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Becoming a young adult with cerebral palsy

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ABSTRACT

Aims: This paper aims to describe the functioning profile of a clinical sample of patients with Cerebral Palsy at the time of transition.
Methods and procedure: For this retrospective observational study, we considered data concerning 389 patients born from 1967 to 1997 with a diagnosis of CP and discharged at the age of 18 ± 3 from "La Nostra Famiglia" Children Care Centres. We reported data concerning: identifiable risk factors, the type of CP, the level of motor, manual and communication abilities, the occurrence of associated impairments and environmental factors, and examine the trends over the decades.
Outcome and results: The disorder was mainly bilateral (86%): 57% of patients had quadriplegia and 43% had diplegia. Most of patients had a spastic disorder (86%). Comorbidities were frequent, mainly intellectual developmental disorder (63.5%). One fifth of patients showed a severe impairment of motor, manual and communicative skills in addition to a severe intellectual development disorder. Over decades, the outcome as expressed by the motor functioning profile improved but the outcome as expressed by the global functioning profile worsened because of the growing number of severe multi-disabilities.
Conclusion and implication: Knowledge about the functioning profile of young adults with cerebral palsy can support health services in the provision of developmentally appropriate care.

What this paper adds

This paper describes the functioning profile (a global description including environmental factors) of a clinical sample of patients with cerebral palsy transitioning to adult age. Available data concern the transition of patients with normal or near to normal intellectual abilities, whereas this study includes participants with severe intellectual developmental disorders or severe motor impairment.

Trends in functional profile, identifiable risk factors and interventions are examined.

1. Introduction

Due to significant improvement in neonatal care, infant mortality has decreased from an estimated rate of 64.8 deaths per 1000 live births in 1990 to 30.5 deaths per 1000 live births in 2016, as reported by the Global Health Observatory (World Health

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Organization (WHO) and Global Health Observatory (2018)). Medical science has enabled an increase of the survival of premature babies over time (Smith, Draper, & Field, 2013). This means that, as reported by While et al. (2004) a growing number of children that previously were not expected to survive, is now able to reach adulthood while dealing with long term disabling conditions.

Among chronic conditions, Cerebral Palsy is one of the most common childhood disabilities and has an incidence of 1 per 500 live births (Oskoui, Coutinho, Dykeman, Jetté, & Pringsheim, 2013). It "describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to a 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" (Rosenbaum, 2017). The clinical phenotype of CP varies from patient to patient and includes children with minimum or mild motor impairment and those with severe quadriplegia. They may suffer from further clinical conditions such as intellectual developmental disorder (IDD), visual and hearing impairments, and seizures…

CP clinical status varies over time, its manifestations may differ from person to person, in different life periods and may be influenced by several aspects such development, therapies, education, familiar and social context (Rosenbaum et al., 2007).

Recently, over 90% of children with CP reaches adulthood (Strauss, Brooks, Rosenbloom, & Shavelle, 2008). Some recent researchers have investigated the long term evolution of CP, and documented an increased deterioration in walking function, greater fatigue (Opheim, Jahnsen, Olsson, & Stanghelle, 2009) and higher rates of complications. As a consequence, a reduced participation in daily activities, social and intimate relationships were also observed (Blackman & Conaway, 2014; Donkervoort, Wiegerink, van Meeteren, Stam, & Roebroeck, 2009). Transition profile has been shown to be related to motor and manual functioning and to the level of education and functioning in activities and participation (Donkervoort et al., 2009).

Patients with CP reported several barriers to transition (Carroll, 2015) and did not often feel ready for that, thus suggesting that transition to adult health services should be flexible with respect to biological age (Björquist, Nordmark, & Hallström, 2015).

All reported papers focused mainly on patients with normal or close to normal intellectual abilities; as per our knowledge, no study includes patients with intellectual and developmental disabilities (IDD), even if its prevalence in CP population is approximately 45% (Yin Foo, Guppy, & Johnston, 2013).

This paper describes patients with CP transitioning to adult age including those with IDD or severe motor impairment, thus avoiding the pre-selection based on motor or cognitive abilities. The profile was described on the basis of the type of CP, the level of functional abilities, the occurrence of comorbidities and identifiable risk factors (Novak et al., 2017). Environmental factors were also considered. In order to examine trends we compared data over three birth decades: from 1967 to 1976, from 1977 to 1986 and from 1987 to 1997.

We believe that understanding the functioning profile of patients transitioning to adult age could support health services in the provision of developmentally appropriate care.

