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Health issues in polyhandicapped patients according to age:

Results of a large French cross-sectional study

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Interpretation of data: MCR, TBV, KB, PA.

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Abstract

Introduction. A better understanding of the natural course of the health status of patients with polyhandicap may optimize preventive and curative care management. From a large sample of patients aged from 3 to 25 years, we reported the description of their health status.

Methods. This was an 18-month cross-sectional study including patients aged from 3 to 25 years with a combination of severe motor deficiency and profound intellectual impairment. The patients were recruited from 4 specialized rehabilitation centers, 9 residential facilities, and a pediatric/neurological department. The following data were collected: polyhandicap etiology, health status (impairments, comorbidities, and neurodevelopmental status), medical devices, and rehabilitation procedures.

Results. A total of 545 patients were included (N=80 [3-5 years], N=166 [6-11 y], N=155 [12-17 y], and N=144 [18-25 y]). The etiology of polyhandicap was unknown for 11.5% of the cases. Behavioral disorders and (orthopedic and digestive) comorbidities were more frequent in the oldest age classes. The neurodevelopmental status of the patients was close to those of a 5- to 7-month-old child without progression across age. Gastrostomy was the most frequent device needed by the patients.

Discussion/Conclusion. Early detection and management of impairments and comorbidities may improve the disease course of the patients.

Keywords

Health status; polyhandicap; disability; comorbidity; mental retardation

Introduction

Polyhandicap (PLH), as a recently defined French concept [1-3], the PLH concept refers to a complex disability condition corresponding to a chronic affliction occurring in an immature brain, leading to the combination of severe/profound mental retardation and serious motor deficit, resulting in an extreme restriction of autonomy and communication. This term is close to the notion of profound intellectual and multiple disabilities, which is used in other countries, and does not systematically refer to a disorder affecting an immature brain. Polyhandicap is a syndromic entity and meets several progressive and non-progressive etiologies. The scarce data available in France enable an estimation of the prevalence of polyhandicap as between 0.7 and 1.3 per thousand, i.e. [4]. Due to the combination of severe motor deficit and profound mental retardation, the patients, completely dependent on human and technical aid, develop with aging, various comorbidities and -impairments [5], which can lead to premature death [6]. Few data examined the evolution of the health status, kinetic occurrence of comorbidities/impairments, and adaptation of care management during the life of the individual, including childhood, adolescence, and adulthood. However, it allows a better understanding of the natural course of life of these patients and consequently optimizing the preventive and curative care management strategies.

Previous studies concerning the description of the health status of patients with polyhandicap have some limitations: performed on a small- and intermediate-sized sample (less than 200 patients), including heterogeneous samples (mixture of severe and less severe patients, such as cerebral palsy and intellectual disability patients), restricting younger children, based on retrospective design, and focusing on very specific issues (such as sensory perception, respiratory disorders, digestive disorders, etc.) [7-15]. For the first time, we reported from a large sample of young polyhandicapped patients aged 3 to 25 years, according to the age-related developmental periods of the children (early childhood 3 to 5, childhood 6 to 11, adolescence 12 to 17, and early adulthood 18 to 25 years old): i) the description of their health status (including severity, comorbidities and impairments,

neurodevelopmental status) and ii) the description of the devices and the rehabilitation procedures they benefit.

Methods

▪ Design and settings

This study was a cross-sectional study including patients from March 2015 to September 2016. The recruitment of patients was performed from 4 specialized rehabilitation centers, from 9 residential facilities of the 17 centers of the French Comité d'Études, d'Education et de Soins Auprès des Personnes Polyhandicapées Association (CESAP), and from a specialized pediatric/neurological department of a university hospital (Service de Neuropédiatrie, UPMC, Hôpital Trousseau, Assistance Publique Hôpitaux de Paris, France) during a routine visit. This study was included in the French national PoLyHandicap (PLH) cohort.

▪ General organization of the PLH cohort

The French national PLH cohort was implemented in various French centers spread over different territories: specialized rehabilitation centers, residential facility, and a university hospital center (Paris, France). The general aim of the cohort was to identify the effect of (socioeconomic, environmental, epidemiologic) determinants on the health status of the patients and the daily life of their (natural and institutional) caregivers (clinical trial registration number NCT02400528). Three different populations were eligible: i. Patients with polyhandicap; ii. Familial referents of the included patients (French legal mention for this type of patients, represented by parents in the majority of cases); and iii. Institutional health care workers of the included patients. The present study focused on patients.

