Comparison of Techniques for Correction of Chin-down Vertical Abnormal Head Position Associated with Infantile Nystagmus Syndrome

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• PURPOSE: **We evaluated the relative effectiveness of combined recession-resection of vertical rectus muscles versus superior rectus recession with inferior oblique weakening for patients who underwent surgical correction of chin-down abnormal head position (AHP) associated with infantile nystagmus syndrome (INS).**

• DESIGN: **Retrospective interventional case series.**

• METHODS: **This is a review of 22 patients who underwent surgical correction of chin-down vertical AHP associated with INS at an academic institution. The primary outcome was collapse of AHP. Unfavorable outcomes included repeat surgery and induced strabismus, in addition to failure of collapse of AHP.**

• RESULTS: **Twenty-two patients had chin-down AHP. Recession-resection (bilateral superior rectus recession 6-9 mm; bilateral inferior rectus resection 5-9 mm) was performed in 11 cases; weakening of both elevators (bilateral superior rectus recession 5-8 mm, bilateral inferior oblique recession or myectomy) occurred in 11 cases. Unfavorable outcome rates were 64% (7/11) compared with 18% (2/11), respectively ($P = .03$). Reoperation was performed for 6 of 22 patients. Five patients were from the recession-resection group, namely 3 for induced V-pattern esotropia, 1 for alternating esotropia, and 1 to correct recurrent AHP. The last of the 6 who required reoperation was in the elevator weakening group, and required correction of a recurrent AHP ($P = .06$).**

• CONCLUSIONS: **While recession-resection of the vertical recti and weakening of both elevators each produce acceptable collapse of chin-down AHP, the former frequently induces a V-pattern esotropia requiring reoperation. (Am J Ophthalmol 2020;213:57–61. © 2020 Elsevier Inc. All rights reserved.)**

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PATIENTS WITH INFANTILE NYSTAGMUS SYNDROME (INS) often adopt an abnormal head position (AHP) to place their eyes in a null position. While most of these AHPs occur in the horizontal plane, they may infrequently occur in the vertical or torsional planes, or in a combination of these positions.[1,2] Regardless of the surgical technique used, the primary purpose for eye muscle surgery in INS with an AHP is to shift the null position of the nystagmus into primary gaze, thus correcting the AHP.[3] The result is an improvement in cosmesis and in visual function, as well as avoidance of such orthopedic issues that arise from muscle contractures in the setting of a long-standing AHP.[4]

In 1953, Kestenbaum[5] described a procedure that entailed weakening of a horizontal rectus muscle (recession) and strengthening of its antagonist (resection) to correct an AHP in the horizontal plane. Though a number of modifications has been proposed,[2,6–10] a large recession-resection procedure applied to the 4 horizontal rectus muscles remains widely accepted as the surgical treatment of choice for patients who have a horizontal plane AHP in the setting of INS.

Some patients with INS have an AHP in the vertical plane—either a chin-down AHP with a null in upgaze or a chin-up AHP with a null in downgaze. For these patients, the surgical technique is less well established. A recession-resection technique on the vertical rectus muscles was shown by Yang and associates[11] and others[6,12] to be superior to recession applied to the vertical rectus muscles alone. Other approaches, such as superior rectus recession with inferior oblique weakening for a chin-down AHP (weakening of both elevators) has also been proposed.[13] However, these reports had relatively few subjects. We reviewed our experience with INS patients requiring surgery for a chin-down vertical AHP (vertical recession-resection or weakening of both elevators) with the aim of exploring efficacy, durability, and limitations of these 2 surgical techniques in correcting a vertical head posture.

METHODS

A RETROSPECTIVE REVIEW OF 150 PATIENTS WITH nystagmus who underwent eye muscle surgery (EMS) for an AHP at our academic center from 1995-2018 was

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conducted. Of these, EMS was performed for a vertical plane AHP (chin-up or chin-down) in 31 patients. Twenty-two patients fulfilled the inclusion criteria for our retrospective observational case series; namely, that they presented with a chin-down AHP that was associated with INS, that they were <18 years of age at the time of surgery, and that they had ≥2 months of follow-up care after their initial surgery. This included 2 patients who had previous EMS for strabismus. Six patients who received surgery for a chin-up AHP were excluded and are reported elsewhere.

