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Original article

Prevalence and clinical characteristics of children with medical complexity in Tottori Prefecture, Japan: A population-based longitudinal study

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Abstract

Objectives: To investigate the prevalence and background of children with medical complexity (CMC) and its secular trend in Japan.

Methods: CMC were defined as patients under the age of 20 years requiring medical care and devices. The patients were enrolled using the national health insurance claims data of three hospitals and two rehabilitation centers in Tottori Prefecture. The study period was divided into three periods: Period 1, 2007–2010; Period 2, 2011–2014; and Period 3, 2015–2018.

Results: A total of 378 CMC were enrolled. The prevalence of CMC was 1.88 per 1000 population among subjects aged <20 years in 2018, and it increased by approximately 1.9 times during the study period. The number of CMC who presented with severe motor and intellectual disabilities did not change from Period 1 to Period 3. Meanwhile, the number of CMC who had relatively preserved motor and intellectual abilities increased from 58 to 98. The proportion of CMC who required respiratory management and oxygen therapy increased by 1.3 and 1.8 times, respectively. By contrast, the proportion of CMC who need tube feeding decreased significantly between periods 1 and 3 ($P < 0.05$).

Conclusions: The prevalence of CMC increased almost twice during the 12-year study period; however, the increase in the number of patients with relatively preserved motor and intellectual abilities was pronounced. This study showed that the need for medical care and devices differed based on the underlying disorders and severity of CMC; therefore, individualized medical, welfare, and administrative services and education about the various types of CMC must be provided.

Keywords: Children with medical complexity; Severe motor and intellectual disabilities; Medical care; Medical device

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1. Introduction

In Japan, the term severe motor and intellectual disabilities (SMID) was introduced by the Ministry of Health, Labor and Welfare in the 1960s to define children with both physical and intellectual disabilities. In 1971, the Ohshima's Classification, a tool utilized to identify children with motor and intellectual disabilities, was proposed as a criterion for identifying SMID administratively and clinically [1]. Suzuki et al. have introduced the term severe motor and intellectual disabilities-medical care dependent group (SMID-MCDG) and developed a scoring system to determine the severity of disabilities according to medical care, treatment, and use of devices (Table S1) [2]. However, with recent advances in neonatal and pediatric intensive care, the proportion of children with the ability of self-mobility and communication who required daily medical care and use of devices has been increasing in Japan. Cohen at al. have proposed a definitional framework of children with medical complexity (CMC) to characterize children with chronic conditions, health care needs, functional limitations, and multiple service needs; this framework has a broader concept than SMID as it involves children with various and multiple complexities [3]. Regarding medical care provisions, the hospitalization rate of CMC has increased rapidly within the past 10–20 years in the US [4], and the medical expenses of CMC account for approximately one-third of the total pediatric medical expenses in Canada and Australia [5,6]. Furthermore, regarding education and welfare, some services and policies do not meet the actual needs of CMC, leading to low satisfaction for CMC [7–10]. CMC has become a medical, welfare, and administrative issue worldwide; however, only a few studies on the number and clinical characteristics of CMC are available in Japan. To understand the needs of CMC and provide individualized medical care, welfare service, and education that meets the needs, it is necessary to understand the number of CMC and the background of CMC, including the severity, underlying diseases, and contents of medical care and devices. In this study, we retrospectively investigated the prevalence and clinical characteristics of CMC and the trend in secular changes in Japan.

2. Methods

2.1. Setting

In this retrospective study, the medical charts of CMC in Tottori Prefecture located in the western part of Japan were reviewed. This prefecture had a population of approximately 560,000 in 2018, and has three center hospitals and two rehabilitation centers for CMC. Almost all CMC in Tottori Prefecture visit and are admitted to these facilities for health management, treatment, and rehabilitation.

2.2. Definition of CMC

The SMID-MCDG scoring system for children has been used for national health insurance claims to add medical fees in inpatients with complex medical care since 1996 (Table S1) [2]. Moreover, outpatients could incur additional home medical expenses from national health insurance claims if they required the items described in the scoring system. In this study, we defined CMC as children who have the items described in the scoring system except for the usage of nebulizer (no. 6), hyperhidrosis due to hypertonicity requiring frequent changing of clothes and position (no. 10), and frequent changing of position (no. 14) as these items could not be covered by national health insurance claims.