▪ Selection criteria

The patient selection criteria were as follows: aged from 3 to 25 years; with a polyhandicap defined by the combination of motor deficiency (quadriplegia, hemiparesis as a predominant hemibody motor impairment, diplegia, extra pyramidal syndrome, cerebellar syndrome, and/or neuromuscular

problems) and severe/profound mental impairment (intelligence quotient <40; for patients older than 5 years old: IQ= developmental age below 2 years old; for children from 3 to 5 years old: IQ= developmental quotient<40%) associated with everyday life dependence (Functional Independence Measure <55), and restricted mobility (Gross Motor Function Scale (GMFCS and GMFCS-ER), III, IV and V [16]); age at onset of cerebral lesion below 3 years old. A written parental / legal guardian consent form was obtained for each participant.

▪ **Data collection**

Data were collected from medical records obtained by a dedicated clinical research assistant and was supported by the referent physician of the patient (a referent physician is designated for each patient). The data included the following items:

1) Socio demographic data: age category based on age at inclusion (4 age classes were defined in accordance with the age-related development periods of children[3-5], [6-11], [12-17], [18-25] years, gender, and structure of care management (specialized rehabilitation center, residential facility, at home),

2) Etiologies of polyhandicap: unknown etiology, known etiology (central nervous system malformations, neurometabolic and neurogenetic encephalopathy, epileptic encephalopathy, pre/perinatal and post-natal causes).

3) Health status:

- Global health severity: i. severe for patients who meet all the following criteria: motor handicap (diplegia or quadriplegia and/or extrapyramidal syndrome and/or severe general hypotonia), IQ <25, FIM <=20, and GMFCS IV and V; ii. less severe for patients who do not meet these criteria,
- Global health stability: i. unstable for patients who meet at least one of the following criteria: recurrent pulmonary infections (>=5/yrs), drug-resistant epilepsy (>=4 seizures/month); ii. stable for patients who do not meet any of these criteria,
- Motor deficiency: quadriplegia, diplegia; movement disorders, severe dystonia, severe hypotonia, extrapyramidal syndrome, ataxia;

- Associated handicaps: i) neuro-sensorial handicaps: visual impairment (partial/complete blindness) and hearing impairment (partial/complete deafness); ii) behavioral disorders (including intermittent screaming, and/or agitation and/or stereotypies and/or intermittent crying and/or self-aggressivity and/or hetero-aggressivity), iii) sleep disturbance (short sleep, night wake up, and difficulties falling asleep).
- Co-morbidities: epilepsy (yes/no, previous status epilepticus, drug-resistant disease), orthopedic (scoliosis, limb deformations, limb fractures, hip luxation, previous arthrodesis and/or other previous orthopedic surgery), pulmonary (pulmonary recurrent infections, aspiration syndrome), digestive (fecal impaction, gastro esophageal reflux, and drooling), urinary (recurrent urinary tract infections-at least once a year and urinal retention), cutaneous (bedsores, pressure sore), chronic pain, chronic diseases (at least one of the following diseases: vascular stroke, myocardial infarction, diabetes, and/or cancer), antibiotic resistant bacteria.
- Neurodevelopmental patterns: i. autonomy: seven facets of autonomy were scored by the referent physician from 1 (worse) to 7 (best autonomy/developmental degree) for the following domains: visual contact, oral language, postural ability, grasping ability, moving ability, cleanliness, and feeding ability; ii. general neuro-developmental status was assessed using the Brunet-Lézine scale, a developmental quotient assessment tool available for infants up to 24-month old. This developmental quotient cannot be used in patients over 2 years old therefore in the present study, only the 4 developmental domains (language, posture-motor abilities, coordination, and sociability) were used. All scores ranged from 0 to 24 months.

4) Medical devices and rehabilitation procedures management:

- Medical devices: at least one, number, and type (invasive mechanical ventilation, non-invasive mechanical ventilation, tracheotomia, nasogastric tube, gastrostomy, permanent urinary probe, cerebrospinal fluid derivation, and central venous catheter).
- Rehabilitation procedures: bed without orthosis, limb orthosis, verticalization device.

- **Ethics**

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, 20/10/2014, reference number 2014-A00953-44). A written consent form was obtained for each participant. Clinical trial registration number NCT02400528.

- **Statistics**

Four age classes were constituted from age at inclusion:[3-5 y], [6-11 y], [12-17 y], [18-25 y]. Demographics, health status, and medical devices/rehabilitation parameters were described for each age class. Trends over age classes were assessed (Jonckheere-Terpstra test and Cochran-Armitage test). Statistical analyses were performed using SPSS software (IBM SPSS PASW Statistics Inc., Chicago, Ill USA). All tests were two-sided. The threshold for statistical significance was established at $P < 0.05$.

Results

- **General characteristics of the sample**

During the 18-month study period, a total of 545 patients (participation rate 90%) were included: 80 patients were aged from 3 to 5 years, 166 from 6 to 11 years, 155 from 12 to 17 years, and 144 from 18 to 25 years. The patients' sex ratio increased with age: 1.0 for the [3-5 y] and [6-11 y] classes, 1.2 for [12-17 y] class, and 1.4 for [18-25 y] class. The youngest patients ([3-5 y] class) were predominantly care managed at home (60%), whereas the oldest age classes were shared between specialized care centers and residential facilities. All of the details are provided in Table 1.