Our study was prospectively approved by the Vanderbilt University Institutional Review Board prospectively and determined to qualify for exempt status. This study is compliant with the Health Insurance Portability and Accountability Act and adhered to the tenets of the Declaration of Helsinki as amended in 2008.

• **CLINICAL EVALUATION:** The primary measure of interest was the degree of AHP in the vertical plane and was typically approximated by observation as being either mild (1-15°), moderate (16-30°), or severe (≥31°). Additional data collected included best-corrected visual acuity, ocular alignment, and ductions. When possible, visual acuity was tested in each eye separately, or, when not, in both eyes simultaneously. Ductions were graded on a scale of +4 to −4. These variables were measured preoperatively and at postoperative visits. Postoperative data were collected from visits at 1 week, 2 months (±6 months), 1 year (±6 months), and last available follow-up. Unfavorable outcomes included repeat surgery, induced strabismus, or lack of collapse of AHP.

• **SURGICAL TECHNIQUE:** Initial surgery for the correction of the AHP, as well any subsequent EMS, were performed by one of the senior authors (SPD or DGM). Combined recession-resection was performed on all patients who presented with a chin-down vertical AHP before August 2011. This procedure involved recession (typically of 8-9 mm) in conjunction with resection of the antagonist (typically of 7-8 mm). Dissection was carried down to bare sclera posteriorly to where the vertical rectus muscles penetrate through Tenon's capsule. After disinsertion, the superior rectus muscle was typically recessed via a modified hang-back technique, securing the muscle pole with a small scleral bite at the recession location and a second scleral pass at the original muscle insertion where the suture was tied. The inferior rectus muscle was resected and sewn back to its original point of insertion. Care was taken to ensure that superior and inferior lid retractors were dissected from their respective rectus muscles at the time of surgery to minimize impact on eyelid position at the time of surgery.

When inferior oblique weakening was performed in conjunction with superior rectus recession (typically of 8-9 mm)—as for patients presenting after August 2011 with a chin-down AHP—recession of the superior rectus was conducted in a similar fashion as previously described. Inferior oblique myectomy was performed after disinsertion of the muscle from the globe. A large section of the muscle was clamped as the muscle emerged from Tenon's capsule. The muscle was then cut just distal to the clamp, cauterized, and allowed to retract backwards into Tenon's capsule. In cases where the inferior oblique was recessed, a standard recession of 10 mm was performed.

The amount of surgery performed on either eye was typically symmetric, depending upon the size of the eye, the length of the muscle available for resection, and the size of the orbit, to achieve a maximal effect. We did not recommend a physical therapy regimen for torticollis after eye muscle surgery.

• **ANALYSIS:** The Fisher exact test was applied to compare 2 groups of patients receiving surgery for correction of a chin-down AHP—those receiving recession-resection of the vertical recti, and those receiving weakening of both elevators—with regard to rates of unfavorable outcomes, as well as rates of reoperation. $P = .05$ was considered to represent a statistical difference between the 2 groups in either measure. Correction for multiple comparisons was not applied.

RESULTS

CLINICAL CHARACTERISTICS OF THE 22 PATIENTS WHO presented to us with chin-down AHP in the context of INS are summarized in Supplemental Table 1. The nystagmus had a predominant horizontal component in 17 patients, while a vertical (downbeat) nystagmus was observed in 4 patients. A description of the nystagmus was not available for 1 patient. The degree of preoperative AHP ranged from mild (3 patients) to severe (19 patients; moderate in 4 patients; not quantified for 1 patient). All 22 patients initially had a null in upgaze in conjunction with their chin-down AHP.

