Vertical transposition of the horizontal rectus muscles to correct head tilt because of infantile nystagmus syndrome – A case series

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**Purpose:** Head tilt associated with infantile nystagmus syndrome (INS) can be corrected by (a) operating the oblique muscles, (b) horizontally transposing the vertical rectus muscles, or (c) vertically transposing the horizontal rectus muscles. We report three cases of INS with head tilt corrected by vertically transposing the horizontal rectus muscles in both the eyes. **Methods:** Three cases of head tilt with INS from an institutional practice operated by a single surgeon were retrospectively reviewed and analyzed. The intervention included full tendon width transposition (upward or downward) of all four horizontal rectus muscles to induce cyclotorsion in the direction of head tilt. The primary outcome measure was the correction of head tilt in the primary position. **Results:** Three patients (boys) of ages ranging from 4 to 7 years with a pre-operative head tilt of 30° were operated upon. Although one patient’s oblique muscles had been operated on to correct head tilt, another patient had an unmasked face turn after the surgery, which was corrected with a modified Anderson’s procedure. Post-operatively, all patients had a reduction of head tilt to a range of 0–10°. **Conclusion:** Vertical transposition of horizontal rectus muscles is a simple surgical option to correct head tilt in INS. However, the results may vary based on individual cases.

**Key words:** Head tilt, INS, nystagmus, surgical outcomes

Patients with infantile nystagmus syndrome (INS) adopt various abnormal head postures (AHPs) for better vision, with head tilt being one of them. Although head tilt is usually corrected by operating on oblique or vertical rectus muscles, we report three cases of correction of head tilt in INS by transposition of the horizontal rectus muscles.

We retrospectively reviewed records of patients with head tilt because of INS who underwent vertical transposition of horizontal rectus muscles at our institute. Prior approval was taken from the ethical committee, and the study adhered to the tenets of the Declaration of Helsinki.

Head tilt was measured with the help of a goniometer with one arm kept vertical and the other arm parallel to the sagittal plane of the patient’s head with the hinge kept at the neck. This was measured with the patient reading the finest possible distant optotypes with full refractive correction.

**Case Series**

**Case 1**

A 4-year-old boy [Table 1] whose parents had noticed a right head tilt since the child was 1 year old presented to the clinic. His refractive error and best-corrected visual acuity (BCVA) were -9.25 DS (diopter sphere)/-4.50 DC (diopter cylinder) at 180° and 20/60 in the right eye and -9.25 DS/-2.50 DC at 180° and 20/60 in the left eye. His near vision was N8 in both eyes. He had an esotropia of four prism diopters (PDs) for distance and near. Ocular motility evaluation showed an abduction deficit of -3 and grade 1 retraction with an up-shoot in the left eye on adduction. He also had INS with fine amplitude right beating nystagmus. A right head tilt of 30° [Fig. 1(1A)] was noted. Anterior segment examination was unremarkable in both the eyes. The retina was normal with myopic tessellations, and there was no torsion. An electroretinogram (ERG) revealed reduced amplitudes of both scotopic and photopic waveforms. Magnetic resonance imaging (MRI) was performed and was normal. The child was diagnosed with Duane retraction syndrome type 1 in the left eye, with INS and cone-rod dystrophy in both the eyes. He underwent right lateral rectus (LR) muscle up-shift with medial rectus (MR) muscle down-shift and left MR up-shift with LR down-shift in the right and left eyes, respectively. The full tendon width of muscles was trans-positioned along the spiral of Tillaux. Two months post-operatively, there was a residual right head tilt of 15°, which improved to 3° by the end of 1 year [Fig. 1(1B)]. The patient maintained the corrected head posture in the most recent follow-up visit (at 20 months).

