Original Article

Clinical features of Helveston syndrome and discussion of individualized surgical design

Man Qin[1#], Guanglong Zhou[2#], Liping Xue[2], Jiarui Liang[2], Li Chen[2], Qiuyu Zhao[1], Yan Wang[3], Yuan Fang[4], Min Hu[2]

[1]School of Medicine, Yunnan University, Kunming, China; [2]Department of Children's Ophthalmology, Affiliated Hospital of Yunnan University, Kunming, China; [3]Department of Children's Ophthalmology, The Fourth Affiliated Hospital, Kunming Medical University, Kunming, China; [4]College of Clinical Medicine, Dali University, Dali, China

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[#]These authors contributed equally to this work.

Correspondence to: Min Hu, PhD. Department of Children's Ophthalmology, Affiliated Hospital of Yunnan University, 176 Qingnian Road, Kunming 650000, China. Email: fudanhumin123@sina.com.

Background: Reasonable personalized surgical design can achieve good treatment results for Helveston syndrome in one surgery, reducing the psychological and economic burden on patients. This article aims to explore the clinical characteristics of Helveston syndrome and the clinical effectiveness and feasibility of individualized surgical design.

Methods: In this retrospective case series study, 28 patients who underwent strabismus correction for Helveston syndrome at the Affiliated Hospital of Yunnan University from June 2018 to December 2020 with complete follow-up data were enrolled. Preoperatively, all patients received standard assessment of vision, intraocular pressure, slit lamp, fundus and refractive status, excluding other eye diseases, as well as detailed special examination of strabismus. These patients were divided into two groups according to the surgical modality: the horizontal muscle surgery alone group and the horizontal muscle surgery combined with superior oblique muscle surgery (combined surgery) group. We used SPSS software for data analysis and compared the postoperative eye position, eye movement, success rate, and reoperation rate between these two groups. Clinical measurement data were compared and analyzed with Fisher's exact test for count data, the *t*-test for normally distributed measurement data, and the Mann-Whitney *U* test for non-normally distributed measurement data. P<0.05 was considered statistically significant.

Results: This study included a total of 28 patients with Helveston syndrome, including 20 males and 8 females. The average age at the time of surgery is 12.04 ± 8.67 years (range, 4–43 years). The postoperative A-pattern degree was significantly greater in the group undergoing horizontal muscle surgery alone [6.23±1.31 prism diopters (PD); range, 0–10 PD] than in the group undergoing combined surgery (0.53±0.32 PD; range, 0–4 PD; P=0.002). Superior oblique muscle overactivity was significantly reduced in the combined surgery group (0.20±0.11+; range, 0–1+) compared to the horizontal muscle surgery alone group (1.31±0.26+; range, 0–2+; P=0.002). However, there was no significant difference in success rate or reoperation rate between the two groups. Additionally, after combining the recession of the superior rectus muscle with the horizontal muscle, the number of A-pattern degrees was greatly reduced.

Conclusions: Helveston syndrome can be improved using a personalized surgical design according to the degree of external strabismus A-pattern, superior oblique muscle overaction, and dissociated vertical deviation (DVD) degree, which improves the success rate of single surgery.

Keywords: Strabismus; personalized surgery; superior oblique overaction; A-pattern; dissociated vertical deviation (DVD)

