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ORIGINAL RESEARCH

Surgical management of congenital superior oblique palsy (CSOP): A retrospective study

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ABSTRACT

Purpose: To study the clinical presentation and surgical outcome of CSOP. **Materials and Methods**: A retrospective analysis of 15 cases of unilateral CSOP in the age group of 7-20 years between 2004-2006 visiting the pediatric ophthalmology department of a tertiary eye care centre in south India was done. **Results**: Patients presented with anomalous head posture (AHP), strabismus and facial asymmetry. Among 15 cases,12 cases underwent one muscle surgery, and 3 cases underwent two muscle surgery. The mean deviation reduced from 15 ± 8 pd preoperatively to 4 ± 2 pd postoperatively which was statistically significant by paired t test. **Conclusion**: Isolated Inferior Oblique (IO) weakening is a safe and effective treatment option for CSOP of up to 20 PD of vertical deviation in primary position and two muscle surgery should be reserved for larger deviations. Good anatomical alignment was obtained in all patients with satisfactory cosmetic results with either total resolution of head posture or only a mild head tilt postoperatively.

Keywords: CSOP, parks 3 step test, ocular torticollis, facial asymmetry

INTRODUCTION

Superior oblique palsy (SOP) is the most frequent form of paralytic vertical strabismus and cranial nerve palsy. It can be unilateral or bilateral and congenital or acquired. Majority of the SOP cases are congenital in origin, even though they presented throughout adulthood.The reason for such delayed presentation is unclear, but it is likely that control of the deviation decompensates over time, possibly exacerbated by factors such as the onset of presbyopia, and the use of downgaze for reading when wearing a presbyopic spectacle Correction (1).

CSOP, browns syndrome and primary inferior oblique overaction (IOOA) are common causes of vertical deviation in childhood. A patient with CSOP can present with ipsilateral hypertropia with contralateral head tilt ,anomalous head posture (AHP), facial asymmetry and rarely amblyopia .No single feature can label a SO palsy as congenital .Characteristic features in the absence of causative factors like trauma and craniosynostosis help diagnose the palsy as congenital .Diagnosis is made based on history of early onset head tilt ,old photographs showing head posture, presence of facial asymmetry , increased vertical fusional amplitudes and absence of diplopia and asthenopia which is a feature of acquired SOP. Normal vertical fusional amplitude is up to 3-4 PD.In an effort to fuse diplopic images, there is a compensatory increase in vertical fusional amplitudes to more than 4 PD up to 20 PD. Parks 3 step test is confirmatory.

