Horizontal Transposition of the Vertical Rectus Muscles to Correct a Head Tilt in 5 Patients With Idiopathic Nystagmus Syndrome

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• PURPOSE: Patients with idiopathic nystagmus syndrome often develop an abnormal head position. A horizontal face turn can be treated with the augmented Kestenbaum procedure, while patients with a chin up or chin down position can be treated with surgery on the vertical recti and/or oblique muscles. Although rare, some patients may have a head tilt with no face turn. We report 5 patients who underwent horizontal transposition of the vertical rectus muscles to correct a head tilt.

• DESIGN: Retrospective case series.

• METHODS: Five cases from 2 different tertiary referral eye centers and 3 different surgeons were reviewed and analyzed. Surgery for the patients consisted of either a 7-mm or full tendon-width transposition of the vertical rectus muscles of each eye to induce cyclotorsion in the direction of the head tilt. The presenting clinical histories, measurements, and surgical outcomes were reviewed. The primary outcome measure was correction of head tilt.

• RESULTS: Of 5 patients, 2 had previous horizontal face turns that were corrected with an augmented Kestenbaum procedure and later developed a head tilt, while 3 patients presented primarily with a head tilt. Age at surgery ranged from 5 to 8 years. Initial tilts were measured between 20-45°. Surgery was performed bilaterally except in 1 patient with history of morning glory disc anomaly and therefore transposition surgery was only performed on fixating eye for that patient. Postoperatively, 3 of 5 patients had near complete (0-5°) resolution of their tilt. One patient had a persistent 5-degree head tilt and a small chin up that was nullified with vertical prisms in spectacles.

• CONCLUSION: Transposition of the vertical rectus to induce cyclotorsion in the direction of the head tilt improves abnormal head titling in patients who have idiopathic nystagmus syndrome. This procedure was safely performed in patients with previous augmented Kestenbaum procedures with no incidence of anterior segment ischemia in our cases. (Am J Ophthalmol 2020;217:68–73. © 2020 Elsevier Inc. All rights reserved.)

PATIENTS WITH IDIOPATHIC NYSTAGMUS SYNDROME often have minimal nystagmus in a null point away from primary position. As a result, they often develop a corresponding abnormal head position to best perceive the world with the least amount of image distortion because of decreased frequency of the nystagmus while assuming the null gaze position. Surgery to correct abnormal head posturing caused by nystagmus follows the general principle of moving the eyes in the direction of the abnormal head position as first described by Anderson[1] and Kestenbaum.[2,3] Von Noorden and Chu[4] first reported the transposition of the vertical rectus muscles to produce a torsional rotation in 2 patients that successfully treated an abnormal head tilt in the setting of cyclovertical strabismus. In our report, we discuss 5 cases that had abnormal head tilt because of null point of nystagmus who were successfully treated with horizontal transposition of the vertical rectus muscles.

METHODS

THIS RETROSPECTIVE CASE REVIEW WAS APPROVED BY THE local institutional review boards of both institutions involved in the study and complied with the U.S. Health Insurance Portability and Accountability Act of 1996. The records of patients who underwent horizontal transposition of the vertical rectus muscles to treat a head tilt that damped nystagmus were reviewed. The preoperative assessment included surgical history, visual acuity, and preoperative head tilt. All patients underwent full tendon-width transpositions of both vertical rectus muscles to rotate the eyes in the direction of the head tilt. For example, a superior rectus nasal transposition placed the temporal edge of the muscle at the prior nasal insertion and the nasal edge approximately 7 mm nasal to that point, which would have the effect of inducing a force vector for extorting the eye. (Figure 1). The postoperative assessment included assessment of vision and head tilt.

