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STUDY OF CLINICAL PROFILE OF PATIENTS WITH CEREBRAL PALSY

Paediatrics

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ABSTRACT

Introduction: *Cerebral Palsy* (CP) *is a diagnostic term used to describe a group of permanent disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems.*

Materials and Methods: Observational cross sectional study. Hospital based study.

Inclusion criteria: Patients admitted in pediatric ward of a tertiary health care center with a diagnosis of Cerebral Palsy.

Exclusion criteria: Repeatedly admitted patients

Conclusions: 68 patients with a diagnosis of cerebral palsy were admitted in the pediatric ward of a tertiary care unit. They constituted 14.2% of all neurological cases. **50** patients were enrolled in the study, rest were the readmissions for the same. *Spastic type* of cerebral palsy was most prevalent [36 (72%)] followed by Dyskinetic, Hypotonic, Ataxic and Mixed. In Spastic type, *Diplegia* was major presentation (47.5%), followed by Quadriplegia and Hemiplegia. 54% of patients presented with CNS complains in form of seizures and not attaining milestones. The patients with epilepsy had recurrent admissions. Most common Antenatal risk factor was Antepartum haemorrhage, most common Perinatal risk factor was Birth asphyxia, most common Post-neonatal risk factor was TBME. Mean Development quotient was (development quotient =33) and maximum delay was observed in Gross motor domain. Microcephaly was seen in 54% patients. Majority of patients with Spastic Diplegia had mild developmental impairment reflected by GMFCS levels I, II, III whereas majority of patients with Spastic Quadriplegia had severe impairment reflected by GMFCS levels IV, V. Most commonly affected joint was knee joint followed by ankle joint. MRI was abnormal in most patients. MRI is the neuro-imaging modality of choice. MRI findings correlate with the topographic distribution of cerebral palsy, but does not aid to etiological diagnosis. Abnormal EEG was also found in patients having epilepsy. Though widely talked multidisciplinary treatment modality for the patients of cerebral palsy, majority of patients in present study were found to be only on Physiotherapy and AEDs, and treatment of acute illnesses were sought.

KEYWORDS

Cerebral palsy, Spastic, Diplegia

INTRODUCTION:

Cerebral Palsy (CP) *is a diagnostic term used to describe a group of permanent disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems*[1,2,5]. Originally reported by Little in 1861 (and originally called 'cerebral palsies').

Overall *prevalence* of cerebral palsy is 2 to 2.5 per 1000 live births[1,2], and the *incidence* is 3.6 per 1000 children with male to female ratio are 1.4:1.0 .

Broadly Cerebral Palsy has certain types such as **S**pastic (monoplegia, diplegia, hemiplegia, quadriplegia) cerebral palsy, **D**yskinetic (choreoethetoid, dystonic) cerebral palsy, **A**taxic cerebral palsy, **H**ypotonic and **M**ixed cerebral palsy.

ETIOLOGY AND PATHOLOGY OF CEREBRAL PALSY

Risk factors for cerebral palsy can be broadly divided in three categories:-

1. Prenatal factors: 75%
2. Perinatal factors: 10-15%
3. Post neonatal factors: 10%

1. Prenatal factors: Prenatal factors are the most contributing factors for cerebral palsy, nearly 75% of all cases, which can be further described in three subheadings :-

a) Maternal factors:-

- Bad obstetric history, very short (<3 months) or very long (>3 years) interval between two pregnancies, positive family history of cerebral palsy
- Iodine deficiency, Iron deficiency, poor nutrition
- High grade fever, Urinary tract infections
- Diseases e. g. diabetes, hypertension, hyperthyroidism
- Intrauterine infections {rubella, toxoplasmosis, cytomegalovirus infection, HIV infection, herpes simplex infection}
- Teratogens e.g. Drugs, Radiations, Smoking, Alcohol, environmental toxins, cocaine and lead
- Fertility problems e.g. advanced age at conception, history of infertility, recurrent foetal wastage
- Poor antenatal care, low socioeconomic status.[2]

b) Foetal factors:-

- Multiple gestation
- Vanishing twin
- Abnormal fetal presentation
- Fetal thrombotic vasculopathy
- Intra Uterine Growth Retardation.[2,6]

c) Placental factors:-

- Chorioamnionitis, chorionic plate thrombi, histological lesions.

