ORIGINAL ARTICLE

Epidemiological profile of patients with cerebral palsy treated at the AACD-SP

Perfil epidemiológico dos pacientes com paralisia cerebral atendidos na AACD - São Paulo

Anny Michelly Paquier Binha [1], Simone Carazzato Maciel [1], Carla Cristine Andrade Bezerra [2]

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ABSTRACT

Cerebral palsy (CP) is a group of multifactorial clinical syndromes characterized by motor deficit, sometimes with postural dysfunction. **Objective:** To characterize a new series of clinical CP patients in the institution from January-2012 to December-2014. **Method:** Retrospective study of 743 electronic medical records of patients screened at CP outpatient facilities. **Results:** 614 cases were considered eligible. 47.4% female and 52.6% male subjects. 29.5% were under 2 years of age, 34% were from 2 to 4, 15.5% from 4 to 6, 16.3% from 6 to 12, 4.6% from 12 to 18 and 0.2% were above 18 years of age. At birth, 50.7% were preterm and 45% term. Regarding weight, 9.1% were classified as extreme low weight, 16.8% very low weight, 21.8% low weight, 43.6% adequate weight, 2.3% macrosomic. The predominant type of delivery were cesarean section (56.5%). Regarding clinical and topographic classification, 13.4% had spastic hemiplegia, 33.9% spastic diplegia, 12.2% spastic tetraplegia, 0.5% spastic monoplegia, 5.9% Dyskinetic / ataxic, 5.7% mixed CP, and 1% were hypotonic. In 55.5% of the families they did not receive any social benefits. Regarding specialized care, for 97.7% of the patients that was the first appointment with a Physiatrist. **Conclusion:** Most pregnant women undertook at least the minimum number of prenatal visits. Cesarean delivery was predominant. Preterm births were slightly higher. The most prevalent type of CP was the spastic diplegic, with GMFCS ranging from 1 to 5. More than half of families did not manage to have access to social benefits.

Keywords: Cerebral Palsy, Hemiplegia, Quadriplegia

RESUMO

Paralisia cerebral (PC) abrange um grupo de síndromes clínicas de causa multifatorial caracterizadas por déficit motor, algumas vezes com disfunção postural. **Objetivos:** Caracterizar a população dos novos pacientes da clínica de PC na instituição, de Janeiro-2012 a Dezembro-2014. **Métodos:** Estudo retrospectivo. Avaliados 743 prontuários eletrônicos de pacientes atendidos em consultas iniciais de PC, sendo elegíveis 614 casos. **Resultados:** Sexo: feminino = 47,4%, masculino = 52,6%. Idade em anos: 29,5% menores de 2; 34% de 2 a 4; 15,5% de 4 a 6; 16,3% de 6 a 12; 4,6% 12 a 18; 0,2% ≥ 18 anos. Ao nascimento 50,7% eram pré-termo e 45% termo. Peso: 9,1% classificados como extremo baixo peso, 16,8% muito baixo peso, 21,8% baixo peso, 43,6% peso adequado, 2,3% macrossômicos. Tipo de parto predominante: cesáreo (56,5%). Classificação clínica e topográfica dos pacientes: 13,4% Hemiparéticos espásticos, 33,9% Diparéticos espásticos, 12,2% Tetraparéticos espásticos, 0,5% Monoparéticos espásticos, 5,9% Discinéticos/atáxicos, 5,7% PC mista, 1% hipotônicos. Em 55,5% das famílias não recebiam auxílio doença. Sobre atendimentos especializados observou-se que para 97,7% dos pacientes tratava-se da primeira consulta com um médico Fisiatra. **Conclusão:** Maioria das gestantes realizaram pelo menos o número mínimo adequado de consultas de pré-natal. Parto cesariano predominou. Nascimentos pré-termo foi ligeiramente superior comparado com a termo. Tipo de PC predominou o tipo Diparético espástico, com GMFCS nas faixas de 1 a 5 equivalente. Mais da metade das famílias ainda sem acesso a benefício social.

Palavras-chave: Paralisia Cerebral, Hemiplegia, Quadriplegia

[1] Physiatrist, Associação de Assistência à Criança Deficiente – AACD.
[2] Medical Resident, Universidade Federal de São Paulo – UNIFESP.

