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Mental wellbeing in non-ambulant youth with neuromuscular disorders: What makes the difference?

Vivienne Travlos[a,*], Jenny Downs[b,c], Andrew Wilson[d], Dana Hince[e], Shane Patman[a]

[a] School of Physiotherapy, University of Notre Dame Australia, PO Box 1225, Fremantle, Western Australia 6959 Australia
[b] Telethon Kids Institute, Roberts Rd, Subiaco, Western Australia 6008 Australia
[c] School of Physiotherapy and Exercise Science, Curtin University, Kent Street, Bentley, Western Australia 6102 Australia
[d] Respiratory Medicine, Perth Children's Hospital, 15 Hospital Ave, Nedlands, Western Australia 6009 Australia
[e] Institute of Health Research, University of Notre Dame Australia, PO Box 1225 Fremantle, Western Australia 6959 Australia

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Abstract

The physical and social challenges associated with neuromuscular disorders may impact mental wellbeing in non-ambulant youth during the more vulnerable period of adolescence. This cross-sectional survey investigated non-ambulant youths' mental wellbeing and relationships with physical health, participation and social factors. The conceptual model was the International Classification of Functioning, Disability and Health (ICF). Thirty-seven youth aged 13–22 years old (mean age 17.4 years; n = 30 male; n = 24 Duchenne Muscular Dystrophy) and their parents provided biopsychosocial data through a comprehensive self-report questionnaire. The primary outcome measure was the Warwick Edinburgh Mental Wellbeing Scale (WEMWBS). Relationships between mental wellbeing and variables within and across each ICF domain were explored using linear regression models. Mean WEMWBS scores (55.3/70 [SD 8.1]) were higher than for typically developing youth and comparable to youth with other chronic conditions. Over half of youth reported severe co-morbidities across all body systems. Multivariable modelling indicated that mental wellbeing was independently associated with academic achievement and perceived family support but not with physical health variables. Beyond management of physical co-morbidities, enabling youths' educational attainment and attending to social support likely optimises youth's wellbeing.

Keywords: Neuromuscular disorders; Youth; Non-ambulant; Well-being; Comorbidity; Participation.

1. Introduction

Severe childhood onset neuromuscular disorders (NMD) such as Spinal Muscular Atrophy (SMA) and Duchenne Muscular Dystrophy (DMD) cause muscle weakness that precludes attainment of walking or necessitates wheelchair dependence usually by adolescence [1]. Physical health is compromised by lack of ambulation and progressive muscle weakness leading to co-morbidities across body systems [2]. Cardiorespiratory system co-morbidities restrict life expectancy [2,3]. Survival well into adulthood is possible with timely provision and uptake of best practice interventions including use of corticosteroids, spinal fusion, and non-invasive ventilation (NIV) [4]. Whilst such intervention optimises physical health, initiation of NIV, for example, signifies the vulnerability of survival and may have adverse impacts on individual and family wellbeing [5].

Wellbeing encompasses domains of an individual's perception of their health, happiness and life satisfaction [6,7]. The International Classification of Functioning, Disability and Health (ICF) [8] provides a framework to conceptualise the physical and social factors that may impact an individual's health and wellbeing. Mental wellbeing during adolescence appears particularly vulnerable, indicated by a sharp rise in mental disorders in typically developing youth [9]. Challenges specific to adolescence include navigating rapid physical changes, identity formation and increasing expectations of autonomy. Physical health and mental wellbeing may be especially vulnerable in non-ambulant youth with NMD whose

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[*] Corresponding author.
E-mail address: vivienne.travlos@nd.edu.au (V. Travlos).

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additional challenges include living with a degenerative health condition, uncertain life expectancy, and limited physical autonomy. Despite an awareness of their physical health needs, non-ambulant youth with NMD described exercising autonomy in their choice to delay uptake of prescribed NIV when this challenged their identity development [10]. Wellbeing in non-ambulant youth is not well understood; studies report wellbeing from poor to comparable to typically developing youth, depending on the measurement tool used [11].

Maintenance of physical health appears linked to health intervention uptake and self-management, which in youth with NMD's is positively associated with psychosocial factors including better parent mental health [12], ethnicity and socioeconomic advantage [13], and in youth with a similarly deteriorating condition (cystic fibrosis) with greater parent support [14] and disease knowledge [15]. Positive findings are published for non-ambulant, ventilator dependent young adults with NMD, with reports that three in four parent caregivers rate their young adult with NMD in "good health" [16] and that wellbeing can be "(very) good" [17], though the latter was associated with having less pain and fatigue [18] and with having opportunities for social, recreational and educational participation [17,19]. A broader baseline picture is needed of youths' own perspective of their mental wellbeing, physical health, participation and social factors. Exploration of associations of these factors may inform what makes the biggest difference to mental wellbeing in youth preparing to transition to adulthood [20].

This study aimed to (i) describe self-reported mental wellbeing in non-ambulant youth with NMD and compare to published data of other groups of adolescents, (ii) identify comorbidities, participation and social factors associated with youth's mental wellbeing and (iii) explore within the ICF framework which factors may be independently associated with youths' mental wellbeing.

