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Dissertation on

# CONGENITAL PTOSIS WITH ASSOCIATED OCULAR AND SYSTEMIC ANOMALIES AND IT'S MANAGEMENT – A STUDY

Submitted in partial fulfillment of requirements of M. S. DEGREE BRANCH III OPHTHALMOLOGY

Of

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# CONGENITAL PTOSIS – IT'S OCULAR, SYSTEMIC ASSOCIATIONS

# AND MANAGEMENT

- AN ANALYTICAL STUDY.

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# **PART – 3**

PROFORMA MASTER CHART KEY TO MASTER CHART BIBLIOGRAPHY SURGERIES PERFORMED

# **AIM OF STUDY**

The main aspect of this study is to analyses the Ocular, systemic associations in congenital ptosis and its management. The purpose of this dissertation is to analyse:

- 1. Incidence of congenital ptosis
- 2. Age and Gender distribution
- 3. Ocular and systemic associations
- 4. Laterality of ptosis
- 5. Clinical parameters and investigations
- 6. Treatment modalities adopted
- 7. Complications of surgery

# **INTRODUCTION**

Ptosis (Blepharoptosis) denotes drooping of the upper eyelid. True congenital ptosis includes developmental dystrophy of levator muscle of unknown cause. In true congenital ptosis, deficiency of striated fibers in the levator muscle is present from birth and remains throughout life. The levator and frontalis muscle act in coordination, the frontalis muscle overacts to compensate the loss of elevating action of levator muscle.

The ptosis is often unilateral but can be bilateral. The choice of management depends primarily on the levator function. There is increase evidence to show that even if a ptosis partially occludes the pupil, there is a risk of amblyopia and surgery forms treatment of choice. Surgery restores stable, normally functioning eyelid and improves superior visual field.

# **ANATOMY OF EYELID**

The eyelids are mobile tissue curtains placed in front of the eyeballs. These act as shutters protecting the eyes from injuries and excessive light. The upper eyelid is unique in that many important structures in several lamellae are contained within a thickness of approximately 2 mm.

# **EXTENT:**

The upper eyelid extends from the eyebrow downward to end in a free margin which forms the superior boundary of palpebral fissure. The lower eyelids below merge in to the skin of the cheek.

# **STRUCTURE:**

From without inwards, each eyelid consists of following layers:

At about 3mm above the eyelid margin, these lamellae arranged from anterior to posterior as:

1. Skin

2. Subcutaneous areolar tissue

3. Pre tarsal orbicularis muscle

4. Post orbicular areolar tissue

5. Tarsal plate

6. Conjunctiva

At about 15mm above the eyelid margin, the lamellae are arranged as 9 layers:

1. Skin

- 2. Subcutaneous areolar tissue
- 3. Pre tarsal orbicularis muscle
- 4. Post orbicular areolar tissue
- 5. Orbital septum
- 6. Pre aponeurotic orbital pad of fat
- 7. Levator aponeurosis
- 8. Mullers muscle
- 9. Conjunctiva

The skin covering the eyelids is elastic, having a fine texture, is extraordinary thin, very mobile, 1mm thick and has thin epidermis. The skin can be divided in to loose mobile portion that overlies the orbital septum and the unmovable portion that overlies the tarsus, fixed to the anterior surface of tarsus by anterior fibres of levator aponeurosis. The skin of medial part differs from temporal part in that it is smoother and more oily with rudimentary hairs. The pretarsal skin demarcated from the preseptal skin at the supratarsal lid crease 7 to 10mm from the lid border. When the eye is open, the loose pre septal skin overhangs the eyelid crease forming upper eyelid fold.

# 2. SUBCUTANEOUS AREOLAR TISSUE:

Beneath the skin, is a layer of loose connective tissue, contains no fat. It is absent near the ciliary margin, palpebral sulci at the canthi where skin adheres to the palpebral ligaments.

#### **3. PRETARSAL ORBICULARIS MUSCLE:**

This layer consists of orbicularis muscle which forms a thin oval sheet across the eyelids. This muscle is most superficial muscle of the eyelid. It can be divided in to:

# a. Orbital part

b. Palpebral part

c. Lacrimal part (Pars lacrimalis)

d. The muscle of Riolan (Pars ciliaris)

# a. The orbital part:

Arises from the medial orbital rim, fans out superiorly and inferiorly, passes around the periphery of the eyelids around 360 degrees to insert near its site of origin at the medial orbital rim.

Musculus superciliaris: The upper fibres of the orbital part which passes to skin of medial part of eyebrow are termed as musculus superciliaris.

Musculus malaris: Inferiorly, the medial and lateral fibres are attached to the skin of cheek called musculus malaris.

# b. The palpebral part:

This part is subdivided in to pretarsal and preseptal portions. They arise from lacrimal fascia, posterior lacrimal crest and anterior part of medial palpebral ligament arcs superiorly and inferiorly and inserts along the lateral horizontal raphe and lateral canthal tendon. The part overlying the tarsal plate is pretarsal portion and that overlies the orbital septum above the tarsus are called preseptal portions.

# c. The pars lacrimalis:

The tensor tarsi often named Horner's muscle attached behind the lacrimal sac to upper posterior lacrimal crest and lacrimal fascia.

# d. The muscle of Riolan:

The fibres of the pretarsal portion which runs along the lid margin behind the ciliary follicle form the pars ciliaris (muscle of Riolan).

**NERVE SUPPLY:** zygomatic and temporal branches of facial nerve.

# 4. POST ORBICULAR AREOLAR TISSUE:

This is composed of loose connective tissue and contains facial nerve fibres. It provides first important surgical landmark in any operation on the eyelids because of its avascularity. The nerves and vessels of the lids lie in this layer.

#### **5. ORBITAL SEPTUM:**

It is a thin, floating membrane of connective tissue which takes part in all movements of eyelids. It is a strong, thin connective tissue. Elastic fibre sheet is absent in this septum. Centrally it becomes continuous with convex border of the tarsi. Peripherally, it originates from the arcus marginalis. In the upper eyelid, the septum terminates inferiorly as a rim which blends in to the fibres of the underlying levator aponeurosis 3 to 4 mm above the tarsal plate. In lower eyelid, it fuses with capsulopalpebral fascia.

# 6. PRE APONEUROTIC ORBITAL FAT:

Is a forward extension of the extraconal orbital fat, which is a very important surgical landmark. It lies beneath the orbital septum, seen as 2 fat compartments in upper lid and 3 in lower lid.

# 7. LEVATOR PALPEBRAE SUPERIORIS:

LPS is a striated muscle. It is about 40mm. takes origin from the lesser wing of sphenoid bone above the optic foramen and from the annulus of zinn. It extends forwards and split in to the aponeurosis and muller's muscle. It lies immediately above the superior rectus muscle and both are supplied by superior rami of occulomotor nerve and with which it functions synergistically. It passes anteriorly in to the orbit as horizontally oriented muscular elements up to the superior transverse ligament of Whitnall's and then it becomes aponeurotic.

# Levator aponeurosis:

Levator aponeurosis is about 10 to 50mm long, is the anatomically unique tendon, delivering force in to horizontally acting muscle in to vertical and posterior motion of lid elevation. This due to unique facial condensation of levator muscle, tendon and Whitnall's ligament. Whitnall's ligament separates the levator muscle and aponeurosis, below the ligament, levator is entirely aponeurotic with few (or) no muscle elements. This ligament is attached to the orbital wall medially at trochlea, laterally to the lacrimal gland capsule and periosteum of orbital wall and is acts as

Check ligament to levator retractor. The aponeurosis fans out and occupies the entire eyelid of width 30 to35mm. It is inserted by fuses with the distal fibres of the orbital septum about 3 to 4mm above the tarsal plate, interdigitate between fibres of orbicularis to insert in to the intermuscular septum and the overlying skin. It also attaches to anterior surface of tarsus and to the superior fornix of conjunctiva. Medially inserted in to medial canthal tendon and forms the medial horn and laterally also inserted in to tendon and forms lateral horn. The aponeurotic fibres form prominent horizontal eyelid crease with elevation of eyelid.