Correspondência
Anny Michelly Paquier Binha
E-mail: apaquier@aacd.org.br

INTRODUCTION

About 15% of the world population has some neuro-musculoskeletal disability, but part of these disabilities are evident only with the child's growth and development, such as Cerebral Palsy (CP), making both diagnosis and epidemiological record more difficult. CP comprise a heterogeneous group of clinical syndromes, all characterized by motor deficit and / or postural dysfunction. These disorders arise as a consequence of a previously non-progressive brain injury generated by a variety of causes ranging from hereditary factors to different types of damage to the binomial maternal-fetal during pregnancy or the newborn (RN) during perinatal or postnatal period up to one year of life. Although neither central nervous system (CNS) injury nor disease is progressive, the onset of neuropathological lesions and their clinical expression may change over time with the maturing of the developing brain that has been affected.[1-4]

The etiology of CP is multifactorial. Prenatal causes (maternal and gestational) include: genetic issues (e.g., higher prevalence among monozygotic twins), brain malformations (schizoencephaly, ventricular dilation, etc.), congenital infections (toxoplasmosis, cytomegalovirus 'CMV', herpes simplex, syphilis, HIV, Zika-virus), exposure to toxic agents especially in the first trimester of pregnancy (tobacco, alcohol, medication, radiation, illicit drugs), nutritional changes (malnutrition), intrauterine hypoxia and vascular lesions (hypoxic-ischemic encephalopathy). The prenatal causes may include prematurity with central nervous system damage, hypoxia / anoxia. Postnatal etiology includes bilirubin encephalopathy, traumatic brain injury 'TBI', stroke, vascular causes, convulsive syndromes, hypoxia / anoxia, metabolic issues, infectious causes, drowning, tumors, and others.[4,5]

The prevalence of moderate and severe forms of CP in industrialized countries is 1.5-2.5 per 1,000 live births.[1] A study using data from three regions in the United States, with no distinction between children with or without a history of prematurity, estimated a prevalence of 3.6 cases / 1,000 children at the age eight.[6] In Europe, a report that used standardized definitions and included 6,000 children with CP from 13 geographically defined populations, found that the overall rate was 2.08 / 1000 live births.[7] In developing countries the prevalence reaches 7 / 1,000 live births if all grades of CP are considered.[1]

The rate of CP is significantly higher in preterm infants than in term infants.[8] Even among preterm or term infants, small variations in gestational age are associated with risks for CP.[9] In preterm infants (<37 weeks of gestation) and also underweight (<2,500g), the prevalence of CP is 20 to 30 times higher than for full-term infants born with adequate weight.[3] Although preterm birth rates are increasing, and being considered the major contributing factor to perinatal and child mortality in developed countries, paradoxically the survival of extreme preterm infants has also increased. Studies indicate that increased survival is due to technological advances and collaborative efforts of obstetricians, neonatologists, nurses and the multidisciplinary team, nonetheless sequelae may arise, mainly from pulmonary and brain functions, such as CP.[10] Epidemiological studies suggest that alcohol abuse by the mother increases the risk of CP more than three times.[11]

The diagnosis of CP is essentially clinical, based on the patient's health and personal history, pregnancy information and the patient's overall physical and neurological assessment. Ideally, the clinical diagnosis should be complemented by an imaging examination of the brain, preferably magnetic resonance imaging (MRI).

In a study conducted with MRI in Australia with 594 CP patients, brain injury occurred predominantly in white matter (45%), followed by gray matter (14%). Normal imaging was found in 13%, malformations 10%, focal vascular injuries 9%, and miscellaneous 7%.

In Brazil, data related to incidence, prevalence, clinical characterization, age of access to diagnosis and rehabilitation treatment are precarious around the country. Therefore this study is justified given the need for knowledge of information concerning the CP patients assisted at a large rehabilitation center. With the results of this study we will be able to improve health planning, care, promotion and rehabilitation actions for patients with CP.

OBJECTIVE

Characterize the population of patients with Cerebral Palsy at the *Associação de Assistência à Criança Deficiente* - AACD (Ibirapuera-SP, Brazil), between January 2012 to December 2014.