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Introduction

The concept of Helveston syndrome was first introduced by Helveston in 1969, with symptoms including A-pattern exotropia, dissociated vertical deviation (DVD), and superior oblique muscle overaction, which is less common (1). At present, the etiology of Helveston syndrome is not clear, and typical cases are easily diagnosed, but atypical cases are difficult to diagnose and can easily lead to missed diagnosis. In addition, due to Helveston syndrome involving abnormal eye movements in three directions: horizontal, vertical, and rotational, the design of surgical plans is more difficult. At the same time, most patients with this disease have abnormal appearance, significant visual impairment, and a heavy psychological burden. Choosing appropriate surgical methods to improve patient satisfaction and reduce their psychological and economic burden is particularly important. The goals of surgery include coordinating the extraocular muscles in both eyes, reestablishing the eyes' normal retinal correspondence, and establishing anatomical conditions that will allow for the regeneration of higher visual functions. Helveston syndrome is usually designed according to the specific situation of the patient, but because the disease is relatively rare. There is very little known information about the clinical characteristics and treatment of Helveston syndrome. Therefore, there are various surgical methods for Helveston syndrome, but there is still controversy over whether the surgery should be performed in one go or in stages. In recent years, some researchers have used different surgical methods to treat Helveston syndrome and compared them. Many methods have shown good efficacy, and there is currently no fixed and accurate surgical method.

To summarize the clinical characteristics of Helveston syndrome and investigate efficient surgical treatment options, we retrospectively evaluated the data of 28 patients with Helveston syndrome who were hospitalized in our ophthalmology department. This article reviews and analyzes the data of 28 patients with Helveston syndrome admitted to our ophthalmology department. We found that as long as personalized surgical plans are designed based on the clinical characteristics of the patients, a single surgery can achieve good results, and there are also various complex or simple surgical methods. There is no fixed surgical method that requires rote memorization. We hope to provide some reference opinions for colleagues in formulating surgical plans for Helveston syndrome. We present this article in accordance with the STROBE reporting checklist (available at https://tp.amegroups.com/article/view/10.21037/tp-23-334/rc).

Highlight box

Key findings

- Helveston syndrome can be customized to increase the likelihood that a single surgery will be successful and that positive outcomes can be obtained with or without coupled superior oblique muscle reduction.

What is known and what is new?

- It is controversial whether Helveston syndrome should be operated on once or several times.
- There was no significant difference in the success and secondary operation rates between the horizontal muscle surgery alone and the horizontal muscle surgery combined with the superior oblique muscle surgery. However, there was a difference in the degree of A-pattern and the degree of hyperfunction of the superior oblique muscle postoperatively.

What is the implication, and what should change now?

- There is no specific template for the surgical design of Helveston syndrome. Personalized surgical design can be carried out according to the degree of exotropia A syndrome, superior oblique hyperfunction, and DVD to improve the success rate of one-time surgery.

Methods

This study was conducted in accordance with the Declaration of Helsinki (revised in 2013). This study has been approved by the Ethics Committee of the Affiliated Hospital of Yunnan University (No. 2019098), and the individual consent to this retrospective analysis has been waived.

Object

This is a retrospective case series study. We retrospectively summarize the surgical data of 28 patients with Helveston

syndrome who were seen, operated on, and followed for at least 1 year following surgery at the pediatric ophthalmology department of the Affiliated Hospital of Yunnan University from June 2018 to December 2020.

Study methods

Inspection method

Preoperatively, all patients received standard evaluations of visual acuity, intraocular pressure, slit lamp, fundus examination, and refractive status to rule out the possibility of additional eye illnesses. Additionally, a strabismus expert examination covering the following components was also performed. (I) Strabismus examination: the objective squint angle of near and distant, including horizontal squint angle and vertical squint angle, was determined using the prism alternating covering method. (II) A/V-pattern measurement: the patient was requested to look directly ahead at a 33 cm point light source, and the objective squint angle of 25° up and 25° down was measured horizontally. (III) DVD measurement: using the prism alternating occlusion method, the objective vertical squint angle of near and distant viewing was measured to determine the degree of upper squint. (IV) Measuring the degree of overactivity of the superior oblique muscle: the patient observes a 33 cm point light source while rotating horizontally; if the internal transposition was less than the exterior transposition, the light spot in the eye was examined. Simultaneously, the Titmus chart was used to evaluate stereopsis, and passive traction tests were applied to identify any limiting issues.