• **RECESSION-RESECTION OF THE VERTICAL RECTI:** Recession-resection of vertical recti [bilateral superior rectus recession (BSRc) 6-9 mm; bilateral inferior rectus resection (BIRs) 5-9 mm] was performed in 11 cases. The primary outcome (complete collapse of the AHP) was achieved in 7 of 11 patients, with mild chin-down AHP persisting in 3 patients (patients 2, 4, and 10) and severe residual chin-down AHP still present in 1 patient (patient 7; Figure).

Seven of 11 patients on whom recession-resection was performed had unfavorable outcomes. Of these, 5 patients developed esotropia. A V-pattern esotropia arose in 3 cases (patients 3, 5, and 10), while a nonaccommodative esotropia arose in 2 cases (patients 6 and 8). As shown in the Table, reoperation was performed to correct the strabismus in all 3 cases where V-pattern strabismus developed, and in 1 of the 2 cases where an alternating esotropia developed (patient 6). In the remaining 2 patients (4 and 7), there was

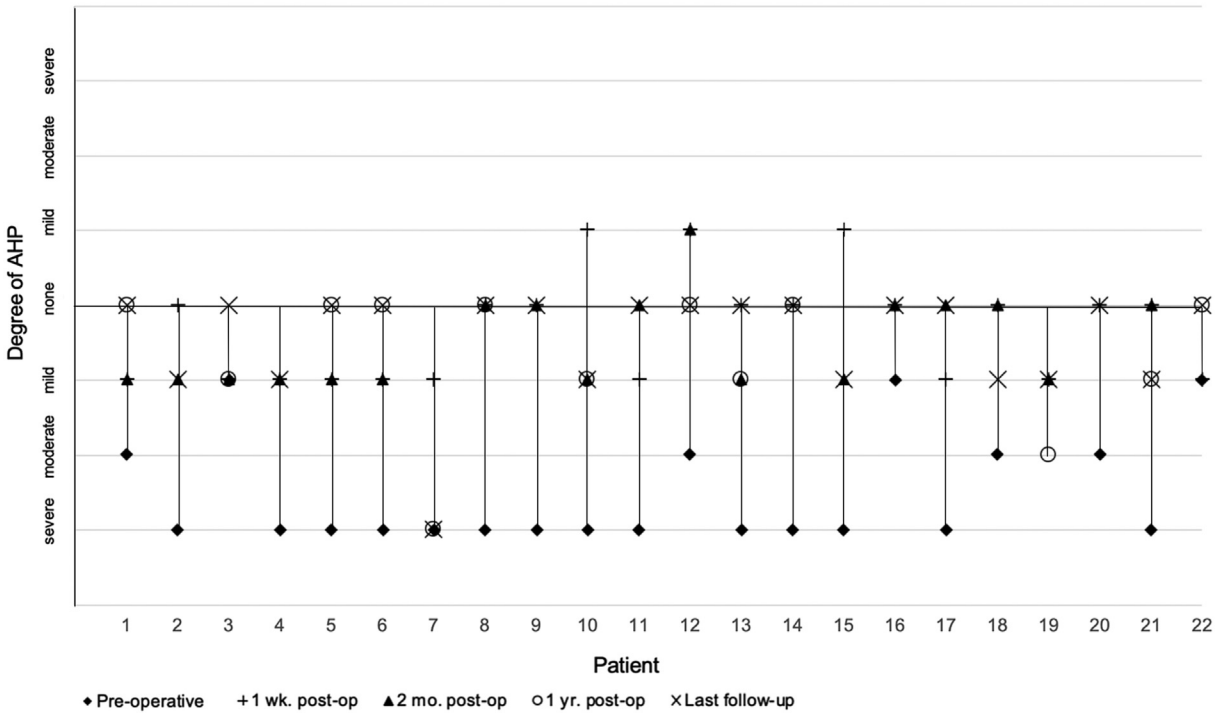

FIGURE. Degree of abnormal head position over time. Degree of vertical head position at preoperative and postoperative clinic visits—at 1 week, 2 months, 1 year, and at last follow-up—displayed for each patient who presented with a chin-down abnormal head position. A chin-down head position extends below the horizontal axis, while a chin-up head position extends above it.