METHODS

This is a retrospective study with data from the electronic medical records of 743 patients at initial consultations in a large AACD unit. The inclusion criteria were the new CP clinic patients attended by the AACD multiprofessional medical team (Ibirapuera / SP) from January 2012 to December 2014. Exclusion criteria were patients without diagnostic confirmation of CP after two years of age, those with a change in diagnosis after investigation of the neuropsychomotor developmental delay (MPSD), those with a significant lack of information about their health history in the medical records. After applying the inclusion and exclusion criteria, data from 614 patients considered eligible for this study were collected and pooled.

This study was submitted to ethical approval of the Institutional Ethics Review Borard, and was approved under registration CAAE 62113616.0.0000.0085.

RESULTS

291 female patients (47.4%) and 323 male patients (52.6%) were found. We distributed patients by age group based on pediatric and physiatry standards, and Gross Motor Function Classification System (GMFCS) concepts with the result of: 29.5% under 2 years of age (A); 34% from 2 to 4 years old (B); 15.5% from 4 to 6 years old (C); 16.3% from 6 to 12 years (D); 4.6% 12 to 18 years (E); 0.2% ≥ 18 years (U).

Other previously diagnosed disabilities and associated with the motor were found, namely: 5.7% visual, 3.9% cognitive, 1.3% auditory, 1.8% two or more motor impairments, whereas 87 (3%) of patients had no disabilities.

Regarding the birthplace of the patients, we found: 55.4% born in the city of São Paulo (SP), 13.8% in the Metropolitan Region (MR) of SP, 14.3% interior of the state of SP, 14.2% in other states, 0.5% from other countries, and 1.8% was not reported. Regarding hometown, we found: 62.7% living in the city of SP - answer 1; 15.8% SP-RM - answer 2; 14.3% of the interior of the state of SP - answer 3; 6.4% from other states - answer 4; and 0.8% had no information - answer 5.

Twinning was reported in 5.9% of cases and trigemellarity in 0.7% of cases. Parental co-bloodiness was identified in 6.0% of the total cases. Regarding maternal data, we found by the total number of prenatal consultations that: 84.2% had 6 or more consultations during pregnancy, 5.4% had less than 6 consultations, 2.9% had consultations without information regarding number of visits, 4.4% did not make any consultation at all. Regarding the gestational age (GA) of the patient at birth we found: 50.7% of preterm patients (<37 weeks

of GA) - answer 1; 45% born at term (37-41 weeks and 6 days) - answer 2; 2.1% post term (≥42 weeks) - answer 3; 2.3% could not inform (e.g., adoption patients) - answer 4.

Among the preterms we found: 15% extreme preterm infants (<28 weeks) - answer 1; 15% severe preterm infants (28-30 weeks) - answer 2; 11.4% moderate preterm infants (31-33 weeks) - answer 3, 9.3% were late preterm infants (34-36 weeks and 6 days of gestational age at birth) - answer 4. The cases considered "non-preterm" corresponded 49.3% of births - answer 5.

Regarding the weight classification of patients at birth, we found: 9.1% with extreme low weight (<1000g), 16.8% very low weight (1000-1499g), 21.8% low weight (1500-2499g), 43,6% with adequate weight (2500-3999g), 2.3% macrosomic (≥4000g) and 6.4% imprecise information. About the place of birth of the child we found: 96.4% born in Hospital / maternity; 1% home birth; 0.7% in other places and 2% could not inform. Regarding the type of delivery we found: 56.5% cesarean section, 37.5% eutocic delivery (normal), 3.1% forceps delivery and 2.9% not reported. Regarding the report of complications during pregnancy / prenatal we had the answers: yes in 71.8%, no in 26.2% of cases and 2% could not report. Regarding neonatal complications we found: 77.9% positive answer; 19.4% did not report any complications and 2.8% could not report complications soon after birth. Regarding the clinical and anatomical classification of patients regarding the types of Cerebral Palsy (CP) we found: 13.4% Spastic Hemiparetics (1), 33.9% Spastic Diparetics (2), 0.5% Spastic Triparetics (3), 12 , 2% spastic tetraparetics (4), 0.5% spastic monoparetics (5), 5.9% dyskinetic / ataxic (6), 5.7% mixed CP (7), 1% hypotonic (8). We classified patients under 2 years as neuropshycho motor development delay (NPMD), which represented 27% of the total of 614 patients surveyed (10).

