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A life worth sharing

Understanding quality of life of persons with profound
intellectual and multiple disabilities



Appolonia M. (Marga) Nieuwenhuijse

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Understanding quality of life of persons with profound intellectual and multiple disabilities
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A life worth sharing

Understanding quality of life of persons with profound intellectual and multiple disabilities

ACADEMISCH PROEFSCHRIFT

ter verkrijging van de graad van doctor
aan de Universiteit van Amsterdam
op gezag van de Rector Magnificus
prof. dr. ir. K.I.J. Maex

ten overstaan van een door het College voor Promoties ingestelde commissie,
in het openbaar te verdedigen in de Aula der Universiteit
op vrijdag 21 januari 2022, te 11.00 uur

door Appolonia Margrietha Nieuwenhuijse
geboren te Ede

Promotiecommissie

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Het levensverhaal van Britt

Geboorte

“Britt werd thuis geboren op vrijdag 23 april 2004. Direct bij de geboorte bleek er al wat aan de hand. De volgende dag werd Britt opgenomen in het ziekenhuis en begonnen de vele onderzoeken om uit te vinden wat er aan de hand was met haar. De artsen konden niet een juiste diagnose stellen. Zij kwamen niet verder dan een atypische vorm van het Aicardi syndroom door onder andere de afwezigheid van het corpus callosum. Wel was al duidelijk dat ze niet zelf zou kunnen lopen, eten, praten. Ze was volledig afhankelijk van zorg dus. Na de vele onderzoeken en teleurstellingen hebben wij in haar 2e levensjaar besloten om Britt mee te nemen naar huis. We hebben haar losgekoppeld van de constante toevoer van antibioticum en hebben alle

onderzoeken gestaakt. We begonnen te kijken naar Britt zelf en niet alleen maar naar haar medische staat. We gingen kijken naar een heel lief meisje met een heel bijzonder en mooi leven in het vooruitzicht. Dit heeft Britt ontzettend goed gedaan. Ze werd onderdeel van ons gezin en knapte zienderogen op. Ze hoefde niet meer aan het antibioticum en wij begonnen haar steeds beter te snappen en leerden dat ze wel degelijk op haar manier met ons communiceerde. Haar kwaliteit van leven verbeterde zienderogen. Wat een ontzettend mooi en bijzonder meisje!”

Nancy & Henk, moeder & vader van Britt



Chapter 1

Chapter 1

General introduction

Introduction

In my work as managing director of specialised day-care centre Omega in Amsterdam, I was regularly confronted with children and adults with profound intellectual and multiple disabilities (PIMD) who passed away. Some years there was only one, some years there were six persons who died that year. I noticed that I, the professional caregivers, and even the parents could react differently to the death of one person or another person with PIMD. In one case, we were shocked and startled. In another case, we were resigned, although, of course, we felt deeply sad, but we accepted. We said to each other, “It is okay; he/she is resting now”.

Why is that? What does this mean? Does this say something about how we valued their quality of life (QoL) at the time they passed away? How did we know about their QoL? Even more surprising was that parents and professional caregivers shared these feelings in most cases, although we had never discussed this. This puzzled me, so I was very enthusiastic when Omega was asked to join a research project about the QoL of persons with PIMD. This was my chance to gain a deeper understanding about the QoL of persons with PIMD. Not only when they have passed away, although questions about QoL are pressing at the end of life, but also in “normal” situations. How do we learn about QoL, and which aspects should we take into consideration? Moreover, how is it possible that parents and professionals seem to share the same feeling about QoL without discussing it? These are questions about QoL and about the assessment of QoL, which I wanted to investigate in this study.

Persons with PIMD

Persons with PIMD have profound intellectual disabilities (IQ not valid or not reliably measurable) and profound neuromotor dysfunctions, sometimes with sensory impairments such as visual and hearing problems and medical problems (including seizures, respiratory and feeding problems) [1]. They are dependent on support from others in all aspects of their daily needs. Communication is mostly presymbolic or protosymbolic, such as movements, sounds, body postures, facial expressions or muscle tensions [2]. This definition is in accordance with the working definition developed by the Special Interest Research Group (SIRG) PIMD of the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD).

Persons with PIMD are not officially registered as such, but it is assumed that approximately 10,000 persons have PIMD in the Netherlands [3]. Because of their severe medical problems, persons with PIMD need much medical care and,

therefore, often encounter medical staff [4]. Furthermore, persons with PIMD are hospitalised frequently [5]. In their younger years, children with PIMD mostly live at home with their parents and attend specialised day-care centres or special schools. When they become older, their parents want/need more respite care such as guesthouses and weekend care and intensification of care in their home situations. Eventually, most adults with PIMD live in a residential care facility, although the living situations and daily activity situations differ per country. In some countries, children with PIMD are more likely to attend special schools.

Quality of Life

Since the beginning of this century, the QoL of persons with intellectual disabilities (ID) has been a subject of academic interest [6] [7] [8] [9]. In 2002, an international panel of experts from the Special Interest Research Group (SIRG) QoL of the IASSIDD described core principles for conceptualisation and for measuring QoL of persons with ID, stating the following for measuring QoL:

1. “QoL measures the degree to which people have meaningful life experiences that they value.
2. QoL measurement enables people to move toward a meaningful life that they enjoy and value.
3. QoL measures the degree to which life’s domains contribute to a full and interconnected life.
4. QoL measurement is undertaken within the context of environments that are important to people: where they live, work and play.
5. QoL measurement for individuals is based upon both common human experiences and unique, individual life experiences” [8].

Cummins described four conceptualisation principles: “a) QoL is multidimensional and influenced by personal and environmental factors and their interactions; b) it has both subjective and objective components; c) it has the same components for all people and d) it is enhanced by self-determination, resources, purpose in life and a sense of belonging” [6].

Petry, Maes and Vlaskamp developed a procedure for evaluating QoL for persons with PIMD [10]. This procedure was implemented in several studies on QoL of individuals with PIMD and resulted in a questionnaire to measure their QoL [11] [12].

Reinders gave a more philosophical view on QoL (‘a life worth living’) of persons with PIMD [13]. He combined theological anthropology and ethics to substantiate the humanity of profoundly disabled persons and their capacity for friendship as a consequence of the friendship God is providing to all human beings.

Proxy assessment of QoL

As verbal communication is impossible and non-verbal signs are hard to interpret, the assessment of QoL of persons with PIMD relies almost exclusively on parents, family members, professional caregivers or other representatives. However, assessment by proxies is challenged in the literature [11] [14] [15] [16] [17]. For example, Cummins stated that proxy measurement is not valid to assess the person's subjective wellbeing [14]. From studies in groups of persons with less severe intellectual disabilities (ID), it appears that proxy outcomes differ from self-assessments [14] [18] [19]. Therefore, the assessment of QoL of persons with PIMD requires attention.

Use of the concept of QoL

Most studies about QoL, both in persons with ID and with PIMD, focus on the use of the concept of QoL “as a conceptual framework for assessing quality outcomes, as a social construct that guides quality enhancement strategies and as a criterion for assessing those strategies” [17]. However, new developments in the past years required other utilizations of the concept of QoL, and especially more attention for both good and poor QoL. One of those new developments is that the lifespan of persons with PIMD has increased [20] [21]. The ageing of persons with PIMD combined with more treatment options also means that end-of-life decisions such as withholding or withdrawing life-prolonging treatments may need to be made more often. The current and expected QoL of the person will play an important role in such decisions. Recent studies revealed that QoL indeed plays a role in end-of-life decisions in persons with PIMD [22].

Also the development of specific, and usually very expensive, new treatment modalities (e.g. medicine for very rare diseases such as for Pompe or Fabry disease) resulted in a different use of the concept of QoL, because QoL became an important part of negotiations on the reimbursement of these treatments. A connection was made between the diminutive improvement in QoL of the patients and the expensiveness of the treatment. This provoked heated discussions, both in politics and in society [23].

Questions about “good” and “poor” QoL

Instead of just focusing on how to improve QoL, or how to use QoL as an outcome measure for interventions or quality of care [24], a study is required in which we also explore when QoL of persons with PIMD is described as “poor QoL” or when concerns about QoL arise. However, exploring concerns about QoL or poor QoL in this group is challenging. First, Reinders et al. warned against using the concept

“poor QoL” because they think that this description may be used to justify the termination of the life of persons with ID [25]. In fact, they labelled this process as the “new eugenics”, referring to World War II. Reinders et al. [25] also brought up this subject during the international congress of the IASSIDD in 2019, which provoked emotional and intense discussions. Second, in discussions with experts about persons with PIMD before this study was undertaken, this possibility of misuse of the description of “poor QoL” was mentioned frequently.

However, we think we should also explore descriptions of poor QoL because, in everyday practice, concerns about QoL do arise, as we have seen in the examples described above about Omega. Furthermore, parents, physicians and professional caregivers are confronted with these subjects and struggle with these questions [26]. Exploring these questions can help parents, physicians and professional caregivers care for persons with PIMD. Furthermore, we think we can explore questions about QoL because of the design of our study as an inductive qualitative study using interviews and focus groups with participants who know persons with PIMD well. Above all, a qualitative design helps to explore reports by individuals without trying to create generalisable standards or quantitative indicators of QoL.

Objective

We aimed to conduct a series of studies in which we not only explored how “good QoL”, but also “poor QoL” is described and when concerns about QoL arise. Because we know from clinical practice that concerns about QoL specifically arise in parents, professional caregivers and physicians who care for persons with PIMD, we focussed on them as our respondents. Our research questions were:

- Which concepts are used for QoL in persons with PIMD? (Chapter 2)
- Which instruments or methods are used, and who determines QoL? (Chapter 2)
- How do physicians describe the good and poor QoL of persons with PIMD? (Chapter 3)
- When do physicians start to have concerns about the QoL of persons with PIMD? (Chapter 3)
- What is the perception of other professional caregivers about good and poor QoL of persons with PIMD? (Chapter 4)
- What do parents think is necessary to assess the QoL of their child? (Chapter 5)
- Who, according to parents, can best assess the QoL of their child? (Chapter 5)

As a part of this thesis, we developed a tool for parents based on the results. This tool may empower parents to think about and describe the QoL of their child.

Methods

To explore the first topic, we conducted a literature search. This search provided an overview of concepts and assessment of QoL in persons with PIMD, which we could compare with our empirical findings. To explore the other topics, we performed an inductive qualitative study. We used qualitative research because we wanted to explore experiences, emotions and perceptions [27]. We relied on interviews as our main research method. We asked physicians and professional caregivers who know persons with PIMD well how they described “good QoL”, “poor QoL”, and when they started to have concerns about QoL. Furthermore, we asked them how they ascertained the QoL of persons with PIMD.

Normally, we would have interviewed parents first because they know persons with PIMD best [28]. However, another study was performed simultaneously, in which parents of children with PIMD were also interviewed [29]. This study explored end-of-life decisions for children with severe disabilities and parents of children with PIMD could be recruited for both studies, which we thought was too much burden for parents. Therefore, we chose to explore the parents’ perceptions at a later stage in focus groups. We opted for focus groups because we wanted to encourage discussion between parents and give parents the opportunity to share their views with each other.

We conducted seven interviews with physicians, 11 interviews with professional caregivers, five interviews with mothers, and 22 parents participated in the focus groups. We used the qualitative data analysis software, MaxQDA, to code the interviews and focus groups. After that, we analysed the interviews and findings from the focus groups thematically [30].

Thesis outline

First, we report the results of our literature search in Chapter 2. Although several articles about QoL of persons with PIMD exist, no overview of the literature about the QoL assessment for PIMD has been provided, including articles published after 2009 and supported by a systematic search of electronic databases. We answered the following research questions: (1) Which concepts are used for QoL in persons with PIMD? (2) Which instruments or methods are used, and who determines QoL?

In Chapter 3, we describe the results of the interviews with physicians. We chose to interview physicians who knew the target group well. We searched for information on physicians’ perceptions of good and poor QoL of persons with PIMD and when they start to have concerns about QoL. More specifically, we searched for the tipping point at which they start to have concerns and for what reasons they have concerns

about QoL. We answered the following research questions: 1) How do physicians describe good and poor QoL of persons with PIMD? 2) When do they start to have concerns about the QoL of persons with PIMD?

In Chapter 4, we describe the results of the interviews with the professional caregivers. Professional caregivers play an important role in the care of persons with PIMD and their QoL. Not only are they one of the assessors of QoL of persons with PIMD, but they also influence QoL of persons with PIMD. Since professional caregivers may be confronted with medical decisions concerning persons with PIMD and the dilemmas associated with this, their perspective not only on good QoL but also on poor QoL of persons with PIMD is important. Therefore, the objective was to explore professional caregivers' perception of good and poor QoL of persons with PIMD.

Chapter 5 describes the results of the focus groups with parents. The perspective of parents on the assessment of QoL of their child with PIMD is lacking in the literature. However, the assessment of the QoL of their child is important, because QoL is a topic in medical decisions concerning their child. Therefore, we explored parents' perceptions on the assessment of the QoL of their child. The central questions were: 1) What do parents think is necessary to assess the QoL of their child? 2) Who, according to parents, can best assess the QoL of their child?

Finally, in Chapter 6, we summarise the main conclusions of the different studies and answer the research questions. Next, we describe the implications of the most important results for daily practice. Subsequently, we reflect on some of the results concerning ethical issues and medical decision-making.

In an appendix, we present the tool that we developed during this study to help parents describe the QoL of their child and empower them in their discussions with physicians and others about the QoL of their child.

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Het levensverhaal van Britt

Familie

“En daar was Britt in ons leven! We wisten nog niet helemaal wat ons te wachten stond en hoe het leven van Britt ons leven zou veranderen. Haar broer Lars was direct heel lief en zorgzaam voor haar. En wij als ouders moesten uiteraard wennen aan het leven met een gehandicapt kind. Al snel merkten we dat we haar goed konden begrijpen. We kregen eigenlijk tegen de verwachtingen in veel van haar terug. Na vier jaar werd haar zusje Suze geboren. Dat was ook een heel mooi proces, waardoor het gezin in evenwicht kwam. Er ging logischerwijs veel aandacht naar Britt, maar door de komst van Suze werd dat rechtgetrokken. En Britt werd ook de lieveling van alle andere familieleden, oma’s, opa, neven en nichten. Vrij snel snapten we Britt beter en namen we haar ook overal mee naar toe. Britt kon zo intens genieten van gezelligheid met en bij familie, vrienden en burens.



Uiteraard waren er voor ons gezin ook af en toe best moeilijke momenten als het met de gezondheid van Britt minder goed ging en ze ook wel met spoed naar het ziekenhuis moest. Dat waren voor met name haar broer en zusje, maar ook voor ons als ouders, heftige ervaringen.

Voor een gezin met drie kinderen en allebei de ouders die werken, was het een hele klus om alles in goede banen te leiden, maar met hulp van haar broer, zusje, de oppassen, dagopvang Omega en de liefde van Britt, hebben wij het gered maar zeker ook enorm genoten van en met elkaar.”

*Nancy & Henk, moeder & vader van Britt,
en Lars en Suze, broer en zusje*



Chapter 2

Quality of life of persons with profound intellectual and multiple disabilities: A narrative literature review of concepts, assessment methods and assessors

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MA PhD Erik Olsman: conceptualisation, methodology, investigation, analysis, writing, review and editing and supervision.

Abstract

Background

The objective of this study was to provide an overview of concepts, methods of assessment and assessors in empirical studies on quality of life (QoL) of persons with Profound Intellectual and Multiple Disabilities (PIMD).

Method

A narrative literature review was conducted by searching electronic databases until December 2016, and the included studies were then analysed thematically.

Results

Twenty-one articles were included in the analysis. Multidimensional concepts of QoL consisting of both subjective and objective components were found, although the articles used different distinctions for each. QoL was assessed by behavioural observations or interviews/ questionnaires. Family members, primary caregivers and more distant observers (or a combination of these three) determined QoL. Triangulation of assessors and methodological pluralism were used.

Conclusions

This review highlighted several components of QoL in persons with PIMD, as well as challenges in researching the QoL of persons who are not able to express themselves verbally. The distinction between objective and subjective QoL was difficult to clarify, with interpretation being both inevitable and necessary. Although triangulation is seen to be good practice in QoL assessment, the role of proxies in assessing the QoL of persons with PIMD needs further investigation.

Introduction

Quality of life (QoL) is often a key factor in the decision-making process for those caring for people with complex care demands [1]. There are decisions to make about major surgeries, tube feeding and withholding or sustaining life support. Clarity in the description and consensus about the assessment of QoL is therefore crucial. Obviously, QoL can best be judged by the individuals themselves, but persons with Profound Intellectual and Multiple Disabilities (PIMD) cannot verbally express how they experience their QoL.

Persons with PIMD are individuals with profound intellectual disabilities (IQ < 20) and profound neuromotor dysfunctions, sometimes with sensory impairments and even medical problems (including seizures, respiratory and feeding problems). Furthermore, these persons seem to have neither an apparent understanding of verbal language nor a symbolic interaction with objects [2]. This is in accordance with the working definition developed by the Special Interest Research Group (SIRG) PIMD of the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD). Persons with PIMD are dependent on support from others in all aspects of their daily needs. Communication is mostly pre- or proto-symbolic and consists of movements, sounds, body postures, facial expressions or muscle tensions [3]. Persons with PIMD are not officially registered as such, but it is assumed that in the Netherlands approximately 0.06% of the total population have PIMD, although most of them were adults [4].

Since 2000, much academic attention has been given to QoL of persons with intellectual disabilities (ID) [5] [6] [7] [8]. Schalock et al. [8] described the core principles for measuring QoL, stating the following:

- (1) QoL measures the degree to which people have meaningful life experiences that they value.
- (2) QoL measurement enables people to move towards a meaningful life they enjoy and value.
- (3) QoL measures the degree to which life's domains contribute to a full and interconnected life.
- (4) QoL measurement is undertaken within the context of environments that are important to people: where they live, work and play.

- (5) QoL measurement for individuals is based upon both common human experiences and unique, individual life experiences.

In addition, several books describe the concepts of QoL. In his overview of the literature, Lyons [9] referred to the Special Interest Group on quality of life (SIRG QoL) of the IASSIDD and the criteria it describes for research on QoL in persons with ID. These criteria include the following: multidimensionality, guiding operational principles, methodological pluralism, a system perspective and a focus on individual and family QoL. In 2009, Petry and Maes [10] wrote an overview chapter in a book in which they specifically addressed the importance, definitions and measurement of QoL in persons with PIMD. However, they did not provide a review of available studies.

Therefore, no overview of the literature on the assessment of QoL in PIMD has been provided, including articles published after 2009 and supported by a systematic search of electronic databases. Such an overview is necessary to clarify the current state of concepts, methods of assessment and assessors in the research on QoL in persons with PIMD. This can be helpful in future research on QoL in persons with PIMD related to medical decisions. In addition, more information on this subject may help physicians and family members to make adequate decisions when they are confronted with medical care decisions related to the QoL of persons with PIMD. Therefore, the main aim of this study was to answer the following research questions: (1) Which concepts are used for QoL in persons with PIMD? (2) Which instruments or methods are used and which persons determine QoL?

Method

A narrative literature review of the empirical studies describing QoL in persons with PIMD was conducted.

Literature search and selection of articles

The studies were identified through searches in the following databases: MEDLINE, EMBASE, CINAHL, PsychINFO and Philosopher's Index, with the help of an experienced clinical librarian. These searches first occurred in March 2015, and the searches were repeated in May 2016 and December 2016. Two sets of keywords were used, the first relating to QoL, including but not limited to QoL, Health-related QoL or wellbeing. The second related to the target group, such as persons with PIMD or persons with profound, severe or multiple mental retardation; those with a

learning, developmental, intellectual disability; or those with a medical complexity, minimal movement or cerebral palsy. A complete description of search terms and strategies is available from the authors.

