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Original Article

The Outcome of Inferior Oblique Myectomy for Apparent Inferior Oblique Overaction Associated with Craniosynostosis

Hye Won Park[1,2], Sueng-Han Han[1,3]

[1]*Department of Ophthalmology, Yonsei University College of Medicine, Seoul, Korea*
[2]*Department of Ophthalmology, Konyang University College of Medicine, Daejeon, Korea*
[3]*Institute of Vision Research, Department of Ophthalmology, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea*

Purpose: Strabismus in patients with craniosynostosis is common, but surgical correction of strabismus in these patients remains challenging. We report our findings in six patients (four of whom were Korean) with craniosynostosis who underwent strabismus surgery to specifically address V-pattern horizontal strabismus with moderate-to-severe inferior oblique (IO) overaction, using IO myectomy at a single tertiary hospital between 2005 and 2016.

Methods: We recorded preoperative characteristics including sex, age, type of strabismus, versions grading, refractive error, and visual acuity. The grading of cyclorotation of horizontal rectus muscles by V-pattern categorized using coronal computed tomography imaging.

Results: Of the six patients, exodeviation was found in four patients and vertical deviation in two patients in primary position. One patient had both horizontal and vertical strabismus. Available computed tomography imaging showed that V-patterns were category 1 (mild) in two patients, category 2 (moderate) in one patient, and category 3 (severe) in two patients. Complete success was defined as absence of IO overaction any more. Overall complete success rate of IO myectomy was 83.3%.

Conclusions: IO myectomy appeared to have some benefits in V-pattern horizontal strabismus with moderate-to-severe IO overaction in patients with craniosynostosis.

Key Words: Craniosynostoses, Inferior oblique overaction, Korea, Myectomy, V-pattern strabismus

Craniosynostosis is a skull deformity defined as the premature fusion of one or more of the cranial sutures [1]. Craniosynostosis can occur as part of a syndrome or as an isolated defect. Syndromic craniosynostosis, such as the Crouzon and Apert syndromes, is less common than nonsyndromic cases. A multidisciplinary team approach including pediatric ophthalmologists, plastic surgeons, and neurosurgeons is needed to address all disability associated with syndromic craniosynostosis [2]. It has been suggested that divergent orbit, displaced extraocular muscles, and extorsion of the obit causes strabismus associated with craniosynostosis [3]. Craniosynostosis occurs in approximately 1 in 2,500 children [4]. Children with craniosynostosis syndromes can have a range of ophthalmic complications,

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Corresponding Author: Sueng-Han Han, MD, PhD. Institute of Vision Research, Department of Ophthalmology, Severance Hospital, Yonsei University College of Medicine, 50-1 Yonsei-ro, Seodaemun-gu, Seoul 03722, Korea. Tel: 82-2-2228-3570, Fax: 82-2-312-0541, Email: shhan222@yuhs.ac

including of visual acuity, refractive error, ocular arrangement and corneal and optic atrophy [5]. Amblyopia due to strabismus and high refractive errors are potential causes of visual loss in these children [6]. Therefore, it is important to check regularly by a pediatric ophthalmologist [7].

In a systematic review on the prevalence, strabismus was the most common ocular anomaly in both nonsyndromic and syndromic craniosynostosis [8]. Strabismus can occur primary, but also as a result of craniofacial surgery [7]. Two-thirds of patients with craniosynostosis have strabismus [9], with V-pattern horizontal strabismus being the most common pattern [10]. The influence of orbital architecture was associated with strabismus in patients with craniosynostosis [11]. Although inferior oblique (IO) weakening procedures could not normalize ocular motility (extorted orbit shifts the medial rectus muscle up and the lateral rectus down, which simulates IO overaction), previous studies have examined the effective surgical techniques including superior oblique (SO) tuck, IO anterior transposition to correct the V-pattern strabismus [4,12]. No previous study has reviewed the management the IO muscle myectomy surgery to correct the V-pattern horizontal strabismus with marked IO overaction in a group of Korean patients with craniosynostosis.

We report our findings in six patients (four of whom were Korean) with craniosynostosis who underwent strabismus surgery to specifically address V-pattern horizontal strabismus with moderate-to-severe IO overaction using IO myectomy.

