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Case Report

Congenital isolated nuclear oculomotor nerve palsy

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ABSTRACT:
Congenital bilateral oculomotor nerve palsy is a rare condition which presents as ptosis, ophthalmoplegia and exotropia.It can be associated with global developmental delay and hypoxic-ischemic encephalopathy. Early diagnosis and interventions improves the outcome of disease. We present a girl child of age 1 year 3 months, having ptosis bilateral (left> right), exotropia, eye movements were restricted except in abduction. She had delayed milestones since birth with four-limb hypotonia and convulsions. Neurological examination revealed global developmental delay with developmental quotient of 53%, with signs of quadriplegic cerebral palsy, and microcephaly. MRI showed gliosis, encephalomalacia, and cortical atrophy in the left temporo-parieto-occipital lobe, reflecting perinatal vascular insult. Ophthalmological examination revealed pupil-sparing bilateral oculomotor nerve palsy without amblyopia or major refractive errors. Thorough studies excluded metabolic and infectious etiologies. This case illustrates the association of congenital oculomotor nerve palsy with other systemic neurologic conditions, such as HIE and GDD. This is mostly due to perinatal hypoxic insult. Treatment involves surgical correction of ptosis and alignment to prevent amblyopia and maximize visual potential

Keywords: Oculomotor Nerve Diseases, Developmental Disabilities, Ptosis, Ophthalmoplegia

INTRODUCTION:

Congenital bilateral oculomotor nerve palsy is a rare neuro-ophthalmologic condition characterized by impaired function of the third cranial nerve on both sides. The oculomotor nerve plays a critical role in eye movement, eyelid elevation, and pupillary constriction.
Dysfunction of this nerve results in significant ophthalmoplegia, bilateral ptosis, strabismus, and often pupillary abnormalities. Unlike acquired cases, which may result from trauma, vascular events, or infections, congenital cases are typically linked to developmental anomalies, genetic factors, or brainstem malformations.Due to its rarity and variable clinical presentation, congenital bilateral oculomotor nerve palsy poses diagnostic and management challenges. Affected individuals often exhibit compensatory head postures to optimize visual function, and many require surgical or rehabilitative interventions to improve ocular alignment and eyelid function. This review aims to provide an overview of the etiology, clinical features, diagnostic approach, and treatment strategies for congenital bilateral oculomotor nerve palsy, with a focus on optimizing visual outcomes and quality of life for affected individuals.

CASE REPORT:

A girl of 1 year and 3 months comes to ophthalmology clinic with a complaint that both eyes deviate outward and cannot open since birth. She has an inclination towards the left side without nystagmus, aberrant regeneration, oculomotor synkinesis, or amblyopia. She was delivered at term as a fourth child by cesarean section weighing 3.4 kg. LSCS was required as there had been prior labor, meconium aspiration, and prolonged labor. The baby cried at birth but required a 5-day NICU stay for respiratory distress and oxygen therapy. No history of consanguineous marriages was found.

Neurological History: There had been a history of convulsions from birth with twitching episodes over the right hand lasting for 5 minutes. She could not sit or stand and was apparently delayed in her development. A

previous episode of fever with pneumonia included uprolling of eyes and frothing at the mouth.

Neurological Examination:

- Four-limb hypotonia.
- Developmental quotient: 53% (indicative of global developmental delay).
- Head circumference: 42 cm (<3 SD), suggestive of microcephaly.
- Diagnosed with quadriplegic cerebral palsy associated with global developmental delay and hypoxic-ischemic encephalopathy.

Brainstem Evoked Response Audiometry (BERA): Hearing sensitivity within normal limits.