2. Materials and methods

2.1. Study design and participants

This study was a cross sectional survey. Eligible participants were youth (13–22 years old) with NMD who used wheelchairs full-time and their parents. Youth who were able to stand transfer and walk a few steps with support were excluded. Recruitment across Australia by mail-out invitation was conducted between September 2014 and July 2016 via eight patient organisations and five tertiary children's hospitals with neuromuscular clinics. Those indicating interest were asked to complete a paper-based questionnaire. All participants provided written, informed consent. Structured interviews were conducted by the first author as necessary to minimise missing data. Approvals were obtained from the Human Research Ethics Committees (HREC) of The University Notre Dame Australia (014122F), Princess Margaret Hospital for Children (2014103EP), the Women's and Children's Health Network (HREC/15/WCHN/23), the Sydney Children's Hospital Network (HREC.15.SCHN.194) and Children's Health Queensland (HREC/16/QRCH/31).

2.2. Measures

A comprehensive questionnaire that was a compilation of validated scales and de novo questions to measure each of the primary and independent variables was developed using the ICF domains as a framework (Fig. 1). Guidance with the content of the questionnaire was provided by expert clinicians and researchers through the Australasian Neuromuscular Network (www.ann.org.au) and the Treat-NMD Registry of Outcome Measures [21]. Four young adults with NMD piloted the questionnaire to identify any ambiguity with instructions or questions. Feedback was sought in regard to readability, clarity, face validity, design and time taken to complete, with minor modification to the survey tool undertaken accordingly.

2.3. Primary outcome – mental wellbeing

The Warwick Edinburgh Mental Wellbeing Scale (WEMWBS) [22] is validated in typically developing youth [23] and asks participants to rate their thoughts and feeling "*in the last two weeks*" in 14 positively phrased questions capturing domains of life satisfaction, positive affect, meaning and purpose. Responses are rated on 5-point Likert scale ranging from "*none of the time*" to "*all of the time*". These were summed to provide a score out of 70, where higher scores indicated greater mental wellbeing.

2.4. Independent variables

2.4.1. Activity (Function)

Activity was measured as physical ability utilising the Egen Klassification Scale Version 2 (EK2) [24]. Shown to be valid and reliable in non-ambulant individuals with NMD [25], responses are rated on a Likert scale and higher scores indicated greater physical ability.

2.4.2. Physical health – co-morbidities of body functions and structure

Participants rated frequency of health complaints on the Health Behaviour of School Aged Children Measurement Question 55 (HBSC MQ55) Symptom Check List [26,27]. This scale asks about eight complaints ("*headache*", "*stomach ache*", "*backache*", "*dizziness*", "*feeling low*", "*irritability or bad temper*", "*nervousness*" and "*difficulties getting to sleep*") over the previous six months on a 5-point scale ("*almost every day*" to "*rarely or never*"). Higher HBSC MQ55 scores indicated less frequent health complaints.

Cardiorespiratory system co-morbidities were categorised by the number of episodes of respiratory illness requiring antibiotic use in the previous 12 months, whether and when NIV had been issued, pattern of daily NIV use and the presence of a diagnosis of cardiomyopathy. Daytime fatigue was measured in three questions modified from a scale for typically developing school aged youth [28] ("*how often do you:*