We excluded the following sources: articles not written in English, Dutch, German or French, as well as editorials and conference abstracts. In addition, articles in which persons with PIMD were not the target group were excluded. We also excluded articles in which QoL was not the subject but an outcome measure in an interventional study. And finally, articles for which the subject was QoL of relatives of persons with PIMD, for example, were also excluded.

The inclusion criteria for the target group were a combination of multiple disabilities including both an intellectual and physical disability with a dependence on support from others in all activities in daily life, as well as no apparent understanding of verbal language. Only peer-reviewed articles were included. From these, 3182 articles were found including 790 duplicates, leaving 2392 usable articles. For the search strategy, see Figure 1 (flow chart).

Figure 1. Flowchart



Procedure

In order to apply the inclusion criteria, the first and the last author independently screened the titles of the 2392 articles and then compared their findings until they reached consensus. The articles that clearly did not fulfil the inclusion criteria were excluded. When there was doubt, the article was included for the next round. The abstracts were then screened, following the same procedure. The two researchers had doubts about 18% of the articles and after discussing these together, there remained doubts about 6%. This 6% was screened by the second author of this paper, who made a final decision. Of the remaining articles, the full text was screened by the first author and articles that raised doubt (21%) were also screened by the last author. The cross-references of the included articles were also screened by the first and the last researcher. Both researchers discussed including or excluding these articles until they reached consensus.

Analysis

First, the articles were screened thematically on the research questions (concepts, instruments/methods and assessors). Then, differences and similarities between descriptions of concepts were compared. Subsequently, an inventory was drawn up, both the instruments and methods of assessment described and an inventory of the assessors. After that, we searched for relationships between the concepts and the assessment methods used. The results of these thematic analyses were discussed by the first and the last researcher, who went back and forth several times between the included articles and the thematic analysis in order to refine the analyses. For example, many articles appeared to refer to both the subjective and the objective component (as one part of the concepts). Therefore, we searched in the articles for the definitions of subjective and objective components and the possible relationships between their definitions and the assessment methods. We found that subjective components were assessed by the use of different methods and objective components mostly by questionnaires. In order to sharpen and critically test the initial analyses, they were also discussed with the other authors of this paper.

Results

Twenty-one articles were included. For a summary of the included articles and results, see Table 1. The included studies were performed in Europe, the US, Canada and Australia. Also included were studies aiming to develop a procedure for

evaluating QoL of people with PIMD, conducted in Belgium, the Netherlands and Germany [11].

Which concepts of QoL are used?

The articles included did not directly answer the question “which concepts are used for QoL?” However, 12 of the included articles referred to Schalock’s principles relating to QoL [11] [12] [13] [14] [15] [16] [17] [18] [19] [20] [21] [22]. Two articles referred to the SIRG QoL [13] [17] in which QoL was conceptualised by the following descriptions: (a) QoL is multidimensional and influenced by personal and environmental factors and their interactions; (b) it has both subjective and objective components; (c) it has the same components for all people and (d) it is enhanced by self-determination, resources, purpose in life and a sense of belonging. The last element was not examined in the studies included in this review.

With respect to multidimensionality, several combinations of domains were found. For an overview of domains, see Table 2. These several combinations of domains were operationalised in items [12] [23] [24] [25], and twice a Delphi round was used [23] [25]. In addition, in the article of Petry et al. [24], the basic domains of Felce and Perry [26] were described as valid and relevant for people with PIMD, which may refer to the third SIRG QoL statement that QoL has the same components for all people.

References to objective and subjective components of QoL were found in 10 studies [14] [15] [16] [17] [18] [20] [21] [22] [27] [28]. Vos et al. [22], p. 441, following Cummins [5], Landesman [29] and Perry and Felce [30], described the objective component of QoL as the “information on the life of the person that has properties that can be measured in the public domain and verified by different parties (e.g., how many visitors one has during the week).” On the other hand, Vos et al. [22], p. 441 defined the subjective component as the information “about the private world of experience of the person, which can be viewed as an estimate of the true subjective experiences and cannot be verified directly through any evaluation of the variable in question beyond the level of the individual.” The following subjective components were examined: happiness [31] [32], pleasure [16], emotions [22], mood [15] [21] [22] [33], life satisfaction [14] and interest [15] [33]. However, the studies made different distinctions. For example, one author includes emotions in the subjective components of QoL [20], [22], whereas the QoL-PMD includes emotional wellbeing in the objective components of QoL [18] [24] [25].

Some of the included articles referred to the (strategic) use of the concept of QoL [11] [15] [18] [21] [22]. They stated that QoL was used as a conceptual framework for assessing quality outcomes, that is, as a social construct that guides quality enhancement strategies and as a criterion for assessing these strategies [34]. Petry, for example, mentioned that persons with PIMD need special care, and consequently that attention should be given to the optimisation and evaluation of the quality of care because its most important result is an enhanced QoL [11].

Table 1. Characteristics of the included articles

1st author	Year	Objective	Persons with PIMD	Study design	Components of QoL	Instruments or methods to assess QoL PIMD	Assessors	Psycho-metric properties
Ouelette-Kuntz	1994	To develop the Quality of life Interview Schedule (QUOLIS) and the intra- and inter-reliability	n=10, age >18	Qualitative (interviews)	Twelve domains of Ouelette	QUOLIS: questionnaire (filled in by interviewer)	Trained interviewer, interview with two people who know the person well, family member or friend and staff member	Intra- and interrater reliability
Green	1996	To operationalise, measure and increase happiness among people with profound disabilities	n=5, age 18-41	Qualitative (observations)	Happiness	Two experiments using behavioural observations & video tapes	Familiar and unfamiliar observers	NA
Favell	1996	To develop and demonstrate methodologies for measuring happiness in people with profound mental retardation and other severe handicaps	study 1: n=7, age not mentioned study 2: n=1, age 30 study 3: n=20, age not mentioned study 4: n=1, age 30	Qualitative (observations of video tapes and other observations)	Happiness	Four studies using observations of video tapes and other observations	Supervisory staff (familiar), trained observer (unfamiliar)	NA
Petry	2001	To develop and implement a valid and reliable procedure for evaluating the QoL of people with profound and multiple learning disabilities	NA	NA (Study protocol)	NA	NA	NA	NA

Table 1. Continued

1st author	Year	Objective	Persons with PIMD	Study design	Components of QoL	Instruments or methods to assess QoL PIMD	Assessors	Psycho-metric properties
Ross	2003	To develop an informant questionnaire on affect for use to adults with PIMD and examine its psychometric properties	n=53, age >18	Quantitative (questionnaires, psychometric analysis)	Mood, Interest and Pleasure	MIPQ: questionnaire	Familiar informants	Intrater reliability; internal consistency; construct validity
Lyons	2005	To underpin the assumptions and face validity of the Life Satisfaction Matrix	n=22, age<18	Qualitative (participant observations, semi-structured interviews)	Satisfaction	Life Satisfaction Matrix: observations and interviews	Most familiar communication partner, next communication partner and researcher (unfamiliar)	Face validity
Petry	2005	To consider the general validity of the basic domains of quality of life in persons with PIMD	n=42, age <18; n=34, age>18	Qualitative (interviews)	Five domains of Felce and Perry	NA	NA	NA
Petry	2006	To explore a procedure for drafting individualised profiles of how people with profound multiple disabilities express pleasure and displeasure	n=6, age 9-29	Mixed method (video tapes, questionnaires)*	Pleasure/displeasure	Behavioural observations, questionnaire, videotape	Parent, direct support staff, supervisory staff and independent researcher (unfamiliar)	NA
André	2007	To describe quality of life in children with severe disability and to look for factors which influence this QoL, in particular gastrostomy	n=28, age 1-18	Qualitative (interviews)	Five domains of André	QUALIN: questionnaire (filled in by interviewer)	Parents	NA

Table 1. Continued

1st author	Year	Objective	Persons with PIMD	Study design	Components of QoL	Instruments or methods to assess QoL PIMD	Assessors	Psycho-metric properties
Petry	2007a	To assess items on QoL and related aspects of support in people with PIMD	NA	Qualitative (Delphi study)	Five domains of Felce and Perry	NA	NA	NA
Petry	2007b	To examine which support characteristics are associated with QoL of people with PIMD	n=42, age <18; n=34, age>18	Qualitative (semi structured interviews)	NA	NA	NA	NA
Hartnett	2008	To explore the contributions of day programmes to QoL	n=8, age 21-32	Mixed method (interviews, questionnaires)*	Eight domains of Schalock	Interviews and questionnaire	Parents and staff	NA
Petry	2009a	To evaluate different dimensions of QoL of people with PIMD and the association between their QoL and personal and setting characteristics	n=49, mean age 23.7 year	Quantitative (questionnaires)	Six subscales, based on five domains of Felce and Perry	QoL-PMD: questionnaire	Parent, direct staff, indirect staff	NA
Petry	2009b	To examine the possibilities for item reduction and the psychometric properties of the QoL-PMD	n=49, mean age 23.7 year	Quantitative (further development of questionnaire)	NA	NA	NA	Internal consistency, construct validity
Petry	2010	To evaluate the psychometric properties of the Mood, Interest and Pleasure Questionnaire	n=360, age>18	Quantitative (questionnaires, behaviour checklist)	Mood, Interest and Pleasure	MIPQ: questionnaire	Direct support staff	Test-retest, inter-rater reliability

Table 1. Continued

1st author	Year	Objective	Persons with PIMD	Study design	Components of QoL	Instruments or methods to assess QoL PIMD	Assessors	Psycho-metric properties
Vos	2010a	To measure a non-interpretative measure of subjective well-being, that is, how the respiratory, cardiovascular and electro dermal response system associate with the valence dimension of emotion	n=3, age 23, 23, 52	Mixed method (video observations, physiological measurements)*	Emotions (physiological)	Physiological measure and behavioral observations, video tape	Direct support staff and researcher (unfamiliar)	NA
Vos	2010b	To explore the client and service characteristics contributing the subjective well-being of persons with severe and profound ID measured by the Mood, Interest and Pleasure Questionnaire	n=360, age>18	Quantitative (questionnaires)	Mood, Interest and Pleasure	MIPQ: questionnaire	Direct support staff	NA
Vos	2013	To examine the relationship between mood and emotions in people with severe and profound intellectual disabilities	n=27, mean age 34.5	Mixed method (video observations, questionnaires)	Mood and emotion	Experiment using behavioural observations, video tapes and MIPQ: questionnaire	Two direct support staff members and independent researcher (unfamiliar)	NA
Bigby	2014	To explore the conceptualisation of good outcomes and support in terms of QoL and staff practices	n=37, age>18	Qualitative (participant observations)	Eight domains of Schalock	Qualitative indicators extracted from participant observation	Author and research fellow	NA

Table 1. Continued

1st author	Year	Objective	Persons with PIMD	Study design	Components of QoL	Instruments or methods to assess QoL PIMD	Assessors	Psycho-metric properties
Verdugo	2014	To provide reliability and validity evidence of the San Martin Scale	n=1770**, age>16	Quantitative (questionnaires)	Eight domains of Schalock	San Martin scale: questionnaire	Parent, family or direct support staff	Reliability and construct validity
Gomez	2015	To contribute to the knowledge and understanding of QoL by developing a set of QoL indicators	NA	Qualitative (Delphi study)	Eight domains of Schalock	NA	NA	NA

Note:

NA, not applicable; PIMD, Profound Intellectual and Multiple Disabilities; QoL, Quality of Life;

* mixed method design but very small sample;

** most participants had PIMD.

Which instruments/methods were used and who determined QoL?

Six instruments and assessors.

Five questionnaires determining QoL in persons with PIMD were found, as well as one instrument using observations and interviews, which are presented in Table 1. According to the authors [27], the QUOLIS measured both objective and subjective components. The questionnaire, filled in by an interviewer, was based on questions posed to a family member, friend or staff member. The QoL-PMD is used to exclusively measure objective components of QoL [18]. This questionnaire is completed by a parent, direct staff and indirect staff. As the authors noted [15] [20] [22] [33], the MIPQ only measures subjective components and is filled in by direct support staff. However, the other questionnaires, the QUALIN, based on interviews with parents [35] and the San Martin Scale, filled in by parents, family or direct support staff [19], did not distinguish between subjective and objective components. The development of the QoL-PMD was based on interviews with parents and professionals [24] and on a Delphi study [25], in which experts selected items based on the domains of Felce and Perry [26]. Most questionnaires were based on a combination of several domains (see Table 2).

Table 2. QoL domains used by different authors

Author	Domains
Schalock	Emotional well-being, interpersonal relationship, material well-being, personal development, physical well-being, self-determination, social inclusion, rights
Felce and Perry	Physical well-being, material well-being, social well-being, development and activity, emotional well-being
André	Psychological conditions, physical conditions, familial context, abilities to communicate, abilities to stay alone
Ouelette	Health services, family guardianship, income maintenance, education training and employment, housing and safety, transportation, social and recreational services, religious/cultural supports, case management, aesthetics, advocacy, counselling

The Life-Satisfaction-Matrix (LSM, [14]) combined participant observations and semi-structured interviews, which assessed the subjective component “satisfaction with life.” The assessors were the most familiar communication partner, the next most familiar communication partner and one unfamiliar partner. The LSM is based on four assumptions for persons with PMD [14], p. 767:

- (a) for persons with PMD, life satisfaction is improved when more time is spent on preferred activities and less on non-preferred activities;
- (b) individuals with PMD express their inner states through consistent behavioural repertoires;
- (c) these behavioural repertoires can be discerned by familiar others, and validated by an independent other;
- (d) an individual’s routine daily activity preferences can be determined from their affective behavioural repertoires.

Other methods and assessors.

To determine subjective components of QoL, most studies used behavioural observations [16] [22] [31] [36]. Green and Reid [36] and Favell et al. [31], for example, selected observable responses (including facial responses such as smiling) to define happiness. This behaviour was observed by supervisory staff and unfamiliar observers. In the study of Petry and Maes [16], staff members identified expressions of pleasure and displeasure and found out that persons with PIMD especially used sounds and facial expressions to express pleasure and displeasure and positive and negative moods. In three qualitative studies [12] [13] [17], in-depth interviews with parents and staff and participant observation were used. Through in-depth interviews, Petry et al. [17] examined which support characteristics were associated with QoL in persons with PIMD. Characteristics with regard to the structure of the setting, such as group size and staff turnover, and with regard to the organisation of the support, such as interdisciplinary, played a central role.

Methodological pluralism and triangulation of assessors.

Answering the question which instruments/ methods were used and who determined QoL, we found that Petry and Maes [16] used methodological pluralism and assessor triangulation. They used several methods to determine QoL in persons with PIMD, such as behavioural observations and questionnaires (methodological pluralism). Triangulation of assessors means determining QoL in persons with PIMD by two familiar observers and a third unfamiliar observer. Although parents and proxies mostly were the assessors of QoL, in five articles their role was mentioned as a limitation [13] [17] [18] [19] [36]. These studies referred to research on other groups, in which proxy outcomes differed from self-assessments [37] [38] [39].

In their study, Petry and Maes [16] used triangulation in the following way: first, staff members identified expressions of pleasure and displeasure, and then parents and direct support staff filled in a questionnaire about how the person expressed these. An independent researcher then coded behavioural signals. Vos et al. [22] used two members of the direct support staff to identify behaviour expressing both positive and negative emotions (affective profile), followed by an independent researcher coding the emotions. For an overview of persons determining QoL in persons with PIMD, see Table 1.

Discussion

In line with the findings of Petry and Maes [10] in their overview chapter, we found no direct answer to our first research question on which concepts were used. We did find that most authors referred to the theory of Schalock and the conceptualisation of the SIRG QoL, a result of a two-year consensus process with a panel of experts [8]. We found several combinations of domains as well as references to objective and subjective components. However, of the four elements of the SIRG conceptualisation described in the articles, one was not explored in the included studies, namely the one stating that QoL is enhanced by self-determination, resources, purpose in life and a sense of belonging.

However, in persons with PIMD, self-determination and purpose in life are concepts that can raise questions due to their developmental level ($IQ < 20$). If someone's IQ is low, does this mean that their QoL is low? The answer to this question is of moral importance because it implies a devaluing of the lives of persons with PIMD cf. [40]; cf. [41]. On the other hand, it may be worth examining in clinical practice and in research what self-determination, purpose in life and being a person (could) mean in everyday life of persons with PIMD. Reinders [42] did this from a theological and anthropological point of view. In this vein, Petry and Maes [10] described self-determination in relation to having choices and control over activities and environment. Furthermore, Vlaskamp [43] developed an educational program for children with PIMD in which the assumptions were that persons with PIMD had an influence on their environment and were able to develop meaningful relationships with others.

Another issue in the included articles and in the overview of Petry and Maes [10] was the distinction between objective and subjective components of QoL. However, the included studies used different distinctions, which is confusing. Vos et al. [20] [22]

included emotions in the subjective components of QoL, whereas the QoL-PMD includes emotional well-being in the objective components [18] [24] [25]. Furthermore, in order to measure subjective well-being, such as pleasure or happiness, researchers often used assumptions. For example, Lyons [14] assumed that preferred activities improve life satisfaction and therefore QoL. However, we doubt the adequacy of this assumption because persons with PIMD may behave in a stereotypical way and prefer a certain behaviour [44], like playing with (their own) hands or compulsively needing a specific toy. Although they seem to prefer this behaviour, it may prevent them from gaining (new) experiences that could improve their QoL [44]. Moreover, stereotypical “preferred” behaviour (such as self-injury) could indicate a low QoL. Another example is the assumption in Green and Reid [36] that smiling reflects happiness. We find that this assumption needs more reflection. In fact, smiling can be a side effect of an epileptic seizure with no relationship to happiness. Additionally, laughing and smiling can be syndrome related, as with Angelman Syndrome [45]. If even parents or direct staff are unsure how they should interpret the behaviour of the person with PIMD, this may be even more difficult for distanced observers.

Based upon the conclusions of Petry and Maes [10], we agree that assessing QoL in persons with PIMD is a challenge for researchers as well as caregivers. We think that one solution to this challenge may be an approach that recognises the importance of interpretation. For example, Goode and Hogg [46] saw gaining information about QoL in persons with PIMD as an art or craft using careful judgment combined with empathy. They articulated that information can neither be easily operationalised nor described in a written document. Such an approach recognises the value of QoL in persons with PIMD without trying to objectify it. The importance of interpretation is also recognised in narrative approaches [47]. Methods like these should be developed further in future studies.

Another issue is triangulation and the use of proxies. When determining QoL in persons with PIMD, researchers and proxies should be critical about their own assumptions and interpretations of behaviour. In this regard, it is worth referring to Petry and Maes [16] who aimed at methodological pluralism and triangulation to confront these difficulties. We found that triangulation was used by other researchers as well. In most studies, we found that parents or direct support staff assessed or determined QoL in persons with PIMD, sometimes as one of the partners in triangulation. Some authors argued that the person who is closest to the person with PIMD should determine his or her QoL [27] [35], whereas many studies consider using a proxy a limitation. These different opinions on the use of proxies (such as

parents) require attention, especially because parents play a significant role in decision-making [1]. More in-depth research on the role of parents and proxies in assessing QoL when making difficult, life-altering decisions is necessary.

In accordance with the SIRG QoL, the concepts found in the included articles were not only used to describe the QoL of persons with PIMD, but they were also used with the strategic aim to be a change agent, with the main aim to improve quality outcomes [9]. This is confirmed by Petry and Maes in their overview chapter of QoL in persons with PIMD [10]. According to them, the main purpose of measuring QoL is to improve QoL by detecting circumstances that decrease it and make changes based upon those findings. Therefore, they consider the use of the concept of QoL in end-of-life decisions very undesirable. “By adopting this value as a central tenet in QoL measurement, the application of QoL in ethical decisions on life and death or in refusing to treat certain people is discarded as an abuse” [10], p. 26. However, as recent research shows, QoL is certainly related to the decision-making processes in persons with PIMD, including end-of-life decisions [1].

The strengths of this study are its systematic research and explicit findings of the different uses and views on concepts in research on QoL of persons with PIMD. A limitation of our study is that the included studies were performed mostly in Western countries, for example, in Belgium and the Netherlands, which may limit the generalisability of the findings and requires further research in other contexts.

