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Development and initial validation of the polyhandicap severity scale

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Abstract

Objectives

Providing a new tool, based on the point of view of experts in polyhandicap, which assesses the global severity of the health status of polyhandicapped persons is necessary. We present herein the initial validation of the polyhandicap severity scale (PSS).

Methods

The initial development of the tool was undertaken in two steps: item selection and validation process. The final set included 10 items related to abilities and 17 items related to comorbidities and impairments. The patient selection criteria were as follows: age > 3 years, age at onset of cerebral lesion under 3 years old, with a combination of motor deficiency and profound intellectual impairment, associated with restricted mobility and everyday life dependence. External validity, reproducibility (20 patients), responsiveness (38 patients), and acceptability were explored.

Results

During the 18-month study period, a total of 875 patients were included. Two scores were calculated: an abilities score and a comorbidities/impairments score (higher score, higher severity). The 2 scores were higher for: older patients, patients with a progressive etiology, patients with more devices and more medications, patients with higher dependency and lower mobility. Indicators of reproducibility and responsiveness were satisfactory. The mean time duration of fulfilling was 22 minutes (standard deviation 5).

Conclusions

Quantifying the health severity of polyhandicapped persons is necessary for both healthcare workers and health decision makers. The polyhandicap severity scale provides the first reliable and valid measure of the health severity status for children and adults.

Keywords

polyhandicap; health status; severity; scale; validation

I. Introduction

Polyhandicap is a chronic complex disability condition that occurs in an immature brain, leading to a combination of profound mental retardation and serious motor deficit, and resulting in an extreme restriction of autonomy and communication [1]. This definition was adopted by the French scientific community and French law (French Law No. 89-798, 1989, October 27th, health policy of care disability) and has been accepted in recent literature [2-5]. Other words may be used to designate these patients. The most frequent term is profound intellectual and multiple disabilities which is close to polyhandicap but does not systematically refer to a disorder affecting an immature brain [6]. Polyhandicap is a syndromic entity affecting the developing brain that is caused by both progressive and nonprogressive etiologies. In France, the incidence is estimated at 880 new cases per year [7].

Children and adults with polyhandicap present various degrees of neurodevelopmental alterations including mobility and posture, communication and language, coordination and sociability. They may develop comorbidities and associated impairments which strongly differ from one patient to another, which increase with aging [2, 5, 8]. The heterogeneity of the disabilities and, the chronicity and expandability of the disease explain the large panel of severity of the health status that the patients present, leading to various disease courses, medical and educative needs [4].

Therefore, quantifying health severity is now necessary both for healthcare workers and health decision makers. According to the severity status, the (educative and medical) care management, the geographical distribution and the health resource allocation would be optimized. A review of the literature has revealed that there is no tool that specifically measures this construct in this population. Some tools allow for a partial picture of the severity. For cerebral palsy populations, the Gross Motor Function Classification System (GMFCS) assesses mobility [9], and the Top Down Motor Milestone Test assesses motor skills [10]. Autonomy may be assessed using the Functional Independence Measure scale [11], even if the measure wrongly discriminates patients with a high level of human and technical dependence. Mental disability is often inaccurately assessable using the intelligence quotient. Neurodevelopmental status may be estimated using an adapted version of the Brunet-Lézine scale, originally used to assess infants up to 24 months old [12]. These scales have some limitations, i.e. they have not been validated on polyhandicapped persons while

they present the most severe profiles regardless of autonomy, motor and mental disabilities; and they never consider comorbidities and impairments that strongly contribute to the severity of the health status. In this context, it is necessary to provide a new tool, based on the point of view of a large panel of experts in polyhandicap, which assesses the global severity of the health status of polyhandicapped persons.

The objective of this research was to develop a severity score for patients with polyhandicap that can be easily applied, in research and clinical practice. We present herein the initial validation of the polyhandicap severity scale (PSS).

II. Material and method

2.1. Design and settings

The initial development of the tool was undertaken in two steps: item selection and validation process. The global process was supervised by a steering committee comprising a neuro-pediatrician, a specialist in physical and rehabilitation medicine, a specialist in epidemiology, a pediatrician, a general practitioner, and a member of patient association.

