

Children with medical complexity and paediatric palliative care: data by a respiratory intermediate care unit.

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Abstract

Pediatric palliative care (PPC) is an active and total approach to the care of children with life-limiting conditions and to their families. PPC programs provide ongoing care for children with medical complexity (CMC), many of whom will reach adulthood. The aim of the study is to describe a population of CMC afferent in 4 months to the Intermediate Care Unit of a tertiary referral hospital for southern and central Italy. We enrolled all CMC patients admitted at our unit in 4 randomized months. We registered pathologies and different categories of childhood diseases, devices and needs, hospitalization and home care plan. Among 195 admitted to our unit, 87 CMC were included. Median age was 9 (0.1 – 35.7) years. The main pathologies recorded were neuromuscular, neurological, respiratory, metabolic, malformative, genetic syndromes and outcomes of prematurity. Comorbidity made by respiratory, digestive, neurological, cardiac and urological involvement was present in a high percentage of cases. Among our patients, only 24 hadn't any devices. The average length of hospitalization was 7.0 (1.0 – 270.0) days with 2 (1.0 – 7.0) admissions per year for patient. Home care activation was not required for only 24 of 87 patients. Children eligible for CPP are increasing and their survival results in a rise in comorbidities and special needs demanding multi-level interventions. Respiratory symptoms are the most recurrent observed demonstrating the need for an expert in CPP to have respiratory skills. Sharing data and knowledge of CMC needs may help to improve care coordination and interventions.

Keywords: Children with medical complexity, Pediatric palliative care, chronic diseases, comorbidity, special needs

Introduction

Children with medical complexity (CMC) are a growing patient population seen increasingly in general pediatric practice [1]. CMC are characterized by presence of one or more chronic clinical condition, either diagnosed or unknown, that are severe and/or associated with medical fragility. Medically fragile children are part of population of children with special health care needs (CSHCN) requiring a high level of ongoing care and support, depending on technology and often on multiple specialists for survival. Furthermore CMC require family-identified health care service needs, such as medical, specialized therapy and education. Functional limitations in CMC are typically severe in fact and may require assistance from technology and use of health resources that may include frequent or prolonged hospitalization, multiple surgeries or the ongoing involvement of multiple subspecialty services and providers [2].

Pediatric palliative care (PPC) is an active and total approach to the care of children and young people with life-limiting conditions, embracing physical, emotional, social and spiritual elements through to death and beyond; it is an active process, not simply the cessation of treatment, and that it is total, that is the multidimensional approach [3] which also encompasses the families of these children. Currently, PPC programs range from consult teams that respond to end-of-life issues to freestanding respite/hospice centers that provide ongoing care for children with chronic complex conditions, many of whom will live into adulthood [4].

In Italy, despite the introduction of legislation (Law 38/2010) stipulating the right of children and families to access appropriate services for pain control and paediatric palliative care, the availability of these services is still limited [5].

At this historic moment, we believe it should be very useful to enrich literature's data by sharing each centre case series. Furthermore few data are available about the management of the constellation of symptoms that are so prevalent in children referred to PPC [6].

Our aim is to describe a population of CMC in clinical situations of incurability and / or chronicity afferent in 4 months to our Unit, that is a referral center for southern and central Italy.

Methods

We conducted a retrospective study enrolling all CMC patients admitted at the Intermediate Care Unit of a tertiary referral hospital, Bambino Gesù Children's Hospital, Research Institute (Rome, Italy) in 4 randomized months (March, April, September, October 2018).

We identified patients and classified them by using the four different categories of childhood diseases reported by the Association for Children with Life-threatening or Terminal Conditions and the Royal College of Paediatrics and Child Health [7] (table 2):

- *Category 1*: life-threatening conditions for which curative treatment may be feasible but can fail, where palliative care is provided together with attempts at curative treatment;
- *Category 2*: diseases which are life-threatening at an early age, where appropriate treatment may prolong life and provide an adequate quality of life;
- *Category 3*: progressive conditions without curative treatment options, where treatment is exclusively palliative;
- *Category 4*: non-progressive, irreversible conditions, with complex healthcare needs, that give rise to many complications and premature death;

Furthermore we recognized patients with rare disease as well it is defined as a condition affecting fewer than 1 in 2,000 people in Canada and Europe or fewer than 200,000 people in the United States [8].