Materials and Methods

Ethics statement

This study was approved by the Institutional Review Board of Severance Hospital (No. 4-2017-0350) and was performed in compliance with the tenets of the Declaration of Helsinki. The requirement for informed consent was waived due to the retrospective nature of the study.

Study design and setting

We retrospectively reviewed the medical records of all patients with craniosynostosis (as diagnosed by plastic surgeons) that had V-pattern horizontal strabismus with marked IO overaction that underwent strabismus surgery, including IO myectomy, at a single tertiary hospital between 2005 and 2016.

For myectomy, the conjunctiva and Tenon capsule were dissected in the inferior temporal zone, and then the IO muscle was isolated with a muscle hook; 8 to 10 mm section of the IO was excised. After complete cauterization of the cut edges, the conjunctival incision was sutured.

Ocular alignment was measured in the primary position at 6 m and 33 cm using accommodation controlling targets. In the patients that could not concentrate on the targets, strabismus was assessed using Hirschberg method. The degree of overaction of the IO muscle was quantified on a scale from +1 to +4, in which 0 indicated normal function.

Coronal computed tomography (CT) imaging from the posterior to the anterior orbit was performed to measure rectus muscle exocyclorotation. Digital imaging and communications in medicine images were exported from the coronal orbit CT scans. Exocyclorotation was identified based on a horizontal line joining the center of the bellies of the medial and lateral rectus muscles and a vertical line joining the center of the bellies of the superior and inferior rectus muscles. In mild V-pattern subjects, the orbital walls surround typically oriented four rectus muscles. In moderate subjects, medial bowing of sphenoid greater wing leads to the lateral rectus down. In severe V-pattern subjects, the shorter and wider orbits was accompanied by lateral displacement of the superior rectus muscles [13]. The severity of V-pattern on CT imaging are structurally categorized in three groups according to the anatomy of the posterior orbit: category 1 (mild, two patients), category 2 (moderate, one patient), and category 3 (severe, two patients).

All patients who were old enough had their visual acuity (VA) measured, while those who were not had their fixation reflex assessed using the central, steady, and maintained method [14]. Stereopsis was measured with the Randot test in patients who were old enough and cooperative. All the patients had their versions and ductions examined to detect any limitations of extraocular movement. Participants also underwent cycloplegic refraction and fundus examinations.

All surgeries were performed by three surgeons (JBL, SHH, and JH). All patients except for one had postoperative follow-up of at least 1 month after strabismus surgery. In all cases, the preoperative and postoperative examinations were performed by the same ophthalmologist.

Results

Patient characteristics

There were five male patients and one female patient in this study. In this series, four patients were referred to us from within Korea (including one half-Korean, half-Vietnamese), one from Canada, and one from Russia. Of these, one patient was diagnosed with syndromic craniosynostosis due to Apert syndrome.

A total of six children with a mean age at the time of surgery of 4 years (range, 2–7 years) were treated for moderate-to-severe IO overaction with IO myectomy with a mean duration from the first visit to surgery of 13.2 months (range, 4–36 months). One patient was scheduled for surgery 5 months after the first visit, but the patient's delay in presentation was attributable to a delay on the part of the parents themselves.

A total of four patients with craniosynostosis underwent at least one craniofacial surgery at a mean age of 14.3 months (range, 7–20 months). Strabismus have occurred primary in three patients, but also as a result of craniofacial surgery in one patient.

VA was documented in four patients. All patients had cycloplegic refraction data and fundus findings. Visual evoked potential data was available for one patient.

The ability to fix and follow in each eye was present in six patients. The mean logarithm of the minimum angle of resolution with correction in the four patients (mean age, 5 years) that completed optotype VA testing was 0.1 (eight eyes of four patients).

A total of four patients were hypermetropic, and there was no one with myopia. Astigmatism of ≥0.75 diopters (D) was present in five patients and anisometropia of ≥0.75 D was present in one patient (Table 1).

Of the five patients able to perform cover-uncover and alternate prism cover testing, six (100%) showed overaction of the IO muscle in preoperative testing. Of these, exodeviation was found in four patients, vertical deviation in two patients in primary position, and both horizontal and vertical deviations were observed in one patient.