# Nerve supply:

Is from the superior division of Oculo- motor nerve.

# Action:

Elevation of eyelid.

# 8. MULLERS MUSCLE:

It is a smooth muscle, superior tarsal muscle, arises from the posterior terminal fibres of lacrimal muscle where the muscle becomes aponeurotic. It extends as a thin strip to the upper tarsal plate then it descends for 8 to10mm as individual fibres mixed with connective tissue, fat and blood vessels.

# Nerve supply:

Supplied by sympathetic nerves.

Action: serves as a accessory elevator of the eyelid.

# **TARSUS:**

Tarsal plates are dense fibrous connective tissue with some elastic tissue, flat and form posterior, firm skeleton of eyelids. It extends nasally from just beneath the punctum to within 4mm of lateral canthal angle. The upper tarsus merges with medial and lateral canthal tendon with maximum width in the center of the lid of about 9 to 10mm. It lodges 30 to 40 parallel holocrine sebaceous glands called meibomian glands in upper lid and 20 to 30 in lower lid. Posteriorly, it is closely adherent to conjunctiva. The free border at the lid margin is thick, attached border is thin, continuous with septum orbitale except where pierced by levator in upper and inferior rectus in lower lid.

#### 9. PALPEBRAL CONJUNCTIVA:

Palpebral conjunctiva, a highly vascular mucous membrane lines the posterior surface of both eyelids. It contains numerous mucous secreting goblet cells accessory lacrimal glands of Krause and wolfring. It may be subdivided in to:

a. Marginal zone: lies between skin and conjunctiva proper, puncta opens in this zone.

- b. Tarsal zone: is thin, reddish and highly vascular through which yellow streaks of tarsal glands visible.
- c. Orbital zone: lies between tarsal upper border and fornix.

# **BLOOD SUPPLY:**

Upper lid from ophthalmic artery, branch of internal carotid artery. Lateral palpebral artery, branch of lacrimal artery and medial palpebral artery forms marginal arterial arcades, which lies in sub muscular plane in front of tarsal plate. Branches from this, supraorbital and dorsal nasal arteries supplied eyelids and levator muscle. Branches from arterial arcades supply orbicularis, skin, fornix and backward to tarsal glands and conjunctiva. The lower lid is supplied by external carotid artery through its facial, maxillary and superficial temporal branches.

#### **VENOUS DRAINAGE:**

Veins are larger and more numerous than arteries of eyelids. They are arranged in 2 layers. They are:

a. Pretarsal venous plexus: It drains structures superior to the tarsus. It drains in to sub- cutaneous veins, angular vein on medial side ultimately in to internal jugular vein. Lateral

side, superficial temporal and lacrimal veins ultimately in to external jugular vein.

 b. Post tarsal venous plexus: It drains structures posterior to tarsal plate and in turn drains in to ophthalmic veins.

# LYMPHATIC DRAINAGE:

Lymphatic drainage from the lateral two thirds of the upper eyelid and lateral one third of the lower eyelid enter the pre auricular lymph nodes. The medial one third of upper eyelid and medial two thirds of lower eyelid drains in to the submandibular lymph nodes.

# **NERVE SUPPLY**

# **MOTOR NERVES:**

Are facial nerve (supplies orbicularis muscle) and occulomotor nerve (supplies levator palpebrae superioris).

# **SENSORY NERVES:**

Are derived from first and second division of trigeminal nerve. The upper eyelid is supplied by supra trochlear, supra orbital, infratrochlear and lacrimal nerves. The lower eyelid is supplied by infraorbital, lacrimal and infratrochlear nerves.

#### **SYMPATHETIC NERVES:**

Supplies the muller's muscle, vessels and glands of skin.

# **INTER PALPEBRAL FISSURE:**

Between the eyelids surface of the globe exposed is the interpalpebral fissure, measuring about 25 to 28mm horizontally and 7 to 11mm vertically. It is almond shape upper eyelid opening curves upward from each angle; highest elevation is just medial to midpoint of the pupil. The upper lid margin lies 1 to 2mm below the superior corneal limbus and lower lid rests at inferior corneal limbus.

# **FRONTALIS MUSCLE:**

Is quadrilateral shaped muscle, arises from epicranial aponeurosis and inserted in to eyebrow's skin mingling with fibres of orbicularis and corrugators.

Nerve supply: Supplied by facial nerve.

Action: It elevates the eyebrows above the line of vision.

# **PHYSIOLOGY OF EYELID**

The orbital rims, eyebrows, eyelids and periorbital soft tissues protect the anterior surfaces of the eye.

## **EYELID MOTILITY:**

The upper and lower eyelids are the opposing motor systems form a complex system movements, one for opening and one for closing the palpebral fissure. These involve both voluntary and involuntary controls, through the occulomotor, facial nerves as well as the sympathetic nervous systems.

# **EYELID OPENING:**

# THE UPPER EYELID:

The upper lid is elevated through the levator palpabrae superioris, Muller's muscle aids the levator in maintaining the elevation of lid. Extension of Herring's law, the levator muscles of both upper lids are bilateral and equally innervated. This is frequently demonstrated in patient with a unilateral ptosis with contra- lateral upper lid retraction. The eyelid muscles, levator and orbicularis muscles obey Sherrington's law of reciprocal innervations. When levator receives maximum innervations during opening, the orbicularis receives minimum innervations and vice versa. Frontalis partly contribute to lid elevation above the line of vision in extreme up gaze.

# THE LOWER EYELID:

The lower lid lacks a voluntary muscle. The capsulopalpebral fascia and inferior palpebral muscle are lower lid retractors.

# **EYELID CLOSSURE:**

Voluntary and involuntary closure of the eyelid is produced by the action of orbicularis oculi. EMG shows that there are 3 functional units, they are:

- Those responding in spontaneous blinking and tactile corneal reflex are pretarsal fibres.
- Those responding to voluntary blinking and sustained activity include preseptal fibres.
- Those responding to forceful closure of eyelid include all 3 parts of orbicularis namely pretarsal, preseptal and orbital fibres.

#### **SUPRANUCLEAR CONTROL:**

The corticobulbar and extra pyramidal systems both contribute to the levator nucleus and levator tones are related to the level of arousal. Levator and superior rectus muscles are linked together in all positions of gaze.

# **BLINKING:**

It refers to co-ordinate opening and closing movements of eyelids. A blink may be complete, incomplete, voluntary and involuntary. The involuntary blinks are further subdivided in to spontaneous and reflex blinks. The principal units of blinking fibres are pretarsal orbicularis. Preseptal fibres involved in blinking to a lesser extent.

Normal blinking rate ranges from 12 to 20 times/ min.

#### **BELL'S PHENOMENON:**

The eye turns upwards and slightly outwards when the lids are closed, so that cornea is removed from the region of palpebral aperture. Bilateral, highly coordinated reflex between the facial and oculomotor nuclei. Electrical activity record reveals that levator action ceases, superior rectus muscle action rises and inferior rectus action is inhibited. Superior rectus action begins later than the orbicularis but persists longer.

