Section: Pediatrics

Original Research Article

A STUDY ON CLINICAL SPECTRUM OF CEREBRAL PALSY IN CHILDREN FROM 6 MONTHS-14 YEARS OF AGE GROUP WITH SPECIAL REFERENCE TO WEST SYNDROME

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Abstract

Background: Cerebral palsy is the most common motor disability in childhood. Various studies have been carried out from all over the world for prevalence, reporting estimates of CP ranging from 1.5 to >4 per 1000 live births or children of a defined age range. Cerebral palsy is not a defined, separate disease classification, but an umbrella term encompassing aetiologically diverse symptoms, which change with age. A group of lifelong mobility and posture issues that limit activities and are related to non-progressive abnormalities in the developing fetal or infant brain is collectively referred to as cerebral palsy. The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior as well as epilepsy and secondary musculoskeletal problems. **Materials and Methods:** This is a Prospective Observational Study was conducted in the Department of Paediatrics, M.K.C.G. Medical college and Hospital, Berhampur. The cases with Cerebral Palsy or/and West syndrome admitted to the Department of Paediatrics from 6 months to 14 years of age. Cases with cerebral palsy from 6 months to 14 years of age. Detailed history including the presenting complaints, associated precipitating factors, significant past history, and admission in SNCU at the time of birth for causes like Perinatal asphyxia, Preterm, NNH, Sepsis, and Meningitis was included in the study. Antenatal history, natal history, post-natal history, and developmental history with the head-to-toe examination are included. **Result:** The majority of the cases were presented as Spastic diplegic type (36.7%) followed by Quadriplegia (28.6%) and Infantile spasms (29%). The above table shows that NCCT is advised in 7 patients out of 98 patients out of which 4 are abnormal (4.1%) This shows that the maximum number of Cerebral Palsy cases 36 cases (52%) are Preterm deliveries and 48 cases (33%) are Term deliveries. But among 29 West syndrome cases only 7% are Preterm and 93% are Term deliveries. This reveals that Preterm is major risk factor for Cerebral Palsy than West Syndrome. This shows that out of 69 Cerebral Palsy cases 62 cases (90%) which is maximum form of seizures is GTCS type and 7 % is Myoclonic type and 3% is of Complex Partial type and no Infantile spasms were noticed. Among 29 West Syndrome cases 25 (86 %) presented with Infantile Spasms type and 4 cases (14%) presented with GTCS type. This explains that in West Syndrome cases most common form of presentation is Infantile Spasms. No Complex type or Myoclonic type are seen in this study. **Conclusion:** The majority of Cerebral Palsy and West Syndrome cases have an association with a significant past history of Birth asphyxia. The majority of Cerebral Palsy and West Syndrome cases were treated with Anti Epileptic Drugs and a combination of ACTH and Anti Epileptic Drugs for controlling seizure episodes. A multidisciplinary approach is the best method in the management of Cerebral Palsy and West Syndrome. Management is not

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curative; however, if provided optimally it can improve the quality of life of these children and their families. Physicians, in cooperation with the child, family, and members of a multidisciplinary team, can coordinate a complex care system to the maximal benefit of each child.

INTRODUCTION

Cerebral palsy is the most common motor disability in childhood.[1] Various studies have been carried out from all over the world for prevalence, reporting estimates of CP ranging from 1.5 to >4 per 1000 live births or children of a defined age range.[2-7] Cerebral palsy is not a defined, separate disease classification, but an umbrella term encompassing aetiologically diverse symptoms, which change with age. A group of lifelong mobility and posture issues that limit activities and are related to non-progressive abnormalities in the developing fetal or infant brain is collectively referred to as cerebral palsy. The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior as well as epilepsy and secondary musculoskeletal problems.[2]

A number of factors contribute to the development of CP including antenatal, natal, and postnatal. The majority of factors affect the brain during the antenatal period. Intrapartum such as perinatal asphyxia also plays an important role in the development of CP. Postnatal causes such as meningitis, head trauma, and hyperbilirubinemia contribute a large proportion of the disease. Maternal malnutrition, infection, and anemia are some of the preventable factors.

