Neurological Subtypes of Cerebral Palsy and Functional Classification

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Abstract
Introduction: Cerebral palsy (CP) is the most common developmental disability in children. There are several classification systems for cerebral palsy. A uniform system based on functional ability is needed for categorization and comparison.

Aim and Objectives: To classify the common neurological subtypes of cerebral palsy, to categorize them functionally as per the Gross motor functional classification system (GMFCS) for mobility and the Manual Ability Classification system (MACS) for manual function and to assess the utility of the functional classification.

Materials and Methods: A hospital - based study conducted over a period of 18 months on hundred children with cerebral palsy aged 2-12 years. The children were categorized into neurological subtypes and functional levels using GMFCS and MACS and the data was analyzed.

Results: Of the 100 cases, spastic quadriplegia, hemiplegia and diplegia, dyskinetic CP and mixed type accounted for 47, 25, 17, 7 and 5% respectively. Spastic quadriplegia was the commonest type. The majority of patients were in GMFCS and MACS levels 5. The association between the neurological subtypes and GMFCS and MACS levels was found to be statistically significant.

Conclusion: A classification system based on functional limitation is practical and easy to use.

Keywords: Cerebralpalsy, neurological subtypes, functional classification, GMFCS, MACS.

Introduction
Cerebral palsy (CP) was first described by Dr Little in 1861 [¹]. There has been significant progress in the evaluation and treatment of cerebral palsy and the associated impairments in the last few decades. Assessment of activity restriction is an important part of CP evaluation [²]. Gross motor functional classification system (GMFCS) and Manual Ability Classification system (MACS) are international systems used to evaluate the functional abilities of patients with cerebral palsy. They are also used in the rehabilitation of the patient and in assessing the improvement after different modes of treatment like physiotherapy and surgery.

GMFCS was introduced by Palisano et al for assessment of ambulation [³]. It classifies children into 5 levels of increasing severity. Assessment is based on the usual activities of the child at school, home or community. MACS, used to assess upper limb and hand function was introduced by Eliasson [⁴]. MACS also classifies patients like GMFS into 5 levels of severity. Grading is based on history and observation of upper extremity.
performance when handling objects in daily activities. Relevant and age appropriate activities such as eating, dressing, and writing, using objects within reach of the child are noted. It does not distinguish between the abilities of individual hands, but assess the overall performance, using both hands.

Methods
This was a cross sectional study conducted in Government Medical College Hospital, Thrissur over a period of 18 months. One hundred consecutive patients aged 2-12 years with a diagnosis of cerebral palsy attending the neurodevelopment clinic were included in the study. Patients with progressive neurological disorders, children with neurological problems due to insults to the brain acquired after 2 yrs of age and those without tone abnormalities and activity limitation were excluded.

Detailed clinical examination included tone and topographic distribution. The children were classified according to their activity limitation using GMFCS and MACS. The relationship of the neurological subtypes with GMFCS and MACS levels was assessed and the association between the type of CP and their GMFCS and MACS levels was analyzed.

Results
Of the 100 children with CP enrolled in the study 79% were born term and 21% were preterms. Birth weight ranged from 1.2 kg to 3.5Kg; the mean birth weight was 2.39.kg. The children were of age 2 to 12 years and the median age was 7 years. 63% were below 5 years. The most common neurological subtype of CP was spastic quadriplegia (47%) followed by spastic hemiplegia (25%) and diplegia (16%). The dyskinetic (7%) and mixed CP (5%) were the least common.

51% of the 47 patients with spastic quadriplegia were at GMFCS level 5, while 8.5%, 23.4%, 17% and 9% were at levels 4, 3, 2 and 1 respectively. Among the 16 cases of diplegic CP, none were in GMFCS levels 4 or 5, the majority (68.7%) were in level 2. Most of the children with hemiplegia were in levels 1 (32%) & 2 (48%). More than half (57.1%) the cases with dyskinetic CP were at level 5 as were 80% with mixed CP. The association between the GMFCS levels and the subtypes of CP was assessed by chi-square test and found to be statistically significant (Pearson chi square test value \(-64.61, df=16, p value = 0.000\)).

