Quality of Life Research (2019) 28:783–794
https://doi.org/10.1007/s11136-018-2057-3

Psychometric properties of the Quality of Life Inventory-Disability (QI-Disability) measure

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Accepted: 16 November 2018 / Published online: 20 November 2018

Abstract

Purpose Children with intellectual disability encounter daily challenges beyond those captured in current quality of life measures. This study evaluated a new parent-report measure for children with intellectual disability, the Quality of Life Inventory-Disability (QI-Disability).

Methods QI-Disability was administered to 253 primary caregivers of children (aged 5–18 years) with intellectual disability across four diagnostic groups: Rett syndrome, Down syndrome, cerebral palsy or autism spectrum disorder. Exploratory and confirmatory factor analyses were conducted and goodness of fit of the factor structure assessed. Associations between QI-Disability scores, and diagnostic and age groups were examined with linear regression.

Results Six domains were identified: physical health, positive emotions, negative emotions, social interaction, leisure and the outdoors, and independence. Goodness-of-fit statistics were satisfactory and similar for the whole sample and when the sample was split by ability to walk or talk. On 100 point scales and compared to Rett syndrome, children with Down syndrome had higher leisure and the outdoors (coefficient 10.6, 95% CI 3.4,17.8) and independence (coefficient 29.7, 95% CI 22.9, 36.5) scores, whereas children with autism spectrum disorder had lower social interaction scores (coefficient − 12.8, 95% CI − 19.3, − 6.4). Scores for positive emotions (coefficient − 6.1, 95% CI − 10.7, − 1.6) and leisure and the outdoors (coefficient 5.4, 95% CI − 10.6, − 0.1) were lower for adolescents compared with children.

Conclusions Initial evaluation suggests that QI-Disability is a reliable and valid measure of quality of life across the spectrum of intellectual disability. It has the potential to allow clearer identification of support needs and measure responsiveness to interventions.

Keywords Quality of life · Intellectual disability · Child · Adolescence · Measurement · Test validation

Abbreviations

ASD	Autism spectrum disorder
CI	Confidence interval
DIF	Differential item functioning
ICF	International Classification of Functioning, Disability and Health
PedsQL	Pediatric Quality of Life Inventory
QI-Disability	Quality of Life Inventory-Disability
QOL	Quality of life
SD	Standard deviation
WA	Western Australia

Electronic supplementary material The online version of this article (https://doi.org/10.1007/s11136-018-2057-3) contains supplementary material, which is available to authorized users.

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Introduction

Intellectual disability occurs in 1.9 per 100 children and approximately 15% of these children have severe impairment [1]. Worldwide the prevalence is higher in low- and middle-income group countries [2]. Children with intellectual disability have greater exposure to the social determinants of poor health such as economic disadvantage [3, 4]. The effects of intellectual disability are pervasive for both physical and mental health.

An important outcome to evaluate the effectiveness of services for children is quality of life (QOL), which refers to satisfaction with a composite of life experiences and includes domains that are universal (e.g. physical and mental wellbeing) with additional domains for particular populations [5]. As part of an evidence-based platform for patient-centred clinical care, service delivery and formulation of policy, it is critical that outcomes are evaluated in terms of individual experiences rather than professional assessment of what is important to children with intellectual disability [6]. We also need outcomes that incorporate the important features of impairment, activity, participation and the environment in which the child lives, as presented in the International Classification of Functioning, Disability and Health (ICF) [7]. Multifaceted interventions for complex conditions are unlikely to impact one outcome, and composite outcomes such as QOL could be suitable and efficient to measure [8]. However, our capacity to measure the impact of interventions in children with intellectual disability is limited because available QOL measures were not developed with the specific issues of children with intellectual disability in mind and did not involve families in their development [9]. For example, two qualitative studies on children affected by cerebral palsy [10] and autism spectrum disorder (ASD) [11] with comorbid intellectual disability found important themes missing from generic scales.

We recently undertook four qualitative studies to investigate the domains of QOL important to children with intellectual disability. In-depth interviews were conducted with parents of 6–18-year-old children with either Down syndrome ($n = 17$), Rett syndrome ($n = 21$; a severe genetic neurodevelopmental disorder mainly affecting females [12]), cerebral palsy ($n = 18$) or ASD ($n = 21$) [13–16]. Together these conditions represent a range of characteristics seen in the broader population of those with intellectual disability including functional, behaviour and socialisation difficulties; medical comorbidities; and different needs for autonomy. QOL domains were consistent across the four groups and included physical health and emotional wellbeing; pleasure in communication, movement and day-to-day routines; and satisfaction derived from social connectedness, leisure activities and the natural environment. Domains such as emotional wellbeing mapped broadly to other QOL measures but included many elements that were unique to our subject group [13–16]. These family-reported data were consistent conceptually with the ICF [7] and could usefully inform the content of a specific QOL measure for children with intellectual disability.

QOL is a concept evaluated through self-reflection, but this is challenging if cognitive abilities preclude self-report. For children, parents often act as proxies particularly if their child does not have verbal skills [17]. Based on our extensive qualitative dataset, this paper describes the development and validation of the Quality of Life Inventory-Disability (QI-Disability).

Methods

Data sources

Participants were parents of children registered with one of five databases. Families with a child with Down syndrome born from 1980 to 2004 who had previously participated in our research [18] were invited to take part, and additional families were recruited through Developmental Disability WA (a community organisation in the disability sector) and advertising on Facebook. Families with a child with Rett syndrome were recruited from the Australian Rett Syndrome Database, an ongoing population-based register established in 1993 that collects longitudinal data [19]. Families with a child with cerebral palsy and intellectual disability were recruited from the Victorian Cerebral Palsy Register, a population-based register of individuals with cerebral palsy born in Victoria since 1970 [20]. Families with a child with ASD and intellectual disability were recruited from the WA Autism Biological Registry [21] or the WA Autism Register [4].