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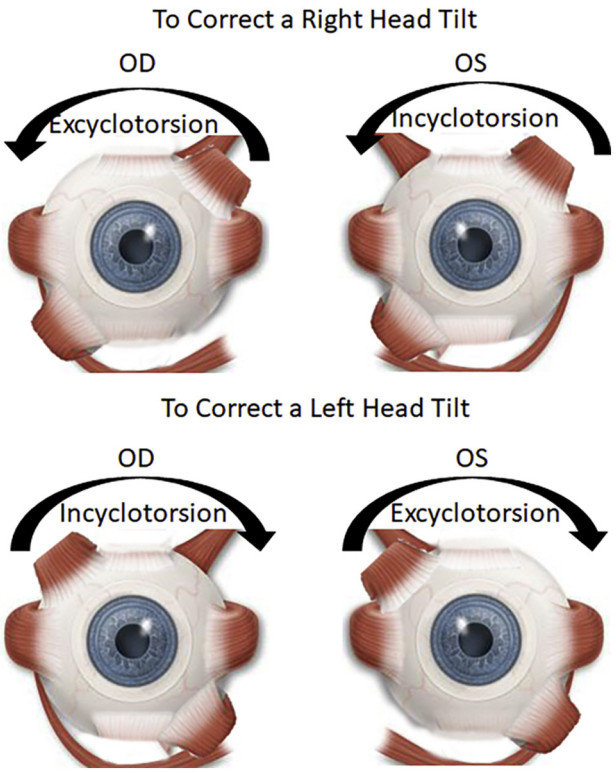

FIGURE 1. To correct a right head tilt, nasal transposition of the right superior rectus and left inferior rectus and/or temporal transposition of the right inferior rectus and the left superior rectus leads to excyclotorsion of the right eye and incyclotorsion of the left eye, respectively. To correct a left head tilt, nasal transposition of the right inferior rectus and left superior rectus and/or temporal transposition of the right superior rectus and left inferior rectus leads to incyclotorsion of the right eye and excyclotorsion of the left eye, respectively.

• **CASE 1:** A 6-year-old girl was born with a morning glory disc anomaly of the left optic nerve. She had an anomalous head posture since infancy. When first examined by one of the authors (B.J.K.), she was noted to have a right head tilt of 30° and a face turn to the left of 5° as measured with a goniometer. She had a manifest horizontal jerk nystagmus that damped in her preferred head position. Her cycloplegic refraction and best corrected visual acuity was OD plano +1.75 × 90, 20/25 and OD −4.75 +2.00 × 75, 5/200. She had a relative afferent pupil defect OS. Other than the left optic nerve anomaly, there were no other structural abnormalities noted. Her OD had no fundus torsion, and it was difficult to assess torsion in her OS because of the optic nerve anomaly. A 35Δ left esotropia was present. The patient underwent surgery in the form of a full tendon-width nasal transposition of the right superior rectus and a full tendon-width temporal transposition of the right inferior rectus to address the head tilt. That surgery was combined with a 5-mm recession of the left medial rectus and a 7-mm resection of the left lateral rectus to address the esotropia. After surgery, her head posture was improved, varying between 0 and 5 degrees of right tilt and 0 to 5 degrees of left face turn. Her left fundus now showed +2 intorsion. She remained stable for 2 years after which she was lost to follow-up.

• **CASE 2:** As an infant, this 8-year-old boy was diagnosed with idiopathic nystagmus. At 5 years of age he underwent a modified Kestenbaum-Anderson (K-A) procedure by another ophthalmologist to correct a right face turn, which consisted of a 7-mm recession of the right medial rectus, an 11-mm resection of the right lateral rectus, an 8.5-mm resection of the left medial rectus, and a 9.5-mm resection of the left lateral rectus. This initially improved his head turn but he gradually began developing an increasing left head tilt. When seen at 8 years of age by one of the authors (B.J.K.) he had a cycloplegic retinoscopy and best corrected visual acuity of OD −2.00 +2.50 × 90, 20/30 and OS −2.25 + 2.75 × 90, 20/40. He had 200 arc-seconds of stereopsis. His pupils were normal, and he did not have a paradoxical response to darkness. His eyes were structurally healthy, and he showed no fundus torsion. He had a preferred head posture of 25° left head tilt, as measured with the cervical range of motion device, which damped his horizontal jerk nystagmus. He underwent surgery for the head tilt that consisted of full tendon-width transpositions of all the vertical rectus muscles with ciliary artery sparing as summarized in Table (temporally for the right superior rectus and left inferior rectus and nasally for the right inferior rectus and left superior rectus). After surgery, his head tilt was eliminated, and he showed +2 to +3 fundus intorsion OD and +2 to +3 fundus extorsion OS. He remained stable for over 4 years after which he was lost to follow-up.