# CLASSIFICATION OF PTOSIS IS TO ARRIVE AT APPROPRIATE TREATMENT

# I. SIDNEY FOX'S CLASSIFICATION -1972:

# **ETIOLOGICAL CLASSIFICATION OF PTOSIS:**

# A. CONGENITAL PTOSIS:

- a. Simple.
- b. Complicated
  - i by other lid anomalies
  - ii. Paradoxical ptosis (neurological)
  - iii. by ophthalmoplegias

# **B. ACQUIRED:**

- a. neurogenic b.myogenic late spontaneous, myasthenia gravis
- c.traumatic d.senile

# C. HEREDO FAMILIAL:

- a. at birth (embryogenic fixation –mongoloid)
- b. late appearing (external ophthalmoplegias)

# II. FRUEH-B- MECHANICAL CLASSIFICATION OF PTOSIS-1980.

# A. NEUROGENIC PTOSIS- 3rd nerve palsy

- Misdirected 3<sup>rd</sup> nerve fibres
- Horner's syndrome

# **B.** MYOGENIC PTOSIS

- congenital development dystrophy
- progressive external ophthalmoplegias
- traumatic levator muscle injuries
- myotonic dystrophy
- oculopharyngeal muscular dystrophy
- myasthenia gravis

- toxic myopathy
- late acquired hereditary ptosis
- non-hereditary acquired myopathy

# C. APONEUROTIC PTOSIS- aponeurotic dehiscence (or) disinsertion

- senile aponeurotic redundancy

# **D.MECHANICAL PTOSIS**-eyelid (or) orbital tumour

- eyelid edema/ infection/ hematoma
- dermatochalasis
- brow ptosis
- cicatrical conjunctival scarring
- upper eyelid skin disease
- redundant tarsal syndrome

# E.PSEUDO- PTOSIS-orbital volume loss

- hypotropia
- blepharospasm and hemifacial spasm
- chronic ocular surface
- irritative disease
- contra lateral eyelid retraction

# **III.BEARDS CLASSIFICATION- 1981:**

# A. CONGENITAL PTOSIS-60%

a. with superior rectus normal function b. with superior rectus weakness

c. with blepharophimosis syndrome d. synkinetic ptosis (Marcus gunn jaw winking ptosis) e. misdirected third nerve ptosis

# **B. ACQUIRED PTOSIS-40%**

# a. Neurogenic

- i. traumatic ophthalmolgia
- ii. lesions of the occulomotor nerve

iii. Ophthalmoplegic migraine
iv. Horner's syndrome
v. multiple sclerosis ptosis
otosis
i. senile ptosis
ii. late acquired hereditary ptosis
iii. Progressive external ophthalmoplegias

iv. Hyperthyroidism

v. steroid ptosis

# c. traumatic ptosis

**b.** Myogenic

# d. mechanical ptosis

i. lid tumour ii. Blepharochalasis iii. Cicatrical ptosis

# e. pseudo- ptosis

i. true to anophthalmia, microphthalmos and phthisis bulbi.

ii. due to hypotropia iii. due to dermatochalasis

# LITERATURE OF CONGENITAL PTOSIS

Congenital ptosis is most commonly caused by a dystrophy of the levator muscle, which elevates the upper eyelid and is present since birth. It is of unknown etiology with strong hereditary tendency and is of eight types illustrated below:

# I. SIMPLE PTOSIS:

It is autosomal dominant, since the LPS and Superior rectus are closely associated until late in development, the anomaly involves both muscles. It is due to failure (or) peripheral differentiation of levator palpabrae superioris. It is of two types:

# a. SIMPLE UNCOMPLICATED PTOSIS:

Affecting the lid only, microscopically the levator muscle and aponeurosis invariably dystrophic, ptosis is secondary to local development defects in muscle structure.

# b. DOUBLE LEVATOR PALSY:

Is the commonest type of congenital ptosis, complicated by ipsilateral dysfunction of superior rectus without other signs of third palsy.

#### **II. ASSOCIATED WITH OTHER LID DEFORMITIES:**

# **Blepharophimosis syndrome:**

Bilateral, autosomal dominant, it contributes to about 3.5%. Clinical features are telecanthus, epicanthus inversus, ptosis with poor levator function, ectropion of lateral portion of the lower lids.

Other anomalies are: flat nasal bridge, mental deficiency, mongolism, microphthalmos, anophthalmia, facial hemi atrophy.

# III. PTOSIS COMPLICATED BY VARYING DEGREE OF EXTERNAL OPHALMOPLEGIA:

It accounts for 9% of ptosis.

# **KEARNE-SAYRE SYNDROME:**

Due to mitochondrial deletion disorder, characterized by progressive external ophthalmoplegias, heart block with Retinitis Pigmentosa, male preponderance in the 1'st or 2'nd decade.

# CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA:

May share ocular manifestation but fewer systemic manifestations, in 2'nd or 3'rd decade. Ptosis with ophthalmoplegias, diplopia, and asymmetrical involvement is present.

# **MYOTONIA DYSTROPHICA:**

Is localized to chromosome 19, in adults. Ptosis with premature thinning or loss of hair.

# IV. ASSOCIATED WITH MYASTHENIA GRAVIS AND MYOTONIA:

Due to marked end plate acetyl choline receptor deficiency caused by mutation. Congenital myotonia is present at birth, appears before the age of 5years.

# **V. SYMPATHETIC PTOSIS:**

Very rare, Congenital Horner's syndrome is characterized by partial ptosis, anhydrosis of ipsilateral face, and anisocoria with heterochromia.

# VI. SYNKINETIC PTOSIS: Is found in following conditions.

- 1. Congenital Marcus gunn jaw winking phenomenon.
- 2. Congenital III nerve palsy with synkinesis.
- 3. Duane's syndrome.
- a. Congenital Marcus gunn jaw winking phenomenon: It was first

Reported in 1885. In its fully developed form, at rest the upper lid covers the upper half of cornea. In a partial ptosis, when jaw is opened, the apparently paretic upper lid shoots upward to a level higher than normal eye. If the jaw is deviated to the affected side, the ptosis is increased, but if moves to opposite side, a maximal retraction occurs.

# **Pathophysiology:**

An aberrant connection between the motor branches of trigeminal nerve innervating external pterygoid muscle and superior division of occulomotor nerve that innervating the levator muscle.

#### EMG:

Shows synkinetic innervations of external pterygoid and levator muscles.

#### **INVERSE MARCUS GUNN PHENOMENON:**

Marin amat- 1924, ipsilateral eyelid closure when jaw moves to the opposite side. It seems to be an interfacial association rather than trigemino- facial association.

- b. Congenital third nerve palsy with synkinesis- central defects involving 3'rd nerve Palsy and aberrant regeneration.
- c. Duane's syndrome- although synkinesis does not involve lid innervations, enophthamos with apparent ptosis may be produced. Lid will descend further on adduction.
- VII. **PERIODIC PTOSIS:** Occurring in cyclic oculomotor spasm.

**VIII. Intermittent pseudoptosis:** associated with retraction syndrome. Ptosis associated with systemic anomalies: Is associated with systemic anomalies like cleft palate, heart disease, Arthogryposis multiplexa congenital, arachnodactaly. In klippel fiel syndrome, there will be other lid anomalies and extrinsic muscle involvement. Marfan's syndrome: tall, arachnodactaly, arm span more than total height, lung aplasia, ocular involvement like myopia, ptosis, megalocornea, lens dislocation, blue sclera and miotic pupil. Noonan's syndrome, fetal alcohol syndrome, Turner's syndrome, fetal drug toxicity like hydantoin, trimethadone.

#### Syndromes associated with cranio-facial anomalies:

Golden- harr syndrome, Saethre- chotzen syndrome, Gorcinchaundry moss syndrome, Apert syndrome, Epibulbar choristoma, Hypertelorism / Teleorbitism, Facial microsomia.

