

# Surgical management of Helveston syndrome (triad of A- pattern exotropia, superior oblique overaction and dissociated vertical deviation) using 'Four Oblique' procedure

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**Purpose:** To report the surgical outcomes in six patients of Helveston syndrome using a "four oblique" procedure. The popular methods for surgical management include superior rectus recessions alone or combined with superior oblique tenectomy. However, large angle exotropia correction would entail a higher risk of anterior segment ischemia when the superior rectus needs to be operated along with the horizontal recti. Hence, we evaluated the long-term results of this uncommon procedure. **Methods:** This was a retrospective review of six patients diagnosed to have manifest dissociated vertical deviation (DVD) with A pattern exotropia with bilateral superior oblique over action. All patients underwent horizontal muscle recessions/resections for exotropia along with bilateral posterior tenectomy of the superior oblique with inferior oblique anterior transpositioning. **Results:** The median age was 10 years (Range 5–26 years). The mean postoperative follow-up was 26 ± 14.02 months (Range 12–48 months). The mean reduction in exotropia was from 36.5 ± 21.06 PD (Range 15–65 PD) to 6.1 ± 3.06 PD (Range 3–10 PD). The procedure corrected the A pattern from a mean 23 ± 7 PD (Range 15–35 PD) to 7.6 ± 3.2 PD (Range 3–10 PD). The average DVD in the right eye reduced from 14 ± 4.3 PD (Range 8–20 PD) to 5.3 ± 1.2 PD and in the left eye from 14.33 ± 3.6 PD (Range 10–18 PD) to 4.1 ± 1.1 PD. The DVD asymmetry reduced from 6.33 ± 3.4 PD to 1.5 ± 1.3 PD. **Conclusion:** "Four oblique" procedure with horizontal muscle surgery seems to be an effective method for significantly correcting the A pattern as well as reducing the DVD with good long-term outcome in our case series.

**Key words:** A-pattern exotropia, dissociated vertical deviation, inferior oblique anteropositioning, superior oblique over action, superior oblique tenectomy

Dissociated vertical deviation (DVD) is a characteristic motility disorder that manifests itself as elevation, abduction, and extorsion of the non-fixating eye. It is estimated that 40% of the patients with DVD may have some oblique over action.<sup>[1]</sup> The triad of A pattern exotropia, bilateral superior oblique over action with DVD was first described by Eugene Helveston in 1969 and is also called the "Helveston Syndrome."<sup>[2]</sup> There is sparse literature regarding the surgical management of this strabismus complex.<sup>[3-7]</sup>

The hypertropia caused by the DVD and the associated superior oblique over action pose a real challenge to the management of this condition. Multiple surgical procedures have been proposed including bilateral superior rectus (SR) recessions alone and bilateral SR recession combined with superior oblique (SO) weakening. Bilateral SR recessions tend to be successful in management of small A pattern with DVD.<sup>[5]</sup> Bilateral SR recessions with superior oblique weakening tend to be effective in incomitant DVD with DVD being greatest in abduction, as well as moderate "A" pattern ranging from 12 to 20 prism dioptres (PD).<sup>[3-5]</sup> Simultaneous weakening of the

SO and SR may convert the existing A pattern into a V pattern by inversion of vertical incomitance.<sup>[8]</sup> In addition, when the exotropia tends to be large or in cases of reoperations, there is a risk of anterior segment ischemia when the superior rectus needs to be operated along with the horizontal recti.

Gamio reported the four oblique muscles weakening procedure for A pattern strabismus with DVD in 9 patients from the hypothesis of the cyclovertical muscles being responsible for DVD.<sup>[7]</sup> Velez *et al.* have shown good outcomes of the procedure in their series of 14 patients.<sup>[5]</sup> We report the long-term results of the four oblique muscles weakening procedure in six patients with a mean follow-up of 2 years.

## Methods

We conducted a retrospective review of six consecutive patients presenting with Helveston syndrome, all of whom underwent surgery for A pattern exotropia associated with DVD and superior oblique overaction (SOOA). This study protocol was

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reviewed and approved by the institutional review board of the hospital in compliance with the Declaration of Helsinki. Patients with severe unilateral amblyopia, previous oblique muscle surgery or vertical muscle surgery, and follow-up less than 12 months were excluded from the study.