2.3. Patient selection

The participants were less than 20 years of age at the time of each study period between 2007 and 2018 and were considered to have underlying disorders that developed when they were under the age of 16 years. The patients were limited to those living in Tottori Prefecture. CMC were identified using the national health insurance claims data.

2.4. Items included in the investigation

We assessed the medical charts of CMC from 2007 to 2018 after they were introduced to any medical care and use of devices. We examined the characteristics of the patients, residence, time when each medical care was introduced and discontinued, underlying disorders, severity, and contents of medical care and use of devices. Patients who required long-term hospitalization were defined as those who were admitted to the hospital or stayed at centers for more than 6 months. Regarding medical care and use of devices, noninvasive positive pressure ventilation (NPPV), tracheostomy positive pressure ventilation (TPPV), biphasic cuirass ventilation, and intrapulmonary ventilation were collectively defined as ventilator treatment. The usage of ventilators, tracheostomy, and airway tubes was collectively defined as respiratory management. In addition, the utilization of nasogastric tube, gastrostomy, and intestinal fistula was collectively referred to as tube feeding. We investigated the proportion of CMC who received medical care and used devices in each study period. The underlying disorder in CMC was defined as main disorders that commonly affected medical care. The disorders were categorized into six groups: congenital disorders, perinatal disorders, acquired neurological disorders, cardiac diseases,

neoplasms, and others. The details of each disorder are shown in Table S2.

2.5. Severity of CMC

In this study, CMC was classified into four categories according to the ability of self-mobility and communication: Groups 1, 2, 3, and 4 (Fig. S1). The ability of self-mobility was evaluated based on the achievement of the best physical function and Gross Motor Function Classification System (GMFCS) score [11]. CMC who have a GMFCS score of 1–4 were considered to have the ability of self-mobility, and those with a GMFCS score of 5 had disability of self-mobility. CMC less than 1 year of age during the last follow-up were excluded from the evaluation of self-movement because they could not be accurately evaluated due to their motor developmental milestones. The ability to communicate was assessed based on verbal comprehension during the final follow-up. CMC aged <3 years during the last follow-up were excluded from the evaluation of the ability to communicate because of their verbal developmental milestones. Group 1 comprised CMC who presented with disability associated with self-mobility and communication (those considered to have SMID). Group 2 comprised CMC who presented with disability associated with self-mobility but were able to communicate. Group 3 comprised CMC who presented with the self-mobility and disability associated with communication. Group 4 comprised CMC who presented with self-mobility and communication (those with relatively preserved motor and intellectual abilities).

2.6. Analysis

Data were collected at the Division of Child Neurology, Tottori University, after anonymizing at each hospital and center. After obtaining data about CMC, duplicate cases were deleted, and the prevalence of CMC was calculated as the number of CMC divided by the size of the population under the age of 20 years in Tottori Prefecture annually. Data were available from the annual reports of population by the Statistics Division of the Tottori Prefecture government. The prevalence was described per 1000 population under the age of 20 years. The study period was divided into three periods: Period 1, 2007–2010; Period 2, 2011–2014, and Period 3, 2015–2018. The severity of new-onset CMC in Period 3 was not classified because it might not be fixed during the final follow-up. All data were analyzed using the Statistical Package for the Social Sciences software version 25 (IBM Japan, Tokyo, Japan). The categorical data during the three study periods and each group were compared with Pearson's chi-square test. In all the analyses, a p-value <0.05 was considered statistically significant. This study was approved by the ethical committee of the Faculty of Medicine, Tottori University, in June 2018 (No.18A067).

3. Results

A total of 378 CMCs (218 boys and 160 girls) were included during the 12-year study period. At the time of analysis, 44 participants died. During the 12-year period, 49 patients reached the age over 20 years, and 114 patients became free from medical care and use of devices; thus, they were excluded from the criteria of CMC during this time. The classification of the underlying disorders of CMC that were excluded from the definition of CMC during the observation period is shown in Table S3.

