Participation and quality of life

in children and adolescents with congenital limb deficiencies

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Content

Participation and quality of life in children and adolescents with congenital limb deficiencies: a narrative review

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Date:	May 2009

Number of word s: 3462

Abstract: 261

Samenvatting

Doel : Kinderen en jongeren met congenitale reductiedefecten van de extremiteiten zijn zichtbaar en fysiek verschillend van hun leeftijdsgenoten. Ze ervaren, afhankelijk van de ernst en uitgebreidheid van de reductiedefecten, beperkingen in activiteiten. Daardoor lopen zij risico tot verminderde participatie in sociale activiteiten en vrijetijdsbesteding. Dit kan een negatieve invloed hebben op hun zij hun kwaliteit van leven ervaren. Het doel van deze literatuurstudie is het beschrijven van de participatie en kwaliteit van leven van kinderen en jongeren met congenitale reductiedefecten van de extremiteiten. Het psychosociaal functioneren wordt ook beschreven omdat het sterk gerelateerd is aan het concept kwaliteit van leven.

Design : Beschrijvende literatuurstudie.

Methode : Een alomvattende beoordeling van de literatuur werd uitgevoerd op participatie, kwaliteit van leven en psychosociaal functioneren in kinderen en jongeren met congenitale reductiedefecten van de extremiteiten (leeftijd 8 tot 18 jaar). Een systematische zoekstrategie werd gehanteerd in verschillende data bronnen.

Resultaten : Vijftien cross sectionele studies werden geïncludeerd. De literatuur tot op heden biedt weinig kennis over hoe kinderen en jongeren met congenitale reductiedefecten van de extremiteiten participeren en hoe zijn hun kwaliteit van leven ervaren. Het psychosociaal functioneren is vergelijkbaar met gezonde leeftijdsgenoten, ondanks dat het beschreven wordt als een risicofactor bij kinderen met reductiedefecten. Factoren die van invloed kunnen zijn op de participatie en kwaliteit van leven bij kinderen en jongeren met congenitale reductiedefecten van de extremiteiten werden niet gevonden.

Conclusie : Meer onderzoek is nodig naar hoe kinderen en jongeren met congenitale reductiedefecten van de extremiteiten participeren en hoe zij de kwaliteit van hun leven ervaren. Een beter begrip zal professionals in de gezondheidszorg helpen in het bepalen van hun therapeutische interventies. Een breder perspectief op hoe de kinderen en jongeren met congenitale reductiedefecten van de extremiteiten participeren en van het leven genieten zal ouders helpen in het maken van de juiste keuzen voor hun kinderen.

Trefwoorden: *reductiedefecten, participatie, psychosociaal functioneren, kwaliteit van leven*

Abs tract

Purpose : Children and adolescents with congenital limb deficiencies are visibly and physically different from their peers. They present limitations in activities, depending on the severity of deficiency. Therefore they are at risk for lower participation in social and leisure activities. This might negatively influence the perception on their quality of life. The aim of this narrative review is to describe participation and quality of life in children with congenital limb deficiencies. Psychosocial functioning, being closely related to the concept of quality of life is described as well.

Methods : A comprehensive review of the literature was conducted on participation, quality of life and psychosocial functioning in children and adolescents with congenital limb deficiencies (ages 8-18 years). The review involved a systematic search using multiple data sources.

Results : Fifteen cross sectional studies were included in this review. The literature to date provides limited knowledge on how children and adolescents with congenital limb deficiencies participate and how they perceive their quality of life. The psychosocial functioning, although described as at risk, appears to be comparable to healthy peers. Factors influencing the level of participation and quality of life in children and adolescents with limb deficiencies could not be extracted.

Conclusion s : More research is needed on how children and adolescents with congenital limb deficiencies participate and how they perceive their quality of life. A better understanding will help health care professionals in targeting their therapeutic interventions. A broader perspective on how children and adolescents with congenital limb deficiencies are involved in and enjoy life will help parents in making the right choices for their children.

Keywords *: limb d eficien cies, partici pation, ps ych oso cial functionin g, quality of li fe*

Introduction

Limb deficiency disorders are heterogeneous, chronic physical conditions with a prevalence of 5-9 in 10.000 births.[1] Most limb deficiencies in children are congenital in origin; acquired limb deficiencies as a result of disease or physical trauma, are less ordinary.[2]

Almost all children with lower limb deficiencies are fitted with prosthetic components to enhance their ability to participate in activities of daily living. The use of prosthetic devices in children with upper limb deficiencies is less evident, depending on level of deficiency and functional gain.[3]

Participation is defined by the World Health Organization as the nature and extent of a person's involvement in life situations.[4] For children and youth, involvement in life situations includes participation in recreational and leisure activities as well as school and work activities.[4] Regular participation in day-to-day activities is an important aspect of children's health, well-being, and development.[5] In general, children and adolescents with disabilities tend to be more restricted in participation than their peers.[6] The most important factors that influence participation are children's functional abilities, environment (e.g. attitudes of community members), family (e.g. parents interest in recreation and family support) and personal characteristics (e.g. gender and social competence).[7-9]

An emerging concept in the psychological functioning of children with chronic disorders is Health Related Quality of life (HRQoL). Quality of life can refer to aspects of a person's well being (physical, psychological, social), as well as aspects of the environment and a person's standard of living.[10] Psychological well being, self esteem, adjustment and happiness are constructs related to quality of life.[11] Evidence from the literature suggests that adolescents with disabilities are at greater risk for psychosocial maladjustment than adolescents without disabilities.[12;13]

The relationship between participation and quality of life is currently not fully understood. It is suggested that participation is associated with increased quality of life and reduction of health and social problems in children with and without disabilities.[7;8;14] However, lower participation by physical limitations does not automatically imply a decreased quality of life.[15] The abovementioned studies focused on children with more complex disabilities: besides physical limitations, cognitive impairments were present.

Children and adolescents with limb deficiencies have limited physical potential but no cognitive impairments. The visible abnormal appearance might negatively influence their participation and quality of life. To what extent they are involved in and enjoy life situations is unclear.

The main objective of this review is to study and describe the current knowledge on participation and quality of life in children and adolescents with congenital limb deficiencies. Furthermore, the literature was searched for risk and protective factors that might influence participation and quality of life in these children.

Methods

While participation and quality of life are relatively new concepts, it is to be expected that limited research is available. To broaden the search, psychosocial functioning as an outcome was included, being closely related to the concept of quality of life. Studies that assessed participation, quality of life and/or psychosocial functioning in children and adolescents with congenital limb deficiencies were considered eligible for this review of the literature.

Search strategy

A comprehensive search from 1980 to October 2008 for relevant studies was performed in CINAHL, the Cochrane library, EMBASE, HaPi, PEDro, PsychINFO and MEDLINE. The literature search was limited to published studies, available in full text English articles. Mesh, thesaurus and text-based search terms included: quality of life, participation, adolescents, children, limb deficiencies, perceived disabilities, well being, personal satisfaction, physical appearance, adjustment, self esteem, self perception, psychosocial outcome, family, sports and recreation.