# **PATHOGENESIS OF CONGENITAL PTOSIS:**

Is deficiency of striated muscle fibres which causes inability of muscle to contract and relax so that elevation of lid to its normal level in primary gaze and relaxation restriction results in lid lag on looking down. Lack of peripheral differentiation or aplasia of the muscle at 20mm stage of development of four recti and two oblique's becomes differentiated and it is only after this that the levator forms by the formation of some of the fibres from medial aspect of superior rectus.

Levator may be:

- Completely absent or may be fused with superior rectus
- ✤ Replaced by a band
- ✤ Anomalous insertion

# **EVALUATION**

Pre-operative history and examination are vital because these decide the choice of surgery.

# History: includes

- ✤ Time of onset- since birth or later.
- ✤ Unilateral or bilateral.
- Improvement or progression or static.
- History of birth and trauma- mode of delivery whether forceps applied.
- ✤ History of associated other illness.
- ✤ Variation during the day.
- ✤ Diplopia.
- Family history of eye abnormalities, ptosis.
- ✤ History of previous surgery.

A detailed history was taken with examination of patient as per the pneumonic mentioned below.

# DESTINY -

- **D** Droop (degree of ptosis).
- **E** Excursion (amount of levator function).
- **S** Superior rectus action.
- **T** Tear Schirmer's test.
- I Iliff test for levator function.
- **N** Nerves corneal sensation.
- Y Yawn Marcus Jaw winking phenomenon.

# **OCULAR EXAMINATION:**

# **VISUAL ACUITY:**

Record of best corrected visual acuity to evaluate presence of amblyopia of ptotic eye.

# **PALPEBRAL APERTURE:**

Difference in palpebral aperture may be used as a measure of ptosis.

#### Vertical inter palpebral fissure height:

Measured at the widest point between the lower and upper eyelid with patient fixating distant object in primary gaze.

# Margin reflex distance- 1:

Is the distance from corneal light reflex to the upper eyelid margin, most effective. The mean measurement of MRD-1 in Indian population is 4.1+/-0.5mm. In severe ptosis, have a value of negative value.

# Margin reflex distance- 2:

Is the distance from the corneal light reflex to the lower eyelid margin.

Vertical palpebral height = MRD-1 + MRD-2.

**LEVATOR ACTION:** estimation of levator function is the single most important aspect for surgical planning.

# 1. Berke's method:

Excursion of upper lid measures from extreme down gaze with frontalis muscle action is blocked. The patient positioned against wall, while surgeon's hand press the forehead above the eyebrows that there is no downward or upward push. The patient is asked to look at extreme down gaze and then in extreme up gaze and the readings are recorded in mm and are classified as:

Good- 8mm (or) more.

Fair - 5mm to 7mm.

Poor - 4mm.

# 2. Putter man's method:

Is the assessment of levator function by measuring the margin limbal distance (MLD) in the extreme up gaze.

# **ASSESMENT IN CHILDREN:**

Measurement in small children is difficult. The presence of lid fold and movement of the eyelid occurs due to the levator action. The anomalous head posture like child moving his head back suggests poor levator action.

#### **ILLIF'S TEST:**

Is applicable in first year of life. The upper eyelid of the child is everted as the child looks down. If levator action is good lid reverts on its own.

#### Margin crease distance:

Is the height of the crease on the normal side should be measured and compared to the ptotic eyelid. When more than one lid creases are present, the most prominent one should be considered. The distance of the lid crease from the margin is measured as it helps in planning the surgical incision.

#### **Bell's phenomenon:**

Confirmation of presence of upward rotation of the eyeball on closure of eye is to indicate whether there is a risk of exposure keratitis in the postoperative period.

#### **Corneal sensation / Tear film abnormality:**

Should also be noted using a cotton wisp.

#### **Ocular motility:**

The extra-ocular muscle function particularly levator function should be recorded. History of worsening of ptosis late in the day should be subjected to levator fatigue test. The definitive diagnostic tests are tensilon test or neostigmine test.

#### **OTHERS:**

Variation in amount of ptosis with extra-ocular muscle in jaw winking movements (Synkinesis) should be assessed.

#### **Pupillary examination:**

Pupillary abnormalities are present in some acquired and congenital condition associated with ptosis like Horner's syndrome, III cranial nerve palsy. Miosis in Horner's syndrome and mydriasis in oculomotor nerve palsy.

#### Associated ocular findings:

In autosomal dominant, Blepharo phimosis syndrome, severe bilateral ptosis may be associated with telecanthus, epicanthus inversus, flattening of superior orbital margin, horizontal shortening of eyelids and hypoplasia of nasal bridge.

# SURGICAL PROCEDURES FOR MANAGEMENT OF SIMPLE PTOSIS

#### Anaesthesia:

In children, general anaesthesia is preferred. In adults, Local anaesthesia is preferable to general anaesthesia , since voluntary movement of levator muscle aids in the identification of lid structures and better intra-operative assessment of lid level .

#### **LEVATOR RESCTION:**

# **\*** EVERBSCH SURGERY (ANTERIOR OR TRANS CUTANEOUS APPROACH):

#### **Principle:**

The septum is divided and when the pre aponeurotic pad of fat is retracted, the whole levator complex can be examined directly for any defects. The muscle is shortened, sutured directly to the tarsus. Excess skin is excised and skin crease reformed which pick up the underlying levator muscle.

Indications: Ptosis with 5mm (or) more of levator functions.

#### Advantages:

Better and more extensive approach, easier to identify all structures (attachments of muscle). Large resection is possible and formation lid crease also possible

# ✤ BLASKOVIC'S SURGERY (POSTERIOR OR TRANS-CONJUNCTIVAL APPROACH)

## **Principle:**

The same procedure but through trans- conjunctival approach.

Indications: Ptosis with 6mm or more of levator function.

Advantages: Speed and sureness of isolating the LPS without injury to it.

# **Disadvantages:**

Lid tissues are turned inside out, orientation to structures become difficult, mobilization difficult, large dissection difficult. Risk of lacrimal gland injury as well as muller's muscle.

# FASANELLA SERVAT SURGERY (TRANS CONJUNCTIVAL MULLERECTOMY)

# **Principle:**

The upper part of tarsus with its attached palpebral conjunctiva, muller's muscle and levator are grasped and excised.

# **Indications:**

- Mild congenital ptosis with levator function more than 10mm.
- ➢ Horner's syndrome.
- Small degree of involutional ptosis not associated with aponeurotic weakness.

# Advantages:

Simple, very useful procedures with fair function of levator.

# \* MULLER'S MUSCLE AND CONJUNCTIVAL RESECTION:

# **Principle:**

The muller's muscle and the underlying conjunctiva are resected. Indications: Congenital and acquired ptosis with positive phenylephrine test response.

# Advantages:

Tarsectomy which can cause lid instability, meibomian secretion loss in Fasanella- Servat procedure complications are prevented here.

# **\*** APONEUROSIS SURGERY

#### **Indications:**

Acquired ptosis with an aponeurotic defect and good levator function, more than 10mm with negative phenylepherine test.

Advantages: correction of anatomical defect.

# **Disadvantages:**

More difficult performed than muller's muscle resection.

# \* BROW SUSPENSION OR TRANS- FRONTALIS SUSPENSION

# **Principle:**

Frontalis muscle lifts the eyebrow and eyelid with a subcutaneous sling, usually bilateral sling preferable.

#### **Indications:**

- Ptosis less than 4mm of levator function.
- Myogenic and neurogenic ptosis.
- Blepharophimosis syndrome.

