SURGERY FOR ABNORMAL HEAD POSITION IN CONGENITAL NYSTAGMUS *

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ABNORMAL HEAD POSITIONS ON AN OCULAR BASIS MAY BE THE RESULT OF several mechanisms that are important for the ophthalmologist to recognize. Among these are ocular muscle palsies, either horizontal or cyclovertical, mechanical restrictions as in floor fractures or Brown's Syndrome, uncorrected astigmatism, A or V syndromes, ptosis of one or both eye lids, and congenital nystagmus with an eccentric rest point. It is the purpose of this paper to discuss our experience with the surgical treatment of this latter group.

Based on the classification of Cogan,[1] these patients have the motor-defect type of nystagmus characterized usually by a jerk nystagmus. Somewhere in the field of gaze the eyes assume a relatively stationary position. It is at this position of gaze that the visual acuity is at its maximum. If this position of least nystagmus is in the primary or straight ahead position the patient is fortunate. His head position for maximum visual acuity causes no cosmetic blemish. However, if the position of least nystagmus is in gaze away from the primary position then an abnormal compensatory head position must be assumed for maximum vision.

Correcting the head position by surgery on the ocular muscles is not a new idea. Apparently J. Ringland Anderson of Australia and Alfred Kestenbaum of New York independently conceived the idea of correcting the abnormal head position by operating on the ocular muscles.

Anderson[2] felt that the tone of the horizontally acting muscles was not balanced and that a recession of one yoke muscle in each eye could restore this balance. Kestenbaum[3] achieved a similar result by a bilateral resection and recession operation, but inexplictly this was performed in two stages, creating a strabismus in the interval. Goto[4] of Japan, in 1954, suggested strengthening of the appropriate muscles without a recession for this type of problem.

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A logical extension of these procedures was applied by Dermot Pierse[5] of England in 1959. He described two patients with nystagmus with the head held backward for maximum vision. He weakened both depressors, the inferior rectus and the superior oblique muscles.

Many authors subsequently have published their results with similar operations.[6-9] In general, all have reported good results, as regards the compensatory head turn. The amount of surgery has been limited by most authors to a 5 mm or less recession of the medial rectus in order to preserve adduction of that eye. Corresponding amounts of surgery have been performed on the other ocular muscles.

Since these cases are not too frequent, the direction that the eyes are to be moved may cause confusion. If there is no tropia in the preferred position of gaze, then both eyes must be moved by surgery in a conjugate manner.

If the face is turned to the left so that the gaze is to the right for maximum acuity, the approprite surgery moves both eyes equally to the left, in the direction of the face turn. Expressed in another manner, the appropriate surgery moves the eyes away from the preferred gaze (See Fig. 1).

If there is a tropia so that one eye is fixing, the position of the head is determined by the preferred position of gaze of the fixing eye. Any change in the head position will be mediated by the fixing eye. Therefore surgery performed on this eye alone will correct the deviated head position. This is in contrast to the usual philosophy in strabismus surgery in which surgery is performed on the nondominant eye. The effect of surgery on the heterotropia will have to be estimated. When the head position is appropriately altered by surgery on the fixing eye, additional surgery on the nonfixing eye may be needed for any residual heterotropia. This may be accomplished at the same time or as a subsequent procedure.

Our experience with surgery for head turning in congenital nystagmus is based upon 19 cases from Wills Eye Hospital and St. Christopher's Hospital for Children. Sixteen of the 19 cases are males. A similar preponderance of males with this type of nystagmus was noted by Anderson and by Cogan. The follow up period ranges from two months to 34 months, with an average follow up period of a little more than 13 months. The age at time of the surgery ranged from 15 months to 43 years with an average age of approximately 8½ years.

The association with mental retardation is of interest. Three of the 19 patients had a mild to moderate delay in psychomotor development. One additional patient has a mild hemiparesis.

FIGURE 1

Surgery for the head turn should move the eyes to the left, toward the face turn and away from the preferred position of gaze.

RESULTS

In spite of having surgery on four of the horizontally acting ocular muscles, the effect on binocular function is minimal (Table 1). The lack of any adverse effect on binocularity can be determined by the absence of diplopia, by cover testing, and by measuring stereo acuity and fusional vergences.

Fourteen of the 19 patients were orthophoric in their position of least nystagmus as judged by cover testing. Ocular muscle surgery on two muscles in each eye to effect a change in head position incurs the risk of producing a heterotropia and diplopia. In none of these 14 patients was a heterotropia or diplopia noticed postoperatively.

In four orthophoric patients the stereo acuity by the Titmus test was the same preoperatively as postoperatively. In an additional two patients in whom stereo acuity was not measured preoperatively, it was 40 and 50 seconds of arc postoperatively.