Reference lists of studies included and reviews have been hand searched for relevant publications. The complete search and screening of titles and available abstracts was done by the first author (AM). Studies that met the inclusion criteria were summarized in terms of purpose, type and setting of study, study sample and outcome measures. Results of all studies on participation, quality of life and psychosocial outcome were independently assessed by the first author Two independent researchers (IvW, MK) examined the summary of all studies for definite inclusion and exclusion and checked the results. Disagreements were discussed until consensus was reached.

Selection criteria

Studies were included if they met the following inclusion criteria: (1) children and adolescents with congenital limb deficiencies (age 8 -18 years), (2) outcome measures concerning quality of life, participation or psychosocial functioning, (3) publication between 1980 and October 2008.

Exclusion criterion was the combination of limb deficiencies with cognitive impairment. Reviews of the literature were excluded as well .

Results

Search results

A total of 87 possible relevant studies were selected from the search (Figure 1). After screening title and abstract, 64 articles did not match the inclusion criteria. Four reviews were excluded; three[13;16;17] narrative and one systematic review[12].Two studies[18;19] were excluded for not being available full text through the Dutch Library.

After screening seventeen full text studies, another two studies were excluded. Reasons for exclusion were: population age above 18 years[20] and unclear study sample[21]. Reference tracking of eligible articles and present reviews[12;13;16;17] did not provide new studies for inclusion.

The fifteen included studies are all classified as cross sectional descriptive studies focusing on children and adolescents with congenital limb deficiencies with measurement outcome on quality of life, participation or psychosocial functioning.

Table 1 summarizes all studies included. Studies are listed in descending order of publication and by name of the first author.

Table I: Summary of studies

Study	Study Design	Setting	Population	N	Mean age (range in years)	Outcome	Variables (measures)
James et al (2006)[22]	CS	LDC USA	UCBED	489	Unkown (2-20)	QoI Participation	Emotional -, social -, school functioning (PedsQol) Sports/physical function, happiness (PODCI)
Hermansson et al (2005)[37]	CS	LDAPC Sweden	ULRD	62(PR) 37(CR)	12.6 (8-18) 14.8 (11-18)	Psychosocial	Behaviour/emotional problems, social competence (CBCL - PR+YSR-CR) Depressive symptoms (CDI)
Vannah et al (1999)[24]	CS	LDC USA	LLD	258	Unkown (2-21)	Participation	Recreational/sports, days of school missed (PES) (non standardized information)
Herring et al (1999)[23]	CS	Hospital USA	LLD (Syme amputation)	21	11.2 (5-18)	Participation Psychosocial	Participation in organized sports (non standardized information) Self concept (PHSC) Behaviour: social and emotional problems (LBC) Resources + stress in families (QRS)
Varni et al (1996) [36]	CS	CANP USA	LD	44	14.7 (13- 18)	Psychosocial	Perceived physical appearance, general self esteem(SPPA)
Varni et al (1993) [35]	CS	CANP USA	LD	54	10.1 (8-13)	Psychosocial	Depressive symptoms (CDI) Self perceived trait anxiety (STAIC) General self esteem(SPPC)
Varni et al (1992a) [33]	CS	CANP USA	LD	49	10.3 (8-13)	Psychosocial	Perceived social support (SSSC)
Varni et al (1992b) [34]	CS	CANP USA	LD	111	10.4 (6-17)	Psychosocial	Behaviour/emotional problems, social competence (CBCL)
Varni et al (1991a) [31]	CS	CANP USA	LD	51	10.3 (8-13)	Psychosocial	Perceived physical appearance (SPPC)
Varni et al (1991b) [32]	CS	CANP USA	LD	80	10.3 children 14.7 adolescents	Psychosocial	Perceived physical appearance (SPPC+SPPA)
Varni et al (1991c) [30]	CS	CANP USA	LD	54	10.1 (8-13)	Psychosocial	Depressive symptoms (CDI)
Varni et al (1989a) [28]	CS	CANP USA	LD	42	8.4 (6-13)	Psychosocial	Behaviour/emotional problems, social competence (CBCL)
Varni et al (1989b) [29]	CS	CANP USA	LD	41	10.5 (8-13)	Psychosocial	General self esteem (SPPC)
Varni et al (1989c) [27]	CS	CANP USA	LD	27	10.2 (8-13)	Psychosocial	Depressive symptoms (CDI)
Rubenfeld et al (1988) [26]	CS	CANP USA	LD	41	10.5 (8-13)	Psychosocial	General self esteem (SPPC)

Abbreviations: CS=cross sectional; CANP=Child and Adolescent Needs Project; UCBED=Unilateral Congenital Below-the-Elbow Deficiency; ULRD=Upper Limb Reduction Deficiency; LD=Limb Deficiencies; LLD=Lower Limb Deficiency; QoL=Quality of Life; PedsQol=Pediatric Quality of Life Inventory; PODCI=Pediatric Outcome Data Collection Instrument; PES=Prosthesis Evaluation Scale; LDAPC=Limb Deficiency and Arm Prosthesis Centre; LDC=Limb Deficiency Clinics; CBCL=Child Behaviour Checklist; CDI=Children's Depression Inventory; YSR=Youth Self Report; PHSC=Piers-Harris Self concept Scale; LBC=Louisville Behaviour Checklist; QRS=Questionnaire on Resources and Stress; SPPS=Self Perception Profile For Adolescents; Beck Depression Inventory; State-Trait Anxiety Inventory; SSSC=Social support Scale for Children; DAS=Dyadic Adjustment Scale; FRI=Family Relationship Index ; CHS=Children's Hassles Scale.

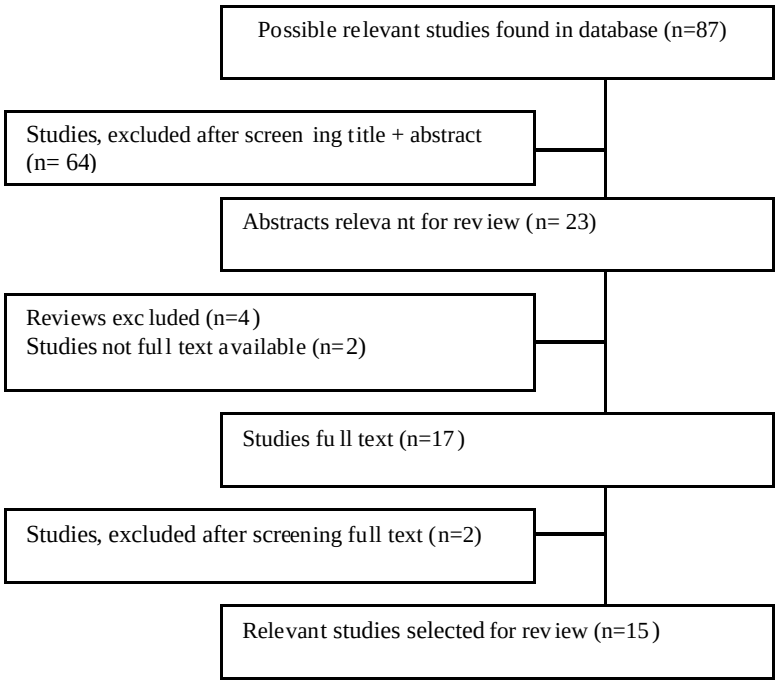

Figure 1 Flow chart of study selection

Evaluation of results

The results of the 15 included studies are described in three different domains representing the primary outcomes of these studies: participation, quality of life and psychosocial functioning.