Ophthalmological Examination:

Feature	Right Eye (OD)	Left Eye (OS)
Vision	Follows light and object	Follows light and object
Lids	Mild Ptosis (2mm droop)	Moderate Ptosis (3mm droop)
Pupil	Central, reactive to light	Central, reactive to light
Lens	Clear	Clear
Anterior Segment	Within normal limits	Within normal limits
Fundoscopy	0.3 c/d, circular, WNL, FR present	0.3 c/d, circular, WNL, FR present

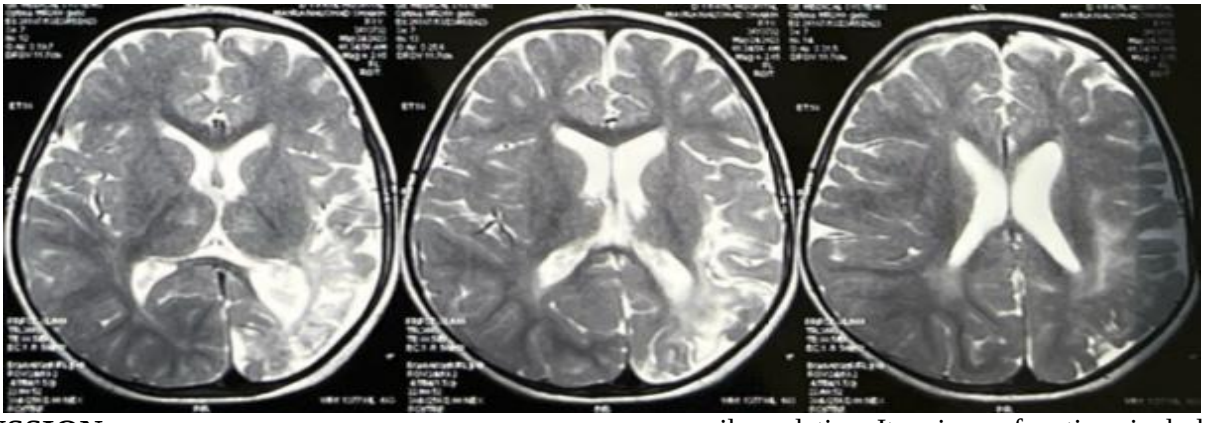
Extraocular Movements: Both eyes showed restricted movement in all gazes except abduction, giving a "down and out" appearance.
Hirschberg Test: 30° exotropia.
Cover Test: Bilateral exotropia.
Cycloplegic Refraction: No significant refractive error.
Laboratory Investigations:

- Hemogram: Normal.
- HIV, HCV, and HBsAg: Negative.
- Chest X-ray: Normal.
- Syphilis, Lyme disease, and collagen vascular diseases: Ruled out

Radiological Findings: MRI Brain + Orbit (Plain):

- Mild ex vacuo dilatation of the occipital horn of the left lateral ventricle.
- Gliosis and encephalomalacic changes.
- Mild atrophy of the left temporo-parieto-occipital lobe, consistent with perinatal vascular insult.

The I**mage 1** below shows axial MRI scans of the brain, taken using T2-weighted imaging, which highlights cerebrospinal fluid (CSF) in bright white. The ventricles in the brain are prominently visible in the center of each image, with an apparent dilation, suggesting potential conditions like hydrocephalus or atrophy. Surrounding brain structures show varying levels of gray and white matter differentiation, which could indicate abnormalities in brain parenchyma or other pathological changes.



DISCUSSION:

Oculomotor Nerve Anatomy, Functions, and Clinical Correlations. The 3rd Cranial Nerve also called as oculomotor nerve plays a role in critical motor and autonomic functions, thus aiding ocular movement and pupil regulation. Its primary functions include:

1. **Autonomic Function:** Innervates the pupil and lens via parasympathetic fibers, facilitating pupillary constriction and lens accommodation **[1]**.
2. **Somatic Motor Function:** Elevates the

upper eyelid and controls most extraocular movements essential for visual fixation and tracking **[2]**.

The muscles supplied by the oculomotor nerve can be categorized as

- Extraocular Muscles:

1. Levator palpebrae superioris: Raises the upper eyelid.
2. Superior rectus: Elevates the eyeball
3. Medial rectus: Adducts the eyeball
4. Inferior oblique: Elevates the eyeball when adducted.
5. Inferior rectus: Depresses the eyeball

