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Problematyka niepełnosprawności dorosłych z mózgowym porażeniem dziecięcym

Issues in disability among adults with cerebral palsy

Streszczenie

Wstęp. Osoby dotknięte mózgowym porażeniem dziecięcym (MPD) stanowią odrębną grupę rezydentów zakładów opieki długoterminowej. Charakterystyczne dla tych pensjonariuszy są zaburzenia motoryki, którym towarzyszą zwykle inne, różnorodne problemy zdrowotne, stwarzające specyficzne potrzeby opieki.

Cel. Określenie częstości występowania mózgowego porażenia dziecięcego (MPD) wśród pensjonariuszy zakładów opieki długoterminowej oraz problemy dotyczące tych osób.

Materiał i metody. W oparciu o analizę dokumentacji i przesiewowe badanie kliniczne spośród 435 pensjonariuszy zakładów opieki długoterminowej powiatu przemyskiego wyodrębniono grupę osób z objawami MPD i oceniono u nich wybrane problemy zdrowotne w aspekcie ilościowym.

Wyniki. Ujawniono 35 osób spełniających przesłanki do rozpoznania mózgowego porażenia dziecięcego w dwóch kategoriach opieki: „dorośli niepełnosprawni intelektualnie" (29 osób) i „osoby przewlekle psychicznie chore" (6 osób). Chorzy ci stanowili odpowiednio 27% i 4% wyżej wymienionych kategorii. Wzmianka o MPD pojawiła się w dokumentacji 10 osób, w tym u 9-ciu rozpoznanie postawiono w okresie dzieciństwa. U 9 osób opisywano inne niż MPD zaburzenia motoryki. Deficyt intelektualny opisano w dokumentacji 29 osób (83%) co stanowi odsetek istotnie wyższy niż oczekiwany na podstawie polskich badan epidemiologicznych.

Wnioski. Deficyt intelektualny będący główną przyczyną kwalifikacji do objęcia opieką długoterminową powoduje pomniejszanie znaczenia lub pomijanie innych dysfunkcji (w tym ruchowej) w ocenie stanu zdrowia chorych z MPD. Uzasadnione jest twierdzenie, ze osoby z MPD stanowią i winny stanowić osobną kategorię podopiecznych w opiece długoterminowej.

Słowa kluczowe: mózgowe porażenie dziecięce (MPD), chorobowość, niepełnosprawność, orzeczenie o niepełnosprawności, deficyt intelektualny.

Abstract

Introduction. Individuals affected by Cerebral Palsy (CP) may be recognized as distinctive group of attendees of long-term care facilities. Motor disturbances, typical in the group are usually accompanied by various disabilities, requiring specific medical and social support.

Aim. The aim was to determine the incidence of cerebral palsy (CP) among the attendees of long-term care facilities,and to characterise the patients with cerebral palsy (CP).

Material and methods. On the basis of medical documentations analysis and screening clinical examination of 435 attendees of Przemyśl district long-term care institutions a group of patients suffering from cerebral palsy was determined and evaluated in regard to quantitative aspects of selected health problems.

Results. Thirty five patients meeting the conditions for diagnosing cerebral palsy were identified in two categories of care: "adults with intellectual impairment" (29 people) and "patients with chronic mental disorders" (6 people). The abovementioned patients constituted 27% and 4% of the related groups respectively. Cerebral palsy was mentioned in medical documentation of 10 patients, in 9 cases it had been diagnosed in childhood. Nine of the patients were diagnosed with other locomotion disabilities. Intellectual impairment was identified in the documentation of 29 people (83%), which gives a higher ratio than expected, taking into consideration Polish epidemiologic research.

Conclusions. Intellectual impairment, recognized as the main reason for provision of a long-term care,results in placing minor importance or neglecting completely other dysfunctions (including locomotive disabilities) in cerebral palsy patients' health examinations. The statement that patients with cerebral palsy (CP) constitute and should be recognized as a separate category of attendees of long-term institutions is justified.

Key words: cerebral palsy (CP), morbidity, disability, medical disability certificate, intellectual impairment.

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INTRODUCTION

Cerebral palsy (CP) is a chronic complex disease lasting throughout the lifespan of an impaired person. The presence and development of CP among adults is the topic of just a handful of medical research and is very seldom a subject matter of clinicists' research, on account of associating it with the developmental medicine. The improving standards of medical care during pregnancy as well as care provided in labour wards had no impact on the CP incidence [1]. A thesis that a frequency of cerebral palsy prevalence increases with the civilisation development of a society is more frequently met. Predicted life expectancy of a patient with CP who reached puberty is close to the average [2]. Therefore, a rise in percentage of CP impaired adults in population is probable, due to the increase in the average life expectancy and demographic changes, and thus it will be an increasingly burning socio-medical problem.