We detected rectus muscle excyclorotation based on the

Table 1. Refractive error and BCVA

Patient no.	Cycloplegic refraction			
	Spherical lens (D)	Cylinder lens (D)	Spherical equivalent	BCVA
1				
Right eye	+0.50	–0.50	+0.25	0.9
Left eye	+0.75	–0.75	+0.38	1.0
2				
Right eye	+3.00	–2.00	+1.00	NR
Left eye	+2.50	–1.50	+1.75	NR
3				
Right eye	+1.00	–1.25	+0.37	0.7
Left eye	+1.00	–1.25	+0.37	0.7
4				
Right eye	+1.00	–1.00	+0.50	NR
Left eye	+1.00	–1.00	+0.50	NR
5				
Right eye	+1.75	NR	+1.75	1.0
Left eye	+2.25	–1.00	+1.75	1.0
6				
Right eye	+3.50	–1.50	+2.75	NR
Left eye	+3.00	NR	+3.00	NR

BCVA = best-corrected visual acuity; D = diopters; NR = not reported.

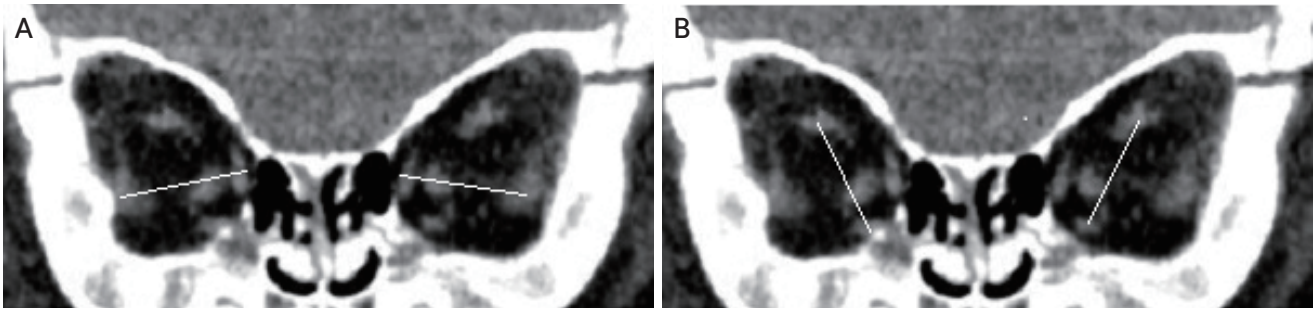

Fig. 1. Coronal views on computed tomography scan showing lateral displacement of the superior rectus muscle and downward displacement of the lateral rectus muscle. (A) A horizontal line joining the center of the belly of the horizontal rectus muscles. (B) A vertical line joining the center of the belly of the vertical rectus muscles.

anatomy on the coronal posterior orbital CT. The severity of the V-pattern of increased excyclorotation of the rectus muscles lead to lateral displacement of the superior rectus and inferior displacement of the lateral rectus muscles (Fig. 1A, 1B). Available CT imaging showed that two patients had category 1 (mild), one patient had category 2 (moderate), and two patients had a category 3 (severe) V-pattern (Fig. 2A–2C). A severe V-pattern with limited

