CHAPTER 1

1.1 INTRODUCTION

Cerebral Palsy (CP) is a group of permanent disorders that occur in the growing fetal or infant brain which include non-progressive set of disorders causing restriction of motion and abnormal posture. (1) It is one among commonly found motor disabilities during childhood. The motor disorders of Cerebral Palsy are usually accompanied by conflict of sensation, cognition, communication, perception, and/or behaviour and/or by a seizure disorder and by secondary musculoskeletal problems. Oculomotor functions and speech are affected due to bilateral involvement of corticobulbar tracts. As corticobulbar system is composed of the upper motor neurons of the cranial nerves and terminates on motor neurons within brainstem motor nuclei. Therefore, it controls the muscles of the face, head and neck. The primary cause of Cerebral Palsy is extreme prematurity and very low birth weight in most of the developed countries.

Overall Prevalence of CP:

World Scenario: The prevalence of Cerebral Palsy (2) from the population-based studies in the world estimates ranging from 1.5 to more than 4 per 1,000 live births. The overall prevalence of Cerebral Palsy was 2.11 per 1000 live birth (3). On an average, Cerebral Palsy occurred 1.2 times more frequently among boys than girls. Among the several types of CP, Spastic Cerebral Palsy (80%) was the majority among other types (4).
Prevalence of Cerebral Palsy was lower in Asians than Caucasians (1.09 versus 1.36 per 1000; relative risk = 0.80, 95% confidence interval [CI] = 0.74–0.87) and ranged from 0.61/1000 in Thai children to 2.08/1000 in Samoan children (5). Several Asian subgroups had low risk with respect to maternal age, educational attainment, and birth weight (6).

**Prevalence of CP by birth weight**

**World Scenario:**

Cerebral Palsy was more prevalent (59.18/1000 live birth) in children with 1000 to 1499g birth weight than in children with 2500g birth weight (1.33/1000 live birth). Three other studies found the prevalence of CP in relation to birth weight per 1000 neonatal survivors (2).

(a) 60.04 (under 1500g)

(b) 8.33 (between 1500-2499g)

(c) 1.16(over 2500g)

In Japan from 1977 to 2000, pattern of incidence in Cerebral Palsy due to gestational age and low birth weight, the incidence of Cerebral Palsy was done. The prevalence of CP was111.80 per thousand neonatal survivors born 28 weeks and 144.72 per 1000 neonatal survivors born between 28 and 31 weeks (7). They concluded that the proportion of Cerebral Palsy had increased due to low birth weight and lower gestational age (7).
Indian scenario:

Prevalence of Cerebral Palsy in India is three per thousand live births. The rate has remained the same for more than two decades (8). In India, since the cause of Cerebral Palsy is most often in term babies or near term that have anoxia and birth related complications, or related to infection post natal, we cannot make assumption about co-occurring conditions and must study them separately.

The risk factors for Cerebral Palsy namely males, premature babies, low birth babies, multiple births were identified by The Indian Children with Cerebral Palsy register report in 2009. Higher rates of Children with Cerebral Palsy were mainly associated with prematurity, low birth weight and multiple births (9). 42% of the Children with Cerebral Palsy in India were born prematurely, 11% of the Children with Cerebral Palsy were from a multiple birth. This might be due to slow intrauterine growth(10).

In Jammu & Kashmir, prevalence of Cerebral Palsy less than 10 years of age was investigated in RS Pura town (11). A total of 11 cases of Cerebral Palsy yield a prevalence rate of 2.27/1000 in the age group of less than 10 years. They concluded that the proportion of Cerebral Palsy occurring in males was higher than in females (12).
Visual Impairments in CP

In the “I Count” study in Chennai, India, Dr. Namita Jacob documented the nature of visual impairments among five hundred children with developmental disability. Over 65% of the children were found to have a vision impairment, with optic nerve atrophy and cortical vision impairment as the most significant causes of impairment among the children (13). In 2000, Bhatia (12) documented one hundred consecutive cases of Cerebral Palsy found that 80% of the children had associated conditions other than locomotors disabilities that were often not addressed by the primary care physician. Dowdeswell et al. 1995, found that prematurity and asphyxia affected the visual system among CP (14). Since prematurity is a major cause of Cerebral Palsy in India, documentation of vision impairment in Cerebral Palsy is important.

