Recession-Resection of the Vertical Rectus Muscles for Chin-up Vertical Abnormal Head Position Associated With Infantile Nystagmus Syndrome

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ABSTRACT

Purpose: To review the correction of chin-up abnormal head position (AHP) due to infantile nystagmus syndrome via surgery on the vertical rectus muscles, typically a combined recession-resection of these muscles.

Methods: This was a review of 6 patients who underwent surgical correction of chin-up vertical AHP in the context of infantile nystagmus syndrome at an academic institution. The correction of AHP, visual acuity, ductions, and ocular alignment were noted both preoperatively and postoperatively. The need for repeat surgery, induced strabismus, or correction of AHP were also noted.

Results: Six patients underwent surgery for chin-up AHP. A combined recession-resection of the vertical rectus muscles (bilateral inferior rectus muscle recession of 5 to 8 mm; bilateral superior rectus muscle resection of 7 to 8 mm) was performed in 4 of 6 patients, and isolated bilateral recession of the inferior rectus muscles was performed in the remaining 2 patients. Four of 6 patients (67%) achieved complete correction of their AHP at the last follow-up visit, with a mild residual chin-up AHP persisting in the other 2 patients. One patient developed large angle exotropia, one had restrictive hypertropia and horizontal plane null position, and another developed an incomitant horizontal strabismus with exotropia in right gaze. Reoperation was performed in the former 2 patients, with successful correction of the strabismus in each.

Conclusions: Surgery on the vertical rectus muscles can reduce or eliminate a chin-up head position in patients with infantile nystagmus syndrome. Care should be taken to avoid producing a restriction of depression in abduction if the amount of resection is too large.

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INTRODUCTION

An abnormal head position (AHP) in the vertical plane can occur in patients with infantile nystagmus syndrome if the null position of the nystagmus is located eccentrically in either up gaze or down gaze.[1,2] Eye muscle surgery aimed at correcting such an AHP is accordingly directed at the cyclovertical muscles to shift the null position into primary gaze.[3,4]

We recently reported our results from a series of 22 patients with infantile nystagmus syndrome and a chin-down AHP who underwent eye muscle surgery to reduce their head position moving the eyes into down gaze.[5] The correction of a chin-up AHP

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on requires moving the eyes into up gaze, typically by weakening the depressor muscles, strengthening the elevator muscles, or both.[2,6-13] Some reports have incorporated superior oblique weakening in the surgical plan, in conjunction with either inferior rectus recession or superior rectus resection.[10-12] We avoid superior oblique weakening because of the potential for induced excyclotorsion, which may preclude postoperative fusion and its relative unpredictability. In the current study, we report our experience with surgery in 6 patients with chin-up AHP in the setting of infantile nystagmus syndrome wherein inferior rectus weakening was performed in combination with superior rectus resection in 4 of the 6 patients.

PATIENTS AND METHODS

An institutional surgical database of pediatric strabismus surgeries performed by a single surgeon (SPD) was reviewed to identify patients who underwent eye muscle surgery for infantile nystagmus associated with an AHP from 1995 to 2018. Of 126 such patients, 24 had eye muscle surgery performed for a vertical plane AHP by this surgeon. Of these, 6 patients fulfilled the inclusion criteria for this retrospective observational case series: they presented with a chin-up AHP that was associated with infantile nystagmus syndrome, they were younger than 18 years at the time of surgery, and they had at least 2 months of postoperative follow-up care. Two of the 6 patients (patients 3 and 4) had had previous eye muscle surgery for strabismus. Eighteen patients with a vertical AHP at presentation were excluded from this study because their head position was chin-down rather than chin-up.

This study was evaluated by the Vanderbilt University Institutional Review Board and determined to qualify for exempt status. It was 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

Our primary measure of interest was the magnitude of AHP in the vertical plane, which was typically approximated by observation as being either mild (1 to 15 degrees), moderate (16 to 30 degrees), or severe (31 degrees or more). Best corrected visual acuity (tested in each eye separately when possible, or tested in both eyes simultaneously when not possible), ocular alignment, and ductions (graded on a scale of +4 to -4) were also noted. These variables were measured preoperatively and at postoperative visits collected at 1 week, 2 months (±6 months), and 1 year (±6 months) after surgery, and the last available follow-up visit. The need for repeat surgery and postoperative induced strabismus or lack of correction of AHP was also noted.