• **CASE 3:** This girl was first examined by one of the authors (B.J.K.) at 7 years of age. She had been diagnosed as having idiopathic nystagmus syndrome at 1 year of age by another ophthalmologist. She was found to have what appeared to be a rotary and horizontal jerk nystagmus that damped with a head tilt to the left of 30° and a small variable right face turn of between 0 and 10° as measured with a cervical range of motion device. There was no manifest strabismus detected. She had a cycloplegic retinoscopy and best corrected visual acuity of OD +0.50 + 0.75 × 60, 20/40 and OS +0.75 + 0.75 × 130, 20/40 with 400 arc-seconds of stereopsis. Her pupil responses were normal and her eyes were structurally healthy. She had no fundus torsion but was just on the normal side of the range for extorsion in both eyes. She underwent surgery to correct the left head tilt consisting of full tendon-width transpositions of all the vertical rectus muscles as summarized in Table (temporally for the right superior rectus and left inferior rectus and nasally for the right inferior rectus and left superior rectus). When last seen 3 years after surgery she had a negligible left head tilt. The right fundus showed a trace of intorsion and the left fundus showed +3 extorsion. The

TABLE. Summary of Results of this Case Series

Case No.	Surgeon	Age at Surgery (Years)	Tilt (Direction Degrees)	Preoperative Vision (OD; OS)	Other Eye Muscle Surgery	Tilt Surgery Summary	Postoperative Tilt (Direction Degrees)	Other Information
1	B.J.K.	6	Right 30	20/25; 20/200		RSR nasal + RIR temporal	Right 0-5	Morning glory disc OS and presented with a concomitant 35 PD left ET and underwent LMRc 5, LLRs 7 at time of transposition. Right fundus demonstrated intorsion post–tilt procedure
2	B.J.K.	8	Left 25	20/30; 20/40	Modified Kestenbaum-Anderson: RMRc 7, RLRs 11, LMRs 8.5, LLRc 9.5	RSR temporal + RIR nasal AND LIR temporal + LSR nasal, with ciliary artery sparing	None	Congenital motor nystagmus right face turn
3	B.J.K.	7	Left 30	20/40; 20/40		RSR temporal + RIR nasal AND LSR nasal + LIR temporal with ciliary artery sparing technique	None	Congenital motor nystagmus with rotary and horizontal jerk nystagmus
4	N.M.B.	5	Left 20	20/30; 20/30		Surgery 1: RSR temporal + RIR nasal AND LSR nasal + LIR temporal Surgery 2: 8-mm resection and replacement of RSR temporal + replacement of LSR nasal	Left 5	Congenital motor nystagmus with ocular torticollis. Second surgery required because of mild chin up position and some mild residual tilt. Second surgery found that there was 2-3 mm of stretch scar over superior rectus, they were replaced the 1 tendon width transposition as originally intended
5	S.P.D.	7	Right 45	20/30; 20/30	Modified Kestenbaum-Anderson: LLRc10, LMRs 8.5, RMRc7, RLRs 10 at age 2	RSR nasal +RIR temporal AND LSR temporal +LIR nasal	Right 5-10	History of prematurity with infantile nystagmus syndrome

ET = esotropia; LIR = left inferior rectus; LLR = left lateral rectus; LMR = left medial rectus; LSR = left superior rectus; OS = left eye; PD = prism diopters; RIR = right inferior rectus; RLR = right lateral rectus; RMR = right medial rectus; RSR = right superior rectus.

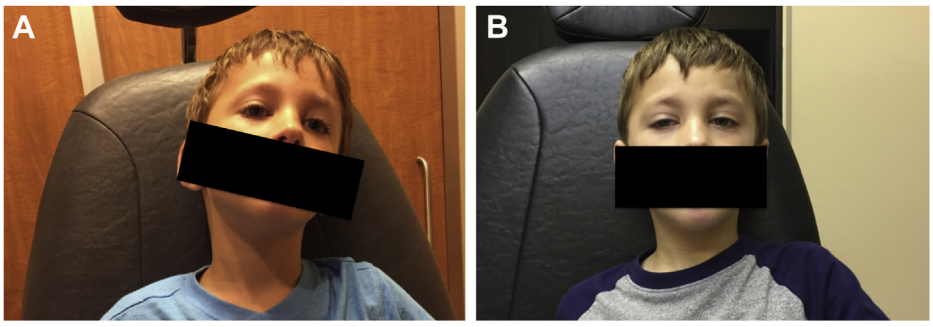

FIGURE 2. (A) Preoperative photograph demonstrating torticollis and left head tilt. (B) Postoperative photograph with small residual tilt.

face turn to the right was usually not noticeable but with intense visual effort was sometimes present at about 5° as determined from photographs provided by the parents.