Diagnostic criteria

(I) A-pattern exotropia: the difference between upward 25° and downward 25° gaze oblique angle ≥10 prism diopters (PD). (II) Superior oblique muscle function: superior oblique muscle function eye position; the inward turning eye is lower than the outward turning eye for superior oblique muscle overaction; ≤5° is 1+, ≤10° is 2+, ≤15° is 3+, and >15° is 4+. (III) DVD: the degree of DVD was determined based on the original in-situ vertical strabismus, and the degree of separated vertical deviation was categorized as 1+ for less than 5 PD, 2+ for 5–10 PD, 3+ for 10–15 PD, and 4+ for more than 15 PD. Normal postoperative eye position was defined as strabismus ≤±10 PD; internal strabismus was defined as strabismus >+10 PD; and exterior strabismus was defined as strabismus >−10 PD. Normal eye position at the final follow-up, absence of evident A-pattern and DVD, absence of head position abnormality, diplopia, dyslexia, and secondary operation for the entire follow-up period signified a successful operation.

Surgery and follow-up

Two surgical procedures were performed: horizontal muscle surgery alone and horizontal muscle surgery combined with superior oblique muscle surgery. The precise surgical procedure depended primarily on the patient's A-pattern and the degree of superior oblique muscle overactivity. When the A-pattern is more severe and the degree of bilateral superior oblique muscle overaction is greater than++, the superior oblique muscle tendon-breaking operation or superior oblique muscle-lengthening operation should be considered. On the other hand, when the A-pattern is less severe and the degree of superior oblique muscle overaction is greater than++, the lateral rectus should be retracted and transposed downward.

Monocular or bilateral superior rectus posterior migration should be performed in equal or unequal amounts based on the degree of bilateral DVD in patients with a high degree of DVD that impairs the appearance and is accompanied by superior rectus overaction but minor superior oblique muscle overaction. If both the A-pattern and DVD degrees were minor and the superior oblique muscle overaction was not apparent, only horizontal muscle surgery, including internal rectus shortening and external rectus regression, was undertaken. The surgical plans for the 28 patients were determined using the aforementioned guidelines. All of the surgeries were designed and completed by a surgeon according to the same standards. All procedures were evaluated at 1, 3, and 6 months postoperatively, and every 6 months thereafter. At each appointment, the eye was inspected using the same techniques as those applied preoperatively.

According to the surgical procedure, the patients were separated into two groups: the horizontal muscle surgery alone group and the horizontal muscle surgery combined with superior oblique muscle surgery (combined surgery) group. The postoperative eye position, eye motions, success rate, and reoperation rate of these two groups were compared.

Statistical analysis

SPSS software (version 22.0; IBM, Chicago, IL, USA) was utilized for the statistical analysis of the data. Clinical measurement data were compared and analyzed with Fisher's exact test for count data, the *t*-test for normally

Table 1 Comparison of postoperative parameters

Factors	Horizontal muscle surgery alone	Combined surgery†	P
Number of cases	13 (46.4)	15 (53.6)	
No. of operated muscles	3.15±0.19	4.80±0.18	0.000‡
Combined with superior rectus recession	4 (30.8)	2 (13.3)	0.372§
Mean follow-up (months)	24.31±1.02	27.33±1.45	0.109¶
Angle of deviation (PD)			
Near	−4.77±2.41	−0.07±1.12	0.047‡
Distance	−2.23±2.14	2.33±1.08	0.170‡
A-pattern amount	6.23±1.31	0.53±0.32	0.002‡
SOOA (+)	1.31±0.26	0.20±0.11	0.002‡
Success	12 (92.3)	15 (100.0)	0.087§
Reoperation	1 (7.7)	0 (0.0)	0.464§

Data are listed as n (%) and mean ± standard deviation. †, horizontal muscle surgery with superior oblique weakening surgery; ‡, Mann-Whitney U test; §, Fisher's exact test; ¶, t-test. PD, prism diopters; SOOA, superior oblique overaction.

distributed measurement data, and the Mann-Whitney *U* test for non-normally distributed measurement data. P<0.05 was considered statistically significant (*Table 1*).