2. Material and method

2.1. Participants

We considered all patients with a diagnosis of cerebral palsy born from 1967 to 1997 and discharged, after a minimum of 3 years of rehabilitation, from a network of seven Child Care Centers in Italy from 1985 to 2015 ($n = 565$). Only patients discharged at age of 18 ± 3 were selected, the final sample consisted of 389 patients (216 male and 173 female). 81 patients born from 1967 to 1976, 145 born from 1977 to 1986 and 163 from 1987 to 1997.

The mean age at the time of admission was 2 years 5 months (SD = 3 years 3 months) and the mean age of discharge was 15 years 6 months (SD = 3 years 4 months).

2.2. Classification of CP

Data were collected from clinical reports and concerned: risk factors, characteristics of motor abnormalities, the level of motor, manual and communication abilities and the occurrence of comorbidities such as IDD, epilepsy, visual and hearing impairments. The movement disorder was classified as unilateral or bilateral. This latter was classified as diplegia or quadriplegia. CP was classified as either spastic, ataxic or dyskinetic (including dystonia and athetosis). Motor, manual and communication functioning was characterized using the following scales: the Gross Motor Function Classification System - Expanded & Revised (GMFCS - E&R) (Palisano et al., 1997); the Manual Ability Classification System (MACS) (Elliasson et al., 2006) and the Communication Function Classification System (CFCS) (Hidecker et al., 2011). The GMFCS is a 5-level classification system that describes the gross motor function. Level I indicates that the patient performs gross motor skills with minimum limitations and it describes the highest level of functioning. Level V is used when patients are transported in a manual wheelchair in all settings.

The MACS is a 5-level classification system that describes the ability to use the hands to handle objects in everyday activities. Level I indicates that the patient is able to handle objects easily and successfully. Level V is used when the patient cannot handle objects and needs total assistance.

The CFCS is a 5-level classification system that describes everyday communication performance. Level I indicates that the patient is able to communicate easily both as sender and receiver with most people in most environments. Level V indicates that the patient is seldom effective both as sender and as receiver even with familiar conversational partners.

If present, IDD was classified as: mild (approximate IQ range 50–69), moderate (IQ range 36–49), severe or profound ($IQ \leq 35$).

Table 1
Distribution of GMFCS levels by subtypes of CP.

		GMFCS I	GMFCS II	GMFCS IIII	GMFCS IV	GMFCS V	
Anatomic distribution	Hemiplegia	1 (0,25%)	51 (13,11%)	1 (0,25%)	0	1 (0,25%)	**54**
	Diplegia	1 (0,25%)	74 (19%)	43 (11,05%)	16 (4,11%)	10 (2,57%)	**144**
	Quadriplegia	0	24 (6,17%)	15 (3,85%)	30 (7,71%)	122 (31,36%)	**191**
		2 (0,51%)	**149** (38,3%)	**59** (15,17%)	**46** (11,82%)	**133** (34,19%)	**389**

Number of cases (percentage in brackets) for each level of GMFCS for each type of CP (spastic, dyskinetic and ataxic). Levels indicates the patient: *Walks without Limitations* (I); *Walks with Limitations* (II); *Walks Using a Hand-Held Mobility Device* (III); *Self-Mobility with Limitations* and *may Use Powered Mobility* (IV); and *Transported in a Manual Wheelchair* (V).

The occurrence of visual impairments (visual acuity loss, saccadic impairments, strabismus, oculomotor problems), hearing impairments (hearing acuity loss, auditory attention and orientation to stimuli) and epilepsy was reported.

Although there is often not just one specific cause (Novak et al., 2017; Rosenbaum et al., 2007), there are some risk factors that may interfere with the normal development of brain structures and functions. Risk factors were classified as: hypoxia, preterm birth (infants < 36 weeks of gestation), hemorrhage, perinatal infection, congenital anomalies, exposure to toxic substances, epilepsy, other and unknown.

With reference to environmental factors of the ICF model (World Health Organization (WHO) (2001)), we reported information concerning rehabilitative and surgical interventions delivered during the incharge period.

The study has been reviewed and approved by the Ethic Committee (Prot. N. 61/17 - CE), the article is adherent to the committee's recommendations and all participants gave their written informed consent.

3. Results

The movement disorder was bilateral in 86% of participants (n = 335). Out of this, 57% (n = 191) had quadriplegia and 43% (n = 144) had diplegia. The rate of bilateral motor disorder gradually decreased from 94% in the '67-'76, to 86% in the' 77-'86 and further reduced to 82% in the' 87-'97. The decrease was due to the reduction of the rate of quadriplegia (from an initial 63% to 43%); rates of diplegia increased (from 31% to 39%).