Every patient underwent visual acuity recording, cycloplegic refraction, anterior segment examination, and fundus evaluation. An orthoptic work up comprised of recording binocular single vision, stereopsis, and alternate cover test. Prism cover test was done in primary position (distance and near) and also in up gaze, down gaze and lateral gazes. On the basis of the difference in measurements in up gaze and down gaze the amount of "A" pattern was measured in prism dioptres. DVD was measured separately in each eye using the prism under cover test where base down prisms were placed under cover to neutralize the vertical movement after neutralizing the horizontal deviation using base in prisms. The amount of DVD was measured in primary position. DVD was considered to be asymmetric when the difference between the two eyes was greater than 7PD.<sup>[9]</sup> All the patients included had a superior oblique over action and none of the patients had reduced elevation in adduction.

All the patients underwent horizontal muscle surgery depending on the amount of exotropia along with bilateral posterior tenectomy of the superior oblique and inferior oblique anterior transpositioning [Table 1].

All the patients were operated using the fornix incision. The inferior oblique was approached through a fornix incision

8 mm from the limbus in the inferotemporal quadrant. The inferior oblique was hooked after identifying the Park's triangle to ensure that all the fibers were completely isolated. After disinserting it close to the insertion, the inferior oblique muscle was secured with 6-0 polyglactin suture. The inferior rectus was then hooked and isolated. The inferior oblique was sutured just adjacent to the inferior rectus at the same level.

The superior oblique was approached through a superotemporal fornix incision. After hooking the superior rectus, the insertion of the superior oblique was identified. A thin spatula (iris reposer) was used to carefully separate all the fibers from the sclera and was then hooked. The posterior 7/8<sup>th</sup> fibers were isolated by splitting the tendon longitudinally and 10 mm block from the insertion till the temporal border of superior rectus waste nectomized from the insertion. There was no frenulum lysis done.

The horizontal muscles were recessed or resected the conventional way using 6-0 polyglactin sutures.

The patients were followed up on day 1, day 10, after 1 month, and thereafter 4 monthly as deemed necessary by the examining ophthalmologist. The measurements obtained at the last follow-up were used to compare the results with the preoperative measurements [Figs. 1-4].

### Results

The median age of the cohort was 10 years (Range 5-26 years). The mean postoperative follow-up was 26 ± 14.02 months

**Table 1: The preoperative characteristics of patients including age, visual acuity, average preoperative deviation in the primary position and the surgical plan for each patient**

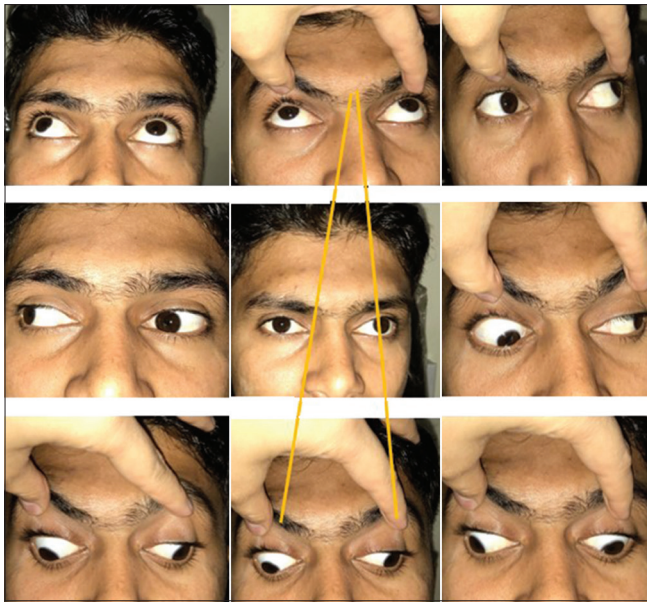
	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age (years)	26	8	8	12	16	5
Vision OD	6/9	6/24	6/12	6/18	6/12	6/9
OS	6/18	6/9	6/9	6/18	6/12	6/9
Horizontal deviation	15 PD XT	57 PD XT	40 PD XT	65 PD XT	17 PD XT	25 PD XT
A pattern	18 PD	25 PD	15 PD	25 PD	20 PD	35 PD
DVD OD	18 PD	12 PD	14 PD	12 PD	20 PD	8 PD
OS	10 PD	14 PD	18 PD	16 PD	10 PD	18 PD
Surgical procedure	BE LR recess 3.5 mm, BE PTSO, BE IOAT	RE 7 mm LR recess with BE 5 mm MR resect, BE PTSO, BE IOAT	BE LR recess 8.5 mm, BE PTSO, BE IOAT	RE 8 mm LR recess with 5.5 mm MR resect with LE LR recess 5.5 mm, BE PTSO, BE IOAT	BE LR recess 4 mm, BE PTSO, BE IOAT	BE LR recess 5.5 mm, BE PTSO, BE IOAT