Conclusion

This review provides insight into several components of QoL in persons with PIMD and in the tools used to assess them, questionnaires and behavioural observations in particular. Methodological pluralism and triangulation of assessors with varying closeness in relation to the person with PIMD are often considered good assessment elements. However, we questioned some elements of the conceptualisation including the following: the difference between objective and subjective QoL, as well as underlying assumptions of behavioural observations. Instead of trying to objectify QoL in persons with PIMD, we argue that the existence of interpretation is recognised.

Research on QoL in persons with PIMD remains a challenge because of their incapability of expressing themselves verbally. To confront this challenge, in their overview chapter Petry and Maes [10] argued for more valid and reliable research

methods to measure QoL in persons with PIMD. We think that a narrative approach, which recognises both the role of interpretation and the storied nature of who a person is, can be helpful in describing QoL in persons with PIMD, especially when we extend QoL to medical decisions in persons with PIMD. In these decisions, the role of parents is essential.

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Het levensverhaal van Britt

Vakanties

“Zeker in haar beginjaren namen wij Britt altijd overal mee naar toe op vakantie. Van vakanties in eigen land o.a. op het strand van IJmuiden, meerdere keren naar Ibiza en Spanje tot skivakantie in Oostenrijk waar ze echt mee de berg op ging in een zelfgemaakte “slee”. Zij kon daar zo van genieten. Maar ook vele uitstapjes gedaan met haar. We gingen ieder jaar met Britt naar de hele bijzondere Dreamnight in Artis, ze ging op kamp met Omega, meerdere keren naar Bio Vakantieoord in Arnhem en eenmalig naar Villa Pardoes in de Efteling. Op latere leeftijd ging het echter allemaal wat minder makkelijk om haar mee te nemen. Er waren steeds meer voorzieningen voor haar nodig en ze werd steeds groter én zwaarder. Maar ze heeft vele mooie reizen, fijne vakanties en heerlijke uitstapjes meegemaakt waar ze voor de volle 100% van heeft genoten. En wij dus ook!! Dit heeft zeker een grote positieve bijdrage geleverd aan haar kwaliteit van leven.”

*Nancy & Henk, moeder & vader van Britt,
en Lars en Suze, broer en zusje*



Chapter 3

Chapter 3

Physicians' perceptions on Quality of Life of persons with profound intellectual and multiple disabilities: A qualitative study

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Abstract

Background

Quality of Life (QoL) may be a factor in physicians' decisions on treatment of persons with Profound Intellectual and Multiple Disabilities (PIMD). We explored the perception of physicians on the QoL of persons with PIMD.

Method

We conducted semi-structured interviews with seven experienced physicians treating persons with PIMD, thematically analysed them and identified clusters of related elements of QoL.

Results

Emotional, physical and relational clusters of elements were found. Physicians named mainly emotional elements to describe good QoL, physical elements to describe poor QoL and they emphasised the role of relational context on QoL. In addition, they mentioned a balance between good and poor elements as good QoL. Physical decline, bad prognosis and increasing problems in the relational context influenced QoL in a negative way.

Conclusions

The influence of the relational context on both good and poor QoL needs more investigation because of its consequences for ethical decision-making.

Introduction

Quality of life (QoL) of persons with Profound, Intellectual and Multiple Disabilities (PIMD) is a challenging subject in research, because these individuals cannot verbally express themselves. Persons with PIMD are individuals with profound intellectual disabilities (IQ <20) and profound neuromotor dysfunctions, sometimes with sensory impairments and even medical problems (including seizures, respiratory and feeding problems). Even more, these persons seem to have neither an apparent understanding of verbal language nor symbolic understanding [1]. This means, for example, knowing that a chair is something to sit on. This description of persons with PIMD is in accordance with the working definition of persons with PIMD developed by the Special Interest Research Group (SIRG) PIMD of the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD).

In research, QoL is mostly used as an outcome measure for interventions or quality of care. In a narrative literature review [2] we found that questionnaires, for example the QoL-PMD [3] [4] and San Martin scale [5], and behavioural observations [6] [7] were frequently used as methods to assess QoL. Parents, family, primary caregivers and researchers were the assessors of QoL. However, QoL is not only used as an outcome measure, but also affects ethical deliberations on major surgery, tube feeding, and with-holding or sustaining life-supporting treatment [8] [9]. In recent years, more research has been conducted on ethical decision-making in clinical practice. For example in end-of-life care in persons with intellectual disabilities [10] or in decisions concerning children with neuro-disabilities [11]. A recent study explored the attitude of physicians to life-threatening events in children with severe neurological disabilities in Switzerland [12]. However, in our literature review we found that knowledge on the role of QoL of persons with PIMD in ethical deliberations was scant [2] and some authors say that the use of the concept of QoL in ethical issues is precarious [13]. In clinical practice, however, ethical deliberations to make decisions on major surgery, tube feeding, and withholding or sustaining life-supporting treatment do occur and in the Netherlands, physicians bear the final responsibility for these decisions. These decisions can feel onerous for those who are responsible for persons with PIMD, such as parents and physicians [8] [14].

Recent research on clinical decision-making in persons with PIMD identified elements that contribute to QoL according to physicians [9]. Apart from this, there is

not much information on physicians' perceptions of good and poor QoL of persons with PIMD and when they start to have concerns about QoL. More specifically, we do not know what the tipping point is at which they start to have concerns and for what reasons they have concerns about QoL of persons with PIMD. Therefore, we conducted this qualitative study in which we explored physicians' perceptions on QoL of persons with PIMD. We answered the following research questions: 1) How do physicians describe good and poor QoL of persons with PIMD? 2) When do they start to have concerns about QoL of persons with PIMD?

Method

We performed an exploratory study using semi-structured interviews with physicians and analysed them thematically [15].

Recruitment

We included paediatricians, paediatric neurologists and intellectual disability physicians because they are the first responsible physicians of patients with PIMD in the Netherlands. Paediatricians and paediatric neurologists work in hospitals and are the first responsible physicians for persons <18 years with PIMD living at home. Intellectual disability physicians, mostly working in institutions, are the first responsible physicians for persons with PIMD living in these institutions. Only physicians with more than 5 years of experience in treating persons with PIMD were included. Sampling aimed at the variation in age of the patients they cared for. We recruited the participants by opportunity and snowball sampling with the help of experienced physicians and psychologists who are on our project group, comprising the authors, three psychologists and a paediatrician. They searched in their networks for physicians in different parts of the Netherlands who met the requirements.

Research ethics

According to the Ethical Review Committee of the Academic Medical Center, our study did not need an ethics review. Eligible participants were informed about the research by telephone and by letter and when they agreed to participate, we sent them a letter to confirm this.

Data collection

The first author conducted the interviews using an interview guide (Supplementary file 1). This guideline was discussed in the research group until we reached consensus. The interviewer started the interview with the description of persons

with PIMD. Then we asked the physicians to describe a patient with PIMD whom they deemed to have a good QoL and a patient with poor QoL and why. After that, we asked the physicians when they started to have concerns about the QoL of a patient with PIMD and to give examples. All the interviews were audiotaped and transcribed verbatim. The face-to-face interviews took place in the hospital or institution in which the physician worked and took approximately 1 hour each (mean, 65 min; range, 57–73 min).

Data analysis

The transcribed interviews were analysed using the qualitative data analysis software, MaxQDA. First, two interviews were independently analysed by AMN and EO to identify fragments relevant to answer the research questions. Then, the first author used a mixed strategy with codes derived from the interview guide, and codes that emerged from the data, such as emotions of the interviewee and balance, leading to a first code tree (Supplementary file 2). After that, AMN discussed this with EO and DW and they regrouped the codes thematically and added an explanation, leading to a second code tree (Supplementary file 3). Next, two (other) interviews were coded by AMN and EO and two members of the project group (EH and GG) using the second code tree and discussed the results.

After that, AMN interpreted the fragments belonging to the codes and discussed the results with EO. Based on this discussion the analyses were refined. For example: the elements of good and poor QoL were first clustered in four different clusters: Emotional, Physical, Relational and Contact. After discussion, Relation and Contact were combined in the same cluster. The final analysis was discussed between AMN, EO and DW until they reached consensus.

Participants

We contacted 8 physicians (2 paediatricians, 2 paediatric neurologists and 4 intellectual disability physicians), all with 5 years or more experience – varying from 5 to 36 years (mean, 16; SD, 11) – with the target group. One paediatric neurologist cancelled participation because of too much work. The 2 paediatricians and a paediatric neurologist who participated, worked in different academic hospitals and were specialised in the target group. The 4 intellectual disability physicians all worked in care organisations for people with Intellectual Disabilities (ID), targeting various subgroups. One of the intellectual disability physicians also worked in a children's day-care centre for the target group. The mean age of the participants was 50 years (SD = 12).

Results

Hesitations of physicians to describe QoL in persons with PIMD

In the interviews with the physicians, all were hesitant to speak about QoL of persons with PIMD. Some were hesitant because they felt that QoL in this target group depends solely on subjective interpretation and mentioned that persons involved with persons with PIMD could have different ideas on QoL. Or the participants mentioned that there was no “standard” for QoL and that QoL of persons with PIMD is not the same as in other persons [numbers refer to participant numbers].

P4: Yes, that is dangerous, because what is discomfort in this group? Yes, [he] has epilepsy. Is motor unrest discomfort? We all think that it is, but – in my opinion – we are unable to interpret it specifically in this group.

P1: Of course quality of life is totally different to yours or mine, these are not [normal] children, they will never become autonomous, they will never go to school or university.

Participants also mentioned that their perception of QoL of a person with PIMD was influenced by the relationship they had with this person.

P5: But if one of my colleague's patients is admitted during the weekend and I think to myself “this child does not have any form of communication with his/her surroundings and is just lying in bed and now he/she is in hospital again with [...]”, then I do think “Gee, is this really relevant?” Because I am looking at him or her as a stranger.

P1: That is the way in which he [intensive care specialist] looks at it: he sees a very, very, very sick child that is very retarded entering the hospital in a resuscitation setting and has no concept of what the child's life was like before this moment.

How do physicians describe good and poor QoL of persons with PIMD?

The analysis of the elements of good and poor QoL led to three central clusters: emotional, physical and relational. For an overview of these clusters and their elements, see Table 1.

Table 1. Three clusters of elements of Quality of Life (QoL)

CLUSTER	GOOD QOL	POOR QOL
EMOTIONAL	Happy, pleasure, enjoyment	Inconsolable crying, appears unhappy, no emotion, empty, shallow, suffering
PHYSICAL	Normalised muscle tone, no pain, no epilepsy or under control, can eat without choking, is able to breathe easily	Tense muscles, choking, short of breath, pain, major epilepsy, sick
RELATIONAL	Is able to make contact and has interaction. Good relationship with parents, parents/family are able to cope, loving family, loving social environment, safety	No contact. No parents, burden too great for parents /social environment, no safety

Emotional cluster

Participants regularly mentioned emotions or feelings when they described good QoL. For example, pleasure and enjoying things or being satisfied were mentioned frequently to describe good QoL. As one of the respondents (P6) said: 'Well, that person is enjoying life, he appears content'. Conversely, they also mentioned emotions or feelings when describing a patient with poor QoL, although this was less frequently referred to.

P6: He can barely be activated or motivated to do anything, he no longer appears to enjoy anything.

Physical cluster

On the other hand, physical elements were mostly mentioned to describe poor QoL. Physicians listed physical elements such as epilepsy, pain, tightness of the chest and problems with feeding or choking, or both. Also, hospitalisation because of the physical problems was mentioned frequently as a negative element. A few physicians described good QoL, as the absence of negative physical elements.

Relational cluster

Besides emotional and physical elements, respondents often referred to relational elements. One of the elements was familial aspects and this was stated as being very important, like a good relationship with parents, which gives the person with PIMD safety and the capability and resources of parents to shoulder the burden of caring for their child.

P7: Why I think that she has a quality of life? She lives in a home situation with a family with younger children, with the parents who care for her with a great deal of love, and where she feels wanted. She is allowed to be who she is. This was a long road for her parents, but now she is allowed to be who she is.

One of the physicians specifically mentioned the safety given by the parents and the social environment as being crucial for good QoL.

P4: For example, the child experiences safety. There is a very clear ... The child has a loving family around her. Or care providers, or whatever. With continuity. If that is missing, yes that does affect you ... What you want most for a child, is safety. That is what it is really about. A child has to feel safe somewhere. A child has to feel safe with certain individuals. Normally this involves the parents, but if there are very regular care providers, then as far as I am concerned, the same applies. But this is about safety and feeling safe.

Some participants stated that the social environment of the person with PIMD was essential to their existence.

P5: Yes, that does define the lives of others. And yes, I do think that you never look at one life on its own ... And in this case ... this category of individuals and children, it is so, you cannot focus on that child alone. You simply have to look at the whole picture

Interviewer: So also at the social environment? ... Interviewee: Also at the social environment. I mean, we are nothing with-out our surroundings. Meaningless beings on a meaningless planet. I am truly convinced of this. I mean, by virtue of the fact that we know what we can expect from each other, yes.

P1: ...these children cannot make their own quality of life...you need someone who is fully committed to the child.

Balance or equilibrium

When answering the question of describing a person with good QoL and a person with poor QoL, sometimes physicians described QoL neither as completely good nor as completely poor, but rather in terms of an equilibrium or balance between good and poor elements.

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P4: And the second point is that I think that it is very much about an equilibrium between the positive things in life and the negative things in life. I also think that if you have relatively few negative things, then there will also not be many obviously positive things ... If you start looking ... But if there is a lot of suffering, problems, etc., that you need a lot of positive things to maintain that equilibrium.

Or they spoke of good QoL in spite of the patient's poor health or in spite of the fact that not all elements were positive.

P3: And that is someone with quite severe physical limitations, with physical problems and despite this she appears happy to me. And as far as I am concerned, she does have quality of life.

The other equilibrium that they mentioned as good QoL, which they referred to as "harmony" or "balance", was an equilibrium between the person with PIMD and the environment.

P5: Yes but, you know, if you base things on the idea that the child has developed a sort of harmony, with the parents, within the family, with other children, at the day-care centre; if that is all in harmony, then it does not really depend on the [cognitive] level of the child.

Concerns

Physicians started to have concerns about QoL when there was a change of the situation in a negative way.

P7: This boy, who you only see deteriorating – he was able to walk, but can now no longer walk ... this is also a boy who is always grimacing, has few moments of enjoyment and in the end was not able to eat very well. So that was all deteriorating too, he used to be able to enjoy his food, but – possibly due to the medication he was receiving for his epilepsy – he started having trouble swallowing, so we had to switch to pureed food ... which he did not like!

P2: And yes, also the fact that she is getting older, has all sorts of ailments, is experiencing pain, is fatigued, yes, I think she is no longer experiencing a good quality of life.

Participants particularly referred to negative changes in the physical and the relational cluster.

Negative change in physical cluster

Some of the participants addressed the shortening of intervals between hospitalisations or between prescriptions of medication.

P6: Yes, if it becomes in and out, in and out, at shorter intervals, then yes ... we happen to have a girl at the moment who goes back and forth to hospital and that makes me think, because that is a brain stem dysfunction, then I think yes ... well [...] Wow, look if there are 3,

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4 courses of treatment per year and with a short interval, then you do think gosh, now I am at this point again ... well, yes then I do start ... And at some point you can say listen, this person is getting respiratory tract infections very frequently – as I just said – or a choking episode [...] or something similar and you give them one course of antibiotics after the other, then you do start to think. Then you do think, yes, isn't it time that we all get together?

In addition, almost all physicians referred to the process of physical decline or having a bad prognosis as factors that influenced their perception about QoL of persons with PIMD in a negative way.

P2: Well, first you start with the idea of when to start such a conversation. And that is often related to the fact that a deterioration has occurred. Deterioration that you think will become progressive.

P3: I do think that the prognosis was bad, considering the danger of choking and the risk of repeated bouts of pneumonia. So in that sense, the prognosis was not favourable.

Negative change in the relational cluster

Some physicians spoke about the decline of the strength, resources or abilities of parents to care for their disabled child.

P1: Too great a burden on the family, they were simply unable to cope, they were caring for this girl literally day and night, she was also in pain, was unable to eat properly, was being bottle-fed and ultimately her surroundings – the sub-optimal environmental factors, the parents' inability to cope – also contributed to the fact that this girl died, because her family simply could not carry on.

In all the interviews, physicians mentioned the death of parents, or the fear of parents to die before their child. Some mentioned that the death of the parents has a negative effect on QoL of the person with PIMD. Within this context, one of the physicians mentioned his wish for parents to outlive their child.

P5: But I do think that – even with a child that is very severe [disabled] – it can play major part in your life, to go through that together with that child, you want to see it through, yes I do think that. Interviewer: What do you mean “see it through”? Interviewee: Well that you are also there at the end. Interviewer: As a parent? Interviewee: And that as a parent, you should not give up at the end and I think that – though of course you are not able to make this choice – just like with healthy children, no parent wants to see their child die before them. Is it actually the other way around with these children, that you say well the journey has been completed now.

Discussion

This study addressed how physicians perceived QoL of persons with PIMD and an emotional, physical and relational cluster were found. In addition, they mentioned a balance or equilibrium between elements as good QoL. The findings suggest that the tipping point to start concerning about QoL was a negative change, mostly in physical elements, such as physical deterioration, or in the relational context, like parents not being able to cope anymore, or when there was a disturbance in the balance/equilibrium.

In the literature, QoL is frequently described in domains. In a literature review, we found several combinations of domains [2]. In our present study, the clustering of elements was an inductive process, based on interviews with seven physicians. However, some of the elements we found were also present in the different domains described by others, such as familial context and capability to communicate [16], basic security and family bonds [17] and emotional well-being [18] [19]. On the contrary, other domains were not mentioned by our interviewees, for example: self-determination, social inclusion and rights [19]. The physical cluster with elements of physical state and health is described in all combinations of domains that we found in the literature [2]. Our findings are also in line with the findings described by Zaal-Schuller, et al. [9].

The importance of physical elements or physical health on QoL is comprehensible, because of the complex medical problems of persons with PIMD. However, when they described good QoL, our respondents did not mention physical elements, but mostly emotional and relational elements. Conversely, when they described poor QoL, they mentioned physical elements. Furthermore, when they started to have concerns, one of the aspects they mentioned was a deterioration in the physical cluster. In this negative change, they frequently mentioned decline and a bad prognosis of the physical state or health as factors that influenced their judgment. One explanation for this finding may be that physicians often see their patients when they suffer from physical complaints or physical deterioration. Another factor they mentioned was the frequency of hospitalisation and the prescription of medications. Information on hospital admission of persons with PIMD [20] and prescription of medicine to persons with PIMD is available [21], although not related to QoL of persons with PIMD. In addition, the literature on the parental burden of care and

QoL of parents and their hospitalised children [22] confirms our findings that hospitalisation has an effect on QoL.

Our respondents emphasised the effect of the relational context (particularly the parents) on QoL of persons with PIMD, which included their comments on the death of the parents. The literature also revealed that the parents are often one of the assessors of QoL of persons with PIMD [2]. This can complicate matters, particularly in situations where the QoL of the person with PIMD plays a role in decisions regarding medical treatment [9]. On the one hand, the parents are the assessors of the QoL of their child, while on the other hand they are an important factor that influences this QoL. This opinion about the mixed roles of family members in medical decision-making has been described in a different context as well [23]. However, in these cases, the patients had been able to discuss their wishes and ideas about QoL before they became ill. This is not the case for persons with PIMD. Parents, or others involved, always have to make the crucial decisions in life for the persons with PIMD. Whether this is more difficult or perhaps easier is not known. Further research is necessary to explore what the perspective of parents is on these different roles.