Regarding the classification by the GMFCS, we found: 13.7% type 1, 9.8% type 2, 12.2% type 3, 9.4% type 4, 12.4% type 5; in 10.1% of the cases it was not possible to conclude and in 32.1% of the cases the classification was not applicable because it is still in diagnostic definition or because of other disabilities associated with the interference with the GMFCS classification.

Regarding the socioeconomic status of these patients we found that in 55.5% of the cases the families did not receive any kind of social benefit, 36.6% received some kind of social benefit and in 7.7% of the cases the information was imprecise.

Considering the questions about access to specialized medical care prior to initial
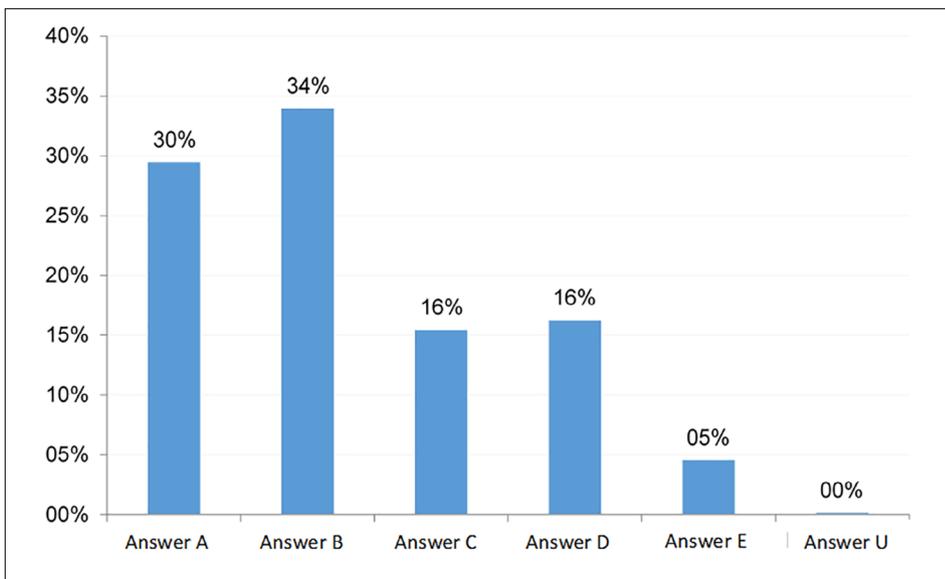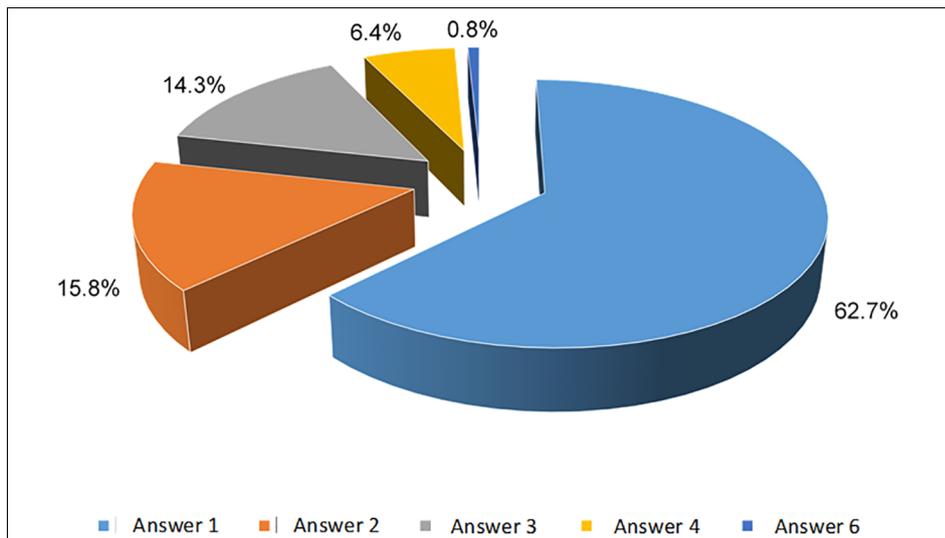