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2. Perinatal factors: These contribute up to 10-25% of total cases, they are :-

- Prematurity or VLBW (very low birth weight) is the single most important determinant of cerebral palsy
- Prenatal Asphyxia or birth asphyxia
- Ischemic perinatal stroke
- Hypoglycemia, dyselectrolytemias
- Hyperbilirubinemia

- Intra vascular and Intra cerebral bleeds
- Sepsis, pneumonia, meningitis
- Premature separation of placenta[1,2,6]

3. Post neonatal factors: First two years of life has utmost significance in postnatal period for developing cerebral palsy. These contribute up to 10% of total cases,

- CNS infections such as viral encephalitis, tubercular meningitis and pyogenic meningitis
- Head injuries and Seizures
- Gastro-enteritis with hypernatremic dehydration
- Hypoxic damage, hyperpyrexic damage.[1,10,13]

CLINICOPATHOLOGIC CORRELATION OF CEREBRAL PALSY[1,2,6,10]

Cerebral palsy subtype	Pathology	Underlying aetiology
1. Spastic diplegia	• Periventricular leukomalacia • Periventricular hemorrhagic venous infarction	• Prematurity • Ischemia • Infection • Thyroid disorder
2. Spastic quadriplegia	• Multicystic encephalopathy with cortical atrophy • Periventricular leukomalacia • Selective neuronal necrosis • Parasaggital cerebral injury • Cerebral malformations	• Perinatal/intrauterine hypoxic ischemic events • Endocrine / metabolic / genetic
3. Spastic hemiplegia	• Cerebral injury MCA territory (infarction necrosis) • Cerebral cortical malformations	• Genetic • Prenatal events like hypoperfusion-haemorrhage
4. Dyskinetic	• Basal ganglion • Status marmoratus • Bilirubin deposition	• Perinatal asphyxia • Neonatal hyperbilirubinemia (kernicterus)
5. Ataxic, hypotonic	• Cerebellar lesions • Enlarged ventricles	• Prenatal (genetic)

MATERIALS AND METHODS:

This was an observational cross sectional study conducted over 2 years at a tertiary care center.

Setting- Hospital based study.
Approval from the **Institutional Review Board (IRB)** was taken.
Inclusion criteria: Patients admitted in pediatric ward of a tertiary health care center with a diagnosis of cerebral palsy.
Exclusion criteria: Repeatedly admitted patients

- Admitted patients with the diagnosis of cerebral palsy were enrolled for the study.
- Written and informed consent obtained from their parents/guardians for using the details.
- Details of the patient collected from the Case Reporting Files (CRF).
- Proforma was filled.
- Data was analyzed by using SPSS software v 14.0
- Conclusion was derived.

OBSERVATIONS:

A total of 2750 patients were admitted in the paediatric ward of a tertiary care institute during, the study period of two years. 478 (17.50%) patients among these had C.N.S. disorders, amongst which Cerebral Palsy constituted 14.2%.

Table 1: Types of Cerebral palsy

Type of Cerebral palsy		No of patients		Percentage	
Spastic	Diplegic	17	36	34%	72%
	Quadriplegic	14		28%	
	Hemiplegic	5		10%	
Hypotonic		5		10%	
Ataxic		2		4%	
Mixed		2		4%	

In the present study spastic type of cerebral palsy was observed in 36(72%) patients followed by dyskinetic in 5(10%), hypotonic in 5(10%), ataxic in 2(4%) and mixed type in 2(4%). In spastic type, diplegia 17(34%) was major presentation followed by quadriplegia 14(28%) and hemiplegia 5(10%).