Of the 378 participants with CMC, 274 had new-onset CMC during the study period. The secular trend in the prevalence and number of CMC is shown in Fig. 1. During the study period, the prevalence of CMC increased approximately 1.9 times from 0.98 to 1.88 per 1000 population under the age of 20 years, and the total number of CMC cases also increased approximately 1.6 times from 112 to 174. The number of individuals with CMC living in their own houses increased from 93 in 2007 to 155 in 2018, accounting for approximately 90% of CMC cases, whereas the number of patients who required long-term hospitalization did not change (approximately 20) during the study periods (Fig. S2).

The distribution of underlying disorders did not change significantly during each study period (Table 1). Approximately 40%–50% of individuals with CMC presented with congenital disorders in each study period, followed by perinatal disorders. Moreover, congenital disorders also accounted for the largest proportion of underlying disorders in individuals with new-onset CMC. The proportion of CMC with cardiac diseases had increased over time with no significant change, accounting for 12.6% of all CMCs in Period 3. In terms of disease classification, other diseases included obstructive sleep apnea syndrome (SAS) (n = 12), tracheomalacia, laryngomalacia, airway stenosis and choanal atresia (n = 10), digestive diseases (n = 9), renal and urinary diseases (n = 6), cleft lip and palate (n = 4), congenital diaphragmatic diseases (n = 3), neonatal drug withdrawal syndrome (n = 2), anterior spinal artery syndrome (n = 1), and diseases of unknown cause (n = 2). In terms of severity, the proportion of patients in Group 1 CMC decreased from 51.7% in Period 1 to 41.6% in Period 3. By contrast, the proportion of patients in Group 4 CMC increased from 32.6% to 41.2%. Moreover, the number of patients in Group 4 CMC increased from 58 in Period 1 to 98 in Period 3. However, the number of CMCs belonging to the other group did not change. The proportion of patients who presented with new-onset CMC in Group 1 CMC

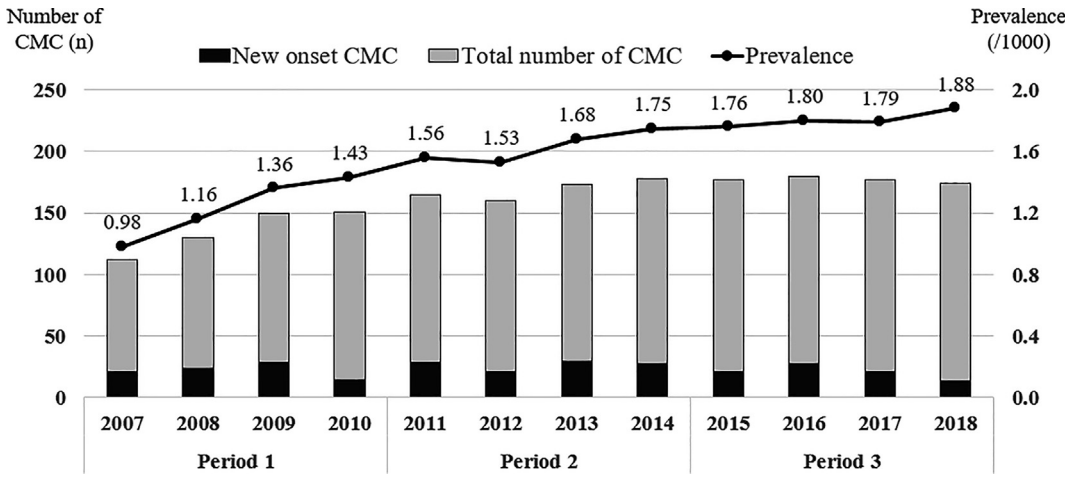

Fig. 1. Trend in the prevalence and total number of children with medical complexity (CMC) in Tottori Prefecture. Prevalence was described per 1000 population under the age of 20 years. Prevalence increased approximately 1.9 times from 2007 to 2018. The number of individuals with CMC increased gradually from Period 1 to Period 2 and flattened.

increased. By contrast, that in Group 4 CMC decreased. All these changes were not statistically significant. Regarding underlying disorders according to the severity of CMC, congenital disorders accounted for more than 50% of cases in all groups, except for Group 4 CMC (Table 2). The proportion of patients who presented with congenital and perinatal disorders was significantly lower in Group 4 CMC than in Group 1 CMC (35.2% and 16.0% vs. 51.0% and 28.6%, respectively, $p < 0.05$). By contrast, the proportion of patients in Group 4 CMC with cardiac and other diseases was significantly higher than that of patients in Group 1 CMC ($P < 0.05$).