Our data described for each patient additionally pathology, complexity, chronicity, comorbidity, respiratory disease, support in ventilation if needed (oxygen therapy (O2tp), invasive mechanical ventilation (IMV), non-invasive ventilation (NIV), nutrition and dependence from devices

(nasogastric tube (NGT), percutaneous endoscopic gastrostomy (PEG), percutaneous endoscopic jejunostomy (PEJ), parenteral nutrition (NPT), neurological impairment and disease (epilepsy, neurocognitive delay, spastic quadriplegia), urologic impairment (cystostomy, intermittent catheterization), motor impairment, psychological symptoms and aids. Moreover, we reported number of admissions per year, length of stay and kind of domiciliary health care required.

We also identified different groups according to their comorbidities:

- *group 1*: respiratory and digestive diseases;
- *group 2*: neurological and respiratory diseases;
- *group 3*: respiratory, digestive and neurological diseases;
- *group 4*: respiratory and cardiac diseases;
- *group 5*: cardiac, neurological and respiratory diseases;
- *group 6*: cardiac, respiratory, digestive and neurological diseases;
- *group 7*: cardiac, neurological, urological and respiratory diseases;
- *group 8*: cardiac, neurological, urological, digestive and respiratory diseases

Statistical analysis

Statistical analysis was performed with MedCalc Statistical Software version 18.2.1 (MedCalc Software bvba, Ostend, Belgium; <http://www.medcalc.org>; 2018). A descriptive analysis was performed for demographic and clinical characteristics. Continuous variables were non-normally distributed, and were summarized as median (min - max), unless otherwise specified. Continuous data were compared by using Mann-Whitney tests. Chi-squared test was used to evaluate the increased presence of feeding, motor, neurologic, and urologic “problems” in children with at least 1 device. For all analysed parameters, $p < 0.05$ was considered statistically significant.

Results

Population

Among the examined period 195 pts were admitted to our Unit. According with the inclusion criteria for pediatric palliative care, the data of 87 CMC, 47 males (54%), were analyzed. 67 of them were planned admissions, 20 patients required an emergency hospitalization. Median age was 9 (0.1 – 35.7) years. We identified 2 infants, 3 children aged within 12 months of life, 55 children with age included within 1 year and 14 years, 14 patients aged within 18 years old, 13 patients over the age of 18, with the oldest patient aged 35 years old. Mean age of onset of symptoms was 9.53 months. Demographic characteristics of studied population are summarized in table 1.

Pathologies and different categories of childhood diseases

All children were affected by chronic pathologies, 72 of them with rare disease. We recognized 27 patients with neuromuscular diseases, 24 patients with central nervous system diseases such as cerebral palsy, leukodystrophies, drug resistant epilepsies, 15 patients affected by respiratory diseases (pulmonary hypertension, diaphragmatic hernia, bronchopulmonary dysplasia, broncho-tracheal malformation, hypoventilation), 6 patients with metabolic disorders, in 7 patients multiple malformation affecting face, heart, bone, finally 15 patients had genetic syndromes (table 2).

Furthermore we classified them in the four different categories of childhood diseases [7] (table 2).

Comorbidity

Comorbidity was present in a high percentage of cases as well as 55 cases had respiratory and digestive diseases (group 1), in others 55 patients there was both neurological and respiratory involvement (group 2). In 45 cases respiratory, digestive and neurological diseases were developed (group 3), 32 patients had both respiratory and cardiac disease (group 4), 21 patients showed cardiac, neurological and respiratory involvement (group 5), 16 others children had cardiac, respiratory, digestive and neurological impairment (group 6), 6 cases presented cardiac,

neurological, urological and respiratory impairment (group 7), finally in others 6 patients was registered cardiac, neurological, urological, digestive and respiratory comorbidity (group 8). Details are reported on Figure 1.

