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Case Report

Surgical Management of Complex Anomalous Head Posture in Idiopathic Infantile Nystagmus

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ABSTRACT

Idiopathic infantile nystagmus (IIN) is usually associated with a null zone, which is the zone of minimal nystagmus intensity. An anomalous head posture (AHP) is adopted to shift the null zone from an eccentric position to primary position. A complex AHP may include head position involvement in different ocular axes-namely face turn, chin elevation or depression and head tilt or a combination of these. Surgically, various procedures have been described for correction of this condition. We evaluated 2 children aged six and ten years, who presented with shaking of eyes along with presence of a complex AHP. First child had 20 degrees right face turn, 20 degrees right head tilt and 10 degrees of chin elevation. She underwent a combination of 2 procedures-augmented Anderson's procedure for correction of right face turn and modification of Kestenbaum procedure for right head tilt along with bilateral IR recession, for correction of chin elevation. Postoperatively, AHP was satisfactorily corrected to 5 degrees face turn and minimal head tilt, and it remained stable for one year. The second child had left face turn 15 degrees, with right head tilt of 20 degrees and chin depression 10 degrees. He underwent a combination of 2 procedures-augmented Anderson's procedure for correction of left face turn and modification of Kestenbaum procedure for right head tilt along with bilateral SR recession, for correction of chin depression. Postoperatively, AHP was satisfactorily corrected to 5 degrees face turn and minimal head tilt, which remained stable over a period of one year. In both cases, AHP was corrected by operating on only 2 muscles in each eye (one horizontal and one vertical) at a time. As both cases presented with combination of both torsional and vertical components of AHP, we decided to treat them both by surgery on a single vertical rectus muscle bilaterally to correct the chin position (elevation or depression), as well as torticollis (transposition of vertical recti, either nasally or temporally as needed). Since a third rectus muscle was not operated upon, there was a lesser possibility of developing anterior segment ischaemia. Additionally, as all components of AHP were corrected in one session, need for a second procedure under general anaesthesia to correct residual AHP was avoided in both cases.

Introduction

Idiopathic infantile nystagmus (IIN) is a disturbance of oculomotor control due to unknown etiology producing horizontal jerky multiplanar involuntary oscillations of eyeballs. It is accompanied by presence of a null zone, which is zone of minimum nystagmus intensity. An anomalous head posture may be adopted, in an attempt to shift null zone from an eccentric position to primary position. Commonest type of AHP described in literature is horizontal face turn to either side, followed by vertical chin up or chin down posture, with head tilt being the least

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common type [1]. These AHPs may exist alone or in combination. Various surgical modalities are described for correction of AHP.

We describe cases of two children with IIN and complex AHP, which were corrected using a combination of augmented Anderson's procedure for face turn and a modification of torsional Kestenbaum procedure, using the transposition of a single rectus muscle for correction of both head tilt and chin position.

Case Reports

Case 1

A 10-year-old female presented with shaking of her eyes since birth. Her parents also complained that she adopted a right head tilt while straining to see. She did not have any other significant ocular or systemic illnesses in the past and her neurological evaluation was normal.

Table 1: Table showing 9 gaze binocular vision of the patient in all gazes, which shows that vision is best in levodepression (null zone).

R		**L**
6/36	6/24	6/24
6/36	6/24p	6/18p
6/36	6/24p	**6/12 p**

Her best corrected vision, measured binocularly (using Snellen's chart), was 6/24, N10 in forced primary gaze; and 6/12, N8 in levodepression. Table 1 shows her binocular vision in all 9 gazes. She had a complex anomalous head posture (AHP) of 20 degrees right face turn, 20 degrees right head tilt and 10 degrees of chin elevation with distance fixation; measured using a goniometer. She had horizontal jerky right beating nystagmus, with decreased amplitude in levodepression (null zone), dampening on convergence and with no latent component. She was diagnosed with idiopathic infantile nystagmus (IIN) and complex AHP. Her anterior and posterior segments were within normal limits, with no evidence of retinal pathology. Preoperatively the child had a normal disc to fovea correlation and no evidence of subjective (double Maddox rod test) or objective (on indirect ophthalmoscopy) torsion. We could not perform electronystagmography.

For correction of AHP, we performed a combination of 2 procedures - augmented Anderson's procedure, which is recession of horizontal yoke muscles considering right eye as dominant eye (RE MR recession 9 mm with LE LR recession 12 mm) for correction of right face turn and 5 mm inferior rectus (IR) recession along with temporal transposition in right eye; and another 5 mm IR recession with nasal transposition in left eye to correct chin elevation and right head tilt.

Surgery Performed: Augmented Anderson procedure

(RE Medial rectus recession 9mm + LE lateral rectus recession 12 mm) — R face turn

+

Torsional Kestenbaum procedure (modification)

(RE- Temporal transposition with 5 mm IR recession + LE Nasal transposition with 5 mm IR recession) — Right head tilt and chin elevation

A schematic representation of the above surgery has been provided, explaining various components of the procedure (Figure 1).