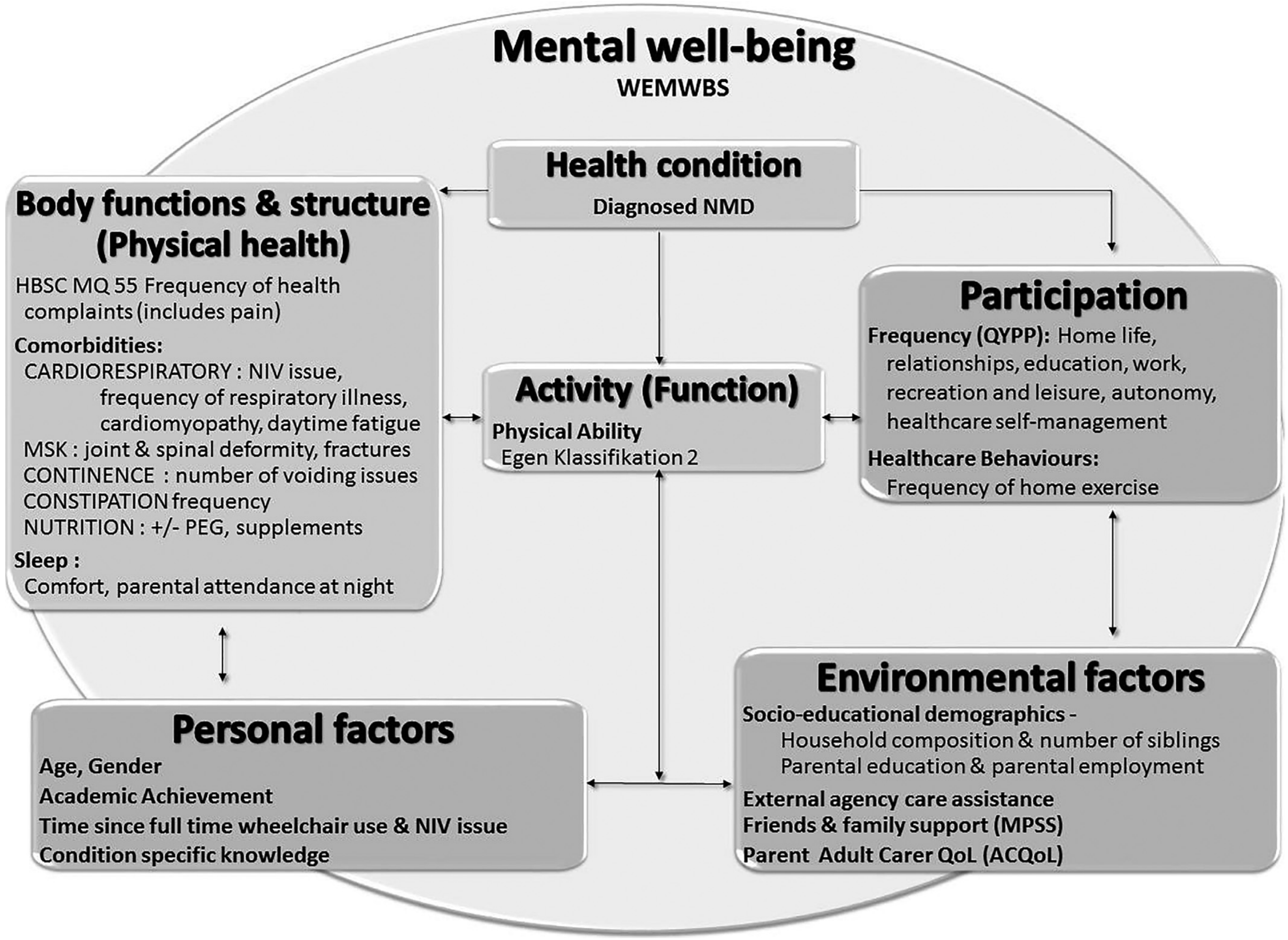

Fig. 1. Questionnaire variables within the International Classification of Functioning, Disability and Health (ICF) framework
ACQoL – Adult Carer Quality of Life; HBSC – Health Behaviour of School Age Children study; MPSS – Measure of Perceived Social Support; MQ – Measurement Question; MSK – musculoskeletal; NIV – non-invasive ventilation; NMD – Neuromuscular Disorder; PEG – Percutaneous endoscopic gastrostomy, QYPP – Questionnaire of Young People's Participation; WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

1. fall asleep during the day, 2. get drowsy during the day, 3. feel you need more sleep."). These questions were added as this information was not captured in the EK2 and MQ55 measures and on face validity they were reflective of fatigue irrespective of physical ability. Responses were summed and a higher score was used to indicate more frequent daytime fatigue.

Musculoskeletal system co-morbidities included: the total of fractures experienced at any age; scoliosis categorised as *"none"*, "*Cobb angle* < *40°"* and *"Cobb angle* > *40°"* with or without spinal fusion; the number of severe joint contractures totalled from responses to "*very tight*" descriptors of range of motion for shoulder elevation, elbow flexion and extension, hip extension, knee flexion and extension and ankle dorsiflexion.

Nutritional status was categorised by the need for oral supplementation and/or Percutaneous Endoscopic Gastrostomy (PEG) feeding to maintain body weight. Urinary continence issues were captured as the count of affirmative responses to five questions of issues experienced by wheelchair dependent youth: limited fluid intake ("*I don't drink so I don't have to go to the toilet"*), bladder emptying issues (difficulty passing urine and use of urinary catheter) and leaking ("*I leak urine because I often leave asking for help too late*" and "*I leak urine because I' m embarrassed/avoid asking for help*"). Constipation was categorised as *"none"* and *"occasional / frequent"* through affirmative answers to questions of frequency and assistance with bowel voiding. These were modified from the Rome III Criteria for Functional Gastrointestinal Disorders [29].

Youth described frequency of sleep discomfort ("*How often is comfort at night a problem*", rated on 5-point Likert scale) and their parent reported frequency of attendance at night.

2.4.3. Participation

Frequency of participation in life situations specific to young people (home life, relationships, education, work, recreation and leisure, and autonomy) was measured using the Questionnaire of Young People's Participation (QYPP) [30]. Scores for each item were totalled for each category and higher scores reflected greater frequency of participation [31].

2.4.4. Personal factors

Descriptive variables of age, gender and date of wheelchair and ventilator issue were collected. Youth self-rated their academic achievement in comparison to their school peers as "*above average*" ("*My marks in most subjects are higher than those of most other kids at my school*"), "*average*" ("*My marks in most subjects are about in the middle compared to other kids at my school*"), "*below average*" or "*life skilled*", where the latter described those enrolled in a modified curriculum in special needs classes or a special school. Specific knowledge around NMD was measured with 11 questions from the Care-NMD survey [32]. The number of affirmative "*yes, sufficiently"* responses were totalled (maximum 11) with a higher total indicating greater condition specific knowledge.

2.4.5. Environmental factors

Youth's perceived social support was measured using the Multidimensional Scale of Perceived Social Support (MPSS) [26] questions specific to friends and family. Responses to statements of support rated on a Likert scale from "*very strongly disagree*" to "*very strongly agree*" were totalled and higher scores indicated greater perceived support. Demographic information collected from parents/primary caregiver included family composition, highest education level, employment status and the number hours of external agency support. Parents reported parent-carer QoL using the Adult Carer Quality of Life Questionnaire (AC-QOL) [33], with higher scores indicating better carer QoL.

The questionnaire is available on request from the corresponding author.

2.5. Statistical analyses

Normally distributed data were expressed as means and standard deviations (SD), skewed data as medians and interquartile ranges (IQR), and categorical variables as frequencies and percentages. One-sample Wilcoxon signed-rank tests were used to compare WEMWBS median scores of youth with NMD in this study with median scores in a sample of typically developing youth reported by Clarke et al. [23] and in samples of youth with chronic conditions reported by Merrick et al. [34]. Ninety-five percent confidence intervals for median differences were determined using bootstrapping procedure (1000 samples) [35].

