

A Preliminary Study on the Outcome of Plication Augmentation of the Augmented Anderson Procedure for Patients with Infantile Nystagmus Syndrome and a Face Turn

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Abstract

Purpose: To report the results of plication augmentation of the augmented Anderson procedure in patients with infantile nystagmus syndrome and face turn.

Methods: In this retrospective study, all patients who underwent plication augmentation of the augmented Anderson procedure between August 2015 and November 2018 were included. Our study included patients older than 6 years with a face turn $>25^\circ$. We also included patients with residual face turns $\geq 15^\circ$ after Anderson-type procedures. The face turn was measured by a goniometer and also quantified with prisms placed with apex in the direction of the face turn. We plicated the medial rectus of one eye by 5.0 mm and lateral rectus of the fellow eye by 7.0 mm based on the direction of the face turn in addition to the augmented Anderson procedure. Patients were reviewed on the 1st postoperative day, 1st month, and every 6 months thereafter.

Results: Eight patients with a mean face turn of $27.5^\circ \pm 6.5^\circ$ underwent plication augmentation of the augmented Anderson procedure. Two patients had residual face turns after a previous Anderson-type procedure. We obtained a mean correction of $25^\circ \pm 6.5^\circ$ with a median prismatic correction of 45 prism diopters (PD) for each eye. The median face turn at the last review was 2.5° , and all patients were corrected to within 10° . Excluding patients operated for residual face turns, we had a mean dose response of 2.7 PD/mm and 1.7 $^\circ$ /mm of surgery on each eye. Five patients had an improvement in null zone visual acuity. Two patients had a restriction in ocular motility of -2 in the direction of the recessed extraocular muscle at the last review, and the remaining had a -1 restriction.

Conclusions: Plication augmentation of the augmentation Anderson procedure appears to be a safe and effective procedure for patients with infantile nystagmus syndrome and a face turn more than 25° . It may also be used for residual face turns more than 15° .

Keywords: Augmented Anderson procedure, Face turn, Infantile nystagmus syndrome, Plication augmentation, Strabismus

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INTRODUCTION

The surgical treatment of nystagmus involves two approaches – one that mechanically aims to move the null position to coincide with/be close to the primary position and another that attempts to broaden the null zone and improve the Expanded Nystagmus Acuity Function (NAFX) by reducing

the small signal gain of the ocular motor system. Toward the former aim was the surgeries proposed by Anderson, Goto, and Kestenbaum. Repka¹ suggested surgical amounts of 5 mm, 6 mm, 7 mm, and 8 mm split between the horizontal recti of the two eyes such that 13 mm of surgery was done on each eye. This approach frequently resulted in undercorrections

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when the face turn was more than 20°. Therefore, a 40% or 60% augmentation of this approach was proposed. Undercorrections may still happen with augmentations.¹⁻⁵ A reversal of face turn may happen with 4 horizontal recti surgery if periodic alternating nystagmus (PAN) has been missed in the preoperative evaluation, and this may be difficult to address.⁶

As another procedure, recessing one horizontal recti on each eye by large amounts (on the side opposite to the face turn – e.g., for a right face turn, the left lateral rectus, and right medial rectus may be recessed) was named as augmented Anderson procedure. This leaves two recti which can be operated in the event of undercorrections. A reversal of face turn caused by a missed PAN can be corrected by recessing the other two recti.⁶ Recessing by small amounts is ineffective as the orbital elastic forces draw the eyes back by the same amount. Large retroequatorial recessions, however, reduce muscle tone and severely weaken the contractility of yoke muscles.⁷ Gupta *et al.* reported on the results of fixed recessions of medial rectus by 9 mm and lateral rectus (of the fellow eye) by 12 mm. He reported a reduction in head posture from a preoperative $32.5^\circ \pm 5.8^\circ$ to $5.0^\circ \pm 8.7^\circ$ at 3 months in their series of 12 patients.⁶ Gräf *et al.* showed a reduction in head posture from a preoperative median of 35° to 10° at 5 months. He did equal recessions of 13 mm on the yoke muscles. None of the patients were overcorrected. Eleven out of eighteen patients had a residual face turn of <10°. Two patients needed further augmentation of the recession.⁸ It appears, therefore, that head turns >25° may not be fully corrected with the augmented Anderson procedure. In all three studies, there was a considerable variability in the correction achieved.⁶⁻⁸