The number of CMC who required respiratory management increased from 77 in Period 1 to 102 in Period 3, whereas those who required home oxygen therapy (HOT) increased from 55 in Period 1 to 99 in Period 3. However, the difference in both results was not significant (Table 3). By contrast, although the number of CMC requiring tube feeding did not change during the study periods, the number of patients in total CMC using tube feeding decreased significantly from 51.7% in Period 1 to 38.7% in Period 3 ($p < 0.05$). The proportion of CMC requiring total parenteral nutrition, colostomy, and dialysis were extremely low, and no obvious change was observed during the study periods. Fig. 2 shows the difference in the need for medical care and use of devices according to the severity of CMC. The proportion of CMC who required respiratory management, TPPV, tube feeding, and suction in Group 1 CMC was significantly higher than that in other groups ($p < 0.05$) (Fig. 2). The proportion of CMC who required HOT in Group 2 was significantly higher than that in other groups ($p < 0.05$). The proportion of patients in Group 4 CMC who required respiratory management, NPPV, tube feeding, and HOT was 28.4%, 21.6%, 24.1%, and 42.6%, respectively.

4. Discussion

This study showed that the prevalence of CMC in 2018 was 1.88 per 1000 population under the age of 20 years, and it increased by about 1.9 times in 2007. Based on these data, the number of CMC cases was approximately 38,000 in Japan. According to a previous survey conducted by the Japan Pediatric Society, the prevalence of CMC in 2015 was approximately 0.77, and it increased by 2.6 times within 9 years in Japan [12]. Moreover, a study conducted by the Japanese Ministry of Health, Labor and Welfare has indicated that the number of CMC increased by 1.8 times from 2005 to 2015 [13]. In the current study, the proportion of CMC was higher than that of the two previous studies. The difference in the prevalence and estimated number of CMC was partly attributed to the survey method and extraction criteria used. In the previous survey, the prevalence of CMC in Tottori prefecture was higher than that in other regions in Japan; therefore, the present estimated number of CMC in entire Japan may be overestimated [12]. However, we consider that this population-based study is valuable because it is the first to assess the total number of individuals with CMC in a relatively large area in Japan.

In other countries, Cohen et al. have proposed a definitional framework of CMC [3]. Moreover, they assessed all hospitalized patients in Ontario for 1 year and reported that the proportion of CMC accounted for 0.67% of the total pediatric population, described as an estimated prevalence of CMC [5]. Because the definition of CMC in the previous study differed from that

Table 1
Underlying disorders and severity of CMC during each study period.

		Period (Year)	Period 1 (2007–2010) n (%)	Period 2 (2011–2014) n (%)	Period 3 (2015–2018) n (%)
Total CMC		Total number (n)	178	242	238
	Underlying disorders	Congenital disorders	86 (48.3)	103 (42.6)	111 (46.6)
		Perinatal disorders	33 (18.5)	59 (24.4)	51 (21.4)
		Acquired neurologic disorders	21 (11.8)	24 (9.9)	17 (7.1)
		Cardiac diseases	15 (8.4)	24 (9.9)	30 (12.6)
		Neoplasms	5 (2.8)	8 (3.3)	6 (2.5)
		Others	18 (10.1)	24 (9.9)	23 (9.7)
	Severity	Group 1	92 (51.7)	106 (43.8)	99 (41.6)
		Group 2	6 (3.4)	8 (3.3)	8 (3.4)
		Group 3	11 (6.2)	13 (5.4)	10 (4.2)
		Group 4	58 (32.6)	104 (43.0)	98 (41.2)
		Unknown or not evaluated	11 (6.2)	11 (4.5)	23 (9.7)
New-onset CMC		Total number (n)	87	105	82
	Underlying disorders	Congenital disorders	42 (48.3)	40 (38.1)	39 (47.6)
		Perinatal disorders	12 (13.8)	27 (25.7)	14 (17.1)
		Acquired neurologic disorders	6 (6.9)	6 (6.9)	3 (3.7)
		Cardiac diseases	9 (10.3)	13 (12.4)	13 (15.9)
		Neoplasms	3 (3.4)	5 (4.8)	0 (0)
		Others	15 (17.2)	14 (13.3)	13 (15.9)
	Severity	Group 1	38 (43.7)	63 (60.0)	
		Group 2	2 (2.3)	3 (2.9)	
		Group 3	6 (6.9)	5 (4.8)	
		Group 4	33 (37.9)	27 (25.7)	
		Unknown or not evaluated	8 (9.2)	7 (6.7)	