Results

A total of 28 participants were included in this study, including 20 (71.4%) males and 8 (28.6%) females. The mean age of these patients at the time of surgery was 12.04±8.67 years (range, 4–43 years). There were 18 (64.3%) children (<14 years), 4 (14.3%) adolescents (14–18 years), and 6 (21.4%) adults (>18 years). Furthermore, there were eight eyes in six cases of amblyopia. The best-corrected visual acuity before surgery was as follows: one patient (adult) had a corrected visual acuity of 0.4 in the right eye and 0.9 in the left eye, and the remaining 27 patients (54 eyes) all had a corrected visual acuity ≥0.5. Preoperatively, the mean value of the near horizontal strabismus in all patients was −84.11±28.77 PD (range, −20 to −130 PD), and the mean value of distance horizontal strabismus was −75.54±24.50 PD (range, −30 to −100 PD). Also, the mean DVD degree was 2.07±1.22+ (range, 2 to 15 PD, 1+ to 4+), with 16 cases (57.1%) of bilateral asymmetric DVD; the mean A-pattern degree was 18.57±15.50 PD (range, 10–70 PD); and the mean degree of superior oblique overaction was 1.86±5.23+ (range, 1+ to 3+), among which 7 patients (25%) had bilateral asymmetric superior oblique overaction (*Table 2*).

The eye position was checked 1 day after the operation. In the horizontal muscle surgery alone group, one patient had exophoria, and the distance and near strabismus angles of another patient were assessed using the alternate occlusion method (−30 and −20 PD, respectively). The corneal reflection of the remaining 26 patients was in the center. One month postoperatively, the horizontal strabismus was re-examined as before. During alternate masking, some patients exhibited a slight movement of both eyes from the external superior to the median, and the majority of patients demonstrated a loss of superior oblique overactivity and A-pattern. A comparison of the nine positions of one patient before and after the operation is shown in *Figure 1*.

In *Table 1*, a comparison of postoperative examinations between the two groups is presented, with 13 cases in the group undergoing horizontal muscle surgery alone and 15 cases in the group undergoing combined surgery. The mean follow-up time in the horizontal muscle surgery alone group was 24.31±1.02 months (range, 20–32 months), which was 27.33±1.45 months (range, 20–36 months) in the combined surgery group (P=0.109). At the most recent follow-up, the mean horizontal strabismus in the horizontal muscle surgery alone group was −4.77±2.41 PD (range, −30 to 0 PD) at a near distance and −2.23±2.14 PD (range, −25 to 5 PD) at a distance. In the combined surgery group, the mean near horizontal strabismus was −0.07±1.12 PD (range, 10–10 PD) and the mean distant horizontal strabismus

Table 2 Preoperative clinical features

Variables	Value
Number of patients	28
Gender	
Male	20 (71.4)
Female	8 (28.6)
Age (years)	12.04±8.67 [4–43]
Follow-up time (months)	25.93±4.97 [20–37]
Mean refraction (diopters)[†]	
OD	–0.50±1.97[‡] [–5.875 to 2.375]
OS	–0.60±2.19 [–6.375 to 4.000]
Amblyopia	
Unilateral	4 (14.3)
Bilateral	3 (10.7)
Exodeviation (PD)	
Near	–84.11±28.77 [–130 to –20]
Distance	–75.54±24.51 [–100 to –30]
A-pattern amount	18.57±15.80 [10 to 70]
Degree of SOOA (+)	
OD	1.86±0.53 [1–3]
OS	1.71±0.54 [1–3]
Asymmetrical	8 (28.6)
Symmetrical	20 (71.4)
Degree of DVD (+)	2.07±1.22 [1–4]
Asymmetrical	16 (57.1)
Symmetrical	12 (42.9)