Materials used for frontalis sling surgery:

 Fascia lata 2.Palmaris longus tendon 3.Non absorbable suture 4. Silicone rods 5.Mersilene mesh.

## **Methods:**

Hildreth- silver suspension
 Double rhomboid sling

- 3. Crawford's triangular and modified Crawford's sling
- 4. Fox pentagon sling.

#### FOX PENTAGON SLING:

Most commonly used procedure. The surgeon passes the sling material from one end of lid incision to other and then passes each end of sling material to the respected brow incision, posterior to orbital septum. Shortening the loops of sling material will elevate the lids.

#### MANAGEMENT OF COMPLICATED PTOSIS

**1. Blepharophimosis syndrome:** carried in two stages.

**Stage-1**- correction of epicanthal fold and telecanthus by number of procedures namely: Blair Spaeth, Y-Plasty and Mustard's double –Z plasty. Mustard's double-Z plasty is preferable for correction.

Double –Z plasty: first mark (A) just medial to medial canthus, (B) is marked so as to make the Intermedial canthal distance half that of inter papillary distance.2 marks are joined and all subsequent lines drawn are 2mm smaller than the line AB.2 lines drawn from point A parallel to upper and lower lid margins(AF). From center point C (of AB) line is drawn medially at 60degree both above and below (CD). Another line is drawn outwards from point D (DE). Lateral canthotomy and canthoplasty is carried out before the skin incisions begin. Flaps are undermined and retracted, cleared of all tissues up to periosteum and medial palpebral ligament identified. Bony opening made and smoothened. MPL of one side is wired with 24G sterile stainless steel wire. The wire is threaded to the other MPL with similar double bite the two ends are tightened, apposition assessed by movements. Wire ends are pushed back in to bony openings. Hemostasis is achieved and the incision closed in layers.

#### Follow up:

Bandage removed after 24 hrs and sutures are removed after 5-7 days. **Stage –2**:

The second stage procedure is performed 6 months after primary surgery. Bilateral fascia lata sling procedures are undertaken.

#### 2. Marcus –gunn ptosis correction:

Depends on whether the jaw winking is cosmetically significant. Choice of procedure is based on amount of ptosis and levator large resection is necessary. In case with significant jaw winking, levator disinsertion with the fascia lata sling surgery is the procedure of choice.

#### **3.** Congenital ptosis with superior rectus weakness:

Is common association as the two muscles develop from same myotome due to myogenic.

- 1. The hypotropia is corrected by surgery on the inferior rectus muscle before ptosis correction. The other procedure is levator excision on the normal eye followed by a bilateral fascia lata sling surgery.
- 2. Knapp's procedure: Transposition of medial and lateral rectus to the insertion of superior rectus is surgery of choice for double levator palsy.
- 3 Months later, ptosis correction is performed by standard technique.

## **COMPLICATIONS OF PTOSIS SURGERY**

#### 1. UNDER CORRECTION:

Is the most frequent complication due to fault of judgment and of techniques. It is most frequently due to resection of too little of levator muscle or aponeurosis and following brow suspension.

#### 2. OVER CORRECTION:

Usually follows levator resection and is rare. Rectified with massaging.

- 3. LID LAG, LAGOPHTHALMOS: more common with brow suspension.
- **4. EXPOSURE KERATITIS:** Lid lag results in keratitis, if not responds to medications, tarsorrhaphy can be done.

#### 5. ENTROPION AND ECTROPION:

Rare complications of ptosis surgery. Entropion following maximal levator resection and following sling surgery. Ectropion occurs following trans conjunctival approach of levator resection.

#### 6. LID CREASE, LID FOLD ABNORMALITIES:

Lid crease may too low results from skin incision. Lid fold may redundant if levator resection has been large and excess skin has not been excised.

#### 7. LOSS OF LID LASHES.

#### 8. CONJUNCTIVAL PROLAPSE:

From upper fornix is rare. It will retract spontaneously within a week or two weeks.

9. **POST OPERATIVE HAEMORRHAGE:** Rare, pressure dressing over the lid for a day or two.

## 10. INFECTION:

Because of abundant blood supply, infection is rare. When occurs, treated successfully with antibiotics except in brow suspension with a non- absorbable sutures, infection recurs. Removal of sutures and reoperation is the procedure of choice.

#### **SECTION – II**

#### AIM OF THE STUDY:

- 1. All children having congenital ptosis associated with congenital ocular and systemic associations were analysed in this study.
- 2. It also analysis the degree of ptosis, the effect of associated anomalies both ocular and systemic on the management of ptosis.

#### **MATERIALS AND METHODS:**

The study was conducted at Regional Institute of ophthalmology, Government ophthalmic hospital, Chennai for a period of 24 months from September 2006 to September 2008. Cases were referred from Chennai, different parts of the state and neibouring states. Out of 155 cases of congenital ptosis taken up for study, 78 cases had ocular and 12 cases had systemic associations.

#### **INCLUSION CRITERIA:**

- All patients who noticed ptosis since birth were included in this study.
- Congenital ocular and systemic anomalies associated with ptosis patients.

#### **EXCLUSION CRITERIA:**

- Post traumatic.
- Post inflammatory.
- Post surgical.
- Apo neurotic.
- Mechanical
- Pseudo ptosis and other acquired causes.

In each case, detailed history was taken with ocular and systemic examination. Ptosis was examined thoroughly as given in the proforma with special emphasis on the degree of ptosis, amount of levator action, Bell's phenomenon, orbicularis muscle action, corneal sensation, staining, presence of jaw winking phenomenon, associated ocular and systemic anomalies were carefully examined and included in this study. Pre and post operative photographic documentation was done for comparing the amount of outcome in surgery. Adequate and appropriate investigations were done before being taken up for surgery. Surgery was decided based on type and clinical nature of ptosis, amount of levator function, margin limbal distance and degree of ptosis. 3 types of surgical procedures were planned and performed for 121 cases:

- 3 cases of Mild ptosis (1-2mm) with good levator action (> 8mm)→Fasanella servat was done.
- 19 cases of moderate ptosis (2-4mm) with fair levator action (5-7mm)
  →anterior transcutaneous approach of levator resection was done.

It was calculated according to marginal limbal distance:

In bilateral cases  $\rightarrow$  9 – MLD of ptotic eye x 3.

In unilateral cases  $\rightarrow$  MLD of normal eye – MLD of ptotic eye x 3.

- 66 cases of severe ptosis > 4mm with poor levator action < 4mm</li>
  →Frontalis sling surgery were done. These cases were operated with non absorbable suture material 4'0 Prolene.
- 2 cases of severe ptosis, Whitnall's sling was done. Whitnall's sling is an internal sling where anterior tarsal plate was sutured to Whitnall's ligament.

Post operative evaluation was done on first and fifth post operative days, monthly follow up for 2 months and 6 months later. Follow up was done to find out the incidence of complications like failure, under correction and success of surgery especially in cases underwent sling surgeries. 7 cases reported with failure due to slipping of knot were re operated after two months from the date of surgery.

Double Z plasty followed by sling surgery was done in 12 cases of Blepharophimosis syndrome. Levator disinsertion followed by sling surgery was done in 14 cases of Marcus gunn jaw winking phenomenon. Out of 12 cases of strabismus, 5 cases underwent squint surgery followed by sling surgery. Crutch glasses were given for 2 patients with defective bell's phenomena with CPEO. Minimal ptosis of 34 cases were not surgically interfered.

#### **OBSERVATIONS AND RESULTS**

155 Cases reported with congenital ptosis in the age group of, from birth to 50 years were studied. Among these, 14 cases had family history of ptosis (9.0%). Only one had history of forceps application (0.64%). 95% of cases came for cosmetic purpose and 5% came for defective Vision. 90 male patients with 58.1% and 65 female patients with 41.9% were included in this study.