Development

A working group (JD, AE, NM, HL) extracted statements from 77 interview transcripts [13–16] to illustrate observable aspects of each QOL domain. The statements were discussed by the group and edited to form questionnaire items. Where possible, items were worded positively to measure wellbeing rather than the converse and to reduce threats to the self-esteem of the parents who were completing [22]. However, negative behaviours were framed to explicitly acknowledge these behaviours as endorsed during consumer consultation. Each item was accompanied by a five-point Likert scale indicating the frequency of each aspect of the child's wellbeing.

The items were reviewed iteratively by the authors and the initial questionnaire draft comprised of 50 items.

We tested the meaning of the questionnaire items with a sample of parents using cognitive interviewing [23], to provide feedback on the comprehensibility and relevance of the items. Sixteen parents registered with one of the five databases were recruited and their children represented different ages, genders, clinical severity and comorbidities. During a recorded telephone interview, each parent was asked to complete the draft QI-Disability, describe their understanding of each item and share other thoughts (e.g. why they chose the rating category). Parents were recruited until thematic saturation was achieved as observed by repetition of responses. Parent wording and rationale for each of the items were tabulated. The wording of 24 items were clarified to better reflect the intended meaning of the item, three items were merged with other items and six items were removed due to a lack of utility (e.g. did not capture the intended meaning). The questionnaire then comprised of 41 items.

Validation

Between November 2016 and April 2017, QI-Disability was administered to 253 parents/primary caregivers using the REDCap (Research Electronic Data Capture) tool, with a paper format or telephone interview also available. With 41 candidate items, this sample was larger than the generally recommended sample size of five participants per item [24]. Of those families contacted, 98.4% (61/62) families with a child with Down syndrome, 95.6% (66/69) families with a child with Rett syndrome, 77.2% (64/83) families with a child with cerebral palsy and 91.2% (62/68) families with a child with ASD responded. Most (89.7%) respondents were biological mothers of whom 53.6% ($n=135$) worked full- or part-time and 17.4% ($n=44$) lived in a rural community. The mean age of the children was 12.2 (SD 4.1 years, range 5–18 years). Data describing the distribution of child and family characteristics are presented in Table 1.

Analyses

Exploratory factor analysis using all available data was performed to identify the factor structure using the iterated principal factor method incorporating promax rotation and pairwise deletion of missing data. A cutoff of 1.0 for the eigenvalue was used to define the domains to be retained and items with a loading <0.4 on any factor were excluded. Confirmatory factor analysis was then performed to verify the factor structure. To provide a basis for acceptance or rejection of the model, goodness of fit was assessed using the following statistics: CMIN/df value, root mean square error approximation, the Comparative Fit Index and the Tucker–Lewis Index. Cronbach's alpha, the Composite Reliability and Average Variance Extracted statistics were calculated for each factor to assess convergent validity. The maximum correlation squared value was calculated for each factor to assess divergent validity. Confirmatory factor analysis and goodness-of-fit analyses were also performed with the sample restricted to those who could walk independently or not, and those who could talk and be understood by those who did not know the child well or not.

Differential item functioning (DIF) comparing various sub-groups was performed using the STATA DIF detect command [25]. This employs ordinal regression models with item score as the dependent variable and the relevant domain score and group membership as independent variables. We used the recommended criterion of a $>10\%$ change in the domain score coefficient when group is added to the model to identify uniform DIF [25]. Non-uniform DIF is identified when the group membership $\times$ domain score interaction coefficient differs significantly from zero at the $P<0.05$ level. In addition to groupings based on ability to walk and to talk, we evaluated DIF by age group at the time of the questionnaire (younger than 12 years vs. 12 years and older).

After reverse coding of relevant items, item scores were transformed to a range of 0–100. Specifically, never was scored as 0, rarely as 25, sometimes as 50, often as 75 and very often as 100. Domain scores were calculated by the sum of item scores divided by the number of items. The total score was calculated by the sum of domain scores divided by the number of domains. Linear regression models were then used to examine the associations between total and domain scores and diagnostic (Down syndrome, Rett syndrome, cerebral palsy, ASD) and age (5–11, 12–18 years) groups. Analysis was restricted to questionnaires with a response to all items for either total or domain scores.

Results

Exploratory and confirmatory factor analyses

Exploratory factor analysis of the 41 items resulted in the extraction of six domains. The factor loadings were <0.4 for seven items and these items were excluded (Table 2). Subsequent confirmatory factor analysis affirmed the same factor structure, but the factor loadings for two items were <0.4 and these items were then excluded (Table 3). The remaining items loaded strongly on domains describing "social interaction" ($n=7$), "negative emotions" ($n=7$), "leisure and the outdoors" ($n=5$), "independence" ($n=5$), "physical health" ($n=4$) and "positive emotions" ($n=4$).