Data are listed as n (%) and mean ± standard deviation [range]. [†], refraction is presented as spherical equivalent; [‡], negative value indicates myopia. OD, oculus dexter; OS, oculus sinister; PD, prism diopters; SOOA, superior oblique overaction; DVD, dissociated vertical deviation.

was 2.23±1.08 PD (range, 0–10 PD) at the last follow-up. Postoperatively, the horizontal strabismus prism degree at a near distance was greater in the group undergoing horizontal muscle surgery alone (–4.77±2.41 PD) than that in the combined surgery group (–0.07±1.12 PD; P=0.047). However, there is no difference in the horizontal strabismus prism degree at a distance between the groups (P=0.170).

Moreover, the postoperative A-pattern degree was significantly greater in the group undergoing horizontal muscle surgery alone (6.23±1.31 PD; range, 0–10 PD) than in the group undergoing combined surgery (0.53±0.32 PD; range, 0–4 PD; P=0.002). Superior oblique muscle overactivity was significantly reduced in the combined surgery group (0.20±0.11+; range, 0–1+) compared to the horizontal muscle surgery alone group (1.31±0.26+; range, 0–2+; P=0.002).

In the horizontal muscle surgery alone and combined surgery groups, superior rectus recession was performed in 4 (14.3%) and 2 (7.1%) patients, respectively, and there was no statistically significant difference between the two groups (P=0.372) in terms of the probability of combined superior rectus recession. Patients with superior rectus recession in the horizontal muscle surgery alone group had a significantly lower mean postoperative A-pattern than those who did not undergo superior rectus recession (P=0.002); however, there was no significant difference in the mean A-pattern with or without superior rectus recession in the combined surgery group (P=0.466).

A total of 10 (76.9%) and 15 (100%) patients were successful in the horizontal muscle surgery alone and combined surgery groups, respectively, with no significant difference between the two groups' success rates (P=0.084). During the follow-up period, one patient (7.7%) in the horizontal muscle surgery alone group and no patients in the combined surgery group underwent secondary surgery, and there was no statistically significant difference between the two groups (P=0.464).

Discussion

Helveston syndrome is a triadic syndrome characterized clinically by A-pattern exotropia, DVD, and superior oblique hyperfunction. A-pattern exotropia is caused by the excessive action of the superior oblique muscle when the eyeball is turned inward and downward, resulting in excessive external rotation. Helveston believed that the hyperfunction of the superior oblique muscle is primary and unrelated to DVD, so the cause of DVD is unknown (1). However, an overactive superior oblique muscle is overactive can cause a patient's eye to turn downward, which diminishes the upward drift caused by DVD, resulting in diagnostic difficulties and a high likelihood of a missed diagnosis (1).

Since Helveston syndrome is characterized by abnormal eye movements in three directions—horizontal, vertical, and rotational—the design of surgical protocols and the timing of surgery have been controversial. In 1991, McCall *et al.* (2)

Figure 1 Photos of the patient's eyeball positions in nine directions before and after surgery. (A) Eyeball position pictures of a patient in nine directions before surgery. (B) Eyeball position pictures of the patient in nine directions after surgery. These images are published with the patient/participant's consent.

proposed for the first time the treatment of superior oblique hyperopia and DVD by superior oblique and superior rectus muscle reduction, followed by secondary surgery to correct horizontal strabismus. Velez *et al.* (3) also reported that bilateral superior rectus regression could correct small degrees of exotropia A-pattern; however, a combined superior oblique muscle reduction is required when the exotropia is severe. They believed that staged surgery is highly feasible but suggested that horizontal muscle surgery combined with superior oblique surgery to correct the external oblique A-pattern should be performed first, and the need for secondary surgery should be determined based on the extent of the DVD after the initial surgery. The primary considerations are as follows: (I) the number of muscle strips involved in the staged surgery is small, and the risk of postoperative anterior segment ischemia syndrome is low; (II) the DVD is reduced or disappears in some cases following superior oblique muscle surgery and horizontal strabismus correction, and can also improve with age.