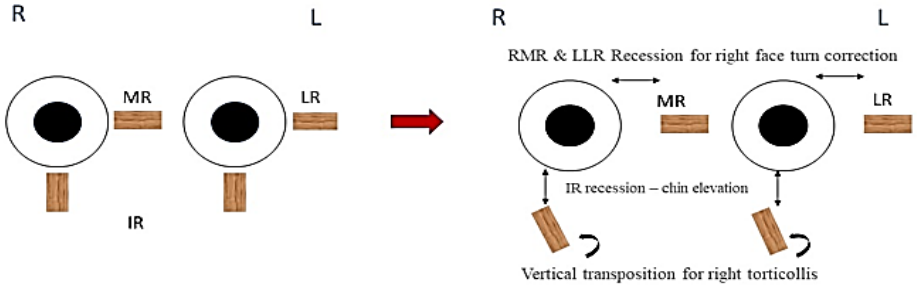

Figure 1: Schematic representation showing Augmented Anderson's procedure involving recession (double sided horizontal arrow) of RMR and LLR for correction of right face turn; along with temporal transposition of right IR and nasal transposition of left IR (curved arrows) for torticollis and IR recession (double sided vertical arrows) for chin elevation.

Figure 2: A) Preoperative photograph showing right face turn and head tilt. **B)** Postoperative photograph showing improvement with very mild AHP.

Postoperatively, AHP was satisfactorily corrected to 5 degrees face turn and minimal head tilt, and she continued to maintain this correction at 1-year post surgery. Figure 2 shows preoperative and postoperative head posture. Parents reported a decrease in amplitude of nystagmus and there

was no documented torsion both on subjective and objective assessments, during the postoperative visits.

Case 2

A 6-year-old male presented with shaking of his eyes since birth. His parents also complained about him adopting a right head tilt and left face turn while straining to see. He did not have any other significant ocular or systemic illnesses in the past and his neurological evaluation was normal.

Table 2: Table showing 9 gaze binocular vision of the patient in all gazes, which shows that vision is best in dextroelevation (null zone).

R		L
6/6p	6/12	6/18
6/12	6/9	6/18
6/12p	6/18	6/24

His best corrected vision, measured binocularly (using Snellen's chart), was 6/9, N8 in forced primary gaze; and 6/6, N8 in dextroelevation. Table 2 shows his binocular vision in all 9 gazes. He had a complex anomalous head posture (AHP) of left face turn 15 degrees, with right head tilt of 20 degrees and chin depression 10 degrees with distance fixation; measured using a goniometer. He had horizontal jerky left beating nystagmus, with decreased amplitude in dextroelevation (null zone), dampening on convergence and no latent component. He was diagnosed with idiopathic infantile nystagmus (IIN) and complex AHP. His anterior and posterior segments were within normal limits, with no evidence of retinal pathology. Preoperatively the child had a normal disc to fovea correlation and no evidence of subjective (double Maddox rod test) or objective (on indirect ophthalmoscopy) torsion. We could not perform electronystagmography.

For correction of AHP, we performed a combination of 2 procedures - augmented Anderson's procedure, which is recession of horizontal yoke muscles considering left eye as the dominant eye (LE MR recession 7 mm with RE LR recession 10 mm) for correction of left face turn and 6 mm superior rectus (SR) recession along with nasal transposition in right eye; and another 6 mm SR recession with temporal transposition in left eye to correct chin depression and right head tilt.

Surgery Performed: Augmented Anderson procedure

LE Medial rectus recession 7mm + RE lateral rectus recession 10 mm) — Face turn

+

Torsional Kestenbaum procedure (modification)

(RE- Nasal transposition with 6 mm SR recession + LE Temporal transposition with 6 mm SR recession) — Right head tilt and chin depression

Postoperatively, AHP was satisfactorily corrected to 5 degrees face turn and minimal head tilt, and he continued to maintain this correction at 1-year post surgery. Figure 3 is a schematic representation of the procedure, explaining its various components. Parents reported a decrease in amplitude of nystagmus and there was no documented torsion both on subjective and objective assessments, during the postoperative visits.

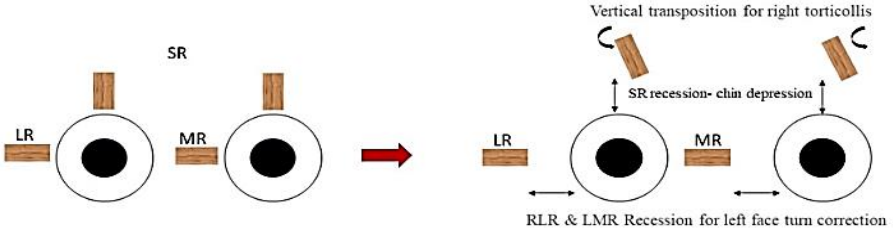

Figure 3: Schematic representation showing Augmented Anderson's procedure involving recession (double sided horizontal arrow) of LMR and RLR for correction of left face turn; along with nasal transposition of right SR and temporal transposition of left SR (curved arrows) for torticollis and SR recession (double sided vertical arrows) for chin depression.