Table 1 Frequency distribution (%) for children in the validation study

	All ($n=253$)	Rett syndrome ($n=66$)	Cerebral palsy ($n=64$)	Down syndrome ($n=61$)	Autism spectrum disorder ($n=62$)
Age (years)					
5–11	115 (45.4)	34 (51.5)	25 (39.7)	21 (34.4)	34 (54.8)
12–18	138 (54.5)	32 (48.5)	38 (60.3)	40 (65.6)	28 (45.2)
Sex (female)	157 (62.1)	66 (100.0)	35 (54.7)	33 (54.1)	23 (37.1)
Verbal communication					
Speaks well and understood	70 (27.7)	1 (1.5)	4 (6.3)	32 (52.4)	33 (53.3)
Difficulty in speech or does not use speech	183 (72.3)	65 (98.5)	60 (93.8)	29 (47.5)	29 (46.8)
Eating					
Feeds self, including finger feeding	148 (58.6)	18 (27.3)	10 (15.6)	60 (98.4)	60 (96.8)
Needs to be fed	62 (24.5)	36 (54.6)	23 (35.9)	1 (1.6)	2 (3.2)
Enterally fed	43 (17.0)	12 (18.2)	31 (48.4)	0 (0.0)	0 (0.0)
Personal needs					
Can look after his/her personal needs or needs checking and reminding	58 (23.1)	0 (0.0)	1 (1.6)	29 (48.3)	28 (45.2)
Is provided with assistance but helps, or is dependent on other persons	194 (77)	66 (100)	63 (98.5)	31 (51.7)	34 (54.8)
Mobility					
Walks independently	148 (58.5)	26 (39.4)	1 (1.6)	60 (98.4)	61 (98.4)
Walks with assistance or unable to walk	105 (41.5)	40 (60.6)	63 (98.5)	1 (1.6)	1 (1.6)
Use of hands					
Manages day-to-day activities involving hands	71 (28.0)	0 (0.0)	1 (1.6)	34 (55.8)	36 (58)
Can pick up objects or pieces of food	114 (45.1)	30 (45.4)	31 (48.4)	27 (44.3%)	26 (41.9)
Unable to pick up objects	68 (26.9)	36 (54.6)	32 (50.0)	0 (0.0)	0 (0.0)
Comorbidities					
Vision problems	89 (35.2)	8 (12.1)	33 (51.6)	40 (65.6)	8 (12.9)
Hearing problems	43 (17.0)	2 (3.0)	15 (23.4)	21 (34.4)	5 (8.1)
Epilepsy	90 (35.6)	47 (71.2)	36 (56.3)	1 (1.6)	6 (9.7)
Respiratory infections	60 (23.7)	12 (18.2)	22 (34.4)	19 (31.2)	7 (11.3)
Poor bone health	31 (12.3)	21 (31.8)	10 (15.6)	0 (0.0)	0 (0.0)
Scoliosis	71 (28.1)	38 (57.6)	31 (48.4)	2 (3.3)	0 (0.0)
Hip pain	29 (11.5)	5 (7.6)	24 (37.5)	0 (0.0)	0 (0.0)
Other	68 (26.9)	16 (24.2)	15 (23.4)	19 (31.2)	18 (29.0)

Reliability, convergent and divergent validity

The inter-factor correlations were of moderate size with coefficients ranging in magnitude between 0.20 and 0.68 (Supplementary Table 1). The six-factor model showed satisfactory indices of relative fit using the CMIN/df and Root Mean Squared Error of Approximation values, although the Comparative Fit and Tucker–Lewis indices were slightly smaller than the recommended cut-point of 0.9 (Supplementary Table 2). Factor loadings (Table 3), correlation coefficient values and goodness-of-fit statistics (Supplementary Table 2) were similar for each of the mobility and communication sub-groups indicating consistency of responses to the questionnaire across different levels of functioning.

Cronbach's alpha values ranged from 0.72 for "physical health" to 0.90 for "positive emotions" and composite reliability values ranged from 0.75 for "physical health" to 0.91 for "positive emotions", each >0.7 and indicative of satisfactory convergent validity (Supplementary Table 3). The average variance extracted values for the "physical health" and "negative emotions" domains were <0.5 giving conflicting evidence for the convergent validity of these domains. For each domain, the average variance extracted values were larger than the maximum correlation squared value, providing evidence for satisfactory divergent validity (Supplementary Table 3).

Table 2 Factor loadings for individual scale items onto each of the six domains from the exploratory factor analysis ($n = 253$)

Item[a]	Social interaction	Positive emotions	Physical health	Negative emotions	Leisure and the outdoors	Independence
Expressed happiness when understood	.545					
Appeared relaxed when making eye contact	.472					
Initiated greetings with people verbally	.605					
Enjoyed being included	.838					
Enjoyed social experiences of mealtimes	.740					
Responded positively when others paid attention to them	.675					
Showed pleasure or excitement when looking forward to activities	.495					
Been in a good mood		.412				
Smiled or brightened their facial expression		.864				
Showed happiness through body language		.768				
Showed cheeky or comical mannerisms		.825				
Had enough energy to participate in routines and activities			.646			
Kept in good general health			.596			
Slept well through the night			.490			
Been alert and aware during the day			.683			
Showed that they are in pain				.416		
Been unsettled without apparent reason				.752		
Showed aggression				.620		
Appeared upset or angry				.873		
Become withdrawn with a low mood				.557		
Deliberately hurt themselves				.507		
Expressed discomfort with changes in routine				.560		
Showed signs of being anxious or agitated				.690		
Enjoyed moving their body					.665	
Enjoyed feeling steady or stable during physical activities					.442	
Enjoyed physical activities					.777	
Enjoyed going on outings in the community					.426	
Enjoyed spending time outdoors					.674	
Expressed their needs						.621
Made their own choices for activities or things they enjoy						.698
Expressed discomfort when not given enough time to complete tasks						.626
Helped to complete routine activities						.702
Enjoyed making things with their hands—can be with help						.432
Enjoyed using technology						.579

[a]Deleted items with factor loading < 0.4: Appeared comfortable or relaxed. Responded to being calmed when uncomfortable or upset. Was willing to do as asked. Appeared uncomfortable with sounds, lights, etc. Enjoyed TV programs, movies, reading or music. Enjoyed eating their favourite foods. Showed an interest in contact with animals

Differential item functioning (DIF)

Uniform DIF was displayed for only one item—"Enjoyed making things with their hands" with higher scores (mean = 3.2) among those who were able to walk compared with those unable to walk (mean = 2.6). There were five instances of non-uniform DIF among the three sets of group comparisons on each of the 32 items (Supplementary Table 4). Taking into account multiple testing, this number of significant results is no greater than would be expected by chance.