Table 2: Presenting Complaints

Presenting chief complaints	No. of patients
Seizures	27 (54%)
LRTI	12 (24%)
Not attaining mile stones	8 (16%)
Others	15 (30%)

Most common presenting complaint was seizures in 27 patients (54%), followed by LRTI 12(24%), not attaining milestones 8(16%).

42(84%) patients had repeated hospitalisations.

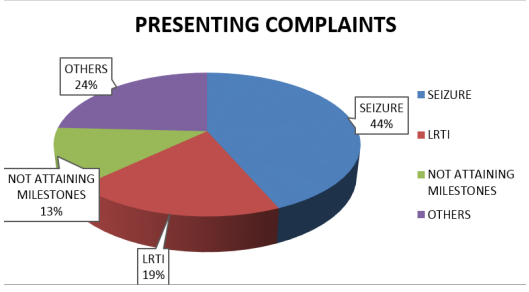


Table 3: Risk factors:

Risk Factors		No. of patients	%
Antenatal (Maternal history)	Antepartum Haemorrhage	11	22
	Fever	9	18
	Other illnesses	9	18
	Preecclamptic Toxaemia	8	16
	Multiple Gestations	5	10
Perinatal	Birth Asphyxia	24	48
	Low Birth Weight	16	32
	Neonatal Septicaemia	13	26
	Prematurity	10	20
	Neonatal Encephalopathy	7	14
	Neonatal Jaundice	6	12
Post-neonatal	TBME	5	10
	Trauma	1	2

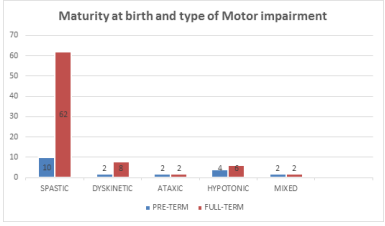
Most common antenatal risk factor was Antepartum haemorrhage 11(22%), followed by fever 9(18%). Consanguinity was seen in 26(52%) patients.

Most common perinatal factor was Birth asphyxia 24(48%) followed by Low birth weight (32%).

Most common post-neonatal factor was TBME 5(10%) followed by Trauma 1(2%)

Table 4: Maturity at birth and type of Motor impairment

Type of motor impairment	Pre-term n (%)	Full-term n (%)	Total no.
Spastic	5 (10)	31 (62)	36
Dyskinetic	1 (2)	4 (8)	5
Ataxic	1 (2)	1 (2)	2
Hypotonic	2 (4)	3 (6)	5
Mixed	1 (2)	1 (2)	2
Total no.	**10**(20)	**40** (80)	**50 (100)**



10(20%) patients had premature delivery while 40(80%) patients were full-term delivered. Out of which 5 (14%) out of 36 spastic cerebral palsy patients and 1(20%) out of 5 dyskinetic cerebral palsy patients were prematurely delivered. 31 (86%) out of 36 patients had spastic type of cerebral palsy and 4 (80%) out of 5 are dyskinetic cerebral palsy patients were full term.

Table 5: Head circumference

Head circumference	No of patients
Normal	23 (46%)
Microcephaly	27 (54%)

In this present study 27 (54%) patients had microcephaly, while 23 (46%) had normal head circumference.

Table 6: Development Quotient [DEVELOPMENT QUOTIENT

Type of motor impairment	Mean Development quotient (according to DDST-II)				
	Gross-motor development Quotient	Fine-motor development quotient	Personal /social development quotient	Language development quotient	Over all development quotient
Spastic	25	28	34	31	29
Dyskinetic	31	34	36	33	33
Hypotonic	37	44	50	35	43
Ataxic	50	55	50	50	51
Mixed	15	10	15	10	12
Overall	31	34	37	32	33

Mean development quotient was (Development quotient =33) and maximum delay was observed in Gross motor domain where mean development quotient was lowest of (Development quotient = 31) whereas personal-social domain was least affected (Development quotient =37). Patients with spastic cerebral palsy had maximum delay in gross motor domain with their mean development quotient being 25, while personal/social domain was least affected with mean development quotient of 34.