Etiology of CP:

Cerebral Palsy has multiple etiologies in antenatal, natal and postnatal periods (17). High risk factors for Cerebral Palsy were older maternal age, male gender, preterm birth, and low birth weight(16). Intrauterine infections, developmental malformations of brain are accountable in some of the cases (18). The main etiologies were brain dysphasia (17%), fatal brain vascular disorders (15%), hypoxic ischemic encephalopathy (14%) (15).
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CATEGORIZATION OF CEREBRAL PALSY:

Cerebral Palsy is often categorized by severity plane as mild, moderate, severe, or no CP. They are also classified based on Physiological, Topographical, Etiological and Supplemental factors.

Mild - The child would move without assistance and with no limitations in active daily living.

Moderate - The child would require adaptive technology to perform daily activities.

Severe - The child would have significant challenges in performing daily activities.
No CP - The impairment would be acquired after achievement of brain enlargement and hence it is classified in the occurrence that caused Cerebral Palsy, such as traumatic brain injury or encephalopathy (20).

Physiological

Physiological means relating to normal healthy functioning and not pathological.

Spastic Cerebral Palsy: It is caused due to the damage in cortex which is the outer layer of the brain. It is the most common type of CP; around 70-80% are of this type of CP. (20).

Athetoid Cerebral Palsy:

It is also known as dyskinetic Cerebral Palsy, which is caused due to the damage caused in the basal ganglion. As the muscle tone would get affected, both hypotonia and hypertonia seen in these individuals. (20),(21)

Ataxia Cerebral Palsy:

It is caused due to the damage in the cerebellum. Ataxia means “incoordination”. These individuals have jerky movement and instability.

Mixed Cerebral Palsy:

It is caused due to damage in the many parts of the motor control centers in the brain. The individuals may have a combination spasticity, dystonia and ataxia.
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**TYPES OF CEREBRAL PALSY**

- **SPASTIC**: tense, contracted muscles (most common type of CP).
- **ATHETOID**: constant, uncontrolled motion of limbs, head, and eyes.
- **RIGIDITY**: tight muscles that resist effort to make them move.
- **TREMOR**: uncontrollable shaking, interfering with coordination.
- **ATAXIC**: poor sense of balance, often causing falls and stumbles.

Fig 2: Types of CP and the area of the brain affected (21)

Fig 3: Areas of the brain involved in various types of CP and the functions affected due to CP (21)
Topographical: Topographical means relating to the arrangement or accurate representation of the physical features of an area.

A. Monoplegia involves one limb; condition is rare.

B. Paraplegia Involves the legs only and practically always of the spastic or rigidity type.

C. Hemiplegia The lateralized one-half of the body is affected and it is usually spastic, although pure athetoid hemiplegias are occasionally seen, as are pure rigidity hemiplegias. There is often sensory involvement in the areas of proprioception to point discrimination and form perception. Aphasias appear more frequently in right than in left hemiplegias and are much more common in the acquired than in the congenital Children with Cerebral Palsy.

D. Triplegia Involves three extremities, usually both legs and one arm, usually spastic. This may represent hemiplegia plus paraplegia, or incomplete quadriplegia. In the latter case, both arms will be equal or nearly equal in length. In the former, the involved arm will be shorter.

E. Quadriplegia (Tetraplegia) Involvement of all 4 extremities. Patients with the greatest involvement of the legs are usually spastic, and patients with greatest involvement of the arms are usually the dyskinetics, including athetoids.

F. Diplegia This term is seldom used. “Paralysis affecting like parts on either side of the body; bilateral paralysis.”

G. Double Hemiplegia This term implies those cases in which the arms are more involved than the legs. These are usually spastic in type.
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Darker shaded are more severely involved region

Fig 4: Various types of CP Vs affected regions of the brain (21)
Etiological:

Etiological means relating the cause or origin of a condition.

A. Prenatal

1. Hereditary:

   It is transmitted through genes, also known as “Cerebral Palsy agenesis”.

   No progression. Generally, symptoms are present at birth.

2. Acquired inutero

   a. Prenatal infection:

      Maternal infection during gestation like toxoplasmosis, rubella

   b. Prenatal:

      Anoxia—the carbon monoxide, or strangulation of mother, maternal anemia, hypotension, e.g., following spinal anesthesia, placental infarcts, or placenta abruption, kinking, knots or prolapses of the cord.

   c. Prenatal children with Cerebral Palsy hemorrhage: maternal toxemia, direct trauma, maternal bleeding diathesis.

   d. Rh factor

   e. Metabolic disturbances: gestational diabetes.

   f. Gonadal irradiation, harmful exposure to x-ray

   g. Maternal malnutrition.

