Evaluation of the role of displacement surgery in the management of congenital nystagmus.

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Abstract: Purpose: To evaluate the effectiveness of displacement surgery in damping of ocular oscillation and management of compensatory head posture in patients of congenital nystagmus. **Methods**: Fifteen 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. **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-value = 0.001 for Right eye & P-value = 0.001 for Left eye). **Conclusion**: The displacement surgeries (e.g. Kestanbaum 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 nystagmusin patients with congenital nystagmus who has null points away from the primary position.

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Introduction

Nystagmus may be congenital or acquired, Visual acuity in congenital nystagmus has proven to be primarily related to the duration of foveation periods, during which the image of a target falls onto the fovea and eye velocity slows down. A high variability of the eye position during the foveations hinders a stable placement of the target image on the centralmost fovea and consequently decreases visual acuity. **(Cesarelli M, 2000)**

Cogan has classified congenital nystagmus into two principal types, which he named *sensory defect nystagmus* and *motor defect nystagmus.* **(Louis F, 2007)**

Motor defect nystagmus is a form of congenital nystagmus in which the primary defect is in the efferent mechanism, possibly involving the centers or pathways for conjugate oculomotor control, and the amplitude and the frequency may decrease or the nystagmus may disappear completely in one position of gaze. This may cause the patient to assume an anomalous head posture to improve visual acuity with the eyes in the position of least nystagmus (null point, neutral zone, privileged area. **(Spielmann A, 2001)**

The goal of extraocular muscle surgery for nystagmus is to reduce abnormal head posture (AHP) and/or nystagmus, the main surgical concept to reduce AHP are dependent on the characteristics of the nystagmus. **(Graf M, 2001)**

Two main interventions have been advocated in the management of congenital nystagmus: the displacement intervention and the immobilization intervention. Kestenbaum displacement surgery is indicated if solely a certain version innervations reduce the nystagmus. Both eyes are turned parallel in the direction of the AHP by combined recess and resect surgery on each eye.thus, the neutral zone of the nystagmus shifts to the straight gaze position, the concept of artificial divergence is promising when not only a certain version innervations but also convergence reduces the nystagmus. **(Graf M, 2001)**

Recently it was hypothesized that tenotomy of the recti muscles and reattachment in their original insertions although neither weaken nor strengthen the muscles, it improves the nystagmus acuity function and dampen the nystagmus amplitude by removal of the tendon organ responsible for proprioception. **(Weil A, 2013)**

Patients and Methods

This study 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.

- Inclusion criteria: Patients with congenital nystagmus that have a null point in which the nystagmus amplitude decrease.

- Exclusion criteria: Patients with acquired nystagmus or who had previous extraocularmuscle surgery.

Presurgical Assesment:

History: Age of the patient: the age was ranged from 4-10 years, Age at onset of the complaint: Most of 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.

Examination:

- Refraction: cycloplegic refraction was done.
- 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).
- 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.
- Presence of null point: it was present in all of cases, eight patients had it on levoversion, five patients had it on dextrovoversion, one patient had it on deorsumversion and one patient had it on convergnce.
- 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 has 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°.
- Complete ocular motility examination: there were no limitation of ocular motility in the cases of the study.
- Cover test and prism cover test for heterophorias: seven patients were orthotropic, one patient was esophoric, six patients were exophoric and one patient had intermittent exotropia.

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.
- 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, 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 extraoculr muscle, and 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.

For recession of the extraocular muscle:

- The anterior insertion of the muscle is cleaned and adequately exposed. A double armed 6-0 Absorbable sutures (Vicryl) on a spatulated needle was used to secure the muscle insertion, Calipers set at a predetermined amount were used to mark the distance posterior from the poles of the original insertion for placement of the muscle, The posterior sclera was marked to correspond with both the superior and inferior aspects of the muscle, The needles attached to the muscle were passed partial thickness through the sclera with visualization of the needle through the superficial sclera lamellae, The muscle was disinserted from the globe by placing traction on the muscle hook and the sutures to keep them away from the insertion. Westcott scissor was then used to disinsert the muscle from the sclera. The sutures were then tied and trimmed. (Figure 1)

For tucking of the extraocular muscle:

- A 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, The sutures were then tied and trimmed. (Figure 2).