Wang *et al.*⁵ pointed out that surgery on two recti may provide only mechanical benefits, and other benefits such as broadening of null zone and improvement in NAFX may not result. Tenotomy and reattachment on all four horizontal recti in both eyes reduced the small signal gain of the ocular motor system.⁴ He hypothesized that this may be achieved even by placing augmentation sutures on the recti (thereby minimizing complications of muscle surgery and sparing the ciliary circulation).⁹ Keeping the above studies in perspective, we propose that adding plications to the augmented Anderson procedures for face turns more than 25° may provide benefits of both worlds – potential reversibility in the event of a large overcorrection/reversal of head posture, sparing of ciliary circulation, and benefits of surgery on all four recti as proposed by Dell’Osso *et al.*⁹ There are very few studies that report on plications in the management of nystagmus. Schild *et al.*¹⁰ reported a success rate of 88% in his series of 42 patients. Equal amounts of recession/plication were done with doses varying from 5.5 to 10 mm depending on the face turn. Sauer *et al.*¹¹ reported good correction in three out of four patients with preoperative mean face turn of 22.5° with the use of plication alone in the yoke muscles. We report our findings in patients with the addition of plications to the augmented Anderson’s procedure in patients with infantile nystagmus syndrome with face turns of more than 25°.

METHODS

In this retrospective study, all patients with infantile nystagmus syndrome >6 years of age with a maximum elicited face turn (for distance) more than 25° who underwent augmentation of the augmented Anderson procedure between August 2015 and November 2018 were included in the study. The study also included residual face turns $\geq 15^\circ$ after a previous Anderson-type nystagmus surgery, where only recessions had been done. An informed consent was obtained from all patients/legal guardians. The study was approved by the institution review board and adhered to the tenets of the Declaration of Helsinki.

All patients underwent a comprehensive ophthalmic evaluation which included a detailed history, vision (monocular and binocular null zone visual acuity), refraction, cycloplegic refraction, comprehensive anterior, and posterior segment examination. A cover test was done for distance and near to elicit any manifest strabismus. Measurement with prisms was done when a manifest deviation was noted. Binocularity was checked with Bagolini striated glasses, and stereopsis was measured with the Randot stereotest. The maximal face turn was elicited for distance by asking the patient to look one line below the smallest line seen on an Early Treatment Diabetic Retinopathy Study chart. In infantile nystagmus, the direction of the nystagmus reverses across the null zone. The head posture in our patients was further confirmed by observing reversal of fast phase across the null zone. For example, if a patient had a left face turn that measured 30°, the nystagmus would be left beating in primary position and in the null zone. If the face was to be turned more than 30° to the left, the nystagmus changes direction to right beating.¹² This point of reversal was taken as the null zone. The face turn was measured with each eye fixing and confirmed over observation periods of over 8 min (the letters on the electronic chart were frequently changed to prevent the patient from memorizing) on at least two occasions. Patients with manifest latent nystagmus, PAN, and more than one null zone were excluded from the study. The face turn was measured with a goniometer. The face turn was also quantified with prisms with the patient fixing on 20/40 target. Equal prisms of increasing strength were progressively added to both eyes until a small reversal of face turn was



Figure 1: (a) Preoperative photograph of patient number 6 showing left face turn with a small chin depression. (b) Postoperative photograph of patient number 6 at last review near-total correction of the face turn

obtained. For purpose of analysis, any face turn that was not reversed with 50 prism diopters (PD) on both eyes was taken as having 60 PD (on each eye) of face turn. 50 PD was the largest prism strength available with us. To calculate the dose response, the face turn or PD (on each eye) was divided by the average millimeters of surgery performed on either eye, i.e., 16.5 mm.