**Case 2**

A 3-year-old boy with left eye dermolipoma since birth presented to our clinic. His parents had noticed a left head tilt for 6 months. The patient had INS with multi-planar...
nystagmus. His refractive error and BCVA were -6.25 DS/-1.00 DC at 180° and 20/200 in the right eye and -6.00 DS/-1.00 DC at 180° and 20/200 in the left eye, respectively. The boy kept his head tilted toward the left at an angle of 30° [Fig. 1(2A)]. He was orthotropic with full ocular movements in both eyes. Slit-lamp examination of the anterior segment was unremarkable in both the eyes. The fundus examination showed a tilted disc with myopic peri-papillary chorioretinal atrophy. Scotopic amplitudes were more reduced than photopic waveforms on

Figure 1: Pre-operative and post-operative head posture of all three cases. Case 1 as seen pre- operatively with the right head tilt of 30° (1A) and small residual tilt after a year of surgery (1B). Case 2 with a persistent left tilt of 25° after oblique muscle surgery (2A) and after 3 months of surgery with no residual AHP (2B). Pre-operative right head tilt of 30° in case 3 (3A) and after 7 months of surgery (3B) with a residual head tilt of 10° and minimal face turn

Table 1: Pre- and post-strabismus surgery: Summary of three cases of infantile nystagmus syndrome

| Case No. | Age at surgery/ Sex | Best Corrected Visual acuity (OD: OS) | Associated findings | Prior surgical procedure | Right tilt pre-operative | Total follow-up in months | Additional procedure after surgery/d for head tilt | Post-operative head tilt | Complications | Residual head tilt | Additional procedure after surgery for tilt |
|----------|---------------------|-------------------------------------|--------------------|--------------------------|------------------------|-------------------------|-----------------------------------------------|------------------------|--------------|-----------------|---------------------------------------------|
| 1        | 4 yr/Male           | 20/60; 20/60                        | High Myopia, Duane retraction syndrome Type 1, Cone-rod dystrophy | None                     | Right tilt -30°         | 20                      | None                                         | Left tilt -25°           | None         | Right MR, Recession 6 mm, Left LR, Recession 6 mm | None                                     |
| 2        | 3 yr/Male           | 20/200; 20/200                      | Dermalpoma, High Myopia, Rod-cone dystrophy | Right inferior oblique resection, left superior oblique tenectomy-corrected 5° tilt from 30° left tilt pre-operatively. | Left tilt -25°          | 13                      | None                                         | Left tilt -25°           | None         | None            | None                                        |
| 3        | 7 yr/Male           | 20/100; 20/80                       | Retinopathy of pre-maturity | Retinal laser procedure in both the eyes for retinopathy of pre-maturity | Right tilt -32°         | 23                      | Right MR, Recession 6 mm, Left LR, Recession 6 mm | Right tilt -32°           | None         | Residual head tilt | Right MR, Recession 6 mm, Left LR, Recession 6 mm | None                                     |

Table 1 notes:
- OD = right eye, OS = left eye
- R= Right, L= Left, LR= Lateral Rectus muscle, MR=Medial Rectus muscle, ↑ = Upshift; ↓ = Downshift
Figure 2: (a) In this scenario, the patient had a right head tilt. (b) Both the eyes have been surgically rotated in the direction of head tilt – to correct right head tilt, the right eye is excyclo-rotated and the left eye is incyclo-rotated. (c) The head tilt was corrected after the induced rotation in both the eyes.

The second effect is a force based on a similar concept, Decker proposed vertical muscle surgery. Based on this, the vertical rectus muscle recession in the right eye and anterior tenectomy of the left superior oblique muscle. A head tilt of 25° (left-sided) persisted for 6 months post-operatively. A right LR down-shift with MR up-shift and a left MR down-shift with LR up-shift was performed to correct the same. Three months post-operatively, he had no AHP [Fig. 1(2B)] and his visual acuity improved to 20/160 in the right eye and 20/125 in the left eye. This is maintained at 13 months of follow-up.

Case 3
A 7-year-old boy presented with a history of retinopathy of pre-maturity in both eyes, for which he was lasered in his infancy. His refractive error and BCVA were -1.25 DS/-2.00 DC at 180° and 20/100 in the right eye and -5.50 DS/-2.50 DC at 180° and 20/80 in the left eye, respectively. He had INS with small-amplitude high-frequency jerky right beating nystagmus and a right head tilt of 30° [Fig. 1(3A)] in addition to an exotropia of 8 PD for distance and near. Slit-lamp examination of the anterior segment was unremarkable. The fundus examination showed a normal disc with the lasered peripheral retina. An LR up-shift and MR down-shift were performed in the right eye, and MR up-shift and LR down-shift were performed in the left eye. Seven months post-operatively, the child had a residual head tilt of 10° [Fig. 1(3B)] with a persistent right face turn of 20°. Subsequently, a modified Anderson’s procedure was performed for the face turn, which corrected it and is maintained in the last follow-up, at 23 months.