On assessing hand function in the 100 children 32% belonged to MACS level 5 with 19, 25.13, and 11% at levels 1 to 4. This difference found in the study was assessed by chi-square test. (test value 55.703 df -16, p value = 0.000) and found to be statistically significant.

Among the cases with spastic quadriplegia, nearly half (48.9%) had MACS level 5. There were 62.5% at MACS level 1 and 37.5% at level 2 among those with diplegia but none at MACS levels 3, 4 & 5. Most children with hemiplegic CP belonged to levels 2 (32%), 3 (28%) and 1 (24%) respectively. Majority of children with dyskinetic and mixed CP were at level 5.

Figure 1- Neurological subtypes of cerebral palsy

SQ – Spastic quadriplegia SD – spastic diplegia SH – spastic hemiplegia .DKCP- dyskinetic CP MCP – mixed CP
Table 1: Gross Motor Functional Classification System (GMFCS) levels of each subtype of Cerebral Palsy

<table>
<thead>
<tr>
<th>Level</th>
<th>Functional limitation</th>
<th>Total 100</th>
<th>SQ = 47 No</th>
<th>HP = 25 No</th>
<th>DP = 16 No</th>
<th>Dys = 7 No</th>
<th>Mixed = 5 No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level 1</td>
<td>Walks without limitations</td>
<td>10</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>32</td>
<td>2</td>
</tr>
<tr>
<td>Level 2</td>
<td>Walks with Limitations</td>
<td>32</td>
<td>8</td>
<td>17</td>
<td>12</td>
<td>48</td>
<td>11</td>
</tr>
<tr>
<td>Level 3</td>
<td>Walks with hand-held mobility device</td>
<td>16</td>
<td>11</td>
<td>23.4</td>
<td>0</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Level 4</td>
<td>Self-mobility with limitations; may use powered mobility</td>
<td>9</td>
<td>4</td>
<td>8.5</td>
<td>4</td>
<td>16</td>
<td>0</td>
</tr>
<tr>
<td>Level 5</td>
<td>Wheelchair bound</td>
<td>33</td>
<td>24</td>
<td>51</td>
<td>1</td>
<td>4</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 2: Manual Ability Classification System (MACS) levels of each subtypes of CP.

<table>
<thead>
<tr>
<th>Level</th>
<th>Upper limb and hand function</th>
<th>Total 100</th>
<th>SQ = 47 No</th>
<th>HP = 25 No</th>
<th>DP = 16 No</th>
<th>Dys = 7 No</th>
<th>Mixed = 5 No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level 1</td>
<td>Handles objects easily and successfully</td>
<td>19</td>
<td>2</td>
<td>4.2</td>
<td>6</td>
<td>24</td>
<td>10</td>
</tr>
<tr>
<td>Level 2</td>
<td>Handles most objects but with reduced quality or speed of achievement</td>
<td>25</td>
<td>9</td>
<td>19.1</td>
<td>8</td>
<td>32</td>
<td>6</td>
</tr>
<tr>
<td>Level 3</td>
<td>Handles objects with difficulty, needs help to prepare or modify activities</td>
<td>13</td>
<td>5</td>
<td>10.6</td>
<td>7</td>
<td>28</td>
<td>0</td>
</tr>
<tr>
<td>Level 4</td>
<td>Handles a limited selection of easily managed objects in adapted situations</td>
<td>11</td>
<td>8</td>
<td>17</td>
<td>2</td>
<td>8</td>
<td>0</td>
</tr>
<tr>
<td>Level 5</td>
<td>Does not handle objects and has severely limited activity to perform even simple actions</td>
<td>32</td>
<td>23</td>
<td>48.9</td>
<td>2</td>
<td>8</td>
<td>0</td>
</tr>
</tbody>
</table>