Comparison of known groups

Descriptive statistics describing QOL total and domain scores are shown in Supplementary Table 5. The mean

Table 3 Factor loading (95% confidence interval) values from confirmatory factor analyses for all children and sub-groups based on capacity to walk or talk

Factor	Item[a]	All children ($n=253$)	Able to walk independently ($n=148$)	Unable to walk independently ($n=105$)	Able to speak ($n=70$)	Unable to speak ($n=183$)
Social interaction	Expressed happiness when understood	.733 (.663, .803)	.711 (.613, .810)	.765 (.669, .862)	.718 (.590, .846)	.746 (.666, .826)
	Appeared relaxed when making eye contact	.702 (.628, .776)	.679 (.577, .782)	.734 (.630, .838)	.772 (.661, .882)	.690 (.601, .780)
	Initiated greetings with people verbally	.619 (.534, .704)	.628 (.518, .739)	.601 (.467, .735)	.471 (.279, .664)	.653 (.559, .748)
	Enjoyed being included	.722 (.654, .791)	.720 (.630, .810)	.739 (.636, .841)	.751 (.639, .862)	.702 (.617, .788)
	Enjoyed social experiences of mealtimes	.646 (.564, .728)	.637 (.528, .747)	.663 (.542, .783)	.629 (.476, .782)	.646 (.549, .743)
	Responded positively when others paid attention to them	.739 (.673, .805)	.738 (.651, .825)	.761 (.666, .856)	.777 (.673, .881)	.734 (.655, .813)
	Showed pleasure or excitement when looking forward to activities	.756 (.689, .824)	.770 (.686, .855)	.749 (.644, .853)	.799 (.698, .899)	.745 (.661, .829)
Positive emotions	Been in a good mood	.756 (.682, .830)	.855 (.785, .925)	.622 (.481, .764)	.856 (.768, .943)	.738 (.644, .832)
	Smiled or brightened their facial expression	.850 (.804, .896)	.847 (.791, .902)	.860 (.789, .931)	.859 (.787, .932)	.845 (.788, .901)
	Showed happiness through body language	.935 (.898, .973)	.931 (.883, .978)	.934 (.874, .993)	.868 (.782, .954)	.951 (.907, .996)
	Showed cheeky or comical mannerisms	.813 (.762, .864)	.812 (.749, .874)	.828 (.748, .907)	.816 (.728, .903)	.800 (.735, .864)
Physical health	Had enough energy to participate in routines and activities	.770 (.700, .841)	.791 (.705, .877)	.774 (.664, .885)	.859 (.763, .954)	.746 (.654, .837)
	Kept in good general health	.550 (.449, .651)	.537 (.404, .669)	.588 (.436, .741)	.574 (.398, .750)	.550 (.427, .673)
	Slept well through the night	.493 (.385, .601)	.483 (.341, .625)	.473 (.299, .647)	.605 (.434, .775)	.451 (.317, .585)
	Been alert and aware during the day	.781 (.711, .851)	.780 (.692, .868)	.784 (.671, .897)	.823 (.716, .930)	.758 (.668, .849)

Table 3 (continued)

Factor	Item[a]	All children ($n=253$)	Able to walk independently ($n=148$)	Unable to walk independently ($n=105$)	Able to speak ($n=70$)	Unable to speak ($n=183$)
Negative emotions	Been unsettled without apparent reason	.730 (.662, .798)	.729 (.639, .818)	.778 (.683, .872)	.713 (.581, .845)	.769 (.697, .842)
	Showed aggression	.521 (.423, .618)	.642 (.537, .747)	.364 (.188, .540)	.639 (.486, .792)	.481 (.360, .602)
	Appeared upset or angry	.819 (.760, .877)	.772 (.689, .854)	.884 (.806, .962)	.768 (.647, .888)	.834 (.767, .900)
	Become withdrawn with a low mood	.730 (.656, .805)	.748 (.658, .838)	.683 (.550, .817)	.809 (.702, .915)	.701 (.607, .796)
	Deliberately hurt themselves	.540 (.444, .635)	.598 (.484, .712)	.453 (.291, .616)	.561 (.388, .734)	.538 (.425, .650)
	Expressed discomfort with changes in routine	.586 (.497, .767)	.615 (.504, .726)	.563 (.419, .707)	.661 (.516, .806)	.563 (.453, .672)
	Showed signs of being anxious or agitated	.719 (.651, .787)	.756 (.676, .837)	.686 (.572, .799)	.789 (.686, .892)	.691 (.606, .777)
Leisure and the outdoors	Enjoyed moving their body	.849 (.795, .902)	.855 (.785, .924)	.840 (.756, .925)	.926 (.864, .989)	.826 (.757, .895)
	Enjoyed feeling steady or stable during physical activities	.783 (.721, .846)	.724 (.630, .819)	.779 (.681, .878)	.775 (.667, .883)	.776 (.699, .853)
	Enjoyed physical activities	.755 (.695, .816)	.790 (.716, .864)	.758 (.665, .850)	.838 (.758, .917)	.744 (.670, .819)
	Enjoyed going on outings in the community	.786 (.705, .866)	.695 (.575, .816)	.864 (.757, .970)	.818 (.702, .933)	.752 (.648, .856)
	Enjoyed spending time outdoors	.553 (.464, .643)	.619 (.510, .729)	.561 (.425, .697)	.643 (.499, .788)	.556 (.449, .663)
Independence	Expressed their needs	.785 (.715, .855)	.707 (.587, .827)	.742 (.617, .867)	.475 (.263, .687)	.773 (.683, .862)
	Made their own choices for activities or things they enjoy	.815 (.748, .882)	.853 (.746, .960)	.719 (.589, .848)	.874 (.724, 1.024)	.766 (.675, .857)
	Helped to complete routine activities	.520 (.425, .616)	.483 (.345, .621)	.310 (.133, .487)	.259 (.023, .496)	.465 (.348, .582)
	Enjoyed making things with their hands—can be with help	.854 (.766, .942)	.887 (.759, 1.016)	.800 (.648, .953)	.884 (.708, 1.059)	.818 (.701, .934)
	Enjoyed using technology	.476 (.377, .576)	.405 (.267, .544)	.482 (.327, .637)	.428 (.238, .618)	.406 (.279, .532)