Cerebral palsy is broadly classified into two types Spastic - the most common ranging from 60% - 80%,[8,9] and Extrapyramidal. Spastic is further classified into Quadriplegia, Diplegia, and Hemiplegia. Extrapyramidal is further classified into Athetoid, Dyskinetic. Among the Spastic type Quadriplegia is the most common (34.9%) followed by Diplegia (28.7%) and Hemiplegia (21.9%) of cases.[10,11]

Cerebral palsy is mostly diagnosed based on motor function and postural issues that start in early childhood and last all the way to the end of life; these issues are non-progressive but do alter with age. The primary symptoms of cerebral palsy, motor function abnormalities, are typically accompanied by additional dysfunctions, including secondary musculoskeletal issues, seizures, and sensory, perceptual, cognitive, communication, and behavioral.[12-21]

Many metabolic and non-progressive genetic disorders may present with motor dysfunction resembling cerebral palsy. Such disorders are often characterized as CP mimics.[22-25]

Although the brain injury in CP is nonprogressive, the co-morbidities and the functional limitations change over time, affecting the functioning and quality of life. Therefore, it is essential for doctors to comprehend the numerous forms and co-occurring impairments of CP in order to assess the severity of the issue, include multidisciplinary treatment, introduce preventative measures, forecast functional limitations, and inform parents of the impact. The disorder was first identified as "West Syndrome" (WS) by William James West in his own son James Edwin (1840–1860),[26] and has since been referred to by a variety of terms, including infantile spasms (IS) to refer to the most pertinent clinical event and epileptic spasms (ES) to refer to the disorder's potential for onset outside of infancy. have been progressively proposed, first adopting the term infantile spasms (IS) in accordance with the most relevant clinical event, and later the term Epileptic Spasms (ES) since the disorder may have its onset outside infancy.[27,28]

West syndrome is an epileptic encephalopathy that starts between the age of 2 months and 12 months mostly >6 months. It consists of a triad of :1. Infantile spasms that usually occur in clusters particularly in drowsiness or about arousal.2.EEG showing Hypsarrhythmia which are high voltage slow chaotic background with multi-focal spikes.3.Neurodevelopmental delay or regression. West syndrome is classified into two types i) Cryptogenic (cause not known) ii) Symptomatic. In symptomatic west syndrome, a cause can be due to perinatal encephalopathies, (hypoxic-ischemic encephalopathy) malformation, or underlying metabolic disorders. An early diagnosis and a shorter lag time to start treatment represent the gold standards to get an effective response.

The Gold standard therapy for Infantile Spasms consists of the administration of Adrenocorticotrophic hormone [ACTH], Prednisolone, and Vigabatrin [VGB].[12,13] Early initiation of therapy with injection ACTH or high dose of prednisolone for 2 weeks and then tapering over 2 weeks resulted in a good outcome in the form of complete or partial cessation of Infantile spasms.

Early diagnosis and proper control of seizures with physiotherapy, occupational therapy, and speech therapy, the outcome remains better. Involvement and education of parents especially Mothers is an important tool as far as the management of a CP Child is considered. Early referral of the Child to DEIC (District Early Intervention Centre) for a multipurpose approach result in a satisfactory outcome.

MATERIALS AND METHODS

Study Design: Prospective Observational Study.
Study setting: Department of Paediatrics, M.K.C.G. Medical college and Hospital, Berhampur.

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Study Population: The cases with Cerebral Palsy or/and West syndrome admitted to the Department of Paediatrics from 6 months to 14 years of age.

Inclusion Criteria

Cases with cerebral palsy from 6 months to 14 years of age.

Children with features of West Syndrome that are admitted to IPD from 6 months-2yrs of age were also included in the study.

Exclusion Criteria

Children < 6months.

Children with Progressive Encephalopathy.

Syndromic children.

Cerebral Palsy with other chronic diseases such as Chronic Liver disease, Chronic Kidney disease, Hypothyroidism, and Congenital Haemolytic Anaemia, are not included in this study.

Methodology

The cases which have more or less satisfied the definition of Little Club Memorandum have been included in the present study. Patients of both sexes from age group 6 months to 14 years attending the Paediatrics Department of M.K.C.G Medical College and Hospital, Berhampur comprised the study group. Children clinically diagnosed with Cerebral Palsy and West Syndrome admitted in dept of paediatrics in MKCGMCH were evaluated in detail. Children who presented with seizures were included in the study.

Detailed history including the presenting complaints, associated precipitating factors, significant past history, and admission in SNCU at the time of birth for causes like Perinatal asphyxia, Preterm, NNH, Sepsis, and Meningitis was included in the study. Antenatal history, natal history, post-natal history, and developmental history with the head-to-toe examination are included.

The Cerebral Palsy Child and West syndrome will be further evaluated for Vision by Complete eye examination, fundoscopy/visual evoked response (VER) whichever is possible and also for Hearing by Brain Evoked Response Audiometry (BERA)/ Oto Acoustic Emissions (OAE).

During the time of hospitalisation detailed history of the type of therapy given to the children (any AED/ACTH+AED/PRED+AED/) is taken and the response to the therapy is also observed by ensuring the number of seizure episodes per day in CP children and number of Infantile spasms per day in WEST SYNDROME children accordingly the outcome of the children is seen.