Some of our respondents stated that persons with PIMD cannot exist without their relational context. This is understandable, because of their total dependency on others in all aspects of daily life. As we found in the literature that QoL (and thus relational context according to these physicians) affects decisions [9], this importance of the relational context can raise moral questions. These moral questions deserve further attention because they may be (highly) controversial in the field of care for persons with PIMD. For example, may or should relational context play a role in decisions on major surgery, tube feeding or withholding or sustaining life-supporting treatment? May or should the absence of a warm and loving environment influence these decisions? Or may or should the fact that parents have passed away influence these decisions? In order to answer these questions, the perspective of family, parents and professionals on the impact of the relational context on QoL of persons with PIMD is worth exploring further in future research.

In this study, we asked seven physicians to describe QoL of persons with PIMD. We found a reticence in our respondents to answer this question. They realised that their perception on QoL of persons with PIMD is subjective, based on interpretation and dependent on their relationship with the patient. The current assessment methods [13] may not be the (only) good response to this hesitation. An alternative response may be the development of narrative approaches to QoL because they

recognise the importance of interpretation and relationship when interpreting someone's QoL.

Furthermore, we found that our participants balance elements of good and poor QoL, and that they start to experience concerns when this balance tends to become negative. This information was obtained from one group of informants of QoL, namely physicians. If and when other informants of QoL of persons with PIMD – such as parents, family and professionals – start to have concerns about the balance of good and poor elements, QoL warrants further investigation.

The strength of this study is that it reflects the struggle of our respondents to assess QoL in a justifiable manner in persons with PIMD, which may offer possibilities for identification for other physicians going through similar processes. A limitation of this research is that only seven physicians were interviewed. Another limitation of this study is that the results, though they may be transferable, cannot be generalised to other settings or cultures.

Our study indicates that physicians may balance good and poor elements when they describe QoL of persons with PIMD and may start to have concerns when physical elements or the relational context deteriorate. The influence of the relational context on QoL and its consequences in ethically difficult decisions needs more investigation.

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Supplementary file 1. Guideline for interview, list of topics interview questions.

In this series of interviews, we will interview physicians.

A. We need to define the target group before we start. The interviewer and the interviewee need to agree on this. Nakken and Vlaskamp defined the group of individuals with PIMD. An important characteristic is: being unable to speak or express themselves clearly in any other way.

The persons who form the subject of our research (persons with PIMD) have the following characteristics;

(a) profound intellectual disability and (b) profound motor disability. They also have a number of additional severe or pro-found secondary disabilities or impairments. In addition to profound intellectual and physical disabilities, sensory impairments are also frequently experienced and especially the prevalence of cerebral visual impairment is high. Individuals with profound multiple disabilities also have an overall risk of developing medical complications and almost all require regularly administered medication (e.g., anti-convulsion drugs, sleep medication, medication to prevent reflux and drugs with a sedative effect). They form a physically vulnerable group of persons with a heavy or total dependence on personal assistance for everyday tasks [1].

B. It is then important to determine, the involvement of the interviewee with the aforementioned target group. How many of these individuals does the interviewee know and how often does he/she see them? What age are these individuals and in what type of setting do they live?

We have two research questions that we will discuss:

Research question 1:

How do physicians describe good and poor quality of life in this target group?

Research question 2:

When and why would physicians start to doubt the quality of life?

It is important to ask the interviewee to think about an existing person/situation. This allows you to ask in-depth questions about tangible aspects. Avoid speaking in general terms!

A few guidelines for the interviews:

Concretise:

Can you give me an example of this? In which situation does this play a role?

Challenge:

[literally repeat a few words of what respondents say]. Can you tell me more about this? How was that for you?

Clarify:

what do you mean by ... ? I am not sure whether I completely understand what you have just said. Could you tell me a bit more about ... ?

Summarise:

so if I understand correctly, you are saying ...

Take the lead: excuse me, but I have to interrupt you there. What I would like to know is ... I am really glad that you have said this, but I would like to return to the topic ...

Personal: I can imagine that. That is quite intense, very beautiful, etc. (be careful here, as this can also determine the direction of the interview)

The interview

1. Introduction

Introduction

- introduce yourself and thank the person for his/her willingness to take part
- aim of the interview / course of events

Aim of the research

- By interviewing physicians, we want to gain an understanding of how they view the quality of life in this group and what their considerations are in this regard.

Factual details

- name
- age
- m/f
- profession
- how long have you been working with this target group and in what role, what age are they, where do they live.
- cultural background
- telephone number and/or e-mail address

Course of events

- start the interview
- record any unusual observations. Things that are not recorded on tape, but could be useful in the analysis of the interview.
- The interviewer should manage the time
- evaluation at the end (would like to know what you thought about the interview)

Any questions?

Research question 1

Can you describe a person from the group that we have just defined that you would say has a good quality of life?

- Describe the situation
- What was the reason that you thought this?
- What did you notice about the child/the adult?
- Did any other factors play a role? For example, the living situation, the parents or professional care providers

Can you describe a person that you would say does not have a good quality of life?

- Describe the situation
- What was the reason that you thought this?
- What did you notice about the child/the adult?
- Did any other factors play a role? For example, the living situation, the parents or professional care providers

Research question 2

Have you ever had doubts about the quality of life of a person from the target group? If so, can you describe someone?

- Describe the situation
- What was the reason that you thought this?
- What did you notice about the child/the adult?
- Did any other factors play a role? For example, the living situation, the parents or professional care providers
- How did you handle these doubts?
- How was that for you?
- What happened next, such as conversations with parents or other doctors?
- Were any decisions made?

Conclusion

- Would you like to receive a literal transcript of our research? (with all the “uhms” and “ahs”); (you can still add information to this version)
- Are there doctors or other people that you think would be worth interviewing? Perhaps because you know that they have a very different opinion or are involved in this subject matter in a different way. Do you have any names?

Supplementary file 2

Main code	Sub code
<i>Emotion of interviewee</i>	
<i>Weighing yes/no QoL</i>	balance concerns
<i>Ethics</i>	
<i>Process</i>	prognosis deterioration process characteristic
<i>Relationship</i>	dependence on others family situation safety relationship parents reciprocity environmental variables
<i>QoL PIMD</i>	QoL general happy relaxed enjoys eating joy comfortable observation QoL contact pleasure
<i>No QoL PIMD</i>	no contact epilepsy no joy inconsolable crying difficulty eating and drinking hospital admissions negative pain difficulty breathing discomfort suffering appears sad/unhappy sick behavioral problems

Supplementary file 3

1. *Yes QoL PIMD*
Description of situations in which QoL PIMD is good.
2. *Weighing yes/no QoL*
Quotes that exhibit concerns about whether or not the patient has good or poor QoL, or where a deliberation is made.
3. *No QoL PIMD*
Descriptions of situations in which QoL PIMD is not good.
4. *Emotion of interviewee*
Statements in which an emotion is mentioned. For example: "I find it difficult to see her crying like that" or "sometimes she struggles to breathe so much that it makes me feel awful and I think poor child".
5. *Determining QoL*
Statements that reveal how the respondent determines or uncovers QoL. For example: "I think that this must be unpleasant" or "I get the feeling/impression that this is unpleasant".
6. *Process*
Statements in which process characteristics, such as deterioration or a poor prognosis are mentioned.
7. *Relationship*
Statements in which QoL is related to relationships with other persons or relationships with a physical context.
8. *Ethics*
Statements in which an ideal is formulated; about good/not good; about rights and obligations; or about doing the right thing.
9. *Interesting statements that cannot be coded.*

Het levensverhaal van Britt

Sociale omgeving

“Praten met je lijf? Ja, hoor. Dat kan heel goed. En dat met geheel eigen geluiden ondertitelen. Britt leerde ons, de woorden voorbij, de zintuigelijke waarneming volop te benutten. Met haar alertheid was zij met haar tactiele en auditieve waarneming volop aanwezig in contact. Haar aanwezigheid en prachtige wijze van communiceren maakte dat iedereen altijd wel even contact met haar wilde. Ik ontmoette in haar een wijze vrouw. In het café, op de stoep, bij mij of bij haar thuis. Zij was er en kleurde ons sociale leven zoals alleen zij dat kon.”

Buurvrouw Christa en haar dochters Lotte en Sanne



“Wat hebben we veel met Britt op het terras en op de stoep met een drankje gezeten. Met Britt als groot voorbeeld dat gezelligheid niet af hoeft te hangen van praten maar van het genieten van met elkaar zijn. En dat bracht Britt naar het bewustzijn. Dank Britt, we missen je enthousiasme en gezelligheid!”

Buren Feyoena & Peter en hun dochters Sophie & Daphne



Chapter 4

The perspectives of professional caregivers on quality of life of persons with profound intellectual and multiple disabilities: a qualitative study

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Abstract

Persons with Profound Intellectual and Multiple Disabilities (PIMD) are supported by professional caregivers for a large part of their lives. However, the perspectives of professional caregivers on good or poor Quality of Live (QoL) of persons with PIMD have hardly been explored.

To explore the perspective of professional caregivers on QoL of persons with PIMD, we performed a qualitative study, conducting semi-structured interviews with eleven professional caregivers and thematically analysed them.

In the interviews, these professional caregivers described examples of good and poor QoL. Good QoL was mostly described in emotional terms such as happiness, pleasure and enjoying things. Poor QoL was mostly described in physical terms such as pain, sickness and shortage of breath. The capability of persons with PIMD to influence the environment was described as an element of good QoL. Furthermore, these professional caregivers described how they assessed QoL. They 'sensed' QoL, they tested it (trial and error) and discussed QoL of persons with PIMD with colleagues. Participants emphasised the dependency of persons with PIMD on professional caregivers. Furthermore, these professional caregivers described feelings of failure and powerlessness if they could not assess or fulfil the needs of persons with PIMD.

We conclude that these professional caregivers use their (tacit) knowledge to understand QoL of persons with PIMD. The relationship between QoL of persons with PIMD and professional caregivers needs more investigation, because of its consequences both for the persons with PIMD and the professional caregivers.

Introduction

Quality of Life (QoL) is a central principle in the support of persons with Profound, Intellectual and Multiple Disabilities (PIMD). Persons with PIMD are individuals with profound intellectual disabilities (IQ < 20) and profound neuromotor dysfunctions, sometimes with sensory impairments and even medical problems such as seizures, respiratory and feeding problems [1]. They are totally dependent on others in all aspects of daily life.

QoL of persons with Intellectual Disabilities (ID) has been a subject of academic interest since the beginning of this century [2] [3] [4] [5]. In accordance with Verdugo et al. [6], Petry and Maes [7] describe that QoL is used as a conceptual framework for assessing quality outcomes, as a social construct that guides quality enhancement strategies and as a criterion for assessing the effectiveness of these strategies. In addition, in a previous study was found that QoL of persons with PIMD also affects ethical deliberations on medical decisions [8]. In those situations not only perspectives about good QoL are important, also views about poor QoL, or concerns about QoL are relevant.

In a literature review, we found that QoL of persons with PIMD is often assessed through questionnaires and behavioural observations [9]. The assessors usually were familiar to the persons with PIMD, most often parents and professional caregivers. Some authors argued that triangulation between familiar and unfamiliar assessors was seen as a good assessment practice [7]. However, the assessment of QoL of persons with PIMD is difficult because these persons cannot describe their QoL themselves, nor can they confirm or deny the outcome of the assessment of their QoL.

Not only the assessment of QoL of persons with PIMD is challenging, also caring for them is difficult, because they cannot communicate when and how support should be given. In addition, caring for persons with PIMD is a heavy task for parents [10] and an informal network is often lacking [11]. Consequently, parents share their care with the professional caregivers of persons with PIMD [12]. Furthermore, because of their medical problems, medical professionals regularly care for them as well [13]. Research showed that the support offered by professional caregivers requires partnership with the parents [14], although a recent study showed that, in the view of mothers, professional caregivers often lack the specialist knowledge or skills

required to understand the needs of persons with PIMD [15].

Other research showed how the support offered by professional caregivers influences the QoL of persons with PIMD [16] [17]. Petry et al. [16] found out that, besides characteristics of the settings and the internal organization of the support, characteristics of the support staff itself influenced QoL of persons with PIMD. Furthermore, research explored the perspectives of professional caregivers on different domains of QoL of persons with PIMD [18]. Petry et al. [18] asked support staff to give examples of elements of good QoL and how QoL of persons with PIMD could be improved.

Hence, professional caregivers play an important role in the care of persons with PIMD and in their QoL. Not only are they one of the assessors of QoL of persons with PIMD, they also influence QoL of persons with PIMD. However, in medical decisions, not only good QoL or the improvement of QoL is important, but also views about poor QoL or reasons for concerns about QoL are relevant. Since professional caregivers may be confronted with medical decisions concerning persons with PIMD and the dilemmas associated with this, their perspective not only about good QoL but also about poor QoL of persons with PIMD is important. Therefore, the objective of this study was to explore professional caregivers' perception on good and poor QoL of persons with PIMD.

Materials and methods

We performed an exploratory study with a qualitative design, using semi-structured interviews with professional caregivers of persons with PIMD and analysed these thematically [19] [20]. A qualitative design was most appropriate because we searched for experiences and perceptions. In the analysis, we followed Braun and Clarke's phases: familiarisation with the data, coding, searching for themes, reviewing themes and defining themes [19].

Recruitment

Professional caregivers in the Netherlands are part of a team caring for persons with PIMD, mostly in residential care facilities and day-care centres. Some of the professional caregivers coordinate the care around the person with PIMD and are the primary person responsible for this care and our study focused on these professionals. They mostly have a pedagogical or nursing background and we used both purposive and snowball sampling to recruit them. We aimed to recruit

participants from both residential care facilities and day-care centres and we wanted to achieve a variety of age, both of the caregiver, and of the person they cared for. We approached psychologists and managers in the networks of professionals working with persons with PIMD and asked them to search for professional caregivers working with persons with PIMD in their institutions, who might be interested in participating in the study. One participant heard about the research through others and signed up. Participants were given detailed information on the study by the first author via email and when they responded that they were (still) interested, the first author called them. In this telephone conversation, we checked whether the participant was responsible for the care of persons with PIMD and whether the information about the research was clear. Once they agreed to participate, this was confirmed by letter and an appointment for an interview was made.

Research ethics

Our study has been conducted in accordance with the relevant passages in the World Medical Association Declaration of Helsinki. The Medical Ethics Committee of the Academic Medical Centre granted a waiver for our study because it was not subject to the Medical Research Involving Human Subjects Act. Professional caregivers gave oral consent after having read the information letter specifying participants' rights, like their right to withdraw from the study at any moment and the assurance of anonymity. We confirmed their oral consent in writing.

Table 1. Summary of the interview guide

<p>The focal question during the interview is: <i>When do professional care providers think that a person with PIMD has a good quality of life and when does a person with PIMD have a poor quality of life. Do they ever have concerns on the quality of life? Why?</i></p> <p>It is important to ask the interviewee to think about an existing person/situation. This allows you to ask in-depth questions about tangible aspects. Avoid speaking in general terms!</p> <p>2. The questions</p> <ol style="list-style-type: none">a. Can you tell me your first thoughts about “quality of life”, for the group that we are discussing?b. Do you know someone who has a good quality of life? Whilst thinking about this person, can you tell me more about them?<ul style="list-style-type: none">• Describe the situation• What do you notice about this person?c. Do you know someone who you think has a poor quality of life? Whilst thinking about this person, can you tell me more about them?<ul style="list-style-type: none">• Describe the situation• What did you notice about the child/the adult?d. Have you ever had concerns about a person’s quality of life?<ul style="list-style-type: none">• Describe the situation• Why did you think that?
--

Data collection

The interviews were conducted by trained interviewers (AMN, first author: 8; EO, supervising author, 2; EH, member of the project group: 1; GG, member of the project group: 1) using the same interview guide (Table 1). The face-to-face interviews took place at the workplace of the participants and took approximately 50 min each (mean 53 min, range 31-71 min). All interviews were audio-recorded and transcribed verbatim.

Data analysis

In analysing the data, we used MaxQDA software. In the analysis process, we used both deductive and inductive coding. We used a coding tree that was composed in previous research about QoL of persons with PIMD (deductive coding), and added new codes that were derived from the data (inductive coding) [21]. First, the first author AMN coded all the interviews and constructed a coding tree based on both the deductive and inductive coding. Second, the supervising author EO coded two interviews and compared his coding with the coding of AMN. This resulted in some refinements of the coding tree. Supplementary file 1 demonstrates the distinction between codes based on previous research and the codes based on the inductive coding process (the codes based on inductive coding are in italics). Then, the first author interpreted the fragments belonging to the codes and grouped them under different themes. After that, she discussed the results of this interpretation and the themes with DLW and EO. Based on this discussion the analysis was refined and this was continued during the whole process. The analysing process finally led to four themes, which will be presented below.

Participants

Thirteen participants were approached by telephone, of which one was excluded because he did not work with the target group. Twelve professional caregivers were interviewed. However, one interview was excluded because the interview revealed that the participant worked with persons with only intellectual disabilities and not with PIMD. Characteristics of the respondents are provided in Table 2.

Table 2. Characteristics of participants (n = 11)

Gender	Female	n = 11
Age	25-34	n = 4
	35-45	n = 6
	46-50	n = 1
Workplace	Residential care facility	n = 6
	Day-care centre	n = 4
	Hospital	n = 1

Results

Four major themes were found. First, descriptions of good and poor QoL. Second, ways of assessing QoL. Third, dependency of persons with PIMD on their environment and its influence on their QoL. Last, how this dependency influenced professional caregivers and their emotions.

Good and poor QoL

In the interviews, we asked professional caregivers to speak about persons with good and poor QoL and their concerns about QoL. Professional caregivers described good QoL in terms of being happy, having the ability to enjoy things, being relaxed, developing and having contact. In their description of good and poor QoL, most professional caregivers uttered hesitations, stressing that it is very difficult to know something about QoL in persons with PIMD, because they are unable to speak for themselves. While speaking about QoL of a person with PIMD, some participants questioned their own interpretations immediately. In the following quote most of these aspects come along [P11 =Participant 11, I = Interviewer].

P11: [An example of a person with a good QoL is] a cheerful girl. She is developing in various fields, such as motor skills and communication. Yes, basically in everything. And she is developing in leaps and bounds, but we also have a girl that is progressing in very small steps. And if development is possible, in other words that they feel safe and comfortable enough for development to be possible.

I: So if I understand you correctly, an important component for you when considering quality of life is that a child develops in small or large steps?

P11: Yes, or that the child Well, that is very tricky when you put it like that, because sometimes you don't see any development. But they are content, they are having a good day basically. That you can see that the child is happy or that they ... Even if the signals are not very strong but you see that the child experiences things as being pleasant. I think that if a child is crying the whole day or the child is only moaning and groaning, then you have to wonder what their quality of life is and how you could improve that by sorting things out.

Another aspect of good QoL mentioned by several professional caregivers was the capability of persons with PIMD to influence their surroundings. They mentioned different examples of having influence such as, making choices in what they want to eat, drive away in their wheelchair, showing that they are not feeling well by moaning. The more a person with PIMD can show what he or she wants and the professional caregiver understands these signals, the better QoL there is according to these professional caregivers.

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P4: Well, how can this client influence his or her own happiness? And that is very difficult, because in most cases this is very minimal, the influence that they can have in their lives. For example, when it comes to making decisions, but also about having a say in what happens to them.

I: In what sense do you mean. Do I understand you correctly, that you mean: if you have more of an influence then this usually means a greater quality of life? Or is that not the case?

P4: Yes, I do tend to think that way, yes. I think it would be great if that were possible. Yes, I am really basing that on my own situation. But place yourself in their situation, everything in their lives happens to them. We decide where you go, what you look like, whether you have a bath or not, what you eat, what you drink. Everything! Whether someone wipes your nose or not. If you have some sort of tool to make yourself understood, whatever that is or however that is achieved, then I think that would be fantastic! Then that is something that belongs to you and not because somebody else thinks that, it is yours.

In their description of poor QoL, these participants mentioned mostly physical elements such as experiencing pain, suffering, shortness of breath, illness or a combination of elements. Apart from these physical elements, some of them mentioned 'unhappy', 'excessive sleeping', 'no contact' and 'not visited by parents'.