2.2. Selection of items

For the first step, 2 neuropsychiatrists who had experimented with the management of patients with polyhandicap for more than 15 years, suggested candidate severity criteria on the basis of an international literature review. The search was performed within the past 20 years (1984 to 2014; 1st included patient in the study: 1/3/2015) using the Medline database. Medical subject headings (MeSH) reflecting the actual key search concept were determined by the following significant terms: "Polyhandicap" or "Profound Multiple Disability(ies)" or "Profound Intellectual and Multiple Disability(ies)" or "cerebral palsy". The first search yielded a total of 436 citations, and a second manual selection was performed based on the titles and the abstracts of the citations, excluded all citations not focusing on the health status. For each citation, items related to symptom(s), disability(ies), comorbidity(ies), and impairments(s) were recorded and classified as 'certainly', 'probably', or 'not probably' related to polyhandicap. The first set of items was screened by a panel of 11 experts including health care professionals with a minimal experience with polyhandicap of 5 years (one neuropsychiatrist, one pediatrician, one physical and rehabilitation medicine physician, one general practitioner, one physiatrist, one nurse, one nurses' aide, one member of patients' association, one psychologist, one epidemiologist, and one expert on the

development of health measures). The experts agreed that the severity of the polyhandicap resulted both from the consequences of the neurodevelopmental delay that resulted in motor and cognitive deficiencies and from the accumulation of impairments and associated comorbidities that degraded the patients' overall health status. Thus, they selected items assessing the neurodevelopmental level of the polyhandicapped persons and items relative to the cumulated impairments. During 3 meetings, the items were discussed regarding their exclusion or retention, their formulation, and the modalities of quotation. An intermediary set was tested on 20 patients for feasibility. During a last consensus meeting, the panel of experts provided the final set of items after item reformulation, if necessary. The final set included 10 items related to abilities (general posture, supine posture, sitting posture, walking/moving, visual contact, language, communication, feeding, continence, and manual grasping) and 17 items related to comorbidities and impairments (main physical disability, other physical disabilities, drug-resistant epilepsy, dorso-lumbar column disability, other orthopedic disabilities, respiratory disability, digestive disability, dental disability, behavioral disorders, sensorial disability, pain, sleep disorders, skin disability, urinary disability, ventilation devices, other invasive devices, other disability). The details of the items and their response modalities are provided in the supplementary material (Appendice. Final set of items of the Polyhandicap Severity Scale).

2.3. Population and validation process

The validation process was included in the French national Polyhandicap study [2, 4, 5]. This study was implemented from March 2015 to September 2016 in various French centers: 4 specialized rehabilitation centers, 9 residential facilities, and a university hospital center (Paris, France). The general aim of the study was to identify the potential (socioeconomic, environmental, epidemiologic) determinants of the health status of the patients (clinical trial registration number NCT02400528; French ethics committee, 20/10/2014, number 2014-A00953-44). A written consent form was collected for each participant. The patient selection criteria were as follows: age > 3 years, presenting with a combination of motor deficiency (tetraparesia, hemiparesis, paraparesia, extrapyramidal syndrome, cerebellar syndrome, and/or neuromuscular problems), profound intellectual impairment (intelligence quotient <40) associated with everyday life dependence (Functional Independency Measure (FIM) <55), and restricted mobility Gross Motor Function Classification System (GMFCS) III, IV, and V); in addition, the age at onset of cerebral lesion was under 3 years old.

2.4. Data collection

The final set of items was included in the data collection of the French national Polyhandicap study. The items were completed by the healthcare workers in charge of the patient. The following data were collected to describe the sample and assess the external validity of the tool:

- 1) Sociodemographic data: age, age categories (child, adult), gender.
- 2) Aetiology status: classified as unknown or known, progressive or non-progressive.
- 3) Health status:
 - Impairments: i) severe motor impairments: tetraparesis, paraparesis, and hemiparesis; ii) other neurologic impairments: movement disorders, severe dystonia, global hypotonia, extrapyramidal syndrome; and iii) behavioral disorders (including withdrawn behavior [13], intermittent screaming and crying, agitation, self-aggressivity or hetero-aggressivity, stereotypies and/or merycism).
 - Comorbidities: epilepsy (yes/no), at least one previous onset of status epilepticus reported in the medical file, drug-resistant epilepsy defined by more than 4 seizures per month despite adapted anticonvulsant treatment, including at least 3 anticonvulsant drugs), orthopedic comorbidities (scoliosis, limb deformations, limb fractures, hip luxation), and pulmonary comorbidities (pulmonary recurrent infections (≥ 5 /yrs), aspiration syndrome).