Figure 1. Patients by age group

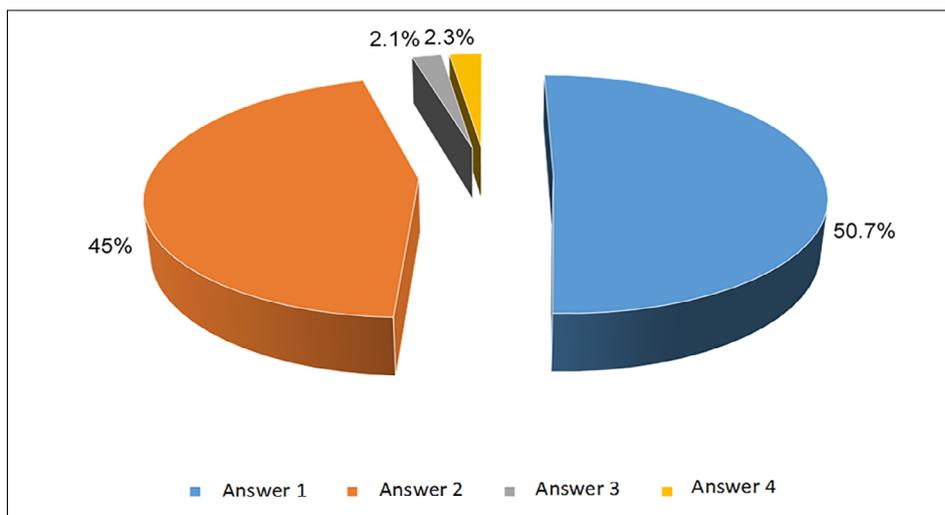**Figure 2.** Distribution per hometown

Figure 3. Distribution by Gestational age at birth

screening / consultation at our institution, we found that 62.4% of the cases had already had access to at least one prior consultation with a Neurologist / Neurosurgeon, 50.3% with a Pediatrician, 15. 1% with Orthopedist and 97.7% never consulted with the doctor Physiatrist / specialist in physical medicine and rehabilitation. Regarding care provided by any of the professionals that make up the multiprofessional team, in 75.4% of the cases, the patients had already received at least one evaluation / care in some type of therapy, as 69.2% in Motor Physical Therapy, 25.7% Speech Therapy, 21.7% Occupational Therapy, 8.1% Aquatic Physiotherapy, 3.9% Psychology, 1.1% Pedagogy, 0.7% Nutrition, and in 24.6% of cases the patient never had previous contact with consultations / therapists of a multidisciplinary team.

DISCUSSION

Some epidemiological publications regarding CP published in Brazil and worldwide report the tendency of a prevalence of male patients when compared to female patients,[13] however other studies in other countries could not conclude the same.[14] Our study revealed that there was higher prevalence of CP patients among the male subjects.

We divided patients' ages into age groups according to the GMFCS revised expanded classification system before 2 years, 2-4 years, 4-6 years, 6-12, and 12-18 years,[15] and we added the range 18 years of age and above. We found the highest number of visits to patients aged 2 to 4 years incomplete (34%), followed by the age group to 2 years incomplete (29.5%), which may show some difficulty in diagnosis or even refer new suspected / confirmed PC cases to a referral institution. Regarding the findings about other disabilities besides the postural-motor disability, due to the young age, many cases were still under investigation on other possible associated disabilities, such as visual impairment, and despite the suspicion, many cases at the time of the consultation, did not have diagnostic confirmation yet, making this data inaccurate. It is essential to seek to establish the etiological diagnosis, determine the specific type of CP and also to investigate the existence of other deficiencies other than postural-motor, so that one can know about the prognosis and the best planning of a rehabilitation treatment program that always needs to be multidisciplinary.