Table 7: Functional classification

GMFCS Level	No of patient n (%)	Sharma R., Sinha A. et al n(%)
I	2 (4)	26 (10.5)
II	8 (16)	20 (8)
III	15 (30)	31 (12.5)
IV	14 (28)	28 (11.5)
V	11 (22)	143 (57.5)

GMFCS (Gross Motor Functional classification System) Level is derived and 15 (30%) patients fall in GMFCS Level-III, 14 (28%) in level-IV, 11 (22%) in level-V, 8 (16%) in level-II and 2 (4%) patients fall in level-I.

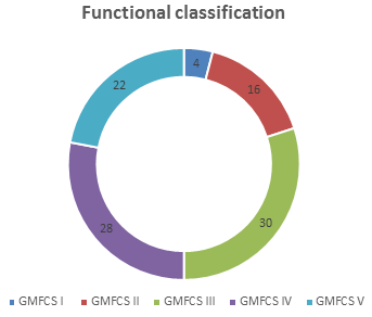


Table 8: Type of motor impairment and GMFCS levels

Type of motor impairment	GMFCS Level n (%)				
	I	II	III	IV	V
Spastic	1 (2.0)	5 (14)	10 (28)	10 (28)	10 (28)
Dyskinetic	0	1 (20)	2 (40)	2 (40)	0 (0)
Hypotonic	1 (20)	1 (20)	2 (40)	0 (0)	1 (20)
Ataxic	0	1 (50)	1 (50)	0 (0)	0 (0)
Mixed	0	0 (0)	0 (0)	2 (100)	0 (0)
Total	2 (4)	8 (16)	15 (30)	14 (28)	11 (22)

[$\chi 2 = 21.12, P = 0.18$]

Maximum severity of motor impairment as seen by GMFCS levels was seen in patients of Spastic and Mixed type of cerebral palsy. Majority of patients with Spastic Diplegia had mild impairment reflected by GMFCS levels I, II, III whereas majority of patients with Spastic Quadriplegia had severe impairment reflected by GMFCS levels IV, V.

Table 9: Co morbidities/ sequelas :

Co morbidities/Sequelas	Frequency	Percentage
No. of joint deformities	87	87%
Feeding dependency	40	80%
PEM	31	62%
Visual disability	13	26%
Behavioural disorders	11	22%
Hearing disability	8	16%

Maximum associated co-morbidity was joint deformities (87%), followed by feeding dependency (80%) and PEM (62%).

Table 10: Distribution of deformities according to topography

Joints (19)	No. of joint involved; n (%)			Total	
	Hemiplegic cerebral palsy (%)	Diplegic cerebral palsy(%)	Quadriplegic cerebral palsy(%)	50×2=100	%
Hip (2)	7 (50)	28 (56)	31 (86.1)	66	66
Knee(2)	11 (78.5)	43 (86)	33 (91.6)	87	87
Ankle(2)	10 (71.4)	40 (80)	28 (77.7)	78	78
Foot(2)	6 (42.8)	20 (40)	22 (61.1)	48	48
Spine(1)	0 (0)	4 (16)	4 (22.2)	8	8
Shoulder(2)	4 (28.5)	11 (22)	15 (41.6)	30	30
Elbow(2)	11 (78.5)	16 (32)	31 (86.1)	58	58
Forearm(2)	5 (35.7)	19 (38)	24 (66.6)	48	48
Wrist(2)	8 (57.2)	24 (48)	28 (56)	60	60
Hand(2)	4 (28.5)	15 (30)	16 (44.4)	35	35

Joints in the lower limb are more affected than the upper limb. Knee joint deformity (87%) was most common followed by ankle joint (78%) followed by hip (66%) and elbow (58%) followed by foot (48%) and forearm (48%).