2.5. Analysis

2.5.1. Scoring

An abilities score was calculated as the sum of the 10 item scores, ranging from 0 (best possible ability) to 60 (worse possible ability). A comorbidities/impairments score was calculated as the sum of the 17 items scores, ranging from 0 (none) to 69 (more comorbidities/impairments).

2.5.2. External validity

To explore the external validity, the relations between the scores of severity were determined by assessing the associations between the 2 scores and the sociodemographic and clinical features. For qualitative variables, the mean dimension scores of the PSS were compared across patient groups that were expected to differ (e.g. age categories, etiology, care management structure, profound mental impairment (IQ <25), GMFCS classes, recurrent pulmonary infections, and drug-resistant epilepsy) using Student's t test.

Quantitative variables (e.g. age, number of medical devices/medications, neurodevelopmental status, and FIM score) were analyzed using Pearson's correlation coefficients. The following hypotheses were made: i) abilities scores should be linked to aetiology status (progressive, non-progressive), number of medical devices, neurodevelopmental status, FIM score, GMFCS level, recurrent pulmonary infection, and drug-resistant epilepsy; and ii) comorbidities/impairment scores should be linked to age, aetiology status (progressive, non-progressive), number of medical devices, neurodevelopmental status, FIM score, GMFCS level, recurrent pulmonary infection, and drug-resistant epilepsy.

2.5.3. Reproducibility and responsiveness

Reproducibility (the ability to produce the same results in the absence of a meaningful change) and responsiveness (the ability to detect a meaningful change) were assessed. To test reproducibility, a second assessment was performed for 20 patients 3 weeks after the first assessment. The patients were excluded in the case of a change of health status (defined by the need of hospitalization (intensive care unit, surgical intervention) or any new significant health event (pneumopathy, iterative epileptic seizures, status epilepticus, aggravation of motor/neurologic impairments, limb fracture) occurring between the 2 assessments. The 3-week interval selected for the test–retest reliability was selected to minimize the likelihood of changes in the patients' clinical status. To test responsiveness, a second assessment was performed on 38 patients, 5 years after the first assessment. Two groups were distinguished: those without change of health status (stable) and those with a change of health status (worsened). A worsened patient was defined according to: i) having a progressive etiology of polyhandicap, and ii) changing to a more medicalized care management modality. The 5-year interval selected for responsiveness was selected to maximize the likelihood of changes in the patients' clinical status.

2.5.4. Acceptability

The number of missing data for each item, and the duration of fulfillment were recorded.

III. Results

3.1. General characteristics of the sample

During the 18-month study period (March 2015 to December 2016), a total of 875 patients aged 3 to 68 years were included. A total of 410 patients were cared for in specialized

rehabilitation centers, 372 were cared for patients in residential facilities, and 83 were cared for at home. All the characteristics are provided in Table 1.

3.2. External validity

All the assumptions formulated a priori were ascertained. Older patients presented higher scores of severity, for both the abilities and the comorbidities scores. The patients with a progressive etiology presented higher severity scores. Patient care managed in specialized reeducation centers was more severe than that of patients cared for at home or those cared for in residential facilities. More medical devices and more medications were associated with higher severity. The Brunet-Lézine scores, FIM score, and GMFCS level were correlated with severity. All the details are provided in Table 2.

3.3. Reproducibility and responsiveness

The reproducibility, tested on 20 patients, was very satisfactory for both dimensions (abilities and comorbidities/impairments). Responsiveness was tested for 38 patients: 22 individuals were defined stable and 16 individuals were defined as worsened. As expected, the scores of the tool were stable for the stable patients. For the worsened patients, the tool only captured a change in the comorbidities score and did not capture the abilities score. All the details are provided in Table 3.

