria, measured in the patients at an older age, was the highest among the four prism-treated groups.

8. No or Uncompleted Treatment (57 cases)

Asked for second opinion	33
Non-compliance, refusing glasses	12
Prism treatment not yet finished	5
Prism treatment broken off	7

Conclusion

1. The Practice

The aim of the study was to establish the best way to treat patients with intermittent exotropia. Sometimes this problem solves spontaneously without any treatment, but only before the age of 2.5 years. To observe a child and wait for longer is a waste of time in my experience. Considering the potential benefit, I would choose to prescribe glasses as a first step, regardless of the type of refraction error because sometimes glasses do help. As a next step I recommend fitting prisms. These stabilize the eyes in the divergent position, which seems paradoxical but in reality bifoveal perception is possible in this position; fusion is trained during the day with a simultaneous elimination of suppression. The prisms should correct the total deviation because complete relaxation of the eye position is necessary to prevent overacting convergence impulses that draw the divergent eyes inside again [1]. Orthoptic exercises for convergence are not advisable at this state of treatment. The child learns by the prisms that the divergence is the “normal” position. After some years binocular functions are established and become strong enough to maintain an almost straight eye position. This gentle and physiologic treatment is effective only for squint deviations up to 10°; more severe cases require an operation. There are many unfavorable reports in the literature [3 and 18] of high rates of recurrence after these operations. Surgery is an intervention which particularly disrupts the equilibrium between the eye pair -as can to be seen in case 4 reported here. Therefore, it is advisable to additionally treat patients with prisms before and, if necessary, after surgery.

Treatment of a child who squints should be started as soon as possible to shorten the time of recovery. Based on experience with my patient cohort, I believe, the longer the prismatic glasses are used, the more stable and lasting are the benefits. It seems to be important to have an as small as possible angle

of exophoria at the end of the treatment. Modern working conditions that involve long periods of concentration on a computer screen are unfavorable for eyes. Many adults -who were operated in childhood – require further surgery because of asthenopic complaints and decompensating divergent squints.

The difficulties in implementing the prism method which are probably the reason why prisms are not the standard method of strabismology, are well-known: Compliance with the wearing of glasses, many years of treatment with periodical controls; the method is time consuming but safe. However, many parents of squinting children prefer the shorter surgical treatment without anything else.

2. The Theory

Some authors have postulated that divergent squints are caused by anatomical hindrances in the orbit. This may be right for extreme cases of divergence like Morbus Crouzon, but not for common cases of intermittent exotropia. If the "position of rest" can be normalized by prisms, the anatomy must be normal. The position of rest seems to be based on the tonic innervation of the eye muscles, tonic vergence, accommodative and fusional vergence.

Returning to refraction, the 67% of emmetropic patients. If hypermetropia of +2D is the common refraction in early childhood and babies have a distance of sight about 1m in daily life, an accommodative effort of 4 – 5 D and adequate convergence are necessary for clear binocular vision. In Donders' opinion [19], myopic babies may develop an exodeviation on the base of an underactive accommodation-convergence mechanism. My patients seemed to confirm this thesis.

I cannot exclude other concepts of squint origin, for example that active innervational impulses cause the turning out of one eye. I have seen a boy of 7 years who suddenly started to squint when his parents got divorced.

The chapter of intermitting divergence is not yet closed. More scientific investigations need to be done.

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