[a]Two items were deleted with factor loadings < 0.4 on any domain: Showed that they are in pain. Expressed discomfort when not given enough time to complete tasks

(SD) total score for all children was 67.9 (14.3) out of a maximum total score of 100 and mean domain scores ranged from 60.4 (24.0) for "independence" to 74.1 (18.6) for "positive emotions". Low and high scores were obtained for total and domain scores across the diagnostic groups. Comparisons of total and factor scores between diagnostic, functional and age groups are shown in Table 4. Compared to Rett syndrome, children with Down syndrome had higher total (coefficient 10.55, 95% confidence interval [CI] 5.70, 15.39), "social interaction" (coefficient 7.13, 95% CI 0.61, 13.65), "physical health" (coefficient 9.10, 95% CI 2.42, 15.78), "leisure and the outdoors" (coefficient 10.6, 95% CI 3.36, 17.83) and "independence" (coefficient 29.70, 95% CI 22.88, 36.52) scores. Compared to Rett syndrome, children

Table 4 Linear regression of the relationships between total and factor scores and predictor variables

	Total score		Social interaction		Positive emotions		Physical health		Negative emotions		Leisure and the outdoors		Independence	
	Coefficient (95% CI)	*P* value	Coefficient (95% CI)	*P* value	Coefficient (95% CI)	*P* value	Coefficient (95% CI)	*P* value	Coefficient (95% CI)	*P* value	Coefficient (95% CI)	*P* value	Coefficient (95% CI)	*P* value
Diagnosis														
Rett syndrome ($n=66$)	Ref		Ref		Ref		Ref		Ref		Ref		Ref	
Cerebral palsy ($n=64$)	−0.67 (−5.45, 4.12)	0.784	−6.52 (−12.96, −0.08)	0.047	0.67 (−5.64, 6.98)	0.834	2.67 (−3.95, 9.29)	0.428	2.94 (−3.47, 9.34)	0.367	−1.54 (−8.71, 5.64)	0.673	−3.38 (−10.12, 3.35)	0.323
Down syndrome ($n=61$)	10.55 (5.70, 15.39)	<0.001	7.13 (0.61, 13.65)	0.032	4.78 (−1.59, 11.14)	0.140	9.10 (2.42, 15.78)	0.008	1.87 (−4.56, 8.30)	0.57	10.60 (3.36, 17.83)	0.004	29.70 (22.88, 36.52)	<0.001
ASD ($n=62$)	−0.02 (−4.82, 4.78)	0.993	−12.81 (−19.28, −6.35)	<0.001	−7.09 (−13.42, −0.75)	0.028	4.12 (−2.53, 10.77)	0.224	−8.07 (−14.47, −1.67)	0.014	0.27 (−6.97, 7.50)	0.942	23.31 (16.55, 30.07)	<0.001
Walking														
No independent walking ($n=105$)	Ref		Ref		Ref		Ref		Ref		Ref		Ref	
Independent walking ($n=148$)	5.42 (1.82, 9.03)	0.003	2.17 (−2.85, 7.18)	0.395	0.01 (−4.67, 4.70)	0.995	5.99 (1.18, 10.79)	0.015	−4.19 (−8.91, 0.53)	0.081	7.98 (2.73, 13.23)	0.003	23.53 (18.22, 28.85)	<0.001
Communication														
No or poor speech ($n=183$)	Ref		Ref		Ref		Ref		Ref		Ref		Ref	
Able to speak ($n=70$)	6.30 (2.37, 10.23)	0.002	5.46 (−0.01, 10.93)	0.051	1.48 (−3.67, 6.63)	0.571	4.53 (−0.79, 9.84)	0.095	−1.41 (−6.61, 3.78)	0.593	4.29 (−1.53, 10.11)	0.148	25.72 (19.85, 31.58)	<0.001
Age group														
5–11 years ($n=115$)	Ref		Ref		Ref		Ref		Ref		Ref		Ref	
12–18 years ($n=138$)	−2.04 (−5.64, 1.56)	0.266	0.84 (−4.13, 5.81)	0.739	−6.14 (−10.71, −1.56)	0.009	−2.86 (−7.66, 1.94)	0.241	0.26 (−4.43, 4.94)	0.915	−5.36 (−10.59, −0.13)	0.045	−0.24 (−6.24, 5.77)	0.938

with ASD had lower scores for "social interaction" (coefficient − 12.81, 95% CI − 19.28, − 6.35) but higher scores for "independence" (coefficient 23.31, 95% CI 16.55, 30.07). Children who could walk independently or talk had slightly higher "physical health" and "independence" scores than if unable to walk or talk. Children able to walk independently had higher "leisure and the outdoors" scores, and children able to speak had higher "social interaction" scores. Scores for the "positive emotions" (coefficient − 6.14, 95% CI − 10.71, − 1.56) and "leisure and the outdoors" (coefficient − 5.36, 95% CI − 10.59, − 0.13) domains were lower for adolescents compared with children (Table 4).

The final item set is shown in Supplementary Table 6.

Discussion

Our recently identified QOL domains and domain elements as observed in children with intellectual disability formed the foundation for the development of QI-Disability, some that were not well represented in available generic QOL measures. These data indicated the need for a measure developed specifically for children with intellectual disability where options are currently extremely limited. Derived from qualitative data, the items in QI-Disability described caregiver observations of behaviours rather than their impression of what was important for the child's QOL, and were constructed to describe QOL rather than functioning to ensure measurement was broader than health-related QOL [26]. Prior to pilot testing, families then informed the final selection of items for QI-Disability and evaluated their wording for clarity and appropriateness. These processes contrast with the development of KidsLife, also a proxy-report measure of QOL for children with intellectual disability, where items were based on QOL domains for adults with intellectual disability and the judging of experts used to determine their relevance to children [27]. Caregivers of individuals with intellectual disability completed the Pediatric Quality of Life Inventory [28] but scores are difficult to interpret because items do not represent all relevant QOL domains [13–16]. Best practice methodologies [29] in the current study explain the intrinsic validity of QI-Disability.