- **Health status**

Less than half of the individuals were defined as severe according to the definition described above independent of the age classes. Thirty-six to 37% of the individuals were defined as unstable for the 3

first age classes (from 3 to 17 years) while only one-quarter of the individuals aged 18-25 years were unstable. All details are shown in Table 1.

- Etiologies

Etiologies of polyhandicap were unknown for 11.5% of the cases. Among the known etiologies, 39% (n=183) of the patients presented central nervous system malformations, 5% (n=25) neurometabolic diseases, 4% (n=19) neurogenetic diseases, 2% (n=10) epileptic encephalopathies, and 163 (34%) patients presented an acquired disease, including pre-perinatal (N=129) and post-natal (N=34) etiologies, and 15% (n=70) other causes. All of the details are provided in Table 2.

- Associated impairments

Eighty to 85% of the patients presented quadriplegia. The proportion of movement disorders was lower (12%) for the oldest individuals (18-25 years) compared with the other age classes (16% for the 3 youngest age classes). Severe dystonia was reported for 18% of the youngest age class (3-5 years) and decreased with aging (10-11% for the oldest classes). Global hypotonia decreased with aging: 49, 38, 33, and 25%, respectively, for [3-5 y], [6-11 y], [12-17 y], [18-25 y] classes. Ataxia was reported at a similar proportion for the 4 age classes. Twenty-four to 36% of the children presented with visual impairment, and 4 to 8% presented with hearing impairment with no evident change over time. Behavioral disorders were less frequent in younger children (40%) compared with older individuals (more than 70% for the children older than 12 years). Sleep disorders were more frequently observed in younger children ([3-5 y] and [6-11 y] age classes) compared with older patients. All details are shown in Table 3.

- Comorbidities

Approximately 60% of the sample, independent of the age class, presented epilepsy. Drug-resistant epilepsy was less frequent for the oldest individuals (18% for the [18-25 y] class) compared with the youngest classes (31% for [3-5 y] and [6-11 y] classes). Although orthopedic comorbidities consequently increased with aging, pulmonary comorbidities (recurrent pulmonary infections and aspiration syndrome) remained stable. Although the proportion of gastro esophageal reflux was

stable over age, the proportion of fecal impaction regularly increased with the age of the patients (29, 33, 41, and 47% in the 4 successive age classes). The proportion of drooling was stable across ages. Recurrent urinary tract infections were more frequent in the older age. The proportion of urinal retention was lower for the youngest age class (3%) compared to the 3 oldest age classes (approximately 4-5%). Cutaneous comorbidities (pressure fragility and bedsores) regularly increased with age. Chronic pain proportions differed between age classes. Antibiotic resistant bacteria occurrence did not change across the 4 age classes. All details are shown in Table 4.

- **Neurodevelopmental status:**

All autonomy scores were low (1 to 4/7); scores tended to decrease or to be stable in the oldest age classes. From Brunet-Lezine, the developmental level of polyhandicap patients was very low (4 to 7 months) for all domains and patients did not demonstrate any progression with age in none of the four domains of the Brunet-Lézine scale. All details are shown in Figure 1.

- **Medical devices and rehabilitation procedures:**

The proportion of patients with at least one medical device increased with age: 32% for the [3-5 y] class and over 40% for the 3 other age classes. Gastrostomy was the most frequent device that patients needed (30 to 40%). Very few patients required permanent urinary probe, central venous catheter, or tracheotomy. Rehabilitation procedures such as daily installation of patients in sitting device (wheelchair sitting and positioning devices such as contoured seat intended to increase postural stability), limb orthosis (ankle-foot orthosis, or knee ankle-foot orthosis, wrist-hand splint, ..) and daily installation in verticalization devices did not differ across the 4 age classes. All medical devices and rehabilitation procedures are shown in supplemental file.

Discussion

The present study provides for the first time a substantial description of the health status and the care management of a large sample of French young patients with polyhandicap according to four age classes, from 3 to 25 years. This description of the 4 age classes presents interesting proposals of

the evolution over time as key elements for health care workers to optimize the preventive and curative care management of these patients.