Surgical Technique

Strabismus surgery for the correction of the AHP and any subsequent procedures were performed by the senior author (SPD). Bilateral recession of the inferior rectus muscle combined with resection of the superior rectus muscles was performed on 4 of 6 patients. The remaining 2 patients had had either previous (patient 4) or simultaneous (patient 6) horizontal rectus muscle recession to correct a coexisting horizontal strabismus; therefore, bilateral inferior rectus muscle recession was performed to decrease the risk of anterior segment ischemia.

Following a limbal peritomy, dissection was carried down to the bare sclera posteriorly to where the vertical rectus muscles penetrate through Tenon's capsule. The muscle was secured with a polygalactin suture and, following disinsertion, the inferior rectus muscle was recessed via a modified hang-back technique. The muscle pole was secured with a small scleral bite at the recession location and with a second scleral pass at the original muscle insertion, where the suture was tied. The superior rectus muscle was resected and sewn back to its original point of insertion. Care was taken to ensure that superior and inferior eyelid retractors were dissected from their respective rectus muscles before either recession or resection of those rectus muscles.

Surgery on each eye was typically performed symmetrically and in an equal amount depending on the size of the eye, the length of the muscle available for resection, and the exposure of the globe within the orbit to achieve a maximum effect. In one patient, a modification of the surgical dose was necessary for the correction of a vertical heterotropia. Surgery on the horizontal rectus muscles was performed concomitantly to correct a coexistent horizontal strabismus in one patient (patient 6) and had been performed previously in another patient (patient 4). None of our patients underwent a physical therapy regimen for torticollis following eye muscle surgery.

TABLE 1

Preoperative Patient Demographics and Clinical Data

Patient No.	Sex	Age (m)	INS Etiology	Null	AHP	Other Eye Dx	Prior EMS	Initial EMS for AHP	Subsequent EMS
1	F	112	Idiopathic	D	CU, severe	–	–	BIRc-8; BSRs-8	–
2	M	53	Cone dystrophy	D	CU, severe	–	–	BIRc-8; BSRs-8	BLRc-8 (1); BMRs-4 (2)
3	F	93	Oculocutaneous albinism	D	CU, severe	Alt XT (corrected)	LLRc-8	BIRc-7; BSRs-8	LSRc-4; LMRc-8; RLRc-10
4	M	89	Ocular albinism	D	CU, severe	ET (corrected)	BMRc-11	RIRc-6, LIRc-7	–
5	M	49	Ocular albinism	D	CU, severe	–	–	BIRc-8; BSRs-7	–
6	M	78	Idiopathic	D	CU	A-pattern ET	–	BIRc-5; BMRc-4.5	–

INS = infantile nystagmus syndrome; AHP = abnormal head position; Dx = diagnosis; EMS = eye muscle surgery; D = down gaze; CU = chin-up; BIRc = bilateral inferior rectus recession; BSRs = bilateral superior rectus resection; Alt = alternating; XT = exotopia; LLRc = left lateral rectus recession; LSRc = lateral superior rectus recession; LMRc = lateral medial rectus recession; RIRc = right inferior rectus recession; LIRc = lateral inferior rectus recession; ET = esotropia

RESULTS

Clinical characteristics of the 6 patients who presented with chin-up AHP in the context of infantile nystagmus syndrome are summarized in **Table 1**. A known afferent abnormality was noted in 4 of 6 patients: ocular albinism in 3 patients (patients 3, 4, and 5), and a cone dystrophy in 1 patient (patient 2). The nystagmus had a predominant horizontal component in all 6 patients. The degree of preoperative AHP was severe in 5 patients, and not quantified for 1 patient (patient 6). All 6 patients initially had a null in down gaze in conjunction with their chin-up AHP. The mean age at time of surgery was 79 ± 24 months, and 4 of 6 patients were male. Two patients (patients 3 and 4) had prior eye muscle surgery for correction of horizontal strabismus before presenting for treatment at our institution.

Surgical Management of Patients With Chin-up AHP

Concurrent recession-resection (bilateral inferior rectus muscle recession of 7 to 8 mm; bilateral superior rectus muscle resection of 7 to 8 mm) was employed in 4 patients (patients 1, 2, 3, and 5) who presented with a chin-up AHP. In 2 patients (patients 4 and 6: bilateral inferior rectus recession of 5 to 7 mm), recession of the inferior rectus muscles alone was performed because of previous or concurrent horizontal rectus muscle recession. A modification of the surgical dose was deemed necessary in 1 patient for the correction of a vertical heterotropia (patient 4). Surgery on the horizontal rectus muscles was performed concomitantly to correct a coexistent horizontal strabismus in 1 patient (patient 6).

