

FRONTALIS SLING SURGERY WITH SILICON ROD IN PTOSIS DUE TO CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIARatul Charan Deka¹, Roonmoni Deka²**HOW TO CITE THIS ARTICLE:**

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ABSTRACT: Chronic progressive external ophthalmoplegia (CPEO) is a rare genetic disorder which affects the ocular muscles producing ophthalmoplegia and ptosis. Mitochondrial DNA is at fault. Due to drooping of the upper eyelid the patient finds it difficult to move around. Besides, the patient adopts a defective head posture for proper viewing. Attempt has been made to elevate upper eyelid by the action of frontalis muscle using silicon rod as a sling. Frontalis sling surgery in CPEO using different materials like fascia lata, muscle tendon, skin muscle strip, supramid suture, silicon rod etc. are tried by different surgeons with varied success. 4 cases suffering from CPEO have been included in this study. For molecular diagnosis help has been taken from Geneticist. In all the cases, after obtaining the consent, upper eye lid of one eye has been elevated by frontalis sling using a silicon rod. Patient is given the option to undergo second procedure for the other eye if he was satisfied and there were no ocular complications. In all the cases satisfactory elevation of upper lid in the operated eye was achieved. Frontalis sling using silicon rod was found to be an easy and effective surgery to assist vision in CPEO where levator action was lost.

KEYWORDS: CPEO, ptosis, silicon rod.

INTRODUCTION: Ptosis and external ophthalmoplegia are characteristic features of Myotonic dystrophy and Chronic Progressive external ophthalmoplegia (CPEO). Mitochondrial DNA mutation is linked to this disorder with varied inheritance. As extra ocular muscles are rich in mitochondria due to high energy demand, selective involvement of ocular muscles in mitochondrial disorders is not uncommon. CPEO is the most common presentation in clinical practice.¹

A gradual onset of bilateral drooping of upper eyelids along with inability to move eye balls is the characteristic presentations of CPEO. Diplopia is usually not found in CPEO as the process is slow and involves both the eye simultaneously. Common age of onset is middle age. Both the sexes are affected. Due to ptosis, patient uses his frontalis muscle to elevate the lid and in doing so adopts a chin-up head position. This study is done to observe the effectiveness of frontalis sling using silicon rod to lift the upper eyelid with the help of frontalis muscle.

MATERIALS AND METHODS: From Feb 2011 to Jan 2014, out of approximately 60, 000 new cases attending the eye OPD of Fakhruddin Ali Ahmed Medical College, Barpeta, Assam, 5 cases of CPEO involving extra ocular muscles (EOM) and Levator Palpebrae Superioris (LPS) were detected. 3 cases were males & 2 cases were females. 4 cases have been included in this study as one male case did not turned up for surgery.

Their chief complaint was drooping of both the upper eye lids causing difficulty in vision.

Thorough ocular and systemic examinations were done as per standard protocol. Main ocular abnormality was near total loss of extra ocular muscle movement and ptosis. No LPS action was

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found. In all the cases, upper lid covered the pupil on both sides in primary position without effort. Patient tried to lift the lid with over action of the Frontalis muscle and slight backward bending of the head. In that position there was partial exposure of pupil but of a different degree on both sides. In no case total pupillary exposure was found even after maximum effort by the patient. Interestingly there was no diplopia in any of the cases.

All the cases were examined by the physicians but no other significant defect was detected. Once clinically diagnosed to be suffering from CPEO, opinions of geneticists were sought and diagnosis confirmed. The eye presenting with higher degree of visual problem was selected for surgery. Patients were operated by single surgeon at different points in time.

Silicon rod is commercially available. Frontalis sling surgery is a method by which a silicon rod of one millimeter diameter with 2 attached long needles was used to anchor the tarsal plate to frontalis muscle. Though LPS is not acting, by the action of frontalis muscle, patient can elevate the upper lid with relative ease. As cosmetic correction is not the priority in these cases, lid elevation is kept in minimum so that there is no corneal exposure and minimal lid lag.

The operative procedure has been performed in the following steps:

1. Informed consent has been taken from the patient and his/her close relative.
2. They were informed that one eye will be operated first and its effectiveness will be evaluated. They have the choice to getting the other eye operated based on the satisfactory outcome.
3. Local infiltrating anaesthesia by 2% lignocaine with adrenaline was used to anaesthetize supra orbital area and upper lid as a whole.
4. 3 small incisions of about 3 mm each were made at the level of upper boarder of the tarsal plate about 5 mm apart. In the same way 3 incisions were also made, app. 8 mm apart at the level of the upper boarder of the eyebrow keeping the middle one at a slightly higher level. One needle of the silicon rod was passed through the lower central incision towards the temporal one and likewise the other needle towards the nasal one.
5. Each of the needles was then passed in upward direction to come out through upper side incisions above the eyebrow.
6. Now, both the needles were passed in towards each other to come out through the upper central incision. A sleeve, a silicon tube about 8mm in length (which is supplied by the manufacturer) was then engaged by pushing both the needles carefully through it. Once the requisite lift of the upper lid is obtained (upper 2mm cornea kept covered by the upper lid), the tube is fixed, rod ends cut, 3 mm distal to the tube and buried under skin muscle through the upper central incision. Skin incisions usually do not require stitches. The upper most skin incision may require a stitch as some amount of manipulation takes place during burying.
7. Pad and bandage applied after putting antibiotic ointment.