P2: [An example of a person with PIMD with a poor QoL] was a very vulnerable girl, who often - almost always - had some physical ailment. She often had a cold or there was a problem with her tube feeding or, well all sorts of physical ailments that she was struggling with. And she did not like being touched, she found that very difficult. Her parents did not visit her often, they also found it very difficult. So yes, a girl who was always very vulnerable and difficult to have any form of contact with. It was hard for her to express what she liked and did not like. One thing that she did not like was being touched, that was very clear. But it was very difficult to see what she did like.

How professional caregivers assessed QoL

When answering the questions in the interviews, participants not only described good or poor QoL, they also described spontaneously how they found out about QoL. Several participants mentioned that they "read their faces", or that they "felt" whether the person with PIMD was feeling well. Other participants described how they "saw" it. In addition, some participants mentioned that they used their intuition to find out about the QoL of a person with PIMD. Stated differently, participants described in various ways that they "sensed" QoL of the persons with PIMD.

P7: I find this a very difficult topic. It is very hard to say whether a client's quality of life is good.

I: What makes it difficult?

P7: Because they cannot express themselves, I think. It is something that we ... We express our feelings. And my feeling may differ from that of a colleague. Because you are really trying to read their faces. And we base our judgement on that.

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P11: Yes, that is very tricky, because that is very often the case with this target group, that you have to do everything based on your intuition.

Besides this "sensing" QoL, participants described other ways of finding out QoL, for example by testing it using trial and error. Some participants mentioned that they discussed QoL of the person with PIMD with colleagues. The following quote is an example of how they used trial and error.

P7: Look, if I see that she is in pain - or I think that she is - then I say: "I will give paracetamol", but that does not help, that makes it tricky. And that to me is the most difficult about PIMD, that you are constantly on the lookout, what could it be? Is it the hip? And then - to be on the safe side - we go to the doctor again to check whether it is not dislocated, because she also has a hip problem. And you start ticking things off to check whether there are no physical problems, whilst in her case it could also be psychological. And yes, that is very difficult to determine. And that is why, with her, we are really trying to rule things out. We had a period in which we tried paracetamol. See whether things improve if she receives regular doses of paracetamol for a month. Now the antidepressants. After that, we will try dipiperon [antipsychotics]. And so, to conclude the entire story, maybe this is simply who this girl is and we need to accept that.

Dependency

Professional caregivers also described QoL of persons with PIMD as dependent on their social environment, which included family members and participants themselves. Participants felt the urge to know or understand the needs of their clients with PIMD and some of them referred to the importance of training to improve this. If they succeeded in finding out the needs, this contributed to good QoL. In other words, 'being understood' in their needs was an aspect of good QoL of persons with PIMD. Consequently, the more the person with PIMD was able to express his needs, the easier it was to fulfil these needs and contribute to good QoL according to participants.

I: Your first thought about quality of life for this group?

P8: Well, that they feel understood in what they express. I find that really important. That you really try to meet their needs. I think that this does give them quality of life. You sometimes hear examples of people who do not experience love. That they are unable to function at all.

I: And what is the difference between a lack of love and not being understood? Is that the same?

P8: Yes, I think that they are related.

I: So, because you simply feel a lot of love for someone?

P8: Yes, but love is also not possible if you are simply not understood. If someone is not understood. If you look at how we use that training to really look at their needs ... then you really start to meet their needs. I find that very beautiful.

Emotions of the respondents

In several interviews participants mentioned emotions related to this dependency or the search for the needs of their clients. These emotions occurred mostly when they could not fulfil the needs of persons with PIMD, or they could not find out the needs for good QoL. Some participants became emotional while describing the dependency of persons with PIMD and the impact this has on the relationship between the person with PIMD and the professional caregiver.

P1: I think that this is mainly due to the fact that these clients are so dependent on us, on our interpretations and our observations. But if you don't know whether someone is able to express himself, because something is holding him back, or whether it has something to do with pain, then what? I find that tricky.

I: But what you describe is that his quality of life could be determined by the relationship that he has with you and by what is possible in that relationship.

P1: Yes, I think so.

I: Yes, that is more or less what you are describing.

P1: But I think that if more - oh, now I am becoming a bit - if you could find more time. Yes, I am becoming emotional.

I: If you could give them even more time, then they would have a better quality of life?

P1: Yes, I think that if you were to see this client.

I: If you had more time?

P1: Well, maybe not so much more time, but greater continuity. If you look at your own family situation - and of course this isn't a family situation, but I think that is what these clients actually need, a small group of people surrounding them and caring for them. And learning to understand them. But of course, that group is quite large. And this is actually great for some clients, this diversity. And of course, you should not compare it directly to a family. But sometimes I do wonder, with all the varying contacts it is sometimes hard to build a relationship. And in the case of that one boy, the second client that I mentioned, I think that is really a shortcoming.

Several professional caregivers mentioned situations in which they knew the needs for good QoL of persons with PIMD but could not fulfil these needs. In these cases, they uttered feelings of frustration and experienced failure. There was usually an organisational reason for not being able to meet the needs of the persons with PIMD, such as staff shortages or lack of time.

P6: Well, living in an institution definitely has an effect on the quality of life, because when you live in an institution you are faced with time constraints.

I: And how is that for you?

P6: Frustrating! Yes. Frustrating. And that has a major effect on me, which in turn impacts on the way in which I interact with that child. In other words, it affects the quality of how I care for someone. Yes, so where the client lives is a major topic when it comes to quality of life. And not only when it comes to time, but a lot of other factors too: type of people, level of education.

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P7: And you really notice that and I think it also has an effect on the quality of life. We have a girl that is quite tense as far as muscle tone is concerned, so it would be ideal if she could take a bath every day. That is great for her, because then she can relax, well then she probably (inaudible), but you do not have that time.

Some participants mentioned emotions when they were no longer able to find ways to achieve good QoL in persons with PIMD. For example, if they did not know how to achieve happiness. Some participants mentioned feelings of powerlessness.

P10: In my opinion, when I think about it, it is the moment when you experience a sense of being powerless. That's what I think.

I: If you start feeling powerless?

P10: Yes, when you no longer know how to make someone happy, like the girl that cried the whole day.

I: Yes. That is quite possible.

P10: That's what I think.

I: And what is that then? In the moment that you feel powerless? (pause) Yes, because what was it you said just now? That you no longer know how to make someone happy?

P10: Yes. That you are no longer able to make the client feel happy, or safe.

P2: Well yes, I think the word "powerlessness" too. Yes. That you can pull out all the stops and if nothing helps. Or if another colleague also does not succeed. Nobody is able to achieve that bit of relaxation or that bit of pleasant life for him or her. Then it feels like a heavy burden.

Discussion

In this study, we explored professional caregivers' perceptions on QoL of persons with PIMD. We found examples of good and poor QoL. The quotes revealed that professional caregivers "sensed" QoL in different ways, and they tested it (trial and error) and discussed QoL of persons with PIMD with colleagues. Working with persons with PIMD, as our findings suggest, can touch their professional caregivers, especially when they do not know how to achieve good QoL for these persons.

In describing good and poor QoL, we found the same elements as we found in previous research on the perception of physicians on QoL of persons with PIMD [21]. Good QoL was mostly described in emotional terms such as happiness, pleasure and enjoying things. Poor QoL was mostly described in physical terms such as pain, sickness and shortage of breath. This is also in line with the QoL domains of emotional well-being and physical well-being, described by others [5] [22]. Furthermore, the findings suggest that the capability to influence the environment of persons with PIMD is an element of good QoL. In these cases, the person with PIMD

is able to express what (s)he wants, or professional caregivers are able to find out the needs of the person with PIMD. This finding highlights an aspect of QoL that is hardly captured by the domains we found in a previous literature review [9]. However, this aspect of QoL may be worth further exploring, because it could be an important aspect of QoL of persons with PIMD.

Our study suggests that professional caregivers assess QoL mainly by sensing it: our participants saw it, read faces, felt it or they based it on their intuition. These findings could be seen as descriptions of how professional caregivers develop knowledge on QoL of persons with PIMD, which is mostly tacit knowledge. These findings are in line with Reinders describing the importance of tacit knowledge in care [23]. Interestingly, our study suggests that not all of their knowledge is tacit: sometimes, professional caregivers verify their knowledge by testing it (using trial and error) and discussing with colleagues. Their use of (tacit) knowledge may be a valuable addition to the current assessment methods of QoL, which focus mostly on objective measurement of QoL, and it deserves further scrutiny in future studies.

Several of our participants described the relatedness between the QoL of persons with PIMD and themselves. Reinders [23] already argued that a high quality of relationship between professional caregiver and client was necessary for good care. Our findings also suggest that QoL of persons with PIMD is dependent on that relationship. In previous research, one of the physicians stated that persons with PIMD 'cannot make their own QoL' [21]. This relationship between QoL of persons with PIMD and the professional caregiver could be described in different ways. In an epistemic sense, it means that the professional caregivers have the knowledge on QoL of persons with PIMD. In an instrumental sense, the professional caregiver knows and uses instruments to improve QoL in persons with PIMD. This is comparable with a surgeon, whose surgical intervention is an instrument to cure a patient. In a constitutive sense, the professional caregiver is the source of QoL of the person with PIMD. Our findings indicate that all these descriptions apply to the relationship between the professional caregiver and QoL of persons with PIMD. On the other hand, maybe these different descriptions cannot be separated from one another and go together.

Being a source of QoL may feel like a huge responsibility for professional caregivers though, and it may explain the emotional reaction of some of them. The experiences of failure and powerlessness illustrate that caring for the QoL of persons with PIMD has an impact on professional caregivers. In the literature, emotional demands are described as one of the predictors of burnout in persons working with people with

intellectual disabilities [24] [25]. In addition, the impact of feelings of powerlessness is described in the literature [26]. Professional caregivers in our study experienced failure when they were unable to fulfil the needs of persons with PIMD, due to circumstances such as lack of time or workload. We are not sure that residential care facilities and day-care centres in our country are always aware of this relatedness between the QoL of persons with PIMD and their caregivers and the impact it may have on caregivers. Our study supports awareness of these issues, which may help to reduce the threats to the continuity of care for those who are fully dependent on others.

Understanding the needs of persons with PIMD was very important according to our participants. Some of them mentioned the benefits of specialised training to ‘understand’ persons with PIMD. The necessity of programmes in which the needs of persons with PIMD are systematically examined and further explored in activities and goals is also mentioned in the literature [27] [28] [29]. In addition, if professional caregivers improve their understanding of the needs of persons with PIMD, this can provide greater satisfaction and enjoyment of work. Furthermore, consultation and intervision or supervision may help professional caregivers to improve their understanding of persons with PIMD and bring emotional responses more into perspective.

The strength of this study is that it not only provides insight into how professional caregivers view QoL in persons with PIMD, but that it also reveals the influence of the relationship between the person with PIMD and the professional caregiver on the QoL of persons with PIMD. Moreover, our findings may provide valuable additions to the current assessment instruments. However, this research was conducted with eleven Dutch respondents and therefore provides information about working in Dutch care organisations. We do not know whether there would be a different outcome in other countries or cultures. A second point to note is that all participants were female. However, because this reflects the field, we do not see that as a limitation.

We conclude that this study provides insight into professional caregivers’ perspectives on QoL of persons with PIMD. They develop and use their (tacit) knowledge to assess QoL of persons with PIMD. The relationship between QoL of persons with PIMD and their professional caregivers deserves further attention in future studies. Paramount to these are the findings on professional caregivers’ experiences of failure and powerlessness, when working with persons with PIMD and trying to assess and fulfil their needs for good QoL. These findings demand

recognition of the work of professional caregivers in caring for persons with PIMD. This means enough training and supervision, organisation models that allow caregivers to build a relationship with persons with PIMD and recognition of any moral distress and its impact on the caregivers and the care they give. Management of both day-care centres and residential care facilities should take their responsibility in providing the necessary conditions so that professional caregivers can do their job properly and without unnecessary moral distress.

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Supplementary file 1

Code System

how do they find out

Oneself as interpretation instrument

emotion of interviewee

feeling of failure

powerlessness

costs

Weighing yes/no QoL

disagree about yes/no QoL

balance

doubt

ethics

Treatment

end of life decision

palliative sedation

doubts about treatment and QoL

role of significant others

Process

prognosis

deterioration

relationship

dependence on others

we see the needs and act on them

difficult to establish contact

family situation

safety

relationship parents

reciprocity

environmental variables

Supplementary file 1 continued

Code System

QoL PIMD

contact
able to develop
happy
relaxed
enjoys eating
joy
comfortable
observation QoL
stable health
pleasure
having an influence on the environment

QoL general
no QoL PIMD

epilepsy
showing little emotion
excessive sleeping
no joy
tense
inconsolable crying
difficulty eating and drinking
hospital admissions negative
pain
difficulty breathing
discomfort
suffering
fearful
appears sad/unhappy
sick
no contact
behavioral problems

Het levensverhaal van Britt

Dagcentrum Omega

"Wanneer ik aan Britt denk, dan zie ik een kwetsbaar, sfeergevoelig, sociaal én wilskrachtig meisje. Britt heeft mij laten zien hoe belangrijk "Totale Communicatie" is. Het is voor mij een methode om de wereld voor kinderen veilig en herkenbaar te maken, voor Britt was het dé manier om mij te laten weten hoe zij haar leven wilde leven."

Paula, persoonlijk begeleider van Britt bij Omega



"En maar wapperen met die tere kleine handjes... Dan wist je, ze is blij! Om haar wereldje (be)grijpbaar te maken brachten we wat we maar konden bedenken binnen bereik van (of "naar") die kleine handjes. En dan straalde ze... steeds maar weer. De complexiteit van haar leventje maakte zij tot het meest simpele: er is hier, er is nu, er is blijheid, in heel haar lijfje. Soms was er even geen blijheid, dan werd haar lijfje ziek en wisten we "het moet anders" en dat gebeurde dan, onontkoombaar. Het vergde wel even tijd en inspanning voor ik al haar signalen leerde begrijpen, maar vervolgens kon zij als geen ander een relatie met je aangaan waarin ze je wist te raken."

Rolia, persoonlijk begeleider bij Omega



"Met een lach, een geluid, of handgebaar liet Britt je weten dat we een fijne band hadden. Met enthousiaste geluiden liet Britt steeds vaker weten dat ze genoot van de gezelligheid om haar heen. Ook genoot ze intens van de liedjes die thuis en door ons werden ingezongen op de "big mack". Zo bijzonder hoe zij feilloos op die grote rode knop kon drukken, waarmee zij zelf die liedjes kon afspelen!"

Andrea, persoonlijk begeleider bij Omega



Chapter 5

Chapter 5

The perspective of parents on the assessment of quality of life of their children with profound intellectual and multiple disabilities

Manuscript submitted.

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Abstract

Assessing Quality of Life (QoL) of persons with profound intellectual and multiple disabilities (PIMD) is difficult. QoL is taken into consideration in many medical decisions that are taken frequently regarding children with PMID. The perspectives of parents of children with PMID on the assessment of QoL have not been studied.

We conducted a qualitative study, forming three focus groups with a total of 22 parents of children with PMID to explore their view on what is necessary to assess QoL of their children and subsequently, who is best suited to assess QoL.

Parents describe a long-term relationship of the assessor with family (child and parents), with trust as important aspect, as a requirement to assess QoL. Parents consider family members, preferably the parents themselves as the best assessors of QoL, followed by siblings. Professional caregivers, mostly mentioned by name, are considered the next alternative. Most parents thought that physicians do not know the child well enough to assess QoL.

In conclusion, the parents of children with PMID in our study consider trust and a long-term relationship essential for assessing QoL and do not consider physicians able to do so, but consider themselves as best assessors.

Introduction

The assessment of Quality of Life (QoL) of persons with profound intellectual and multiple disabilities (PIMD) is difficult. Persons with PIMD have profound intellectual disabilities (IQ<20) and severe physical disabilities [1]. Most persons with PIMD have additional sensory impairments such as visual and hearing problems and medical comorbidities [2]. Furthermore, they are dependent on others for all activities in daily life and they cannot verbally express themselves.

Several instruments have been developed to assess QoL [3]. Questionnaires have been developed such as the QoL-PMD [4] [5], the MIPQ [6] [7] and the San Martin Scale [8] but also behavioural observations with regard to happiness [9] [10] pleasure [11] and emotions [12] have been used. The assessors were mostly familiar to the persons with PIMD, with parents and direct support staff mentioned most frequently. In addition, unfamiliar observers were mentioned and triangulation with familiar and unfamiliar assessors was seen as a good assessment practice [11]. However, the role of parents in assessing QoL of persons with PIMD is sometimes critically questioned [4] [8] [10] [13] [14]. A background of this critique is that studies in groups of persons with less severe intellectual disabilities suggested that proxy outcomes differed from self-assessments [15] [16] [17].

Parental perspectives on support characteristics that influence the QoL of persons with PIMD and on the operationalisation of domains of QoL [14] [18] have been described. The perspective of parents on the assessment of QoL of their child with PIMD, however, has not been described before, while it is important. For example, QoL is a central topic in many medical (end-of-life) decisions [19]. Furthermore, the perspective of parents on the role of other (familiar and unfamiliar) assessors has not been explored. Therefore, the objective of this study is to explore parents' perceptions on the assessment of QoL of their child. The central questions were: 1) What do parents think is necessary to assess the QoL of their child? 2) Who, according to parents, can best assess the QoL of their child?

Materials and methods

We performed a qualitative study with three focus groups consisting of 22 parents of children with PIMD. They were asked about their perception on the assessment of QoL of their child, who can assess it and why. We used focus groups because we

wanted to explore similarities and differences between parents and to provoke discussion on these topics [20].

Recruitment and research ethics

We used purposive sampling to search for parents with a child with PIMD, aiming to obtain variation in age of participants' children with PIMD and their living place (either at home or in a residential care facility). Parents were approached via psychologists from our professional network and via national networks of parents and professional caregivers. Psychologists searched for parents with children with PIMD whom they thought would be willing to talk about QoL of their child. One parent suggested another parent and three parents (including one parent whose child was deceased) heard about our study and signed up. When parents had expressed an interest in the study, they were approached by the first author via e-mail or telephone and they received detailed written information. Remaining questions were answered, after which the appointment for the focus group was made. All parents signed a letter of consent, in which they confirmed that they had been informed about the study and their rights, including the right to withdraw at any moment and assurance of anonymity. The Medical Ethics Committee of the Academic Medical Centre granted a waiver for our study, confirming that it was not subject to the Medical Research Involving Human Subjects Act (WMO).

Data collection

Before we conducted the focus groups, we interviewed five mothers of children with PIMD to set up a guideline for the focus group (the mothers did not participate in the focus groups). Besides asking them to tell us about the QoL of their child, we shared our view on the topics of the focus groups with them and asked if they had suggestions and additions. In addition, we used our previous literature review study and qualitative research [3] [21], which together with the input from the mothers, led to a guideline (Supplementary file 1).

The first author, accompanied by the second or the fourth author of this paper conducted the three focus groups, which all took place at a day-care centre for persons with PIMD. Focus groups took between 2 and 2.5 hours each. They were audiotaped and transcribed verbatim.

Analysis.

The transcribed focus groups were analysed using the qualitative data analysis software, MaxQDA. First, AMN and EO read all the transcripts and searched for themes in an inductive coding process. For example, the role of physicians revealed during this phase. In the second round AMN and EO coded the focus groups using

codes derived from previous empirical research on QoL of persons with PIMD and the questions asked in the focus groups [3] [21]. Supplementary file 2 demonstrates the distinction between the codes derived from previous research and the codes based on the questions of the focus groups. The codes in italics are the codes derived from previous research. After coding the focus groups, the fragments belonging to the codes were placed per code in an excel file and studied thoroughly. Then AMN grouped the fragments in themes and discussed them with EO and DW. In these discussions, they addressed different aspects of the themes until consensus was reached. The analysis led to four themes, which will be presented below.