Hospitalization

The average length of hospitalization was 7.0 (1.0 – 270.0) days. Each patients had 2 (1.0 – 7.0) admissions per year in the last previous 12 months.

Devices and needs

Among our patients, only 24 hadn't any devices, 55 patients received PEG, but 1 had PEG/PEJ, and 31 had tracheostomy. We observed in 28 patients both tracheostomy and PEG, 9 patients were carriers of tracheostomy, PEG and bladder catheter. In 12 cases was necessary bladder catheterization. Central venous access were required in 5 patients. The studied population showed need for motor aids in 67 subjects. Furthermore, 69 patients had motor and chest physiotherapy (PT) program both at hospital and at home, 6 only chest PT and 1 only motor PT. As well as reported, 52 patients were treated with mechanical ventilation and supplemental O₂tp was prescribed in 79 cases as needed at home, only 2 patients among the analyzed period were oxygen dependent. In our center we prescribed pulse oximeters for home monitoring to 80 patients. Enteral feeding pump was normally used in 59 patients. In 1 of 24 patients without devices was reported difficult feeding and 56 of 63 patients with at least one device showed digestive diseases ($p < 0.0001$). Motor problems were registered in 18 of 24 patients without devices and in 58 of 63 patients with at least one device ($p = 0.033$). Neurologic damage was reported in 18 of 24 patients devices free, instead it was found in 50 of 63 children having one device or more ($p < 0.0001$). Urologic impairment didn't affected any patient without devices, while it affected 18 of 63 pts needing one device or more ($p = 0.0035$). Psychological disorders, as well it couldn't be assessed in 17 pts, affected 3 of 24 patients without devices, but 6 of 46 pts with at least one device showed psychological symptoms ($p = 0.9490$). Hospital admissions per year was 1.5 (1.0-4.0) in patients

without devices and 2.0 (1.0-7.0) in children with at least one device ($p = 0.0044$), with 5.5 (2.0-20.0) days spent in hospital in the first group and 10.0 (1.0-270.0) in the second group ($p = 0.0002$). Among patients with a known home care assistance program, 3 of 13 hadn't any devices, but 29 of 43 required home care activation ($p = 0.005$). Differences between patients group without device and with at least one device are reported in details in table 4.

Clinical aspects

- Motor impairment: 12 patients were paraplegic, 29 patients tetraplegic, 13 showed spastic quadriplegia. In 14 patients were registered neuromotor delay, but hypotonia in 12 patients, 52 patients needed wheelchair.
- Respiratory aspects: 84 patients showed respiratory symptoms. Among the 51 vented subjects, 28 had NIV <12h/die, 6 had VMI <12h/die, 17 patients were treated with VMI>12h/die. During the hospitalization O2tp was administered to 11 of them. In 31 patients was not reported neither support in ventilation nor in oxygen need.
- Nutrition and food issues: in 54 patients was positioned PEG, 1 child was reported as carriers of both PEG and PEJ. 2 children needed NGT, during the hospitalization no one required NPT neither at home.
- Cardio-circulatory system: in 32 subjects were registered comorbidity with cardiological involvement and treatment.
- Neurological disease: 42 patients were affected by epilepsy and required specific pharmacological treatment, 30 subjects showed neuromotor delay and 2 patients had severe spastic hypertonia treated with baclofen pump.
- Urologic diseases: 17 children required intermittent bladder catheterization and 1 patient had cistostomy.

- Psychological disorders: as well as in 17 patients there was no possibilities to evaluate psychological disease because of their severe conditions and their neurological impairment, we observed depression in 7 subjects and behavior disorder in 2 patients, apparently no symptoms in other cases were reported.