All surgeries were done by the first author under general anesthesia for patients <14 years of age. For older patients, the surgery was done under topical anesthesia with conscious sedation and anesthetist supervision. Surgery aimed to move the eyes in the direction of the face turn. For example, in a patient with right face turn, the left lateral rectus was recessed by 12.0 mm, the left medial rectus was plicated by 5.0 mm, the right medial rectus recessed by 9.0 mm, and the right lateral rectus plicated by 7.0 mm. Adjustment for strabismus, when needed, was done by adjusting the surgical dosages in the nonfixing eye to correct the strabismus. For example, in patient number 3 [Table 1], the lateral rectus plication in the nonfixing left eye was increased to 8.5 mm (from 7 mm) to correct for the esotropia. Postoperatively, patients were placed on topical antibiotics and loteprednol tapered over 10 days. Patients were reviewed on day 1, 1st postoperative month, and every 6 months thereafter. At each postoperative visit, the maximum head turn was elicited by the method described above. From the 1st month onward, vision (monocular and binocular), refraction, cover test, tests for binocularity and stereopsis, and anterior and posterior segment examination were also done.

Patient data were entered in Microsoft® Excel 2010. Paired *t*-test was used to calculate the statistical significance between observations.

RESULTS

A total of eight patients (3 male and 5 female) underwent the surgery. Of these, two patients had surgery for a residual head turn. The median age was 14 years (range, 6–24 years). Patient characteristics are mentioned in Table 1. The mean preoperative face turn was $27.5^\circ \pm 6.5^\circ$. The median postoperative face turn at the 1st postoperative month and at last follow-up was 2.5° (range, -5° – 15°). The differences between the 1st postoperative month and the last review were not significant ($P = 0.43$). The median prismatic correction achieved for each eye was 45 PD, and the mean correction in head turn was $25^\circ \pm 6.5^\circ$. Excluding the patients operated for a residual face turn, we had a dose response of 2.7 PD/mm and 1.7°/mm of surgery on each eye. The median follow-up was 13 months (range, 6–32 months). Patient number 3 had a small esotropia of 16 PD which was fully corrected by the surgery [Table 1 - increasing lateral rectus plication to 8.5 mm]. The dose response in this patient was calculated by dividing the correction achieved (in degrees/PD per eye) by the average surgery per eye, i.e., 17.25 mm.

Three out of eight patients were fully corrected, while all patients were corrected to within 10° . One patient had a consecutive

face turn of 5° . This patient also had a chin elevation that was fully corrected by posterior tenectomy of the superior oblique. Three patients had a one-line improvement in their visual acuity, and two patients had improvement of over two lines at the last review. No patient had a loss of the best corrected visual acuity (BCVA). No patient had an induced strabismus due to surgery, and no other complication was noted. Patient number 3 had left suppression on Bagolini glasses with no recordable stereopsis. Three patients had stereopsis 400 s or arc or less, and the rest had good stereoacuity. This remained unchanged after surgery. Two patients had -2 restriction of ocular motility in the direction of the recessed muscle (symmetric without any induced deviation), and the rest had -1 restriction of ocular motility at the last follow-up. None of the patients needed further surgery. An example is shown in Figure 1.

DISCUSSION

The reported success rates of nystagmus surgeries in treating patients with infantile nystagmus syndrome and a head turn have varied substantially from 32% to 100%. Most studies have, however, included only a small number of patients with short periods of follow-up. It is known that the success rates drop with increasing follow-up. Good success rates have been reported with relatively small surgical dosages.^{9,13} Gräf *et al.*¹⁴ suggested forced BCVA testing (i.e., getting the patient to read 1–3 lines below the BCVA level) to bring out the maximum head posture.¹⁴ In an earlier study, Gräf *et al.*⁸ suggested noting the reversal of fast phase across the null zone as a means to reliably determine the maximum head posture.⁸ Unless these measures are followed, it is possible to underestimate the head posture and over report the success rates. In addition, we also used measurement by prisms to note a reversal of head posture.