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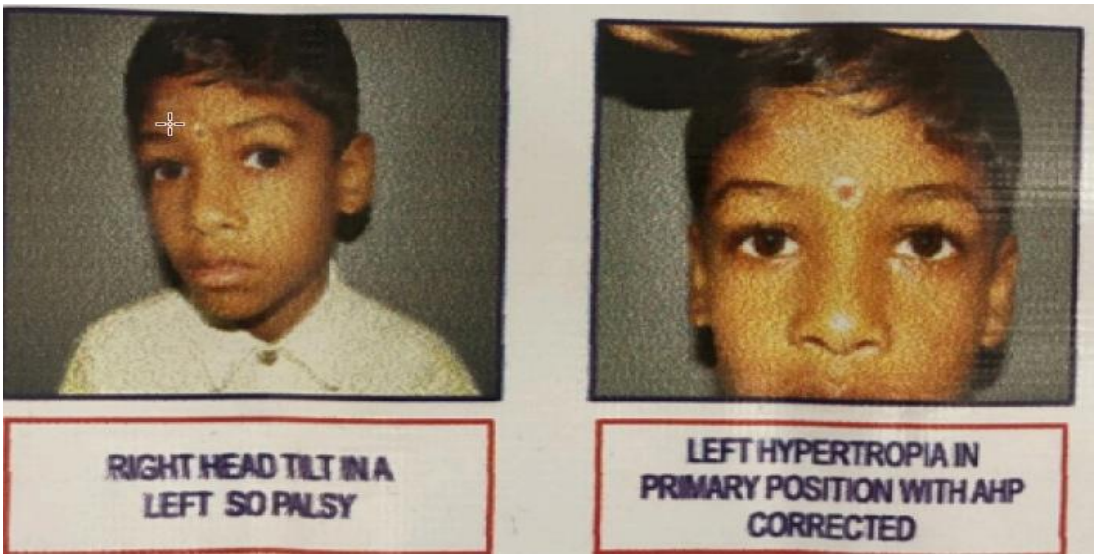
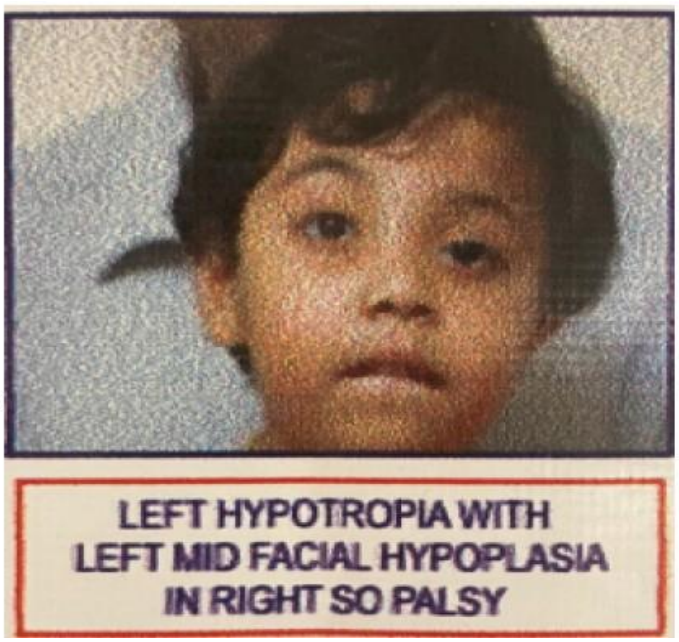


The complete three-step test fails to detect 30% of cases of superior oblique palsy (superior oblique muscle atrophy as confirmed by MRI). Often only two of three steps are positive in superior oblique palsy (2) AHP can be caused by various ocular or non-ocular diseases. AHP for ocular reasons is usually assumed to maintain binocularity and / or to optimize visual acuity. The prevalence of ocular causes of AHP was reported to be 18%–25%. Ocular AHP occurs for a variety of reasons, the most important of which include nystagmus, superior oblique palsy, and Duane's retraction syndrome. The first step in evaluating a patient with AHP is a correct differential diagnosis between non-ocular and ocular sources by performing comprehensive eye examination and ruling out other causes of AHP such as orthopedic and neurological conditions. The accurate diagnosis of the cause of ocular AHP and timely treatment can prevent the development of facial asymmetry and secondary muscular and skeletal changes in these patients. (3)Kushner et al studied ocular causes of abnormal head postures and concluded that incomitance accounted for 62.7% of head postures and nystagmus for 22.2 %. (4)

Severe torticollis is the main indication for surgical treatment in children with CSOP as the persistence of significant head tilt could lead to progressive facial asymmetry and tightness of the neck muscle namely, sternocleidomastoid .

Facial asymmetry: It develops because of chronic head tilt of any cause from young age. The facial asymmetry in congenital superior oblique palsy is typically manifested by midfacial hemihypoplasia on the side opposite the palsied muscle, with deviation of the nose and mouth toward the hypoplastic side. When the head is tilted, the dependent side of the face becomes smaller with reduced distance between lateral canthus and the corner of the mouth on the side of the tilt.