Participants

The parents of nineteen children were approached for the focus groups. One parent did not respond to the email, and the parents of two children had to be excluded because their son or daughter did not belong to the target group. For characteristics of participants' children (n=16), see Table 1. Twenty-two parents participated in the focus groups and in six cases, both the father and mother participated (fathers: 12, mothers: 10).

Table 1. Characteristics of participants' children (n=16)

Living at home	7	Age: mean 16, range: 10-22
Living in residential care facility	7	Age: mean 29, range: 15-51
Deceased	2	Age: 13 and 43

Results

Four major themes were found: what is necessary to assess QoL, who can assess QoL, the role of physicians, and difficulties in assessing QoL.

What is necessary to assess QoL

When discussing "what is necessary to make statements on the QoL of your child," parents mentioned two main aspects: there had to be a relationship with the child and a relationship with the parents.

Relationship with the child

Parents in the focus groups mentioned that persons who can assess the QoL of their child had to know their child very well or thoroughly, of which knowing the child for a long time was an aspect. "The physiotherapist who has been coming here for 15

years” or “*The home carer who has been coming here for 35 years.*” Another aspect was contact with their child on a regular basis, preferably several days a week. Mostly, parents referred to persons who took care of their child on a daily basis. [I is the author leading the focus groups, P 1 through to P 22 are the parents in the focus groups]

P 10: I choose the first responsible person for the day-care facility and the first responsible person for the home, all the home aides, why these three? Because I think they’re best equipped to assess our daughter’s daily wellbeing, because they see her at least a few days per week, care for her, sit with her... unlike the doctors and hospitals, the physiotherapists surrounding them, the general practitioners... they may have an impression of her, but it’s too fragmented, so I don’t think that these doctors and therapists have an adequate understanding of what daily life entails for a child like our daughter.

However, not only the quantity of time spent with their child was a requirement, but also the quality, for example, reciprocity was mentioned. Some parents described it as a sensitivity a person had for their child and the reciprocity there was between this person and their child. One parent called it love.

P 13: I think you can also look at your child, how your child responds to that care provider. I think that also plays a role: the interaction we as parents observe. Very simple, some people love their job and perform tasks in a loving way and others simply think: well, I need to put food in that child and then I’m done.

P 14: Yes, that is being very blunt, but you can notice quite quickly when someone...

P 13: ... whether they’re doing it with love.

P 9: What I’d like to add is - I see that with one or two, they really have a click with [name child], where you really see them connect with each other... Eye contact and you can see her enjoying the care that she’s receiving at that time. You see a lot more interaction.

Relationship with the parents

The parents mentioned several times that they have to trust persons who assess the QoL of their child, when the assessors are not themselves. They frequently mentioned that they had a relationship with these persons. One parent explained the necessity of trusting the assessor because of the advocacy role that he had to take for his daughter.

P 16: These are people whom I trust and have a relationship with.

P 19: But I think that we try to put ourselves in that position [as a substitute for the daughter] that we say that because: my [name child] cannot tell you that, so someone has to say it for her: ‘I do trust you and I don’t trust you.’ So, I think that we say that on behalf of the child, because we think: that is someone who is important or valuable to

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our child. Speaking for myself, I say these things because I try to observe from her perspective: who is valuable to her.

Conversely, if there was no relationship with the parents, the parents did not want him or her to say anything about the QoL of the child. The quote below is an example of this.

P 16: What I really hate is officials. And by that I mean persons who think they can have an opinion about my daughter or about me because of their official position. 'I'm your daughter's supervisor and I think'... or 'I am your doctor and I think...' Or 'I am...' whatever the case may be. Their reasoning is based on their position and not on: I am in this together with you, I want to build a relationship with you or whatever you tell me; I am visiting you, I also want to get to know your daughter, you know.

Who can assess QoL?

We asked the parents in the focus groups “who can make statements about the QoL of your child”. This question means both “who is capable to make statements” and “who is allowed to make statements”. When answering this question all parents in the focus groups mentioned themselves first. Next, they mentioned familiar others such as brothers and sisters or other family members or friends of the child or family. Parents also mentioned professionals. These professionals knew the child well, such as home cares or someone from the day-care centre or the residential facility. In general, the professionals were mentioned by name. Table 2 shows the individuals who can assess QoL in sequence of frequency mentioned.

Table 2. Who can assess QoL in sequence of mentioning?

Who, in sequence of mentioning	Specification
Parents ¹	Not applicable
Siblings	Not applicable
Close family members	Grandparents or parents' brothers, sisters or sisters-in-law. Sometimes nieces or nephews.
Professionals	Almost all parents mentioned one or more professionals, mostly by name
Others	Friends of the parents, respite care parents, the child's care coach,
Physicians	Some mentioned the parents' general practitioner, 1 mentioned the paediatrician.

¹including spouses who were not the biological parent

When asked who *cannot* say something about the QoL of their child, some parents mentioned care institutions, municipality, officials and authoritative sources, and with emphasis: physicians. The role of physicians will be described in detail below.

The role of physicians

Parents in all the focus groups spontaneously mentioned the (role of the) physicians. This role of physicians provoked a lot of discussion in the focus groups. Some of the parents were very explicit in their hesitation about the involvement of physicians in the assessment of QoL of their child, whilst some of them took a slightly more nuanced approach. According to most of the parents in the focus groups, physicians can say nothing or very little about the QoL of their child. The main reason for their stance was that physicians did not know their child sufficiently enough because they only saw him or her during office hours or when (s)he was very ill. In other words, physicians experience only unusual moments and never ordinary life.

I: How much do you actually need? I mean: a doctor does not see your child often enough. But how much time do you need to get a good impression? Do you need to observe the child for 24 hours?

P 9: I think that you should have almost daily interaction.

P 10: Yes, at least a few times per week and I do think that it would be good for doctors to spend a week with parents like us, with these children. Just spend a week from early morning to late in the evening: what is that like?

P 12: So, the problem is that it's not only fragmented, but also that they experience the unusual moments/situations, but never ordinary life. That's the point.

Another reason was that physicians did not have experience with the target group to which their child belonged. Therefore, these physicians did not know the particular details of persons with PIMD. Furthermore, parents felt that most physicians understood only one small component and not the whole child.

P 3: I am not sure whether you have the same experience, but in an emergency situation, you first need to get through this army of medics in training, who completely overreact, because [child's name] has all these abnormal values, the saturation is never good enough, the blood test results are not good. Everything is abnormal and you need to explain every single time:
a. Read the medical file. Ask us for advice. Sounds familiar?

P 7: But these doctors, you generally have a whole team of them, they only understand one small component, be it a neurologist or a pulmonologist or whatever.

However, the conversations between parents in the focus groups revealed that physicians do communicate their view on QoL of the child to the parents. For some parents this was puzzling and seemed difficult because in their point of view, physicians cannot assess QoL of their child. This was even more difficult when

physicians did not ask the opinion of the parents on the QoL of their child first. Furthermore, physicians' opinion on QoL of their child felt like a judgement for some parents.

P 6: But I think, what bothers us is that a doctor already has an opinion or a judgement on your child's quality of life. Without asking any questions first. I think that this is a very important aspect. But I do think that it's very important for doctors to acknowledge this and to be open to discuss it, but in a way that it's not presented as a judgement or an opinion about your child's quality of life, because I think all the parents agree with me that they [parents] are most capable of understanding their child's quality of life.

This seemed even more complicated when QoL was used in combination with decisions on medical treatment. Parents shared their experiences with each other in the focus groups and did not always agree on this subject. One parent had the feeling that physicians decided in end-of-life-decisions and based their decision only on *their* view on QoL of the child. Other parents however, disagreed and experienced a shared decision with physicians. The quote below reveals that in end-of-life-decision this mother had a different experience.

P 10: What makes me angry, is that in the lives of our children, the doctors and therapists give you so many responsibilities about medicines, about making decisions about: how is your child doing and what should or should not happen to your child? They give you all this responsibility, you have to deal with everything yourself and call us if you can't manage and as soon as you say, 'Wait a second, stop. We want to talk about this: what is the quality of life and what are we going to pursue, what are we not going to pursue? What are the options?' Then they often reply: 'Yes, but that is for us to decide.' And then I think: yes, but...

P 11: We don't share that experience. [Name child] received anti-seizure medication at some point he completely turned in on himself and then I said: We can live with the seizures, but we are going to stop the medication and the only thing that the neurologist said was: 'We'll increase the dose one more time, just to be sure that it is not working.' Then we said: 'Ok, one more step.' And after one week: if it's not working, we will stop the medication and that was fine.

P 10: Those types of decisions too. We've always been able to discuss these decisions with the doctor, but then I'm really referring to end-of-life decisions. In those cases, you as a parent are called to a halt, you're allowed to share your opinion, but the final decision rests with the doctor, whilst all those other responsibilities [are yours to deal with].

Following on from this, most parents did see an important role for physicians, which was as critical counterpart. Stated more specifically, as an experienced medical counterpart. Some parents wanted to discuss medical situations and decisions concerning their child with the physicians from different viewpoints. In most cases, the physicians mentioned the various medical options and the parents weighed these options from the perspective of QoL of their child.

P 6: It said so in his protocol. I said: 'But I'm not going to accept that. This is not a child like all the other children that you see, who plays outside in the school playground and that sort of thing'. So, then I had a very long conversation with this doctor and what it boiled down to was that that medicine was life-prolonging. I said: 'But her life expectancy with this syndrome is normal, as far as you know, so she could live up to 80 years old, but I don't know if I would want that, because then she'll outlive me. So can we talk about that?' In the end, we made the joint decision not to give that medicine. I said: 'If it were to improve her quality of life right now, if she were short of breath at the moment and she would be given medicines and no longer be short of breath, then I would want the treatment now', but he said: 'That's not what it is for at all.' And then I said: 'Then it's very clear to me.' And fortunately, he agreed with me. So I think that's a very good example of a doctor, how he can empathise with you as a parent and respect and accept that you have the right to an opinion on this.

Difficulties in assessing the QoL of their child

Although all parents mentioned themselves as the one who could assess the QoL of their child best, in their discussions with each other, they spoke about difficulties in describing or assessing the QoL of their child. The most important reason was that these children cannot verbalise their QoL. Parents mentioned that they had to 'read' their child. Parents usually learned to do so, but it remained difficult, especially when their child was not well. In fact, some parents described their concerns whether they understood their child well and one parent admitted that sometimes, he made a guess.

P 19: And I agree with you, actually we are the ones who need to determine whether that is still quality of life or not, because you cannot ask her. At the same time, this is the most difficult and frustrating part, that you cannot communicate properly, that you need to trust your instincts of who she is and I think that we - as parents, because we have known her the longest - are most likely to sense what is going on, but quite often we are also just making a guess. I find that tricky sometimes. Yes.

Another aspect that was discussed in the focus groups was the relationship between the QoL of their child and their own QoL. Parents reacted differently to these theses. Some of them described that their QoL was complete entangled with the QoL of their child. Some parents mentioned that some decisions they made had more to do with their own QoL than with the QoL of their child. Mostly these were decisions on staying in a guesthouse or living in a residential care facility. All parents mentioned the prevailing of the QoL of their child above their own QoL when their child was in hospital. Some parents described that their QoL and the QoL of their child could be seen as separated from each other.

P 6: I do not think that my quality of life depends entirely on my child's quality of life. Of course these things are related, but my quality of life is also related to that of my husband

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and my other child and my husband's children and a lot of other factors. So she is certainly a factor, but it is not true that you cannot examine (name child) quality of life without examining my quality of life... I think that these are separate aspects.

One parent mentioned other persons as a critical counterpart, which helped her to sharpen her opinion on QoL of her child. Furthermore, these persons could help her to disentangle her own QoL from that of their child or helped her to make decisions. This mother emphasised that she was the one making the decisions concerning her child, but that a critical view of someone else may help.

P 20: I would like to respond to that, because I'm also aware that I sometimes have a blind spot regarding [name child], that I think that I always know better and of course that is true in 99% of cases, but for that one percent I would like to receive input from others, including others not listed on the 'yes list.' That it makes you think about things again and discussing with your friends: yes, what is that actually, Quality of Life? And for whom? For me? For him? Just for anyone in general. Where are the boundaries? It is always good to receive input from a different angle.

Look, ultimately, I decide, but I have no problem with somebody suggesting a completely different idea that makes you think: yes, I'm going to think about that. Does that affect my opinion and my decision? Sometimes it does, sometimes only partially. So that's important, that I don't develop tunnel vision about how the treatment or other aspects are progressing. I think that this is important for me personally.

Discussion

In this study, we asked parents in focus groups what is necessary to assess QoL and consequently, who can assess QoL of their child with PIMD. We found that all parents mentioned themselves first as the ones who could assess the QoL of their child best. After that, they mentioned brothers and sisters of the child, relatives of the parents and lastly, professional caregivers, mostly mentioned by name. The persons who parents think can assess QoL are those who have a relationship with their child and have a relationship with themselves. The relationship with their child is not only built on time and frequency (quantity), but also on, for example, reciprocity between the caregiver and the child (quality). In the relationship with the parents, trust appears to be the most important aspect.

These findings differ from assessment practices we found in our earlier literature review [3]. In six of the included articles, QoL in persons with PIMD was assessed by familiar assessors as well as unfamiliar assessors [9] [10] [12]

[22] [23] [24]. Furthermore, we found that triangulation of assessors, which included unfamiliar assessors, was mentioned as a good assessment method [23]. The parents in the current study did not seem to think unfamiliar assessors are necessary. In fact, they mentioned the opposite namely, that the assessor has to know the child very well, otherwise (s)he cannot assess QoL of the person with PIMD. Stated differently, building a relationship with the child is necessary to assess the QoL of that child in the parents' view. Triangulation of assessors who know the child thoroughly however, may help the assessment process. After all, assessing the QoL of their child was difficult according to some parents in our study. In addition, one parent described that a critical counterpart could help to sharpen her opinion about QoL of her child and the unravelling of the disentanglement between the QoL of her child and her own QoL.

The role of physicians was mentioned frequently and spontaneously, both during the focus groups and in the interviews with the five mothers preceding this study. Therefore, this topic deserves scrutiny here. Why mentioned parents physicians so often and apparently had mixed feelings about them? Although we did not explore the reason for this attention for physicians, there are some examples described in literature that might contribute to the difficulties parents experience in relation to physicians. For example, persons with PIMD are hospitalised frequently and hospital is a place where parents and children often meet with physicians. This hospitalization places a burden on both the child and the parents [25], together with the cause of hospitalization. Other health care providers than the known physician may confront parents with the necessity of the treatment of their child, while the child may also be in a critical condition. From a medical perspective, a pop-up appears in the medical file at every hospital admission, asking the doctor to fill in whether all medical treatments are deemed necessary or whether there is any restriction in medical treatment. That encourages every attending physician, also those who do not know the child or the parents, to ask parents if the child should be admitted to the intensive care, should be ventilated, resuscitated and so on. Questions about QoL will automatically arise during those discussions.

Another example of why parents may have mixed feelings about their interaction with the physicians might be that the transfer from childhood to adulthood does not progress smoothly between physicians and therefore many different physicians may be involved in the medical care of adult persons with PIMD [26].

Furthermore, most participants thought that physicians could not assess QoL of their child. The reasons they mentioned were that physicians did not know the child well enough and did not see the child on a regular basis and not long enough. Other reasons were that, several physicians do not know the target group and that most of them are highly specialised, and might lack a holistic overview. Consequently, if physicians did communicate their view of QoL of the child to parents, this was not always appreciated. This was even less when the opinion of the parents was not asked before. Some parents mentioned that statements about QoL made by physicians felt like a judgement or verdict of the QoL of their child. Why did they experience it in such a way? We think of two possible reasons for this. The first may be that the view of physicians on QoL is not without consequences due to the role of physicians in decision-making on medical treatment. After all, the final responsibility of the consequences of these decisions lies, by law, with the treating physician, who should weigh all the aspects of medical decision-making. Perhaps, parents fear that a negative statement about the QoL of their child by a physician may lead to less medical treatment of their child. Another reason may be that a physician's negative view of the QoL of their child evoked feelings of indignity or worthlessness of the life of their child.

Some parents experienced negative feelings when physicians made statements about the QoL of their child, predominantly in end-of-life decisions. Probably, this had to do with the position of parents and physicians in this process. Other literature regarding the position of parents and physician in decision-making processes both confirm and contradict our results. Sullivan et al. [27] confirm our findings, namely the wish of some parents in our study for the physician to be their medically experienced counterpart and support them to be the end-of-life decision-maker for their child. Other research shows that there is no consensus in the parents' preferences regarding their influence on treatment decisions for their child, although this research was not limited to end-of-life decisions [28]. Zaal et al. [19] recommended that parents and physicians should more frequently discuss their view of QoL of persons with PIMD. Our findings however, did not confirm this. We found that these parents did have hesitations to discuss QoL of their child with physicians. However, Zaal et al. [29] reported, among other aspects, that a long-lasting relationship between parents and physician contributed to mutual trust and facilitated the decision-making process. This finding is in line with our finding that a long relationship and mutual trust are important, also for assessing QoL of persons with PIMD. We think that physicians should be careful to share their view of QoL of persons

with PIMD and ask parents first. Furthermore, they need to be sure, that their relationship with the parents is good enough to allow these conversations.

A limitation of this study is that parents in the focus groups were willing to talk about the QoL of their child, which can be an inclusion bias. This could have been created, because we asked psychologists to search for parents whom they thought would be willing to talk about QoL of their child. Furthermore, some parents signed up themselves when they heard about this study. We do not know if there are parents who do not want to talk about the QoL of their child and if so, why not. Could that invalidate our results? It does not seem likely, because it is probable that such parents would be at least as concerned as our respondents about trust and about others deciding about the QoL of their child. In addition, the findings of this study can only very cautiously be generalised to other contexts. The liberal Dutch climate, when it comes to medical-ethical issues, influences the context of this study. On the one hand, that may be an advantage because parents feel free to discuss these topics. On the other hand, this climate is perceived by some other societies as reprehensible, because of the risks of the slippery slope. Consequently, future studies are required to explore the experiences and ideas of parents in other countries and contexts. A strength of our study was the methodology, which included pre-discussions with parents and involved focus groups, in which parents stimulated and supported each other to talk about their own ideas and experiences. This empowered them to raise their struggles with physicians and at the same time, they felt free to talk about their own experiences, even when these contradicted the experiences of other participants.

Our conclusion is that, according to parents in our study, only persons who know their child thoroughly, have a relationship with their child and with them, and whom they trust are allowed to assess the QoL of their child. If a physician does not have this relationship, he or she is not allowed to assess QoL of their child. When physicians nevertheless share their view about QoL, this may feel as a verdict or judgement for parents. This conclusion requires future studies that explore the roles of parents and physicians (including physicians' final responsibility by law) in shared decision-making, the mutual sharing of views about QoL and the development of the role for physicians as an experienced medical counterpart. These roles should be explored further whilst ensuring that both physicians and parents retain their responsibility.

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Supplementary file 1

Guideline focus groups parents

Introduction

Welcome

Background

We have created an overview of the literature and conducted interviews with physicians and professional caregivers on QoL of persons with PIMD. We now want to ask parents on their view on QoL and the assessment of the QoL of their child. We think that it is important that you - as parents – have a significant voice in this process. Therefore, we have interviewed 5 mothers and have asked you to participate. We are pleased that you are willing to take part in this study!

Objective

Sharing experiences on the QoL of your child, on your QoL and the relationship between them and your view on the assessment of the QoL of your child. Who can assess QoL of your child and why.

Important

There are no right or wrong answers. The aim is to gain a deeper understanding of the similarities and differences between you.

Practical

Your name will be mentioned regularly for the recordings. This is essential for typing a transcript of the recordings. Your names will ultimately be removed from the typed transcripts.

Provide an overview of the evening:

Introductions and a conversation about quality of life of your children.

Who can make statements about the quality of life of your child and why.

Hopefully there will be some time at the end of the meeting for points that have not been discussed yet, but which you deem important to mention. Please use the yellow Post-It notes if your comment/question is not discussed during the evening.