Our success rate at 6 months is better than other studies on the augmented Anderson procedure [Table 2]. Only one patient had a small consecutive face turn. We also included two patients with residual face turn after previous nystagmus surgery. One patient had adjustments for strabismus and was fully corrected. A dose-effect relation of 2.7 PD/mm/eye and 1.7°/mm/eye was obtained with surgery. We are not aware of any other study that reports the dose-response effect with prism measurements. Gräf *et al.*¹⁴ reported a median dose-effect response of 1.42°/mm in patients who underwent the Kestenbaum Anderson procedure.¹⁴ Schild *et al.*¹⁰ reported a median dose-effect response of 1.35°/mm with the Kestenbaum Anderson procedures.¹⁰ It is likely that adding plications to the augmented Anderson procedure resulted in a higher dose-response effect. Recessions in the augmented Anderson procedure are more than the 60% augmentation of the Kestenbaum Anderson procedure.¹ This could cripple muscle function to a greater extent, resulting in a greater dose-response effect.⁸ Thus, plication augmentation of the augmented Anderson procedure may yield a greater surgical effect than equal dosing on the Kestenbaum Anderson procedure, and this needs to be studied in a larger number of patients. Our study shows that the procedure is safe and

Table 1: Clinical characteristics of our study patients

Age (years)	Diagnosis	Preoperative face turn	Preoperative null zone visual acuity	Preoperative prism measurement of face turn (PD) on each eye	Surgery done	1 st -month face turn (°)	Face turn at last review (°)	Prism quantification of the face turn at last review	Follow-up (months)	Null zone visual acuity last review	Ocular motility restriction in the direction of the recessed muscle	Result of Bagolini stereopsis (seconds of arc)
24	INS	30° left	20/32 OU	40	OD 12/5 and OS 9/7	0	0	0	6	20/20 OU	2-	Fusion, 50
23	INS	30° right	20/20 OU	45	OS 12/5 and OD 9/7	0	0	0	6	20/20 OU	1-	Fusion, 50
23	INS, left anisometropic amblyopia, left optic nerve hypoplasia	30° left	20/32 OD, 20/200 OS	>50	OD 12/5 and OS 9/8.5 mm	0	0	0	24	20/32 OD, 20/200 OS	1-	Left suppression no stereopsis
6	INS	35° right	20/40 OU	>50	OS 12/5 OD 9/7	15° right	10° right	20	32	20/32 OU	1-	Fusion, 400
14	INS, residual face turn	20° left, 20° chin elevation	20/32 OU	25	OS 12/5, OS 6→9/7 with OU PTSO	5° right	5° right	-14	6	20/32 OU	1-	Fusion, 40
8	INS	30° left, 5° chin depression	20/50 OD, 20/40 OS	>50	OD 12/5, OS 9/7	10° left	5° left	8	21	20/25 OU	1-	Fusion, 400
6	Cone rod dystrophy	30° left	20/80 OU	45	OD 12/5 OS 9/7	5° left	5° left	14	6	20/50 OU	1-	Fusion, 400
14	INS, OCA, residual face turn	15° right	20/32 OU	25	OD LR 7 mm plication, OS MR 5 mm plication	10° right	5° right	10	19	20/32 OU	2-	Fusion, 100

12/5: 12 mm lateral rectus recession and 5 mm MR plication, 9/7: 9 mm MR recession and 7 mm lateral rectus plication, 9/8.5: 9 mm MR recession and 8.5 mm lateral rectus plication, 9→12/5: 9 mm of lateral rectus recession increased to 12 mm and 5 mm of MR plication, 6→9/7: 6 mm of MR recession increased to 9 mm and 7 mm of lateral rectus plication. INS: Infantile nystagmus syndrome, PD: Prism diopters, OCA: Oculocutaneous albinism, LR: Lateral rectus, MR: Medial rectus, PTSO: Posterior tenectomy of the superior oblique