People with CP suffer from locomotion and posture disturbances, most often these are coupled with other medical conditions such as intellectual impairment, epilepsy, and perception stimuli disorders (sight, auditory, equilibrium and proprioception), speech and deglutition disorders. Psychological disturbances and conduct disorders, caused by primary central nervous system impairment, anomalous psycho-intellectual development and insufficient environment adaptation, are often present in these pathological complexes. During adulthood and in elderly age further constitutional disorders develop, such as: osteoporosis, locomotor system functional pain impotence, predominantly caused by degenerative joints disease and the clinical picture of the disorder evolve together with the development of an individual [3].

Currently there are no guidelines regarding the treatment of an adult suffering from CP. This situation results partly from the concentrated attention of medical personnel towards this disease during the period of growth (childhood and middle childhood), and partly due to the insufficient clinical characteristics of adult patients suffering from CP.

AIM

The aim of the research conducted was to determine the incidence of occurrence of CP among the adult attendees of long-term care facilities from a specific geographical area (quantitative assessment of the scale of the problem discussed), as well as to identify their health problems (qualitative assessment). Another aim was to prepare a cohort for a long term observation that aims at better understanding of the course of CP in adults.

MATERIAL AND METHODS

The research was conducted in Przemyśl district among the residents of social care facilities (SCF) and health care and curative institutions (SCCI). There are 5 SCFs in the district and in 2009 they were inhabited by 435 people (according to the data collected by the departments responsible for medical statistics in these facilities). In SCCIs that are authorised to care for elderly people and people with dysfunctions and long-term somatic diseases, there were no residents with neurological disorders of congenital nature or that developed in childhood.

After the analysis of medical documentation, neurological examination and community interview, a group of patients who suffered from paresis and locomotion impairment from early childhood was identified from among the SCFs residents. Patients with poliomyelitis were excluded from the group, as well as the people with diagnosed progressive causes of neurological disorders and uncertain cases (for example when it was impossible to determine the time of paresis development). Thus a group of 35 people was distinguished; each person suffered from locomotion/posture impairment that developed or occurred within the first years of life and was accompanied by other neurological deficits – according to the definition of CP [4].

Those residents were subject to a detailed clinical examination – the qualification of the type of the injury according to Ingram was performed, as well as the qualification of locomotion abilities according to Gross Motor Function Classification System (GMFCS). The analysis of their medical documentation was conducted in order to get to know whether CP had been identified, and to determine the type of care provided, the type of medical disability certificates, as well as other medical problems – mental disorders, speech and visual disorder, hearing impairment, the presence of epilepsy and operations the patient had undergone.

RESULTS

Medical disability certificate is the most important document for an attendee of SCFs qualifying them for admission. Since 1999 this certificate has been issued by disability evaluation boards that may assign maximum of 3 out of 11 "symbolic impairment codes" to the assessed person. Six persons from the researched group possessed such certificates – data available in Table 1, symbolic codes were standardized to comply with the version bounding since 2003 [6]. Code 05R (musculoskeletal/movement) was assigned to two people with unidentified "mental impairment" stated in their medical documentation. None of the patients with profound or severe mental impairment had 05R code assigned, though locomotion disabilities were described in their documentation and were visible during clinical diagnosis (Table 1).

After issuing of acceptance to SCF, a medical record of an attendee is created. Data from this record are the basis for determining the type of care according to article 56 of the "Social Welfare Act" [10] and for preparing listings/reports for fiscal settlements with payers (local authorities, National Health Fund). Diagnosis from these records were standardised to fulfil the ICD-9 norms (International Statistical Classification of Diseases and Related Health Problems) and were analysed – the obtained data is presented in Table 2. In the case of 6 out of 35 patients psychological impairment and behaviour were described – they were assigned to a group "attendees with chronic psychological disorder" (Article 56 of the "Social Welfare Act", item 3). An intellectual deficit of various levels of severity was recorded in 29 cases, these people were categorised as "adults with intel-

TABLE 1. List of patients with "medical disability certificate".