B. Natal

1. Anoxia

   a. Mechanical respiratory obstruction.

   b. Atelectasis.
c. Narcotism (due to drugs).

d. Placenta previa or abruptio.

e. Maternal anoxia or hypotension.

f. Breech deliveries with delay of the after-coming head.

g. Bleeding in the first trimester

C. Postnatal

1. Trauma (accidental): Subdural hematoma, skull fractures, wounds and contusions of the brain

2. Infections: meningitis, encephalitis, brain abscess which is more common in children than adults

3. Toxicity: Lead, arsenic, coal tar derivatives, streptomycin.

4. Vascular accidents: congenital aneurysms, hypertensive encephalopathy, emboli due to bacterial endocarditic or fat embolism, cerebrovascular thrombosis, in debilitated infants, sudden pressure changes which is more common in adults than children.

5. Anoxia: Carbon monoxide poisoning, strangulation, high altitudes, and deep pressure anoxia, hypoglycemia.

6. Neoplastic, or late development defects: Brain tumors, brain cysts, internal hydrocephalus, and hydrocephalus.

V. Neuroanatomical

VI. Functional Capacity (degree of severity)

Class I: no practical limitation of activity.

Class II: slight to moderate limitation of activity.
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Class III: moderate to great limitation of activity.

Class IV: unable to carry on any useful physical activity.

VII. Therapeutic

Class A: not requiring treatment.

Class B: needs minimal bracing and minimal therapy.

Class C: needs bracing and apparatus, and the services of a Cerebral Palsy treatment team.

Class D: require long term institutionalization and treatment.

VISUAL ISSUES IN CEREBRAL PALSY

FIXATION IN CEREBRAL PALSY:

Evaluation of fixation is important in Children with Cerebral Palsy. Fixation capacity of a child may vary during different tasks (22). It can be stable, or unstable, and can be disturbed by spastic movements. Although it is possible for a child to read with poor fixation, it is quite challenging for most as the complex demand of motor functions required in reading may be too high for mastery. Head control and posture affects the eye movements and thus also the use of vision. For many Children with Cerebral Palsy, body control may require so much of their motor capacities that the children cannot maintain erect posture during the lessons when listening and looking at the teaching materials and so they are bent over the desk. In that position, the use of vision is ineffective (23). In some children, backward head movement may trigger a spasm in the oculomotor muscle and other extra-ocular muscles so that the eyes turn in (convergence), the pupils become
smaller (miosis) and there is a myopic change in refraction of up to 10-15 dioptres making it hard to immediately bring their eyes to focus on the object of interest.

**FAST EYE MOVEMENTS OR SACCADES IN CHILDREN WITH CEREBRAL PALSY:**

Saccades are quick, precise eye movements from one fixation target to the other. It is vital to perform many visual tasks especially while eyes have to fixate at the fast moving objects to get clear vision. The quality of fixation, the speed and the accuracy of the saccades need to be observed, as they are important in enabling fluent reading. Saccades are often abnormal in children with motor problems, especially in children with Cerebral Palsy (24). Children can usually follow and track moving objects using smooth eye movements. The following movements of children with impaired vision are often not smooth pursuit movements but composed of irregular saccades. Head movements can be used to compensate for insufficient following movements and if they are good enough, poor following movements will not affect the child’s activity.

**STRABISMUS IN CHILDREN WITH CEREBRAL PALSY:**

There is increased prevalence of squint in Children with Cerebral Palsy. In children with oculomotor problems, large angle strabismus may be present early during the first year of life. Although strabismus or squint may look disturbing, it seldom disturbs a child’s activity (25). The image received by the squinting eye is prevented from being used in the processing of visual information or only its peripheral parts are used.
NYSTAGMUS IN CHILDREN WITH CEREBRAL PALSY:

Nystagmus often takes the form of horizontal pendular eye movements, which seem to disturb fixation. Usually there is a direct relationship between the amplitude of the nystagmus and visual acuity. If visual acuity is only moderately decreased, nystagmus amplitude is small. If visual acuity is poor the amplitude is large.

ACCOMMODATION IN CHILDREN WITH CEREBRAL PALSY:

Oculomotor functioning, especially accommodative function is affected in Children with Cerebral Palsy as it is a motor function disorder. Accommodation in the normal visual system occurs as a combination of mechanical, visual and psychological stimuli and not solely linked with the motor function. Therefore, reduced accommodation is more associated with reduced visual acuity, presence of squint and high refractive errors (26, 27). In uncorrected myopia the child is able to see the near targets clearly and the distant ones blurredly.