Participation

No studies specifically report on participation as outcome or used a standardized measure for participation. Aspects of participation are described in three studies[22;23;24]. James et al[22] examined a large group of children and adolescents with Unilateral Congenital Below-the-Elbow Deficiency (UCBED), age range between 2 and 20 years. Parents and children above eleven years (n=184) reported on 'sports/physical function', 'happiness' and 'global function' as a part of a musculoskeletal health questionnaire addressing activity and participation components of function (PODCI)[25]. No significant differences on these domains were found in children with UCBED compared with the general population and between prosthesis wearers and non wearers. No exploration was made on factors related to participation.

Herring et al[23] and Vannah et al[24] reported the frequency of recreational and sports activities in children with Lower Limb Deficiencies (LLD). In the study of Vannah et al[24], more than half of the 258 children were participating in regular sports: swimming 60%, indoor chores 59%, running 58%, outdoor chores 55%, bicycling 52%. Basketball was done by 42% of the children, comparable with 43% of 21 children in the study of Herring[23] who were participating in competitive school athletics. Missing more than ten days of school per year due to their limb deficiency was reported by Vannah et al[24] in 7% of the children.

Quality of life

Assessment of Health Related Quality of Life was done in the study of James et al[22] in which children with UCBED were compared with the general population. Within the group of children with UCBED comparison of prosthesis wearers with non wearers was also made. Both parents and children for those who are five to twenty years of age reported the quality of life of the child: no significant differences were found. When comparing children wearing a prosthesis with children not wearing a prosthesis also no significant differences were found, except for the quality of life with regard to school functioning: significant higher scores were reported for prosthesis wearers compared with non wearers. Eleven to twenty-year-old children with UCBED felt significantly happier than children in the general population, regardless of prosthetic use, as reported in the Happiness domain of the questionnaire addressing activity and participation (PODCI).[25]

Psychosocial functioning

All, but two studies[22;24] focussed on psychosocial adaptation in children and adolescents with congenital limb deficiencies. Eleven studies[26-36] were executed by Varni et al within the Child and Adolescent Needs Project (CANP). They investigated psychological and social adjustment in children with Limb Deficiencies (LD) in order to identify risk and protective factors. The children participating in the CANP project were diagnosed with deficiencies of upper and/or lower limbs of which the majority was congenital. Description of subgroups was not provided. Age range of the children was between 8 and 18 years. Outcomes in the series of studies were perceived physical appearance, self esteem, depressive symptoms, anxiety, perceived social support, behaviour and social competence.

In general, children with LD are not significantly different in how they perceive their physical appearance[32] and social support[27;33], and in their self esteem[29;32] compared with the general population. The children with LD are not more depressed[27;30] and they do not experience a greater number of hassles[27] than physically healthy peers. A comparison of congenital and acquired limb deficiencies, which has been performed in two studies[30;36], showed no significant differences.

Varni et al[34] demonstrated greater behavioural and emotional problems and lower social competence in children with LD than the normative sample. Through parent report 23% of the children in abovementioned study appeared to function in the clinically significant maladjustment range for behavioural and emotional problems; 14% of the children functioned in the clinically significant social maladjustment range. These percentages were compared to the proportion of maladjustment in the general population which is 10% for both behavioural and emotional problems and social competence.

A contrasting result is described in the study by Hermansson et al[37] in which parents and their children with ULRD who had initially been fitted with myoelectric prosthetic hands reported on social competence, emotional or behavioural problems and depressive symptoms. No significant differences compared to normative samples were observed. All children though, irrespective of age or gender, had significant higher scores on the withdrawn behaviour subscale. Girls with ULRD (13-17 years) had lower, although not significant, social competence compared to girls in the normative sample.

To identify risk and protective factors, researchers of the CANP project examined associations of the primary outcome measures. Correlations of study outcomes are presented in Table 2.

None of the demographic factors, age, sex and socio economic status (SES), were significantly associated with the outcome variables, except for higher SES being significantly correlated with lower perceived physical appearance[29] and for sex which was significantly correlated with internalizing behaviour problems[28]. Degree of limb loss was not significant correlated with most of the dependent variables, except for self esteem.[34] In adolescents, degree of limb loss was significantly associated with lower general self esteem, while it was not in children.[34]

Table II: Summary of correlations CANP

Predictor variables (measures)	**Outcome (measures)**						
	Anxiety (STAIC)	Behaviour (CBCL)	Depressive Symptoms (CDI)	Perceived Physical Appearance (SPPA)	Perceived Social Support (SSSC)	Self esteem (SPPC)	Social Competence (CBCL)
Age	-(-)[35]	-(+)[28;34]	-(+)[30;35]	-(-)[31;32;36]	-(-)[33]	-(+)[26;29]	-(-)[34]
Degree of limb loss	-(-)[35]	-(-)[28;34]	-(+)[30;35]	-(-)[31;32;36]	-(+)[33]	+(-)[36] -(+)[26;29]	-(+)[34]
Sex	-(+)[35]	+(-)[28] -(+)[34]	-(-)[30;35]	-(-)[31;32;36]	-(?)[33]		-(-)[34]
SES	-(?)[35]	-(-)[28;34]	-(-)[30;35]	+(-)[31]	-(?)[33]	-(-)[26;29]	-(+)[34]
Anxiety (STAIC)		+(+)[34]	+(+)[35]	+(-)[31;36]	+(-)[33]	+(-)[36]	
Behaviour/social competence (CBCL)	+(-)[35]	+(+)[34]				+(+)[29]	
Depressive symptoms (CDI)		+(+)[34]		++(-)[31]	+(-)[33]		
Depressive symptoms (BDI)	+(+)[35]	+(+)[34]	+(+)[35]	+(-)[36]		+(-)[35;36]	
Emotionality (EAS)		++(+)[28]					++(-)[28]
Family relationship (FRI)	+(-)[35]		+(-)[35]			+(+)[35]	
Family functioning (FES):							
• Cohesion		+(-)[28]				-(+)[29]	+(+)[28]
• Organisation		+(-)[28]				+(+)[26;29]	-(+)[28]
• Moral-religious emphasis		++(-)[28]					+(+)[28]
• Conflict		+(+)[28]				+(-)[26;29]	-(-)[28]
• Intellectual-cultural orientation		+(-)[28]					+(+)[28]
Marital relationships (DAS)	+(+)[35]		+(+)[35]	+(-)[31]		+(-)[35]	
Perceived Appearance(SPPC)							
• General self esteem			++(-)[30]	++(+)[31;36]	+(+)[33]		
• Social competence				+(+)[31]		+(+)[26;29;36]	
• Scholastic competence				++(+)[31]		+(+)[29;36]	
• Athletic competence				+(+)[31]		+(+)[29;36]	
• Close friendship				+(+)[31]		++(+)[36]	
Social support (SSSC):							
• Classmate	+(-)[35]		+++(-)[27;30;35]	+(+)[31]		++(+)[26;29;35]	
• Friends	-(-)[35]		++(-)[27;30;35]	+(+)[31]		+(+)[26;29;35]	
• Parent	+(-)[35]		++(-)[27;30;35]	+(+)[31]		+(+)[26;29;35]	
• Teacher	+(-)[35]		++(-)[27;30;35]	+(+)[31]		+(+)[26;29;35]	
Stress (CHS)			+(+)[27;30]	+(-)[31]		+(-)[29]	