Horizontal and vertical fusional vergences were normal in four patients both preoperatively and postoperatively. Furthermore, it was normal in

TABLE 1

Patient	Age surgery yrs.	Follow up months	Preop. dev. prism diopters	Postop. dev.	Surgery	Results	Comments
1	3½	23	0	0	Recess RLR 7, LMR 5 Resect RMR 6, LLR 8	No imp.	Mild mental retardation
2	11	22	0	0	Same as #1	Imp.	
3	4½	2	0	0	Same as #1	Cure	Lost to follow up
4	4	34	0	0	Same as #1	Imp.	
5	8	18	0	0	Same as #1	No imp.	
6	43	17	RET = 50	RET = 25	Recess MR OU 5 mm	Imp.	
7	8	29	0	0	Same as #1 Plus recess IO OU 8 mm	Imp. Imp.	
8	15	27	LET = 30	LET = 20	Recess RMR 5 mm Resect RLR 8 mm	Imp.	Brother of #7
9	5	27	0	0	Recess RMR 5, LLR 7 Resect RLR 8, LMR 6	Imp.	
10	4	13	0	0	Same as #9	Cure	Optic N. hypoplasia, mild
11	1½	2	RET = 45	RE(T) = 30	Recess LMR 5 Recess LLR 8	Imp.	Mild mental retardation Mild optic N. hypoplasia
12	2½	3	0	0	Recess RMR 6, LLR 8 Resect RLR 8, LMR 6	Imp.	Lost to follow up
13	6	22	0	0	Recess RMR 6, LLR 8 Resect RLR 9, LMR 7	Imp.	Unilateral high myopia
14	1½	2	0	0	Same as # 13	Cure	Lost to follow up
15	2½	2	0	0	See (2) below	Imp.	Prior recess MR OU 5 For cong. ET
16	12	10	0	0	Recess RLR 10, LMR 7 Resect RMR 8.5, LLR 11	Imp.	See (1) below
17	12	5	0	0	Recess RMR 7, LLR 10 Resect RLR 11, LMR 8.5	Cure	
18	3	5	LET = 30	0	Recess RMR 7, LLR 5 Resect RLR 11, LMR 4	Cure	Mild mental retardation
19	13	3	LET = 30	0	Recess RMR 7 Resect RLR 11	Cure	Moderate RLR O.U. VA OS C.F., hemiparesis

two patients postoperatively where comparative measurements had not been taken preoperatively.

From these results it can be concluded that simultaneous surgery on four horizontal rectus muscles can be undertaken in an orthophoric patient without undue risk of producing a tropia, diplopia, or disturbing binocularity.

Our indication for surgical correction of a preferred eccentric position of gaze is a cosmetically unacceptable torticollis that is not corrected by glasses. In general the more severe the head turn the earlier the surgery is performed. Since the indication is largely for cosmesis, a cure is defined as no head turn while attaining maximum visual acuity.

Using this criterion, six of the 19 patients have been cured with no apparent head turn remaining. Two showed no improvement and the remaining ten showed varying degrees of improvement of their head position. Some of those that are now classified as improved had an apparent cure or more nearly normal head position for several weeks to months after surgery but gradually turned back toward their preoperative preferred position of gaze. Therefore, short term results may be misleading and overly optimistic. For the six patients which were designated as cures the average follow up period was five months, ranging from two to 13 months. Perhaps with more time some of these patients may have their head and eyes revert toward their preoperative position.

DISCUSSION

Our results and those of other authors have demonstrated that surgery on all four horizontal muscles is a safe and effective means of altering the abnormal head position in congenital nystagmus. Our experience indicates that varying degrees of improvement may be expected in most patients but that complete elimination of the head turn occurs in only one third of the patients.

Cooper and Sandall suggested that the amount of ocular surgery be one half of that amount calculated to correct the head turn. In other words if the head turn were 30 degrees, one should operate to correct 15 degrees. In their series of seven patients the angle of head turn was measured with a perimeter.

Parks has for several years suggested a modified Kestenbaum procedure. He differs from Kestenbaum's original method in two ways. First he operates on all four muscles at the same time, instead of performing two separate unilateral recession and resection operations a proposed by Kestenbaum. Kestenbaum suggested that the same amount of resection

and recession be done on each muscle. Being fully aware that the same amount of surgery on the medial and lateral rectus muscles has different effects, Parks proposed the following:

5 mm recession of the medial rectus
6 mm resection of the medial rectus
7 mm recession of the lateral rectus
8 mm resection of the lateral rectus

These amounts were accepted as the upper limit in order to preserve full ductions. With this technique four of his ten patients would be classified as cures by our criterion of no residual head turn.

For reasons that are not clear most of these patients have much less head turning at near. Measurement of the angle of turning using a perimeter may falsely underestimate the degree of head turning unless particular attention is paid to maintaining distant fixation by having the patient look over the perimeter. Often the patient becomes sensitive to their abnormal head position by the attention of parents and physicians. They may tend to hold their heads straight even though there is blurred vision as a result. Only when maximum distant visual acuity is required can the full extent of the head turn be elicited. These patients will gradually turn their heads as smaller optotypes are presented.

Usually the immediate postoperative results are very gratifying, sometimes with an overcorrection of the head turn. In too many patients there is a gradual return of the head position toward the preoperative position.

We have tried to avoid possible sources of error by recording photographically the head position during a time of maximum distant vision, and thereby maximum head turning. By comparing sequential pictures one can easily note the effect of surgery.