DVD=Dissociated vertical deviation, PD=Prism dioptres, XT=Exotropia LR=Lateral rectus, MR=Medial rectus, RE=Right eye, LE=Left eye, BE=Both eyes, PTSO=Posterior tenectomy of the superior oblique, IOAT=Inferior Oblique Anterior Transpositioning

**Table 2: The postoperative follow-up for each patient with the postoperative correction of exotropia, A pattern and DVD**

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age (years)	26	8	8	12	16	5
Vision OD	6/9	6/24	6/12	6/18	6/12	6/9
OS	6/18	6/9	6/9	6/18	6/12	6/9
Follow-up (months)	12	36	48	24	12	24
Horizontal deviation	3 PD XT	9 PD XT	2.5 PD XT	7 PD XT	5.5 PD XT	10 PD XT
A pattern	6 PD	10 PD	3 PD	12 PD	6 PD	9 PD
DVD OD	4 PD	6 PD	6 PD	4 PD	7 PD	5 PD
OS	3 PD	6 PD	4 PD	5 PD	3 PD	4 PD

DVD=Dissociated vertical deviation, PD=Prism dioptres, XT=Exotropia, OD=Right eye, OS=Left eye



**Figure 1:** A preoperative photograph of a patient with large angle exotropia with bilateral superior oblique overaction with A pattern