Associations between WEMWBS scores and the independent variables were explored using linear regression models. A three-step modelling procedure was used: Step 1 investigated univariable associations between the independent variables and WEMWBS scores; Step 2 comprised a multivariable linear regression model between independent variables and WEMWBS scores *within* each ICF domain (Fig. 1); Step 3 comprised a multivariable linear regression model including significant variables from those identified in Step 2 *across all ICF domains*. A variable was considered significantly associated with the outcome if the Wald Chi-Square test of model effects returned $p < 0.05$. Statistical analyses were calculated using SPSS® Statistics (Version 24, IBM Corp, New York: USA).

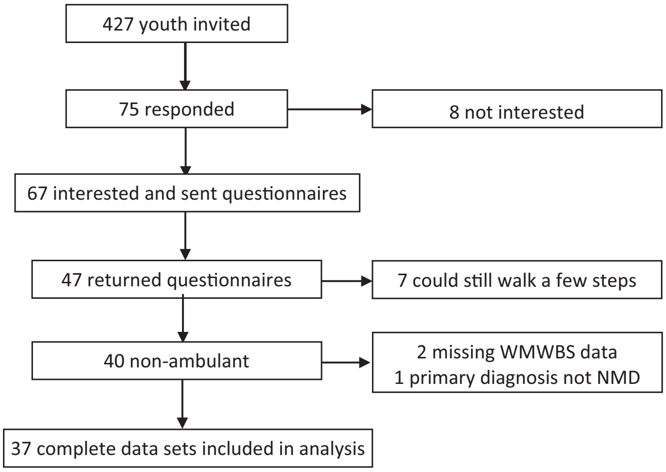

Fig. 2. Participant flowchart
WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

3. Results

3.1. Participant characteristics

Thirty-seven youth non-ambulant youth from across Australia returned complete data sets and were included in this analysis (Fig. 2). Participant characteristics are shown in Table 1. Sixty-five percent of participants lived with DMD ($n = 24$). Youth grouped as 'other NMD's' included those diagnosed with SMA II ($n = 3$, 2 female) and Nemaline Rod Myopathy ($n = 3$, female) and Congenital Muscular Dystrophy ($n = 7$, 2 female). Youth had been full-time wheelchair users for at least 5 months prior to questionnaire completion. The EK2 scores did not significantly differ between diagnostic group, gender or age grouped by above and below Australian school leaving age (18 years old) (Table 1).

3.2. Mental wellbeing

The WEMWBS scores were high with a mean of 55.3/70 (SD 8.1, range 39–68). Median WEMWBS scores for youth with NMD were more than five points higher than for typically developing youth and for youth with Autism Spectrum Disorder, but were similar to scores for youth with Cerebral Palsy or Diabetes (Table 2).

3.3. Co-morbidities, participation and social factors

Over half of youth reported severe comorbidity in one or more body systems (Table 3). Feeling nervous ($n = 10$, 27%), backache and difficulty getting to sleep ($n = 9$, 24% each) and

Table 1
Participant characteristics ($N=37$).

Characteristic	Group	*n* (%)	EK2[a] Mean /53 (SD)	Test for difference, *t* (36)
Diagnosis	DMD	24 (65%) [all male]	33.4 (8.7)	0.27, $p=0.79$
	Other NMD	13 (35%) [7 female]	32.6 (8.3)	
Gender	Male	30 (81%)	33.0 (8.9)	−0.20, $p=0.84$
	Female	7 (18%)	33.7 (6.7)	
Age (years)	13–17	21 (57%)	34.0 (8.0)	0.71, $p=0.49$
	18–22	16 (43%)	32.0 (9.2)	

[a] EK2 – Egen Klassifikation 2 Scale, measure of physical ability (maximum score 53).

Table 2
Mental wellbeing median scores for youth with NMD compared with typically developing youth[a] and youth with longstanding chronic conditions.[b]

		WEMWBS score Median [IQR]	Median difference: NMD and group of comparison	
			Median difference (95% CI)	*p*
	Youth with NMD (n=37)	54 [49-62]		
Groups of comparison	Typically developing youth[a] (n=1517)	49 [43-54]	5.0 (3.0 to 10.0)	<0.001
	Autism Spectrum Disorder[b] (n=118)	47 [41-52]	7.0 (4.0 to 12.0)	<0.001
	Cerebral Palsy[b] (n=106)	53 [48-60]	1.0 (−2.0 to 6.0)	0.116
	Diabetes[b] (n=150)	53 [47-58]	1.0 (−2.0 to 6.0)	0.116

[a] Group data derived from Clarke et al [23].
[b] Group data derived from Merrick et al [34]
CI – Confidence Interval; WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

Table 3
Univariable effects of ICF physical health variables on mental wellbeing.