Some scholars argue that, within the appropriate range of strabismus, a single surgery can be designed to correct both the horizontal and vertical strabismus, thereby avoiding additional surgery (4). From the experimental results of this study, it is evident that there are no significant differences in the success and reoperation rate between the horizontal muscle surgery alone group and the combined surgery group, indicating that there is no specific and unique template for the surgical treatment of Helveston syndrome. The design should be based on the A-pattern, the degree of superior oblique muscle overaction, the DVD, and the degree of fundus rotation. Different surgical approaches should be employed, and appropriate surgical methods should be chosen to increase patient satisfaction and decrease the psychological and financial burdens of patients (5). The present study aimed to discuss the individualized surgical approach according to the degree of

overaction of the superior oblique muscle and the degree of DVD by reviewing the medical records of Helveston syndrome patients who have been seen and operated on at our institution in recent years.

Correction of the superior oblique overaction and A-pattern

It is generally believed that overaction of the superior oblique muscle is the main cause of the A-pattern. The superior oblique muscle is responsible for incyclo, external rotation, and downward rotation. An overactive superior oblique muscle can cause increased external rotation when the eye is gazing downward, manifesting as the A-pattern. Grading of superior oblique muscle overaction: corneal reflection is lower in internal transposition compared to external transposition; 5° is +, 10° is ++, 15° is +++, and >15° is ++++. The A-pattern is usually corrected by superior oblique muscle reduction, and the data from this study also showed that superior oblique muscle reduction is very effective at correcting the A-pattern. Superior oblique tendonectomy, superior oblique lengthening, and superior oblique intrathecal tendonectomy are typical superior oblique decompression procedures.

Superior oblique tendon dissection

This procedure is simple to perform, highly stable, and has a low risk of recurrence; consequently, it is the most frequently used clinically. However, the superior oblique tendon dissection cannot be quantified, and the risk of postoperative complications involving superior oblique muscle paralysis is high. Therefore, the surgical indications for superior oblique tendonotomy must be strictly regulated, and this procedure should only be performed when there is clear incyclo of the eye, an A-pattern, and obvious overactivity of the superior oblique muscle. To avoid the postoperative loss of stereopsis or rotational diplopia, patients with stereopsis should undergo this procedure with caution.

Superior oblique muscle lengthening

Multiple researchers have confirmed that lengthening the superior oblique muscle can significantly correct its overactivity. Pollard *et al.* stated that silicone tendon lengthening was utilized to correct the A-pattern (6). However, silicone, as a foreign body, can result in a more severe inflammatory response and an increased risk of adhesion syndrome (7). Bardorf *et al.* (8) found that Z-lengthening of the superior oblique muscle significantly improved the superior oblique hyperfunction and decreased the degree of external oblique A-pattern and DVD in 19 patients with Helveston syndrome.

Intrathecal tendon dissection of superior oblique muscle

Sun *et al.* (9) weakened the superior oblique muscle via intrathecal tendon dissection of the superior oblique muscle. They made a 6–8 mm incision along the long axis of the muscle sheath to expose and cut the tendon fibers. This surgical approach method preserves the integrity of the deep muscle septum, so that the tether between the superior rectus and the superior oblique continues to have a restrictive effect, thereby avoiding peripheral adhesion. This allows for a more accurate prediction of the surgical effect, a complete reduction of the superior oblique's overactivity, and the prevention of superior oblique paralysis. The nasal superior oblique tendon rupture of the superior rectus is superior to the temporal tendon rupture, and the closer the tendon rupture is to the trochlear, the more beneficial the effect. However, the more severe the A-pattern prior to surgery, the more obvious the tendon rupture.

In addition, superior oblique muscle weakening carries the risk of secondary paralysis of the superior oblique muscle, and some surgeons have successfully used horizontal rectus muscle displacement to correct the A sign. However, there is limited pertinent data and further confirmation is required (10).