ASSESSMENT OF VISUAL FUNCTIONS:

In general, assessment of visual acuity is done with high contrast test-charts; but, in real life, visual information is seldom at high contrast level, as we use intermediate and low contrast levels (26). For example, visual functioning at low contrast is very essential for communication as facial expressions require low contrast in order that we see thin lines at the edges of the mouth and eyes.

Assessment of visual field in Children with Cerebral Palsy is often a difficult task, where it demands cognitive skills and eye-hand coordination.
Diplegic or tetraplegic Cerebral Palsy conditions often cause problems in measurement of visual fields, as they have postural and fixation difficulties. In general, subjectively experienced field is better than the field depicted by clinical tests. Clinical tests measure visual field at a very low luminance level, 5-15 cd/m², which is a luminance level border between photopic and mesopic vision, but we use our vision at day light luminance levels in most tasks. The interpretations of perimetry examinations may or may not depict the functional visual field. As a result, usual clinical measurements are quite often misleading. Lighting conditions and varying colors ensure that few things are ever similar to testing situations. Hence when the test situation cannot be standardized; visual fields can be assessed approximately by confrontation technique in Children with Cerebral Palsy.

Children from their early ages start using colors in many daily activities. Children may experience a very mild defect that is rarely noticeable in daily activities. As colour vision defects can be caused by optic-nerve disorders, common in Children with Cerebral Palsy, it is vital to assess the colour vision(25).

In addition to reduced visual acuity, specific visual-perceptual impairment is also frequent in Children with Cerebral Palsy. In addition, poor control of extra-ocular muscles, inaccurate fixation, and the high prevalence of strabismus and nystagmus are all expected to be the causal factors for visual impairment, signifying an intricate multifactor problem(25).

The proportion of prematurity related Children with Cerebral Palsy is lower, birth and infection related factors are higher than in the Northern/ the
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developed country is from where we have most research. Therefore we must study the children from our population to understand the visual issues specific to them. Efficient visual functioning is very important for learning and development of children. Therefore, this study would focus on assessing the visual functions in Children with Cerebral Palsy. Since the prevalence of Cerebral Palsy is higher than other disabilities worldwide, we started looking at their associated problems. In that we found that there is higher prevalence of visual impairments which integrated us to conduct the study in Children with Cerebral Palsy (27)

Studies of economically developed countries documented that visual impairment occurs commonly in children with developmental disability. Health status of children in developed and developing countries like India is not same due to low income and poor early health services in the developing countries.

Data on normal visual development is not available on an Indian population. Data on Indian children are likely to be different from our Western counterparts due to genetic profiles and environmental factors. Hence we also planned assessing all the visual functions in normal children in our population with the age and sex match of Children with Cerebral Palsy data.
1.2 REVIEW OF LITERATURE

CAUSES FOR CEREBRAL PALSY IN INDIA:

In India, since the cause of Cerebral Palsy is most often in term babies or near term that have anoxia and birth related complications, or related to infection post natal, we cannot make assumption about co-occurring conditions and must study them separately.

In most developing countries, the primary cause of Cerebral Palsy is extreme prematurity and very low birth weight and co-occurring conditions are often sequela of related to these two conditions.

A study by Srivatsava et al in 1992 describes the causes for Cerebral Palsy India. He concluded that anoxia was consistently the most common natal or postnatal etiological factor for 0.55% monoplegia 1.29% paraplegia, 11.76%, quadriplegia, 6.07% diplegia and 0.55% ataxia from the study conducted at Safdarjung Hospital in New Delhi (29),(30).

K.Himmelmann et al (31),(32) in a population-based Indian study on 411 children with various types of Cerebral Palsy with 4 to 8 years of age found that 19% had severe visual impairment 40% had learning disability, 33% had epilepsy.
VISUAL PROBLEMS IN CHILDREN WITH CEREBRAL PALSY:

VISUAL FUNCTIONS & REFRACTIVE ERROR:

Further detailed study by Locasio et al in 1977 among 128 children with Children with Cerebral Palsy assessed visual acuity, refractive state, oculomotor functions, sensory fusion, and ocular health. Vision or ocular defects were found to be present in 86% of the children studied. Significant refractive errors and strabismus were the most common defects, being present in 50% or more of the children (28).

Several studies underline the high incidence of refractive errors and disturbances in the oculomotor system of children with Cerebral Palsy (29). Optometric examinations by Schieman et al (2005) at the Pennsylvania College of Optometry among 73 children with normal intelligence but with Cerebral Palsy revealed a higher than normal incidence of strabismus, significant refractive errors, amblyopia, nystagmus, and optic atrophy (30).