		N=37	Mental Wellbeing WEMWBS scores	
Predictor Variable (/scale max)		**n (%) /Median [IQR]**	**β coefficients (95% CI)**	**p**
Health condition				
Type of NMD	DMD	24 (65%)	0.0 (reference)	–
	Other NMD	13 (35%)	1.01 (−4.40 to 6.42)	0.715
Activity (function)				
Egen Klassifikation 2 score (/53)		34.0 [26.5 – 39.0]	0.17 (−0.14 to 0.47)	0.282
Physical health				
HBSC MQ55 Frequency of health complaints (/40)		34.5 [29 – 36.5]	**0.56 (0.17 to 0.95)**	**0.005**
NIV issue	Yes	25 (68%)	0.0 (reference)	–
	No	12 (32%)	1.82 (−3.68 to 7.32)	0.516
Cardio-myopathy	Yes/unsure	27 (71%)	0.0 (reference)	–
	No	11 (29%)	3.63 (−1.91 to 9.16)	0.199
Daytime fatigue (/9)		2 [1 – 3]	**−1.86 (−2.97 to −0.75**	**0.001**
Respiratory illness	None	21 (57%)	0.0 (reference)	–
	1-2	12 (32%)	0.67 (−4.99 to 6.32)	0.817
	≥ 3	4 (11%)	−2.58 (−11.11 to 5.94)	0.553
Deformity – severe contractures		1 [1–2]	−0.43 (−1.83 to 0.97)	0.550
Deformity - Spinal scoliosis	No scoliosis	10 (27%)	0.0 (reference)	–
	<40°Cobb angle	20 (54%)	−0.85 (−6.61 to 4.91)	0.772
	>40°Cobb angle	7 (19%)	**−7.24 (−14.57 to 0.81)**	**0.053**
Fractures (total number)		1 [0–2]	**−1.46 (−2.80 to −0.08)**	**0.039**
Nutritional supplement	None	31 (84%)	0.0 (reference)	–
	PEG/Oral supplement	6 (16%)	**−8.08 (−14.60 to −1.56)**	**0.015**
Urinary continence issues total no.		1 [0–1]	−2.25 (−5.52 to 1.02)	0.177
Constipation	Occasional / frequent	20 (54%)	0.0 (reference)	–
	Never	17 (46%)	3.96 (−1.07 to 8.99)	0.123
Sleep discomfort	Often	13 (35%)	0.0 (reference)	–
	Sometimes	20 (54%)	2.15 (−3.06 to 7.36)	0.764
	Never	4 (11%)	**10.25 (1.88 to 18.62)**	**0.016**
Parental assistance at night (n=35)	Never/occasional	18 (49%)	0.0 (reference)	–
	≥ twice a night	17 (46%)	−4.34 (−9.45 to 0.77)	0.096

Bolded numbers indicate significant association (95% CI excluding zero).
HBSC MQ – Health Behaviour of School Age Children Measurement Question; NIV – non-invasive ventilation; PEG – Percutaneous Endoscopic Gastrostomy; WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

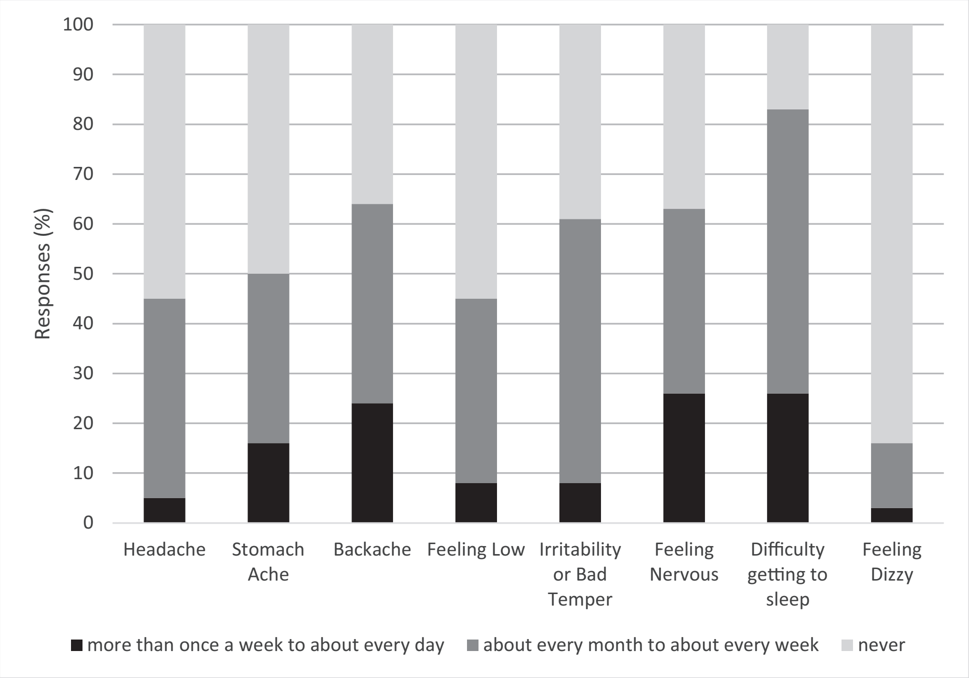

Fig. 3. Frequency of health complaints ($N = 37$) as per Health Behaviour of School Aged Children Measurement Question 55 (HBSC MQ55).

stomach ache ($n = 6$, 16%) were frequently reported as occurring "*more than once a week*" and "*about every day*" (Fig. 3). Two thirds ($n = 25$) used NIV, with time since NIV issue ranging from 1 month to 14.7 years. Over half ($n = 21$, 57%) reported no acute respiratory illness requiring antibiotic use in the prior 12 months. Twelve youth (32%; 7 with DMD, 5 with other NMD's) were unsure of their cardiac status. Over half ($n = 21$, 57%) had experienced fractures ranging from one to seven fractures in any bone. A minority ($n = 6$) required nutritional supplementation to maintain their body weight. Sufficient condition specific knowledge was reported by most youth for "*breathing and chest health*" ($n = 34$, 92%) and by a small proportion of youth for "*gastric tube/PEG placement in case of weight loss and difficulties with eating*" ($n = 11$, 30%).

