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Evaluation of the Role of Displacement Surgery in the Management of Congenital Nystagmus

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Abstract

Introduction: The aim of this study was to assess the effectiveness of displacement surgery in damping of ocular oscillation and management of compensatory head posture in patients of congenital nystagmus.

Methods: This study was carried out in 2014. The participants were 50 patients with congenital nystagmus that dampens in a certain direction of gaze with or without abnormal head posture and with or without strabismus. Patients underwent the augmented modified Kestenbaum (augmented parks) procedure. Data were analyzed by SPSS version 15, using Chi-square text, exact test, and Mann Whitney U test.

Results: In this study, 12 patients (80%) were noticed clinically to have damping of nystagmus while 3 patients (20%) had no damping of nystagmus. Twelve patients (80.0%) had no post-operative abnormal head position, while 3 patients (20.0%) had residual abnormal head position. There was a statistically significant difference between the pre-operative BCVA in primary position and post-operative BCVA in primary position (p = 0.001 for both right and left eyes).

Conclusion: The displacement surgeries (e.g. Kestenbaum procedure) for the treatment of patients with congenital nystagmus is an effective procedure for correction of abnormal head posture, improving the visual acuity in the primary position and damping of nystagmus in patients with congenital nystagmus who have null points away from the primary position.

Keywords: Nystagmus, Abnormal head posture, Tenotomy, Immobilization surgery, Displacement surgery

1. Introduction

Nystagmus can be congenital or acquired, visual acuity in congenital nystagmus has been found to be principally related to the duration of foveation periods, at which time, the target image falls onto the fovea, and eye velocity reduces. A high changeability of the eye position during the foveations, encumbers a stable placement of the target image on the intermediary fovea and consequently reduces visual acuity (1). Cogan has categorized congenital nystagmus into two principal types, termed sensory defect nystagmus and motor defect nystagmus (2). Motor defect nystagmus is a type of congenital nystagmus in which the main defect is in the efferent mechanism with possible involvement of the centers or pathways for conjugate oculomotor control, and the amplitude and frequency may lessen, or the nystagmus can completely disappear in a random position of gaze. This can cause the patient to adjust head posture in order to recover visual acuity with the eyes in the area of least nystagmus (null point, privileged area, neutral zone) (3). The main objective of extraocular muscle surgery for nystagmus is to lessen incidences of abnormal head posture (AHP) or nystagmus, as the main surgical concept to reduce AHP is determined by the characteristics of the nystagmus (4). The two main interventions which have been promoted in the management of congenital nystagmus are the immobilization intervention and the displacement intervention. If only a certain version innervation reduces the nystagmus, Kestenbaum displacement surgery is indicated. By combined recess and resect surgery on each eye, both eyes are turned directly towards the AHP. Then the neutral zone of the nystagmus moves to the straight gaze position. The concept of artificial divergence is encouraging when, not only certain

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version innervations but also convergence reduces the nystagmus (4). Recently, it was hypothesized that tenotomy of the recti muscles and reattachment in their original insertions, although neither weaken nor strengthen the muscles, improves the nystagmus acuity function and dampens the nystagmus amplitude by removal of the tendon organ responsible for proprioception (5).

2. Material and Methods

2.1. Patients and selection criteria

This study was conducted in 2014 and included fifteen patients with congenital nystagmus that dampens in a certain direction of gaze with or without abnormal head posture and with or without strabismus. The following were set as the inclusion and exclusion criteria:

1) Inclusion criteria: Patients with congenital nystagmus that had a null point in which the nystagmus amplitude decrease
2) Exclusion criteria: Patients with acquired nystagmus or who had previous extraocular muscle surgery

2.2. Pre-surgical Assessment

The following methods were applied in examination of all patients prior to the surgery:

1) Refraction: cycloplegic refraction was done.
2) Assessment of visual acuity and best corrected visual acuity in the primary position and position of minimal nystagmus using the illiterate E test (the E game), the average of pre-operative BCVA in primary position was 0.3 ± 0.15 ranging (0.1-0.6) and BCVA in null zone position was 0.6 ± 0.1 ranging (0.4-0.8).
3) Types of Nystagmus: it was bilateral, conjugate, jerky, to the right in eight patients, to the left in six patients and upwards in one patient.
4) Presence of null point: it was present in all cases, eight patients had it on levoversion, five patients had it on dextroversion, one patient had it on deorsumversion and one patient had it on convergence.
5) Assessment of abnormal head posture: the pre-operative abnormal head positions were assessed while the patient fixated on a small distant object and estimated to the nearest 15° preoperatively, one patient had no AHP and the nystagmus dampens on convergence, eight patients had face turn to right which was 30° in five patients, 45° in three patients, five patients had face turn to left which was 30° in four patients, 45° in one patient, the last patient had chin elevation 20°.
6) Complete ocular motility examination: there were no limitations of ocular motility in the cases of the study.
7) Cover test and prism cover test for heterophoria: seven patients were orthotropic, one patient was esophoric, six patients were exophoric and one patient had intermittent exotropia.