In our earlier surgery on those patients we were doing the modified Kestenbaum procedure as recommended by Parks, hereafter called the classic maximum. Eight patients had this amount of surgery on all four muscles (Fig. 2, 3, 4A, 4B). Two with esotropia had the classic maximum surgery on the fixing eye (Fig. 5A, 5B). Of these ten patients, two were considered to have cures. Two patients who were orthophoric underwent the "classic plus one" surgery, 6 and 9, 7 and 8 mm in each eye (Fig. 6A, 6B). One of these patients was considered as having a cure.

Four patients underwent "classic plus 40%" surgery (Fig. 7A, 7B, 8). In these patients the classic maximums of 5, 6, 7, 8 were each increased 40% to 7, 8.4, 9.8 and 11.2 mm respectively. The tenths of a millimeter are not meant to suggest that muscle surgery is that accurate, but only to show how the figures were derived. Of the four patients, three are considered to be cured, that is their best vision is in the primary position. In

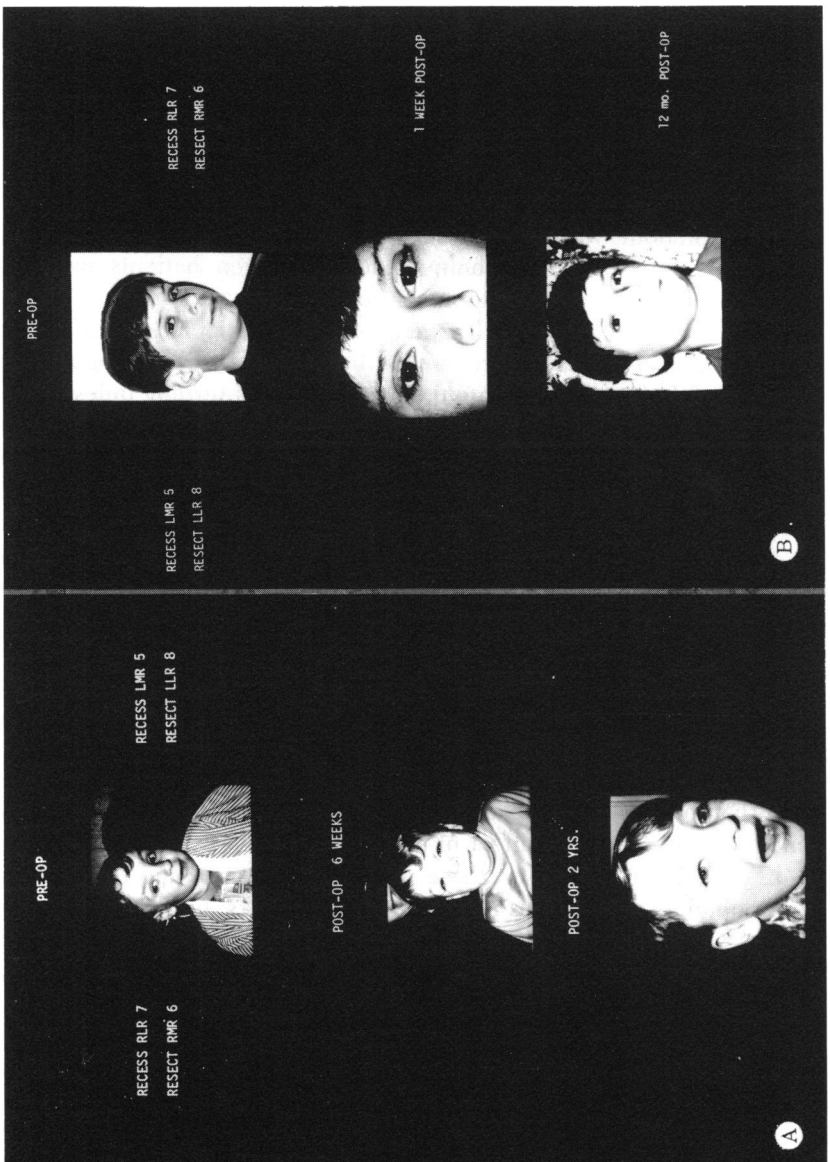

FIGURE 2

A: Patient 1. No effect two years after a modified Kestenbaum procedure. B: Patient 2. Moderate effect one year after a modified Kestenbaum procedure.