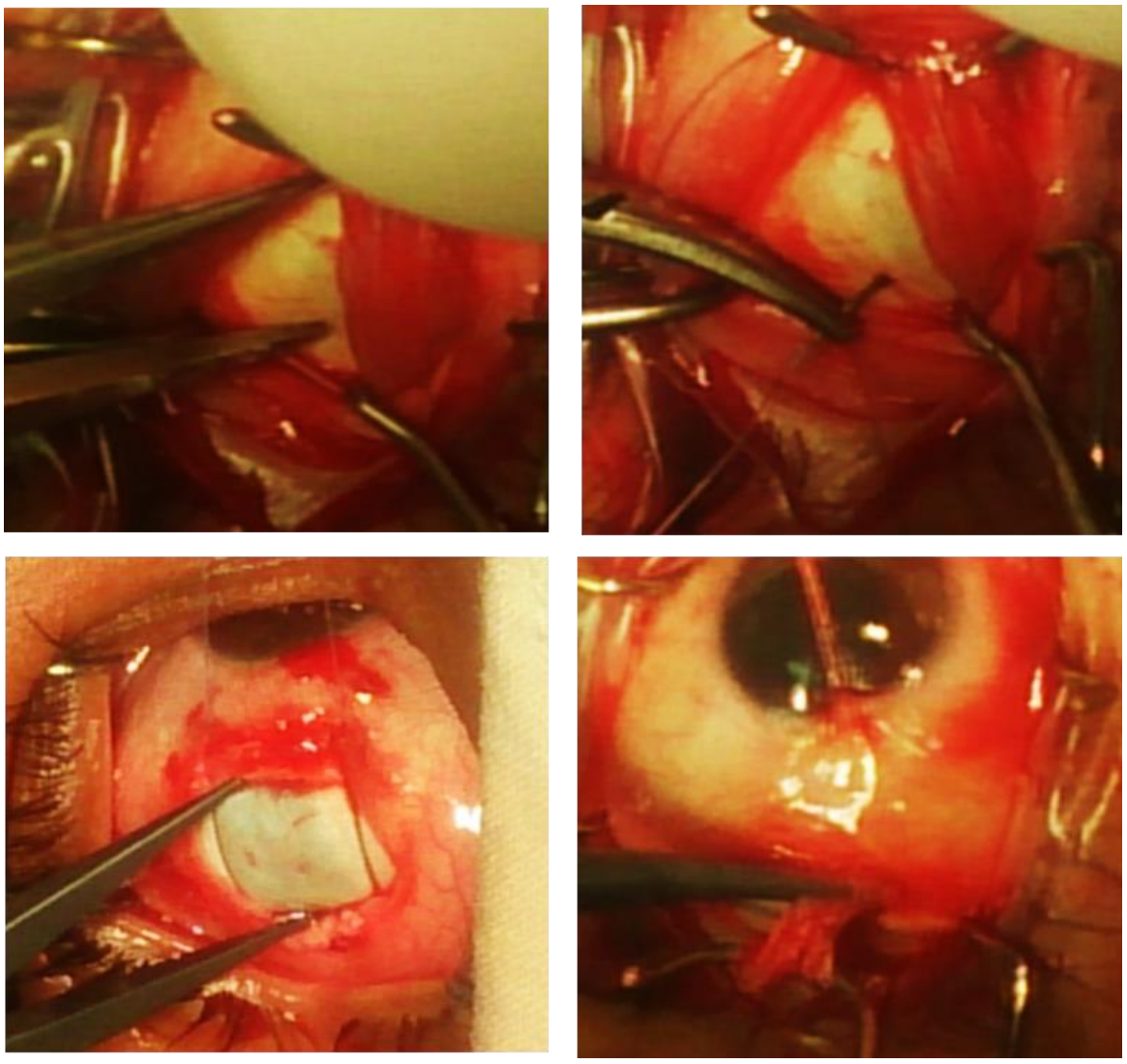

Figure (1) Extraocular muscle recession.