A higher proportion of the youngest children were managed in home care. It is probable that families succeed in keeping their child at home during the first years of life, even those who presented with severe health status and a higher level of dysautonomy. However, the families were most likely no longer able to take care of them when the child grows or becomes an adult (due to their own aging, psycho-social, and organizational reasons). The instability of health status and the progressive nature of the disease, more frequently reported for patients cared for at home, can result in important difficulties to face in these singular situations at their own home and induce parents and families to orientate the patient, after some years, towards a structure offering continuous medical monitoring. The proportion of patients with an unknown etiology of the polyhandicap (11.5%) was relatively lower than the proportion provided in previous studies, which ranged from 15 to 60% [17, 18]. This finding may be partially explained by the fact that we focused on a population including young individuals while other studies included mixed (children and adult) populations. The spreading of etiological investigation and genetic counseling during the last years contribute to the decrease in the part of unknown etiologies [19]. Nearly 50% of the known etiologies were constitutional diseases (including malformations of the central nervous system and genetic disorders), while acquired etiologies constituted a third of the total cases. The acquired etiologies had more likely prenatal or perinatal origins, as previously highlighted in a European study showing a decline in post-natal cerebral palsy cases [20]. Approximately 6% of the etiologies were the consequences of rare diseases, as progressive encephalopathies well-known diseases leading to premature death. A (very) low proportion of our patients presented epileptic encephalopathy, a well-known disease with severe cognitive deficiency and difficulty to control the seizures [21]. We are convinced that this proportion was underestimated due to the difficulty to rebuild the entire medical history of these patients since birth.

As an informative finding, we found that the global severity of the patients remains stable over the age classes. This stability over time may be due to the definition of the global severity, which we have based on non- or few-progressive clinical aspects (including motor impairment and/or cognitive deficiency). Developing a severity index comprising both fixed and progressive clinical aspects as comorbidities and associated impairments may better reflect the global severity of the patients. However, the stability notion varied throughout growth: older age classes were less unstable. In this study, the stability notion was partially based on drug-resistant epilepsy. Two explanations may concur: first, drug-resistant epilepsy is known to be less frequent when the child is growing (particularly for children with epileptic encephalopathy) and second, children with progressive etiologies, such as neurometabolic etiologies frequently associated with resistant epilepsy, usually die before adulthood [21].

Not surprisingly, a large majority of our patients presented a heavy motor impairment (quadriplegia 85%), which is consistent with the pre-defined selection criteria of our cohort. For other neurological impairments, movement disorders, severe dystonia, and global hypotonia were more frequent in younger. Movement disorders and dystonia are often linked to progressive diseases leading to premature death. And it is known that early cerebellar lesions were associated to hypotonia which decreased throughout growth.

Expectedly, sensorial (visual and hearing) deficiencies remained stable throughout aging because the deficiencies directly result from the etiology of polyhandicap. Nevertheless, the proportion of patients presenting visual impairment was lower than in other studies performed on children with intellectual disability and motor impairment, and children with cerebral palsy [22-24], which can be explained by an under/misdiagnosis of sensorial deficiency due to the very little communication ability of the patients. Systematic detection and diagnosis of these deficiencies may improve the wellbeing of the patients. Similar to cerebral palsy patients, behavioral disorders (40-72%) were very frequent in polyhandicapped children and largely increased during adolescence [25]. The presence of

noisy behavioral disorders may have an important consequence for the patient, prompting the family to select a non-familial care structure when the patient is growing and aging.

People with severe or profound intellectual and motor disabilities experience multi-comorbidities that complicate health needs [6, 24]. As one of the most frequent comorbidity of polyhandicap, epilepsy (particularly uncontrolled epilepsy) contributes to an increase in brain damage. In our study, epilepsy (55-60%) and drug-resistant epilepsy (18-31%) were a little less frequently reported than in previous studies (70-80% for epilepsy, 66% for drug-resistant epilepsy) [17, 24, 26], and nearly one quarter of patients already presented previous status epilepticus. The availability of recent anti-epileptic drugs, allowing for a better control of epilepsy, may be an explanation. Although the proportion of epilepsies remained stable through the age classes, the proportion of drug-resistant epilepsy was lower for the oldest class, and thus this finding is most likely linked with progressive neurometabolic etiologies before adulthood and frequent stabilization of epilepsy after adolescence. Not surprisingly, as a consequence of the neurological damage and tone disorders, orthopedic comorbidities were very frequent and increased during growth, in particular during the pubertal period. Detection and taking care of these comorbidities, source of chronic pain and mobility deterioration, may limit their health consequences. Dislocations of the hips and scoliosis associated with thoracic deformity may benefit from curative and preventive measures[27, 28], such as spasticity treatment, arthrodesis, and reduction of scoliosis before adolescence.

The proportion of recurrent pulmonary infections was around 10-15% over age as a probable consequence of aspiration syndrome (32%) [8]. The deterioration of the respiratory function probably occurs later due to aggravation of scoliosis and subsequent repeated episodes of pulmonary infections caused by chronic bronchial congestion, associated with aspiration syndrome is the leading cause of death in patients with polyhandicap[6, 29, 30].

Other comorbidities, such as fecal impaction, recurrent urinary tract infections, renal failure, cutaneous defects, thyroid disorders, chronic anemia increased with age inducing discomfort and chronic pain, potentially lead to premature death. Strategies of detection, prevention, and treatment

previously showed interesting efficacy to improve well-being of the patients and should more systematically be implemented.