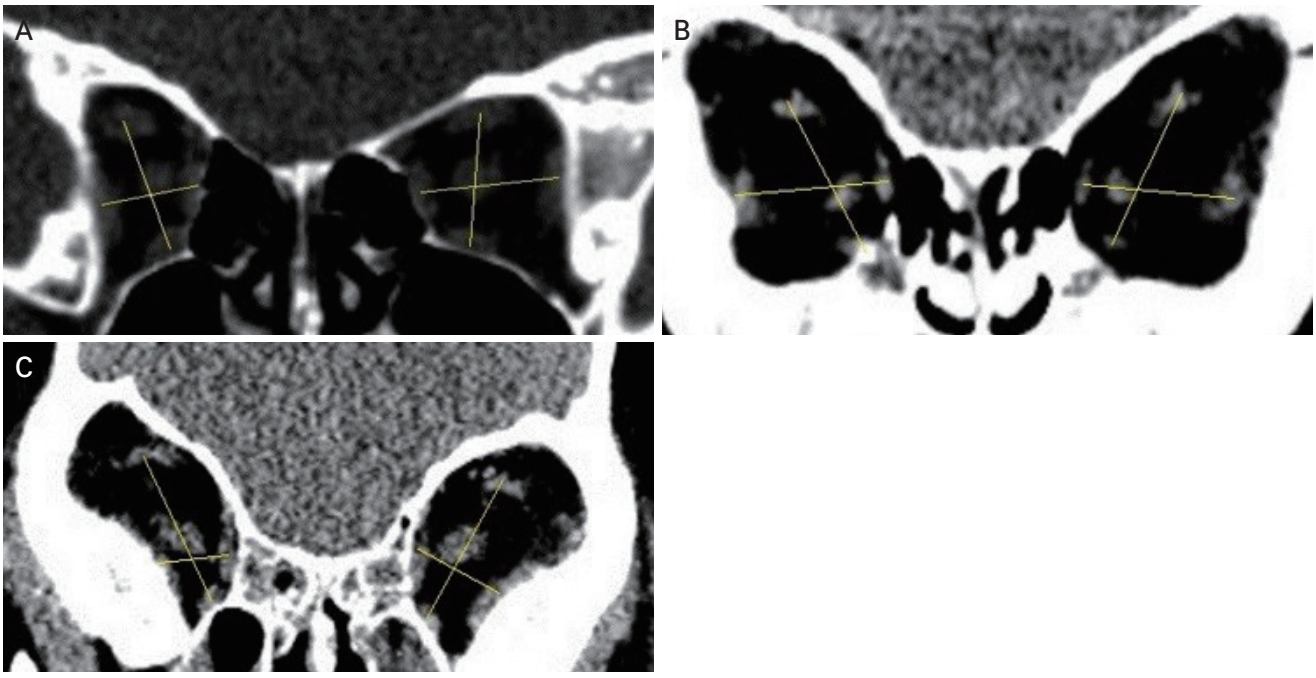

Fig. 2. Coronal orbital computed tomography scans. (A) In mild V-pattern subjects, the orbital walls surround typically oriented four rectus muscles. (B) In moderate subjects, medial bowing of sphenoid greater wing leads to the lateral rectus down. (C) In severe V-pattern subjects, the shorter and wider orbits was accompanied by lateral displacement of the superior rectus muscles. Horizontal lines joining the center of the belly of the horizontal rectus muscles and vertical lines joining the center of the belly of the vertical rectus muscles.