In the territory analysis of the territoriality as to the place of birth and hometown of the patients, we considered the IBGE geographical divisions standard and divided the patients into 5 groups according to the regions: SP-
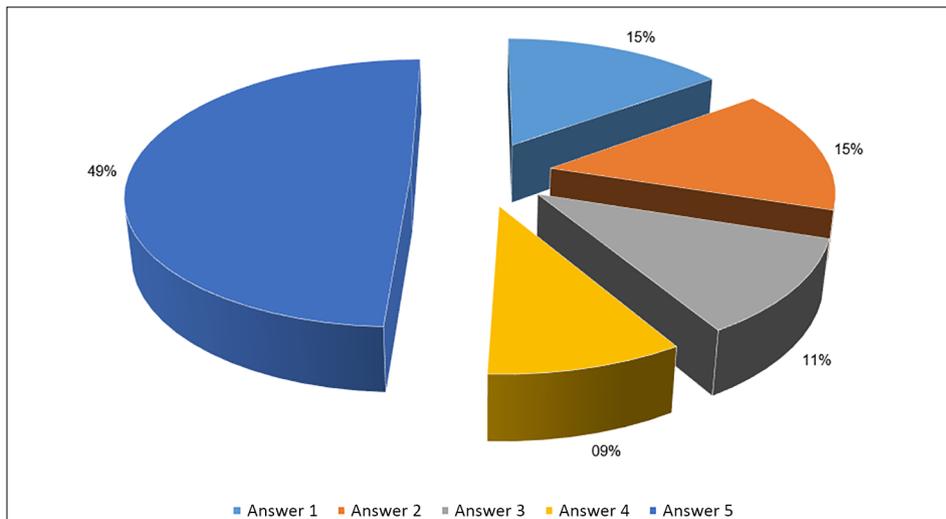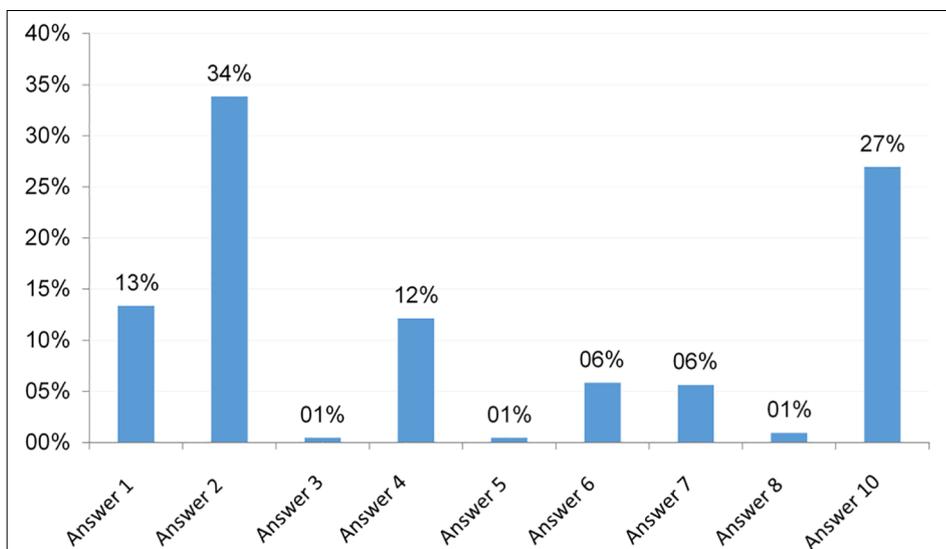

Figure 4. Distribution by pre-term classification

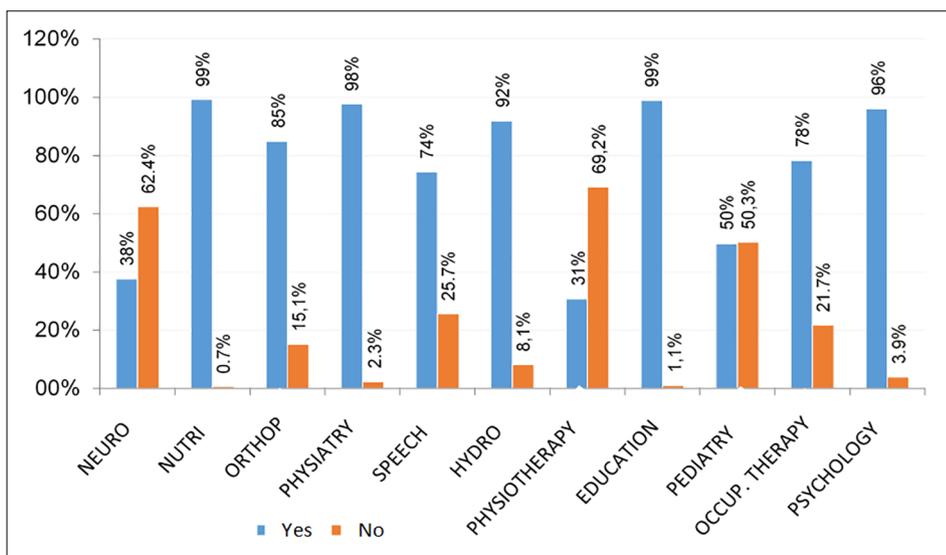**Figure 5.** Distribution per gestational age at birth