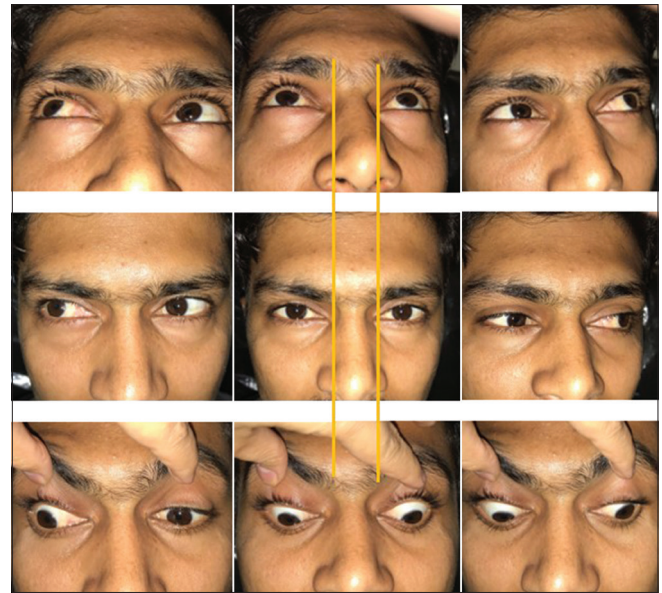


**Figure 2:** Presence of large DVD under the translucent occluder in the same patient

(Range 12–48 months). The mean reduction in exotropia obtained was 30.5 PD, reducing from preoperative mean of  $36.5 \pm 21.06$  PD (Range 15–65 PD) to  $6.1 \pm 3.06$  PD (Range 3–10 PD) postoperatively. The surgical success was defined as primary position correction of exotropia with a residual of less than 10 PD of exotropia or esotropia. All the patients got acceptable correction of exotropia in the primary position [Table 2]. None of the patients underwent any resurgeries. The procedure collapsed the A pattern from a preoperative mean of  $23 \pm 7$  PD (Range 15–35 PD) to  $7.6 \pm 3.2$  PD (Range 3–10 PD) giving an average correction of 16 PD [Fig. 5]. In two patients (patient number 2 and 4), there was a residual A pattern of 10 PD and 12 PD, respectively. The average DVD in the right eye was  $14 \pm 4.3$  PD (Range 8–20 PD) and in the left eye was  $14.33 \pm 3.6$  PD (Range 10–18 PD), and the mean asymmetry was  $6.33 \pm 3.4$  PD. Three patients had asymmetrical DVD with difference between two eyes being 10 PD. They also underwent the same surgical procedure with good outcomes and no residual hypertropia. Postoperatively the average DVD in the right eye was  $5.3 \pm 1.2$  PD (Range 4–7 PD) and that in the left eye was  $4.1 \pm 1.1$  PD (Range 3–6 PD) [Fig. 6], while the mean reduction in asymmetry of DVD was from  $6.33 \pm 3.4$  to  $1.5 \pm 1.3$  prism dioptres [Table 2]. None of the patients developed fusion or stereopsis postoperatively.

## Discussion

The combination of A pattern strabismus with DVD is a complex strabismus where good results can be obtained by choosing



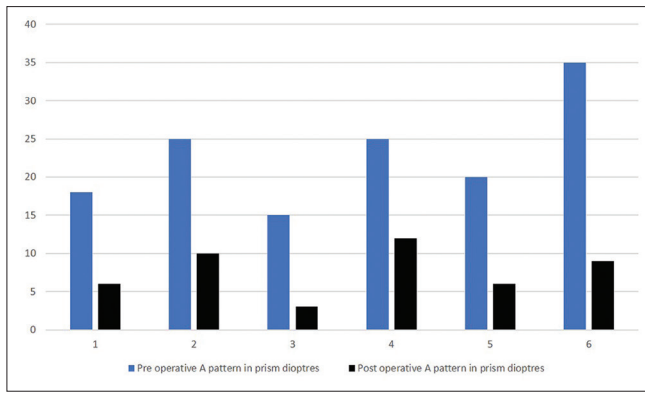
**Figure 3:** A postoperative photograph of the patient depicting the correction of exotropia with collapse of A pattern



**Figure 4:** Good control of DVD under the translucent occluder postoperatively

the right surgical plan. The popular choice for management would be SR recession with SO tenectomy. Small to moderate “A” patterns measuring less than 20 PD may respond well to superior rectus weakening with superior oblique weakening.<sup>[3,5]</sup> However, this procedure of weakening the superior rectus and superior oblique is fraught with the risk of eyelid retraction, vertical incomitance with relative over action of the inferior oblique, and inversion of pattern. Melek *et al.* in their series of three patients reported correction of “A” pattern from 39 to 15 PD using superior rectus weakening with superior oblique tenectomy.<sup>[8]</sup> Postoperatively, all patients had an inversion to a “V” pattern measuring 5–12 PD. When this strabismus complex is associated with a large angle exotropia the eyes are prone to the risk of anterior segment ischemia if a horizontal recess-resect procedure has to be performed along with superior rectus recession. Four oblique muscles weakening procedure markedly reduces this risk by avoiding adjacent rectus muscle surgery and has showed the greatest impact in collapsing the “A” pattern as well as the correction of DVD.