Left eye was involved in 64 patients with 41.2%, right eye in 56 patients with 32.9% and both eyes were involved in 40 cases with 29.1%.

#### CONGENITAL PTOSIS WITH ASSOCIATED OCULAR ANOMALIES:

Out of 155 patients, 78 had simple congenital ptosis with 50.3% and 77 were associated with ocular and systemic anomalies(49.7%). Among ocular associations, 20.6% patients had superior rectus weakness, 10.3% had Marcus gunn jaw winking phenomena, 12(7.7%) had Blepharophimosis syndrome and 7 (3.2%) had strabismus were reported. 2 (1.3%) cases each of bilateral congenital nystagmus, epicanthus, Duane's syndrome, coloboma choroid with iris and congenital myasthenia gravis were reported. 1 case of Euryblepharon and 2 cases of CPEO were included in ocular associations.

PTOSIS WITH OCULAR ASSO	NO. OF PATIENTS	PERCENTAGE
MARCUS GUNN JAW	16	10.3%
BLEPHAROPHIMOSIS SYN	12	7.7%
SUPERIOR RECTUS PALSY	32	20.6%
STRABISMUS	5	3.2%
EPICANTHUS	2	1.3%
CONG NYSTAGMUS / STRABISMUS.	2	1.3%
DUANE'S SYNDROME	2	1.3%
CONG MYASTHENIA GRAVIS	2	1.3%
COLOBOMA CHOROID& IRIS	2	1.3%
EURYBLEPHARON WITH CPEO	1	0.64%
CPEO	1	<i>O.64%</i>

# CONGENITAL PTOSIS ASSOCIATED WITH ASSOCIATED SYSTEMIC ANOMALIES:

Among 155 patients, 4 had delayed milestones (2.6%), 3 (1.93%) patients reported with congenital heart disease – Atrial septal defect and each one case of Noonan's syndrome, cerebral Palsy, Arthogryposis multiplexa congenita, patent urogenital sinus, craniofacial anomaly, congenital hypothyroidism with developmental delay, cleft palate, cleft lip with bifid uvula reported were included in this study. 1 patient found to have Oculo cerebro dento auricular malformation

#### ANALYSIS

The present analytical study on evaluation and surgical management of all congenital blepharoptosis was conducted at Regional institute of ophthalmology, Government Ophthalmic hospital, Chennai. 155 cases were evaluated and examined in detailed manner, Ocular and systemic anomalies associations were looked for in this study. The analytical data are as follows:

#### **INCIDENCE:**

Over a period of 24 months, 155 congenital ptosis cases were collected totally, the number of cases indicated that we should be alert for the possibility of co existing defects in patients with blepharoptosis. Out of this, at birth to 20 years formed major group of 128 cases with 82.5%, more than 50% of the patients belong to the age group of 5-15 years.

#### **GENDER DISTRIBUTION:**

Out of 155 patients, 90 were males (58.1%) and 65 were females (41.9%). This study shows that there is a male predominance.

AGE GROUP (in years)	MALE	FEMALE
AT BIRTH- 10	33	30
11 - 20	38	26
21 - 30	12	7
31 - 40	4	1
41 - 50	3	-
>50	-	1
TOTAL	90	65
PERCENTAGE	58.1%	41.9%

**AGE** - This table shows incidence of ptosis according to the age group:

# LATERALITY :

Among the laterality, left eye was noted in 64 patients with 41.2%, right eye was predominant in 56 patients with 32.9% and bilateral in 40 patients with 29.1%.

Among ocular associations, simple ptosis were found in78 patients (50.4%), superior rectus weakness were involved in 32 patients (20.6%), 16 had Marcus gunn jaw winking phenomenon (10.3%), 12 had Blepharophimosis syndrome (7.7%). Strabismus was found in 7cases (4.5%), 2 cases each had epicanthus, congenital nystagmus, coloboma of choroid and iris, Duane's syndrome and 2 had Euryblepharon. CPEO was reported in 2 cases.

#### **VISUAL ACUITY:**

Out of the cases included in this study, 121 had 6/6 vision, 27 cases had 6/12 but corrected to 6/6 with spectacles and 7 reported with amblyopia of 6/60 NIP vision.

#### **STRABISMUS:**

7 Cases 0f strabismus were reported, out of that 3 cases had exodeviation, 2 had esodeviation and 2 had vertical deviation.

#### SYSTEMIC ASSOCIATIONS:

Out of 155 cases, 12 were associated with systemic anomalies. Among 12 cases (7.7%), Congenital heart disease - atrial septal defect was associated in 3 cases (1.93%), 1 case each of Noonan's Syndrome, cerebral palsy, patent urogenital syndrome, cranio facial anomaly, congenital Hypothyroidism with developmental delay, cleft palate and cleft lip with bifid uvula were associated. 1 case of Arthogryposis multiplexa congenita reported with genu valgum and mild left eye

ptosis (0.64%). Arthogryposis multiplexa congenita is a condition found in a child, born for a maternal myasthenia gravis mother. Noonan's syndrome is a syndrome consists of ptosis, low set ears, cataract, webbed neck, frog facies, atrial septal defect, patent ductus arteriosus and mental retardation. 1 case came with a syndrome of Oculo cerebro dento auricular malformation having congenital ptosisblepharophimosis Syndrome, cerebral palsy, mental retardation, dental malformation, bat ears and convergent squint.

#### **DEGREE OF PTOSIS:**

Among the degree of ptosis, 40 patients had an average of 2mm mild degree of ptosis (25.8%), 3.5mm of moderate was found in 20 cases (12.9%) and 5mm of severe were reported in 95 patients (61.3%). Severe degree of ptosis formed the major group.

#### **SURGERY:**

Among 121 of 155 patients underwent surgery (78%), 45 were done under general anaesthesia after appropriate investigations. Fasanella servat surgery was done in 3 cases of mild ptosis (1.9%). Levator resection was done in 20 patients reported with mild and moderate ptosis, (12.9%) and frontalis sling in 68 patients with severe ptosis (42.6%). Internal sling surgery (Whitnall's sling) was done in 2 patients severe ptosis.

Age In Yrs	Fasanella Servat	Levator Resection	Frontalis Sling	Zplasty +Sling	Squint +Sling	Levator Dis+Sling	Total
At birth-10	2	3	25	4	1	4	39
11-20	1	11	25	4	4	7	52
21-30	-	4	9	3	-	3	21
31-40	-	2	4	-	-	_	6
41-50	-	-	2	-	-	-	2
>50	-	-	1	-	-	-	1
	3	20	66	11	5	14	119
Internal slign (whitnall's sling) 21- 30 yrs	-	-	2				121

Levator resection was done in 9 males (5.8%) and 11female patients (7.1%) were operated through transcutaneous approach. Depending on degree of ptosis and residual action of LPS, amount of resection was performed. Double `z' plasty followed by frontalis sling was done in 11 cases of blepharophimosis syndrome. All had good results with mild under correction of frontalis sling. Levator disinsertion followed by frontalis sling was done in 14 cases of Marcus gunn jaw winking phenomenon. In patients with poor levator action, frontalis sling was done by fox pentagon technique in all patients.39 male and 27 female in 68 cases of severe ptosis underwent frontalis sling. The suture material used as suspensory material was 4-0' Prolene. Frontalis sling formed major group of all surgeries.

#### **COMPLICATIONS:**

Under correction formed the major group of complications found in 10 cases (6.9%). Failure of surgery was noted in 7 patients who underwent frontalis sling surgery (4.5%), most probably due to slipping of knot or failure of anchoring the knot to the frontalis muscle. Mild exposure keratitis was noted in 1 case (0.65%) and was treated with lubricants. All cases recovered well within one week. Infection was not noted in this study following ptosis surgery, because of abundant blood supply, adequate antibiotics and strict aseptic precautions.