Correlation strength(r):+ = fair (r=0.25 to 0.50); ++=moderate to good (r=0.50 to 0.75); +++=good to excellent (r=.75 to 1); - = no significant correlation. Correlation direction:(+)= positive; (-)=negative; (?)=unknown.
Abbreviations: BDI= Beck Depression Inventory; CBCL=Child Behaviour Checklist; CDI=Children's Depression Inventory; DAS=Dyadic Adjustment Scale; EAS=Emotionality, Activity, Sociability Temperament Survey; FES=Family Environment Scale; FRI=Family Relationship Index; SPPA=Self Perception Profile for Adolescents; SPPC=Self Perception Profile for Children; SSSC=Social Support Scale for Children; STAIC=State-Trait Anxiety Inventory

How children with LD perceived their physical appearance was predictive for their self esteem, level of depression and anxiety.[31;36] A trend was noted toward lower perceived physical appearance in children as they approached adolescence, suggesting a risk status for lower perceived physical appearance in adolescents with LD.[32] A significant correlation was found between perceived class mate social support and teacher social support on perceived physical appearance.[31] Multiple perceived social support domains were found to be statistically significant predictors of psychological adjustment. Higher perceived social support was associated with lower depressive and anxious symptoms and higher self esteem.[33] Especially classmate social support appeared to be highly predictive and was therefore represented as a risk factor for children with LD.[33]

A significant amount of variance in psychological adaptation was explained by familial (parental discord, marital discord, family support, parent social support,) and non-familial (class mate, teacher, friend social support) social environment factors.[35] In general, more family cohesion, moral-religious emphasis and organisation in combination with less family conflict, predicted better psychological and social adaptation.[28]

Significant predictors for behaviour problems in children with LD were family functioning and parental adjustment.[28;35] This is confirmed by the results of the study done by Herring et al.[23] All children in this study had an amputation of the forefoot for the treatment of a variety of disorders of the lower extremity. Family stress was found to have the greatest influence on psychological functioning through its relationship with behaviour, self concept and intelligence. No single variable like sex, intelligence, age at time of study, age amputation, family stress, behaviour or combinations predicted self concept.

Discussion

The literature provides limited knowledge concerning participation and quality of life in children and adolescents with congenital LD. In general, it appears that children and adolescents with congenital LD are not different from normative samples. No data were found to distinguish factors which influence either the domains participation or quality of life in these children.

Participation as a concept of overall functioning has not been measured in children with LD. In James' study[22] children with UCBED were only partially questioned on their

participation. The questionnaire (PODCI)[25] addresses both activity (upper extremity physical function; mobility/transfers; pain/comfort) and participation (sports/physical function; happiness) aspects of function. Main purpose of the PODCI is to assess the efficacy of orthopaedic interventions. This provides only a limited perspective on participation in children with UCBED and none in children with LD in general. Currently, different instruments are available for measuring participation in children and adolescents with and without disabilities.[38] How appropriate those measures are in assessing participation in children and adolescents with LD needs to be studied.

More than 50% of the children and adolescents with LLD participate in sports activities. This is comparable to children and adolescents in the Dutch population: 62% of the children up to 15 years and 42% of adolescents between 15 and 25 years are participating in sports activities.[39] Encouraging children to participate in sports and recreational activities is important considering the physical, psychological and emotional benefits.[40] Due to advances in prosthetic technology and design, opportunities are available for children and adolescents with LD to be physically active in sports or recreation.[17;41]

No factors that influence participation in children and adolescents with LD have been found in the literature. In children and adolescents with cerebral palsy, determinants of participation are related to the child (age, gender, motor function, interest), environment (physical, social, attitudinal) and family (socio economic status, educational level parents, family functioning).[42] Whether these factors are generic for children with various physical disabilities, including limb deficiencies, requires further research.

Only one study[24], reporting on children with UCBED, addressed quality of life. Ten patients with LLD from the study of Herring et al[25] were evaluated on their physical and psychological functioning, several years later in their adulthood.[22] As a part of the psychological functioning quality of life was measured through a questionnaire (QLQ). The results showed no significant difference from the normative sample and supported the idea that adults who have had an amputation of the forefoot in childhood perform as well as the average adult on measures related to quality of life, self concept and psychological adjustment.

Research on how children and adolescents with LD perceive their quality of life is important for understanding the perception on their disability. In Dutch adolescents, 12 to 19 years olds with chronic disabilities, factors significantly related to quality of life

were degree of physical impairment, number of hospital visits, visibility of condition, experiences with visibility, body image, general health and self-efficacy.[44] Age was not related to quality of life.[44] Girls rated their quality of life lower than boys, which is confirmed in other studies.[45;46] Similar results were found in adults with amputation of the upper and lower extremities: factors related to their quality of life were physical disability and pain, and men had higher quality of life than women.[47] In this population, a young age at the time of amputation and an upper-limb amputation were associated with a better quality of life.

Ensuring and improving quality of life of children with disabilities necessitates understanding their psychosocial functioning.[48] The studies of CANP showed that demographic factors and degree of limb loss were not predictive of adjustment in children and adolescents with LD. This lack of relationship between physical impairment and psychosocial adjustment has been found in other studies concerning paediatric groups with physical disability.[16;49]

Adolescents with LD tend to be more vulnerable for negative social perceptions and behaviours.[35] In children with ULRD more withdrawn behaviour was considered to be a possible result from societal attitudes towards visible physical differences.[37] How to deal with peer teasing and curiosity is a concern dealt with by parents of children with ULRD.[50] These findings indicate the importance of assessing social emotional functioning in children and adolescents with LD. It will help health care providers to identify potential needs and areas of support in the challenges children and adolescents with LD face associated with the visible physical difference.

Methodological issues

All studies included in this review are descriptive and exploratory and have a cross sectional design. No causal effect can be drawn from cross sectional studies and therefore the level of evidence is limited.[51]

Most studies have used well developed psychometric measurement tools with demonstrated validity and reliability, except for two studies in which the measures used to describe physical[23;24] and psychological functioning[23] were poorly described, namely lacking psychometric information and references. Except for the abovementioned two studies, all measurements allowed comparison with population norms to control for the non-specific effects of limb deficiency.

Although the population in all studies is described as limb deficient children and adolescents, direct comparison between the studies is difficult due to age range (5-20 years) and lack of knowledge on heterogeneity of diagnoses. While some studies[22-24;37] investigated a subgroup within the diagnosis limb deficiency, the CANP project included limb deficient children without proper description of the variety within the population. It appears that the sample from CANP was collected from the same site, solicited from the UCLA Child Amputee Prosthetics Project.

Although in the CANP studies a small number of children had acquired limb deficiency, a comparison of congenital versus acquired limb loss differences on adjustment measures is mentioned in only two studies[30;36] and proper analysis is lacking. Also gender or age subgroups were not explored due to small sample sizes.

All but one study by Varni and associates[32] used correlates and multiple regression analysis to statistically predict the dependent variables by the independent variables. Despite small sample sizes many variables were tested, without proper power analysis. The common rule of requiring at least 15 subjects per predictor for a reliable equation[52] is ignored in 3 studies[29;31;35] limiting the generalisability with other samples.