Discussion
Head tilt associated with INS is corrected by surgically inducing a torsion in the individual eye similar to the globe position in the preferred AHP. Thus, to correct the right head tilt, the right eye is excyclo-rotated and the left eye is incyclo-rotated by extra-ocular muscle surgery [Fig. 2].

In 1988, de Decker and Conrad published the results of 98 patients operated on all four oblique muscles (recession and resections) for correction of head tilt.[9] Leuder and Galli performed anterior tenectomy of the superior oblique muscle with a contra-lateral recession of the inferior oblique muscle to correct head tilt in six patients with good success.[10]

The superior/inferior oblique muscles can be selectively weakened/strengthened to decrease/exaggerate the effect of incyclo-/excyclo-torsion. For example, to correct the right head tilt, weakening of the right superior oblique muscle and weakening of the left inferior oblique muscle can be performed [Fig. 3a].

Another method is the horizontal transposition of the vertical rectus muscle as performed by Von Noorden and colleagues.[11] In the above example, to excyclo-rotate the right eye, nasal transposition of the superior rectus muscle and temporal transposition of the inferior rectus muscle can be performed in the right eye and vice versa for the left eye [Fig. 3b].

Spielmann demonstrated a similar globe torsion with the help of a slanting recession of all eight rectus muscles in both the eyes.[12] Based on a similar concept, de Decker proposed vertical transposition of the horizontal rectus muscles.[3] Excyclo-torsion can be induced in the right eye by superior transposition of the lateral rectus muscle and inferior transposition of the medial rectus muscle. Similarly, superior transposition of the left medial rectus muscle and inferior transposition of the left lateral rectus muscle will induce incyclo-torsion in the left eye [Fig. 3c]. These procedures performed on both the eyes can correct the right head tilt.

The transposition of the horizontal rectus muscle produces three effects. First, it decreases the action of the muscle when the eye is turned in the direction of the muscle shift. For example, when the lateral rectus is supra-positioned, the abduction is weaker when the eye turns upward. This effect is beneficial in treating A-V pattern strabismus.[10] The second effect is a force vector that is created in the direction of the muscle placement. A new upward force vector is created when the lateral rectus muscle is shifted superiorly. This corrects small monocular hypotropia/hyperptropia associated without oblique muscle
overaction. The final effect is inducing a cyclo-torsion of the globe in the direction from where the muscle was transposed. This effect is the rationale behind shifting of the rectus muscle to treat cycloptopia. While performing simultaneous horizontal rectus muscle transposition in the opposite direction, these first and second effects cancel out each other, whereas a torsional component is added. Sharma and his colleagues studied this torsional effect of monocular vertical shifting of horizontal muscles to treat A-V pattern strabismus.

The primary advantage of the procedure we performed is that horizontal rectus muscles can be approached easily as compared to vertical and oblique muscles, thereby making the surgery less challenging. Induced vertical deviations as seen with horizontal transposition of vertical muscles are not present. Additionally, associated face turn can be managed during the surgery. This could have been an option for case 3 of our series to tackle the head tilt and face turn simultaneously. In older patients, it is better to transpose previously operated horizontal muscles to prevent the risk of anterior segment ischemia. Additionally, the intensity of the nystagmus could decrease because of the tenotomy and re-attachment effect of horizontal rectus muscles. However, the disadvantage is having an unpredictable dose–response curve as there is a difference in the length of the lateral and medial rectus muscles.

Potential complications of this procedure include new vertical or horizontal deviations, tilting of the subjective visual environment, torsional diplopia, and anterior segment ischemia. There were no new deviations noted in our series, and none of the patients complained about torsional diplopia. These findings are similar to the series published by de Castro-Abegar A and colleagues where horizontal transposition of vertical rectus muscles was performed to correct head tilt.

There are a few limitations to the present study. Being retrospective and having a small sample size, in this series, documentation of the nystagmus waveforms was not performed and diagnosis of INS was made clinically. All three cases were myopic, and objective documentation of torsion was not performed.

**Conclusion**

In conclusion, vertical transposition of the horizontal rectus muscle is an unexplored option to correct head tilt in INS. However, further studies have to be performed to establish a dose–response curve.

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**Conflicts of interest**

There are no conflicts of interest.

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