TABLE. Descriptions of Unfavorable Outcomes and Additional Surgeries Performed for Select Patients

Patient No.	Unfavorable Outcome	Additional Surgery
3	V-pattern ET (14 mos)	BMRc-4 mm with inferoplacement
4	Recurrent chin-down AHP (11 mos)	BIRs-6 mm
5	V-pattern ET (1 wk)	BMRc-4 mm; BIOc
6	Alternating ET (11 mo)	BMRc-4.5 mm
7	Recurrent chin-down AHP (15 mos)	—
8	Alternating ET (2 mos)	—
10	V-pattern ET (57 mos)	BMRc-5 mm; BIO myectomy
14	Recurrent chin-down AHP (30 mos)	BSRc to 20 from limbus; BIRs-8
19	A-pattern LXT (24 mos)	—

AHP = abnormal head position; BIR = bilateral inferior rectus resection; BMRc = bilateral medial rectus recession; BSRc = bilateral superior rectus recession; BIOc = bilateral inferior oblique recession; ET = esotropia; LXT = left exotropia.

a recurrence of the chin-down AHP. Repeat resection of the bilateral inferior recti was performed in 1 of the 2 cases (patient 4; Table).

• **WEAKENING OF BOTH ELEVATORS:** Weakening of both elevators [BSRc 5-8 mm, bilateral inferior oblique recession (BIOc) or myectomy] was performed in 11 cases. The primary outcome was achieved in 7 of 11 patients, with mild chin-down AHP persisting in 4 patients (patients 15, 18, 19, and 22; Figure).

As seen in the Table, unfavorable outcomes included 1 patient (patient 14) who had a recurrence of the AHP, as well as 1 patient (patient 19) who developed A-pattern exotropia. Further recession of the superior recti successfully corrected the head posture in 1 patient (patient 14) for whom the AHP recurred (Table).

• **COMPARISON OF TECHNIQUES:** Both groups had similar rates of achieving a complete collapse of their chin-down AHP, with 7 of 11 patients attaining having no appreciable

residual AHP at last follow-up in each group. However, unfavorable outcomes were noted in 7 of 11 patients (64%) in the recession-resection group, compared with 2 of 11 patients (18%) in the weakening of both elevators group ($P = .03$). Reoperation was performed in 5 of 11 patients (45%) in the recession-resection group compared with 1 of 11 patients (9%) for weakening of both elevators; $P = .06$).

• **LONG-TERM FOLLOW-UP:** Mean follow-up among the 22 patients examined in this series was 56 ± 51 months. The response to surgery for each patient in our series is shown in the Figure. At last follow-up, 21 of 22 patients (95.0%) had an acceptable collapse of the AHP, defined as being at most a "mild" AHP within 15° of primary in the vertical plane, or none at all. Recurrence of the AHP occurred in 3 cases (patients 4, 7, and 14). Pre- and postoperative (last available follow-up) findings concerning visual acuity, alignment, ductions, and duration of follow-up are reported in Supplemental Table 2. No patient had more than a 2-line change in visual acuity.

DISCUSSION

NUMEROUS TECHNIQUES HAVE BEEN SUGGESTED FOR correction of a chin-down vertical plane AHP, including recession of the vertical recti alone,[6,11,14] recession-resection of the vertical recti,[2,6,11,14] and weakening of both elevators.[13] We initially used a recession-resection technique on the vertical rectus muscles—similar to that described by Parks and Mitchell[2] and Parks[6]—for correction of any form of AHP in the vertical plane. Approximately 8 years ago, 2 of the authors (SPD and DGM) anecdotally noted that several patients had recurrent AHP and therefore both began to preferentially perform superior rectus recession along with inferior oblique myectomy.