Home care plan

- Domiciliary health care required: home care activation was not required for 24 patients.

Local Health Authority was asked to activate a home care plan, respectively characterized by the following professional roles:

-nurse, doctor and physiotherapist for 21 patients

-nurse, doctor, physiotherapist and physiatrist for 1 patient

-nurse, doctor, physiotherapist and social worker for 1 patient

-physiotherapist for 5 patients

-physiotherapist and speech therapist for 1 patients

- social worker for 3 patients

Informations about home care are not available in 31 patients.

Discussion

Our study analyzed a pediatric population of CMC requiring palliative care. Around half of patients admitted at our Intermediate Care Unit in four months were eligible for CPP. Our results showed that CMC are increasing and reaching an age that goes beyond the expected, now very different from the early deaths previously recorded. This survival results in an increase in comorbidities and special needs of these kind of patients that demand multi-level interventions, providing a holistic approach that concerns all caregivers involved.

Chronic conditions and survival

The study described clinical aspects and needs in pediatric chronic patients managed in our Unit and showed their health condition's severity and complexity. Our data clearly exposed as the number of children and young people (CYP) living with life-limiting and life-threatening conditions is rising with continuing advances in clinical medicine. We reported most of our patients were >1 < 14 years old, but a significant part of population was older than 14 and even > 18 years demonstrating that many CMC reach adulthood. The achievement of a higher average age means to discover and to treat long term side effects of treatments and interventions not appreciated before due to advancements in critical care, oncology, surgery, bronchopneumology and nutrition that make children with serious illnesses living longer, as reported in literature [9; 10; 11].

Over the last half century is reported in fact child mortality has declined substantially [12] and, specially with improved survival rates of premature infants and those born with congenital diseases, the prevalence of CMC is increasing [13]. CMC now are a heterogeneous group representing 13%–18% of the total pediatric patient population [14]. Our case studies was made up of patients with chronic pathologies, with no oncologic disease nor terminal illness, but CYP with a clinical long-term evolution requiring inclusion in a palliative program.

Pathologies, symptoms and different special needs

The analyzed population was significantly heterogeneous, with variable types of pathologies including neuromuscular, respiratory, cardiological, neurological, chromosomal pathologies and effects of prematurity, with many patients suffering from rare diseases. The diversity of the diseases involved confirmed that the spectrum of conditions in CMC requiring pediatric palliative care is really broad requiring different needs [16]. Often in current strategies for pain and symptom assessment and management are focused on one symptom at a time, but CMCs will often have, for example as it is often observed in our CYP, pain, spasticity and airway secretions simultaneously.

Particularly we registered in 96% of our patients respiratory symptoms and this demonstrated the need for a caregiver to have respiratory skills.

However it is really important to consider the interdependence of symptoms and one drug's effect on other symptoms and other drug effects remains to be addressed because even all these aspects make together patient's hospital and out-of-hospital needs.

Functional limitations and technology dependence

Our CYP presented comorbidity and impairment evolving of multiorgan systems often requiring multiple aids and medical technology. We observed difficult in feeding, neurologic, urologic and motor disorders significantly affects the needs of at least one device. The majority of patients showed utilization of and dependence on medical technology according with the successes in survivorship, increasing care coordination needs. Children enrolled had high rates of gastrostomy tube (62%) and tracheostomy tube-dependence (36%), in addition we found bladder catheter and central venous access. We found that digestive and swallowing problems are significantly associated with the need to have at least one device, particularly PEG or PEJ. In addition, neurological impairment could present itself as an obstacle to effective ventilation and / or the ability to feed orally or manage bladder emptying and therefore we described a statistically significant require in need for further aids. In our population patients with motor disabilities needed a significantly higher number of devices compared to patients with normal motor skills. We observed urologic disorders too was significantly registered in our pts needing at least one devices.

Quantification of the need for CMCs is complicated by uncertainty defining both population and specialty.