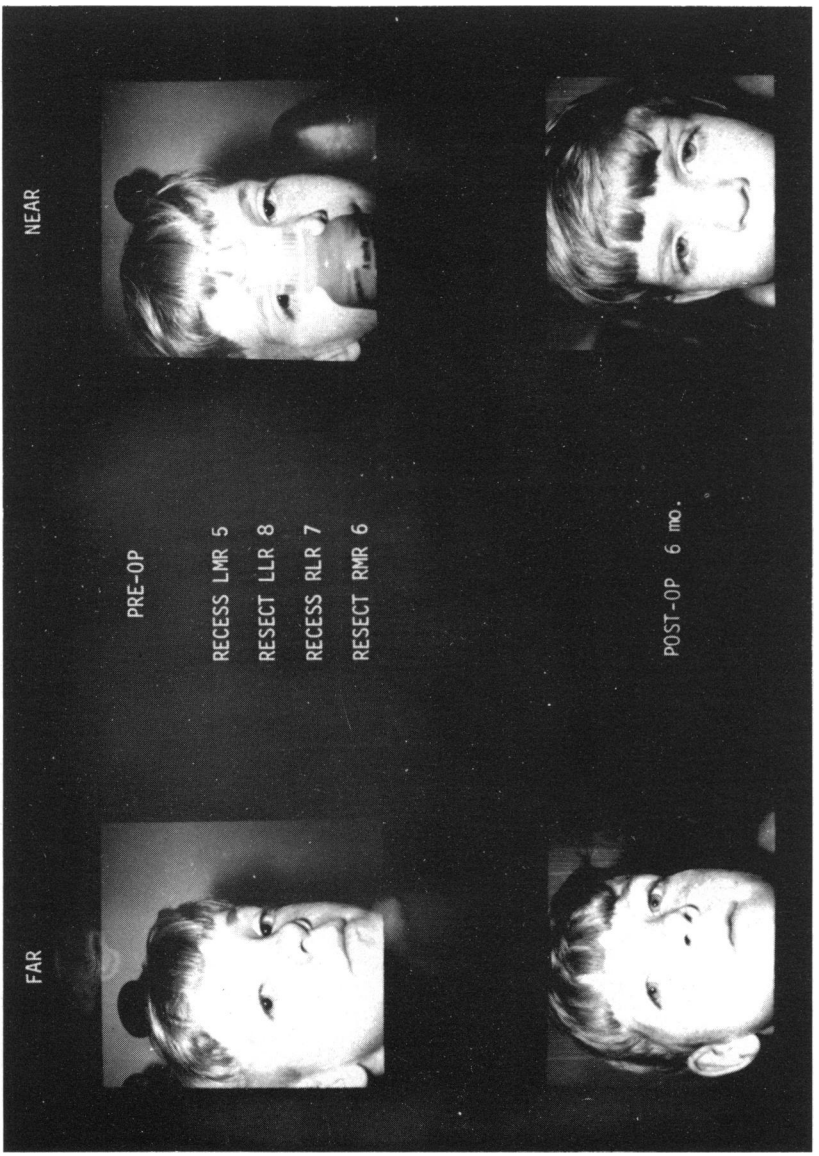

FIGURE 3

Patient 5. Moderate effect six months after a modified Kestenbaum procedure. At last follow up the head turn appeared as preoperative.

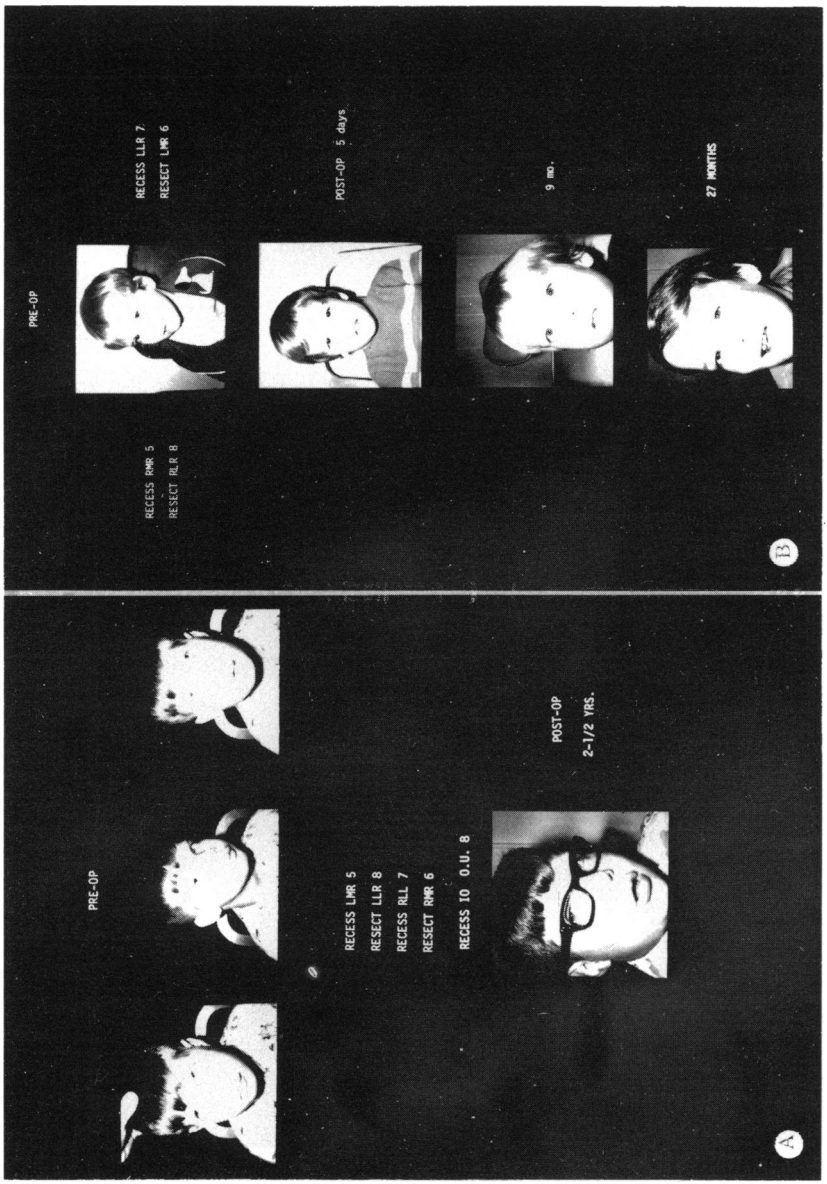

FIGURE 4

A: Patient 7. Moderate improvements at 2½ years with modified Kestenbaum procedure.
B: Patient 9. Slight improvement at 27 months by a modified Kestenbaum procedure. Note the gradual return with time toward the preoperative position.