At the last follow-up visit, a null in primary gaze was achieved in 4 patients, with mild chin-up AHP persisting in 2 patients (patients 1 and 4). One patient (patient 2) developed a large angle exotropia, the correction of which required two subsequent surgeries: bilateral lateral rectus recession followed by bilateral medial rectus resection performed 7 years and 10 years after the initial procedure to correct the AHP, respectively. One patient (patient 3) had an asymmetric bilateral restrictive hypertropia in abduction (probably due to the resection effect of the superior rectus) with a horizontal plane null position requiring additional surgery (left superior rectus recession, medial rectus recession, and right lateral rectus recession), performed at 5 months (**Table 1**). A final patient (patient 4) developed an incomitant horizontal strabismus with exotropia in right gaze, but subsequent surgical correction was not performed.

Long-term Follow-up Care

Mean follow-up time among the 6 patients examined in this series was 39 ± 49 months. Three patients (patients 2, 3, and 4) were observed for at least 2 years after their initial surgery. At the last follow-up visit, the complete correction of AHP was

achieved in 4 of 6 patients, and an acceptable reduction in AHP, defined as being at most a "mild" AHP within 15 degrees of primary in the vertical plane (two patients), was achieved in all 6 patients. Preoperative and postoperative (last available follow-up visit) findings concerning visual acuity, alignment, ductions, and duration of follow-up are reported in **Table 2**. Visual acuity remained stable (or slightly improved) for all patients for whom data were available (patients 1 to 5) to provide comparison between initial visual acuity and that at the last follow-up visit (**Figure 1**). Visual acuity data were not available at the last follow-up visit for patient 6.

DISCUSSION

The current study documents the successful reduction of a chin-up AHP in 6 patients with infantile nystagmus syndrome. Inferior rectus recession was performed in all 6 patients. Superior rectus resection was also performed in 4 patients. We successfully corrected or reduced the AHP in all 6 patients, and we believe that this represents the largest such series where combined surgical correction was confined to the vertical rectus muscles.

Two of our patients required surgery on the horizontal rectus muscles to correct a horizontal strabismus that was manifest in addition to the surgery for the head position. Patient 4 had had previous bilateral medial rectus muscle recessions, whereas patient 6 had an A-pattern esotropia and underwent concurrent medial rectus weakening at the time of the nystagmus surgery. We intentionally avoided surgery on the superior rectus muscles in these two patients to limit the likelihood of anterior segment ischemia. Recession of the inferior rectus muscles alone was performed for the correction of chin-up AHP, with acceptable correction of the AHP at the last follow-up visit in both patients. Thus, maximum inferior rectus recession alone may be sufficient initial treatment for a chin-up AHP. However, a previously reported series of isolated inferior rectus recession found that additional surgery was often needed to fully correct the chin-up AHP.[8] Our observation that our surgery in these two patients did not fully correct their AHP is consistent with the results of Yang et al.[8]

Our surgical preference for correcting chin-up AHP is to perform maximal inferior rectus recession along with superior rectus resection, similar to recommendations by Mitchell and Parks[2] and Yang

TABLE 2

Visual Acuity, Alignment, and Ductions

Patient No.	Visual Acuity		Alignment in Primary		Ductions[a]		AHP		
	Preop	Last Follow-up	Preop	Last Follow-up	Preop	Last Follow-up	Preop	Last Follow-up	Last Follow-up (m)
1	20, 20, _	20, 20, _	Ortho	Ortho	Full	-1 depr w/ abd, -1 depr w/ add OU	Severe	Mild	9
2	_, _, 300	100, 100, 100	–	Ortho	Full	Full	Severe	None	128
3	100, 80, 80	100, 80, 80	Ortho	8 ET	Full	-1 abd OD; -1 elev, -2 elev w/ add, -1 add OS	Severe	None	65
4	80, 125, 80	80, 70, 70	10 ET	10 ET 6 LhypoT	Full	Full	Severe	Mild	25
5	80, 100, 70	70, 60, 50	Ortho	Ortho	Full	+1 elev w/ abd, -2 depr w/ abd OS	Severe	None	6
6	100, 100, 60	_, _, _	18 LET	Ortho	Full	Full	[b]	None	3

AHP = abnormal head position; preop = preoperative; ortho = orthotropic; depr = depression; abd = abduction; add = adduction; OU = both eyes; ET = esotropia; OD = right eye; elev = elevation; LhypoT = left hypotropia; OS = left eye

[a]Ductions: overactions are denoted by "+," whereas limitations are denoted by "-." The term "w/" associates components of an oblique gaze direction (eg, elevation with adduction).

[b]Preoperative degree of chin-up AHP not quantified for patient 6 prior to operative repair.