Factor analyses streamlined the item set and consolidated the qualitative themes into six domains. In broad terms, the domains have conceptual validity because they represent aspects of physical and mental wellbeing, social and recreational functioning illustrated in other child QOL measures [9] and are consistent with the ICF structure [7]. More specifically, items describing QOL in relation to physical health, and positive and negative emotions were extracted from qualitative data representing those domains. Otherwise, items from different qualitative themes were grouped to form the domains "social interaction", "leisure and the outdoors" and "independence". However, these groupings also made conceptual sense. For example, items describing communication experiences in social settings loaded together to represent the child's social interactions. Items describing the pleasures of movement and balance loaded with items describing a range of leisure activities and spending time in the natural environment, providing a comprehensive picture of aspects of participation. The factor "independence" comprised of items necessary for day-to-day communications, routines and everyday tasks in daily living.

The diagnoses of the children together represented the range of health and functioning issues that are observed in children with intellectual disability and a wide range of scores were calculated across each of the domains and within each diagnostic group. In our sample, children with Rett syndrome or severe cerebral palsy were more likely to experience comorbidities such as epilepsy and scoliosis [30, 31], whereas most children with Down syndrome or ASD could walk independently and feed themselves [32, 33]. In this diverse group, some goodness-of-fit analyses were slightly lower than recommended but taken together, the model appears satisfactory. Statistics indicating convergent and divergent validity were also satisfactory, except the average variance extracted values were slightly lower than the recommended cut-point for two of the six domains. There was only one instance of uniform differential item functioning when evaluating each of the 32 item responses by three different sub-groupings of our sample. When replicating factor analyses and validity testing across different levels of communicative and mobility functioning, the validation held. These findings suggest that QI-Disability will be useful across diverse groups of children with intellectual disability and within different groups who experience different impairments and severity.

Variation in QI-Disability factor scores for the diagnostic and age groups was consistent with known between-group heterogeneity and conceptually in alignment with the difficulties experienced. For example, children with Down syndrome had significantly higher total scores than those with Rett syndrome. With regard to specific domains, individuals with Down syndrome where disability is milder scored higher for the "independence" factor [32] than individuals with Rett syndrome who are dependent for most activities of daily living [34]. Alternatively and consistent with other literature [35], scores for "social interaction" were higher for children with Down syndrome who often have a more sociable nature in contrast to children with ASD who experience social difficulties.

Adolescents scored lower for "positive emotions" compared to the younger children, possibly reflecting changes experienced by adolescents in the general population [36] or emotional disorders as reported in adolescents with intellectual disability in a national survey in the United Kingdom

[37]. Interestingly, "leisure and the outdoors" scores were also lower for adolescents, perhaps consistent with lower "positive emotions" scores or with encountering barriers to participation such as issues of access, limited opportunities or attitudes of others of discrimination or exclusion. These data suggest that important differences are identifiable, and some point to opportunities for interventions to increase QOL in adolescents with intellectual disability.

Pilot testing of QI-Disability involved a sample derived from population-based databases representing a range of child and family characteristics. Rett syndrome is caused by a pathogenic mutation in the *MECP2* gene located on Xq28 and almost exclusively affects females [38] and so our sample was entirely female. The gender distribution in the other diagnostic groups was broadly consistent with the literature with some differences. For example, autism is more prevalent in males [39] as reflected in our sample. Epidemiological studies of children with cerebral palsy and comorbid intellectual disability [40] and those with Down syndrome [41] indicate a slightly higher prevalence in males, whereas our samples include slightly more females. Our sample enabled factor analysis and related validation but it will be important to ensure representativeness in future studies when investigating the determinants of QOL. Within each diagnostic group there was a range of strengths and difficulties as seen in clinical care. There were high recruitment fractions with little missing data. We acknowledge that QI-Disability collects proxy-reported data and that there may be differences between parent and child reports [17]. Whilst self-report is preferable where feasible, there is still substantial reliance on parent/proxy reports in the paediatric literature and practice, and particularly in the field of intellectual disability. For intellectual disability, it is necessary to develop a proxy-report measure of QOL that would enable population-based investigations and include the substantial proportion of children unable to self-report. Importantly, the development of QI-Disability is a vital step in the preparation for the development of a child-report measure based on child-reported domains of QOL and appropriate for children with communication difficulties who can self-report.

Conclusion

More validation studies will be an area for future research [42], but the developmental processes, theoretical underpinnings and psychometric testing provide evidence that QI-Disability can be used as an outcome measure to support evaluation in children with intellectual disability. With complex needs, multifaceted outcome measures such as QI-Disability are necessary to assess practice and enable new lines of inquiry on the determinants of QOL and novel interventions.

