

Original Research Article

Pain symptoms in patients with severe cerebral palsy: Prevalence among patients with higher degree of locomotor impairment

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Abstract

Purpose: To evaluate the presence of pain in patients diagnosed with severe cerebral palsy (CP) according to the degree of motor function impairment.

Methods: A cross-sectional study was conducted on students of the Association of Parents and Friends of Exceptional Children (APAE) diagnosed with cerebral palsy and with severe locomotor disability (GMFCS levels IV and V). The study included students of both genders and of any age. After signing the consent form, a structured questionnaire was administered to parents or caregivers to collect data on the history of the illness and the Paediatric Pain Profile (PPP). A score ≥ 14 out of a possible 60 points suggests the presence of pain.

Results: A total of 93 subjects were evaluated. Of the 44 subjects classified with GMFCS level IV, 12 (21.4 %) suffer pain and among the 49 subjects classified with level V, 44 (78.6 %) had pain symptoms (ICDDN ≥ 14 , PR = 3.29, 95 % CI 2.01 - 5.38, $p < 0.01$).

Conclusion: The results show that pain is prevalent in individuals with severe CP. Furthermore, it is more prevalent in patients with a higher degree of locomotor impairment (GMFCS – level V).

Keywords: Cerebral palsy, Pain measurement, Locomotor disability, Gross motor function classification system (GMFCS)

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INTRODUCTION

Cerebral palsy (CP) is defined as a permanent disorder of movement and posture due to a non-progressive brain defect or lesion early in life. Considered as the most common form of physical impairment in the pediatric age group [1], the disorder has a high prevalence, affecting approximately 4 out of 1,000 children born alive in developed countries [2].

Regarding the etiology, there is still no determining factor for the occurrence of CP. However, the conditions that contribute most to its occurrence are hypoxemia and ischemia due to endogenous or exogenous factors, such as genetic and chromosomal abnormalities, complications during pregnancy or childbirth, infections, parasitoses, among others. Depending on the level, there may be impairment of cerebral neurological development. As a consequence, cognitive development is limited

and does not accompany chronological age, reflecting a poor quality of life over time [3-6].

The clinical condition of the patient with CP may progress to the development of muscle contractures and musculoskeletal manifestations such as hip subluxations/dislocations, joint and spinal disorders, bone deformities, osteopenia, osteoporosis, esophagitis, gastroesophageal reflux, recurrent infections, otitis and convulsions [3,7].

The use of instruments for assessment and classification of patients with CP is very important for planning clinical interventions aimed at improving the quality of life of these patients [3,8]. The Gross Motor Function Classification System (GMFCS), which uses patients' locomotion as a parameter, classifies the patients affected by CP according to their functional independence [9]. This instrument uses a five-level ordinal scale to determine, in descending order, the level of independence and functionality of individuals with CP. The impairment levels of individuals classified as levels I and II by GMFCS are considered as mild, those of level III, as moderate, and those of levels IV and V, have their functional impairment classified as severe [10,11].

In addition to the functional impairment presented by CP patients, pain, caused by the progression of clinical conditions in these patients, is a prevalent subjective symptom, lasting at least 9 h per week, in approximately 50 % of the cases. In the patient, this symptom can result in changes in behavior, appetite, sleep patterns, ambulation, leisure and mood. Therefore, it is important to detect pain to treat this symptom adequately [5]. Although self-report is considered the gold standard method for assessing subjective states, individuals with CP, because they have limitations in communication skills, depend on the reports of parents or caregivers and assessment instruments to detect pain [12].

Currently, the Paediatric Pain Profile (PPP) is applied to assess pain in children with severe CP [12]. PPP was recently adapted and cross-culturally validated for Portuguese [13] under the name of Inventário de Comportamentos da Dor na Deficiência Neurológica (ICDDN). Although this instrument was developed for the classification of pain in children, it is known that currently the ICDDN can be used for classification of pain in individuals with CP of all ages. The literature demonstrates that, because the lesion that causes CP is non-progressive, the patient remains at the same level of classification

over the years, which also makes ICDDN a useful tool for the classification of pain in adults [13,14]. The ICDDN is a questionnaire, answered by parents and caregivers, consisting of twenty items that describe pain-related behaviors in patients. Each item was evaluated using a scale, in which the sum of scores ≥ 14 suggests the presence of pain, providing subsidies for planning clinical interventions aimed at the relief of this symptom in patients [13].