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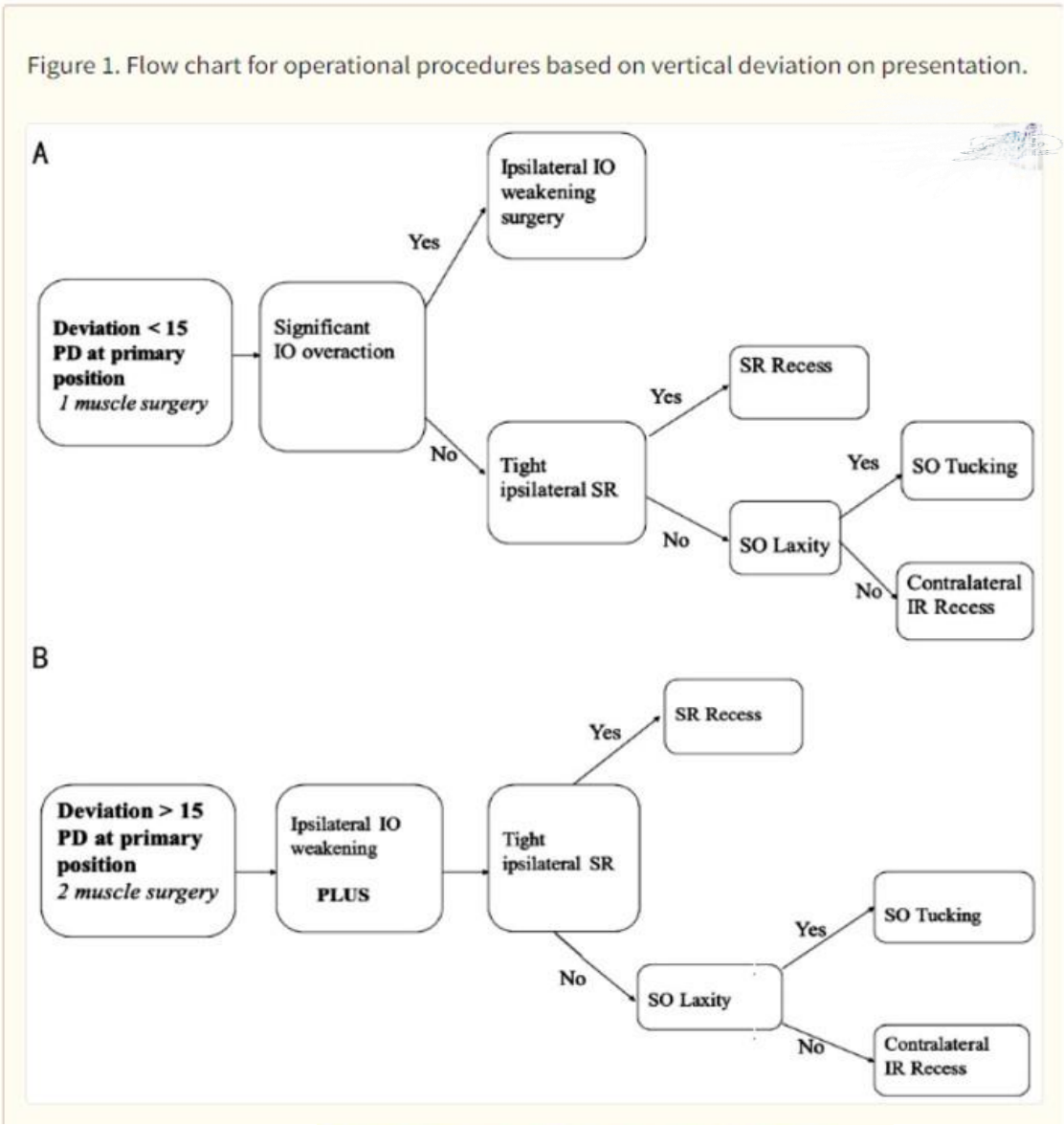
The indications for surgery in CSOP include correction of ocular torticollis and prevent development of facial asymmetry and to improve the sensory status. Various surgical options include inferior oblique (IO) weakening, superior oblique (SO) tuck, IO weakening combined with SO tuck, Contralateral inferior rectus (IR) recession and ipsilateral superior rectus (SR) recession.