The 85.6% of participants (n = 333) had spastic disorders, the remaining had dyskinesia (8.7%, n = 34) or ataxia (5.7%, n = 22). These rates did not substantially change over the three decades: 84%, 87% and 85% for spastic disorders; 12%, 7% and 9% for dyskinetic disorders and 4%, 6% and 6% for ataxic disorders.

Results concerning the GMFCS are reported in Table 1; about half of participants (mainly affected by quadriplegia) used a wheelchair and/or needed complete assistance to be transported (Level IV and V). Almost all patients with hemiplegia (98.15%) or diplegia (82%) were able to walk independently or with an aid (from Level I to III).

Concerning the MACS, about forty percent of all participants presented high levels of impairment in the bimanual upper limb functioning and needed assistance for the achievement of the activity (Level IV-V). Communication performance was less impaired and most patients (68.9%) were effective both in the transmission and in the understanding of messages at least with familiar partners (CFCS level I-III). On the whole, the 63.5% (n=247) presented with IDD, mainly severe or profound (27.65%, n=107). Visual impairments were diagnosed in 59.5% of cases (n=230), epilepsy in 37.5% (n=145). Hearing was impaired in less than 5% of participants (n=17). Results concerning the MACS, the CFCS and rates of IDD in the overall sample are represented in Fig. 1.

Over the decades, the rate of patients scoring from Level I to III in the GMFCS gradually increased from 47%, to 55.17% and finally to 56.44%. In contrast with this trend, both the manual and communication performance, as well as the intellectual ability seemed to impair over the decades. The rate of patients scoring Level IV and V in the MACS gradually increased from 37.04%, to 37.93% and finally to 41.10% in the last decade. Considering the CFCS, this rate increased from 24.69%, to 30.34% and finally to 34.97%. The rate of patients affected by IDD increased from 56.79% to 61.38% and finally reached the 68.71%.

Forty three percent of patients had preterm birth; about one third (30.1%) suffered from hypoxia, whereas less than 15% presented other risk factors. No risk factor was identifiable for 39 patients (10%).

Preterm birth and hypoxia were the more frequent identifiable risk factors in all decades, but from 1987 to 1997 preterm birth rates decreased (from a mean of 48% to a previously observed 36.8%) and rates of unidentified factors increased (from 0 to 15.9%), see Fig. 2.

Treatment lasted a mean of 15 years 5 months (SD = 3 years 3 months) and for 98% of patients it consisted of physical interventions. 70% of these patients received also occupational therapy and about one third (31.8%) speech therapy. Other therapies (as neuro-visual rehabilitation, psychological and psychomotor rehabilitation) were delivered to less than 2% of patients. This trend did not substantially change over decades. During the rehabilitation, an orthopedic surgery was used for 242 patients (62.2%) to reduce deformation and to correct or improve movement and alignment in legs, ankles, feet, hips, wrists and arms. These interventions were mainly: Baker's procedure and/or muscle lengthening (48%), adductor tenotomy (27%) and, to a smaller extent, osteotomy and arthrodesis. Botulinum toxin treatment was used for 8.22% of patients. Over the decades a decrease in the rate of surgeries and an increase in non-surgical approaches were observed (see Fig. 3).