The patient was examined on the next day to evaluate the outcome of surgery. Special attention was given for corneal exposure on attempted closure as they had no bells phenomenon. Systemic antibiotic, analgesics and ocular lubricants were prescribed. Patient was advised to come for frequent checkups. Adequate pupillary exposure without corneal exposure was considered the desired goal.

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RESULTS AND OBSERVATIONS:

Case 1: A female patient, aged about 30 years, presented with drooping of both the upper eyelids. Right eye was selected for surgery. No operative complications encountered. On the first post-operative day there was mild lid edema. Corneal exposure was adequate. No corneal erosions observed. On the second post-operative checkup after two weeks, upper lid skin fold noted which gradually diminished on subsequent follow up. Patient is doing fine in one year follow up with minimal lid lag.



Figure 1



Figure 2

Case 2: A 55 year old housewife with a similar presentation as above. The left eye was selected for surgery. Intra operatively, there was difficulty in passing the needles due to thin muscles and deep socket. There was also difficulty in burying the silicon tube. An infective granuloma was noted after one month in the area of the upper middle incision (Fig.3). Granuloma was removed and systemic antibiotics prescribed. Unfortunately, a granuloma appeared on the second incision site as well after some time and patient complained of pain. Silicon rod was removed. A second surgery was performed on the same side after 3 months which was uneventful. However slight under correction was noted.



Figure 3

Case 3: The third patient was a 70 years male with drooping of both eyelids, more so on the right side (fig 4). Right eye was selected for operation. No operative problem was encountered. Artificial tear prescribed. Immediate post-operative result was good (Fig. 5). However, mild (1mm) drooping of the lid was observed after six months post-operative. Patient is happy.



Figure 4



Figure 5

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Case 4: A 71 year old male patient presented with drooping of both upper eyelids (Fig 6). Right eye was operated. No operative complications encountered. Corneal exposure noted in the lower part on first post-operative day. Lubricants and antibiotics were prescribed. Patient did not turn up for early post-operative checkups. After 9 months patient came for operation of the second eye. Operated eye was normal (Fig 7). Now, the left eye has also been operated recently. Elevation of the lid kept at minimum as per the patient's request (Fig: 8).



Figure 6



Figure 7



Figure 8

DISCUSSION: CPEO is a rare disease. In this study we found the prevalence to be 0.00008% amongst the ocular diseases. CPEO is a disorder characterized by slowly progressive paralysis of the extraocular muscles. Patients usually experience bilateral, symmetrical, progressive ptosis, followed by ophthalmoplegia months to years later. CPEO is the most frequent manifestation of mitochondrial myopathies.¹ CPEO in association with mutations in mitochondrial DNA (mtDNA) may occur in the absence of any other clinical sign.

Mitochondrial DNA encodes for essential components of the cellular respiratory chain. Extraocular muscles are affected preferentially because their fraction of mitochondrial volume is several times greater than that of other skeletal muscles.¹ Impaired protein synthesis in these mitochondria accounts for the histological hallmark of the mitochondrial myopathies. The balance of oxidative demands of a given tissue and the proportion of deleted mtDNA it contains will ultimately determine whether the tissue is affected clinically.²

Ptosis is primarily myogenic in nature. Though all the age groups are vulnerable, middle age affection is common. No effective medical remedy is yet available. Only thing that give some help is to lift the lid by different surgical techniques like levator advancement, use of fascia lata as a sling, use of prop, etc.^{3,4} In Levator resection, subsequent adjustment of the lid position is not always easy. As the LPS muscle is atrophic, mechanical lift by silicon rod is an easy and effective procedure.⁵

Several materials have been tried for correcting paralytic ptosis. Patient's own Fascia lata is a time tested material for sling surgery. As it involves a separate invasive surgery with its own morbidity Silicon rod is selected for frontalis sling in this study. Lid lag is a known complication of sling operation. Silicon rod also produces lid lag but is kept in minimum by not trying to elevate the lid too much. The same principle protects the cornea.

Besides, silicon rod use keeps the adjustment of lid position open in post-operative period. Minimal decrease of lid elevation with time is described in literature. Two causes are- progressing pathology and sagging of the elevating material. In this study, lid elevation has been found satisfactory, stable and confidence boosting for the patient.

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Though the number of patients is less, considering the extreme rarity of the condition, as has been found, study on the effectiveness of the procedure described is clinically helpful.

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