One key point should draw specific attention. While a large proportion of the patients received analgic medication, chronic pain was still sub-controlled. It is well-known that polyhandicapped patients are a high-risk pain-related medical condition (spasticity, joint dislocation, and pressure sores) [24, 31]. We have to promote systematic assessment and management of pain using specific methods developed for these patients with communication difficulties. It remains difficult to totally control pain due to the need to be careful and cautious with the use of level III opioids.

The low proportion of chronic diseases may partially be explained by the young age of the individuals and the absence of exposure to some cardiovascular risk factors (tobacco and alcohol, overeating).

Patja et al. have previously reported a similar finding, demonstrating a reduced mortality from chronic pathology, specifically from neoplasm, in patients with intellectual disability [32]. As previously reported in cerebral palsy children, sleep disorders were more frequent for youngest patients, particularly in difficulties initiating and maintaining sleep [33, 34].

Concerning the neurodevelopmental aspects, patients aged from 3 to 5 years hardly reached the level of development from 5 to 7 months of normal development, both in motor (posture, coordination) and cognitive domains (language, socialization), and the developmental process no longer evolved during adolescence and adulthood. Tonus disorders result in a delay or an impossibility of acquisition of the movements, and consequent development of praxis and gnosis; heavy motor impairment leads to poor sensorimotor experimentations that reduce the development of spatial somatognosic and temporal references and the construction of these representations. The general benefit of poor and inappropriate educational management aggravated motor and cognitive deficits while individualized and intensive educative care should be promising of interesting progression [35, 36].

Expectedly, administered treatments and medical devices were more frequent in the highest age classes according to the various comorbidities that the patients developed. The most common

medical device was gastrostomy (30%), a well-known device useful for preventing recurrent pulmonary infections and gastroesophageal reflux [11, 37] and for treatment of swallowing disorders.

Strengths and limitations

Our study provides health characteristics across ages from a large and homogeneous population PLH children and young adults as previous studies focusing on this population of patients are smaller with inhomogeneous sample. The results of this cross-sectional study have to be considered with caution and should be confirmed by future researches using longitudinale design (Appendix 1 and 2).

Conclusion

In the polyhandicap, heavy cerebral damage occurring on an immature brain explains the severe health condition of the patients, resulting in the entanglement of multiple impairments and comorbidities. Early detection and management of impairments and comorbidities may improve the disease course of the patients.

The authors declare that they have no vested interests.

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References

1. Rousseau MC, Billette de Villemeur T, Khaldi-Cherif S et al. Adequacy of care management of patients with polyhandicap in the French health system: A study of 782 patients. *PloS one* 2018; **13**: e0199986.
2. Rousseau MC, Mathieu S, Brisse C, Billette de Villemeur T. Le parcours de santé de la personne en situation de polyhandicap. *Annals of Physical and Rehabilitation Medicine* 2013; **56**: 271-2
3. Rousseau MC, Baumstarck K, Leroy T et al. Impact of caring for patients with severe and complex disabilities on health care workers' quality of life: determinants and specificities. *Developmental medicine and child neurology* 2017; **59**: 732-7.
4. Rumeau-Rouquette C, du Mazaubrun C, Cans C, Grandjean H. [Definition and prevalence of school-age multi-handicaps]. *Archives de pediatrie : organe officiel de la Societe francaise de pediatrie* 1998; **5**: 739-44.
5. Billette de Villemeur T, Brisse C, Afenjar A et al. Comment la loi Leonetti s'applique t'elle à l'enfant polyhandicapé? *Med Therap Pediatrie* 2012; **15**: 34-7.
6. Rousseau MC, Mathieu S, Brisse C et al. Aetiologies, comorbidities and causes of death in a population of 133 patients with polyhandicaps cared for at specialist rehabilitation centres. *Brain injury* 2015; **29**: 837-42.
7. Vlaskamp C, Cuppen-Fontaine H. Reliability of assessing the sensory perception of children with profound intellectual and multiple disabilities: a case study. *Child: care, health and development* 2007; **33**: 547-51.
8. Yoshikawa H, Yamazaki S, Abe T. Acute respiratory distress syndrome in children with severe motor and intellectual disabilities. *Brain & development* 2005; **27**: 395-9.
9. Idaira Y, Nomura Y, Tamaki Y et al. Factors affecting the oral condition of patients with severe motor and intellectual disabilities. *Oral diseases* 2008; **14**: 435-9.
10. Calis EA, Veugelers R, Sheppard JJ et al. Dysphagia in children with severe generalized cerebral palsy and intellectual disability. *Developmental medicine and child neurology* 2008; **50**: 625-30.
11. Lee L, MacPherson M. Long-term percutaneous endoscopic gastrostomy feeding in young adults with multiple disabilities. *Internal medicine journal* 2010; **40**: 411-8.
12. Poppes P, van der Putten AJ, Vlaskamp C. Frequency and severity of challenging behaviour in people with profound intellectual and multiple disabilities. *Research in developmental disabilities* 2010; **31**: 1269-75.
13. Ohtsuka E, Hayashi M, Hamano K et al. Pathological study of bronchospasms/tracheomalasia in patients with severe motor and intellectual disabilities. *Brain & development* 2005; **27**: 70-2.
14. Veugelers R, Benninga MA, Calis EA et al. Prevalence and clinical presentation of constipation in children with severe generalized cerebral palsy. *Developmental medicine and child neurology* 2010; **52**: e216-21.
15. Yoshikawa H. The difficulties of diagnosing VPA-induced pancreatitis in children with severe motor and intellectual disabilities. *European journal of paediatric neurology : EJPN : official journal of the European Paediatric Neurology Society* 2004; **8**: 109-10.
16. Palisano R, Rosenbaum P, Walter S et al. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Developmental medicine and child neurology* 1997; **39**: 214-23.
17. Kurihara M, Kumagai K, Noda Y et al. Prognosis in severe motor and intellectual disabilities syndrome complicated by epilepsy. *Brain & development* 1998; **20**: 519-23.
18. Motawaj M, Brisse C, Ponsot G, Billette de Villemeur T. Le Décès Des Patients Polyhandicapés: L'expérience Du Service de Pédiatrie Spécialisée Pour Polyhandicapés de La