Also, mothers are being counselled regarding the importance of physiotherapy and occupational therapy in CP and WEST children and are directed for DEIC for a better quality of life.

Data Collection

Data was collected from a reliable informant, from Mother, details of data were recorded in a structured CIF (Case Investigating Proforma) after taking consent from parents. Then the participants were subjected to necessary investigations and results were recorded systematically in a master chart.

Statistical Analysis Plan

The collected data in the master chart were taken for analysis and variables were subjected to percentage and mean SD. Data analysis was performed using Microsoft Excel and SPSS Software. Statistical significance was considered if the p-value is < 0.05.

RESULTS

[Table 1] shows the age of presentation of seizures which shows that the maximum number of cases presented with seizures within 6 months (58.2%) followed by 7-12 months (38.8%).

Table 1: Age-wise distribution of Infantile Spasm's (IS) / seizures: (n=98)

Age at onset of IS/SEIZURES	Number Percent (%)
(0-6) Months	57 (58.2%)
(7-12) Months	38 (38.8%)
(1-2) Years	2 (2.0%)
>2 Years	1 (1.0%)
Total	98 (100%)

Table 2: Sex distribution of cases (n=98)

Gender	Number PERCENT (%)
Male	60 (61.2%)
Female	38 (38.8%)
Total	98

Table 3: Distribution of Cerebral Palsy and West Syndrome as per Gestational age : (n=98)

Gestational Age	Number Percent (%)
PRE-TERM	38 (38.8%)
TERM	60 (61.2%)
Total	98

From the above table maximum cases are of Term Gestational age (61.2%) followed by pre term (38.8%)

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Table 4: Various stages of HIE in Cerebral Palsy and West Syndrome (n=98)

Stages of HIE	Number Percent (%)
NO HIE	37 (37.8%)
I	11 (11.2%)
II	46 (47%)
III	4 (4.1%)
Total	98

The above table shows that out of the children admitted for HIE majority graded under HIE 2 46 cases (47%) followed by HIE 1 11 cases (11.2%) followed by HIE 3 4 cases (4.1%).

Table 5: Types of CP (n=98)

Type of cerebral palsy		Number Percent (%)
Spastic extrapyramidal	Hemiplegia	2 (2.0%)
	Diplegia	36 (36.7%)
	Quadriplegia	28 (28.6%)
	Total	66 (67.3%)
	Choreoathetoid	4 (4.1%)
	Infantile spasms	29 (29.6%)
Total	N	98

The above table shows that majority of the cases were presented as Spastic diplegic type (36.7%) followed by Quadriplegia (28.6%) and Infantile spasms (29%).
The above table shows that NCCT is advised in 7 patients out of 98 patients out of which 4 are abnormal (4.1%)

Table 6: MRI findings in in Cerebral palsy and West Syndrome (n=98)

MRI Brain	Number Percent(%)
Normal Study	5 (5.1%)
Abnormal Study	87 (88.8%)
Not Done	6 (6.1%)
Total	98

The above table shows that out of 98 cases of my study 92 were advised to do MRI BRAIN out of which 87 (88.8%) were abnormal in the form of periventricular leukomalacia, intraventricular haemorrhage, cerebral atrophy, changes of HIE sequelae, followed by Normal (5.1%).

Table 7: Comparison

	Cerebral Palsy	Percentage	West Syndrome	Percentage
Rural	46	67%	19	66%
Urban	23	33%	10	34%
Total	69	100%	29	100%

This shows that out of 69 Cerebral Palsy cases maximum cases 46 (67%) cases are from Rural areas and 23 (33%) are from Urban areas. And Among 29 West Syndrome cases 19 cases (66%) are from Rural areas and 10 cases (34%) are from Urban areas. This shows that the maximum number of cases admitted to the hospital were from rural rather than urban backgrounds.

Table 8: Comparison

	Cerebral Palsy	Percentage	West Syndrome	Percentage
Pre-Term	36	52%	02	7%
Term	33	48%	27	93%
Total	69	100%	29	100%

This shows that the maximum number of Cerebral Palsy cases 36 cases (52%) are Preterm deliveries and 48 cases (33%) are Term deliveries. But among 29 West syndrome cases only 7% are Preterm and 93% are Term deliveries. This reveals that Preterm is major risk factor for Cerebral Palsy than West Syndrome.