Impact on family and Health care use

The service needs have a significant impact on the family unit, specifically time devoted to direct care, frequent provider visits, care coordination, and financial burden. The type, intensity, and

consistency of these manifestations in fact may change dynamically over the life of the child depending on a variety of medical, psychosocial, and community factors. In fact number number of admission per year and days spent in hospital during the admissions were significantly associated with to the presence of 1 or more device, affecting both family stress and health care expenses. In our population it has been observed that these patients, in particular those who depend on technologies, needed at least 2 hospitalizations per year and that the average length of stay was about 10 days, confirming clearly higher needs also in terms of economic resources for the health system ($p = 0.0002$). The presence of at least one device among tracheostomy, PEG, central venous access and bladder catheter increases the psychological disorders, could be correlated to the degree of stress that involves the patient in the first place and certainly the families. This data could be underestimated in the considered population as in some patients it was not possible to evaluate psychological stress due to severe neuromotor delay.

Timing of CPP

At present it is still unclear when the right moment to introduce CMC in CPP is. Pediatric Palliative Screening Scale (PaPaS Scale) is the only tool identified to assess eligibility for CPP in literature [15]. This scale is conceived as an instrument dedicated to the pediatric patient and it was found to be able to recognize children and adolescents who can benefit from PPC effectively and early, favoring their timely handling and their correct inclusion in the most appropriate level of assistance. As well as PaPaS Scale is a precious tool to detect CMC patients who need to be included in a path of palliative care based on trajectory of the disease, expected result of the treatment, symptoms, needs and estimate of life expectancy, however it emerges that some items are particularly aimed at cancer patients and / or at the end of their life but that they are not suitable for assessing the characteristics of patients with chronic diseases, which make up the entire population of our research. This observation reinforces the idea that even in palliative care there are not available protocols for management, classification and inclusion of CMCs.

The palliative care network

CMCs health and quality of life depend on integrating health care between a primary care medical home, tertiary care services, and other important loci of care such as transitional care facilities, rehabilitation units, the home, the school, and other community based settings. Our CYP in fact required, it is in addition to in-hospital care, home assistance involving different professional roles as well as nurse, doctor, physiotherapist, physiatrist, social worker and speech therapist. Home care assistance significant correlated with needing in one or more devices, according with complexity and comorbidities in our patients, as showed in table 4. It is important to underline that none of the patients had in the home team an expert in CPP even if they were eligible to CPP and that this figure, even if it was not complete, was covered by the pediatric pulmonologist. We can report that an extremely qualified health staff made by doctors, nurses, physiotherapists, psychologist, and every element of the assistance chain addressed to CMCs have an important burden of care with family members managing their medical fragility and complex chronic conditions to improve their quality of care and optimize health outcomes.

Limitations

One limitation in our study is that informations about home care and type of assistance involved at home are not available in 31 pts probably because it was prescribed in other referral centers, so it could be underestimated. More studies are needed to establish home care considering devices, complexity and depending in technology. Furthermore our ward is typically a semi-intensive care unit and therefore not a reference unit for oncology patients who could therefore have been undervalued. The months we randomly analysed did not include the winter period, when chronic patients could more frequently need to be hospitalized urgently because of the increased recurrence of infectious episodes, especially respiratory infections, miscalculating the request for urgent hospitalizations compared to the planned hospitalizations.

Moral and Ethical Issues

Finally ethical issue plays a key role because families need help in ethical dilemmas. Specialists are required to have knowledge and experience of managing disease, trajectories of life limiting conditions in CMC, of death and dying process. When is right to introduce and to continue to assist a patient with mechanical ventilation? When is appropriate to subject the patient to the placement of a tracheostomy? And when is not correct to offer the family and the patient to an invasive ventilation? What impacts clinical management have on the patients and families? What effects on the healthcare services and the allocation of resources? Health staff must respect the wishes made by patient and family, quality of life, dignity and equal access to treatment, while guaranteeing compliance with principles of ethics that is autonomy, beneficence, non-maleficence and justice. To establish proportionally of care medical staff have to help patients and their families to understand the consequences related to their choices, to consider all the positive and negative aspects in terms of the patient's life condition and presumable complexities. Therefore, palliative care clinicians should anticipate conducting multiple prognostic and goal-focused conversations, often over many years, regarding a family's concerns and hopes for the future [9].