- Roche-Guyon. *Journées Parisiennes Pédiatrie*. Boulogne Billancourt: Nex & Com Medical Events 2010:267–71.
19. Srivastava S, Cohen JS, Vernon H et al. Clinical whole exome sequencing in child neurology practice. *Annals of neurology* 2014; **76**: 473-83.
 20. Germany L, Ehlinger V, Klapouszczak D et al. Trends in prevalence and characteristics of post-neonatal cerebral palsy cases: a European registry-based study. *Research in developmental disabilities* 2013; **34**: 1669-77.
 21. Nordli DR, Jr. Epileptic encephalopathies in infants and children. *Journal of clinical neurophysiology : official publication of the American Electroencephalographic Society* 2012; **29**: 420-4.
 22. Sasmal NK, Maiti P, Mandal R et al. Ocular manifestations in children with cerebral palsy. *Journal of the Indian Medical Association* 2011; **109**: 318, 23.
 23. van den Broek EGC, van Eijden A, Overbeek MM et al. A Systematic Review of the Literature on Parenting of Young Children with Visual Impairments and the Adaptions for Video-Feedback Intervention to Promote Positive Parenting (VIPP). *Journal of developmental and physical disabilities* 2017; **29**: 503-45.
 24. van Timmeren EA, van der Schans CP, van der Putten AA et al. Physical health issues in adults with severe or profound intellectual and motor disabilities: a systematic review of cross-sectional studies. *Journal of intellectual disability research : JIDR* 2017; **61**: 30-49.
 25. Guyard A, Michelsen SI, Arnaud C, Fauconnier J. Family adaptation to cerebral palsy in adolescents: A European multicenter study. *Research in developmental disabilities* 2017; **61**: 138-50.
 26. Matsumoto A, Miyazaki S, Hayakawa C et al. Epilepsy in severe motor and intellectual disabilities syndrome (SMIDS)--a clinical and electroencephalographic study of epileptic syndromes. *Epilepsy research* 2007; **77**: 120-7.
 27. Saito N, Ebara S, Ohotsuka K et al. Natural history of scoliosis in spastic cerebral palsy. *Lancet* 1998; **351**: 1687-92.
 28. Majd ME, Muldowny DS, Holt RT. Natural history of scoliosis in the institutionalized adult cerebral palsy population. *Spine* 1997; **22**: 1461-6.
 29. Hanaoka T, Mita K, Hiramoto A et al. Survival prognosis of Japanese with severe motor and intellectual disabilities living in public and private institutions between 1961 and 2003. *Journal of epidemiology* 2010; **20**: 77-81.
 30. Hogg J, Juhlberg K, Lambe L. Policy, service pathways and mortality: a 10-year longitudinal study of people with profound intellectual and multiple disabilities. *Journal of intellectual disability research : JIDR* 2007; **51**: 366-76.
 31. van der Putten A, Vlaskamp C. Pain assessment in people with profound intellectual and multiple disabilities; a pilot study into the use of the Pain Behaviour Checklist in everyday practice. *Research in developmental disabilities* 2011; **32**: 1677-84.
 32. Patja K, Iivanainen M, Vesala H et al. Life expectancy of people with intellectual disability: a 35-year follow-up study. *Journal of intellectual disability research : JIDR* 2000; **44 (Pt 5)**: 591-9.
 33. Newman CJ. Sleep: the other life of children with cerebral palsy. *Developmental medicine and child neurology* 2014; **56**: 610-1.
 34. Atmawidjaja RW, Wong SW, Yang WW, Ong LC. Sleep disturbances in Malaysian children with cerebral palsy. *Developmental medicine and child neurology* 2014; **56**: 681-5.
 35. Kozulin A, Lebeer J, Madella-Noja A et al. Cognitive modifiability of children with developmental disabilities: a multicentre study using Feuerstein's Instrumental Enrichment--Basic program. *Research in developmental disabilities* 2010; **31**: 551-9.
 36. Nilsson LM, Nyberg PJ. Driving to learn: a new concept for training children with profound cognitive disabilities in a powered wheelchair. *The American journal of occupational therapy : official publication of the American Occupational Therapy Association* 2003; **57**: 229-33.