Clinical practice suggests an association between the degree of functional impairment and the intensity of painful symptoms presented by patients with CP. However, although valid and reliable instruments for assessment of these parameters are available, studies on the presence of pain and the correlation of this symptom with the degree of functional impairment presented by these patients are still scarce. As far as we know, only the study conducted by McKearnan *et al* [15] which report events related to the presence of pain in children with CP. Nevertheless, the occurrence of pain in patients with CP presenting impairment of motor function, regardless of age, has not been investigated.

This study evaluated the presence of pain in patients with a diagnosis of severe CP according to degree of motor function impairment.

METHODS

Study design and subjects

This observational cross-sectional study was conducted following approved by Research Ethics Committee of the University of Southern Santa Catarina (ref no. 25088313.4.0000.5369 - CAI/CEP/UNISUL 491.102) and are in accordance with the recommendations of the International Ethical Guidelines by the Council for International Organizations of Medical Sciences (CIOMS/2016) [16]. It was conducted at the Association of Parents and Friends of Exceptional Children widely known by the acronym APAE in Brazil) of the cities of Tubarão and Orleans, State of Santa Catarina, Brazil, over a six-month period (April to October, 2014). After permission was obtained from the relevant authority at the EPFA, all the patients with a diagnosis of CP were identified. They were then classified according to the GMFCS scale. Patients with severe locomotor impairment, classified as GMFCS levels IV and V regardless of gender and age, were selected for this study. Excluded were patients with diagnosis of severe CP, those whose parents or caregivers did not understand the content of the consent form

and hence unable to respond adequately to the questions asked by the researchers, or those who did not agree to participate in the study.

The parents or caregivers of the selected patients signed the consent form to participate in the study and a structured questionnaire was applied to each by the authors, in an interview to obtain demographic information and the conditions presented by the patient.

The information obtained included: gender, age, pre-, peri- and post-natal history, history of surgical procedures and previous hospitalization, presence of gastroesophageal reflux, pathological postural deviations, other pathological conditions, use of medications, personal care skills, and verbal communication skills appropriate for the age. Information was also obtained regarding the presence of pain in these patients using the ICDDN.

Data analysis

The data were subjected to statistical analysis using Epi-info 3.5.2[®] software. Results were expressed as absolute numbers, proportions, means and standard deviations. The variables were compared between groups using the Chi-square test for categorical variables, Student's t-test and one-way analysis of variance (ANOVA) followed by Bonferroni test whenever appropriate. $P < 0.05$ was considered as statistically significant.

RESULTS

Demographic data

A total of 93 patients with severe CP participated in the study 52 (55.9 %) were male while 41 (44.1 %) were female 44 patients in level IV and 49 classified in level V, according to the GMFCS. The average age of the patients was 23 years in the range of 1 to 57 years with 53.8 % of patients aged > 18 years.

Characteristics of pre-, peri- and post-natal periods

With respect to the history of the period prior to birth, 54 (58.1 %) patients had regular prenatal follow-up. 57 (61.2 %) patient of the total presented some complication during their development in pregnancy, including hemorrhage (35 %), prolonged expulsive period (21 %) and premature rupture of the membrane associated with preterm labor (15 %), which were the main gestational intercurrents observed.

Considering the type of delivery, 26.9 % of the patients studied were born by cesarean delivery. 23.7 % were born by vaginal delivery at home, 11.8 % were born by vaginal delivery at the hospital level using forceps, and 37.6 % were born by vaginal delivery at the hospital level without the use of forceps. The mean gestational time was 37 ± 3.38 weeks.

The mean Apgar scores at the first and fifth minutes were 5.9 ± 2.23 and 6.6 ± 1.47 points, respectively, being considered intermediate [17]. Mean birth weight was 2.559 ± 1.063 g. Most of the patients (64.8 %) presented normal weight and 35.2 % presented low birth weight (< 2,500 g). Neonatal complications were reported in 85 patients (91.4 %), with neonatal hypoxia/perinatal anoxia described for 60 patients (70 %). The mean age at which CP-related symptoms appeared was 10.8 ± 2.61 months.