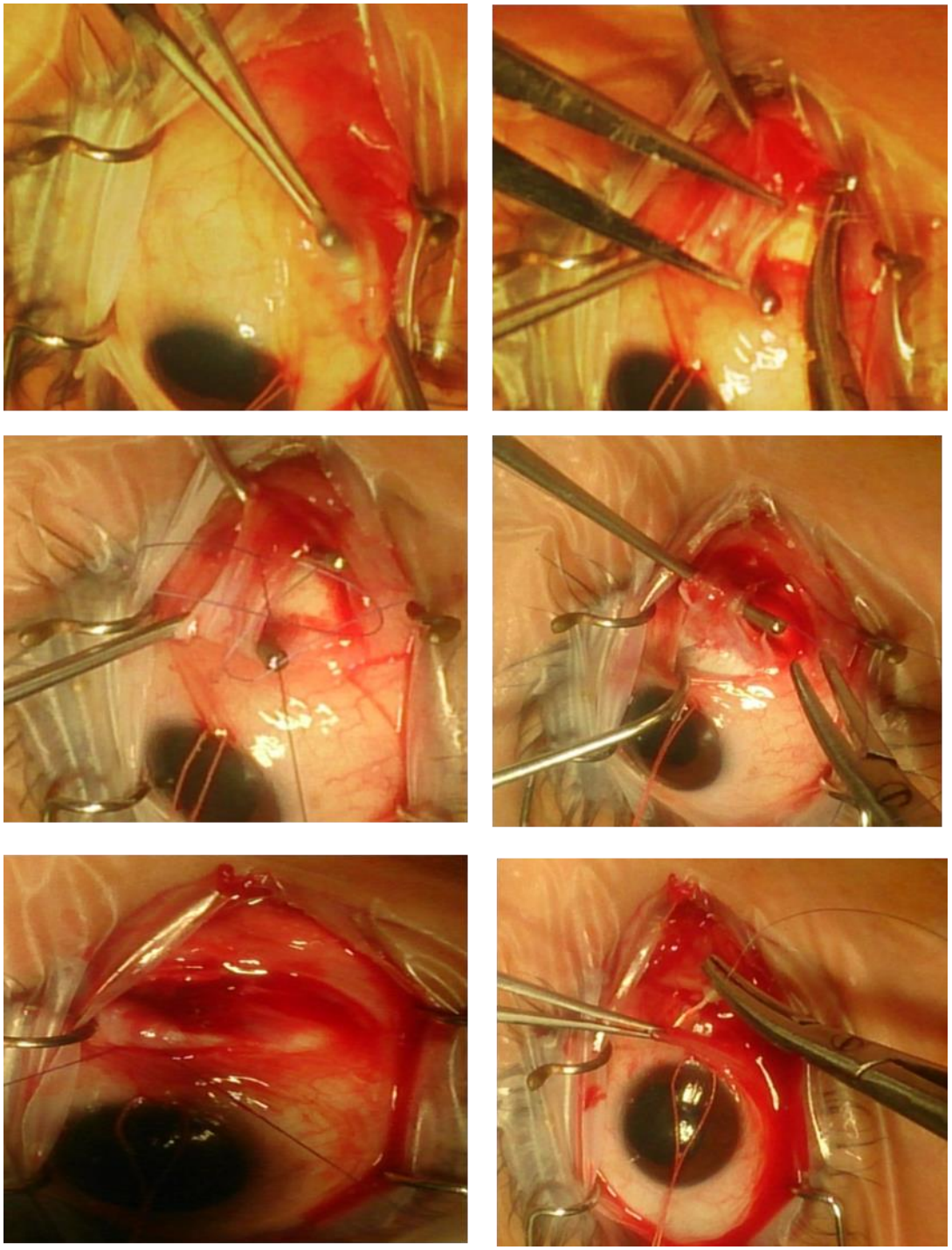

Figure (2) Extraocular muscle tucking

***Post-surgical Follow up*:**

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

- Refraction: using autorefractometer.
- 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).
- Assessment of abnormal head posture: the post-operative abnormal head positions were assessed while the patient fixated on a small distant objectand estimated to the nearest 15°, twelve patient has no

residual AHP, one patient had residul 15° of face turn to the left and two patients has residual 15° of face turn to the right.

- Complete ocular motility examination: restriction of motility after operation varied in our series.
- Cover test and prism cover test for heterophorias: six patients were orthotropic, one patient was esophoric, five patients were exophoric and three patient had exotropia.