Acknowledgements We thank the families for their participation in this study. We acknowledge the database infrastructures necessary for the current study. The authors thank the Disability Services Commission WA in establishing the Down syndrome database, and community organisations Developmental Disability WA and the Down Syndrome Association of Western Australia for their support. We thank the Australian Paediatric Surveillance Unit (APSU) and the Rett Syndrome Association of Australia for their ongoing support in case ascertainment for the Australian Rett Syndrome Database. The Australian Rett Syndrome Study was previously supported by the National Institute of Health Grant 5R01HD043100-05 and National Health and Medical Research Council project Grants #303189 and #1004384. The Victorian Cerebral Palsy Register receives funding from the Victorian Department of Health and Human Services and from the Victorian Government's Operational Infrastructure Support Program for support for register staff. We acknowledge the support of Dr Sue Reid in the recruitment of families with a child with cerebral palsy and Dr Emma Glasson who supported the recruitment of families from the WA Autism Registry. This study is currently funded by the National Health and Medical Research Council (#1103745). ED was previously supported by a Victorian Health Promotion Public Health Fellowship. AW and HL are each supported by a Senior Research Fellowship from the National Health and Medical Research Council (#1077966 and #1117105, respectively). KW in funded by the Apex Foundation for Research into Intellectual Disability and the Royal Children's Hospital Foundation. DR is funded by a University of Melbourne Award. The funders of this research have had no roles in the study design, data collection, data analysis, manuscript preparation and/or publication decisions.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Leonard, H., Petterson, B., Bower, C., & Sanders, R. (2003). Prevalence of intellectual disability in Western Australia. *Paediatric and Perinatal Epidemiology, 17*(1), 58–67.
2. Maulik, P. K., Mascarenhas, M. N., Mathers, C. D., Dua, T., & Saxena, S. (2011). Prevalence of intellectual disability: A meta-analysis of population-based studies. *Research in Developmental Disabilities, 32*(2), 419–436.
3. Bigby, C. (2012). Social inclusion and people with intellectual disability and challenging behaviour: A systematic review. *Journal of Intellectual & Developmental Disability, 37*(4), 360–374.
4. Leonard, H., Glasson, E., Nassar, N., Whitehouse, A., Bebbington, A., Bourke, J., et al. (2011). Autism and intellectual disability are differentially related to sociodemographic background at birth. *PLoS ONE, 6*(3), e17875–e17875.
5. Verdugo, M. A., Schalock, R. L., Keith, K. D., & Stancliffe, R. J. (2005). Quality of life and its measurement: Important principles and guidelines. *JIDR. Journal of Intellectual Disability Research, 49*(10), 707–717.
6. Stewart, M. (2001). Towards a global definition of patient centred care. *BMJ: British Medical Journal, 322*(7284), 444–445.
7. World Health Organization. (2001). *International classification of functioning, disability and health: ICF*. Geneva: World Health Organisation.
8. Terwee, C. B., Bot, S. D., de Boer, M. R., van der Windt, D. A., Knol, D. L., Dekker, J., et al. (2007). Quality criteria were