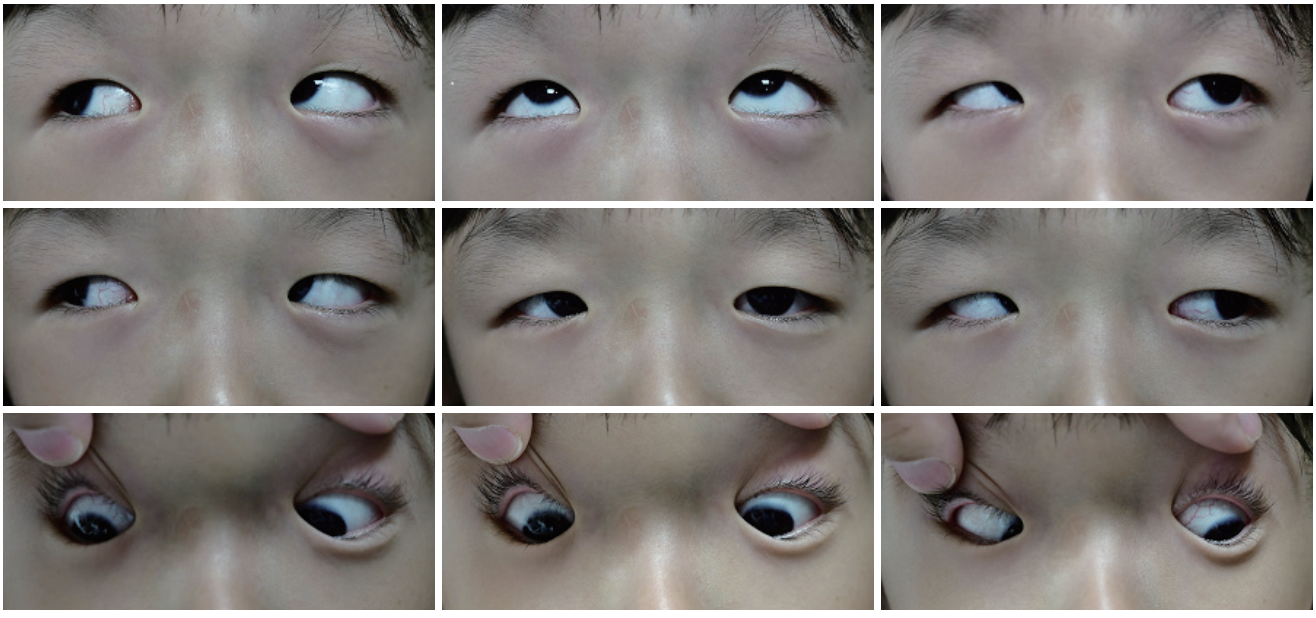

Fig. 3. Preoperative nine cardinal exam showing apparent bilateral inferior oblique overaction (patient 5). The patient's parent provided written informed consent for publication of the clinical images.

abduction in both eyes was found in one patient. In our study, only one patient with severe V-pattern showed IO overaction of 4+ in both eyes.

Surgical outcome

Complete success was defined as absence of IO overaction any more. Overall complete success rate of IO myectomy was 83.3%. IO myectomy appeared to have some benefits in V-pattern horizontal strabismus with moderate-to-severe IO overaction in patients with craniosynostosis (Figs. 3, 4). However, there was no correlation between increased excyclorotation of the rectus muscles and IO overaction in mild-to-moderate and moderate-to-severe V-pattern patients. The extent of both the increased excyclorotation of the rectus muscles and the IO overaction is shown in Table 2.

There was no one with optic disc pallor in fundus exam. The fundus photograph of one patient showed excyclotorsion of the right eye. All patients, except for one, were un-

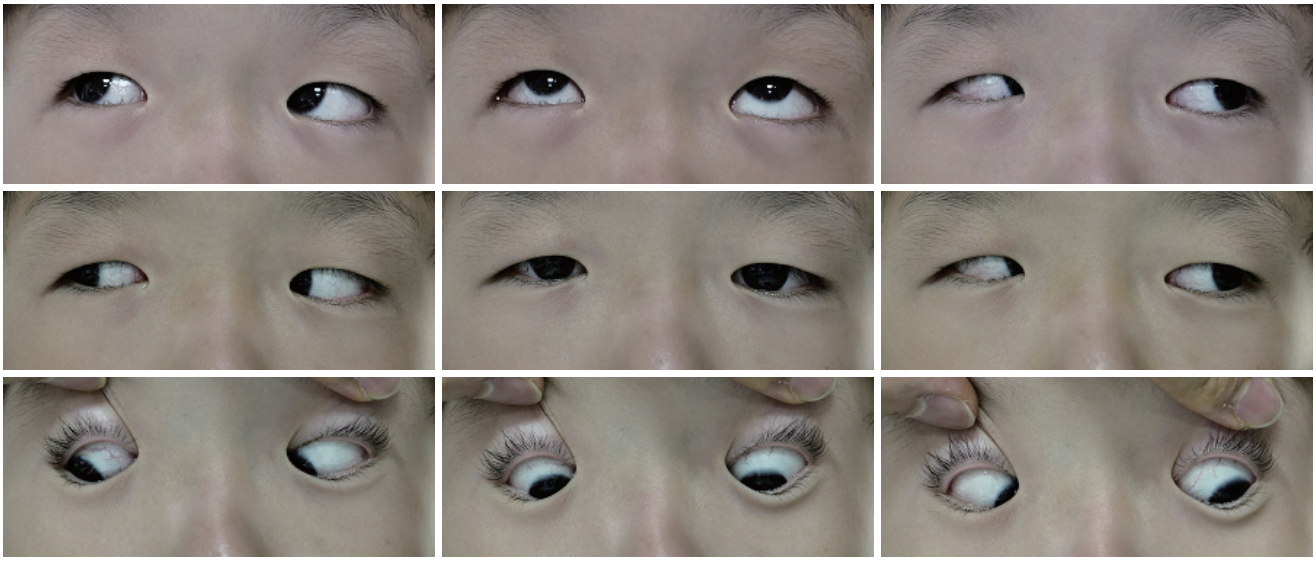

Fig. 4. Postoperative nine cardinal exam showing apparent bilateral inferior oblique overaction (patient 5). The patient's parent provided written informed consent for publication of the clinical images.