You can then raise this point at the end of the evening.

Introductions

Start with a round of introductions. Also ask the parents what their thoughts were prior to this meeting when it comes to the quality of life of their child.

Discuss theses:

The literature reveals that there are varying opinions about the role of the parents in measuring quality of life. It is also evident that physicians assign a high priority to the client's surroundings (parents, family) when assessing the child's quality of life and when making decisions. This raises a number of questions, which we will discuss based on a number of statements. If you agree with the statement, please stand on this side (point). If you disagree, please stand on the other side.

Statements

1. I think that my child and I are so interconnected that our quality of life is difficult to disentangle.
2. My quality of life plays a role in decisions about my child.

Who can assess the quality of life of your child and why?

Who can make statements about the quality of life of your child? We will use yellow Post-Its for this topic, so that the parents can each write their thoughts down individually on a separate Post-It.

These will then be clustered on a sheet of paper and discussed – what are the correct clusters?

Next, we will discuss the specifics:

- Why are these people allowed to make statements about the quality of life of your child?
- Who is not allowed to make statements about this and why not?

Point that have not been discussed

Afterthoughts and specifics.

Supplementary file 2

Code System

QoL non

emotional

relational/contact

environment

physical

costs

QoL

physical

relational/contact

environment

emotional

determine yes/no QoL

dependence on others/environment

interconnection QoL child and parents

effect of parents on QoL of child

effect of child on QoL of parents

no QoL

yes QoL

when

phase in life

age

hospital

who

family

care provider

parents

role of physicians

other

husband/wife

grandparents

brothers/sisters

What

balance/imbalance

deterioration

process

Het levensverhaal van Britt

Laatste jaar

“Britt had een bijzondere eigenschap. Als er een vervelende gebeurtenis in aantocht was of iets wat zij niet wilde, dan kreeg ze koorts of epileptische aanvallen. Zo is een oogoperatie uiteindelijk afgeblazen omdat ze tot 3 keer toe koorts kreeg de dag voor de operatie. Om die reden is ze op Omega ook nooit overgeplaatst naar een andere groep, wat de gewoonte is na een aantal jaren op de groep. Dan werd ze ziek en als we toch doorzetten, en ze werd overgeplaatst, bleef ze ziek totdat ze uiteindelijk teruggeplaatst werd naar haar oude groep en daar werd ze dan weer dat vrolijke en gezellige meisje. Ook stond ze opgelijnd voor een hele zware scoliose operatie, waarvan steeds meer artsen en wij als ouders begonnen te twijfelen of Britt dat wel zou overleven. Ook deze operatie werd dus iedere keer uitgesteld.

In november 2019 kreeg Britt longontsteking en belandde ze op de IC van het AMC. Na een maand intensieve zorg en steeds meer complicaties, werd het steeds moeilijker voor Britt. Haar kwaliteit van leven rende achteruit. Toen echt niets meer hielp om haar koorts te verminderen en deze zelfs bleef stijgen, begrepen we dat Britt er klaar voor was en hebben haar moeten laten gaan. Op 27 november 2019 is Britt op 15-jarige leeftijd uiteindelijk overleden. Maar uiteindelijk heeft Britt ons echt veel geleerd en heel veel teruggegeven. Ze hield ons een overduidelijke spiegel voor, waar wij nog steeds in ons dagelijks leven veel aan hebben.”

Nancy & Henk, moeder & vader van Britt



“She always had a quiet presence that filled the room.”

Chapter 6

General discussion

General discussion

We wanted to gain more insight into the quality of life (QoL) of persons with profound, intellectual and multiple disabilities (PIMD). For that aim, we explored the full spectrum of QoL, which means not only the meaning of good QoL in this group and ways to improve it, as was the focus of most studies in this area until now, but also poor QoL and when concerns about QoL arise. In addition, we explored the assessment of QoL in persons with PIMD: how can QoL of persons with PIMD be assessed and what is necessary for that? Until now, studies of QoL, both on persons with intellectual disabilities (ID) and on persons with PIMD, focused on the use of QoL “as a conceptual framework for assessing quality outcomes, as a social construct that guides quality enhancement strategies and as a criterion for assessing those strategies” [1]. We know that especially questions on “poor QoL” of persons with ID, evoke intense ethical discussions in politics and society, for example, some years ago in the Netherlands, in the case of expensive treatments for patients suffering from Pompe or Fabry disease. Reinders et al. [2] warned against using the concept “poor QoL”, as, in their view, this description may be used to justify termination of life of persons with ID.

The questions we investigated arose from our practice as well, for example, in specialised day-care centre Omega, where questions on good and poor QoL of the persons they cared for came up, especially when these persons had passed away (see General Introduction). Furthermore, it has been reported by previous studies in our country that parents, professional caregivers and physicians often struggled with medical decisions such as withholding intensive care treatment or administration of antibiotics, and concerns related to QoL played a role in these struggles [3] [4]. When encountering the sensitivities associated with exploring poor QoL, it was important to be attuned to the experiences of participants in the different studies, in which my background has proven to be helpful because I have contact with the target group, their parents and professional caregivers on a daily basis. We chose to ask physicians and professional caregivers to describe good QoL and poor QoL of persons with PIMD they cared for and how they tried to understand their QoL. Furthermore, we explored the views of parents on the assessment of QoL of their

children and – more specifically – whom they regard to be capable of making such assessments and why. Our research questions were:

- Which concepts are used for QoL in persons with PIMD? (Chapter 2)
- Which instruments or methods are used, and which persons determine QoL? (Chapter 2)
- How do physicians describe good and poor QoL of persons with PIMD? (Chapter 3)
- When do physicians start to have concerns about the QoL of persons with PIMD? (Chapter 3)
- What is the perception of other professional caregivers on good and poor QoL of persons with PIMD? (Chapter 4)
- What do parents think is necessary to assess the QoL of their child? (Chapter 5)
- Who, according to parents, can best assess the QoL of their child? (Chapter 5)

During this study, we developed a tool that may empower parents to think about and describe the QoL of their child. The development of this tool together with the tool itself will be presented in an appendix.

In the next sections, we will summarise the main findings of the different studies. Next, we will reflect on the results and their consequences for the care of persons with PIMD, including the use of QoL-related considerations in medical decision-making.

Main Findings

This section highlights the answers to the research questions posed in the empirical studies and reflects on them.

Physicians' and professional caregivers' descriptions of good and poor QoL

Physicians and professional caregivers mostly used emotional terms such as happiness, pleasure and enjoying things and having good relationships or good contact with the environment to describe good QoL. Physicians mentioned a balance between good and poor elements as good QoL. For example, when there is a loving environment, while the person with PIMD is not feeling well, this can still be described as good QoL, because the loving environment can balance “not feeling well”. In addition, physicians emphasised the influence of the relational context, especially the relationship with parents, on both good and poor QoL. Professional

caregivers added the capability of persons with PIMD to influence their environment as an element of good QoL.

Poor QoL, according to physicians and professional caregivers, consisted of elements such as pain, sickness and shortness of breath, not feeling well and lack of contact with the environment.

Based on our findings, we clustered these elements of QoL in four groups: emotional, physical, relational elements and “capability to influence the environment”. In our literature review, we found several domains of QoL, of which the eight domains of Schalock are used frequently in care practice [5]. These domains are emotional wellbeing, interpersonal relationships, material wellbeing, personal development, physical wellbeing, self-determination, social inclusion and rights. Our findings confirm the domains of emotional wellbeing, interpersonal relationships and physical wellbeing. The “capability to influence the environment” may be linked to self-determination. Petry and Maes [6] described self-determination in relation to having choices and control over activities and environment. The other domains however (material wellbeing, personal development, social inclusion and rights), were not mentioned in our interviews when we asked participants to describe good and poor QoL of persons whom they cared for. Although the Schalock domains are based on scientific research, it may be that the four groups of elements (emotional, physical and relational and “capability to influence the environment”) are more applicable in practice and appropriate enough to describe good and poor QoL in this target group. This needs further exploration.

What makes physicians concerned about QoL

According to physicians, concerns about QoL arise when there is a change in the situation. This could be a physical decline, bad prognosis and increasing problems in the relational context. More specifically, they mentioned a negative change, when compared to earlier, in the situation as the tipping point when they start to have concerns about QoL. For example, a situation where initially there was good contact with the person with PIMD, and now there is less, can lead to concerns about QoL. Equally, the fact that a person with PIMD who could crawl or ride in a wheelchair recently lost these abilities may lead to concerns about QoL. Consequently, this may lead to the conclusion that concerns about QoL (and good and poor QoL) are always linked to the history of this particular and unique individual. Therefore, good or poor QoL cannot be described in absolute terms. For example, two children can exhibit the same elements of QoL: they cannot roll over by themselves, have very active epilepsy, have little contact with their parents, but do not seem to be unhappy.

However, their QoL could be valued differently. For instance, if the first child could previously crawl and had more contact with its environment because of less frequent seizures, his or her QoL could be described as being deteriorated in comparison to the past. Conversely, the other child could have been this way throughout his/her whole life and, because she/he seems happy, be described as having good QoL. This implies that QoL in persons with PIMD can only be assessed individually because it is connected with that person's life history. This corresponds to one of the principles of measuring QoL: "QoL measurement for individuals is based upon both common human experiences and unique, individual life experiences" [7]. Consequently, no generalising statement about the QoL of persons with PIMD can and should be made.

Physicians' and professional caregivers' assessment of QoL of persons with PIMD

Our interviews with physicians revealed that they were hesitant to describe the QoL of persons with PIMD. They described their perception of QoL of persons with PIMD as subjective, because it is based on interpretation and dependent on their relationship with the patient. These physicians mentioned that if they had no relationship with the child, they could not make statements about the QoL of that child. For example, one physician mentioned the difference in his interpretation of QoL of his own patients and of a patient of a colleague who attended hospital in the weekend.

The other professional caregivers (not being physicians) in our study also uttered hesitation when they were asked to describe the QoL of persons with PIMD. The main reason was that persons with PIMD could not speak for themselves.

Professional caregivers described how they assessed QoL. Several participants mentioned that they "read their faces", or that they "felt" whether the person with PIMD was feeling well. In addition, some professional caregivers mentioned that they used their intuition to find out about the QoL of a person with PIMD. Stated differently, professional caregivers described in various ways that they "sensed" QoL of the persons with PIMD. Furthermore, they tested it (trial and error), for example, they would hypothesise that the meaning of a certain sound is "yes" and respond accordingly and then would check whether or not the child reacts in the affirmative. Finally, they discussed QoL of persons with PIMD with colleagues.

Who, according to parents, can best assess the QoL of their child and why?

Parents also mentioned that assessing the QoL of their children with PIMD is very difficult and that even they were not always sure about their own interpretations. Despite this, parents thought that they, as parents, were most equipped to assess the QoL of their child. Siblings and other relatives and professional caregivers who were very close to their child followed them as possibly good assessors. In most

cases, parents mentioned physicians as ill-equipped to assess the QoL of their child. Parents mentioned that the most important requirement for assessing QoL is having a relationship with their child that needed to conform to both quantitative aspects and qualitative aspects. Quantitative aspects were duration (at least a few years), and frequency (for example, on a weekly basis), and qualitative aspects were, for example, reciprocity between the professional caregiver and the child. Stated differently, having a long-term relationship of good quality with their child was necessary to assess the QoL of their child. This is in line with physicians and professional caregivers in our study said: they mentioned that having a relationship with the child was necessary to make statements about QoL.

Furthermore, parents mentioned that they had to be able to trust the persons who assessed the QoL of their child in order to attach importance to their judgment. This means that according to the parents in our study, not only is a relationship with their child essential to assess QoL, but the parent's relation to the assessor is also important and must be of good quality.

Reflections on the results

In this part, we reflect on QoL as a relational concept and discuss what the consequences are. After that, we reflect on the results in relation to ethical issues that we mentioned in our general introduction and in medical decision-making for persons with PIMD.

QoL as a relational concept

Our empirical studies revealed that relationships are very important for the QoL of persons with PIMD. This involves both the impact of relationships on good and poor QoL and the importance of relationships for assessing QoL.

The importance of relationships is in line with findings reported by others. Watson et al. [8], for instance, found that a strong association between supporter responsiveness and relational closeness is important in supported decision-making in persons with severe and profound intellectual disabilities. Wightman and colleagues [9] also described the importance of relationships. They introduced the “relational potential”, referring to the potential of persons with PIMD to be in caring and loving relationships with their parents. They describe that this capacity of a loving relationship between parents and child can be an ethical justification for the life-sustaining treatment of children with PIMD if parents ask for this instead of focusing on capabilities or cognitions. Axelson and colleagues [10], in their study on improving

factors for participation of persons with PIMD in activities, mentioned that professional caregivers knowing persons with PIMD for a long time were one of the factors that improved participation. Finally, Maes et al. [11] recognised the importance of (good quality) staff interaction and support for good QoL of persons with PIMD and mentioned the dependency of persons with PIMD on others as common characteristic of this group.

This dependency on others for all activities in daily life is obvious because of the disabilities of persons with PIMD [12]. Consequently, their dependency on a relationship with others who care for them is also obvious. Vorhaus [13] mentioned several aspects of the relationship between persons with profound disabilities and their (professional) caregivers, such as reciprocity, love and affection, dignity and trust. In our interviews and focus groups, the word ‘love’ was mentioned several times when the relationship between professional caregivers and a person with PIMD was described. In other words, these relationships can be described as affective relationships. Consequently, what our study adds is that such a relationship has to be a long-term relationship of good quality. This is supported by a recent integrative review on parental knowledge of their children with PIMD [14]. This leads us to the conclusion that long-term relationships of good quality are a prerequisite for good QoL and for the assessment of QoL of persons with PIMD.

Consequences

This emphasis on long-term relationships of good quality should have, in our opinion, several consequences.

First, it affects the assessment of QoL. In Chapter 2, we questioned the possibility of objective assessment. Instead, we pleaded for an intersubjective approach, which recognises both the role of interpretation and the storied nature of who a person is. My colleagues and I described this in our practical-philosophical approach on witnessing QoL in persons with PIMD. We used the word “testimonies” when an individual who knows the person with PIMD well describes the QoL of that person. Stated differently, the assessor of QoL has to know the person with PIMD well to “give testimony” on the QoL of this person [15]. This is in line with our suggestion in Chapter 5 for triangulation as a method to assess the QoL of persons with PIMD by those who know these persons thoroughly. In addition, we agree with parents in that we think that it is very difficult for individuals who do not know the person with PIMD very well to assess their QoL, because their behaviour is not easy to interpret. For example, laughing is not always laughing, but it can be epilepsy or stress. It takes time to learn the nuances, and even parents in this study mentioned that they are not always sure of their interpretations.

Second, this emphasis on a long-term relationship of good quality is important for professional caregivers. They emphasised the QoL dependency for persons with PIMD on the close relationship with their daily, professional caregivers, including themselves. Understanding and fulfilling the needs of persons with PIMD for good QoL was very important, according to our participants. Not being able to see how QoL could be maintained or improved led to feelings of failure and powerlessness. In Chapter 4, we introduced three aspects that are necessary to care for persons with PIMD: knowledge, methods and personal competencies. They need knowledge about persons with PIMD, such as epilepsy, swallowing and choking and hypertensive muscles. Furthermore, professional caregivers need to know different methods, such as specialised communication methods, use of video analysis to understand and interpret behaviour, and methods for sensomotoric training. In addition, they need personal competencies, which are necessary in caring for persons with PIMD, for example, the competence to build a relationship with a person with PIMD. These competencies may require certain personality traits, in other words, belonging to the professional caregiver itself. For example, such professional caregivers like to care for other people; even if these persons with PIMD do not give a clear, visible response, they are very patient and can value very small reactions. However, we do not know enough about the personality traits that are necessary to care for persons with PIMD. They need further exploration in future studies.

Third, the emphasis on a long-term relationship of good quality as a prerequisite for good QoL of persons with PIMD has consequences for institutions. It is important that institutions create the necessary conditions for professionals so that they can build up this long-term relationship with persons with PIMD. However, this is not always in accordance with everyday practice because employee turnover and absenteeism are common problems faced by institutions [16]. Furthermore, moving staff from one department to another after a few years is viewed as good managerial practice. Moreover, a long-term relationship of good quality between professional caregivers and persons with PIMD and their parents may become too close, according to managers. This is sometimes labelled as unprofessional by organisations [17]. Therefore, these long-term relationships between professional caregivers and the persons they care for can bring up questions. After all, when is the relationship of good quality, and when is it starting to get too close or too involved? Furthermore, whose ideas are these? Maybe parents hold a different opinion on what constitutes a good relationship than managers in institutions? These questions deserve to be scrutinised in future studies.

Furthermore, long-term relationships of good quality may be a source of QoL for both patient and caregiver, but they may also lead to strong feelings of failure and powerlessness, which can be a burden for professional caregivers and may lead to absenteeism and even burnout. Such feelings require attention from managers in institutions. Offering intervision or supervision may support professional caregivers in dealing with these feelings [18] [19].

Fourth, the emphasis on a long-term relationship of good quality may result in a new description of QoL in persons with PIMD. A description in which the dependency on relationships is more obvious. Reinders [20] described QoL ('a life worth living') of persons with PIMD and connected it to (the gift of) friendship with a person with PIMD. My colleagues and I introduced "a life worth sharing" and "a life worth caring about" [21]. "A life worth sharing" refers to the dependency of persons with PIMD on others for all aspects of daily life and thus makes others responsible for the QoL of persons with PIMD. This acknowledges how important persons living or working with persons with PIMD are for those with PIMD, and vice versa. Using the concept of "a life worth sharing" may challenge others, to enter into this relationship with persons with PIMD. "A life worth caring about" refers to a caring society, a society in which it is normal to care for others, even when this requires time, energy and financial investments. Stated differently, it is a society in which unrelated citizens regard the life of persons with PIMD as worth caring about. A concept from which persons with PIMD could clearly benefit.

Ethical issues and medical decision-making

In our general introduction, we mentioned that exploring poor QoL was challenging. We described two examples of situations in which the use of QoL was sensitive: the use of QoL in discussions on the payment of medication for Pompe disease and Fabry disease and Reinders et al. [2] and their hesitation for using the concept of poor QoL, as, in their view, poor QoL could be the justification to terminate lives of persons with ID and PIMD. Furthermore, in our study with parents, this sensitivity of using QoL in medical decision making, especially in end-of-life decisions, became obvious. Our results may explain this sensitivity and provide some prerequisites for using the concept of poor QoL.

Our study involving physicians revealed that concerns about QoL arose when there was a negative change in the situation of a person with PIMD. We concluded that they compared the current QoL of a person with PIMD with the history of that person. This comparing with the history of a person could be more necessary in the case of persons with PIMD because they cannot verbalise their QoL themselves, nor can they confirm or reject interpretations of their QoL assessed by others. However,

this implies that QoL is highly personal and individual. Stated differently, statements about QoL cannot be generalised and applied to a whole group of persons with PIMD. However, in the discussion on the payment of medication for Pompe disease and Fabry disease, the assumption was that the QoL of a group of patients could not be improved (enough) by using medication. This led to an intense discussion. The people involved intuitively felt that this is an individual concept and not generalisable. However, politicians have to make decisions about issues such as the payment for expensive treatments for people with very rare metabolic disorders. One option they have is to exclude persons with PIMD from group decisions and decide individually about whether expensive treatments should be reimbursed. The vindication to exclude this group is that this group is exceptional; they cannot speak for themselves or express themselves about their QoL, and they have never been able to. If we compare this with other groups, such as persons with severe dementia, then these persons also cannot express their QoL themselves, but they could say things about their QoL before the onset of dementia. The only group that is perhaps comparable to persons with PIMD is the group of persons with profound intellectual disabilities. However, they are usually capable of walking and of other physical ways of expressing what they want or do not want. In contrast, questions on what this person with PIMD needs for good QoL are always answered based on interpretations by others. This could mean that persons with PIMD should be seen as a fundamentally different group and treated as such in decisions on reimbursements of medicines. In this context, it is an advantage that this group is rather small, so exclusion is possible.