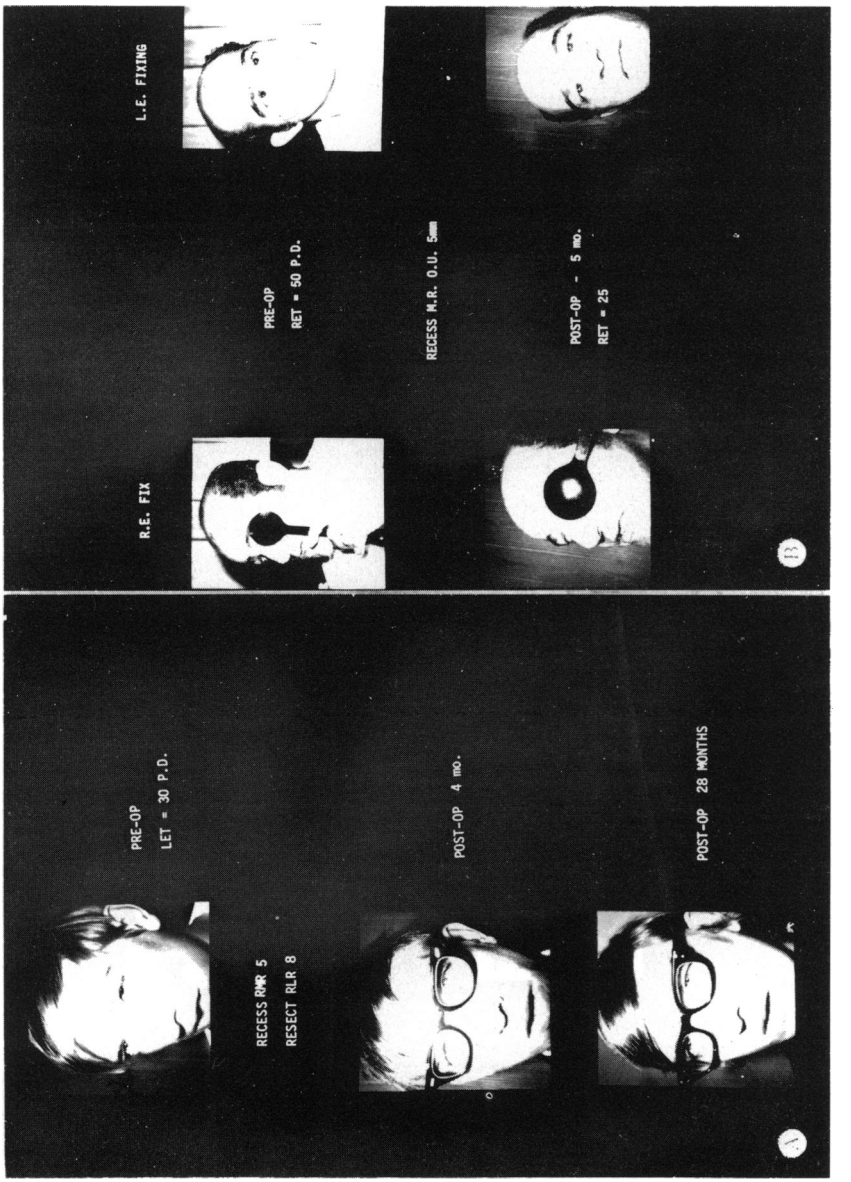

FIGURE 5

A: Patient 8. Classic maximum surgery on the fixing eye with moderate improvement.
B: Patient 6. Recess MR 5 mm O.U. with slight improvement in head position.