DVD correction

DVD, also referred to as alternating upward strabismus, is a condition in which both eyes are alternately covered, with the covered eye being upwardly oblique and more noticeable from a distance. Its cause is currently unknown. Recent studies support the hypothesis that the subcortical center of both eyes, which controls vertical convergent and divergent movements, is alternately and intermittently stimulated as the cause of DVD; however, the cause of the abnormal stimulation of vertical separation remains unknown. Some have hypothesized, based on the external rotation movement of the upward-turning eye and the internal rotation movement of the gazing eye, that the vertical separation movement is coordinated primarily by the oblique muscles, but they cannot explain the initial position and external rotation position of the upward strabismus.

According to the degree of superior obliquity grading:

≤5° as +, 6–10° as ++, 11–15° as ++++, ≥16° as ++++, quantitative surgery: no surgery below 1+, 2+ to 4+ respectively posterior migration of superior rectus muscle 4, 6, and 8 mm.

DVDs are often bilateral, and some operators recommend equal or unequal posterior migration of the superior rectus muscle in both eyes, depending on the degree of separation and strabismus. Even if there is no significant upward obliquity in one eye, a minimal amount of posterior migration of the superior rectus muscle needs to be given according to an unequal surgical design. If only a significant upward oblique eye is operated on, it is likely to cause postoperative upward strabismus in the contralateral eye and a reversed compensatory head position.

In recent years, some scholars have operated on four oblique muscles to correct the A-pattern and DVD. The A-pattern was corrected by breaking the upper oblique muscle tendon of both eyes, DVD was corrected via anterior transposition of the lower oblique muscle of both eyes to the end of the lower rectus, and horizontal muscle surgery was performed to correct horizontal strabismus. The theory of overactive oblique muscle serves as its theoretical foundation. Recessive nystagmus is believed to frequently accompany Helveston syndrome. To control nystagmus and improve the visual acuity of the fixation eye, the superior oblique muscle of this eye may become overactive, causing eyeball incyclo. At the same time, in order to balance the overactivity of the superior oblique muscle, the inferior oblique muscle produces a secondary deviation caused by excessive innervation. Hering's law states that excessive innervation of the ipsilateral inferior oblique muscle will result in excessive contralateral superior rectus muscle activity, which will cause DVD, especially when the eyeball is turned outward. This surgical method avoids the risk of anterior segment ischemia that may result from a single-eye triple rectus surgery (11). Fu *et al.* (12) also carried out a retrospective study on bilateral isolated DVD cases, which were divided into two groups: a bilateral superior rectus recession group and an inferior oblique anterior transposition group. The eye movement, eye position, DVD volume, and complications of the two groups were compared before and after surgery. The study found that both surgical methods can effectively treat isolated DVD without inferior oblique overactivity. However, some academics have questioned whether the anterior transposition of the inferior oblique muscle weakens the function of the inferior oblique muscle and may exacerbate the incyclo and A-pattern. According to this deduction, it is also appropriate for adjusting the V-pattern. Therefore, this concept does not comply with the accepted strabismus correction rules (13), and this surgical method requires additional verification.

Moreover, in some cases, DVD is reduced or eliminated after superior oblique muscle weakening and horizontal strabismus correction and can also improve with age. Consequently, the DVD of some patients can be evaluated for the necessity of fractional surgery.

Horizontal muscle correction

Given that the superior oblique muscle has an excyclo effect, the effect of the superior oblique muscle on the correction of horizontal strabismus should be considered when performing bilateral superior oblique muscle reduction. In one study, bilateral superior oblique muscle weakness shifted the horizontal muscle inward by 9.5 PD (14), while in another study, the horizontal muscle was shifted inward by 7.7 (8.5) PD (15). External strabismus should therefore be appropriately undercorrected when performing bilateral superior oblique weakening. Moreover, surgery on three rectus muscles in one eye will be required when the external strabismus triad must be corrected at the same time, which will increase the risk of ischemia in the anterior segment of the eye, and the internal rectus muscle can be folded instead of shortened (16). The risk of anterior segment ischemia is drastically reduced, and the inflammatory response is mild.