Peter Black, (1992), examined 120 Caucasian school children with severe Cerebral Palsy. Among the examined children, only 20% had ocular adnexa.52.5% had squint, 50% had significant refractive error, 11% had visual defects, 15% had anisometropic and strabismic amblyopia (31). The study emphasized the need for complete ocular assessment as a part of routine assessment in Children with Cerebral Palsy.

In 2007, Nikos Kozeis et al conducted a study in 105 Children with Cerebral Palsy aging 6 to 15 years in Hippokrateion Hospital, Thessaloniki on
visual function and perception. On assessing the best corrected distance visual acuity, he found that 59% had mild visual loss and 25.5% had moderate visual loss (28). Near visual acuity was also recorded, 70.1% had <6/6 and 38.4% had <6/9. Higher prevalence of hyperopia with 43.8% was recorded. The incidence of strabismus was 26.7% esotropia and 27.6% with exotropia. They found that 80.95% were with normal visual field and 94.28% had normal colour perception and 89.52% were free of ocular pathology (32). Stereopsis was abnormal in 85.71% of the children and 57.14% had markedly reduced visual perception. In the University of Helsinki, Finland, Erkkila et al conducted a study with 48 Children with Cerebral Palsy documenting squint and amblyopia status. They found that congenital esotropia was the commonest type and accommodative component of squint was present in few cases of their study. Amblyopia was found in 34 cases and was given necessary treatment. Given the range of visual issues in these studies, it is important to conduct a complete visual examination of all visual functions for Children with Cerebral Palsy.

Apart from the Western studies mentioned above, study by KatochSabita in the year 2007 in India help us to analyze the present scenario of Cerebral Palsy prevalence and incidence of ocular abnormalities in India (33). The study evaluated the ocular functions in 200 Children with Cerebral Palsy aging from 8 months to 21 years and concluded that 40% had visual problems. Myopia was recorded in 13.5% children. 5.5% had horizontal jerky nystagmus and 39% had strabismus. The study concluded that refractive error corrections could play a major role in treating Children with Cerebral Palsy.
ACCOMMODATIVE RESPONSE:

On literature search, there is evidence that accommodative function would be impaired in Children with Cerebral Palsy. Duckman assessed accommodative facility in 5 to 14 years age group of Children with Cerebral Palsy. He found that as this assessment was subjective, he had to exclude children with poor cognition ability as they may not express the response to the given stimulus (40). Therefore we planned assessing accommodative response objectively.

In 1996, Susan J. Leat could overcome this limitation by using dynamic retinoscopy to study accommodation in 43 individuals with Cerebral Palsy in 3 years to 35 years age group (41). She found that 42% had abnormal accommodative response for their age whereas 29% had 4 D or lesser than estimated amplitude of accommodation. They concluded that reduced accommodative function may be associated with reduced visual acuity. In our study, we adopted this method of assessment to ensure the objective measurement of accommodative status as we anticipated that many children would not be able to provide reliable subjective responses.

Paloma et al (1999), illustrated refractive status of 290 Children with Cerebral Palsy aging between 7 and 81 months from ASTRAPACE in Murcia (34). They found higher percentages of hyperopia in the spastic type of Cerebral Palsy. They concluded that normal emmetropization was less effective among these children.
The above literature points to a high incidence of visual issues in children with Cerebral Palsy around the world. However, only limited information is available on children with Cerebral Palsy in India (35). Different studies pointing to a range of visual issues make it imperative to gather data using a comprehensive vision examination using both subjective and objective measures to identify the visual issues among Children with Cerebral Palsy providing the motivation to conduct this study.
1.3 AIM AND OBJECTIVES

STATEMENT OF THE PROBLEM

For learning and development of children, good visual functioning is very important. Data on normal visual development is not available on an Indian population. Therefore, assessment of visual functions in Children with Cerebral Palsy and comparing with age matched normal children in the hospital-based Indian population (control group) is focused in our study.

AIM

To understand the type of refractive status and accommodative response among the children with Children with Cerebral Palsy on comparing with age-matched normal children in the hospital-based Indian population.

OBJECTIVES:

- To find out the visual functions namely, visual acuity, refractive status, visual fields and oculomotor status mainly accommodative response in children with Cerebral Palsy.
- To compare all the visual parameters collected in children with CP with the age-matched normal children in the hospital-based Indian population.
- To analyze the effectiveness of optical intervention on visual acuity and accommodative response.
SCOPE OF STUDY

This study mainly focuses on assessment of visual functions of children with Cerebral Palsy.

- Correction of their visual problems (refractive error) will assist them in pursuing their activities of daily living.