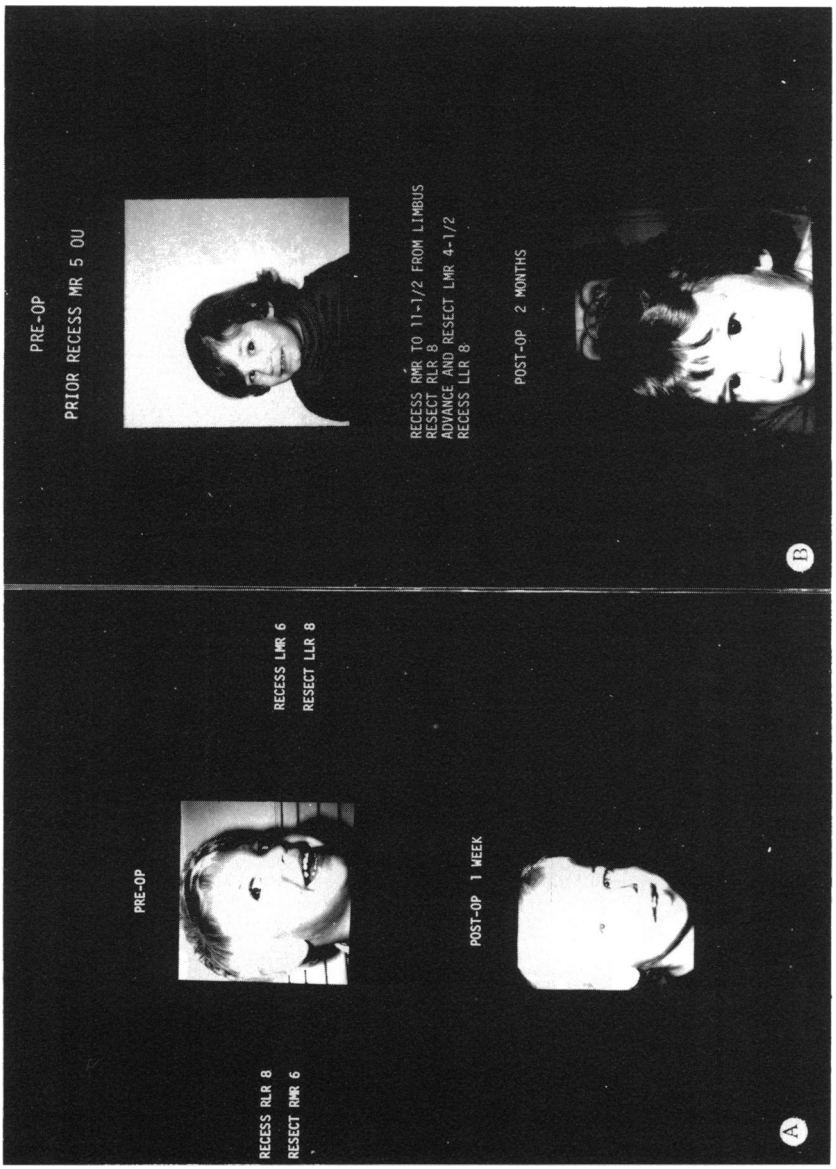

FIGURE 6

A: Patient 12. Slightly more than the classic maximum surgery with only moderate improvement. B: Patient 15. Slightly more than the classic maximum surgery with moderate improvement.

FIGURE 7

A: Patient 16. There is less turning at near pre and post operatively. The surgery of classic maximum plus 40% resulted in moderate improvement with retention of a close NPC but a slight left gaze palsy was created. B: Patient 17. Classic maximum plus 40%. Surgery eliminated the head turn without affecting the NPC but created a gaze palsy to the left.