It was shown that 87 of the patients (93.5 %) were admitted to the Intensive Care Unit and 44 (47.3 %) were submitted to a surgical procedure. Among other comorbidities, the prevalence of epilepsy was observed in 76 patients (81.7 %). Of the 93 patients evaluated, 56 (60.21 %) presented behaviors suggestive of pain (ICDDN ≥ 14), 12 (27.3 %) of them were classified as level IV and 44 (89.7 %) as level V, according to GMFCS (Table 2).

In relation to the use of medication, 80 (86 %) patients used medication therapy, and anticonvulsants represented the main class of medication used (76 %). Among other drug treatments, 51 (62 %) patients used gastric protector and 48 (59 %) patients used some drugs with sedative action. No therapy with direct or specific analgesic property was found.

Considering personal care skills, 68 (73.1 %) patients were unable to maintain personal hygiene independently and none of them had adequate verbal communication skills for the age.

Demographic characteristics and clinical conditions of patients with severe cerebral palsy

Among the demographic variables evaluated (Table 1), the age and sex of the patients had no significant association with the occurrence of pain (data not shown).

However, the higher degree of locomotor impairment (GMFCS - level V) indicated the presence of behaviors suggestive of pain

(ICDDN ≥ 14) in patients with severe CP (PR = 3.29, 95 % CI 2.01 - 5.38, $p < 0.01$; Table 2).

Table 1: Presence of behaviors suggestive of pain (ICDDN ≥ 14) detected in patients with severe cerebral palsy (CP) according to demographic characteristics

Variable	Presence of behaviors suggestive of pain (ICDDN ≥ 14)
Age < 18	62 %
Age ≥ 18	58.1 %
Female	70.7 %
Male	51.9 %

ICDDN = Inventory of Pain Behaviors in Neurological Disability

Furthermore, a greater presence of symptoms suggestive of pain (ICDDN ≥ 14) was observed in the patients with clinical conditions such as postural deviation (PR = 2.13, 95 % CI 1.08 - 4.22, $p < 0.01$), hip dislocation (PR = 1.9121, 95% CI 1.39 - 2.62, $p < 0.01$), snoring (PR = 4.4767, 95 % CI 0.72–27.67, $p < 0.01$), difficulty in swallowing (PR = 7.39, IC 95% 1.97 - 27.74, $p < 0.01$), gastroesophageal reflux disease (PR = 2.2831, 95 % CI 1.14 - 4.53, $p = 0.01$) and bruxism (RP = 1.8076, 95 % CI 0.87 - 3.71, $p = 0.05$).

DISCUSSION

The main data of this study showed that pain is a prevalent symptom in individuals with severe CP and evidence shows that the presence of pain was more prevalent in the patients with a higher degree of locomotor impairment (GMFCS – level V).

The prevalence of male individuals with severe CP is in agreement with that found by Chagas *et al* and Cotter *et al* [8,17]. However, although there was a higher number of males among the

individuals with CP, it was observed that the gender of individuals had no influence on the presence of pain in these patients. Likewise, the age of the patient also had no influence on the prevalence of this symptom. As observed, only a small percentage of the evaluated patients were born preterm. The literature shows that, despite the vulnerability of the premature newborn, prematurity is not essential for the occurrence of CP [1]. The data corroborate previous findings that CP presents a high prevalence in term newborns, with the main cause being perinatal hypoxia/anoxia, which is responsible for the largest contingent of cerebral impairment, reflected in a low Apgar score presented by the newborn [1,5].

Most patients studied herein were born with weight classified as normal. Similarly, Surman *et al* [18] showed that individuals with higher birth weight developed CP in the most severe form.

These authors indicated that, although the risk of CP is higher in individuals with low birth weight, there is a clear relationship between the severity of motor impairment and the birth weight of the patients, with a higher proportion of those born at the ideal weight or greater being more severely affected.

The median age of symptom onset indicative of cerebral palsy in our study was 10.8 months. The findings are similar to a study conducted in Denmark, which showed that the mean age of diagnosis for CP, associated with the onset of symptoms such as high degree of motor disability, abnormalities on cerebral ultrasound and the presence of epilepsy, was around 11 months of age [19]. As verified in our study and reported in other studies, epilepsy is a prevalent comorbidity in patients with CP [4,19-21].