3.4. Acceptability

The number of missing data for each item was low (<10%) and the mean time duration of fulfilling was 22 minutes (standard deviation 5).

IV. Discussion

Polyhandicap, as a complex disability condition, includes a heterogeneous group of patients in terms of health severity. The consequences of neurodevelopmental retardation (inducing various deficiencies of motor skills, praxis, autonomy and communication abilities) and the occurrence of comorbidities and impairments (epilepsy, pulmonary/urinary infections, scoliosis, food and/or salivary aspiration, gastrointestinal reflux, etc.) should be more accurately captured. It is essential, both for health care providers and health decision makers, to have a marker of the severity of the individuals. For the first time, a measure of health severity of polyhandicapped individuals is available. By allowing for better profiling of the severity of the polyhandicapped individuals, it would be easier to optimize their care

management, to adapt their care organizations and to guide them towards more appropriate care structures.

In France, the care of polyhandicapped individuals is rarely care managed at home; those who are cared for at home are essentially the youngest children. There are two main care management modalities: specialized rehabilitation centers and the residential facilities. For these two modalities, units are dedicated for adult and pediatric populations. The rehabilitation centers are supposed to manage the more severe cases by offering a high level of medical and paramedical physical rehabilitation and the residential facilities are supposed to manage the less severe cases by offering a high level of psychosocial education. A recent study has shown that a large proportion of patients are probably not care managed for in the most appropriate care structure with regard to their health status and medical/educational needs. This inadequacy could not only have consequential impacts on the health but well-being of patients and families and also on the optimization of health expenditures. Therefore, we are convinced that the use of a more robust definition of health severity as measured by the polyhandicap severity scale, may help i) to more precisely capture this inadequacy, and ii) to measure effect of future specific actions that could be implemented by health care authorities to correct this inadequacy .

Health-care providers should also benefit from this severity scale. Regularly measuring the health severity should drive therapeutic and preventive strategies, even surgery interventions, and adjust health and educational needs, thereby offering to polyhandicapped persons a coherent, adequate and integrative life plan during the disease course. The polyhandicap severity scale, as a useful tool in daily clinical practice, will provide an accurate health severity level regarding abilities, comorbidities and impairments.

Last, researchers, who are always looking for robust measures, may consider the polyhandicap severity scale as an interesting valid measurement of objective effects of various educational or medical strategies. We want to believe that, in the future, promising therapies should be assessed through clinical studies, using this kind of tool.

Strengths and limitations

- The study design allowed us to assess core psychometric properties, such as reproducibility and sensitivity to change, which are two core psychometric properties of a measuring instrument. These two properties are rarely reported due to longitudinal

data requirements. Our findings confirmed that the polyhandicap severity scale has a satisfactory ability to produce the same score between two assessments and a satisfactory ability to detect a meaningful health change, which are core psychometric properties of a measuring instrument [14 , 15]. This measure could be used to monitor the response to any intervention.

- The content of the scale was mostly based on experts' points of view. The group of experts included only one member of the parents association. While in the specific context of polyhandicap, the "voice" of the patients was not available, we intend to take into account the families' point of view in the future. Minor changes may be incorporated during the process.
- The proportion of persons who were care managed for at home was small. Future works could explore more carefully the validity of the tool in a higher sample of homecare persons.

Conclusion

Quantifying the health severity of polyhandicapped persons is necessary for both healthcare workers and health decision makers. The polyhandicap severity scale provides the first reliable and valid measure of the health severity status for children and adults.

Ethics approval and consent to participate

Regulatory monitoring was performed according to the French law that requires the approval of the French ethics committee (Comité de Protection des Personnes Sud Méditerranée V, Centre Hospitalier Universitaire de Nice, Hôpital de Cimiez, Nice, France, Approval date 20/10/2014, reference number 2014-A00953-44). A written consent form (from written parental / legal guardian) was obtained for each participant.

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- **Declaration of interests:** none.