#### **DISCUSSION:**

155 Patients who noticed ptosis since birth with congenital ocular and systemic anomalies were included in this study. Post traumatic, Myogenic, Post inflammatory, Post surgical, Apo neurotic, Mechanical and Pseudo ptosis were excluded in this study. Familial association was found in 9.1% and it showed that there was familial association. 1case had history of forceps delivery.

#### **GENDER DISTRIBUTION:**

Majority were male (58.1%) than female (41.9%).

Majority of patients belongs to the age group from birth to 20 years. This may be due to cosmetic consciousness and awareness in this age group and lack of cosmetic correction in older age group.

#### **AMOUNT OF PTOSIS:**

Severe degree of ptosis formed the major group compared to mild and moderate (61.3%)

#### **ASSOCIATED OCULAR ANOMALIES:**

Simple congenital ptosis formed 50.3%, out of which 63 patients (80.9%) were unilateral; this correlates with other studies of Beard series-75%, Yanoff study-69%, Levine -75% and Spaeth- 76%. The involvement of left eye was common in this study. Congenital ptosis associated with congenital ocular and systemic associations formed 49.7%.

Superior rectus weakness was found in 20.6% patients, which was slightly higher side with studies like Berks-5%, Beard-5%, Levine- 5% and corresponds with Yanoff-30% and Spaeth- 21%. The incidence of Marcus gunn jaw winking phenomenon was 10.3%, which was slightly higher sided when compared with Beard study-4.6%. 1 case of inverse Marcus gunn jaw winking with poor bell's phenomenon was noted for which surgery was not done. 11 Patients with Blepharophimosis Syndrome comprises of 7.7%, which

was slightly higher than Beard -3-4% and corresponds with Nelson study-6.9%. Blepharophimosis family was reported in this study. Primary ovarian failure in FOX  $L_2$  mutation was reported in several studies with menstrual irregularity and infertility in female blepharophimosis patients. No such irregularities were reported in this study. 7 cases had strabismus (4.5%), of which 3 were associated with exodeviation, 2 had esodeviation and 2 had vertical deviation.

2 patients each had Nystagmus, epicanthus, Duane's syndrome, congenital coloboma of choroid and iris and Euryblepharon with 1.3% and 1 case of congenital myasthenia gravis. In this study, 7 cases were found with amblyopia due to anisometropia and occlusion. Amblyopia in congenital ptosis was noted in 6.9% in case of Nelson study and Drayer et al study.

#### PTOSIS WITH SYSTEMIC ASSOCIATIONS:

Out of 155 cases, 12 cases of systemic associations were noted. 3 had congenital heart disease, out of that atrial septal defect formed the major group with 1.93%. 1 case of Noonans syndrome consisting of congenital ptosis, cataract, atrial septal defect, patent ductus arteriosus, webbed neck, low set ears with short stature and pectus excavatum was reported. Noonans syndrome was named after Dr. Jacqueline Noonan. One child reported with congenital ptosis, cleft lip, cleft palate and bifid uvula (0.64%). One patient came with patent urogenital sinus (0.64%). One case of Arthogryposis multiplexa congenita was reported with mild ptosis in the left eye. It was associated with genu valgum,

occurred usually in maternal myasthenia gravis. One patient (0.64%) came with congenital blepharophimosis syndrome, telecanthus, cerebral palsy, mental retardation, dental malformations, bat ears, convergent squint with developmental delay and came to diagnosed as a syndrome of Oculo cerebro dento- auricular malformations. Surgery was deferred due to protein energy malnutrition and low weight. A case of cretinism and 4 cases of delayed milestones were reported with congenital ptosis.

#### **MANAGEMENT OF PTOSIS:**

Out of 155 cases, 121 underwent surgery (78%), 45 cases were done under general anaesthesia. 76 cases were done under local anaesthesia. Local anaesthesia gave better assessment of lid level per operatively compared with other eye. Fasanella servat was done in 2 cases of male and 1 female, who had mild ptosis with good levator function. The main complication occurred in this surgery was central peaking. This was prevented by proper placement of haemostasis. This procedure was done in the early period of our study. To avoid excision of the normal structures, it was not followed now. 41 males and 27 females of 68 cases underwent frontalis sling surgery. It was done with fox pentagon technique that gave good results. In recent advances, non- absorbable sutures such as silicone rods and mersilene mesh were used in ptosis surgery. But in this study, 4-0 Prolene was used as suspensory material with good results. Even though bilateral sling produce a symmetrical result on down gaze for cosmetic similarities, in this study only the affected eye was operated since the patients were not willing for surgery on the other normal eye. Lid lag on down gaze following sling surgery were informed to all the patients. The reasons for selection of non absorbable suture as the suspensory material were:

- Most of the patients were not willing for surgery on another part of body, though they were explained about the size of scar and success rate with fascia lata.
- ✤ Easy availability of material.
- Can be done in any age and no need to wait in children to grow up, for the leg long enough to furnish adequate fascia lata.

2 patients with severe ptosis underwent internal sling procedure. Whitnall's sling is an internal procedure in which anterior surface of tarsal plate is sutured to Whitnall's ligament with 6-0 non absorbable suture, tied them to raise the lid. Out of 19 cases, 9 males and 10 females were operated for mild and moderate ptosis. Levator resection was done for these patients through anterior trans- cutaneous approach with good results. Levator resection was planned according to Geoffrey J Gladstone and Sydney fox ptosis surgery based on marginal limbal distance formula:

#### Amount of resection:

Bilateral ptosis  $\rightarrow$  9 – MLD of ptotic eyelid x 3

Unilateral ptosis  $\rightarrow$  MLD of normal eyelid –MLD of ptotic eyelid x 3

Trans- cutaneous approach were followed in all the cases because of its advantages. They were:

- Easier and better exposure of tissues.
- More access to levator, where full correction is needed.
- Cornea need not be injured and conjunctivas need not to cut.
- Useful in estimation of how much to resect.
- Lid fold and excess skin excision and possible positioning of lid crease are easier.

Levator disinsertion followed by frontalis sling was performed in 14 patients with Marcus gunn jaw winking phenomenon. Out of that, 6 were male and 8 were female (9.0%). In 11 patients of blepharophimosis syndrome, 2 stages of surgeries were carried out. First stage, epicanthal fold and telecanthus

were corrected with mustard's double z plasty followed by second stage of correction of ptosis with mild under correction of frontalis sling (7.1%). Crutch glasses were given for 2 patients with CPEO and frontalis sling was not done due to absence of bell's phenomenon.

Under correction formed the major complication reported in this study. It was noticed in 10 patients (6.45%). 7 patients reported with failure of surgery due to slipping of knot, for which re surgery was done after 2 months. 1 case of exposure keratitis (0.64%) was found in the immediate post operative period, which recovered well with lubricants and pad and bandage.

# CONCLUSION

- 1. The incidence of ptosis was found to be more common in the age group of 11 to 20 years. This shows the cosmetic consciousness in this age group.
- 2. Majority of patients were male
- 3. Left eye predominance and familial associations were noted.
- 4. Among the congenital ptosis, simple congenital ptosis formed major group.
- Superior rectus weakness, Blepharophimosis syndrome and Marcus gunn Jaw winking phenomenon formed the major ocular anomalies associations. Congenital heart disease formed the major systemic associations.
- Three different surgical procedures were followed in most of the cases.
  Among this, frontalis sling surgery forms the major group.
- 7. As ptosis leads to un- acceptable cosmetic appearance and defective vision, the fight against ptosis by surgical procedures were successful. A surgery restored a stable, normal functioning eyelid and improves superior visual field, thereby forms the main treatment of choice.