37. Strauss D, Shavelle R, Reynolds R et al. Survival in cerebral palsy in the last 20 years: signs of improvement? *Developmental medicine and child neurology* 2007; **49**: 86-92.

Table 1. Sociodemographics and health status of polyhandicapped patients (N=545)

		3-5 years	6-11 years	12-17 years	18-25 years	p
		N=80	N=166	N=155	N=144	
1) Sociodemographics						
Gender	Boys/Men	41 (52.2)	80 (48.2)	86 (55.5)	83 (57.6)	0.134
	Girls/Women	39 (48.8)	86 (51.8)	69 (44.5)	61 (42.4)	
Care modality structure	Spec. Reeduc.center	18 (23)	59 (36)	56 (37)	65 (45)	<0.001
	Residential facility	14 (17)	85 (52)	81 (54)	75 (52)	
	Home care	47 (60)	20 (12)	12 (8)	4 (3)	
2) Etiologies						
	Unknown	7 (9)	23 (14)	13 (9)	23 (14)	0.653
	Known	72 (91)	138 (86)	138 (91)	122 (86)	
3) Health status						
Severity	Less severe	40 (52)	86 (51.8)	77 (50.7)	74 (51.4)	0.896
	Severe	37 (48)	80 (48.2)	75 (49.3)	70 (48.6)	
Stability	Stable	49 (63.6)	104 (62.7)	96 (63.6)	106 (75.2)	0.041
	Unstable	28 (36.4)	62 (37.3)	55 (36.4)	35 (24.8)	

Table 2. Main etiologies of polyhandicapped patients

Unknown etiologies	N=63	
Central nervous system malformations	N=183	<ul style="list-style-type: none"> - Callosum agenesis N=3 - Lissencephaly N=3 - Holoprosencephalia N=2 - Microcephaly N=1 - Cerebellar atrophy N=1 - Grey nuclei malformation N=1 - Pallister Kilian syndrome N=1 - Non-identified N=171
Other causes	N=70	- Non identified
Neurometabolic encephalopathies	N=25	<ul style="list-style-type: none"> - Lysosomal diseases N=6 (Metachromatic leucodystrophia N=2, Gangliosidosis N=1, Tay Sacks N=1, Glucocerebrosidasis (Type III Gaucher disease) N=1, Sandhoff disease N=1) - Neuronal ceroid lipofushinosis N=8 - Mucopolysaccharidosis II and III N=3 - Peroxysomal diseases N=1 - Adrenoleucodystrophy N=1 - Other metabolic syndroms N=3 (Aicardi Gouttières N=1, Sjogren Larsson syndrom N=1, CDG syndrom N=1) - Mitochondriopathy N=4
Neurogenetic diseases encephalopathy	N=19	<ul style="list-style-type: none"> - Cytogenetics N=5 (Trisomic 21 N=1, Monosomy chromosom18 N=1, Wolf-Hirschhorn syndrom N=1, Others N=2) - Rett syndrome N=5 - Angelman syndrome N=3

		<ul style="list-style-type: none"> - Demyelinising affection of central nervous system N=1 - Progressive degenerative encephalopathy N=1 - Sensitive neuropathy type 2 N=1 - Dygue-Melchior-Clausen syndrome N=1 - Moebus syndrome N=1 - Marinesco Sjogren syndrome N=1
Epileptic encephalopathy	N=10	<ul style="list-style-type: none"> - West syndrome N=7 - Familial convulsive encephalopathy N=2 - Dravet syndrome N=1
Pre/perinatal	N=129	<ul style="list-style-type: none"> - Neonatal anoxia N=51 - Prematurity N=34 - Infectious N=28 - Neonatal hypoglycemia N=2 - Toxic N=4
Postnatal	N=34	<ul style="list-style-type: none"> - Post traumatic causes N=20 - Anoxia N=6 - Tumors N=2 - Deshydratation N=1 - Hypothermia N=1 - Fulminant purpura N=1 - Subacute Sclerosing Panencephalitis N=1 - Immune encephalitis N=1 - Reye's syndrome N=1
Missing data	N=12	