Discussion

The most common type of cerebral palsy was spastic quadriplegia, which accounted for 47% of the cases followed by hemiplegic CP (25%) and diplegia (17%) the remaining 11% were constituted by dyskinetic and mixed cerebral palsy.. Spastic quadriplegia is the commonest subtype of cerebral palsy reported from India and other developing countries \[5,6,7,8\]. Studies from the West report a higher incidence of spastic diplegia and hemiplegia reflecting improved neonatal care and greater survival of preterm babies \[9,10,11\]. Singhi et al compared a large cohort of 1212 children with CP registered in the last 10 years in their rehabilitation center with their previous study of 1000 children from the same center. Spastic quadriplegia was the commonest type of CP (51.5%) although lesser in number (61%) than in the previous decade .This indicates a change in spectrum with an increase in diplegic and a decrease in quadriplegic CP in developing countries also \[12\]. Data obtained from a cohort of 374 children from the Victorian Cerebral Palsy Register (VCPR) Australia by Howard et al showed the topographical distribution as hemiplegia (35%), diplegia (28%) and quadriplegia (37%) \[13\]. Gross motor function varied from GMFCS level I (35%) to GMFCS level V (18%). They had fewer patients at level 5 compared to our levels of 10% at GMFCS level 1 and 33% at level 5 reflecting higher morbidity here. Howard et al have commented that comparison of motor types of CP and topographical distribution is difficult because of lack of consensus with classification systems but easier with GMFCS as gross motor function is similar in populations of children with CP and GMFCS provides a valid and reproducible method to describe gross motor function in children with CP using a universal language.

The GMFCS levels of the cases belonging to each subtype when assessed, among the 47 patients with spastic quadriplegia, more than half were in GMFCS levels 5 and none at GMFCS level 1.
This shows that most of the quadriplegic patients were unable to maintain antigravity head and trunk postures and could not control their limb and trunk movements. A similar distribution in the GMFCs levels among patients with spastic quadriplegia with none at level 1 has been reported by Gunel M.K et al.[14]

Among the 16 cases of diplegic CP, 68.7% belonged to GMFCS level 2. These children were able to walk with limitation and needed the help of hand held devices. The remaining diplegic patients belonged to level 3 and level 1. There were no cases at GMFCS levels 4 & 5. A similar pattern has been observed by others also.[14] The majority of the cases with spastic hemiplegia were in GMFCS levels 1 & 2 similar to those with diplegia. They had better gross motor function than hand function and most were able to walk without limitation or with the help of a stick.

Among the cases with dyskinetic CP, more than half (57.1%) had GMFCS level 5 and those with mixed CP, 80% were at level 5 indicating severe motor disability. Because of the abnormal tone and posture these children are usually severely incapacitated.[15]

MACS level was most affected in children with more neurological impairment as in spastic quadriplegia, dyskinetic and mixed CP. Upperlimb function was least affected in children with spastic diplegia. As MACS levels assess the overall performance of the child in doing activities with both hands the hemiplegics were at levels 1,2,3 (84%) with fewer at higher levels.[15] That limitations in hand function are common in all types of CP with characteristics of the disability varying considerably between different CP subtypes was also observed by Camahan et al.[16]

In a population of 367 children with CP aged 4 to 14 years, they have reported that 60% of children had more than minor problems with hand function. (>MACS I) and (MACS I-II) was noted in 87% of children with spastic hemiplegia and in 63% of children with spastic quadriplegia CP. Most of the children at MACS 5 were also at GMFS level 5 suggesting a strong correlation between the two classifications a fact observed in other studies too.[17]

Conclusion
GMFCS and MACS grading of cerebral palsy correlates well with topographic classification. Being easy to use the functional classification should be adopted universally for comparison, assessment of improvement and for research purposes.

References