No	Age	GMF CS	Diagnosis	Neurol. impairment tab 3	Type of CP acc the trial	Impairment Code* (max. 3)
17	27	1	Severe mental impairment F72	no	diplegic	01U
21	36	1	Severe mental impairment F72, spastic tetraparesis G82.4	spastic tetraparesis G82	hemiplegic	01U
36	70	3	Mental impairment F79, epilepsy G40	tetraperesis G82	diplegic	02P 05R 10N
43	45	1	Mental impairment F79	no	hemiplegic	01U 05R
66	21	2	Severe mental impairment F72, cerebral palsy G80	Cerebral palsy G80.	diplegic	01U
82	31	4	Profound mental impairment F73, epilepsy G40	spastic tetraparesis G82	bilateral hemiplegic	01U

* 01U intellectual impairment. 02P psychiatric, 05R musculoskeletal/movement, 10N neurologic disorders

TABLE 2. Main diagnosis based on medical records, standardised to ICD-9 (n=35).

Diagnosis acc ICD-9	Count	Percent
F71	4	11.4
F72	4	11.4
F73	11	31.4
F79	10	28.6
F7*	29	82.9
F69	4	11.4
F0*	2	5.7
Mental disturbances (F0* + F6*)	6	17.1
G40	8	22.9
G80	6	17.1
G8*	10	28.6

One attendee could be diagnosed with more than one disease

TABLE 3. List of patients with neurological deficits noted in medical record.

No	Neurol.impairment (as reported in files)	Diagnosed in	Definite diagnosis of CP
11	Spastic quadriplegia, right eye strabismus, upper limb athetosis.	neurol. ward.	no
12	Cerebral palsy (of spastic type); post-cerebral palsy state.	ped. long term care unit	yes
18	Cerebral palsy.	ped. long term care unit	yes
20	Cerebral palsy. Tetraparesis. Deaf-mutism.	neurol. outp. clinic	yes
21	Spastic tetraparesis	ped. long term care unit	no
22	Spastic quadriplegia	orthopaedic ward	no
23	Cerebral palsy.	ped. long term care unit	yes
24	Cerebral palsy.		
28	Cerebral palsy.		
33	Encephalopathy in the shape of spastic quadriplegia.	neurol. ward	no
35	Hydrocephalus ex vacuo. Grip strength decrease in left upper limb.	neurol. outp. clinic	no
36	Encephalopathy perinatal. Quadriplegia.	neurol. outp. clinic.	no
37	Cerebral palsy	ped. neurol. outp. clinic	yes
39	Brain inflammation in childhood. Global contracture of upper and lower limb.	neurol.ward	no
40	Cerebral palsy	ped. neurol. outp. clinic	yes
42	Early childhood cerebral palsy.	ped. rehabil. ward	yes
44	(Old) right-side hemiplegia.	rehabil .ward	no
66	Cerebral palsy.	ped. long term care unit	yes
82	Spastic tetraparesis.	neurol. outp. clinic.	no

lectual impairment" (idem, item 4). CP occurred in 6 medical records as the main diagnosis (out of these 4 attendees were moved to SCFs from children care facilities) (Table 2).

A number of documents are attached to the medical record of an attendee. These are only for internal use, eg. planning of care and medical treatment of the resident, and include GP's and specialists' consultation results, psychological opinions, hospitalisation records, etc. These were analysed, and it was found out that within the group there were: 9 cases of clinical diagnosis of paresis and locomotion impairment, 10 cases of diagnosed CP (9 of which were given in childhood). The data are presented in Table 3. The correlation between the diagnosis of CP (Table 3, column 4) and treatment by pediatricians (idem, column 3, units marked with „ped." for paediatrics) was assessed by means of χ^2 test giving the result of p=0.0001 (Table 3)

Table 4 depicts the structure of the group in terms of the sex, age and clinical types of CP. High percentage of males in the research group was observed, which is confirmed by literature [1,8]. In assessing according to Ingram classification, diplegia was observed in almost every second person (45.7%) [5]. Diplegia was understood as a neurologic disorder (spastic paresis) affecting only lower limbs (5 cases) or more severe in lower limbs than in the upper limbs (11 cases) (Table 4).

The majority of the group researched (60%) was capable of independent locomotion within the place of living (GMFCS=1 or 2). The correlation between the age and locomotion capabilities measured in GMFCS scale was high in the whole group though it was statistically insignificant (p=0.11 for Spearman's correlation). However, after excluding 5 people who remained in bed (GMFCS=5), the correlation was statistically significant with p=0.047.