2.3. Surgical procedure

Fourteen of fifteen patients underwent the augmented modified Kestenbaum (augmented parks) procedure, with the 40% augmentation formula used for patients with 30° face turn, and the 60% augmentation formula used for patients with 45° face turn or more, the eyes were moved in the direction of the face turn, using combined recession-tucking of the horizontal recti. The patient with chin elevation underwent combined 6 mm tucking-recession respectively of superior and inferior recti of both eyes, to move eyes upwards in the direction of abnormal head posture. Extra ocular muscles recession was used as a weakening technique, and extra ocular muscles tucking was used as a strengthening technique. All patients were operated under general anesthesia, prior to surgery the periorbital skin was cleaned with povidone-iodine and draping of the face and head was performed. A lid speculum was used to retract the lids as well as to keep the lashes out of the field. The globe was fixed with a traction suture to prevent rotation and to maintain the field of view. An incision through conjunctiva and tenon's capsule was made at the limbus or the fornix, and a surgical plane was created down to bare sclera, with both the conjunctiva and tenon's retracted. A muscle hook was then used to isolate the extraocular muscle, and a larger muscle hook was then passed behind the first hook to ensure all muscle fibers were isolated. The insertion of the muscle was then exposed by removing any tenon's attachments. The muscle was carefully cleaned of all its fascial attachments and check ligaments. The anterior insertion of the muscle was cleaned and sufficiently exposed for recession of the extraocular muscle. To secure the muscle insertion, double armed 6-0 absorbable sutures (Vicryl) on a spatulated needle were used. Then, in order to mark the distance posterior from the poles of the initial insertion for placement of the muscle, calipers which were set at a predetermined amount were used. The posterior sclera was marked to correspond with both the superior and inferior aspects of the muscle. The needles which were attached to the muscle were passed through partial thickness of the sclera with visualization of the needle through the superficial sclera lamellae. By adding traction to the muscle hook and the sutures to keep them away from the insertion, the muscle was disinserted from the globe. To disinsert the muscle from the sclera, Westcott scissors were used and sutures were tied and trimmed (Figure 1). For tucking of the extraocular muscle, double armed 6-0 Absorbable sutures (Vicryl) on a spatulated needle were used to secure the muscle at a point marked with calipers set at a predetermined amount. The needles attached to the muscle were passed through the original insertion of the muscle, then the sutures were then tied and trimmed (Figure 2).

2.4. Post-surgical follow up

The patients were seen postoperatively one day, one week and one month following surgery, and were examined for:

1) Refraction: using autorefractometer.
2) Assessment of visual acuity and best corrected visual acuity in the primary position using the illiterate E test (the E game), the average of post-operative BCVA in primary position was 0.6 ± 0.1 ranging (0.4-0.8).
3) Assessment of abnormal head posture: the post-operative abnormal head positions were assessed while the patient fixated on a small distant object and estimated to the nearest 15°, twelve patients had no residual AHP, one patient had residul residual 15° of face turn to the left and two patients had residual 15° of face turn to the right.
4) Complete ocular motility examination: restriction of motility after operation varied in our series.
5) Cover test and prism cover test for heterophorias: six patients were orthotropic, one patient was esophoric, five patients were exophoric and three patients had exotropia.

2.5. Statistical methods

Data were statistically described in terms of mean ± standard deviation (± SD), median and range, or frequencies (number of cases) and percentages when appropriate. Comparison of numerical variables was done using Mann Whitney U test for independent samples. For comparing categorical data, Chi square test was performed. Exact test was used instead when the expected frequency was less than 5. P values less than 0.05 was considered statistically significant. All statistical calculations were done using SPSS (SPSS Inc., Chicago, IL, USA) version 15.