Gordon shing kin Yau et al conducted a retrospective study on surgical outcomes for unilateral superior oblique palsy in Chinese population and concluded that most of the cases of CSOP without SO tendon laxity can be successfully treated by weakening of ipsilateral IO muscle. For those presenting with large primary deviations >15-20 PD, it is recommended to perform a 2 or 3-muscle surgery. They suggested the following flow chart for operational procedures (5)

Figure 1. Flow chart for operational procedures based on vertical deviation on presentation.



A: Smaller deviation; B: Larger deviation.

In our study, we have done IO recession with or without contralateral IR recession depending on the amount of vertical deviation in primary position .

MATERIALS AND METHODS

The source of data were cases of CSOP visiting the pediatric ophthalmology department in a tertiary eye care center in south India from 2004-2006. A retrospective analysis of 15 cases of unilateral congenital SO palsy which presented in the age group of 7- 20 years who underwent surgical correction was done. Cases of SOP documented as congenital in origin were included in the study. The goal of surgery was to correct hypertropia and head tilt. The diagnosis of CSOP was made based on photographic evidence of torticollis since childhood, in the absence of other causes. Each patient's case file was reviewed to collect demographic data (age,gender etc.), history

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regarding onset and duration of signs and symptoms, presence of facial asymmetry, any tightness of neck muscles and motor and sensory status. The following patient details were noted. Visual acuity, versions, ductions, worths 4 dot test, stereoacuity testing with Titmus test/TNO, cycloplegic refraction, orthoptic evaluation including parks 3 step test, alternate prism bar cover test to measure preoperative deviation in primary and nine gaze positions, when possible, post operative deviation, anterior and posterior segment evaluation and dilated fundus examination to look for objective torsion. One or two muscles were tackled depending on the amount of primary deviation in primary position. Standard operative procedure was followed for squint surgery. The second surgery was performed 3 months after the first surgery. Postoperative follow up at 1 day, 1 week, 1 month and at 6 months was done. Minimum follow up was for 6 months. Ophthalmic and orthoptic measurements were repeated at each postoperative visit. Surgical outcome was measured in terms of elimination of strabismus and head tilt or reduction of these findings to an insignificant level. Statistical analysis of the results was done using paired t test.