Self-rated academic achievement was high, with 43% of youth ($n = 16$) rating their academic achievement as above average compared with their peers (Table 5). Frequency of participation was low across life areas, highest in 'school or university life' and lowest in 'work life' (Table 4). Only four participated in the workplace (2 paid and 2 volunteer), each for less than 10 hours per week. None of the five youth who had completed university, or Technical and Further Education held a formal job. Participation in 'relationships' was reported most frequently in using "*on-line communication*" ($n = 29$, 78%) and "*helping friends or family when they are upset (e.g. by listening to & supporting them)*" ($n = 24$, 65%) and least in "*spending time with a boyfriend/girlfriend*" ($n = 6$, 16%).

Youth perceived high levels of support; 21 (57%) "*very strongly agreed*" with all statements of their family's support and 13 (35%) "*very strongly agreed*" with all statements of their friends' support. There was diversity in social demographics, external agency care service provision and parents' AC-QoL (Table 5).

3.4. Associations with mental wellbeing

The univariable relationships between WEMWBS scores and independent variables within each ICF domain are shown in Tables 3–5. The WEMWBS scores were not associated with diagnostic group, EK2 scores, NIV issue (Table 3), age or gender (Table 5). Perceived above average academic achievement was associated with higher WEMWBS scores, on average almost 10 points higher compared to youth with average achievement (coefficient 9.50; 95% CI 4.87–14.13; $p = < 0.001$). The WEMWBS scores of those reporting below average academic achievement was an average of six points lower than those reporting above average academic achievement ($p = 0.059$) (Table 5). More frequent participation in relationships was associated with greater WEMWBS scores (coefficient 0.41; 95% CI 0.07 to 0.75; $p = 0.017$) (Table 4).

Within the *physical health* domain, more frequent health complaints, greater daytime fatigue, higher fracture frequency, need for nutritional supplementation, and discomfort during sleep were all associated with lower WEMWBS scores (Table 3). Accounting for the combined effects of these variables, none were independently associated with lower WEMWBS scores (Table 6).

For the environmental factor variables, having fewer siblings and greater perceived support from family and friends were associated with higher WEMWBS scores. There were very weak relationships with socioeconomic variables such as household composition and parental employment or education

Table 4
Univariable effects of participation variables on mental wellbeing.

		N = 37	Mental Wellbeing WEMWBS scores	
Predictor Variable scale max)		n (%) / Median [IQR]	β coefficients (95% CI)	p
QYPP Home life	(/25)	2.0 [0–5.0]	0.06 (−0.48 to 0.59)	0.841
QYPP School/ university life (N = 29)[a]	(/25)	15.4 [11.3–18.5]	0.17 (−0.68 to 1.02)	0.699
QYPP Work life (N = 4)[b]	(/16)	0.0 [0.0–0.0]		
QYPP Relationships	(/48)	20.2 [12.5–24.6]	**0.41 (0.07 to 0.75)**	**0.017**
QYPP Leisure and recreation	(/84)	28.0 [23.0–32.0]	0.28 (−0.05 to 0.60)	0.095
QYPP Autonomy	(/16)	12.6 [10.0–13.8]	0.51 (−0.38 to 1.52)	0.239

QYPP – Questionnaire of Young People's Participation; WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

[a] Missing responses by 8 youth in older age group no longer in formal education.

[b] Domain not subjected to regression analysis due to low cell count.

Table 5
Univariable effects of social context variables on mental wellbeing.

		N = 37	Mental Wellbeing WEMWBS scores	
Predictor Variable (/x scale max)		n (%) / Median [IQR]	β coefficients (95% CI)	p
Personal factors				
Age		16.9 [15.7–18.8]	0.26 (−1.06 to 1.12)	0.963
Gender	Male	30 (81%)	0.0 (reference)	–
	Female	7 (19%)	−1.21 (−7.81 to 5.38)	0.718
Academic Achievement	Above average	16 (43%)	0.0 (reference)	–
	Average	16 (43%)	**−9.50 (−14.13 to −4.87)**	**<0.001**
	Below average/Life skilled	5 (14%)	−6.45 (−13.16 to 0.26)	0.059
Years since full-time wheelchair use		6.0 [4.5–11.0]	0.06 (−0.50 to 0.63)	0.825
Years since NIV issue (n = 25)		2.2 [1.3–6.5]	−0.01 (−0.77 to 0.76)	0.990
Condition specific knowledge (/11)		8.0 [5.0–9.0]	−0.26 (−1.20 to 0.69)	0.593
Environmental factors				
Household composition	Two parents	32 (86%)	0.0 (reference)	–
	Single parent	5 (14%)	−0.24 (−8.80 to 6.32)	0.748
Number of siblings (range 0–4)		1 [1–2]	**−3.32 (−5.43 to −1.21)**	**0.002**
Responding parent employed (N=35)[a]	Yes	24 (65%)	0.0 (reference)	–
	No	11 (30%)	−3.08 (−8.70 to 2.54)	0.282
Responding parent's education (N = 35)[a]	University trained	18 (49%)	0.0 (reference)	–
	Vocation trained	9 (24%)	−1.39 (−7.12 to 4.94)	0.667
	School only	8 (22%)	−3.04 (−9.63 to 3.55)	0.365
Perceived social support family (/28)		28 [26–28]	**1.59 (0.76 to 2.42)**	**<0.001**
Perceived social support friends (/28)		26 [23–28]	**0.89 (0.26 to 1.53)**	**0.006**
External agency care assistance hours (N =35)[a]	Daily	11 (30%)	0.0 (reference)	–
	A few times a week	15 (41%)	−3.76 (−9.85 to 2.34)	0.227
	None	9 (24%)	−1.42 (−8.33 to 5.48)	0.686
Parent Carer ACQoL (N = 35)[a] (/114)		79.0 [64.5–88.5]	0.13 (−0.10 to 0.27)	0.069

Bolded numbers indicate significant association (95% CI excluding zero).