No details are presented on interventions children received. As stated before, children with limb deficiencies present a heterogeneous condition with a variety of interventions and treatment options. The kind of treatment may influence the relationship between psychosocial factors and participation and overall quality of life.

Conclusion

This review reveals a lack of knowledge on how children with congenital limb deficiencies participate and how their quality of life is perceived. Their psychosocial functioning, although described as at risk, appears to be comparable to healthy peers. Further studies are required to describe how children and adolescents with congenital LD participate and how they perceive their quality of life. Furthermore, identification of factors that influence participation and the relationship with quality of life in children and youth with limb deficiencies needs to be explored. Understanding the interaction between the psychological adjustment of children with LD and participation is important to guide the development of interventions to promote optimal functioning and quality of life in this population.

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Parent and self reported participation and quality of life in children and adolescents with congenital lower limb deficiencies

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Date:	July 2009

Number of word s: 3259

Abstract: 249

SAMENVATTING

Doel : Deze studie beschrijft, vanuit het perspectief van de ouder en het kind, de participatie en gezondheidsgerelateerde kwaliteit van leven (GKvL) van Nederlandse kinderen met congenitale reductiedefecten van de onderste extremiteiten. De resultaten worden vergeleken met de participatie en GKvL van kinderen en jongeren in de algemene Nederlandse populatie. Verschillen tussen de perceptie van ouders en kinderen werden bestudeerd.

Design : Cross sectionele studie.

Methode : 64 kinderen en jongeren met congenitale reductiedefecten van de onderste extremiteiten in de leeftijd van 8 tot 18 jaar en hun ouders hebben vragenlijsten ingevuld. Participatie werd gemeten met de Children's Assessment of Participation and Enjoyment (CAPE) en voor het meten van GKvL werd gebruik gemaakt van de KIDSCREEN-52 vragenlijsten. Nederlandse referentie waarden waren beschikbaar.

Resultaten : De bevindingen indiceren een brede diversiteit en intensiteit van participatie zoals gerapporteerd door de kinderen en jongeren en hun ouders. Jongeren met congenitale reductiedefecten van de onderste extremiteiten (12-18 jaar) participeren in minder diverse activiteiten en doen minder frequent sociale activiteiten en activiteiten waar vaardigheden voor nodig zijn in vergelijking met de Nederlandse normgroep.

Volgens ouders en kinderen is de GKvL van kinderen en jongeren met congenitale reductiedefecten van de onderste extremiteiten gelijk aan de GKvL van de Nederlandse referentiegroep. Ouders rapporteren een lagere GKvL in vergelijking met hun kinderen op de domeinen "fysiek welbevinden" (p=0.045), "stemming en emotie" (p=0.006) en "zelf perceptie" (p=0.001).

Conclusie : Nederlandse kinderen en jongeren met congenitale reductiedefecten van de onderste extremiteiten participeren in diverse activiteiten en ervaren hun GKvL gelijk aan gezonde leeftijdsgenoten. De perceptie van ouders op de participatie en GKvL is grotendeels gelijk aan de perceptie van hun kinderen.

Trefwoorden: *jongeren, kinderen, reductiedefecten, participatie, kwaliteit van leven*

Abs tract

Objective : This study describes the participation and health-related quality of live (HRQoL) in Dutch children and adolescents with congenital lower limb deficiencies (LLD), and compares the results with the participation and the HRQoL of the general Dutch population. Differences between parent and self reports were explored.

Design : Cross-sectional study

Methods : This study assessed participation with the Children's Assessment of Participation and Enjoyment (CAPE) and HRQoL using KIDSCREEN-52 questionnaires through self reports and parent proxy reports of 64 children and adolescents with congenital LLD aged 8-18 years. Dutch reference data from a representative national sample were used.

Results : Findings indicate a broad range of diversity and intensity of participation as reported by children and adolescents with LLD and their parents. Significant differences in diversity and intensity of skill based and social activities were found in adolescents with LLD in comparison with the reference group. Parents reported lower intensity of physical and self improvement activities in their children aged 12-18 years in comparison with the children's self report.

HRQoL as reported by the children and adolescents with LLD and their parents was comparable with the Dutch reference group. Significant differences in comparing parent and self reported HRQoL were found in the domains "physical well being" (p=0.045), "moods and emotions" (p=0.006) and "self perception" (p=0.001).

Conclusion : Dutch children and adolescents with LLD participate in diverse activities and perceive their HRQoL similar to their healthy peers. Parents' perception of the participation and HRQoL is largely in accordance with the perception of their children.

Keywords *: adole sc ents, child ren, lower limb defici encie s, particip ation, quality of li fe*

Background

Congenital lower limb deficiency (LLD) is a chronic disorder occurring in 2 out of 10.000 live births in The Netherlands.[1] In 40% of the children with congenital LLD both legs are affected and 30% also have defects of the upper limb.[1] Although it appears that children with LLD live a life comparable to peers, limitations in the performance of physical activities are present due to dependency on prosthetic devices.[2;3] Apart from physical limitations children with LLD also differ from healthy peers in outward appearance. For being physically and visibly different, it is suggested that children and adolescents with congenital limb deficiencies are at risk for psychosocial maladjustment.[4;5]

Participation and quality of life (QoL) are two key concepts in paediatric research for understanding what parents, professionals and policy want to achieve for children with disabilities.[6] Therefore, both concepts can be considered essential outcomes in describing health status and assessment of interventions.[7]

In the framework of the International Classification of Functioning, Disability and Health (ICF) participation is one of the components describing function and health status and is therefore influenced by environmental and personal factors.[8] Participation is defined by the WHO as involvement in life situations or being involved in everyday activities.[8] Children's participation is influenced by perceptions of self and competence and the presence of supportive environments.[9-14] Participation in activities increases during childhood due to maturation and development of skills.[15;16] However, when children grow into adolescence diversity of participation decreases.[17]

Another concept overarching all domains of the ICF framework is QoL or health related QoL (HRQoL).[18] HRQoL refers to the assessment of various aspects of health from the patient's point of view and includes physical, mental, and social well-being and functioning.[19] Self reported measures of HRQoL are recommended even in children when possible.[20] Evidence has shown that children can reliably report on their HRQoL, taken into account their emotional development, cognitive ability and reading level.[21] Report on the HRQoL of the child through both parent and self report is considered to be valuable complementary information, while it can reflect meaningful differences.[19] Parents of healthy children tend to report higher HRQoL than the children themselves, whereas parents of children with chronic conditions have a tendency to rate children's HRQoL lower than the children's own rating.[22] No consistent findings are reported on factors influencing the child-parent agreement.[23] Poor parental well being and child pain negatively influences parents' perception of their child's quality of life.[23]

To date, no studies are available on how children and adolescents with LLD participate and how they perceive their HRQoL. The objective of the present study is to describe participation and HRQoL in children and adolescents with congenital LLD in comparison to Dutch children in general. In addition, differences between parent and self reports will be studies as well..

Methods

This study was a cross sectional study of children and adolescents with LLD based on questionnaires completed through self report by the children and proxy report by their parents.