Figure 6. Distribution by previous consultations – Yes / No

SP, Metropolitan Region of SP, interior of the state of SP, other states of the state. Brazil and other countries. We observed that the number of patients from the city of SP was the majority in both cases. Regarding the place of birth compared to the hometown we also observed a predominance of the state of SP, which corroborates with reports found by some families who claimed to have moved to the state of SP in search of some or better access to specialized and multiprofessional medical care.

More than 90% of the pregnant women had access to prenatal care, and more than 80% had the appropriate number of consultations (according to the Ministry of Health parameters) and most complications were reported during this period.

With the advances made in fetal medicine and its methods, the frequency of multiple pregnancies (twins, trigemellar, etc.) has been increasing. In Brazil, according to DATASUS / 2012, twins represented 19.7% and triplets / other multiples represented 0.05% of all live births.[16] In this study there was an incidence of twin pregnancy of 5.9%. of CP cases and 0.7% born of trigemellar pregnancy. Recent publications concludes that the gestation of two or more fetuses would not necessarily result in an increased risk for CP-leading disorders, but there is still a consensus in the medical community that the risks of adverse birth-related events increase the incidence of CP in twins.[5,17]

Regarding the classification of types of Cerebral Palsy (CP) we found a predominance of the Spastic Diparetics type (33.9%), followed by Hemiparetic (13.4%) and Tetraparetic (12.2%). This result agrees with the same findings as Cassefo et al. in a previous study with 236 patients in SP- Brazil.[14] However, it differs from a study conducted in 2012 by the Teleton Institute of Santiago, Chile, in which Hemiparesis is the most frequent presentation (29.9%), followed immediately by Diparesia (29.3%) and Tetraparesis (21.3%).[2]

According to a study in Finland,[8] the incidence of CP is higher in late and moderate preterm infants compared to full-term infants, and this is in line with the findings of this study: 50.7% of preterm infants (<37 weeks); 45% born at term (37-41 weeks and 6 days); 2.1% post-term (≥42 weeks), with the lowest incidence being 11.4% moderate (31-33 weeks) and 9.3% being late premature (34-36 weeks and 6 days birth).

Regarding the classification of the patient's weight at birth, we found a predominance of low weight, which totaled 47.7% of cases, almost 40% above the percentage found in a previous study[4] where 35.6% of patients born with low weight were identified.

The prevention of new cases of CP involves issues related to family planning, prenatal care, childbirth, postnatal care, careful observance of risk factors and overall attention to child health in the first year of life. Notably, the historical difficulties in the Brazilian Health System (*Sistema Único de Saúde* - SUS) are evidenced by the inability to receive patients, the lack of a unified system of information on health data, the inadequacy of family planning actions, the quality of prenatal, postnatal and postnatal care, up to the challenge of an adequate longitudinal attention to children's health. This items are also present in our study, given the apparent need for greater and faster changes, especially in the public health system, at all levels of health and attention, because we found equivalent numbers of pre, peri and post natal causes for CP. Another challenging difficulty is underdiagnosis in mild cases of CP.

From our data, we found that the construction of a municipal and state database for the diagnosed cases of CP could generate significant advantage in helping prevention plan, care and rehabilitation.

It is also essential to expand access to multiprofessional rehabilitation at an early age. Only quality comprehensive rehabilitation can prevent further clinical complications and the worsening of motor ability in these patients, from turning children with CP into adults well-integrated into the family and society, with the potential to be economically active, and most importantly, the possibility of being happier people.

CONCLUSION

In this study, we observed that in the population of new patients treated at the institution's Cerebral Palsy clinic, the predominant were: the 2-4 year age group, the spastic Diparetic CP subtype, with no marked differences in GMFCS distribution. Cesarean section was the most frequent type of delivery, and preterm infants had a slightly higher percentage than those born at term.

We also observed that more than half of the patients / families did not receive any kind of governmental social assistance / benefit previously. Almost all the population has never had prior access to care with a physiatrist.

The most accessed therapy previously was Physical Therapy.

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