Statistical method:

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 ($\chi 2$) test was performed. Exact test was used instead when the expected frequency is less than 5. P values less than 0.05 was considered statistically significant. All statistical calculations were done using computer program SPSS (Statistical Package for the Social Science; SPSS Inc., Chicago, IL, USA) version 15 for Microsoft Windows.

Results:

Fifteen patients with 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.

***Pre-operative and post-operative visual acuity**

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-value = 0.001for Right eye & P-value = 0.001for Left eye). The average of vision gain in primary position was 0.3±0.1 SD ranging (0.1-0.4). (table 1)

Table (1): Pre-operative and post-operative visual acuity

Pre-operative	Right eye	Left eye	Post-operative	Right eye	Left eye
	Mean ± SD			Mean ± SD	
BCVA Primary position	0.3±0.1	0.3±0.1	BCVA primary position	0.6±0.1	0.6±0.1
BCVA NZP	0.6±0.1	0.6±0.1			

*** Null Point position**

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.(table 2)

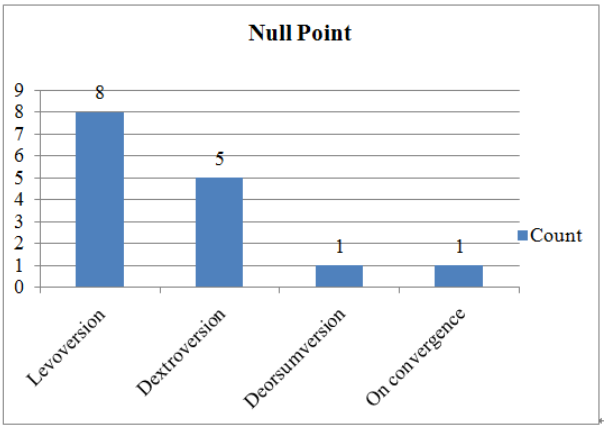

Figure 3: Null point position

Table (2): Null Point position:

		NO	%
Null point	Levoversion	8	53.30%
	Dextroversion	5	33.30%
	Deorsumversion	1	6.70%
	On convergence	1	6.70%

***Abnormal head position:**

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, on patient (6.7%) had residual 15° Face turn to the left, two patients (13.3%) had residual 15° Face turn to the right.80.0% of our patients had no residual AHP and 20.0% had residual AHP. (table 3)

Table (3): Pre-Operative and Post-Operative Abnormal head Position:

	Pre-operative AHP	Number of patients	%	Post-operative AHP	Number of patients	%
AHP	No AHP	1	6.7%	No AHP	12	80.0%
	Face turn to the left 30°	4	26.7%			
	Face turn to the left 45°	1	6.7%	Face turn to the left 15°	1	6.7%
	Face turn to the right 30°	5	33.3%			
	Face turn to the right 45°	3	20.0%	Face turn to the right 15°	2	13.3%
	Chin elevation 20°	1	6.7%			

By using Chi-square test, there was a statistically significant difference between pre-operative and post-operative Abnormal Head positions (P-value = 0.044).