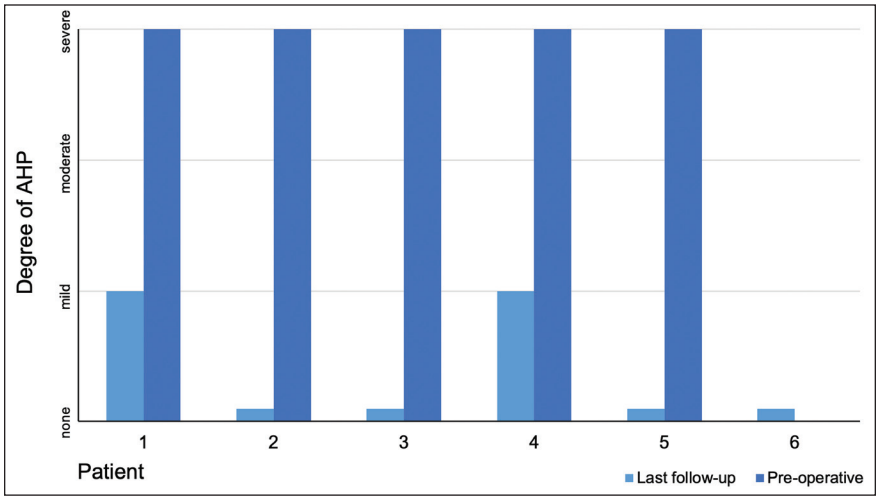

Figure 1. Degree of abnormal head position (AHP) over time. The degree of vertical head position at the preoperative and postoperative (last follow-up) visits displayed for each patient who presented with a chin-up AHP. Note that the degree of chin-up AHP was not quantified for patient 6 preoperatively.

et al.[8] The latter suggested a total of 12 to 20 mm of surgery on the vertical rectus muscles be evenly split between recession and resection depending on the degree of AHP: 10 mm recession alone for an AHP up to 20 degrees.[8] Similar to Yang et al, we encountered significant postoperative strabismus (horizontal strabismus that required additional surgery in 2 patients and bilateral restrictive hypertropias in abduction) that we believed was caused by extensive superior rectus resection, and this was corrected with additional surgery. We also encountered mild postoperative gaze restrictions in 3 patients (patients 1, 3, and 5), although these limitations were similarly non-symptomatic. We did not note any instances of symptomatic postoperative torsion in our series, and that may be because we did not operate on the superior oblique muscle. The restrictive component caused by excessive resection of the superior rectus muscle suggests that surgery be more weighted toward recession of the inferior rectus muscle instead of evenly splitting resection and recession.[2,8]

Our experience was more straightforward than that of Roberts et al,[11] who encountered difficulty in achieving a satisfactory correction of the AHP in 4 patients with a chin-up position. Their treatment via a recession-resection technique was initially met with failure to elicit correction of the AHP, requiring additional surgery in 2 patients. All 4 patients "manifested some degree of residual torticollis postoperatively" between 8 and 15 degrees.[11] The minimal asymptomatic depression deficiencies Roberts et al observed in 3 of 4 of their patients was thought to be suggestive of successful correction of the AHP, with it being a byproduct of the sufficiently large surgical doses employed on the vertical muscles.

The current study was limited by its retrospective nature and varying amounts of postoperative follow-up. Although we were unable to report multiple year follow-up for all 6 patients, the head position attained for those patients with at least 2 years of follow-up beyond the initial surgery appears to be stable. Thus, as has been the experience of others, we expect that late postoperative changes are unlikely following recession-resection for a chin-up AHP.[8,12,13] Owing to the infrequency with which patients with chin-up AHP due to infantile nystagmus syndrome present, larger studies with longer follow-up periods are required to further characterize and to evaluate and improve on surgical techniques applicable to this patient population. Furthermore, our measurements for degree of AHP were estimated, rather than quantified via the use of a goniometer or prisms to straighten the head. Although the latter techniques provide much more accurate information on the degree of AHP and are the more acceptable standard, their use in clinical practice is not typical. Selection bias was unlikely because all operations were performed by the same surgeon.

Our experience with correcting chin-up AHP in patients with infantile nystagmus syndrome demonstrates that surgery applied solely to the vertical rectus muscles, with large bilateral recession of the inferior rectus muscles, combined with resection of the superior rectus muscles where possible, produces durable correction of the head position. Although this technique can induce strabismus or gaze limitation due to the large degrees of eye muscle manipulation required, suitable correction of the former can be achieved through subsequent surgery.

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