Table 11: Neuroimaging:

MRI findings	No of patients
Periventricular leukomalacia	17 (38%)
Cortical atrophy	9 (20%)
Basal ganglia lesion	4 (9%)
Multicystic encephalopathy with cortical atrophy	2 (4.5%)
Perirolandic encephalomalacia	2 (4.5%)
Periventricular hemorrhagic venous infarct	2 (4.5%)
Miscellaneous	9 (20%)

MRI was done in 45 patients; Out of these 45 patients, 42 had an Abnormal M.R.I. and 3 had a normal.
Periventricular leukomalacia was observed in 17 (38%) patients, cortical atrophy in 9 (20%), basal ganglia lesion in 4 (9%).
CT scan was done in 14 patients, 09 had an abnormal report.
EEG was done in 40 patients; 32 had abnormal EEG.
BERA was done in 19 patients, 11 had normal hearing and 8 had SNHL.

Table 12: Treatment Modalities

TREATMENT RECEIVED	Number of patients
Physiotherapy	39(78%)
Muscle relaxant	18(36%)
Orthotics	5(10%)
OTHERS	17(34%)

All patients with CEREBRAL PALSY were receiving one or more type of treatment modalities; Physiotherapy in 39(78%) patients, muscle relaxants in 18(36%), Orthotics in 5(10%) and 17(34%) were on vitamins-supplements and tonics or some alternative therapy like homeopathic remedies, Ayurvedic remedies, treatment with traditional healers. 37(74%) patients had epilepsy and were taking AEDs.

DISCUSSIONS

- 68 patients with a diagnosis of cerebral palsy were admitted in the paediatric ward of a tertiary care unit. They constituted 14.2% of all neurological cases. **50** patients were enrolled in the study, rest were the readmissions for the same.
- Indian study done at Jalandhar district **Sharma R., Sinha A. et al**[5];

most common type of presentation was spastic CEREBRAL PALSY in 207(83.5%) followed by dyskinetic in 29(11.7%), ataxic in 5(2.3%) and mixed type observed in 7(2.5%) patients.48(96%) patients had H/o hospital delivery and 2(4%) had home delivery, 36(72%) patients had h/o normal vaginal delivery and 14(28%) were delivered by C.S. Results are consistent with the present study , in which most common type of presentation was spastic type of cerebral palsy, in which diplegic was the most common, followed by hypotonic, ataxic and mixed in that order.