Conclusions

This study is demonstrative that PPC is not only the work to reduce suffering at the end of life but it includes care and management for CMC. Depending on variety of medical conditions that affect CMC, it increase use of inpatient and outpatient resources. Knowledge of CMC needs may help to improve intensive inpatient and outpatient care coordination clinics and targeted interventions in the way to reduce future health care utilization and hospital readmission for CMC. Sharing data may be a step forward the holistic knowledge of these patients in order to identify children eligible for CPP and to plan the most appropriate assistance to guarantee their needs. Thus, can promote the proper training of personnel involved in PPC and CMC's assistance in the way

to create a home team with trained health professionals and guided by an expert in CPP but with respiratory skills too, as evidenced by our data. From descriptive studies to multicenter research trials we can improve the evidence based for common inpatient clinical issues. A shift is now occurring in the PPC literature to a more difficult and resource-intensive investigation of both PPC processes such as symptom management and psycho-social care, and outcomes, for example, the impact of specific interventions.

Future aim should be to improve care coordination and create partnerships between primary care physicians and the tertiary care center for CMC, by making easier reliance on technology, polypharmacy, and home care to maintain a basic quality of life, to reduce the risk of frequent and prolonged hospitalizations and elevated need for care coordination.

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Table 1. Demographic data

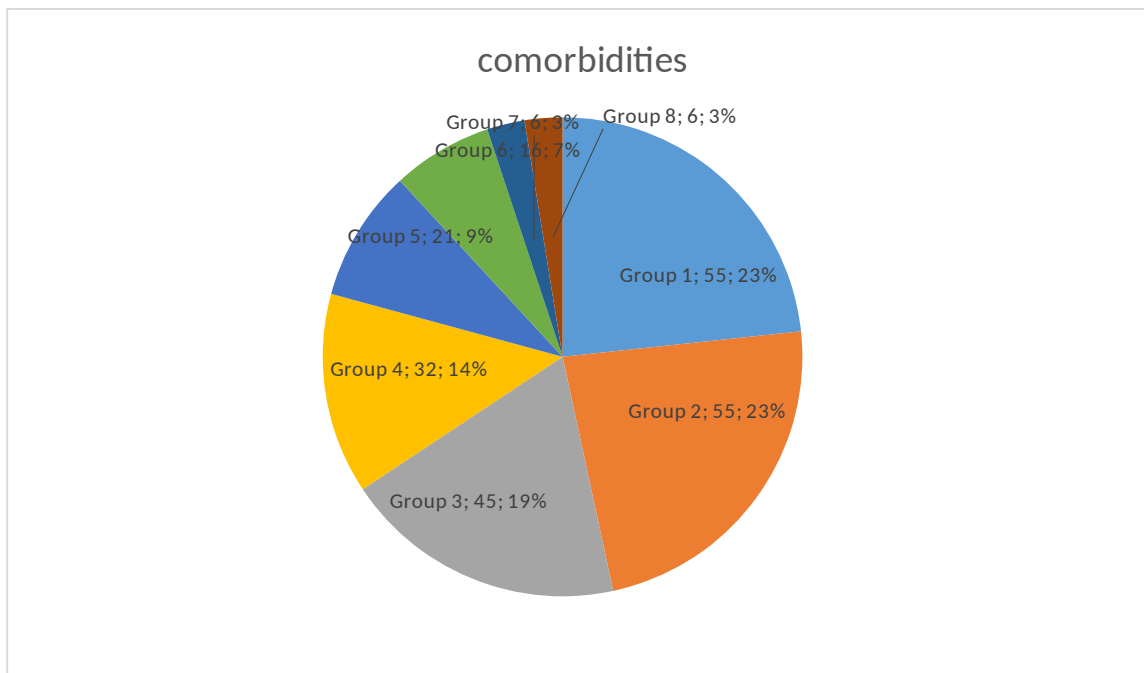
Patients	87
Male (%)	54
Mean age (months)	118,86 ± 93
Age < 6 months (%)	2.3
Age >6 months ≤ 12 months (%)	3.4
Age >1 years <14 years (%)	63.2
Age >14 years ≤ 18 years (%)	16.1
Age > 18 years (%)	14.9