The following is a summary of our individualized surgical plan for Helveston syndrome.

DVD correction

When the DVD is between 5° and 10°, the superior rectus muscle retreats 5–6 mm; when the DVD is between 10° and 15°, the superior rectus muscle retreats 6–7 mm; and when the DVD is greater than 15°, the superior rectus muscle retreats by a maximum of 8 mm. Asymmetric superior rectus retraction is feasible when the degree of DVD differs between the two eyes, regardless of head position. Moreover, the DVD is not corrected temporarily when the DVD in both eyes is less than 5°.

Correction of superior oblique muscle overaction

When the superior oblique muscle function is mildly overactive less than 5° (+), the superior oblique muscle can be temporarily not reduced; when the superior oblique muscle is moderately overactive (5–10°, ++ to +++), superior oblique muscle lengthening should be performed; and

when the superior oblique muscle is severely overactive (>10°, ++++ to ++++), superior oblique muscle tendon severing should be performed. If preoperative binocular vision is present, severing the superior oblique tendon is contraindicated.

Horizontal muscle correction

When DVD, superior oblique hyperopia, and exotropia need to be corrected simultaneously, if the horizontal strabismus is less than 40–50 prism degrees, excluding the convergence insufficiency esotropia, it is feasible to correct the exotropia by receding the external rectus muscle in both eyes, to avoid the risk of ischemia of the preoptic segment induced by the surgery of three rectus muscles; when the external obliquity is greater than 50 prismatic degrees, involving monocular triple rectus surgery, requiring internal rectus shortening as well as external rectus regression, internal rectus folding (instead of shortening) is feasible to reduce the risk of ischemia in the anterior segment of the eye. When the DVD is less than 5° and the superior oblique muscle is mildly overactive (+), correction of horizontal strabismus is sufficient. In cases where bilateral superior oblique muscle reduction and horizontal muscle surgery are performed simultaneously to correct exotropia, the amount of horizontal strabismus surgery is recommended to undercorrect by 10–15 prism degrees.

Given that Helveston syndrome has an early onset, a long duration, and the coexistence of multiple strabismuses, it has a greater impact on the patient's visual function, and fewer individuals have normal visual function prior to surgery, so early surgical treatment is recommended. Surgery can make the visual axes of both eyes parallel in all directions and coordinate the movement of both eyes in each gaze's eye position, which promotes the development of visual acuity, binocular monovision, and binocular stereovision. We determined the surgical approach based on the degrees of superior oblique muscle overaction and DVD and established a customized surgical design. Up to now, we have successfully completed nearly 100 cases of corrective surgery for Helveston syndrome. This reasonable individualized surgical design avoids the risk of anterior segment ischemia and reduces some complications that may result from the surgery, even though the summary of this procedure is based solely on the clinical experience of the surgeon and needs to be confirmed in a large sample of cases, and long-term follow-up on eye position, vision, and optical coherence tomography angiography (OCTA). In addition, this study is a retrospective analysis that may lead to some bias in the results. This requires further confirmation from multicenter clinical trials.

Conclusions

Treatment outcomes for Helveston syndrome can be enhanced using a personalized surgical design according to the degree of external strabismus A-pattern, superior oblique muscle overaction, and the DVD degree to improve the success rate of a single operation. Successful results can be achieved with or without combined superior oblique muscle reduction.

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work, including ensuring that any questions related to the accuracy or integrity of any part of the work have been appropriately investigated and resolved. This study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). This study has been approved by the Ethics Committee of the Affiliated Hospital of Yunnan University (No. 2019098), and the individual consent to this retrospective analysis has been waived.

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