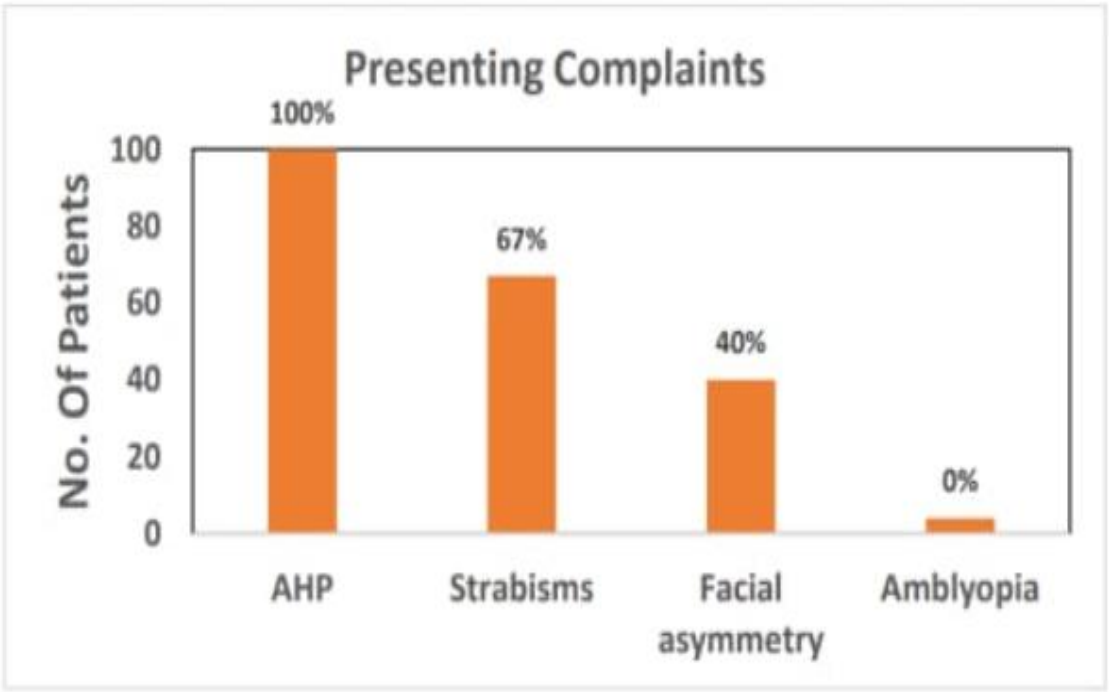
RESULTS

Our study population comprised of 15 cases. Mean age at surgery was 8 yrs. The Right eye was involved in 9 cases (60 percent of cases) and the left eye in 6 cases (40 percent of cases). Our study noted almost equal sex distribution with a male to female ratio of 8:7. No amblyopia was noted in any case.

Table 1

Avg. age at surgery	8 yrs
Sex	M:8 F:7
BCVA	6/6
Refraction	1 case of simple myopic astigmatism
Laterality	R:9 cases L:6 cases

The presenting complaints were AHP, strabismus and facial asymmetry as depicted in the bar graph.



One muscle was operated in 12 (80 Percent) cases and two muscles in 3 (20 percent) of cases as depicted in the pie chart

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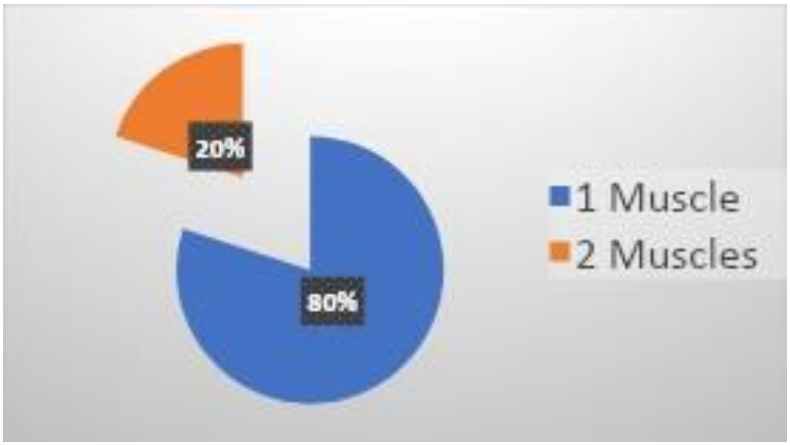
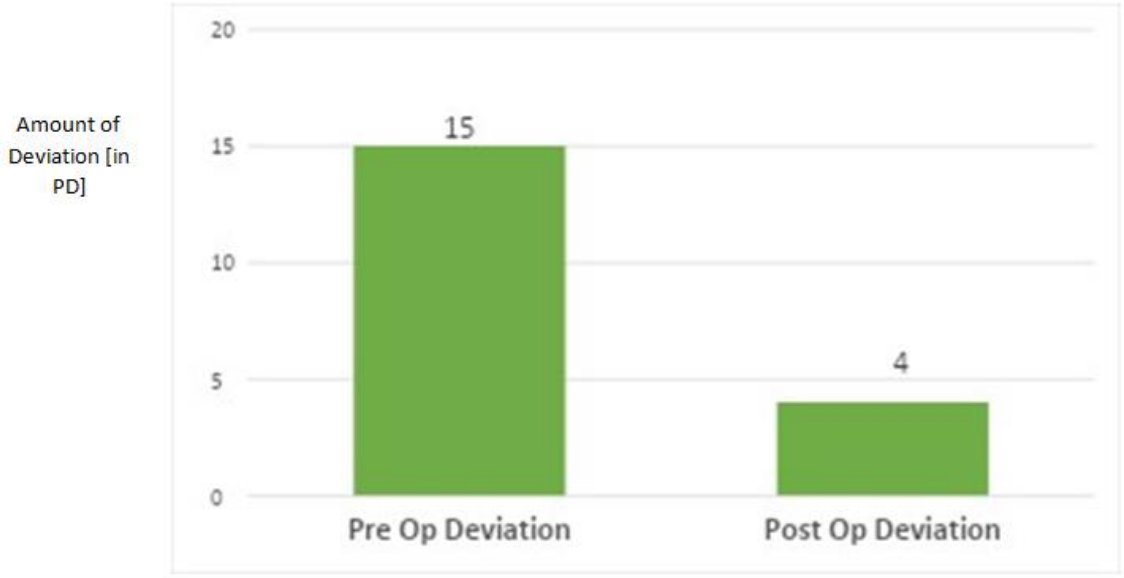