Table 2. Clinical and epidemiological data

Rare disease (%)	82.8
Neuromuscular disease (%)	32
Central Nervous System Disorders (%)	31
Respiratory diseases (%)	17
Metabolic disorders (%)	7
Malformations (%)	8
Genetic syndromes (%)	17
Category 1 (%)	7
Category 2 (%)	14
Category 3 (%)	45
Category 4 (%)	34

Legend: Category 1. life-threatening conditions for which curative treatment may be feasible but can fail, where palliative care is provided together with attempts at curative treatment. Category 2. diseases which are life-threatening at an early age, where appropriate treatment may prolong life and provide an adequate quality of life. Category 3. progressive conditions without curative treatment options, where treatment is exclusively palliative. Category 4. non-progressive, irreversible conditions, with complex healthcare needs, that give rise to many complications and premature death [7].

Fig. 1. Comorbidity



Legend: group 1: respiratory and digestive diseases; group 2: neurological and respiratory diseases; group 3: respiratory, digestive and neurological diseases; group 4: respiratory and cardiological diseases; group 5: cardiac, neurological and respiratory diseases ; group 6: cardiac, respiratory, digestive and neurological diseases; group 7: cardiac, neurological, urological and respiratory diseases; group 8: cardiac, neurological, urological, digestive and respiratory diseases

Table 3: Devices and needs

No device	27%
Tracheostomy	36%
PEG	63%
PEG/PEJ	1.1%
central venous access	6%
bladder catheter	14%
PEG+Tracheostomy	32%
PEG+Tracheostomy+ bladder catheter	10%
PEG+ central venous access+bladder catheter	2.3%
PEG+ central venous	1.1%
PEG+ bladder catheter	1.1%
Motor aids	77%
Motor PT	80%
Chest PT	86%
Mechanical ventilation	60%
Oxygen therapy as need	91%
Oxymeter	92%
Enteral feeding pump	68%

Table 4 - Children's Characteristics according to the presence of at least 1 device

Characteristics	No Devices (N° of Patients = 24)	At least 1 Device (N° of Patients = 63)	P values
Age	10.9 (1.0 - 22)	8.0 (0.1 - 35.9)	P = 0,9168
Male/Female	10/14	30/33	χ^2 0.245, p = 0.6206
Age at symptoms onset	0.2 (0.0 - 10.0)	0.0 (0.0 - 10.0)	P = 0,1773
Feeding problems (yes/no)	1/23	57/6	χ^2 57.589, p < 0.0001
Motor problems (yes/no)	18/6	58/5	χ^2 4.529; p = 0.033
Neurologic problems (yes/no)	18/6	50/13	χ^2 22,138 ; p = <0,0001
Urologic problems (yes/no)	0/24	18/45	χ^2 8.547; p = 0.0035
Psychological disorders	3/24	6/46	χ^2 0.004098; p = 0.9490
Hospital admission per year (N°)	1.5 (1.0 - 4.0)	2.0 (1.0 - 7.0)	P = 0.0044
Days spent in hospital during the admissions (N°)	5.5 (2.0 - 20.0)	10.0 (1.0 - 270.0)	P = 0.0002
Domiciliary health care	3/13	29/43	χ^2 7.879; p = 0.005

Values are expressed as media (min - max), unless otherwise specified;