Table 2. Preoperative and postoperative data

Patient no.	Preoperative alignment			Postoperative alignment			V-pattern on CT*	Surgery
	Primary position (PD)	IO overaction		Primary position (PD)	IO overaction			
		Right eye	Left eye		Right eye	Left eye		
1	14△XT	4	2	16△XT	0	1	NR	BIO myectomy
2	10△RHT	3.5	2	Straight	0	0	Category 3	BIO myectomy
3	20△XT	3	3	14△XT	0	0	Category 2	BLR recession 6.5 mm BIO myectomy
4	50△XT	3.5	3.5	Straight	0	0	Category 1	BLR recession 10 mm BIO myectomy
5	14△RHT	3.5	0	Slightly RHT	0	0	Category 1	RSR recession 4.0 mm RIO myectomy
6	45△AXT	4	4	14△XT	0	0	Category 3	BLR recession 9.0 mm BIO myectomy

CT = computed tomography; PD = prism diopter; IO = inferior oblique; △ = prism diopter; XT = exotropia; NR = not reported; BIO = bilateral inferior oblique; RHT = right hypertropia; BLR = bilateral lateral rectus; RSR = right superior recuts; RIO = right inferior oblique; AXT = alternative exotropia.
*Category 1, mild; category 2, moderate; category 3, severe.

cooperative during the stereopsis test. Only one patient completed the Randot stereopsis test, earning a score of nil arcsecond.

Clinical characteristics

Other ophthalmic findings revealed that one patient had epiblepharon, two patients had entropion, one patient had nasolacrimal obstructions in both eyes. Associated systemic findings included two patients who had cardiac diseases (ventricular septal defect and atrial septal defect). Although detailed inspection was not routinely performed, no specific extraocular muscle abnormalities, such as absence, anomalous insertion, or anomalous anatomy, were found in this series.

Discussion

This study is the first to show the surgical outcomes of IO myectomy in children with craniosynostosis in Korea over a mean follow-up period of 12 months. Contrary to expectations, this study showed overall complete success rate of IO myectomy as 83.3%, which means some benefits in V-pattern horizontal strabismus with moderate-to-severe IO overaction in patients with craniosynostosis.

There are both similarities and differences between the patient population in the previous studies and those in ours [13,15,16]. The V-pattern, where changes in vertical misalignment are assessed based on alternative prism cover tests, and the available orbital CT imaging, through which excyclorotation of the rectus muscles is assessed, are consistent with the previous studies, However, unlike the postulation by Dagi et al. [13] (that the severity of the V-pattern is associated with the degree of excyclorotation), three of the six patients with V-pattern strabismus in our study showed category 2 to 3 excyclorotation of the rectus muscles with no definite correlation to the degree of overaction of the IO muscle. However, one of the six patients with syndromic craniosynostosis showed category 3 excyclorotation of the rectus muscles and +4 overaction of the IO muscle. This finding is in agreement with Dagi et al. [13]'s finding, which showed that a more severe V-pattern was noted in patients with Apert syndrome.

One patient (patient 6) in our study was found to have a *FGFR2* mutation, c.758C>G (p.P253R), on chromosome 10q26 using polymerase chain reaction sequencing of peripheral blood. The patient was diagnosed with Apert syndrome, which typically involves multiple cranial sutures [17]. She had mental retardation, an ostium secundum atrial septal defect, a submucosal cleft palate, and bilateral pansyndactyly of the hands and feet. She had limited (–4) bilateral ocular movement on abduction. Other ophthalmic findings included epiblepharon, entropion, and nasolacrimal duct obstruction. She underwent an additional Hotz operation, correction of entropion, and intubation of a silicone tube in both eyes. The patient did not exhibit abnormal insertion or absence of extraocular muscles as described in previous studies [18,19].