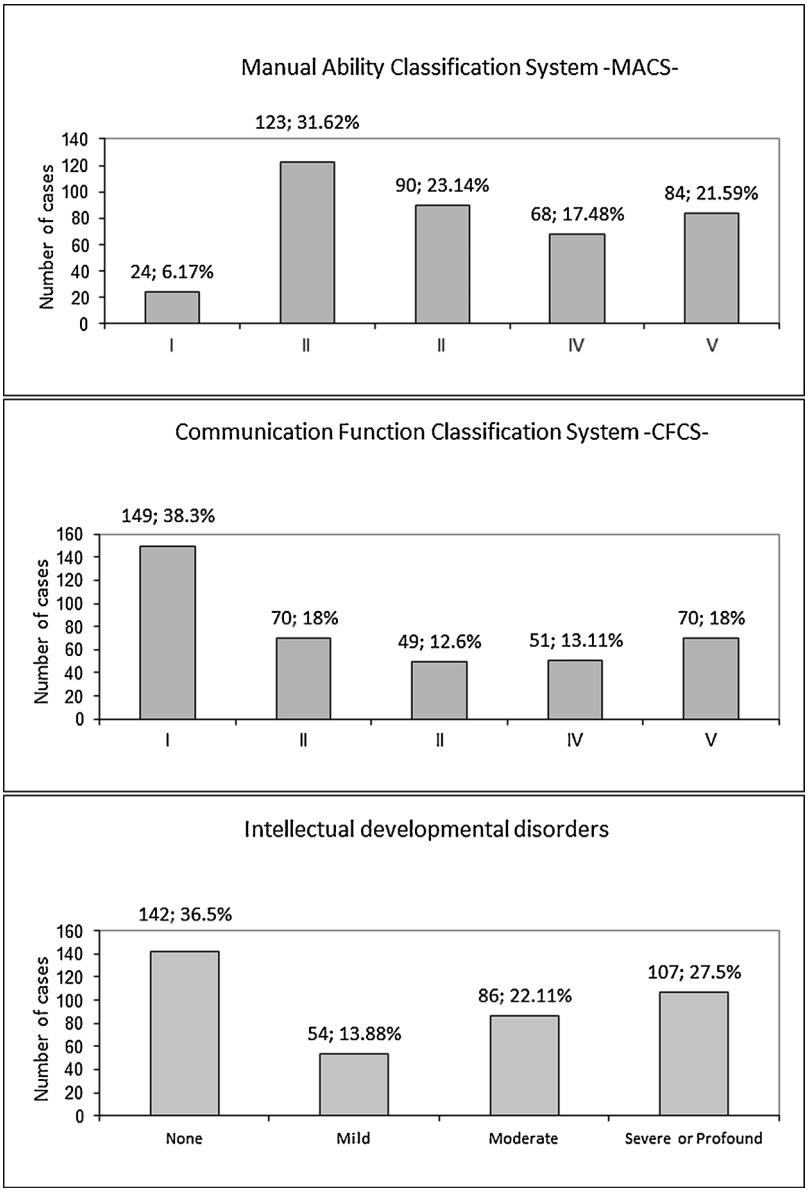

Fig. 1. Number and percentage of cases with different level of MACS, CFCS and intellectual developmental disorders (IDD).
In the first panel MACS Levels indicates the patient: *Handles objects easily and successfully* (I); *Handles most objects but with somewhat reduced quality and/or speed of achievement* (II); *Handles objects with difficulty and needs help to prepare and/or modify activities* (III); *Handles a limited selection of easily managed objects in adapted situations* (IV); *Does not handle objects and has severely limited ability to perform even simple actions* (V).
In the second panel CFCS Levels indicates the patient is: *Effective Sender and Receiver with unfamiliar and familiar partners* (I); *Effective but slower paced Sender and/or Receiver with unfamiliar and/or familiar partners* (II); *Effective Sender and Receiver with familiar partners* (III); *Inconsistent Sender and/or Receiver with familiar partners* (IV); *Seldom Effective Sender and Receiver even with familiar partner* (V).
The third panel displays cases with no intellectual developmental disorders and with different level of IDD: mild (approximate IQ range 50–69), moderate (IQ range 36–49), severe or profound (IQ ≤ 35).

4. Discussion

As reported by Rosenbaum, "the role of aging in changing the clinical phenomenology of CP has been little studied" (Rosenbaum, 2017); in this paper, we presented the functioning profile of a clinical sample of CP patients transitioning to adult age in order to support health services in the provision of developmentally appropriate care.

The sample consisted mainly of patients with a bilateral motor impairment; spasticity was the predominant motor disorder (85.6%), followed by the dyskinesia (8.7%) and ataxia (5.7%). Comparing these results with the Australian Cerebral Palsy Register Report (Australian Cerebral Palsy Register Group, 2016), based on children aged from 0 months to 5 years or later, rates are similar

Fig. 2. The trend of risk factors on the basis of the patient birth decades.