- In the study **Singhi P.D., Ray M., Suri G. et al (n=1000)**[8], most common presenting complaint was delay in motor milestones (88.8%) followed by delayed speech (47.6%) and seizures is (28.1%) patients. In the present study seizure was the most common presenting complain in these patients followed by LRTI followed by not attaining milestones.
- Most common antenatal risk factor was antepartum haemorrhage, followed by fever. Most common perinatal factor was Birth asphyxia followed by Low birth weight. Most common post-neonatal factor was TBME followed by Trauma.
- In **Jacobsson, B., & Hagberg, G. (2004)**[15] about 28% of cerebral palsy cases are born very preterm, compared to 1% of all births. The main form of cerebral palsy related to low gestational age is spastic diplegia; congenital malformations are exceptional in these children. In this study 10(20%) patients had premature delivery while 40(80%) patients were full-term delivered. Out of which 5 (14%) out of 36 spastic cerebral palsy patients and 1(20%) out of 5 dyskinetic cerebral palsy patients were prematurely delivered. 31 (86%) out of 36 patients had spastic type of cerebral palsy and 4 (80%) out of 5 are dyskinetic cerebral palsy patients were full term.
- In the study **Singhi P.D., Ray M., Suri G. et al**[16], where it was seen in 886 (88.6%) patients had microcephaly. Similar results were seen in this present study also were more than half of the patients had microcephaly.
- Mean development quotient was 33 and maximum delay was observed in Gross motor domain where mean development quotient was lowest of 31 whereas personal-social domain was least affected 37. Patients with spastic cerebral palsy had maximum delay in gross motor domain with their mean development quotient being 25, while personal/social domain was least affected with mean development quotient of 34.
- In the study **Sharma R., Sinha A.et al**[5] majority of children were in GMFCS level V (57.5%). In this present study majority of patients fall in GMFCS Level-III & IV, followed by level-V, level-II and level-I respectively.
- Maximum severity of motor impairment as seen by GMFCS levels was seen in patients of Spastic and Mixed type of cerebral palsy. Majority of patients with Spastic Diplegia had mild impairment reflected by GMFCS levels I, II, III whereas majority of patients with Spastic Quadriplegia had severe impairment reflected by GMFCS levels IV, V. Similar data were seen with Distribution of 'type of motor impairment' by GMFCS; **data from Ontario Motor Growth study (Rosenbaum et al. 2002)**[17].
- In **Pruitt, D. W., & Tsai, T. (2009)**[18] study maximum associated co-morbidity was epilepsy and visual impairment followed by cognition, hearing impairment, sleep, pain and GI problems in these order. In the present study maximum associated co-morbidity was joint deformities, followed by feeding dependency and PEM.
- In the study **Sharma R., Sinha A.et al**[19]**;** in upper limb, forearm deformity was most common (42.3%) followed by wrist (39.3%) and in lower limb; ankle (59.8%) was most commonly affected followed by knee (55%). In the present study joints in the lower limb are more affected than the upper limb. Knee joint deformity was most common followed by ankle joint followed by hip and elbow.
- In **Martin Bax, Clare Tydeman, Olof Flodmark**[20] studies among children with clinical evaluation, 351 (84%) had a brain MRI scan assessed for the study. The MRI scans showed that white matter damage of immaturity (WMDI, including PVL) was the most common finding (42.5%), followed by basal ganglia lesions (12.8%), cortical/sub-cortical lesions (9.4%), malformations (9.1%), focal infarcts (7.4%), and miscellaneous lesions (7.1%). Normal MRI findings were present in 11.7%.In this study periventricular leukomalacia was observed in 17 (38%) patients, cortical atrophy in 9 (20%), basal ganglia lesion in 4 (9%). CT Scan was done in 14 patients, 09 had an abnormal report. EEG was done in 40 patients; 32 had abnormal EEG.BERA was done in 19 patients, 11 had normal hearing and 8 had SNHL.
- All patients with CEREBRAL PALSY were receiving one or more type of treatment modalities; Physiotherapy in 39(78%) patients, muscle relaxants in 18(36%), Orthotics in 5(10%) and 17(34%) were on vitamins-supplements and tonics or some alternative therapy like homeopathic remedies, Ayurvedic remedies, treatment with traditional healers. 37(74%) patients had epilepsy and were taking AEDs. In **Krigger KW**[21] study also have similar results showing physiotherapy as an effective treatment modality.

CONCLUSIONS:

Spastic type of cerebral palsy was most prevalent followed by dyskinetic, hypotonic, Ataxic and Mixed. In spastic type, diplegia was major presentation, followed by quadriplegia and hemiplegia. More than half of patients presented with CNS complaints in form of seizures and not attaining milestones. The patients with epilepsy had recurrent admissions. Most common antenatal risk factor was Antepartum haemorrhage, most common perinatal risk factor was birth asphyxia, and most common post neonatal risk factor was TBME. Microcephaly was seen in more than half of the patients.

- Majority of patients with Spastic Diplegia had mild developmental impairment reflected by low GMFCS levels whereas majority of patients with Spastic Quadriplegia had severe impairment reflected by high GMFCS levels. Most commonly affected joint was knee joint followed by ankle joint.
- MRI was abnormal in most patients. MRI is the neuroimaging modality of choice. MRI findings correlate with the topographic distribution of cerebral palsy, but does not aid to etiological diagnosis. Abnormal EEG was also found in patients having epilepsy.
- Though widely talked multidisciplinary treatment modality for the patients of cerebral palsy, majority of patients in present study were found to be only on Physiotherapy and AEDs, and treatment of acute illnesses were sought.

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