of ours, the prevalence of CMC cannot be compared directly.

This study presented the underlying disorders of CMC and its secular trend. The proportion of CMC with underlying disorders did not change significantly. Meanwhile, the number of CMC who presented with congenital disorders, perinatal disorders, and cardiac diseases had increased during the three study periods. During these periods, the proportion of individuals with CMC who presented with congenital disorders accounted for 40%–50% of all cases. Furthermore, CMC who presented with congenital disorders were most common among CMC both who died and who became over the age of 20 years during 12-year study period. Because the survival rate of major congenital disorders, such as 21, 18, and 13 trisomy partly depends on their complications, treatment, and management [14–16], it is expected to change with time. By contrast, in developed countries, the number of children with cerebral palsy, a representative example of perinatal disorders, has decreased in 2000 compared to that in the 1970s and 1980s [17–19]. Furthermore, Brooks et al. have reported that the mortality rate in children with cerebral palsy under the age of 15 years decline by 1.6% annually in California [20]. Since the prognosis of patients with congenital disorders or cerebral palsy is commonly based on their underlying diseases, complications, and management, the distribution of each underlying disorder may change in the future. Regarding congenital heart disease (CHD), the number of patients with CHD increased rapidly worldwide [21]. In addition, the number of patients with CHD who underwent surgery has increased in Japan, whereas the mortality rate did not change at about 4–5% [22]. Therefore, the number of children who required HOT after CHD surgery is expected to increase. Thus, continuous investigation is required because the prognosis of various disorders will change with time.

The novel findings of this study showed the trend in the severity of CMC. CMC with relatively preserved motor and intellectual abilities but requiring frequent medical care and use of devices must be considered based on the definition of Cohen et al. [3]. However, a survey grasping this wide range of CMC had not been conducted in Japan. The total number of patients in Group 4 CMC increased to approximately twice between Period 1 and Period 2 and then remained the same, although that of new-onset CMC in Group 4 unchanged. It is estimated that the increase in the total number of Group 4 CMC can be partly attributed to the high proportion of patients with underlying disorders and low mortality who require long-term medical care. By contrast, the number of new-onset CMC in Group 1 increased. However, the total number of CMC in Group 1 between Period 1 and Period 3 had

Table 2
The proportion of patients with underlying disorders according to the severity of CMC.

	Severity	Group 1 n (%)	Group 2 n (%)	Group 3 n (%)	Group 4 n (%)
	Total	147	19	13	162
Underlying disorders	Congenital disorders	75 (51.0)	11 (57.9)	9 (69.2)	57 (35.2)
	Perinatal disorders	42 (28.6)	2 (10.5)	2 (15.4)	26 (16.0)
	Acquired neurologic disorders	26 (17.7)	0 (0)	0 (0)	4 (2.5)
	Cardiac diseases	1 (0.7)	4 (21.1)	1 (7.7)	30 (18.5)
	Neoplasms	2 (1.4)	2 (10.5)	1 (7.7)	4 (2.5)
	Others	1 (0.7)	0 (0)	0 (0)	41 (25.3)

Table 3
Usage of medical care and devices among individuals with CMC in each period.