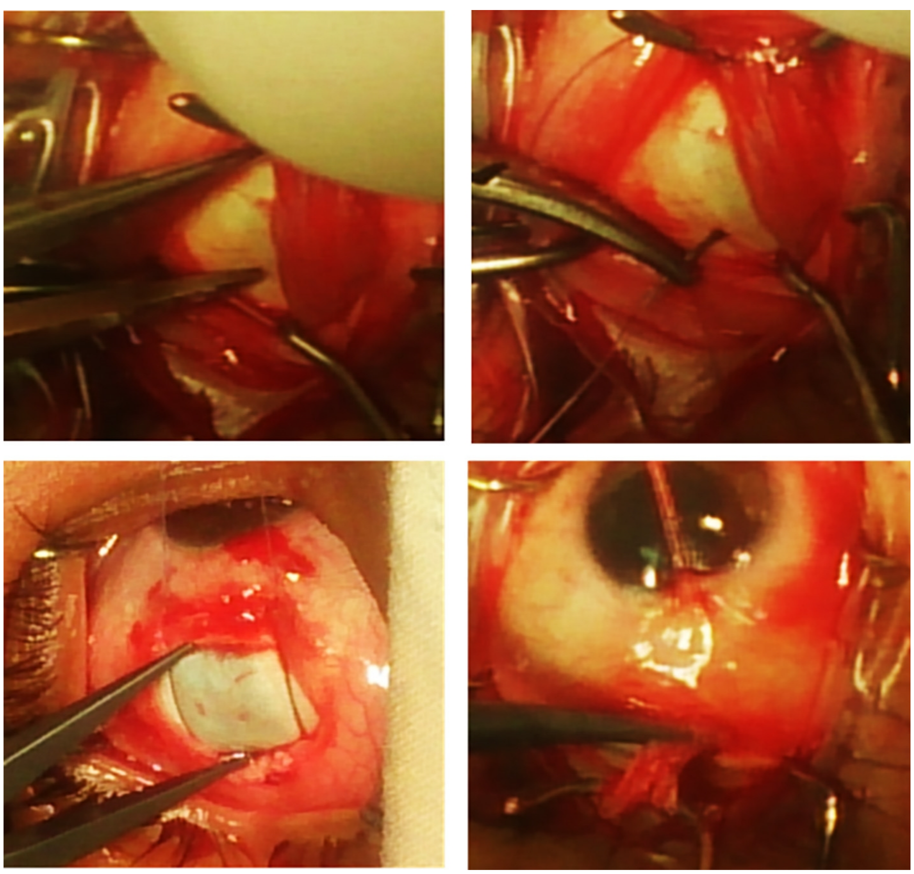

Figure 1. Extraocular muscle recession

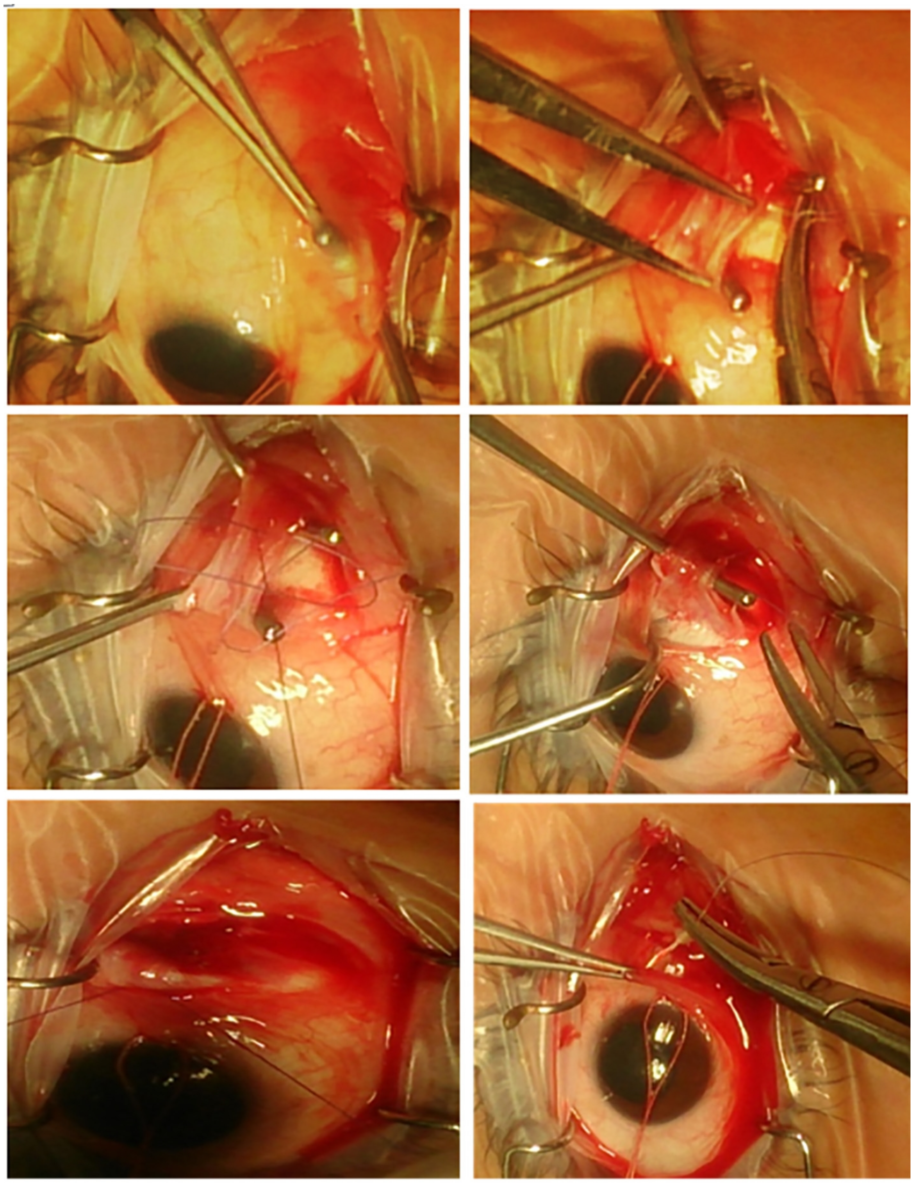

Figure2. Extraocular muscle tucking

3. Results

The age of patients was ranged from 4-10 years. Age at onset of the complaint: Most cases (73.30%) started at birth, the remaining cases (26.70%) started within 6 months of life, family history of nystagmus was positive in one case and maternal history of prematurity was positive in four cases. Fifteen patients had congenital nystagmus that blocks in a certain direction of gaze with or without abnormal head posture and with or without strabismus. The mean age at the time of surgical intervention was 8 years ± 2SD ranging (4-10) years. The majority of cases (73.30%) started at birth, the remaining cases (26.70%) started within 6 months of life. The average of pre-operative BCVA in primary position was 0.3 ± 0.15 ranging (0.1-0.6) and BCVA in null zone position was 0.6 ± 0.1 ranging (0.4-0.8) while the average of post-operative BCVA in primary position was 0.6 ± 0.1 ranging (0.4-0.8). There was a statistically significant difference between the pre-operative BCVA in primary position and post-operative BCVA in primary position ($p = 0.001$ for both right and left eyes). The average of vision gain in primary position was 0.3 ± 0.1, SD ranging from 0.1 to 0.4. The null zone position was on levoversion in eight patients (53.30%), on dextroversion in five patients (33.30%), one patient (6.70%) had it on deorsumversion and the last patient (6.70%) had it on convergence. Regarding the Abnormal head position (AHP), There were no pre-operative AHP in one case (dampens on convergence), four patients had face turn to the left 30°, one patient had face turn to the left 45°, five patients had face turn to the right 30°, three patients had face turn to the right 45°. In addition, in the post-operative follow up period twelve patients (80.0%) had no AHP, one patient (6.7%) had residual 15° face turn to the left, and two patients (13.3%) had residual 15° face turn to the right. Eighty percent of our patients had no residual AHP and 20.0% had residual AHP (Table 1). Chi-square test showed that there was a statistically significant difference between pre-operative and post-operative abnormal head positions (p=0.044). With regard to the pre-operative ocular alignment, seven patients were orthotropic, one patient was esophoric, six patients were exophoric and one patient had intermittent exotropia. In the post-operative follow up period six patients were orthotropic, one patient was esophoric, five patients were exophoric and three patients had exotropia. By using the Chi-square test, there was a statistically significant difference between pre-operative and post-operative ocular alignment ($p \leq 0.001$) (Table 2).