Table 9: Comparison

	Cerebral Palsy	Percentage	West Syndrome	Percentage
GTCS	62	90%	4	14%
CPS	2	3%	0	0
Infantile Spasms	0	0	25	86%
Myoclonic Seizures	05	7%	0	0
Total	69	100%	29	100%

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This shows that out of 69 Cerebral Palsy cases 62 cases (90%) which is maximum form of seizures is GTCS type and 7 % is Myoclonic type and 3% is of Complex Partial type and no Infantile spasms were noticed. Among 29 West Syndrome cases 25 (86 %) presented with Infantile Spasms type and 4 cases (14%) presented with GTCS type. This explains that in West Syndrome cases most common form of presentation is Infantile Spasms. No Complex type or Myoclonic type are seen in this study.

DISCUSSION

In this study the total number of patients admitted to the department of pediatrics in M.K.C.G were 9375 out of which Cerebral Palsy and West Syndrome cases were 98 which accounts for 1% of prevalence this is consistent with Winter et al studies.[7] Whereas the studies of El-Tallawy et al., 2014, and Durkin et al. 2016 showed the prevalence of Cerebral Palsy ranges from 2.04/1000 live births and 2-10/1000 live births. This can be because they have taken prevalence in live births but this study shows prevalence in hospital admissions.

In this study, the most common presentation of all my 98 cases is Seizures (100%) followed by Fever for 22 cases (22.4%). As my study consists of both Cerebral palsy and West Syndrome. In the West Syndrome cases main presenting complaint is Seizures (Infantile Spasms) which is compatible with the study of Lux Al et al (2004) and Fisher RS et al (2017). Similar studies were seen by Kwong KL (1998) et al and Gururaj AK et al (2003). Epilepsy can be an indicator of the severity of neurological injury (quadriplegic CP) or cortical insult (hemiplegic CP).

In this study, the most common type of presentation of Seizures is GTCS type 62 cases (63.2%) followed by Infantile spasms 26 cases (26.5%) which is one of the triads to satisfy West Syndrome. Some children also presented with Complex (focal) type and Myoclonic type seizures, accounting for 2% and 5% respectively. But the studies of Jan MMS et al (2002) show Focal seizures with or without secondary generalization are most common with frequently focal EEG abnormalities. And Infantile Spasms being the most common were compatible with the studies of Lux Al et al (2004) and Fisher RS et al (2017). Out of 98 cases in this study, 89 cases (90.8%) were not associated with any loss of consciousness during the time of presentation which is considered to be one of the neurological manifestations, and 9 cases (9.2%) were associated with loss of consciousness for about 5 minutes.

In this study, among 98 cases about 89 cases (90.8%) were having a history of SNCU admission out of which the most common being perinatal (birth) asphyxia (delayed cry) 61 cases (62.2%) which is in near similar to the studies done by Sharma et al, Anwar S et al, and Singhi et al. Birth asphyxia is one of the perinatal risk factors which had a significant association with the development of CP, and it was noticed that out of 98 children 61 children had a history of birth asphyxia in cases which was found to be independently associated with risk of developing CP. The results supported the studies done by ozturk et al. (2006) and Erkin et al. (2007). The longitudinal study done by Lagunju et al. (2009) declared that birth asphyxia is the leading cause of the development of CP. Even though many old publications in 1993 and 1994 have been suggested that birth asphyxia was the leading cause of CP, early publications in 2006 reveal that birth asphyxia plays a less significant role in the development of CP. Also, the studies of Osborne JP et al (2010) show that the most common cause of West Syndrome was a perinatal brain injury, especially perinatal asphyxia. In contrast, prenatal causes which include cortical malformations, neurocutaneous syndromes, and genetic–metabolic disorders are the predominant aetiologies in the West. Followed by Preterm 38 cases (38.8%) followed by neonatal hyperbilirubinemia 4 cases (4.1%) followed by Septicemia 2 cases (2.0%) and Hypoglycemia 1 cases (1.0%) and 9 cases (9.2%) were without a history of SNCU admission.

CONCLUSION

Cerebral palsy and West Syndrome are one of the most common causes of Developmental delay in the childhood period, attracting researchers from different disciplines. Seizures were the most common presenting symptom (100%). Similarly, Generalised Tonic-Clonic Seizures (63%) were the most common type of seizure followed by Infantile Spasms (26.5%) The majority of Cerebral Palsy and West Syndrome cases have an association with a significant past history of Birth asphyxia. The majority of Cerebral Palsy and West Syndrome cases were treated with Anti-Epileptic Drugs and a combination of ACTH and Anti-Epileptic Drugs for controlling seizure episodes. A multidisciplinary approach is the best method in the management of Cerebral Palsy and West Syndrome. Management is not curative; however, if provided optimally it can improve the quality of life of these children and their families. Physicians, in cooperation with the child, family, and members of a multidisciplinary team, can coordinate a complex care system to the maximal benefit of each child.

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