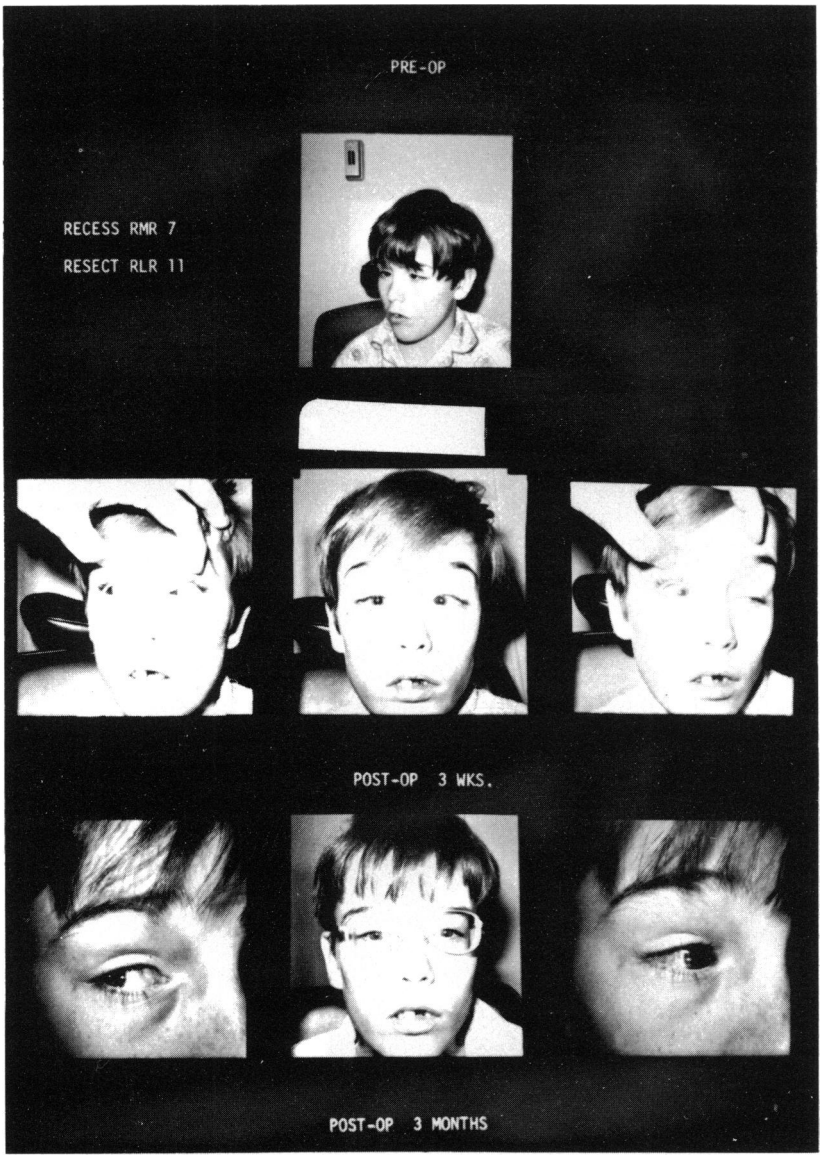

FIGURE 8

Patient 19. Classic maximum plus 40%. Surgery on the fixing eye eliminated the head turn but restricted abduction slightly.

three of the four patients, this classic plus 40% surgery has caused a mild gaze palsy, but to date none have complained of it. Each of the three cured patients is pleased. It is our feeling that the mild defect of ocular rotations that is iatrogenically created is better than a life long head turn.

The data suggests, although the series is small, that more than the classic maximum surgery is often indicated to eliminate the head turn.

SUMMARY

In patients with congenital nystagmus and an eccentric rest point for maximum visual acuity, surgery on the ocular muscles is an effective and safe method of reducing or eliminating the head turning. There are no adverse effects on binocular function as a result. In our series of 19 patients, six were considered as having a cure with no apparent head turn postoperatively. Four of these six patients had more than the classic maximum surgery performed. Two patients had no apparent effect from the classic maximum surgery.

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DISCUSSION

DR THOMAS P. KEARNS. Doctor Calhoun and Doctor Harley are to be congratulated for presenting this interesting and instructive paper to our Society. Their accumulation of 19 patients is remarkable and justifies their recognition as authorities in this aspect of ocular muscle surgery.

I was pleased that they changed the title of their paper from "Surgery in Congenital Nystagmus", as listed in your program, to "Surgery for Abnormal Head Position in Congenital Nystagmus". This change removed my only criticism of their fine paper. This new title is more descriptive of the true purpose of this type of surgery. As the authors point out, this operation is to correct a cosmetic blemish, the head turn, and does nothing to improve vision or lessen the nystagmus. Nevertheless, when a head turn secondary to congenital nystagmus with an eccentric rest point is cosmetically unacceptable to the patient or parents, such surgery is usually justifiable.

This slide tabulates the surgery and results on five patients with head turn secondary to congenital nystagmus seen at the Mayo Clinic over the last six years. Two differences are immediately apparent. First is the small number of patients compared to the authors'. This prompts me to ask the authors how long a period of time does their collection from the two hospitals represent? I ask this not to justiby our small series, but to allow us to obtain a better estimate of just how frequent this operation is required. The second difference is the small number of the authors' patients that had an associated strabismus. As is seen in this slide, four of our five patients had strabismus, as esotropia in each of the four instances. One of Kestenbaum's two patients had strabismus, an exotropia. Four of seven of the patients described by Cooper and Sandall had strabismus, three having an esotropia and one an exotropia. These examples are in contrast to the five of 19 ratio in the series reported today. I wonder if the authors may have an explanation for this. It should be noted that their patients with strabismus all had esotropia, which appears to be the more frequently associated deviation.

The authors point our that confusion may arise on which way to move the eyes in order to correct the head turn. Their suggestion to move the eyes away from the preferred gaze is a good one. This is analagous to remembering which way the prism should be placed in front of an eye to correct a certain phoria. A mistake with prisms may prove to be embarrassing, but its correction is a simple matter. A mistake in the direction of movement of the eyes in this operation would be a catastrophe. This slide illustrates a method I use to clarify this possible confusion. Since this operation is directed toward the head turn, one may forget about the eyes and straighten the head. By leaving the eyes fixed in the deviated direction and thinking about turning the head to the straight ahead position, the proper recession and resection on each of the four horizontal acting muscles becomes obvious. This may seem like the tail wagging the dog, but it does work.

One final point seems worthy of comment. The authors note that some of their patients have gradually reverted to their original position of head turn. This warn-

ing was mentioned by Dr Norman Schatz in a discussion of this type of surgery at the recent Pediatric Ophthalmology Symposium in Philadelphia. He stated that the brain seems to "catch on" and the head turn may return in two years. The authors, with their large series of patients, have an opportunity to do a follow up study on this possibility. It is hoped that they will bring such a follow up report to a meeting of this Society in a few years.

DR ANGUS L. MACLEAN. This is a very timely and instructive paper. I am convinced, after dealing with four or five cases, that one can help most of these unfortunate individuals by improving the visual efficiency and the cosmetic appearance both as regards head turn and nystagmus, but I don't believe one can ever completely cure the nystagmus. I think it can by improved.