Table 2: Presence of symptoms suggestive of pain (ICDDN ≥ 14) detected in patients with severe cerebral palsy (CP) according to degree of motor function impairment (GMFCS IV and V) and clinical conditions presented

Variable	Presence of behaviors suggestive of pain in % (ICDDN ≥ 14)	PR (CI 95%)	P-value*
GMFCS IV	27.3		
GMFCS V	89.7	3.2900 (2.01 - 5.38)	<0.01
Bruxism	64.6	1.8076 (0.87 - 3.71)	0.05
Snoring	64.0	4.4767 (0.72 - 27.67)	0.01
Postural deviation	67.6	2.1300 (1.08 - 4.22)	<0.01
Gastroesophageal reflux disease	68.5	2.2831 (1.14 - 4.53)	0.01
Difficulty in swallowing	74.0	7.3900 (1.97 - 27.74)	<0.01
Hip dislocation	85.7	1.9121 (1.39 - 2.62)	<0.01

ICDDN = Paediatric Pain Profile, translated in Portuguese language as: Inventário de Comportamentos da Dor na Deficiência Neurológica; GMFCS = Gross Motor Function Classification System; PR = Prevalence ratio; CI = 95% Confidence interval. *Chi-square test

It therefore, justifies the use of anticonvulsants by most of the studied population.

Some clinical conditions observed in patients with CP can cause pain, which justifies the high prevalence of pain found in the patients presenting these manifestations. Pain affects negatively the quality of life by causing discomfort and being an important limiting factor CP patients from developing the skills to carry out daily activities [12,13,21,22]. Spasticity, for example, leads to muscle contractures that can be very painful and to compensatory deformities such as scoliosis, cyphosis and hip dislocation, which result in difficulties in locomotion, positioning and, consequently, the pain [23].

Besides spasticity, gastroesophageal reflux disease, prevalent in more than 59 % of patients, if not treated, can cause pain, in addition to recurrent esophagitis and pneumonia [24,25]. Oral dysfunctions, in turn, can result in poor bone mineralization due a low ingestion of calcium, leading to the development of rickets and osteomalacia in these patients, whose major symptoms are diffuse bone pain and muscle weakness. Moreover, in addition to oral dysfunction, anticonvulsants, often used by these patients, may also favor the decrease in bone mineralization [26]. Patients with CP usually require hospitalization and are submitted to painful diagnostic and therapeutic procedures, such as botulinum toxin injections, gastrointestinal endoscopies, venoclysis, stomatologic treatment and surgical procedures [27,28].

Although individuals classified in level IV and V, according to GMFCS, have their functional impairment classified as severe, level V individuals present a more compromised clinical condition because they are even more likely to be affected by complications related to CP [10,11]. Thus, the association between the higher degree of functional impairment and the presence of behaviors suggestive of pain in these patients, demonstrated for the first time, may be related, at least partially, to the clinical conditions presented by these patients that lead to the development of this symptom.

It is worth noting that even with a high prevalence of pain, this symptom is not treated in these patients. As observed with regard to the use of drugs to relieve this symptom, no evaluated patient used drugs with analgesic properties. According to data in the literature, the use of drugs by CP patients is generally limited to the use of anticonvulsants, which was also verified in our study [4,20,21].

Limitations of the study

The limitations of this study include a low number of subjects and the short duration of the study which was performed only in a determinate region of South Brazil. However, such limitations did not make our work unfeasible, since the data were accurate and consistent. The effectiveness of the research instruments used may encourage its application in future studies, carried out in other regions and with a greater number of subjects.

CONCLUSION

The data show that patients with severe CP who had the highest degree of locomotor impairment, are more likely to present with pain. Thus, these patients should be assisted with the objective of identifying and treating this symptom, and aiming at improved quality of life. For this, GMFCS and ICDDN have proven to be effective tools that can be used to assess locomotor impairment and presence of pain, regardless of age, in patients with CP.

DECLARATIONS

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Conflict of Interest

No conflict of interest associated with this work.

Contribution of Authors

The authors declare that this work was done by the authors named in this article and all liabilities pertaining to claims relating to the content of this article will be borne by them.

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