• **CASE 4:** This 5-year-old boy presented to one of the authors (N.M.B.) with idiopathic nystagmus and ocular torticollis with a null point during left head tilt ranging from 20-40° and no manifest strabismus. Cycloplegic retinoscopy and best corrected visual acuity was OD +1.25 20/30 +2 and OS +1.25, 20/30+3 with fusion on Worth-4-dot testing but no appreciable stereopsis. His pupil responses were normal, and the eyes were otherwise structurally normal without any fundus torsion. He underwent initial surgery to correct the ocular torticollis with full tendon width transpositions of the vertical rectus muscles (Table). He had initial good head position from the surgery; however, about 6 months postoperatively, he was noted to have a chin up position with a small residual left tilt as well. The plan was to correct for the chin up posture with bilateral superior rectus resections. At the time of surgery, the superior rectus muscles were found near the original point of insertion. Consequently, he underwent an 8-mm resection and re-establishment of the full tendon-width transpositions as intended in the first surgery, temporal in the right eye and nasal in the left eye. At his initial postoperative appointment 1 week after surgery he had an improved, but small chin up position and essentially 0 degrees of head tilt (Figure 2). The patient then moved out of state and was lost to follow-up.

• **CASE 5:** This 6-year-old boy presented to one of the authors (S.P.D.) after a modified K-A procedure as a 2-year-old (left lateral rectus recession 10 mm, left medial rectus resection 8.5 mm, right medial rectus recession 7 mm, right lateral rectus resection 10 mm) for a right face turn. He continued to have a horizontal jerk nystagmus which damped with a right head tilt of approximately 50° (subjectively determined by the surgeon). Cycloplegic retinoscopy and best corrected visual acuity were OD +2.00 + 0.50 × 090, 20/30+2 and OS +2.00 + 0.50 × 090, 20/40+2. He had 400 arc-seconds of stereoacuity. His pupil responses were normal, and his eyes were otherwise structurally normal without appreciable torsion on fundus examination. He underwent transposition of the vertical rectus muscles in the direction of the head tilt as summarized in Table. After surgery, he did well and had a small residual head tilt to the right of 5-10°. He has been followed for >1 year and has remained stable with a small residual head tilt.

RESULTS

THIS REVIEW IDENTIFIED 5 PATIENTS WITH INFANTILE nystagmus syndrome who underwent transposition of the vertical rectus muscles to rotate the eye(s) in the direction of a head tilt to correct the tilt. Of note, the patients did not have other obvious reason for the adoption of a tilted head position such as ocular tilt reaction caused by brainstem pathology; there was no history of demyelination episodes or other signs or symptoms of brain stem pathology. In all patients the nystagmus damped in the preferred head posture. None of the 5 patients had fundus torsion before surgery, and all had fundus torsion after surgery in the direction one would expect after the vertical rectus muscle transpositions. The surgery was done bilaterally in all cases except the case with morning glory disc anomaly, in whom it was done in the fixating eye only. The surgery improved the head tilt in all 5 patients, although 2 had a small residual head tilt. There were no changes in postoperative visual acuity and no complications. Patient 4 required a second procedure with the intention of correcting a secondary chin up position and the previously transposed superior rectus muscles were found to have migrated back to their original position, which may explain the recurrence of the initial head tilting.

DISCUSSION

IN 1982, CONRAD AND DE DECKER REPORTED 66 PATIENTS with head tilts caused by nystagmus that they treated successfully with oblique muscle surgery to tort the eyes in the direction of the head tilt.[5] Since then, there have

been other reports of doing oblique muscle surgery in this setting to correct a head tilt, but they are usually limited to 1 or 2 cases of each,[6–9] until 2012 when Lueder and Galli[10] reported a series of 6 patients. However, operating on the oblique muscles as described in previous reports had induced unintended vertical deviations after surgery and were sometimes prone to other complications. Despite the difficulty and possible complications of operating on the obliques, if concurrent surgery was planned on horizontal rectus muscles, then oblique muscle transpositions might theoretically reduce the risk of anterior segment ischemia.