proposed for measurement properties of health status questionnaires. *Journal of Clinical Epidemiology, 60*(1), 34–42.
9. Solans, M., Pane, S., Estrada, M. D., Serra-Sutton, V., Berra, S., Herdman, M., et al. (2008). Health-related quality of life measurement in children and adolescents: A systematic review of generic and disease-specific instruments. *Value in Health, 11*(4), 742–764.
10. Young, B., Rice, H., Dixon-Woods, M., Colver, A. F., & Parkinson, K. N. (2007). A qualitative study of the health-related quality of life of disabled children. *Developmental Medicine & Child Neurology, 49*(9), 660–665.
11. Tavernor, L., Barron, E., Rodgers, J., & McConachie, H. (2013). Finding out what matters: Validity of quality of life measurement in young people with ASD. *Child: Care, Health & Development, 39*(4), 592–601.
12. Neul, J. L., Kaufmann, W. E., Glaze, D. G., Christodoulou, J., Clarke, A. J., Bahi-Buisson, N., et al. (2010). Rett syndrome: Revised diagnostic criteria and nomenclature. *Annals of Neurology, 68*(6), 944–950.
13. Davis, E., Reddihough, D., Murphy, N., Epstein, A., Reid, S. M., Whitehouse, A., et al. (2017). Exploring quality of life of children with cerebral palsy and intellectual disability: What are the important domains of life? *Child: Care, Health and Development*. https://doi.org/10.1111/cch.12501.
14. Epstein, A., Leonard, H., Davis, E., Williams, K., Reddihough, D., Murphy, N., et al. (2016). Conceptualizing a quality of life framework for girls with Rett syndrome using qualitative methods. *American Journal of Medical Genetics Part A, 170A*, 645–653.
15. Epstein, A., Whitehouse, A., Williams, K., Murphy, N., Leonard, H., Davis, E., et al. (2017). Parent-observed thematic data on quality of life in children with autism spectrum disorder. *Autism*. https://doi.org/10.1177/1362361317722764.
16. Murphy, N., Epstein, A., Leonard, H., Davis, E., Reddihough, D., Whitehouse, A., et al. (2017). Qualitative analysis of parental observations on quality of life in Australian children with Down syndrome. *Journal of Developmental and Behavioral Pediatrics, 38*(2), 161–168.
17. Davis, E., Nicolas, C., Waters, E., Cook, K., Gibbs, L., Gosch, A., et al. (2007). Parent-proxy and child self-reported health-related quality of life: Using qualitative methods to explain the discordance. *Quality of Life Research, 16*(5), 863–871.
18. Bourke, J., Ricciardo, B., Bebbington, A., Aiberti, K., Jacoby, P., Dyke, P., et al. (2008). Physical and mental health in mothers of children with Down syndrome. *The Journal of Pediatrics, 153*(3), 320–326.
19. Downs, J., Torode, I., Wong, K., Ellaway, C., Elliott, E. J., Christodoulou, J., et al. (2016). The natural history of scoliosis in females with Rett syndrome. *Spine (Phila Pa 1976), 41*(10), 856–863.
20. Reid, S. M., Meehan, E., McIntyre, S., Goldsmith, S., Badawi, N., & Reddihough, D. S. (2016). Temporal trends in cerebral palsy by impairment severity and birth gestation. *Developmental Medicine & Child Neurology, 58*(Supplement 2), 25–35.
21. Taylor, L. J., Maybery, M. T., Wray, J., Ravine, D., Hunt, A., & Whitehouse, A. J. (2013). Brief report: Do the nature of communication impairments in autism spectrum disorders relate to the broader autism phenotype in parents? *Journal of Autism and Developmental Disorders 43*(12), 2984–2989.
22. Davis, E., Waters, E., Mackinnon, A., Reddihough, D., Graham, H. K., Mehmet-Radji, O., et al. (2006). Paediatric quality of life instruments: A review of the impact of the conceptual framework on outcomes. *Developmental Medicine & Child Neurology, 48*(4), 311–318.
23. Baars, R. M., Atherton, C. I., Koopman, H. M., Bullinger, M., & Power, M., & DISABKIDS GROUP. (2005). The European DISABKIDS project: Development of seven condition-specific modules to measure health related quality of life in children and adolescents. *Health and Quality of Life Outcomes*. https://doi.org/10.1186/1477-7525-3-70.
24. Streiner, D. L. (1994). Figuring out factors: The use and misuse of factor analysis. *Multivariate Behavioral Research, 39*, 135–140.
25. Crane, P., Gibbons, L., Jolley, L., & van Belle, G. (2006). Differential item functioning analysis with ordinal logistic regression techniques. *Medical Care, 44*(11), S115–S123.
26. Rosenbaum, P., Paneth, N., Leviton, A., Goldstein, M., & Bax, M. (2007). A report: The definition and classification of cerebral palsy April 2006. *Developmental Medicine & Child Neurology, 109*, 8–14.
27. Gomez, L. E., Alcedo, M. A., Arias, B., Fontanil, Y., Arias, V. B., Monsalve, A., et al. (2016). A new scale for the measurement of quality of life in children with intellectual disability. *Research in Developmental Disabilities, 53*, 399–410.
28. Viecili, M. A., & Weiss, J. A. (2015). Reliability and validity of the pediatric quality of life inventory with individuals with intellectual and developmental disabilities. *American Journal on Intellectual and Developmental Disabilities, 120*(4), 289–301.
29. Waters, E., Davis, E., Ronen, G. M., Rosenbaum, P., Livingston, M., & Saigal, S. (2009). Quality of life instruments for children and adolescents with neurodisabilities: How to choose the appropriate instrument. *Developmental Medicine & Child Neurology, 51*, 660–669. https://doi.org/10.1111/j.1469-8749.2009.03324.x.
30. Colver, A., Fairhurst, C., & Pharoah, P. O. (2014). Cerebral palsy. *Lancet, 383*, 1240–1249.
31. Leonard, H., Cobb, S., & Downs, J. (2017). Clinical and biological progress over 50 years in Rett syndrome. *Nature Reviews Neurology, 13*(1), 37–51.
32. Lin, H. Y., Chuang, C. K., Chen, Y. J., Tu, R. Y., Chen, M. R., Niu, D. M., et al. (2016). Functional independence of Taiwanese children with Down syndrome. *Developmental Medicine & Child Neurology, 58*(5), 502–507.
33. Ming, X., Brimacombe, M., & Wagner, G. C. (2007). Prevalence of motor impairment in autism spectrum disorders. *Brain and Development, 29*(9), 565–570.
34. Leonard, H., Fyfe, S., Leonard, S., & Msall, M. (2001). Functional status, medical impairments, and rehabilitation resources in 84 females with Rett syndrome: A snapshot across the world from the parental perspective. *Disability and Rehabilitation, 23*(3–4), 107–117.
35. Moss, J., Nelson, L., Powis, L., Waite, J., Richards, C., & Oliver, C. (2016). A Comparative study of sociability in Angelman, Cornelia de Lange, Fragile X, Down and Rubinstein Taybi Syndromes and Autism Spectrum Disorder. *American Journal on Intellectual and Developmental Disabilities, 121*(6), 465–486.
36. Sawyer, S. M., Afifi, R. A., Bearinger, L. H., Blakemore, S. J., Dick, B., Ezeh, A. C., et al. (2012). Adolescence: A foundation for future health. *Lancet, 379*(9826), 1630–1640.
37. Emerson, E., & Hatton, C. (2007). Mental health of children and adolescents with intellectual disabilities in Britain. *The British Journal of Psychiatry, 191*(6), 493.
38. Amir, R. E., Van den Veyver, I. B., Wan, M., Tran, C. Q., Francke, U., & Zoghbi, H. Y. (1999). Rett syndrome is caused by mutations in X-linked MECP2, encoding methyl-CpG-binding protein 2. *Nature Genetics, 23*(2), 185–188.
39. Lai, M. C., Lombardo, M. V., & Baron-Cohen, S. (2014). Autism. *Lancet, 383*(9920), 896–910.
40. Reid, S. M., Meehan, E. M., Arnup, S. J., & Reddihough, D. S. (2018). Intellectual disability in cerebral palsy: A population-based retrospective study. *Developmental Medicine & Child Neurology*. https://doi.org/10.1111/dmcn.13773.
41. Leonard, S., Bower, C., Petterson, B., & Leonard, H. (1999). Medical aspects of school-aged children with Down syndrome. *Developmental Medicine & Child Neurology, 41*(10), 683–688.

42. U.S. Department of Health and Human Services, FDA, Center for Drug Evaluation and Research (CDER), Center for Biologics Evaluation and Research (CBER), & (CDRH)., C. f. D. a. R. H. (2009). Guidance for industry: Patient-reported outcome measures: Use in medical product development to support labeling claims. In U. S. D. o. H. a. H. S. Food and Drug Administration (Ed.). Silver Spring.
43. Hu, L., & Bentler, P. M. (1999). Cutoff criteria for fit indexes in covariance structure analysis: Conventional criteria versus new alternatives. *Structural Equation Modeling: A Multidisciplinary Journal, 6*(1), 1–55.
44. Fornell, C., & Larcker, D. (1981). Evaluating structural equation models with unobservable variables and measurement error. *Journal of Marketing Research, 18*(1), 39–50.