Ethical approval was received from the ethical committee of Rehabilitation Centre De Hoogstraat. All parents and children gave written informed consent before participating in the study.

Participants

Children and adolescents with LLD, aged between 8 and 18 years, were recruited from Rehabilitation Centre De Hoogstraat in Utrecht, The Netherlands. The inclusion criterion was congenital deficiency of the lower limb(s) either with or without deficiency of the arm(s). Exclusion criteria were single deficiencies of arm(s) and/or combination with mental retardation or syndromale disorders.

In total 64 children/adolescents and their parents were contacted by letter with information about the study. Those who did not wish to participate in the study could return a reply note. After returning informed consent by both the children and the parents the questionnaires were sent by mail to the participants. Assistance by their parents was allowed for those children requiring help in filling out the questionnaires. Information about the degree of limb loss was obtained from the medical records of Rehabilitation Centre De Hoogstraat; for the purpose of this study they were registered as unilateral LLD, bilateral LLD and LLD in combination with deficiency of arm(s).

Measures

Participation was measured using the Children's Assessment of Participation and Enjoyment (CAPE).[24] The CAPE is a self report measure of children's participation in recreation and leisure activities outside mandated school activities. It can be used in children 6-18 years with and without disabilities.[24] The CAPE contains 55 items which are

presented as drawing and text providing information regarding three aspects of participation: diversity, intensity and enjoyment. Also information is asked about with whom the child participates and where. The items measure discretionary participation, which are situations not essential for life but they represent what children can choose to do. The Cape has five scales: recreational, active physical, social, skill-based and self improvement activities. The CAPE also assesses multiple dimensions of participation, including activity diversity and intensity. Diversity scores indicate the number of activities (maximum of 53) performed by the child over the past four months. Intensity scores indicate the average amount of time the child spends participating in activities. More diverse and intense activity participation are reflected by higher activity diversity and intensity scores. For the purpose of this study overall diversity and intensity scores of the five types of activities are used to describe the level of participation in children and adolescents with LLD. It takes approximately 30-45 minutes to fill out the CAPE.
Psychometric assessments of the CAPE have demonstrated satisfactory internal consistency (Cronbach's alpha=.30-62 for activity type scores), test – retest reliability (ICC=.64-.77 for overall participation) and convergent and discriminant validity.[24] Comparative normative data for participation scores were obtained from a convenience sample of 158 Dutch children, aged 6 to 18 years (mean age 12 years; SD 3) without physical disabilities, recruited from four mainstream schools in The Netherlands.[25]

Quality of life was measured using KIDSCREEN, a 52-item generic health related quality of life measure.[26] This questionnaire is designed for child and parent report and is applicable to healthy and chronically ill children and adolescents aged 8-18 years.[26] KIDSCREEN is a well validated measurement tool that allows comparison with the Dutch general population.[27] Reference data are available for gender and two age groups: 8-11 years and 12-18 years.[28] Ten domains of HRQoL are assessed: physical well being, psychological well-being, moods & emotions, self-perception, autonomy, parental relations & home life, financial resources, peers & social support, school environment, bullying. On a five point Likert scale two different sets of responses are: never, seldom, quite often, very often, always and not at all, slightly, moderately, very, extremely. For each domain, the relevant items are summed and scaled to yield a score in the range of 0-100, with higher scores indicating better quality of life. No global score can be calculated. Reliability for the domains is high, with Cronbach's alpha greater than 0.77 for all ten HRQoL domains. Item internal consistency and item discriminant validity of both

KIDSCREEN-52 proxy and self report version are satisfactory. Agreement between youth and proxy report were satisfactory (ICC: 0.45-0.62).[28] The time required for administration is 15 to 20 minutes.

Statistical analysis

Descriptive statistics (frequency, means, SD) were performed to describe the sample and the children's diversity and intensity of participation. Comparison between children and adolescents with LLD and reference data was done using paired Student *t*-tests.

The child characteristics age (two age groups: 8-11 years and 12-18 years) and degree of limb loss (three subgroups: unilateral, bilateral and combinations of limb deficiencies) were studied using Mann-Whitney *U*-tests.

HRQoL scores for each domain for both parents and children are described as mean and standard deviation. For the purpose of this study the recommended thresholds are used which are fixed at a value of the mean (50), plus or minus half a standard deviation.[28] Comparisons were made between the scores of the children and their parents with independent *t*-tests to determine if there were statistical significant differences between these groups. Correlations (Pearson correlation coefficient) between scores in each domain for children and parents were computed. All analyses were performed by using SPSS version 15.

Results

Sixty four children and their parents were invited to take part in the study. One family declined participation in the study by returning the reply note, seven families declined after contacting them by telephone. The questionnaires sent return by two families got lost by mail. Both families agreed to fill out the KIDSCREEN-52 a second time but not the CAPE for taking to much time. One family refused to complete the CAPE for being of opinion that the questionnaire did not fit the lifestyle of their daughter (18 years). As a result KIDSCREEN-52 questionnaires were completed by 56 parents and their children with LLD (88%) and the CAPE was completed by 53 parents and their children (83%). Nineteen (35.8%) children required help in filling out the CAPE of which 76.2% were children younger than 12 years. In all these cases assistance was provided by the mothers who also administered the parent report.

Mean ± SD age of children with LLD was at time of completing the questionnaires 12.6 ± 3.1 years; 23 children were in the age of 8-11 years and 33 adolescents in the age of 12-18 years (Table I). There were 34 boys (61.4%) and 22 girls (38.6%). Unilateral LLD was present in 34 children (60.7%), bilateral LLD in 9 children (16.1%) and 13 children had combinations of LLD with either unilateral or bilateral deficiency of the arms (23.2%). Of the study participants 94.3% attended mainstream schools (primary and secondary schools), 8.9% attended special schools and one boy of 18 years did not go to school. Parent-proxy responders were 49 mothers (87.5 %) and 7 fathers (12.5%). The demographic characteristics of the participants with LLD are summarized in Table I.

Table I: Demographic characteristics of participants with LLD (n=56)

Demographic characteristic	Number	%
Gender		
Boys	34	60.7
Girls	22	39.3
Age (years)		
8-11	23	41.1
12-18	33	58.9
Degree of lower limb loss		
Unilateral	34	60.7
Bilateral	9	16.1
Combination with upper limb loss	13	23.2
Use of		
Prosthesis	40	75.5
Orthosis	7	13.2
Combination (prosthesis + orthosis)	4	7.6
None	2	3.8
Type of school		
Mainstream	50	94.3
Special	5	8.9
None	1	1.9
Parent proxy responders		
Mother	49	87.5
Father	7	12.5

Participation

Children and adolescents with LLD (8-18 years) participated in many different activities, as reported by themselves and their parents (Table II). No differences in diversity of activities were reported in children with LLD (8-11 years) in comparison with Dutch children without physical disabilities. Adolescents (12-18 years) with LLD perform statistically significantly fewer diverse activities than their healthy peers (p=0.001).