Additional health problems – intellectual impairment, epilepsy, as well as speech, vision, hearing and deglutition disorders were recorded during further analysis. Table 5 presents the results of this analysis arranged in numerical order. Intellectual impairment was the predominant pathology present in the researched group with the other pathologies occurring in the following order: speech disorders, epilepsy and deglutition disorders (dysphagia caused by bulbar palsy). An orthopaedic surgery was done only in 2 cases (in both of them it was Achilles tendon elongation). Table 5 shows the results of χ^2 (chi-square) test correlation between the rate of occurrence of pathologies and expected rate of occurrence of respective health problems for CP population of age 3-19 as determined by Polish epidemiologic research [8]. Significantly higher percentage of intellectual impairment in the researched group correlates with the higher rate of hearing disorders and epilepsy. Speech disorders were similarly frequent, however, the low rate of vision impairment (p=0.09) in the researched group was unexpected (Table 5).

TABLE 4. Selected qualitative and quantitative parameters of the research group (n=35).

Age (mean, SD)	47.71 (14.99)
Females (n, %)	11 (34%)
Males	24 (66%)
Diplegic	16 (45.7%)
Bilateral hemiplegic	7 (20.0%)
Hemiplegic	8 (22.9%)
Ataxic	2 (5.7%)
Dyskinetic	2 (5.7%)
GMFCS (median, min-max)	2 (1-5)
GMFCS 1-2 (n, %)	21 (60.0%)
GMFCS 3	5 (14.3%)
GMFCS 4-5	9 (25.7%)

DISCUSSION

In this research CP was identified with 35 attendees of Przemyśl district long-term care institutions. In 10 cases previous medical diagnosis was confirmed. Noticeably these cases are significantly correlated with preceding treatment in children's neurology clinics or remainąing at children's and youth care facilities. Individuals admitted to SCFs in adulthood and being under health care institutions supervision had symptom based diagnosis (upper limb athetosis – Table 3, patient number 11) or diagnosis of complex disease (spastic tetraparesis – idem, patient number 21). This may result from the very nature of the disease itself, which is a complex of disorders originating from (substantially) varied cause, defined in time – the medical diagnosis is therefore conditioned by identifying the actuation time of a particular cause of the disease. However, although this variable ("perinatal

TABLE 5. Coexisting health problems in the research group.

Dysfunction	Analyzed group		Reference group acc.T.Mieszczanek		p-level (chi-square test)
	count	percentage	count	percentage	
Intellectual impairment	34	97.1	196	51.4	<0.000
Speech impairment	22	62.9	278	62.5	0.964
Epilepsy	17	48.6	147	33.0	0.062
Bulbar dysfunction	13	37.1	-	-	-
Vision impairment	10	28.6	192	43.1	0.093
Hearing impairment	6	17.1	21	4.7	0.002
History of orthopaedic surgical intervention	2	5.7	27	6.1	0.933

TABLE 6. CP presence among the residents of long-term care facilities within the borders of the district.

Category	Total	CP count acc tab 2	CP count acc tab 3	CP count acc the study	Chi-square p-level	
					(col.3 vs 5)	(col.4 vs 5)
All	435	6 (1.4%)	10 (2.3%)	35 (8.0%)	<0.0001	0.0001
Intellectual impairment	109	5 (4.6%)	8 (7.3%)	29 (26.6%)	<0.0001	0.0002
Mental disturbance	152	1 (0.7%)	2 (1.3%)	6 (3.9%)	0.055	0.152

encephalopathy") and complex of symptoms ("tetraparesis") were both recognised correctly in the case of patient number 36, Table 3, the CP was not diagnosed. Common association of CP with developmental period and paediatrics appears to be the reason for this situation (usage of term "state after CP" describing type of anamnesis instead of medical diagnosis proves the hypothesis).

In the existing health care system, CP is diagnosed during medical consultations of children having anomalous clinical or developmental pattern revealed during screening well-child care. The diagnosis is based on recognising the anomalous pattern [7], after the central nervous system was (in childhood) affected by a deleterious factor and therefore does not have to be stated in childhood. Patient number 20 is an excellent example of this hypothesis. The patient was admitted to SCF at the age of 25 with "profound intellectual impairment, tetraparesis", after losing the ability to locomote the patient received neurological consultation and was diagnosed with CP at the age of 40.

Independently from the abovementioned conditions it can be stated that the real number of adults in SCFs suffering from CP is underestimated. In the researched group, 16 people had locomotion disabilities recognized beforehand. Table 6 shows the correlation between the rate of CP diagnosed in this research and the rate of CP revealed based on medical records for the group of attendees of SCFs in Przemyśl district. High percentage of CP among the patients with intellectual impairment (27%) is significant (Table 6).