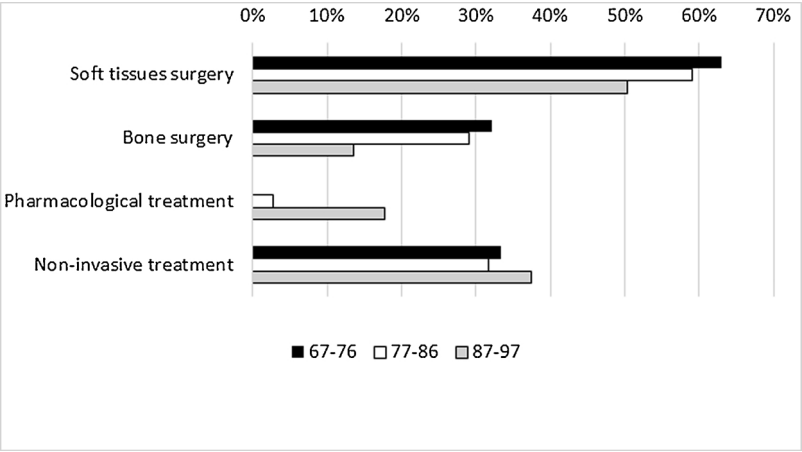

Fig. 3. The trend of orthopedical surgeries delivered by the patient birth decades.

(85.8%, 6.4% and 4.8% for spasticity, dyskinesia and ataxia, respectively). The frequency of quadriplegia is more than twice the Australian one (49% *versus* 22%) and the rate of hemiplegia is one third (13.9% *versus* 39%). This register however is based on children born from 1993 to 2009 and it is therefore more recent. In accordance to this data, also in our sample, the rate of quadriplegia gradually reduced over the decades whereas rates of diplegia and hemiplegia increased. This suggested an improving trend in the movement disorder. An opposite trend was reported by Stanley and Watson (1992) who collected data from 1967 to 1985 and found a 3% fall per period in total hemiplegia rates; a 17% fall in diplegia and a 22% increase in quadriplegia. Both results are in contrast with Pharoah (Pharoah, Cooke, Cooke, & Rosenbloom, 1990) who found rates for quadriplegia and hemiplegia similar throughout the period and a consistently lower prevalence of diplegia with respect to the others. These contradictory results may also reflect changes in definition and classification of CP that continue from 150 years ago to the present time (Morris, 2007).

Premature birth is still the leading risk factor (Stavsky et al., 2017) but there is not just one specific cause for the development of CP (MacLennan, Thompson, & Gecz, 2015). More than three-quarters of patients with quadriplegia used wheel mobility or required complete assistance to be transported, whereas most of patients with diplegia or hemiplegia were able to walk without physical assistance or using a hand-held mobility device. Even if the rate of patients needing complete assistance to be transported decreased over the years, our results show a higher rate of these patients with respect to the Australian Report. This may be explained by the evidence of a severe IDD in about one third of our sample, who were unable to use a wheelchair or other aids independently. About forty percent of patients presented a severe impairment (Level IV and V) in the manual ability and about thirty percent in the communication performance. Over the decades, rates indicating a severe impairment increased both in the manual ability and communication performance.

Consistent with the Australian Report, the most frequently observed comorbidity was the IDD, followed by visual impairments,

epilepsy and hearing impairments. Before discharge, most of patients were treated with an orthopedic surgery but over the decades the rate of surgeries decreased and an increase in non-surgical approaches, such as botulinum injection, was registered.

Transition is more difficult in presence of a severe cognitive disability (Reiss, Gibson, & Walker, 2005). A severe or profound IDD in patients with a complete impairment of walking function (level IV and V of GMFCS) was observed in 50.6% of cases, this rate increased to 65% when considering manual ability (IV and V of MACS) and to 78.5% for communication (IV and V of CFCS). On the whole, about 20% of patients showed a severe impairment of motor, manual and communicative skills in addition to a severe IDD; this means that, even if adults, about one fifth of the sample was totally dependent on others for daily activities. Over the years, this rate slightly increased from 17.2% (from 1967 to 1976) to about 22% (for the following two decades). This suggests that, despite the outcome as expressed by the motor functioning profile by itself improved over years, the outcome as expressed by the global functioning profile appeared more impaired due to the co-occurrence of severe manual, communication and intellectual impairments. Across the decades an increased number of severe multi-disabilities was observed. This could be related to the increased survival rates of severely impaired infants, as demonstrated by Stanley and Watson (1992). On the other hand, the survival into adulthood of CP patients is still negatively affected by the co-occurrence of accompanying impairments (as epilepsy and cognitive impairment) and no trend over time was observed (Himmelmann & Sundh, 2015)

4.1. Limitation

This study focused on a cohort of patients discharged from our network of rehabilitation centres, thus it lacks of epidemiological strength. This study did not include patients discharged before the cut off time (3 years of rehabilitation), thus the rate of patients with mild motor impairment (such as hemiplegia) are likely to be underrepresented. Data were obtained from printed medical records and for this reason it was not possible to count the number of treatment hours each patient received. Measures of perceived quality of life were not available.

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