Gamio<sup>[7]</sup> published his results of four oblique muscles weakening procedure on 9 patients of DVD to reduce bilateral cyclotorsion. Of these, 5 patients had A pattern with SOOA and A pattern decreased from  $19.4 \pm 8.8$  PD to  $3 \pm 3.7$  PD. Velez *et al.*<sup>[5]</sup> reported that the “A” pattern collapsed from  $31.8 \pm 5.5$  PD to  $1.7 \pm 2.4$  PD at a follow-up of 1 year with the four oblique weakening procedure while in our series “A” pattern collapsed from  $23 \pm 7$  PD to  $7 \pm 3$  PD using the four oblique weakening



**Figure 5:** A bar diagram depicting the collapse of A pattern in each patient with the X axis depicting the patients and Y axis denoting the A pattern in prism dioptres

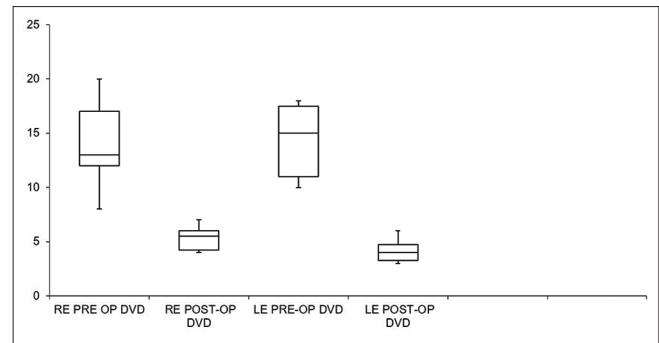
procedure at a mean follow-up of 2 years. Velez *et al.*<sup>[5]</sup> also reported average DVD reduction from  $13.3 \pm 2.4$  PD to  $3.9 \pm 2.5$  PD and reduction in DVD asymmetry from  $4.6 \pm 4.6$  PD to  $3.7 \pm 4.2$  PD. In our series, DVD reduced at an average of  $14 \pm 4.3$  PD in the right eye to  $5.3 \pm 1.2$  PD postoperatively and in the left eye from  $14.33 \pm 3.6$  PD to  $6.3 \pm 3.4$  PD, while the DVD asymmetry dropped from  $6.3 \pm 3.4$  PD to  $1.5 \pm 1.3$  PD at an average follow-up of 2 years.

The exact elucidation of the efficacy of the four oblique muscles weakening procedure is not very clear. Guyton had studied six patients of DVD using scleral search coils.<sup>[10]</sup> He suggested that a vertical vergence movement which was oblique muscle induced occurred with the fixing eye tending to intort and depress and the covered eye extorting and elevating manifesting as DVD. Guyton proposed that a normal oblique muscle produced cyclovergence/vertical vergence in an exaggerated form probably as a learned response, which helped in improving vision in the fixing eye. Enright in 1992 had also suggested a disparity induced vertical vergence in humans associated with binocular torsion and concluded that the oblique muscles in both eyes are largely responsible for vertical vergence movements induced by small vertical disparities.<sup>[11]</sup> The determination of the vertical movement being produced by an oblique muscle or vertical muscle was decided by detection of the torsional movement that occurred simultaneously with the vertical movement. These studies implicate the role of oblique muscles in the etiology of DVD and were a basis for doing the four oblique procedure.

This study has its limitations of being a retrospective review with a small sample size and no control group. Our study did not consider the incomitance of DVD across the horizontal gazes. It could be possible that SR recessions would have given good results in patients with incomitant DVD with DVD being greatest in abduction. However, our experience with four oblique muscles weakening surgery suggests that satisfactory results can be accomplished in this challenging disorder with respect to control of DVD in the primary position along with collapse of the "A" pattern and mitigating the risk of anterior segment ischemia and pattern inversion postoperatively.

## Conclusion

"Four oblique" muscles weakening procedure with horizontal muscle surgery seems to be an effective method for significantly correcting the A pattern exotropia as well as reducing the



**Figure 6:** Box and whisker plot depicting the maximum, minimum, and mean DVD correction of the cohort in prism dioptres in the right eye and left eye. RE- Right eye, LE- Left eye, Pre-op DVD- Pre operative DVD, and Post-op DVD- Post operative DVD

amount of DVD with good long-term outcome. It is definitely indicated in patients having a large angle exotropia with a large A pattern and DVD, in patients with risk of anterior segment ischemia as well as to reduce the chances of inversion.

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## Conflicts of interest

There are no conflicts of interest.

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