ACQoL – Adult Carer Quality of Life Questionnaire; NIV – Non-Invasive Ventilation; WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

[a] Missing parent completed data of 15yo and 19yo with other NMD.

(Table 5). Greater perceived support from family remained independently associated with higher WEMWBS scores when the model accounted for the effects of number of siblings and friend support (Table 6).

Combining all significant variables identified in the multivariable within-ICF-domain models, the final multivariable model across all ICF domains found that WEMWBS scores were higher for above average academic achievement (Wald Chi-Square test of model effects 19.6 [2df]; $p = < 0.001$) and perceived support by family (Wald Chi-Square test of model effects 12.6 [1df]; $p = < 0.001$). Neither frequency of health concerns nor fracture frequency were associated with WEMWBS scores when accounting for the effects of personal (academic achievement) and environmental factors (family support) (Table 6).

4. Discussion

This study sought to describe non-ambulant youths' mental wellbeing and factors that may make the biggest difference to their wellbeing. We found that youth reported relatively high mental wellbeing compared with the general population, despite more than half experiencing severe co-morbidities across cardiorespiratory, musculoskeletal, renal and gastroin-

Table 6
Multivariable effects on mental wellbeing.

		Mental Wellbeing WEMWBS scores			
		Multivariate STEP 2 findings combining significant variables WITHIN ICF domains		**Multivariate STEP 3 findings combining significant variables ACROSS ICF domains**	
Predictor Variable		**β coefficients (95% CI)**	**p**	**β coefficients (95% CI)**	**p**
Physical health					
HBSC MQ55 Health complaints		0.34 (−0.07 to 0.76)	0.106	–	–
Daytime fatigue (/9)		−0.52 (−1.73 to 0.69)	0.401	–	–
Fracture frequency total no		−1.01 (−2.14 to −0.13)	0.083	–	–
Nutritional supplement	None	0.0 (reference)	–	–	–
	PEG/Oral supplement	−4.83 (−10.42 to 0.76)	0.090	–	–
Sleep discomfort	Often	0.0 (reference)	–	–	–
	Sometimes	1.12 (−3.63 to 5.88)	0.644	–	–
	Never	5.89 (−1.53 to 13.30)	0.120	–	–
Participation					
QYPP Relationships		**0.41 (0.74 to 0.75)**	**0.017**	0.24 (−0.01 to 4.93)	0.057
Personal factors					
Academic Achievement	Above average (n=16)	0.0 (reference)	–	0.0 (reference)	–
	Average (n=16)	**−9.50 (−14.13 to −4.87)**	**<0.001**	**−7.84 (−11.34 to −3.53)**	**<0.001**
	Below average / Life skilled (n=5)	−6.45 (−13.16 to 0.26)	0.059	−3.38 (−9.04 to 2.28)	0.242
Environmental/social context factors					
Number of siblings (range 0–4)		**−2.32 (−4.16 to −0.47**	**0.014**	–1.52 (−3.17 to 0.13)	0.071
Perceived social support family		**1.12 (−0.34 to 1.08)**	**0.005**	**1.17 (0.52 to 1.81)**	**<0.001**
Perceived social support friends (/28)		0.52 (−0.03 to 1.01)	0.066	–	–

Bolded numbers indicate significant association (95% CI excluding zero).
HBSC MQ – Health Behaviour of School Age Children Measurement Question; WEMWBS – Warwick Edinburgh Mental Wellbeing Scale.

testinal systems. Above average academic achievement and greater perceived family support influenced mental wellbeing the most, beyond effects of physical co-morbidities.

Mental wellbeing scores of youth in this study were comparable to youth with other chronic physical conditions such as cerebral palsy and diabetes [34] and higher than reported in typically developing youth [23] and youth with autism spectrum disorder [34]. Whist this finding might seem somewhat surprising, it reaffirms the ‘disability paradox’ whereby mental wellbeing does not have a linear association with severity of physical disability [16,17]. One explanation suggested by youths’ qualitative accounts is that youth with chronic conditions who are aware of their physical difference, vulnerability and uncertain life expectancy from an early age use very different reference points by which to judge their health, happiness and life satisfaction [10,36]. An alternative consideration is that physically dependent youth may also experience psychological dependence, thus being less inclined to demonstrate typically adolescent oppositional behaviour such as through reporting lower life satisfaction or experiencing less autonomy to express negative aspects of their wellbeing [37]. Youths’ WEMWBS score distribution in our study was consistent with the comparable studies [23,34], suggesting a believable range of mental health experiences.

We found that perceived family support had the strongest association with youths’ mental wellbeing. This finding is of interest alongside our univariable association of lower wellbeing in larger families. When perceived family support was higher, number of siblings was no longer an influential factor. Provision of individualised support may be more complex in larger families, but any attempt to explain how families managed their unique contexts is to undermine the diverse ways in which they learn to live with severe disability. With growing knowledge of caregiver burden in youth with NMD [38], studies that aim to disentangle how families manage youths’ health and support their mental wellbeing are fertile ground for future research.