I find that determining if the quick motion of the eyes is to the right or left is helpful in analyzing these cases and deciding about the therapy. For example, if the quick motion is to the right, it means that the weak action is to the right side, or the weak muscles are those that move the eyes to the right, and of course this means that the head is going to be turned to the right in order to bring the stronger muscles into play for a better steadying effect on the eyes. It would be necessary to resect the right lateral rectus, recess the right medial rectus, resect the left medial rectus, and recess the left lateral rectus. I think every case presents a quick motion, but it is sometimes difficult to determine which side this is on. I wonder if Dr Calhoun has or knows of any device for taking tracings of the eye movements in these cases. He did not mention this in his paper and I think it would be appreciated if he would comment on it in his discussion.

In operating, there is always a tendency to err on the conservative side. One naturally starts thinking: Isn't this a lot of surgery to be doing on this patient — surgery on all four of the horizontal muscles?

In my first case, I decided on four mm for each muscle. The outcome was satisfactory, but afterwards, I wished I had done more. I think 5 mm for the medial recti and six to seven mm for the lateral recti is a pretty good standard. One never completely cures the nystagmus in these cases, but one always improves and may completely correct the abnormal head turn; but I for one am opposed to ten mm or more of recession or resection of any eye muscle.

I was pleased to hear Dr Calhoun's ideas on the management of nystagmus in a patient with heterotropia. He advocates operating on just one eye — the fixing eye first, and I believe he tries to completely correct the tropia if possible with that single procedure. That idea is completely new to me. In one of our recent conferences I was asked by Dr Stewart Wolff how I would deal with nystagmus in a patient with an esotropia of 30°. I could not then give him an intelligent answer, but think I could now after reading a copy of Dr Calhoun's paper which he kindly sent me in advance of this meeting. Dr Calhoun might want to elaborate on this last point in his closing discussion.

This has been an excellent presentation, and I want to congratulate the authors.

DR MARSHALL M. PARKS. Thank you, Dr Calhoun, for an excellent paper. Since I am apparently responsible for the classic measurements recorded here, I would like to speak to support the continuation of those measurements in the patient who has diffusion and binocularity.

I do agree with these authors that we tend to get under-corrections. Some of the patients look good to begin with. The head malposition return is a difficult problem to deal with. I have had to contend with this in a few patients, and in the patients who have had head return in under-corrections I have offered to the parents of these patients the option of either accepting a limited field of movement of their eyes as a result of doing more than the classic surgery as reported here, or continuing with the head posture. So far every parent has turned me down in accepting the option of accepting a limited field of action.

Therefore I would like to ask the authors: Do you discuss this possibility and complication with the parents before you do the surgery? If so, how do they react to it, and how do you sell something I have been unable to sell to the family?

DR ROBISON D. HARLEY. I would like to thank all the discussers.

Our patients for this study were seen over the past 3 years, to answer on of Dr Kearns' questions, and I do not know why they had less esotropia than other series. I know we are very acutely aware of head turning problems, and perhaps we have picked up more head turners and proportionately we have more head turning without strabismus than with strabismus.

Dr MacLean's suggestion about doing nystagmography or head tracings is a good one. This might be done pre- and postoperatively, because we do have the facilities, and I would like to thank him for the idea. We have not had to repeat on the surgery. I suppose one could consider this, but when you perform the classic surgery +40 percent, one has made extensive muscular alterations. As Dr Parks points out, any more is going to severly limit movement, and therefore, I have been reluctant to consider any further surgery.

As to the question about whether we discuss this with the parents I can not recall discussing it at any great length, except to tell them that in our limited exexperience we have had no serious complications. We feel they will not be any worse, and certainly they have a good chance of being better. We do not go into any further discussion than that.

In the few seconds remaining I would like to discuss a differential diagnostic problem to which Dr Calhoun referred. Head turning in congenital nystagmus must be differentiated from head turning due to astigmatism.

Dr Guy H. Chan working in our department has studied six patients age 2 to 10 years with a face turn induced by astigmatism (refractive torticollis).

His conclusions are as follows:

1. All 6 children had minus cylinder at axis 180 or with-the-rule astigmatism.
2. Head tilting or face turn improved vision in all 6 patients.
3. All face turns were eliminated by corrective glasses.

4. The astigmatic correction varied from −1.00 diopters to −5.00 diopters at axis 180.
5. Associated findings of nystagmus, strabismus, and hyperactive disposition of the children were obstacles to clinician's awareness that astigmatism was the underlying cause for the head tilting or face turn.
6. Face turning in patients with astigmatism apparently afforded better vision. It was suggested that head turning affects the refraction of light entering the eye ball and the Sturm interval could be reduced or eliminated; hence vision improved. A pinhole effect created by the "fixing eye" sighting across the bridge of the nose must be carefully considered.
7. On an optical bench by rotating a combination of lenses along the vertical axis, the collapse of Sturm's interval could be demonstrated.
8. In a hyperactive child who has an unexplained face turn or head tilt associated with nystagmus and strabismus, one should delay surgery and search for hyperopic astigmatism with-the-rule.
9. Even though the amount of astigmatism may appear to be insignificant in a child, one should try to fully correct the fixing eye.
10. The face was invariably turned to the side of the fixing eye that had better vision.
11. Corrective glasses for hyperopic astigmatism with-the-rule have eliminated *face turn* even though the visual improvement was not dramatic or could not be assessed because children were too young for reliable subjective response.