Table 1. Pre-Operative and Post-Operative Abnormal head Position

AHP		n	%
Pre-operative AHP	No AHP	1	6.7
	Face turn to the left 30°	4	26.7
	Face turn to the left 45°	1	6.7
	Face turn to the right 30°	5	33.3
	Face turn to the right 45°	3	20.0
	Chin elevation 20°	1	6.7
Post-operative AHP	No AHP	12	80
	Face turn to the left 15°	1	6.7
	Face turn to the right 15°	2	13.3

Table 2. Pre-operative and post-operative ocular alignment

Ocular alignment		n	%
Pre-operative	Orthotropia	7	46.7
	Esophoria	1	6.7
	Exophoria	6	40.0
	Intermittent Exotropia	1	6.7
Post-operative	Orthotropia	6	40.0
	Esophoria	1	6.7
	Exophoria	5	33.3
	Exotropia	3	20.0

4. Discussion and conclusions

This study was conducted to evaluate the effectiveness of displacement surgery in damping of ocular oscillation and management of head posture in patients of congenital Nystagmus and null zones. The study group included fifteen patients with congenital Nystagmus that dampens in a certain direction with or without abnormal head posture. Five patients (33.30%) had null point in Dextroversion, eight patients (53.30%) in Levoversion. One patient (6.7%) in Deorsumversion, and 1 patient (6.70%) had nystagmus that dampens in convergence. One patient had no AHP but his nystagmus intensity dampens on convergence, nine patient had face turn 30° which was to left in four patients and to right in five patients, four patients had face turn 45° which was to left in one patient and to right in three patients, the last patient had chin elevation 30°. Fourteen patients with abnormal head posture underwent augmented modified Kestenbaum surgery, with the 40% augmentation formula used for patients with 30° face turn, and the 60% augmentation formula used for patients with 45° face turn or more, the eyes were moved in the direction of the face turn using combined recession-tucking of the horizontal recti. The patient with chin elevation underwent combined 6 mm tucking-recession respectively, of superior and inferior recti of both eyes to move eyes upwards in the direction of abnormal head posture. The patient who had null point in convergence underwent artificial divergence surgery (bilateral medial rectus recession). In this study, 12 patients (80%) were noticed clinically to have damping of nystagmus while 3 patients (20%) had no damping of nystagmus. Twelve patients (80.0%) had no post-operative abnormal head position, while 3 patients (20.0%) had residual abnormal head position. The results of our study are consistent with the results of a retrospective study in which the clinical findings for eight patients who consecutively underwent treatment in the University Eye Hospital of Cologne between 2001 and 2007 were investigated. The patients were aged 6 to 16 years; median age was 6.5 years. For all patients, surgery was to correct abnormal head posture due to infantile nystagmus; Surgery was successful in seven of the eight patients (87.5%), with a reduction of the head posture to less than 10 degrees (6). There is a difference between the result of our study and the study performed on fifteen patients at the Pediatric Ophthalmology Department of Wills Eye Hospital in which there were no residual AHP in 53.5% of patients and 46.5% had residual AHP 15° or less. Pre-operative best corrected visual acuity in the primary position was 0.3 ± 0.1 and post-operative best corrected visual acuity was 0.6 ± 0.1, and this means that vision gained in primary position post operatively was 0.3 ± 0.1 and there was a statistically significant difference between post-operative visual acuity and visual acuity in primary position pre-operatively (p=0.001). The improvement in visual acuity in our study is consistent with the results of the study performed on twenty-eight patients at the Department of Pediatric Ophthalmology and Strabismus, Bombay City Eye Institute and Research Centre in which the average null zone logarithm of the minimum angle of resolution acuity was 0.42 preoperatively, which improved significantly to 0.33 postoperatively (7). The results of our study regarding the efficacy of different forms of displacement surgery in reducing nystagmus intensity, correcting abnormal head posture and improving visual acuity is consistent with the results of several studies like the study performed at the ocular motility clinic of the Royal Victorian Eye and Ear Hospital and published by Taylor JN on Feb 1987 titled Surgical Management of Congenital Nystagmus (8). Also, the results in this study were in agreement with results achieved by Lee IS et al. in 2000, the average preoperative face turn in the 63 patients with horizontal nystagmus was 31.9 degrees with an average postoperative face turn of 5.2 degrees (9). The average net change in face turn was 26.7 degrees. The average duration of time from surgery to final examination was thirteen months. Fifty-six out of 63 patients (89%) achieved a straight head position or a residual face turn of 10 degrees or less. Similar results seen in Chang YH study in 2007 in which 45 out of 51 patients (88.2%) who underwent Parks' modified procedures showed face turn less than 10 degrees. In the follow-up of an average 29 months, 36 out of 41 patients (87.8%) with 6-7-6-7 mm procedure had face turn less than 10 degrees (10).

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Conflict of Interest:
There is no conflict of interest to be declared.

Authors' contributions:
All authors contributed to this project and article equally. All authors read and approved the final manuscript.

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