Discussion

The underlying principle involved in surgical correction of anomalous head posture in idiopathic infantile nystagmus is shifting of null zone from an eccentric position to primary gaze. This in turn results in improvement of visual acuity and decreased intensity of nystagmus in the primary gaze. Surgical procedures described for correction of horizontal strabismus include Anderson procedure (involving horizontal yoke muscle recession) with or without augmentation, Goto procedure (horizontal yoke muscle resection), Kestenbaum procedure (recess-resect procedure in both eyes) with or without augmentation [2-4]. For correction of chin position, bilateral vertical recti recession may be performed (bilateral IR recession for chin elevation and bilateral SR recession for chin depression). Roberts *et al.* describe a successful correction of chin down position associated with INS, by performing large SR recessions combined with inferior oblique transposition [5].

Torticollis or head tilt is the least common type of all AHPs. Head tilt to either side is accompanied by ocular rotation in the opposite direction, intorsion of ipsilateral eye and extorsion of contralateral eye. The resultant AHP may be rectified by excyclotorting the ipsilateral eye and incyclotorting the contralateral eye, which may be achieved by operating either on obliques or recti muscles. This was first described by Conrad and de Decker, who operated on all oblique muscles, slanting their insertions in an attempt to produce the above cyclotorsional effect [6]. Spielmann *et al.* described a technique involving slanting of insertion of horizontal recti muscles for the same purpose [7].

Surgeries on obliques involving weakening of SO or strengthening of IO on ipsilateral side and/or strengthening of SO or weakening of IO on the contralateral side have also been attempted by multiple authors. Kraft reported a single case of a 7-year-old boy with congenital nystagmus and

right head tilt who underwent a left IO recession to incycloduct the left eye and a tenotomy of the RSO on the nasal side with insertion of a retinal band to excycloduct the right eye [8]. Similar principles were employed by Prem Prakash *et al.* (IO strengthening by advancement procedure) and Pehere *et al.* (superior oblique anterior tenectomy in the ipsilateral eye, Harada-Ito procedure in the contralateral eye, and inferior oblique recession in the contralateral eye); in their individual case reports dealing with correction of pure torticollis [9, 10].

A Kestenbaum-like surgery has been described, involving transposition of vertical recti muscles in order to effect cyclovertical correction. This would require nasal shifting of superior rectus (SR) and temporal shifting of inferior rectus (IR) on the side ipsilateral to the tilt (producing extorsion); and temporal shifting of SR and nasal shifting of IR on the side contralateral to the tilt (producing intorsion).

Patients may sometimes present with an AHP involving combination of above varieties, necessitating combination of more than one surgical technique to be performed in order to correct the same. Leuder *et al.* reported a case series of 6 patients, most of whom had both horizontal components and a head tilt as a part of the AHP [11]. They underwent a 2 step procedure where the horizontal component of AHP was corrected first followed by anterior 50% tenectomy of the superior oblique tendon on same side and recession of the inferior oblique muscle, on the contralateral side for correction of head tilt. Hertle *et al.* describes another case series of 24 patients, with vertical head postures [12]. Thirteen patients with a chin-down posture underwent a bilateral superior rectus recession, inferior oblique myectomy and a horizontal rectus recession or tenotomy. Eleven with a chin-up posture had a bilateral superior oblique tenectomy, inferior rectus recession and a horizontal rectus recession or tenotomy. In each of the eyes one horizontal and one vertical recti muscle were operated along with one oblique muscle.

In both the cases reported by us, we managed to correct AHP by operating only 2 muscles in each eye (one horizontal and one vertical) at a time. In each of the cases face turn was managed by an Augmented Anderson's procedure, involving large recession of MR ipsilateral to the side of face turn and large recession of LR on the contralateral side. As both cases presented with combination of both torsional and vertical components of AHP, we decided to treat them both by surgery on a single vertical rectus muscle bilaterally to correct the chin position (elevation or depression), as well as torticollis (transposition of the vertical recti, either nasally or temporally as needed). This combination afforded us satisfactory correction of the AHP. No subjective or objective torsion was noted in either of the patients, following surgery.

Conclusion

As the third rectus muscle was not touched, in either of the eyes, there was lesser possibility developing of anterior segment ischaemia. Additionally, since all components of AHP were addressed in one session, need for a second procedure under general anaesthesia to correct residual AHP was also avoided. Hence, we recommend that shifting a single vertical rectus muscle along with recession in each eye would be sufficient to correct head tilts up to 20 degrees, when accompanied by chin elevation or depression.

Institution

Aravind Eye Hospital, Coimbatore, India.

Conflicts of Interest

None.

Funding

None.

Consent

Consent to publish case material and pictures was obtained from the patient's parents.

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