All 15 cases (100 percent) underwent IO recession, and 3 cases (20 percent) underwent IO recession with IR recession as a two staged procedure after a gap of 3 months.

The Mean preoperative vertical deviation (VD) was 15 ±8 PD and mean postoperative deviation was 4 ±2 PD. Vertical deviation reduced from an average of 15 PD to 4 PD,as noted in the bar graph and was statistically significant as per paired t test (P<0.01). In the 3 patients who underwent 2 muscle surgery, the vertical deviation reduced from 27 PD to 4 PD.None of the patients had any associated horizontal deviation.



The final postoperative deviation in primary gaze was ≤ 5 PD in all patients. All patients had significant reduction in head tilt which was subjective to examiner's assessment.

DISCUSSION

Facial asymmetry in ocular torticollis

Facial asymmetry is a condition in which the two sides of the face are not completely alike and similar.When present in an adult, facial asymmetry with an origin of ocular torticollis should help to confirm the chronicity of the defect and prevent unnecessary neurologic evaluation in patients with an uncertain history. Correcting torticollis through strabismus surgery before a critical developmental age may prevent the development of irreversible facial asymmetry. During the developmental stages of children, congenital muscular torticollis, with a prevalence of 0.3–2%, is the most important cause of the development and progression of facial asymmetry. The importance of this type of torticollis is that it is the most common cause of torticollis in children, and its effect on the development and progression of facial asymmetry in children is very similar to ocular torticollis. Therefore, it is necessary to be familiar with nature and causes of congenital muscular torticollis to differentiate it from facial asymmetry due to ocular torticollis. Neck traumas during labor are the most common cause of sternocleidomastoid (SCM) abnormality with a prevalence of 1 in 300 babies. In this condition, the baby shows some degrees of congenital torticollis. About 50–70% of the abnormalities of SCM resolve within the first year of life without requiring any special treatment therefore, the best age for surgical treatment is between 1 and 4

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years of age. The presence of torticollis during sleep or despite closing one eye is the most important finding used for the detection of non-ocular origin of torticollis since in torticollis, with an ocular origin, there is no preference to sleep on one side, and on the other hand, the head returns to its natural position upon closing one eye. In addition, in congenital muscular torticollis, some limitations in the neck movement and loosening of the neck on the affected side are noted while neck and muscular movements are perfectly normal in ocular torticollis. (6)