As for craniofacial surgery, surgical management includes remodeling of the skull vault during infancy, surgery for facial and orbital correction at an age of 5 to 7 years, and advancement surgery for maxilla/mandible during teenage life [6]. In staged surgical approach, one patient (patient 2) has treated at 3 months for decompression of the vault of the cranium and suture release. Three patients in our study have treated for reshaping of the orbitofrontal area plus advancement using techniques such as strip craniectomy or midface distraction osteogenesis. This study is a short-term follow-up observation study, so there is no progression data on the surgery for facial and orbital correction.

Since one early study [20], case reports describing craniosynostosis in Korean patients have been rare. No single study exists which assesses the outcome of IO myectomy in children with V-pattern strabismus.

We have assumed that anomalous lateral displacement of the superior rectus muscle and downward displacement of the lateral rectus muscle were involved in the moderate-to-severe overaction of the IO muscle observed in the development of V-pattern strabismus in craniosynostosis patients. There are several other possible explanations for the etiology; the overaction of the IO muscle and/or the underaction of the SO muscles may contribute to V-pattern strabismus in craniosynostosis [21]. The decrease in the anteroposterior dimension of the medial orbital wall may mechanically inhibit SO function. In addition, it can be assumed that an abnormal rectus muscle pulley position contributes to the underaction of SO function, resulting in V-pattern strabismus in craniosynostosis [14,16,22]. In accordance with these possible mechanisms, a previous case study showed that the abnormal rectus muscle position was

indeed associated with V-pattern strabismus in craniosynostosis [12].

There is no confirmed surgical method for V-pattern strabismus in craniosynostosis [6]. So different surgical approaches have been used on V-pattern strabismus. Several studies have examined including anterior transposition IO muscle, SO tuck, denervation/extirpation, muscle transposition surgery and IO myectomy. Previous study has reported that anterior transposition is preferred when large V-patten, while myectomy is preferred for moderate V-pattern [23]. In cases with V-pattern exotropia, vertical upshifting of the lateral rectus could be the surgical option [24]. However, previous studies have reported that unpromising outcomes after weakening surgeries the IO muscle [25,26]. A possible explanation for this result might be related to management of only the IO muscle while the angle of the rectus muscle excyclorotation remained unchanged [13]. It seems possible that V-pattern strabismus may be due to multifactorial etiology. However, IO myectomy appeared to have some benefits in our study. Though, caution must be applied to the interpretation of these results due to the small sample size, especially because no single surgery has yet proved to be more effective in V-pattern strabismus patients with craniosynostosis. In addition, it is important to bear in mind the possible absence of extraocular muscle insertions in patients with craniosynostosis, especially the superior rectus and oblique muscles. Therefore, a detailed preoperative examination should be performed before surgery.

There are some limitations to our study. Our study was a retrospective case series. Therefore, we could not perform a comparative analysis between craniosynostosis patients with and without a history of other IO weakening surgery. Furthermore, we could not determine the relationship between the excyclorotation of the rectus muscles and the overaction of the IO muscle in the development of V-pattern strabismus in craniosynostosis patients. Further studies with larger sample size are required to determine this relationship. Another limitation was the small sample size, because we studied only patients with overaction of the IO muscle associated with craniosynostosis who were treated with IO myectomy.

Despite the limitations, our study has several significant strong points. This is the first Korean study to investigate the outcome of IO myectomy for V-pattern strabismus in craniosynostosis patients. Furthermore, through available CT imaging, we have investigated the degree of excyclorotation of the rectus muscles in these patients, including two patients with category 1, one patient with category 2, and two patients with a category 3 V-patterns. Significantly, our study included a confirmed case of syndromic craniosynostosis due to Apert syndrome, which is rare in the Asian population. We have made an effort to determine the association, if any, between the mechanical and functional aspects of V-pattern strabismus in craniosynostosis patients.

The mean age at the time of surgery 4 years, and only one patient underwent stereopsis testing (where the result was 0), and sensory testing through long-term follow-up as well as a comparative analysis of the timing of surgery are important in future studies.

In conclusion, our results suggest that although IO weakening surgery may not normalize excyclorotation of the globe, moderate-to-severe IO overaction associated with craniosynostosis may still show some improvement after this procedure.

Conflicts of Interest: None.
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