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Table 1. Sample characteristics

		N=875
		N (%)
1. Sociodemographics		
Age	M±SD	24.5±16.8
Age categories	Children	401 (45.8)
	Adults	474 (54.2)
Gender	Boys/Men	466 (53.3)
	Girls/Women	409 (46.7)
2. Etiology of polyhandicap		
Known		744 (86.2)
Unknown		119 (13.8)
Progressive		235 (27.4)
Non progressive		622 (72.6)
3. Impairments		
Severe motor impairments	Tetraparesis	611 (78.6)
	Paraparesis	142 (18.3)
	Hemiparesis	24 (3.1)
Other neurologic impairments	Movement disorders	107 (12.8)
	Severe dystonia	79 (9.6)
	Global hypotonia	233 (28)
	Extrapyramidal syndrom	193 (23.3)
Behaviorial disorders [^]		622 (72.2)
4. Comorbidities		
Epilepsia	Presence of epilepsia	478 (55.5)
	Previous status epilepticus	188 (26.8)
	Drug-resistant epilepsia	165 (19.1)
Orthopedic	Scoliosis	474 (57.2)
	Limb deformation	599 (70.5)
	Limb fracture	57 (6.8)
	Hip luxation	219 (26.3)
Pulmonary	Recurrent pulmonary infections	90 (10.5)
	Aspiration syndrome	212 (24.8)

^ Behavioral disorders: intermittent scream 64%, agitation 62%, stereotypies 32%, intermittent crying 50%, self-aggressivity 15%, and hetero-aggressivity 8%

Table 2. External validity

		Abilities score*	Comorbidities score*
Age	R	0.15	0.22
	p-value	<0.001	<0.001
Age categories	Children	41.6±11.5	9.5±4.3
	Adults	45.1±10.1	11.3±4.4
	p-value	<0.001	<0.001
Etiology	Non progressive	42.0±11.0	9.6±4.1
	Progressive	47.5±9.4	12.8±4.4
	p-value	<0.001	<0.001
Care management structure	Specialized rehab. center	47.3±8.9	12.9±4.1
	Residential facility	40.0±11.6	8.5±3.4
	Home	40.3±10.7	7.2±3.2
	p-value	<0.001	<0.001
Number of medical devices*	R	0.38	0.33
	p-value	<0.001	<0.001
Number of medications	R	0.32	0.44
	p-value	<0.001	<0.001
Neurodevelopmental status			
Postural	R	-0.57	-0.28
	p-value	<0.001	<0.001
Coordination	R	-0.51	-0.25
	p-value	<0.001	<0.001
Language	R	-0.56	-0.23
	p-value	<0.001	<0.001
Sociability	R	-0.62	-0.26
	p-value	<0.001	<0.001
Profound mental impairment IQ <25	No	30.7±10.1	7.4±3.3
	Yes	45.9±9.3	11.0±4.4
	p-value	<0.001	<0.001
FIM score	R	-0.58	-0.20
	p-value	<0.001	<0.001

GMFCS level	III	31.6±11.3	8.0±3.8
	IV	40.1±11.5	10.0±4.1
	V	47.0±8.3	11.1±4.4
	p-value	<0.001	<0.001
Recurrent pulmonary infections	No	42.8±10.9	10.1±4.3
	Yes	50.1±9.1	13.8±4.7
	p-value	<0.001	<0.001
Drug-resistant epilepsy	No	43.0±10.7	10.6±4.3
	Yes	45.9±11.4	10.0±4.6
	p-value	0.002	0.16

R correlation coefficient; *Higher the scores, higher the severity

*

Table 3. Reproducibility and sensitivity to change

	Reproducibility	Sensitivity to change N=38			
	N=20	Stable		Worsened	
		ICC	N=22	N=16	Delta
		Delta	ES	Delta	ES
Abilities score	0.99	0.54±2.44	0.02	0.87±3.99	0.02
Comorbidities score	0.98	-0.05±2.08	0.02	3.94±4.36	0.62

ICC, intraclass correlation coefficients ; Delta, delta change between baseline assessment and 3 weeks assessment ; ES, effect size (final score – initial score)/initial score (|0,2| small, |0,5| moderate, and |0,8| large change [Algina J, Keselman HJ, Penfield RD: Effect Sizes and their Intervals: The Two-Level Repeated Measures Case, Educational and Psychological Measurement 2005, 65(2):241-258]),