	Period (Year)	Period 1 (2007–2010) n (%)	Period 2 (2011–2014) n (%)	Period 3 (2015–2018) n (%)	Total (2007–2018) n (%)
	Total (n)	178	242	238	378
Medical care and devices	Respiratory management	77 (43.3)	99 (40.9)	102 (42.9)	108 (28.6)
	TPPV	27 (15.2)	39 (16.1)	39 (16.4)	55 (14.6)
	NPPV	35 (19.7)	44 (18.2)	50 (21.0)	88 (23.3)
	Tube feeding	92 (51.7)	105 (43.4)	92 (38.7)	201 (53.2)
	Suction	90 (50.6)	106 (43.8)	105 (44.1)	149 (39.4)
	Oxygen therapy	55 (30.9)	94 (38.8)	99 (41.6)	167 (44.2)
	TPN	4 (2.2)	1 (0.4)	2 (0.8)	7 (1.9)
	Urethral catheterization	21 (11.8)	35 (14.5)	28 (11.8)	46 (12.2)
	Colostomy	0 (0)	2 (0.8)	3 (1.3)	3 (0.8)
	Dialysis	3 (1.7)	3 (1.2)	4 (1.7)	5 (1.3)

TPPV: Tracheostomy positive pressure ventilation, NPPV: Noninvasive positive pressure ventilation, TPN: Total parenteral nutrition.

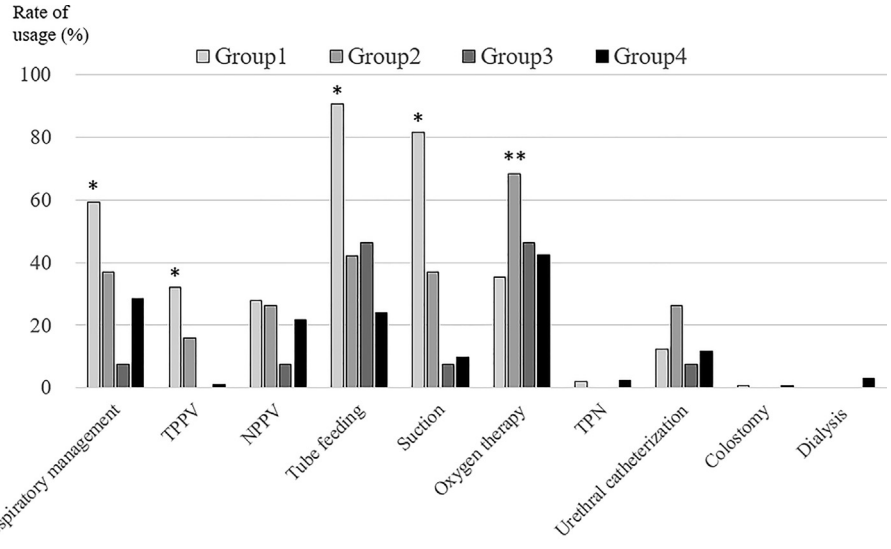

Fig. 2. Usage rate of medical care and devices according to the severity of children with medical complexity (CMC). The rate of CMC requiring respiratory management, tracheostomy positive pressure ventilation (TPPV), tube feeding, and suction in Group 1 CMC was significantly high among all the groups, whereas that requiring home oxygen therapy in Group 2 CMC was significantly high among all groups. *$p < 0.001$, **$p < 0.05$.

not changed significantly. According to previous reports, the number of patients with SMID has been increasing in Japan [23,24]. The lack of significant change in the total number of patients in Group 1 CMC might be partly attributed to the high mortality rate of severe CMC [25,26]. In summary, we propose increasing the number of CMC by approximately 1.9 times over 12 years as an improvement in the quality of medical care for children with relatively preserved motor and intellectual disabilities as well as an improve-

ment in the survival rate of severely disabled children and the management of their complications.