Although it seems that medical disability certificates [6] as a method of data standardisation could improve the quantitative estimation of population suffering from locomotion disabilities, the data shown in Table 1 do not confirm this. Patients with profound or severe intellectual impairment were described with one symbolic code, although they also demonstrated neurological deficits. At the same time, patient number 43 with hemiplegia and lower right limb deformation was described properly, even though the latter fact was not mentioned in the medical record. The use of the term "intellectual impairment" without a quantifier (preventing thus the determination of the degree of disability) might be the reason for the observed situation (similar case happened with patient number 36). A number of privileges, including the financial ones, are based on locomotor disability certificates (these include orthopaedic care and rehabilitation therapy, subsidiaries which are substantial from the SCFs' perspective). Therefore it is very unlikely that patients/caretakers are not interested in receiving such certificates. Consequently, although the size of the researched group is too small to be statistically valid, profound and severe intellectual impairment seems to marginalize the importance of other significant health problems thus distorting the subjective evaluation of the patient.

Epidemiological research on the age group 3-19 conducted on the area of the former Zielona Góra province in the year 2000 revealed 445 cases of CP, among which the rate of intellectual impairment amounted to 51% [8]. Among the adult attendees of SCFs this percentage was close to 83% and the frequency difference was statistically highly significant ($p<0.0001$ while comparing both groups with the χ^2 test). This supports the view that intellectual impairment is of vital importance for the lack of life independence of people suffering from CP and thus for the need of residential care for them. The CP recognition itself seems not to affect the admittance to SCF, which additionally results in undervaluation of the number of CP cases.

The lack of guidelines regarding treatment and rehabilitation of patients with CP seems to be yet another reason for the lack of diagnosing CP in the majority of the members of the research group. In the literature, one can meet opinions about the need of extended rehabilitation in adulthood in order to prevent the loss of the locomotion abilities and delaying the development of the degenerative changes of locomotors system. On the other hand, it is evident that anomalous locomotion may facilitate the development of pain insufficiency and degenerative changes of the locomotors system. Therefore, encouraging of attendees to learn the independent locomotion by all means regardless of the stage of neurologic dysfunction is often negated. Alternatively, it is proposed to correlate the expected result of rehabilitation with the neurological state of the patient [9]. In this research, a statistically significant correlation between the age and the decrease of locomotion abilities in GMFCS scale was observed (as attendees with the most severe dysfunctions, i.e. GMFCS 5, who did not develop the ability to locomote were excluded from the group), which conforms to both of the concepts presented above. However, being the subject of scientific discussion, these concepts may not be the basis for the systematic solutions. Therefore, diagnosing an adult with CP does not necessitate any specific treatment and thus it does not bear any significance for the medical personnel/caretakers.

Current standards of long-term care [10] demand that the people with intellectual impairment and with mental disorders should be accommodated in separate units (buildings) with specific types of care. The features distinguishing patients with CP from both of the abovementioned categories should be stressed here. The main problems and the nature of the disease that last since childhood are locomotion disorders. Coexisting stimuli perception disorders negatively affect the psycho-intellectual development. Disartry and disphasy hinder contacts with the environment and the development of social functions. Additionally, locomotion deficits are the reasons for isolating the patient from others (in the development period – from peers).

Considering the abovementioned facts, no appropriate conditions seem to exist that would compensate for the intellectual deficit caused by the primary brain defect [11], and the episodes of convulsions (as well as treating them) may even make it worse. Additionally, it is possible that the patients' level of intelligence will be underestimated in psychological evaluation, especially if s/he suffers from tetraplegia [12]. The lack of quantitative and qualitative assessment of a disease results in the improper treatment of the patient, e.g. resigning from rehabilitation or group therapy due to the expected unwillingness to cooperate. The thesis of the lack of the correct assessment of the disease is indirectly confirmed by the percentage of people with visual disorders, which is lower in the research group than in the reference group (Table 5, line 5). Visual disorders exhibit less obvious clinical symptoms than speech disorders or epilepsy and do not

affect an attendee's contacts with the caretakers to such a degree as audio impairments. It is then possible to put forward a hypothesis that within the research group there are people with visual disorders that have not been diagnosed (and treated), which can restrict locomotion abilities and thus their functioning in the group. This issue may necessitate further research.

CONCLUSIONS

Adults suffering from cerebral palsy are a group distinct from other attendees of social care facilities due to the diversity of body dysfunctions. This should be a reason enough to form a separate organisational, medical and social category of long-term care. Such dispensarisation is purposeful as confirmed by the observed tendency to marginalise coexistent health problems, liable to treatment, in patients with profound intellectual impairment.

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