Our study showed almost equal incidence in males and females even though some studies have shown male preponderance. Facial asymmetry was seen in only 6 of our cases even though it was considered an important criterion for diagnosis. Amblyopia in CSOP is rare because strabismus is controlled with increased vertical fusional amplitudes as noted in our study also with none of the cases showing any amblyopia. We recommend tackling one muscle when vertical deviation in primary position was < 20 PD and two muscles when vertical deviation in primary position was > 20 PD.Our primary choice of muscle was IO as all cases had inferior oblique overaction (IOOA). It also had added advantages over SO tuck,like SO tuck requiring surgical expertise and 60-100 percent chances of acquired browns syndrome postoperatively.IO being a very forgiving muscle self titrates the result based on the amount of IOOA present with minimal complications. IR muscle was chosen when the second muscle needed to be tackled with a large primary deviation of >20 PD as none of the cases showed any evidence of SR contracture based on 9 gaze measurements and FDT intraoperatively. Two muscle surgery was done as a two staged procedure to avoid overcorrection in moderately large deviations. Favorable Outcome defined as less than 5 PD of hypertropia, and subjective improvement of head tilt was obtained in all cases. Main limitations of our study was its retrospective design causing lack of specific data like torsion in all cases, details on SO traction test and objective measurement of head tilt preoperatively and postoperatively was not done.

Michal Blaumost et al studied clinical outcomes of inferior oblique myectomy in age categorized patients with unilateral superior oblique palsyand concluded that the most common surgical approach for symptomatic patients is weakening of the ipsilateral antagonist inferior oblique (IO) muscle .In cases with large vertical deviation, this can be combined with contralateral inferior rectus recession or ipsilateral superior rectus recession .IO weakening procedures are described as self-titrating or self-graded with preoperative hyper deviation strongly correlating with the amount of correction achieved by the same amount of weakening [7]. Helveston et al studied surgical treatment of SO palsy and concluded that successful treatment of SOP can be accomplished in majority of cases by selective surgery usually beginning with IO weakening plus additional vertical rectus and horizontal rectus surgery as needed, with SO strengthening used only for lax tendons or when torsion is the main problem (8).Abbas Bagheri et al studied clinical features and outcomes of treatment for fourth nerve palsy and concluded that the most common form of SO palsy requiring surgical intervention was congenital which occurred most frequently in young males. Most cases of SO palsy can be successfully treated with a single surgical procedure. The most common operated muscle was the inferior oblique (83.6%) and the most common type of operation was inferior oblique myectomy (83.6%). The success rate for initial surgery was 84% and was increased to 96% with a second intervention. Its surgical treatment is highly successful if tailored according to the severity of the primary deviation and addressing gazes with the most significant deviation. Surgery directed to the SO which is the main involved muscle should be reserved for cases with significant tendon laxity or when torsion is the predominant problem (9) .Kaeser et al compared surgical results in patients with CSOP , treated with IO recession(IOR) alone v/s IOR + superior oblique tuck(SOT) and concluded that IOR alone is a suitable procedure for most CSOP's with moderate to large deviation in adduction, resulting in lower incidence of consecutive brown pattern than with IOR+SOT. (Consecutive Brown's pattern occurred in 18 of 20 patients who underwent IOR+SOT v/s 5 of 20 patients who underwent IOR alone (10). Pilar merino Sanz et al in their study on surgical treatment of superior oblique palsy mentioned that most published studies report favorable outcomes with inferior oblique muscle surgery, hence this muscle is frequently operated on. (11,12,13,14,15,16)

CONCLUSION

CSOP can be easily missed in young patients but with proper history and clinical examination, more cases can be diagnosed and treated early for better sensory and motor outcomes. Isolated inferior oblique muscle weakening is a safe and effective treatment option for CSOP with up to 20 PD of vertical deviation in primary position. Two muscle surgery should be reserved for larger deviations either as a two staged procedure or combined procedure depending on the amount of deviation in primary position. Good anatomical alignment was obtained in all patients with reduction of vertical deviation to ⩽ 5 PD in all patients ($p \leqslant 0.01$ which is statistically significant).All patients had satisfactory cosmetic results with either total resolution of head posture or only a mild tilt postoperatively

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