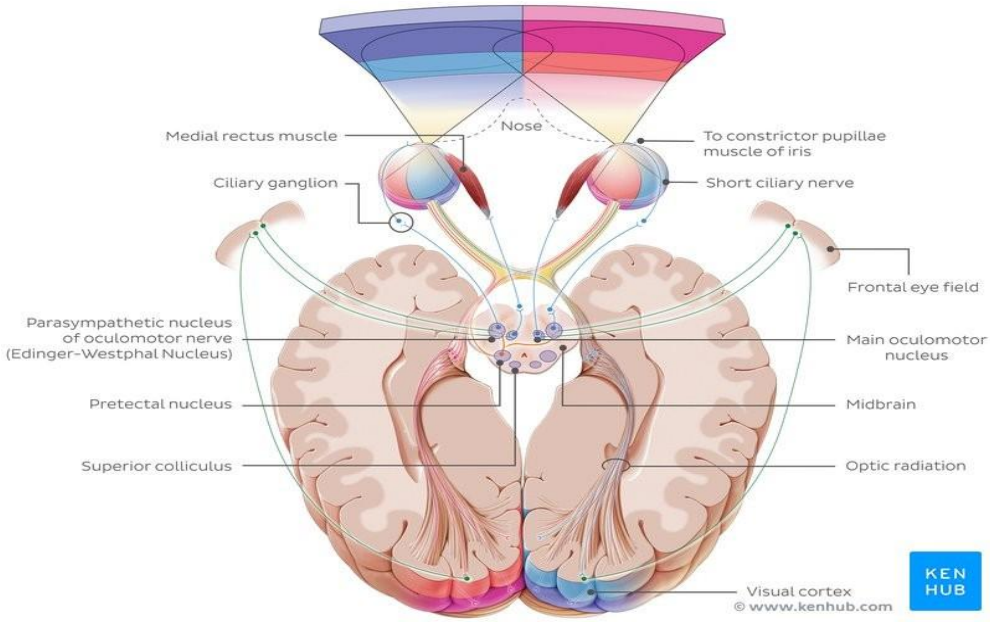
- Intraocular Muscles:

1. *Ciliary body:* For accommodation of the lens.
2. *Sphincter pupillae:* Constricts the pupil

The oculomotor nerve is divided into six key segments:

1. Nuclear Complex**:** Located in the midbrain.It includes:
   - Oculomotor nucleus: Supplies all extraocular muscles.
   - Edinger-Westphal nucleus: Provides parasympathetic innervation to intraocular muscles. Subnuclei within the nuclear complex controls contralateral superior rectus **[3]**.
2. Fascicular Segment**:** The efferent fibers pass through the midbrain, traversing the red nucleus and cerebral peduncles. **[4].**
3. Basilar Segment: This segment emerges from medial cerebral peduncle and is susceptible to trauma or aneurysms of posterior communicating artery **[5].**
4. Intracavernous Segment: In this area, the oculomotor nerve traverses the cavernous sinus, commonly involved in diabetes, pituitary apoplexy, or carotid cavernous fistulas **[6].**
5. Intraorbital Segment: It is divided into superior and inferior segments. This segment supplies the extraocular muscles **[7].**
6. Pupillomotor Fibers: Critical in distinguishing surgical from medical lesions, these fibers regulate pupil constriction **[8].**

Image 2 below shows the course, relationships, and regions of potential clinical involvement of the oculomotor nerve.



Clinical Features of Oculomotor Nerve Palsy:

1. Bilateral Ptosis: If the levator subnucleus affected.
2. Pupillary Abnormalities: Dilated pupils may be seen due to involvement of parasympathetic fibers.
3. Diplopia and Ocular Misalignment: Due to loss of coordinated action of the muscles.

Treatment and Advancements:

The main goal of treatment is to provide ocular alignment and ptosis correction. Techniques include:

1. Non-Surgical Techniques:

A. Occlusion therapy and corrective glasses.

B. Botulinum toxin injections.

2. Surgical Techniques:

- Lateral rectus recession with medial rectus resection

- Transposition of vertical recti muscles.

Advancement: Techniques such as Faden's operation and intramuscular bupivacaine injections into paretic muscles are thought to improve muscle function.

CONCLUSION:

Congenital forms of oculomotor nerve palsy are rare and have incidence of 7.6 per 100,000 cases. They mostly present as pupil sparing and may be associated with global developmental delays. MRI, CT angiography, and cerebral angiography are important to identify the underlying cause.

CONFLICT OF INTEREST: None

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