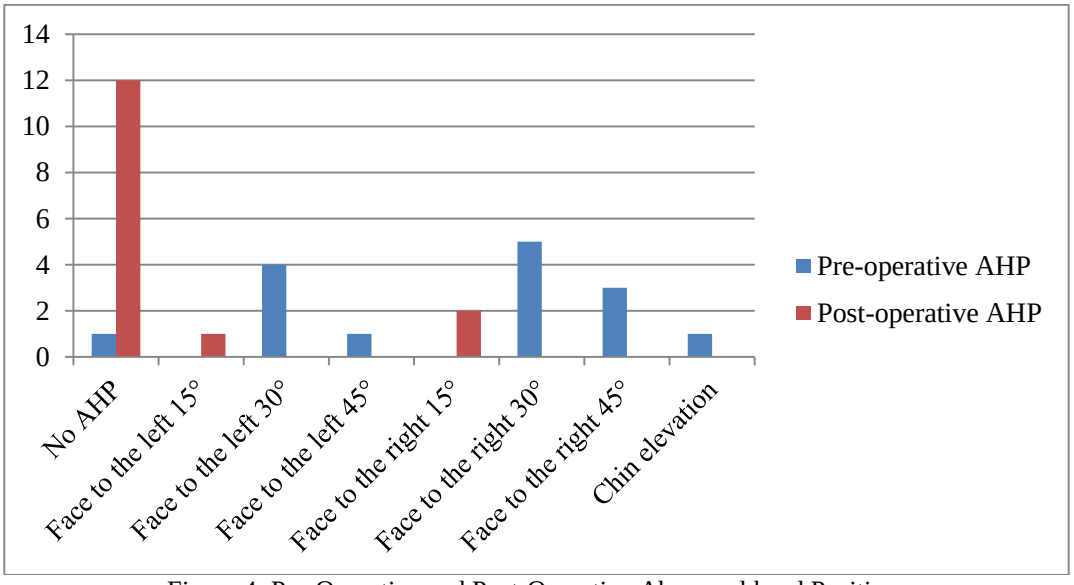

Figure 4: Pre-Operative and Post-Operative Abnormal head Position:

***Ocular alignment:** As regard 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 patient had exotropia. By using the Chi-square test, there was a statistically significant difference between pre-operative and post-operative ocular alignment (P-value = <0.001). (table 4)

Table (4): pre-operative and post-operative ocular alignment:

	Pre-operative	NO	%	Post-operative	NO	%
Ocular alignment	Orthotropia	7	46.7%	Orthotropia	6	40.0%
	Esophoria	1	6.7%	Esophoria	1	6.7%
	Exophoria	6	40.0%	Exophoria	5	33.3%
	Intermittent Exotropia	1	6.7%	Exotropia	3	20.0%

Discussion:

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. 5 patients (33.30%) had null point in Dextroversion, 8 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 is 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 (Baghari et al., 2009).

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% has 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 value=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. (Kumar A, 2011)

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. (Taylor JN, 1987).

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. 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.

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References:

1. Bagheri A, Aletaha M and Abrishami M: Kestanbaum procedure for nystagmus related abnormal head position. Graefes Arch Clin Exp Ophthalmol. 2009 Oct; 247(10):1395-400. Doi: 10.1007/s00417-009-1083-9,2009.

2. Cesarelli M, Bifulco P, Loffredo L and Bracale M: Relationship between visual acuity and eye position variability during foveations in congenital nystagmus. Doc Ophthalmol. 2000 Jul;101(1):59-72.
3. Chang YH, Chang JH, Han SH, et al.: Outcome study of two standard and graduated augmented modified Kestenbaum surgery protocols for abnormal head postures in infantile nystagmus. Binocul Vis Strabismus Q; 22: 235-41, 2007.
4. Graf M, Droutsas K, Kaufmann H: Surgery for nystagmus related head turn: Kestenbaum procedure and artificial divergence. Graefes Arch Clin Exp Ophthalmol. 2001 Jun;239(5):334-41, 2001.
5. Kumar A, Shetty S, Vijayalakshmi P and Hertle RW: Improvement in visual acuity following surgery for correction of head posture in infantile nystagmus syndrome. J Pediatr Ophthalmol Strabismus. 2011 Nov-Dec;48(6):341-6. doi: 10.3928/01913913-20110118-02, 2011.
6. Lee IS, Lee JB, Kim HS, Lew H, Han SH: Modified Kestenbaum surgery for correction of abnormal head posture in infantile nystagmus: outcome in 63 patients with graded augmentaton. Binocul Vis Strabismus Q.; 15(1):53-8, 2000.
7. Louis F. Dell'Osso, Richard W. Hertle and Robert B: Daroff: Sensory and motor nystagmus erroneous and misleading terminology based on misinterpretation of David Cogan's observation. Arch Ophthalmol;125(11):1559-1561, 2007.
8. Spielmann A: Congenital nystagmus: clinical types and their surgical treatment. Ophthalmologica (Basel); 182:65-72, 2001.
9. Taylor JN and Jesse K: Surgical management of congenital nystagmus. Aust NZ J Ophthalmol; 15:25-34, 1987.
10. Weil A: Dealing with dancing eyes. Optom Vis Sci; 69:447-450, 2013.

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