Surgery was typically performed on either the appropriate vertical rectus muscles or the inferior obliques for all patients. However, surgery on the horizontal rectus muscles was performed concomitantly to correct a coexistent horizontal strabismus in 2 cases (patients 9 and 20), with success in both. Of note, 1 of these patients (patient 20) had a V-pattern horizontal strabismus at initial consultation. This patient's AHP was associated with a null position of nystagmus and orthotropia. Though unlikely, it is possible that this patient had manifest latent nystagmus (fusion maldevelopment syndrome) rather than INS because formal eye movement recordings were not performed. Similarly, inferior oblique overaction as a cause of the AHP was unlikely.

Our study presents the largest series of patients receiving surgery for chin-down vertical plane AHP associated with INS. In our hands, recession-resection of the vertical recti and weakening of both elevators appear to be similarly effective in collapsing the AHP, with similar rates of success in the 2 groups with regard to achieving that primary outcome. When the AHP recurred (patients 4, 7, and 14), repeat surgery was sufficient to collapse the AHP to within 15° of primary gaze. We also found that recurrence of the AHP tends to present variably (11-30 months postoperatively).

Our results indicate that recession-resection for a chin-down AHP is accompanied by a significantly greater likelihood that an unfavorable outcome—either need for repeat surgery, induced strabismus, or lack of collapse of the AHP—develops postoperatively ($P = .03$). Unexpectedly, we found that recession-resection of the vertical recti often induced a V-pattern strabismus when used for correction of a chin-down AHP. This finding has not been reported in other series. The tendency toward this postoperative complication may be explained via an examination of the various functions of the vertical recti. Though the primary action of the superior and inferior rectus muscles is to elevate and depress the globe, respectively, the secondary action of both sets of these muscles is to adduct the globe. It is likely that the V-pattern esotropia induced in these patients is caused by a strengthening of the adducting force of the inferior rectus muscles in downgaze by resection. In addition, recession of the superior recti weakens adduction on upgaze. The resultant V-pattern esotropia required further surgical correction—bilateral medial rectus muscle recession and bilateral inferior oblique myectomy in 3 patients (patients 3, 5, and 10)—with success all cases.

Though there was some concern that weakening both elevators to correct a chin-down AHP would produce a significant postoperative limitation of elevation, this does not appear to be the case (Supplemental Table 2). Instead, our data suggest that the elevation deficit associated with a recession-resection technique may be greater; this may have been attributable to a restrictive strabismus produced by large resections.

There are several limitations to this study, including its retrospective nature and differing amounts of long-term follow-up among our patients. Our measurement of the degree of AHP was only available as an estimate. The use of a goniometer (or even prisms to straighten the head) would provide much more accurate information on the degree of AHP, and while this is a more acceptable standard, it is not typically used in clinical practice. That we were unable to detect a difference between the 2 groups—recession-resection versus weakening of both elevators for correction of a chin-down AHP—with regard to the rate of repeat surgery ($P = .06$) is most likely related to a limitation of power by the small sample size in this study. Selection bias was unlikely as our initially used recession-resection technique for correction of a chin-down AHP was later replaced by weakening of both elevators. However, because our change in practice was a result of perceived poor success of the former technique, the results are likely biased to favor the success of the latter.

Studies that are larger or that have longer follow-up periods might result in different findings. Given the

infrequency with which an AHP in the vertical plane caused by INS is observed, however, further exploration of differences in outcomes between these 2 techniques may benefit from a prospective study or from data compiled in a multicenter registry of patients with INS receiving surgery for an AHP in the vertical plane. Predictors of long-term prognosis, as well as clinical characteristics that may influence the surgeon's choice of technique could similarly be examined.

In conclusion, either recession-resection of vertical recti and weakening of both elevators each produce acceptable outcomes with regard to collapse of a chin-down AHP. However, recession-resection is associated with a greater likelihood of an unfavorable outcome, specifically the development of a V-pattern esotropia, which often requires a second surgery. In absence of other indications, weakening of both elevators is the preferable procedure for the correction of a chin-down AHP.

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