#### **<u>REGIONAL INSTITUTE OF OPHTHALMOLOGY AND</u>** GOVERNMENT OPHTHALMIC HOSPITAL – CHENNAI

# STUDY OF CONGENITAL PTOSIS WITH ASSOCIATED

# ANOMALIES

# **PROFORMA**

NAME & ADDRESS	M/F	M/F	
<b>OCCUPATION</b>	D.O. A	D.O.S	D.O.D
COMPLAINTS:			
History of present illness:			
1. Onset of age Notice	ed on Mo	de (sudden / Gro	ıdual)
2. Duration and progression	n		
3. Pre disposing Factors	a. History (pr	regnancy and m	ode of delivery)
	b. Trauma at	Birth	
4. Associated symptoms:			
a. Jaw winking b. Diplopia	c. Dysphagia d. Fa	cial asymmetry	e. Squint.
5. Variability:			
a. Time of day	b. Progression (Int	ermittent or con	ttinuous)
6. Lid position during sleep			

#### PAST HISTORY:

a. Birth history – Normal / forceps / caesarian section.

b. old photographs.

#### FAMILY HISTORY:

Surgery for ptosis

Facial asymmetry or abnormalities

#### GENERAL PHYSICAL EXAMINATION: RIGHT EYE LEFT EYE

1.Head posture

2. Facial asymmetry (fore head wrinkling)

3. Lids and adnexa

4. Palpebral conjunctiva

5. EOM

6. Conjunctiva

7. Cornea

8. Anterior chamber

9. Pupil size

10. Lens

11. Visual acuity w/o glass

With glass

12. Retinoscopy

14. Tension

#### **EXAMINATION OF PTOSIS:**

- 1. Exclude pseudo ptosis, Brow ptosis and confirm ptosis
- 2. Forehead wrinkling
- 3. Position of eyebrow
- 4. Lids- scars, pigmentation

Spontaneous lid movements, Epicanthus

5. Amount of ptosis – Mild / Moderate / Severe Mild / Moderate / Severe

6. Levator function- Good / Fair / Poor Good / Fair / Poor

7. Upper lid crease level

8. Margin reflex distance –I, II.

- 9. Lid position on down gaze Lid lag / Lid drop
- 10. Jaw winking phenomenon
- 11. Orbicularis muscle action
- 12. Function of muller's muscle
- 13. Bell's phenomenon
- 14. Ocular motility
- 15. Strabismus / nystagmus
- 16. Schirmer's test

18. Force duction test

19. Associated features of syndromes.

Systemic anomalies Ocular like - telecanthus, epicanthus

20. Photographs

a. Primary gaze

b. Up gaze

c. Down gaze

d. Movement of upper eyelid and jaw

e. Post operative- early and late

OTHER SYSTEM INVOLVEMENT: CVS RS ABDOMEN CNS

DIAGNOSIS

TREATMENT

# INVESTIGATIONS: HAEMOGLOBIN BLEEDING TIME CLOTTING TIME

POST OPERATIVE EVALUATION AND FOLLOW UP

**COMMENTS** 

#### **KEY FOR MASTER CHART**

SIM–SIMPLE PTOSIS.

SR - SUPERIOR RECTUS WEAKNESS.

MJW – MARCUS GUNN JAW WINKING PHENOMENON.

BPS-BLEPHAROPHIMOSIS SYNDROME.

STS – STRABISMUS.

LPS- LEVATOR PALPABRAE SUPERIORIS

OMA- ORBICULARIS OCULI MUSCLE ACTION

EOM- EXTRA OCULAR MUSCLE

**BP- BELL'S PHENOMENON** 

**CS- CORNEAL SENSATION** 

ST- SCHIRMER'S TEST

CVA- CORRECTED VISUAL ACQUITY

**MM- MILLIMETER** 

F- FULL

**NEG-NEGATIVE** 

**RES- RESTRICTIVE** 

N-NORMAL

FAS SER- FASANELLA SERVAT

LEV RES- LEVATOR RESECTION

F S- FRONTALIS SLING

**U C- UNDER CORRECTION** 

NIL IN- NIL INTERFERENCE

CPEO- CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA

M R- MENTAL RETARDATION

**RDS- RIGHT DIVERGENT SQUINT** 

ADS- ALTERNATE DIVERGENT SQUINT

**RE- RIGHT EYE** 

LE- LEFT EYE

F- FEMALE

M- MALE

ASD- ATRIAL SEPTAL DEFCT

PDA- PATENT DUCTUS ARTERIOSUS

# C P- CEREBRAL PALSY

ARTHO MULTI CONG- ARTHOGRYPOSIS MULTIPLEXA CONGENITA

EXP KER- EXPOSURE KERATITIS

DUANES- DUANES RETRACTION SYNDROME

A-ABSENT

**U C- UNDER CORRECTION** 

FAIL- FAILURE

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# LIST OF SURGERIES PERFORMED

Sl No	Name	Age	Sex	Hos No.	Diagnosis	Surgery
1.	Lakshmi	70	F	40568	RE-Mature Cataract	ECCE With PCIOL
2.	Mani	45	М	49502	LE-Corneo Scleral Tear With Uveal Prolap	Wound Repair
3.	Kamala	60	F	6570	LE- Lower Lid Tear	Wound Repair
4.	Kumar	25	М	5670	RE-Corneal Tear	Wound Repair
5.	Kuppusamy	57	М	41232	LE-Nuclear Cataract	ECCE With PCIOL
6.	Arumugam	66	М	4567	RE-Recurrent Pterygium	Excision With Auto Graft
7.	Manjula	40	F	2578	RE-Recurrent Pterygium	Excision With Auto Graft
8.	Kuppu	69	М	56778	LE-Lagophthalmos	Lateral Tarsorrhaphy
9.	Sarasu	70	F	43215	RE-Dacryocystitis	Dacryocystec tomy
10.	Sangili	30	М	42364	RE-Panophthalmitis Evisceration	
11.	Iyammal	58	F	41325	LE-Immature SICS With Cataract PCIOL	
12.	Kuppu	65	М	2345	RE-Chalazion Incision &Curettage	
13.	Padma	30	F	4563	RE- Pterygium	Excision

Sl No	Name	Age	Sex	Hos No.	Diagnosis	Surgery
14.	Kaliammal	69	F	4321	LE-Upper Lid Tear	Wound Repair
15.	Pencillaya	56	М	42444	LE-Immature Cataract	SICS With PCIOL
16.	Chandran	45	М	5783	RE-Dacryocystitis	Dacryocystorh inostomy
17.	Kumar	50	М	40251	RE- Non Healing Corneal Ulcer	Therapeutic Keratoplasty
18	Robert	60	М	45623	RE-Immature Cataract	SICS With PCIOL
19.	Kuppamma 1	65	F	40020	LE-Nuclear Cataract	SICS With PCIOL
20.	Kannan	70	М	6543	RE-Entropion	Lateral Stripping
21.	Mohan	25	М	43254	RE-Congenital Ptosis	Frontalis Sling
22.	Vijaya	50	F	7529	RE-Conjunct. tear	Wound repair
23.	Lalitha	66	F	41121	LE-Mature cataract	PHACO with PCIOL
24.	Pandian	60	М	45623	RE-Chronic angle closure glaucoma	AG Surgery
25.	Latha	68	F	46573	LE-Dacryocystitis	Dacryocystorh inostomy
26.	Murugan	30	М	40020	RDS	RE-LR recession& MR resection