The importance of supportive relationships to mental wellbeing is undisputed. Supportive relationships are likely also formed through social participation, reinforced by our finding that greater frequency of participation in relationships was associated with youth’s greater mental wellbeing. Youth and families with NMD call for as much attention paid to participation as to medical intervention [39,40] and it is likely that meaningful participation such as in team sports and social activities strengthens supportive relationships. These activities can contribute to individuals’ health self-management through sharing ideas on managing and normalising the experience of living with NMD [41] with supportive peers. Youth with NMD have described balancing social participation with health self-management, for example by limiting their participation to avoid exposure to acute infections and to manage fatigue [10]. The finding that only 6 of the 37 youths had a boyfriend of girlfriend is in keeping with the reported experience of other youth with chronic conditions and consequently healthcare professionals are encouraged to have meaningful conversations with youth about developing healthy romantic relationships [42].

The association of greater wellbeing with higher academic achievement in non-ambulant youth with NMD in our study replicates findings by others [37,43]. This finding may reflect the positive experiences in cognitively able youth who accept that their physical limitations preclude them from competing for physical achievement, and instead focus on and achieve success in academic pursuits. We did not collect intelligence quotient data and cannot be confident that our sample reflects the approximate quarter of youth with DMD with intellectual disability or specific learning disabilities [44,45]. We also acknowledge that our opt-in sampling strategy likely biased our study to more able individuals but the small difference in wellbeing between the above average and below average academic achievers suggests the needs of those with greater educational difficulties were met. Since youths' mental wellbeing is strengthened by their perceived support by their friends who may be their intellectual peers, then it is important that individuals be challenged to achieve according to their intellectual abilities. Enabled educational attainment appears a protective factor of health and wellbeing in youth with chronic conditions in the period of transition to adulthood [46]. It is positive that all but one older youth in our study participated in tertiary education, yet the minimal participation in the workplace in volunteer and paid employment is disappointing, given that all but two youth (who were under the age of 14 years old) were eligible for paid jobs. Accounts of contributions of employed, cognitively able young adults with NMD [41,47] suggest efforts made to enable education and workplace participation are worthwhile.

Contrary to our study hypothesis, respiratory co-morbidities and use of NIV were not associated with mental wellbeing. In terms of frequency of respiratory illness, the current sample appeared fairly healthy and condition specific knowledge was reported highest in relation to this body system. Youths' accounts of self-management in respiratory care may reflect greater uptake of best practice care, or the relatively more robust evidence informing respiratory care [48] than that informing other body systems. We found association of lower mental wellbeing in individuals with a greater number of fractures and this may be related to associated pain and lengthy recovery periods. Associations of other musculoskeletal pain, sleep discomfort and daytime fatigue with lower wellbeing in youth with NMD have been previously reported [18,49] and may be interrelated. The finding that the wellbeing score of youth with no sleep discomfort was on average almost 10 points higher than those with sleep discomfort suggests benefits can be gained from specifically attending to youths' pain, comfort and sleep. Taken together with problems with continence and constipation reported in over half of the sample, calls for coordinated, multidisciplinary patient education and management of body systems issues are reinforced [50,51]. Further research should explore how multidisciplinary teams can provide such co-ordinated care that also maximises available time for educational attainment and meaningful participation, and strengthens supportive relationships.

4.1. Strengths and limitations

A strength of this study is the comprehensive description of the biopsychosocial profile of a group uniform by developmental age and physical ability receiving care in different health care contexts from across Australia. Patient reported outcome measures provide data from youths' own perspective that can be a powerful tool to guide shared decision making [52] and are favoured by regulatory agencies for monitoring the impact of healthcare on functioning and wellbeing [53]. Data collected in this study provide a baseline for further exploration of how youth with NMD manage age-specific issues faced in transition to adulthood.

There are however several limitations of this study. No studies could be sourced exploring the WEMWBS's minimal clinically important difference. Whilst the difference in WEMWBS scores of youth in this study compared to typically developing youth was statistically significant, it is not yet known whether or how this difference is clinically meaningful. Though youth were instructed to complete the WEMWBS independently and offered the option of electronic completion, we did not audit how many youth used an adult's physical assistance with completing the paper version and cannot know whether an adult's presence swayed youths' answers. Future studies with youth should audit and document the level of participants' physical independence in survey completion to minimise risk of socially desirable answers.

Choice of validated, standardised self-report measures of physical health (for example fatigue, bladder and bowel function) was restricted by their lack of relevance to individuals with limited physical ability. Cross sectional data cannot infer causative relationships, only associations.

The risk of type two error is increased with small sample size, and other associations may be found or negated in larger samples. Opt-in recruitment required by Ethics Committees for this study may have favoured participation of youth and their families who were very motivated and had capacity to respond to a research invitation. That is, the sample may be over-representative of those within more confident, organised, supportive social environments in periods of stable health. Opt-out recruitment strategies may have given more youth and their families opportunity to consider study participation [54]. Development of robust measures relevant for youth with limited physical ability and their application to the same and similar populations are needed to validate current findings.

5. Conclusion

Our data provide a comprehensive picture of health and wellbeing across ICF domains in non-ambulant youth with NMD. Findings indicate that despite severe physical health co-morbidities and low frequency of participation, mental wellbeing can compare favourably to that of typically developing youth. The findings that severity of co-morbidity had little relationship with mental wellbeing should encourage healthcare professionals to review time spent focusing on co-morbidities; the proposition is strengthened that equal attention paid to youths' personal, educational and social contexts optimises wellbeing in non-ambulant youth with NMD in transition to adulthood.

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