Table 3. Associated impairments of polyhandicapped patients (N=545)

		3-5 years	6-11 years	12-17 years	18-25 years	p
		N=80	N=166	N=155	N=144	
Severe motor impairments	Quadriplegia	47 (79.7)	114 (85.7)	123 (84.8)	113 (85)	0.716
	Diplegia	10 (17)	17 (12.8)	17 (11.7)	20 (15)	
	Hemiplegia**	2 (3.4)	2 (1.5)	5 (3.4)	0 (0)	
Other motor impairments	Movement disorders	12 (16.4)	25 (16)	21 (15)	16 (11.6)	0.266
	Severe dystonia	13 (18.3)	19 (12.4)	14 (10)	14 (10.2)	0.104
	Global hypotonia	37 (49.3)	59 (37.6)	47 (33)	34 (24.8)	<0.001
	Extrapyramidal syndrom	13 (18.3)	33 (21.6)	40 (27.8)	37 (26.2)	0.123
	Ataxia	4 (5.5)	7 (4.2)	4 (3)	5 (3.8)	0.469
Sensorial disorders	Visual impairment	19 (24.4)	54 (33.1)	54 (35.8)	38 (27.1)	0.889
	Hearing impairment	5 (6.7)	12 (7.6)	11 (7.5)	5 (3.5)	0.259
Behaviorial disorders*		32 (40)	99 (61)	106(70.7)	102 (72.3)	<0.001
Sleep disorders	Short sleep	5 (6.8)	15 (9.4)	8 (5.6)	10 (7.6)	0.748
	Night wake up	19 (25)	41 (25.6)	20 (13.4)	19 (14.2)	<0.001

Difficulties falling asleep	14 (18.4)	36 (22.5)	15 (10.1)	18 (13.4)	0.039
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* behavioral disorders including intermittent screaming, and/or agitation and/or stereotypies and/or intermittent crying and/or self-aggressivity and/or hetero-aggressivity

** predominant hemibody motor impairment

Table 4. Comorbidities of polyhandicapped patients (N=545)

		3-5 years	6-11 years	12-17 years	18-25 years	
		N=80	N=166	N=155	N=144	
Epilepsia	Presence of epilepsia	47 (59.5)	88 (54.7)	90 (60)	86 (60.6)	0.521
	Previous status epilepticus	17 (25.4)	36 (27)	45 (33.3)	26 (24.8)	0.847
	Drug-resistant epilepsia	24 (31.2)	52 (31.3)	39 (25.5)	25 (17.5)	0.005
Orthopedic	Scoliosis	12 (16.2)	55 (34.6)	96 (65)	108 (77.1)	<0.001
	Limb deformation	29 (38.2)	90 (56.3)	106 (71.1)	114 (81.4)	<0.001
	Limb fracture	0 (0)	8 (5)	14 (9.6)	12 (8.7)	0.009
	Hip luxation	7 (9.2)	28 (17.6)	50 (34.2)	41 (31.1)	<0.001
	Arthrodesis	0 (0)	4 (2.5)	33 (23)	52 (38.2)	<0.001
	Other surgery	1 (1.3)	30 (18.5)	52 (34.7)	78 (55.7)	<0.001
Pulmonary	Recurrent pulm. inf.	8 (10.4)	26 (15.8)	25 (16.6)	15 (10.6)	0.774
	Aspiration syndrome	24 (31.6)	40 (24.7)	39 (26.4)	32 (22.9)	0.268
Digestive	Drooling	19 (24.1)	56 (34.1)	57 (37.5)	49 (35)	0.138
	Fecal impaction	22 (28.6)	53 (33.1)	62 (41.1)	64 (47.1)	0.002

	Gastroesophageal reflux	33 (43.4)	68 (41)	69 (45.7)	58 (42.6)	0.871
Urinary	Reccurent urin. tract inf.	1 (1.3)	8 (4.8)	11 (7.5)	17 (12)	0.001
	Urinal retention	2 (2.6)	8 (4.8)	7 (4.7)	6 (4.2)	0.748
Cutaneous	Bedsore	1 (1.3)	4 (2.4)	12 (7.9)	11 (7.7)	0.007
	Pressure fragility	9 (11.8)	35 (21.3)	42 (27.6)	53 (37.3)	<0.001
Chronic pain		3 (3.9)	19 (11.4)	12 (8)	18 (12.5)	0.154
Chronic diseases*		1 (1.3)	3 (1.9)	3 (2)	3 (2.1)	0.703
Resistant bacteria		0 (0)	6 (3.6)	7 (4.5)	6 (4.1)	0.199

* chronic diseases: at least one of the following diseases: vascular stroke, myocardial infarction, diabetes, and/or cancer

Figure captions

Figure 1. Neurodevelopmental status of polyhandicapped patients according to age classes