The current study revealed that the number of CMC who used TPPV and NPPV had increased by 1.4 times and that of patients using HOT by 1.8 times between Period 1 and Period 3. The number of patients undergoing home mechanical ventilation (HMV) in developed countries has been increasing. However, the number of patients using HMV varies widely according to regions [27–30]. A significant increase in the number of pediatric patients using HMV is significantly attributed to the increase in the number of children using NPPV [30], which was consistent with the results of the current survey. The increase in the number of patients using HOT was correlated to the increase in the number of HMV and CHD patients. Because surgery for CHD is increasing, the number of patients using HOT is expected to increase in the future [22].

In this survey, although the number of CMC who required tube feeding decreased from 51.7% in Period 1 to 38.7% in Period 3, the absolute number did not change. The increase in the number of patients using tube feeding has been reported in other countries [31,32], as well as a data from Japanese administrative claims database [33]. The decrease in the proportion of CMC requiring tube feeding is presumed to be a relative decrease due to an increase in the total number of CMC; moreover, the number of CMC using temporary tube feeding varied on a yearly basis, which might be partly attributed to the decrease in the proportion. Because tube feeding is indispensable for the growth and development of children, the need for tube feeding is expected to increase the survival rate of children with severe disabilities.

Furthermore, this study validated the content of medical care among CMC who have relatively preserved motor and intellectual abilities. The need for medical care in Group 4 CMC was lower than that in Group 1 CMC. This result was consistent with that of the survey by Caicedo showing that the use of medical devices in children with moderate disability is significantly lower than that with vegetative state [10]. In the present study, 27% of patients in Group 4 CMC required respiratory management, particularly NPPV. This relatively high usage rate was attributed to disease specificity. Of the 44 patients, 15 presented with muscular diseases, 11 with SAS, and 6 with airway stenosis.

The diversity of medical care and devices and the underlying disorders have a significant effect on the daily life of each CMC. Regarding education, children with SMID often enter schools for special need education with well-equipped facilities and numerous teachers and staffs involved in their care in Japan. Meanwhile, CMC who present with relatively preserved motor and intellectual abilities hope to enter regular kindergartens and schools in their neighborhood. Although these children may establish sociality and develop with other healthy children, they can be exposed to physical risks in their surroundings; moreover, there is a constant need for essential medical care and use of devices. The number of CMC going to general schools is increasing in Japan; however, they are also facing problems of provision with safe environment, teachers' involvement of medical care, and lack of school nurses [7]. As the resources available vary according to the region, their choices of schools often do not meet their wishes. Furthermore, it has been reported that some medical, welfare, and administrative services do not meet the needs of patients because they sometimes differ according to the region, cannot be adapted due to the difference in severity or disease category, and have restrictions of usage time [8–10]. One of the factors that do not meet the needs of patients is that there have been few surveys about the multifaceted evaluation of CMC. By classifying and investigating the underlying disorders, severity, and the contents of medical care and devices as described in this present study, it may be partly possible to design and provide satisfactory individualized services and education according to the severity of CMC to correspond to the wide range of CMC.

This study has several limitations. First, the prevalence could not be compared directly to that of other studies because the definition of CMC was not uniform. In terms of special consideration for medical and welfare services and education, we included children requiring medical care and use of devices. Thus, a definition that is in accordance with the aim of each study as well as a common unified definition for comparison among regions must be established. Second, this was a retrospective study. The severity of CMC was examined using data in the final follow-up. Because the condition of CMC may change over time, a prospective survey must be conducted for a more detailed classification of severity in each study period.

This survey first validated the prevalence of CMC with various severities and underlying disorders in Tottori, Japan. The details of medical care and use of devices required in CMC must be validated. As primary care physicians and pediatric neurologists are most familiar with CMC, it is necessary not only to provide medical care according to various types of CMC but also to collaborate with welfare and education to share the accurate needs of CMC. In addition, it is necessary to establish a common definition of CMC in the community, to perform prospective multifaceted evaluation of CMC in each region, and to simultaneously investigate the utilization, satisfaction, and actual needs of medical, welfare, and administrative services according to the times. Further studies must be conducted to provide appropriate individualized medical, welfare, and administrative services and education about the different types of CMC.

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Conflict of Interest

The authors declare no potential conflicts of interest with respect to the research, authorship, and publication of this article.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.braindev.2020.06.008.

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