Table II: Means (standard deviations) of diversity and intensity of CAPE activities in children (8-11 years) and adolescents (12-18 years) with LLD compared to Dutch children and adolescents without physical disabilities and parents perspective

CAPE activities	**LLD 8-18 year (n=53)**	**Parents 8-18 year (n=53)**	**Dutch reference (n=158)**	**LLD 8-11 year (n=21)**	**Parents 8-11 year (n=21)**	**Dutch reference (n= 92)**	**LLD 12-18 year (n= 32)**	**Parents 12-18 year (n=32)**	**Dutch reference (n=66)**
Diversity	25.80(6.41)	26.28(6.09)	27.13(5.82)	28.05(6.79)	29.19(5.37)	26.16(6.04)	**24.24*** (5.75)	24.17 (5.77)	28.63(5.17)
Intensity:									
Recreational	3.23 (1.20)	3.28 (1.27)	3.38 (1.13)	4.10 (1.00)	4.27 (0.86)	3.91 (0.99)	2.66 (0.97)	2.62 (1.05)	2.63 (0.89)
Physical	1.53 (0.59)	1.42 (0.60)	1.58 (0.77)	1.54 (0.64)	1.52 (0.77)	1.45 (0.72)	*1.52*** *(0.56)*	*1.34*** *(0.46)*	1.75 (0.73)
Social	3.04 (0.86)	2.95 (0.63)	2.85 (0.99)	2.79 (0.87)	2.83 (0.59)	2.36 (0.88)	**3.21*** (0.82)	3.03 (0.66)	3.54 (0.68)
Skill-based	1.03 (0.88)	0.99 (0.65)	1.13 (0.80)	1.33 (0.97)	1.34 (0.65)	1.02 (0.77)	**0.83*** (0.77)	0.77 (0.54)	1.29 (0.81)
Self-improvement	2.17 (0.86)	2.44 (0.91)	2.36 (1.07)	1.92 (0.70)	2.23 (1.02)	2.09 (1.07)	*2.34*** *(0.90)*	*2.59*** *(0.81)*	2.73 (0.97)

CAPE: Children's Assessment of Participation and Enjoyment; LLD: Lower Limb Deficiency
mean*: significant difference LLD and Dutch norms (p<0.05); *mean italic***:* significant difference parent versus self report (p<0.05)

Intensity of participation reported by children and adolescents with LLD (8-18 years) in comparison with healthy peers showed no statistical significant differences. However, adolescents with LLD have statistical significant lower intensity of skill based activities (p=0.008) and social activities (p=0.041) when comparing them with adolescents without physical disabilities.

When comparing parent and self reported diversity and intensity of participation in children with LLD (8-11 years), no differences were found. Statistical significant differences in parent and self report were found in adolescents with LLD (12-18 years) in intensity of physical activities (p=0.03) and self improvement activities (p=0.017). Although not statistical significant, parents reported lower intensity in social activities (p=0.055) in their children aged 12-18 years. Children and adolescents with LLD and the parents reported the same diversity of activities. There was good to excellent agreement between parents' and children's CAPE scores: correlations were all significant (p<0.001), ranging between 0.68 for the intensity scores of physical activities and 0.92 for intensity scores of recreational activities.

No differences were found between children and adolescents with different degrees of limb loss in either diversity or intensity of participation.

Table III: Means (standard deviations) parent and self report children and adolescents with LLD (8-18 years) and Dutch Reference Group

KIDSCREEN-52 dimensions	Self report LLD 8-18	Dutch Reference Group	Parent report LLD 8-18	Dutch Reference Group	Self / parent report *p*-value
Physical Well-being	52,42 (10.75)	52.88 (10.02)	49.68 (10.20)	53.18 (10.10)	**0.045***
Psychological Well-being	53.42 (8.93)	53.32 (8.93)	53.01 (8.87)	52.72 (9.80)	0.745
Moods & Emotions	52.41 (10.10)	51.38 (9.85)	47.65 (10.77)	48.09 (9.87)	**0.006***
Self Perception	51.65 (8.44)	52.22 (9.89)	47.44 (7.30)	50.86 (10.05)	**0.001***
Autonomy	53.11 (7.57)	54.37 (8.92)	50.96 (7.25)	54.64 (8.01)	0.112
Parent Relation & home Life	52.78 (8.51)	53.16 (8.99)	52.90 (8.58)	53.21 (9.29)	0.925
Social Support & Peers	51.51 (12.13)	52.37 (9.33)	50.35 (10.78)	53.29 (9.00)	0.428
School Environment	52.66 (9.70)	53.21 (9.78)	51.94 (8.92)	53.40 (9.99)	0.541
Social Acceptance & Bullying	51.38 (9.88)	48.39 (10.20)	48.61 (11.28)	46.73 (11.11)	0.078
Financial Resources	54.60 (8.67)	52.09 (9.48)	55.39 (8.63)	51.90 (8.99)	0.461

LLD: Lower Limb Deficiency

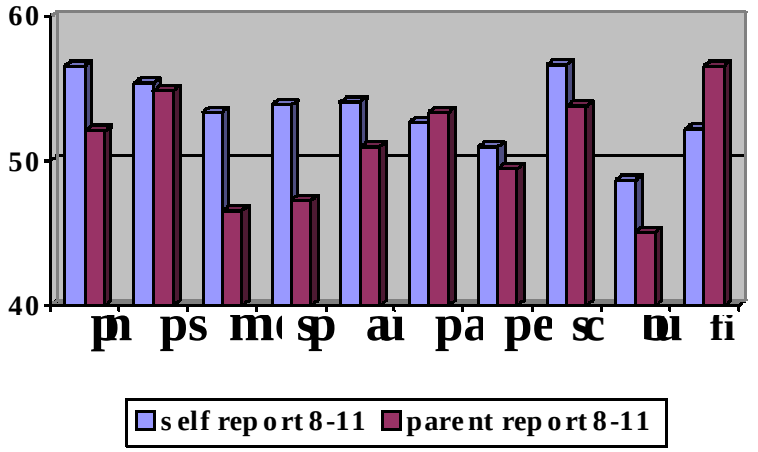
***p*-value:** statistical significant difference between self report and parent report (p<0.05)

Quality of life

The perception of children and adolescents with LLD (8-18 years) on their HRQoL is comparable with the Dutch reference group (Table III). Parents overall reported good HRQoL for their children (8-18 years) on all domains compared with Dutch reference data. Parents of children with LLD (8-11 years) reported lower HRQoL in comparison with the Dutch reference group on the domains “self perception”, “physical well being”, “autonomy” and “peer support”. No differences were found in relation to degree of limb loss. Good HRQoL on all domains was reported by the parents of adolescents with LLD (12-18 years).

When comparing parent and self reported HRQoL in all children and adolescents with LLD statistical significant differences were found in the domains “physical well being” (p=0.045), “moods and emotions” (p=0.006) and “self perception” (p=0.001).

In the younger age group (8-11 years) statistical significant differences were found in the domains “physical well being (p=0.023), “moods and emotions” (p=0.031), “self perception” (p=0.024) and “bullying” (p=0.028). No statistical significant differences were found comparing parent and self report of adolescents with LLD. Correlations between the children’s and parents’ HRQoL scores were significant (p<0.005), ranging between 0.28 for the scale “Moods and Emotions” and 0.56 for the scale “School” indicating fair to good agreement. Agreement on the scales “Autonomy” (r=0.10) and “Financial Resources” (r=0.15) was poor. Figure I presents the results from self and parent report in the two different age groups.