Table 2: A review of studies on the augmented Anderson procedure and plication in infantile nystagmus syndrome

Author (year)	Number of patients	Surgery done	Preoperative face turn (°)	Postoperative face turn at last follow-up (months)	Success rates for residual <10°	Resurgery rate	Follow-up	Dose response	Effect on null zone visual acuity
Arroyo-Yllanes <i>et al.</i> (2002)	21	2 mm retroequatorial recession of MR on one eye and LR on the fellow eye	≥35-40	15° - -15°	12/21 5 consecutive of which 3 were within 10°	Nil	6-84 months	Not mentioned	Not mentioned
Gupta <i>et al.</i> (2006)	12	9 mm MR recession, 12 mm LR recession yoke muscles	32.5±5.8	5.0±8.7°	6/12, 1 consecutive	Nil	3 months	Not mentioned	Borderline significant improvement on ETDRS (<i>P</i> =0.053)
Schild <i>et al.</i> (2013)	42	Plication + recession based on KA principle equally 5.5-10 mm	30	10°	72%	2/42	6 weeks-11 years	1.35°/mm/eye	Not mentioned
Gräf <i>et al.</i> (2019)	29	13 mm yoke muscles OU	35° at 5 m/20° at 0.3 m	12° at 5 m/6° at 0.3 m	75%	3/29	4-15 months	Not mentioned	Not mentioned
Sauer <i>et al.</i> (2019)	4	Plication on yoke muscle 5-7.5 mm MR, 7-12 mm lateral rectus	22.5	2.5	75%	1/4	6-18 months	Not mentioned	Not mentioned
Our study	8	9 mm MR recession, 7 mm LR Plication on one eye and 12 mm LR recession, 5 mm MR Plication on the other eye	27.5±6.5	3.75	87.5%	Nil	6-32 months	2.5 PD/mm/eye 1.6°/mm/eye	5/8 patients had 1-2 line improvement

KA: Kestenbaum Anderson, MR: Medial rectus, LR: Lateral rectus, ETDRS: Early treatment diabetic retinopathy study, PD: Prism diopter

effective and produces only minimal motility restrictions with no induced strabismus.

Sharma¹² had suggested adding resections of 6 mm for the medial rectus and 9 mm for the lateral rectus to the augmented Anderson procedure for head turns larger than 45°. They also recommended more surgery on the lateral rectus when compared to the medial rectus (when treating patients with infantile nystagmus syndrome and a head posture) to maintain competence as surgery on the latter is more effective.¹² Our numbers were more conservative as the face turns were also <45°. We are not aware of any studies reporting on augmentation of the augmented Anderson procedure. It may also be done as a second procedure if only two recti have been recessed in the previous procedure. Five of eight patients in our study including one patient with cone-rod dystrophy had 1-2 line improvement in the BCVA. Other studies have also reported an increase in null zone visual acuity.⁷

One of our patients had a vertical head posture which was fully corrected by posterior tenectomy of the superior oblique. Arroyo-Yllanes *et al.*⁷ reported resolution of head posture in all three axes after modified Anderson procedure.⁷ These results were not reproduced by other authors who reported persistence of the head posture.⁸ Plication of the yoke muscles has a distinct advantage in this situation by sparing the ciliary circulation. This would permit surgery on one more vertical rectus muscle if a chin up or chin down head posture persists after horizontal muscle surgery. Plication also is potentially reversible in the event of severe overcorrection/reversal in a missed PAN.

The limitation of our study includes a small number of patients and lack of eye movement recordings. Therefore, we could not demonstrate a broadening of the null zone with plication and an increase in NAFX as reported by Wang *et al.*⁵ Five of eight patients had an improvement in null zone visual acuity that may represent a response to four muscle surgery.

To sum up, plication augmentation of the augmented Anderson procedure is an effective and safe procedure for patients with face turns >25°. Its results need to be evaluated in a larger number of patients with eye movement recordings.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for images and other clinical information to be reported in the journal. The parents understand that names and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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