In 1987, Spielmann[11] reported that rotating the eyes in the direction of a head tilt could also be accomplished by operating on the rectus muscles. The technique as described induced a torsional effect on the globe by slanting the insertion of all 4 rectus muscles.[11] A few years later, von Noorden and Chu[4] reported that horizontal transposition of the vertical rectus muscles can rotate the eyes around the sagittal axis to correct a torsional problem and then later applied the same principle to successfully treat 4 of 5 patients with head tilts associated with nystagmus.[12] Our case series confirm the efficacy of horizontal transposition of the vertical rectus muscles suggested by von Noorden, which as a procedure is prone to fewer complications than oblique muscle surgery as proposed by others. Further, for reasons outlined below, we feel that vertical rectus muscle surgery to address a head tilt can possibly be limited to only the fixating eye, and surgery to address any concomitant horizontal strabismus could be done in the fellow eye. In the case of a concomitant nystagmus-driven face turn, chin up or chin down posture, surgery to correct those would also need to be done on the fixating eye.

The mechanism by which rotating the eyes in the direction of a nystagmus null point mediated torticollis is not completely clear. The principles underlying the K-A procedure[1–3] for a face turn are similar in that the eyes are rotated in the direction of the abnormal head posture, but there is a difference in that K-A procedure the fovea is rotated away from the primary position; therefore, in order to foveate on an object, the patient must apply the same innervation to muscles to place the eyes in a primary position that is closer to the innervation needed in their preoperative null point. However, when rotating the eyes along the sagittal axis, the fovea is not rotated away and therefore there is no compensatory rotation of the eyes to correct for a surgically induced disparity. In our series, none of our patients had fundus torsion before the operation. Consequently, we feel that the creation of a torsional rotation to correct a head tilt does not work by correcting an underlying torsional problem. Therefore, the mechanisms of success must be different between the 2 procedures.

In our series, 4 of 5 patients had bilateral surgery. Postoperatively it was noted that the torsional surgery induced fundus incyclotorsion of one eye and excyclotorsion of the other. Therefore, after successful surgery, there was typically more of a torsional misalignment or disconjugacy between the 2 eyes, yet our patients were not symptomatic in this regard. Conrad and de Decker, according to von Noorden and associates,[12] suggested that the mechanism of surgical correction for a head tilt by surgically torting the eyes, worked by correcting a tilting of the subjective visual environment. We do not believe this to be the case as none of our patients reported a titling of the subjective visual environment either before or after surgery. Even patients with symptomatic torsion typically do not describe a tilting of the subjective environment under monocular conditions of the affected eye as is seen in unilateral superior oblique palsy with torsion when the contralateral eye is occluded. Therefore, the sagittal rotation of the eyes must somehow alter the feedback mechanism between the eyes and proprioceptive receptors in the brainstem and vestibular apparatus.

Just as a face turn with a K-A procedure performed on the fixating eye alone can be successful in correcting an abnormal head posture, we feel that this torsional surgery can probably be performed unilaterally on the fixating eye alone with success, as was done with our patient with morning glory disc anomaly. For our other patients, there was an induced torsional disconjugacy by operating on both eyes, with the result of 1 eye being intorted and the other eye extorted, but our patient population did not appear to have torsional symptoms as a result. Therefore, at a minimum, surgery ought to be performed on the fixating eye.

In summary, torticollis associated with infantile nystagmus syndrome can be corrected with the horizontal transposition of the vertical recuts muscles along the spiral of Tillaux. Moreover, this surgery can be done for an adopted head tilt that occurs following a previous standard or augmented K-A procedures that were done to correct an abnormal face turn with a low risk of anterior segment ischemia. If there is coexisting strabismus in the setting of an adopted head tilt, the transposition at least must be performed on the fixating eye, while the strabismus can be corrected by operating on the fellow eye. Our report expands the use of transposition surgery to correct an abnormal head tilt associated with infantile nystagmus syndrome.

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