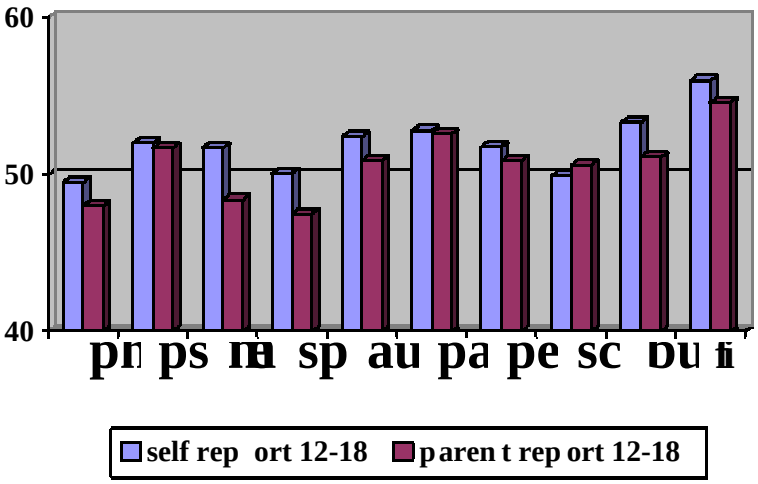


***Figure I*:** KIDSCREEN-52 HRQoL scores by dimension for children and adolescents with LLD and their parents. Scales for each dimension: mean=50 in the reference population
Abbreviations: ph=physcial well being; ps=psychological well being; me=moods & emotions; sp=self-perception; au=autonomy; pa=parent relation & home life; pe=social support & peers; sc=school environment; bu=bullying; fi=financial resources.

Discussion

Objective of this study was to describe participation and HRQoL in children and adolescents with congenital LLD in comparison to Dutch children in general using parent and self reports. Key findings indicate that the participation and perceived HRQoL of Dutch children and adolescents with LLD are similar to the general Dutch population. No differences have been found comparing parent and child report on the level of participation except for intensity of physical activities and self improvement activities in adolescents. Although parents of adolescents with LLD reported their children's HRQoL to be comparable to healthy children, they reported lower self perception, autonomy, peer support and physical well being in their children aged 8-11 years.

Increased participation is associated with higher levels of physical, cognitive and communicative function.[12] This might explain the findings of this study that children and adolescents with LLD participate fully. In general children with LLD have a high level of physical activity and participate in sports although most of them are dependent on prosthetic devices.[2;3]

In this study adolescents with LLD reported to perform fewer diverse activities and less intensity in skill based activities and social activities in comparison with Dutch adolescents without physical disabilities. It is known from various studies that with increasing age the level of participation is decreasing[29] and that frequency of physical activity engagement is reduced as adolescents grow older.[30] The fewer diversity in participation of adolescents with LLD in this study might be a result from the time of measurement with the CAPE. The CAPE asks for activities done over the past four months which results in seasonal effects. This study was performed during the winter perhaps causing fewer diverse activities outside.

Varni et al reported that adolescents with limb deficiencies tend to be more vulnerable for negative social perceptions and behaviours.[31] In children with cerebral palsy negative reactions by others, like bullying and staring, are considered factors negatively influencing their participation.[32] Negative attitudes by others may explain why adolescents with LLD participate in fewer diverse activities.

It appears that diversity and intensity of participation of Dutch children in general is lower than participation levels of children with different diagnostic groups as described in studies from North America.[24;29;33-35] Next to demographic, environmental and functional factors, cultural factors seem to have an influence on

participation as well. Comparison of participation levels between diagnostic groups in different countries should be interpreted with caution.

Like other children with physical disabilities[36-38], children and adolescents with LLD perceive their HRQoL comparable to their healthy peers. Parents rated their child's (8-11 years) self perception, physical well being, autonomy and peer support lower compared to the child's rating. This suggests that parents had a more negative perception on how the limb deficiencies has an affect on their children. This difference in parent and self report is shown in other studies in which parents of children with chronic conditions proxy report lower HRQoL than the children themselves.[22] On the other hand and in contrast with previous findings, in this study the parents rated the adolescents HRQoL similar to the adolescents themselves. This suggests a valid self report by the adolescents and perhaps a less valid self report by the children with LLD. To further explain the differences in child self report and parent proxy reports it is recommended to include variables like parental well being, socioeconomic status, family structure and mental health in future studies.[23]

Furthermore some remarks can be made about the outcome measures CAPE and KIDSCREEN-52. The CAPE in contrast to the KIDSCREEN-52 does not provide a specific parent questionnaire. For the purpose of this study both the children and the parents used the same version of the questionnaire. Parents were asked to fill out the CAPE from their perspective on the participation of their children. Children were allowed to get assistance in administering the CAPE. All mothers who assisted the child also administered the parent report. This may have diminished differences between parent and self report in the children with LLD (8-11 years).

Age appropriateness of the CAPE is doubtful for the broad age span of 6-18 years. Adolescents do not always recognise themselves in the activities as described in the CAPE. Measurement of participation in at least two age bands seems more appropriate given the growing autonomy experienced by the adolescents.[39]

Although the CAPE measures objectively the level of participation it does not provide an indication to which this level is perceived as good enough or as a limitation from the perspective of the person. As argued by different authors[6;9], the subjective experience of participation is covered sufficiently by the construct of HRQoL which underlines the use of KIDSCREEN-52 in this study. In the study by Young[40], the dimensions of KIDSCREEN-52 were compared to disabled children's accounts of their well being. The findings suggested that KIDSCREEN-52 mapped well to the

children's accounts of their lives, but some specific aspects of life were missed. These included: home life, neighbourhood, siblings, pain and discomfort, inclusion and fairness in relationships, particularly peer relationships.[40] Whether all aspects of life which are important to children with LLD are represented by the KIDSCREEN-52 is unclear.

Strength of this study is that it is the first one exploring participation and HRQoL of children and adolescents with congenital LLD using validated measurement tools. Data were collected through self report and through parent report. Both reports can be considered as complementary to one another, providing a broad perspective on both concepts in children with LLD. All participants were recruited from a single-centre convenience sample resulting in a high response rate. A broad geographic diversity is represented due to the supra-regional rehabilitation services of Rehabilitation Centre De Hoogstraat in The Netherlands. Limitation of this study is the lack of exploration on factors influencing participation and HRQoL. More research is needed to identify factors influencing participation and HRQoL in children and adolescents with LLD. It can optimise service planning and development of intervention programmes for both families and children with LLD

Conclusion

Dutch children and adolescents with LLD participate in diverse activities and perceive their HRQoL similar to their healthy peers. Parents' perception on the participation and HRQoL is largely in accordance with the perception of their children. Parents tend to perceive the HRQoL of the children aged 8-11 years lower as the children do themselves. Degree of limb loss did not affect participation and the perception on the different dimensions of the HRQoL.

These findings provide a better understanding of the participation and HRQoL of Dutch children and adolescents with congenital LLD. It can reassure families and service providers that children and adolescents with congenital LLD have full potential to participate in society and that they in general perceive their lives as satisfactory.

Routine assessment of participation and HRQoL in children and adolescents with LLD should be considered as a part of rehabilitation services to provide a basis from which health care professionals, the children and their families can plan their goals on.

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