The impossibility of generalising (poor) QoL to the whole group of persons with PIMD also echoes Reinders et al. [2] when they warn against the judgements of, for example, professionals, scientists or other observers, about the life of persons with PIMD (as a group) as having poor quality. Furthermore, we substantiated that QoL in persons with PIMD only can be assessed by persons who have a relationship with them and their parents. Therefore, we agree with Reinders in that we do not think medical decisions can be based on a generalised idea about poor QoL of persons with PIMD assessed by unfamiliar others. However, concerns on the QoL of a person with PIMD can arise, and it can sometimes be assessed as poor. As a consequence, in individual cases, it is possible that medical treatment of persons with PIMD may be withdrawn for reasons of poor QoL. However, we plead that this should never occur based on the judgment of poor QoL by an unfamiliar party, but on the judgment of familiar others, with the child's parents being the most important ones. This echoes Cummins, who concluded that in decisions on life, next of kin or other surrogates should decide, under the condition that their decision is based on the best interest of the person they care for. "In this light it seems timely to ask professional bodies to

consider whether proxy estimates of SBW (subjective wellbeing, author), made by professionals for people who are unable to respond for themselves, constitute ethical conduct” [22] page 201.

Lastly, we want to discuss the prerequisite of a long-term relationship of good quality for assessing QoL in medical decision-making. After all, most physicians do not have a long-term relationship of good quality with the person with PIMD and therefore would not qualify to assess the QoL of persons with PIMD. Therefore, we think that in discussions on medical decisions, especially end-of-life decisions, the concept of QoL can only be used under certain conditions. Zaal et al. [3], in their study, also mentioned trust and a good relationship as factors for improving good medical decision-making. Furthermore, their advice is to discuss QoL properly between parents and physicians, which means with respect and recognition of each other’s expert knowledge. We think that physicians should be wary of discussing their view on QoL of the child with the parents. If physicians do not know if parents trust them, it might be good if they only mention their view on the QoL of the child if parents ask for it and when parents do not ask for it, to ask parents if they want the physician to say something about it or not. Furthermore, whenever QoL is discussed, physicians should be convinced that the judgement on QoL should usually be a judgement that is obtained from others than themselves. Parents, siblings or health care workers with a longstanding and good relationship with the person with PIMD are the ones who are able to provide that information. Only in very rare circumstances, physicians are able to do so.

However, physicians can perfectly well and should discuss medical treatments with parents without referring to their view on the QoL of the child. For instance, an intensive care specialist can express his doubts about someone being able to come off a ventilator. They can also discuss the need for creating a tracheostomy. It is then up to the parents to assess what that means for the QoL of their child. If the outcome of the parents’ assessment supports the same decision about continuing or stopping the treatment as the physician’s envisaged course of treatment, then there is no problem. A problem arises when the physician is of the opinion that the treatment is futile from a medical perspective, yet the parents feel that the child still has sufficient QoL (or the other way around). In that case, extensive discussions need to take place between the physicians and the parents. Perhaps it can be helpful in these situations for parents to use the tool that we developed during this study (appendix). They can answer the questions about the QoL of their child by using the tool and use those answers in these extensive discussions with physicians. Ultimately, it is best if the physician and the parents can achieve consensus, from the physician’s perspective of medically futile treatment and from the parents’

perspective of the QoL of their child. However, in cases where parents and the health care professional team have opposite ideas about some treatment option, shared decision-making is essential. Where at the end medical decisions should be taken by the treating physician, final QoL decisions should be with the parents. Together these should turn into a shared decision on treatment options.

Conclusion

In this study, we explored not only good QoL or ways to improve QoL, as was usual until now, but also poor QoL and when concerns about QoL arise in persons with PIMD. One of our conclusions was that assessments of good or poor QoL and concerns about QoL are connected with the history of persons with PIMD and, therefore, QoL in persons with PIMD only be assessed individually and is not generalisable to a group of persons with PIMD. Based on our results we think that generalised statements about QoL of the whole group of persons with PIMD (as they have been and continue to be made in public discussions) have no ground and should be opposed. Another conclusion was that long-term relationships of good quality are both a prerequisite for good QoL and for assessing QoL of persons with PIMD. We described the consequences of these conclusions for future research, such as exploration of the balance between becoming too close and remaining professional in long-term relationships between professionals and persons with PIMD and their parents. Furthermore, institutions should pay more attention to the emotional consequences for professional caregivers of such long-term relationships. More research is needed into the required competencies or personal traits that professional caregivers need to care for persons with PIMD. This thesis confirmed that decision-making based on poor QoL is a very sensitive matter for this target group. There may be several reasons for this. Perhaps, one reason is that saying that someone with PIMD has a poor QoL can feel like a judgement not only about the quality but also about the value of that person's life. Therefore, we plead for very cautious use of poor QoL as an argument in medical decision-making. It would be good clinical practice that only persons who have a loving relationship with the person with PIMD may judge the QoL of that person as being of poor quality.

In closing

We started this thesis with questions that arose from the specialised practice of the Omega day-care centre. Why did we react differently when children or adults passed away, such as having feelings of shock in one case, and resignation in another? Does this say something about how we valued their QoL at the time that they passed

away? And if so, how did we know about their QoL? Even more surprising was that, in most cases, this seemed to be a shared feeling of parents and professional caregivers, although we had never discussed this feeling. The answer to these questions that I deduce from this study is that, at Omega, we sometimes succeed in building long-term relationships of good quality with the children and adults with PIMD and a trust-based relationship with their parents.

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Summary

Summary

Chapter 1

In the general introduction, Quality of life (QoL) was highlighted as an important concept for every human being, including persons with profound intellectual and multiple disabilities (PIMD). Persons with PIMD have profound intellectual disabilities (IQ<20) and severe physical disabilities. Most persons with PIMD have additional sensory impairments such as visual and hearing problems and medical comorbidities. Furthermore, they are dependent on others for all activities in daily life and they cannot verbally express themselves. Because persons with PIMD cannot verbally express themselves, assessing QoL of persons with PIMD is difficult. Until now, QoL was mostly brought up in questions about how to improve QoL, or as an outcome measure for either interventions or quality of care.

However, medical decisions such as withholding or withdrawing life-prolonging treatments, in which QoL plays a role, require a study of the full spectrum of QoL. This means that we not only need to explore ways to improve QoL, but also need to explore when QoL of persons with PIMD is described as “poor QoL” or when concerns about QoL emerge. Therefore, we conducted a qualitative study using interviews and focus groups to answer our research questions.

Our research questions were:

- Which concepts are used for QoL in persons with PIMD?
Which instruments or methods are used and which persons determine QoL?
(Chapter 2)
- How do physicians describe good and poor QoL of persons with PIMD?
When do they start to have concerns about QoL of persons with PIMD?
(Chapter 3)
- What is the perception of other professional caregivers of good and poor QoL of persons with PIMD? (Chapter 4)
- What do parents think is necessary to assess the QoL of their child?
Who, according to parents, can best assess the QoL of their child? (Chapter 5)

As a part of this study, we used the results to develop a tool for parents to help them to describe the QoL of their child. In conversation with professionals, for example physicians, this could be helpful in making the right decisions. However, this tool is not a part of this thesis, but is presented in an Appendix.

Chapter 2

A literature review was necessary because no overview of the literature on the assessment of QoL in PIMD had been provided since 2009. Such an overview was necessary to clarify the current state of concepts, methods of assessment and assessors in the research of QoL in persons with PIMD. Twenty-one articles were included in the analysis. Multidimensional concepts of QoL consisting of both subjective and objective components were found, although the articles used different distinctions for each. We found questionnaires such as the QoL-PMD, the MIPQ and the San Martin Scale. Furthermore, QoL was assessed through behavioural observations, in which, for example, happiness, pleasure, and emotions were studied.

The assessors were mostly familiar to the persons with PIMD, with parents and direct support staff mentioned most frequently. In addition, unfamiliar observers were mentioned frequently and triangulation with familiar and unfamiliar assessors was seen as a good assessment practice. However, in our discussion section, we questioned some elements of the conceptualisation including the following: the difference between objective and subjective QoL, as well as underlying assumptions of behavioural observations. Instead of trying to objectify QoL in persons with PIMD, we argued that the added value of interpretation should be recognised. We think that a narrative approach, which recognises both the role of interpretation and the storied nature of who a person is, can be helpful in describing QoL in persons with PIMD, especially when QoL plays a role in medical decisions.

Chapter 3

In the first empirical study, we explored physicians' perceptions of QoL of persons with PIMD. We asked seven physicians to describe QoL of persons with PIMD. However, we found a reticence to answer this question, because according to participants, their perception about QoL of persons with PIMD is subjective, based on interpretation and dependent on their relationship with the patient. Despite this, they did answer the question, and emotional, physical and relational clusters of elements of QoL were found. Physicians named mainly emotional elements such as happiness, pleasure and enjoying things to describe good QoL, and physical elements such as pain, sickness and shortness of breath to describe poor QoL. Furthermore, relational elements were mentioned, such as good contact and a loving family as an element of good QoL and no contact or no parents as part of poor QoL. In addition, they mentioned a balance or equilibrium between good and poor elements as good QoL. Our respondents emphasised the effect of the relational context (particularly the parents) on QoL of persons with PIMD. The findings suggest that the tipping

point of starting to have concerns about QoL was a negative change, mostly in physical elements, such as physical deterioration, or in the relational context, like parents not being able to cope with their situation anymore, or when there was a disturbance in the balance/equilibrium.

Chapter 4

Next, we explored professional caregivers' perceptions of QoL of persons with PIMD. Professional caregivers are responsible for the daily care of persons with PIMD in residential care institutions and day-care centres. In describing good and poor QoL, we found the same elements as in the study on the perceptions of physicians. Good QoL was mostly described in emotional terms and poor QoL was mostly described in physical terms. In addition, professional caregivers said that the capability of the person with PIMD to influence the environment is an element of good QoL. Describing how they assessed QoL, professional caregivers said they "sensed" QoL, tested it (trial and error) and discussed QoL of persons with PIMD with colleagues. Several of our participants described the relationship between the QoL of persons with PIMD and themselves and emphasised the dependency of persons with PIMD on them. This relationship between QoL of persons with PIMD and the professional caregiver could be described in different ways. In an epistemic sense, it means that professional caregivers have the knowledge about QoL of persons with PIMD. In an instrumental sense, the professional caregiver knows and uses instruments/methods to improve QoL in persons with PIMD. In a constitutive sense, the professional caregiver is the source of QoL of the person with PIMD. Our findings indicate that all these descriptions apply to the relationship between the professional caregiver and QoL of persons with PIMD. However, being a source of QoL may feel like a huge responsibility for professional caregivers and it may explain the emotional reaction of some of them during the interviews. In fact, these professional caregivers described feelings of failure and powerlessness if they could not assess or fulfil the needs of persons with PIMD. Working with persons with PIMD, as our findings suggest, can be emotionally challenging for their professional caregivers, especially when they do not know how to achieve good QoL for these persons.

Chapter 5

In this study, we asked parents in focus groups what is necessary to assess QoL and consequently, who can assess QoL of their child with PIMD. We found that all parents mentioned themselves first as the ones who could assess the QoL of their child best. After that, they mentioned brothers and sisters of the child, relatives of the parents and professional caregivers, mostly mentioned by name. The persons who parents believe can assess QoL are those who have a relationship both with their child and with them. The relationship with their child is not only built on time

and frequency (quantity), but also on, for example, reciprocity between the caregiver and the child (quality). In the relationship between the assessor and the parents, trust appeared to be the most important aspect. If physicians did not have this relationship with the child and/or parents, parents considered them as ill equipped to assess QoL of their child. When physicians nevertheless shared their view on QoL, this could feel as a verdict or judgement for parents. However, parents mentioned an important role for physicians as an experienced medical counterpart. One of the conclusions of this study was that further exploration of the roles of parents and physicians (including physicians' final responsibility by law) in shared decision-making including the mutual sharing of views on QoL of the parents' child is necessary.

Chapter 6

The general discussion presented two main conclusions. The first conclusion is that long-term relationships of good quality are a prerequisite for good QoL and for the assessment of QoL of persons with PIMD. We mentioned some consequences of our first conclusion for the practice of care.

First, it has an effect on the assessment of QoL. This implies that only persons, who know the person with PIMD very well, can assess their QoL.

Second, it is very important that professional caregivers can build this relationship with persons with PIMD. However, we do not know much about the personality traits that are necessary to build a relationship with persons with PIMD. This needs further exploration in future studies.

Third, the emphasis on a long-term relationship of good quality has consequences for institutions. It is important that institutions create the necessary preconditions for professionals so that they can build up this long-term relationship with persons with PIMD. Furthermore, questions remain about when a relationship between professional caregivers and the person with PIMD and its parents is good or when is it becoming too close.

Finally, the necessity of a long-term relationship of good quality for assessing QoL in medical decision-making has implications for physicians. After all, most physicians do not have a long-term relationship of good quality with the person with PIMD and therefore would not qualify to assess the QoL of persons with PIMD. We think that physicians should be wary to discuss QoL of the child with the parents. It might be good if they only mention their view on the QoL of the child if parents ask for it and when parents do not ask for it, to ask parents if they want the physician to say something about it or not.

Our second conclusion is that assessment of QoL is highly personal and individual, because it has to be related to the history of a person with PIMD, who cannot

express him or herself. We confirmed in this study that the use of poor QoL is a very sensitive issue. Based on our results we think that generalised statements about QoL of the whole group of persons with PIMD (as they have been and continue to be made in public discussions) have no ground and should be opposed. Furthermore, we think that decisions based on poor QoL can only be made based on the assessment made by persons who have a loving relationship with the person with PIMD.

Samenvatting

Samenvatting

Hoofdstuk 1

In de algemene inleiding werd Kwaliteit van leven (KvL) onderstreept als een belangrijk concept voor ieder mens, inclusief personen met (zeer) ernstige verstandelijke en meervoudige beperkingen ((Z)EVMB). Personen met (Z)EVMB hebben (zeer) ernstige verstandelijke beperkingen (IQ<20) en ernstige lichamelijke beperkingen. De meeste personen met (Z)EVMB hebben ook zintuigelijke beperkingen, zoals visuele of auditieve beperkingen, en medische comorbiditeiten. Bovendien zijn zij afhankelijk van anderen voor alle dagelijks activiteiten en zijn zij niet in staat om zichzelf verbaal uit te drukken. Het feit dat personen met (Z)EVMB niet verbaal kunnen communiceren maakt het lastig om de KvL van personen met (Z)EVMB te beoordelen. Tot nu toe werd KvL meestal besproken in de context van het verbeteren van KvL, of als een uitkomstmaat voor interventies of de kwaliteit van zorg.

Medische beslissingen, zoals het stoppen of niet starten van levensverlengende behandelingen, waarin KvL een rol speelt, behoeven echter onderzoek naar het volledige spectrum van KvL. Dit betekent dat we niet alleen moeten zoeken naar manieren om KvL te verbeteren, maar dat we ook moeten onderzoeken wanneer KvL van personen met (Z)EVMB wordt omschreven als “slechte KvL” of wanneer zorgen over KvL ontstaan. Daarom hebben wij een kwalitatief onderzoek uitgevoerd met interviews en focusgroepen om onze onderzoeksvragen te beantwoorden.

Onze onderzoeksvragen waren als volgt:

- Welke concepten worden gebruikt voor KvL van personen met (Z)EVMB?
- Welke instrumenten of methodes worden gebruikt en welke personen bepalen KvL? (Hoofdstuk 2)
- Hoe beschrijven artsen goede en slechte KvL van personen met (Z)EVMB?
- Wanneer gaan zij zich zorgen maken over KvL van personen met (Z)EVMB? (Hoofdstuk 3)
- Hoe is de perceptie van andere professionele zorgverleners van goede en slechte KvL van personen met (Z)EVMB? (Hoofdstuk 4)
- Wat is er volgens de ouders nodig om de KvL van hun kind te beoordelen?
- Wie is er volgens de ouders het meest geschikt om de KvL van hun kind te beoordelen? (Hoofdstuk 5)

Als deel van dit onderzoek hebben wij de resultaten gebruikt om een hulpmiddel voor ouders te ontwikkelen, om ze te helpen de KvL van hun kind te omschrijven. Dit zou nuttig kunnen zijn in gesprekken met professionals, bijvoorbeeld artsen, om de juiste beslissingen te maken. Dit instrument vormt echter geen deel van dit proefschrift, maar wordt wel in een Bijlage gepresenteerd.

Hoofdstuk 2

Een literatuurstudie was noodzakelijk, omdat de literatuur omtrent het beoordelen van KvL van mensen met (Z)EVMB sinds 2009 niet verder was onderzocht. De literatuurstudie was noodzakelijk om de huidige status van concepten, beoordelingsmethodes en beoordelaars in het onderzoek van KvL van personen met (Z)EVMB te verduidelijken. Eenentwintig publicaties werden opgenomen in de analyse. Multidimensionale concepten van KvL met zowel subjectieve als objectieve elementen werden gevonden, hoewel de publicaties verschillende onderscheidingen gebruikten voor deze elementen. Wij vonden vragenlijsten zoals de QoL-PMD, de MIPQ en de San Martin Scale. Verder werd KvL beoordeeld door middel van gedragsobservatie, welke bijvoorbeeld gelukkig zijn, plezier en emoties onderzocht. De beoordelaars waren voornamelijk bekenden van de personen met (Z)EVMB, waarbij ouders en dagelijks verzorgend personeel het vaakst genoemd werden. Onbekende observatoren werden ook vaak genoemd en triangulatie van bekende en onbekende beoordelaars werd beschouwd als goede beoordelingspraktijk. In onze discussie twijfelen wij echter aan bepaalde elementen van de theorievorming, inclusief: het verschil tussen objectieve en subjectieve KvL, evenals de onderliggende aannames over gedragsobservaties. In plaats van te proberen de KvL van personen met (Z)EVMB te objectiveren stellen wij dat de toegevoegde waarde van interpretatie erkend zou moeten worden. Wij denken dat een narratieve benadering, welke het belang van de rol van interpretatie en het levensverhaal van een persoon erkent, nuttig kan zijn in het omschrijven van KvL van personen met (Z)EVMB, vooral wanneer KvL een rol speelt in medische beslissingen.

Hoofdstuk 3

In de eerste empirische studie onderzochten wij de opvattingen van artsen over KvL van personen met (Z)EVMB. Wij vroegen zeven artsen om de KvL van personen met (Z)EVMB te omschrijven. De deelnemers waren echter terughoudend in het beantwoorden van deze vraag en gaven aan dat hun opvattingen over KvL van personen met (Z)EVMB subjectief is, gebaseerd op interpretatie en afhankelijk van de relatie met de patiënt. Ondanks deze terughoudendheid beantwoorden zij toch de vraag en werden emotionele, fysieke en relationele clusters van elementen van KvL gevonden. Artsen noemden voornamelijk emotionele elementen zoals gelukkig zijn, plezier hebben en van dingen kunnen genieten om goede KvL te omschrijven, en

fysieke elementen zoals pijn, ziekte en kortademigheid om slechte KvL te omschrijven. Verder werden relationele elementen ook genoemd, zoals goed contact en een liefdevolle familie als elementen van goede KvL en geen contact of geen ouders als onderdelen van slechte KvL. Bovendien omschreven zij goede KvL als een balans of evenwicht tussen goede en slechte elementen. Onze respondenten benadrukten het effect van de relationele context (vooral de ouders) op KvL van personen met (Z)EVMB. Deze bevindingen suggereren dat het kantelpunt waarop zorgen over KvL ontstaan een negatieve verandering is, meestal van fysieke elementen, zoals fysieke verslechtering, of in de relationele context, zoals ouders die de situatie niet meer aankunnen, of een verstoring in de balans/ het evenwicht.

Hoofdstuk 4

Vervolgens onderzochten wij de opvattingen van professionele verzorgers over KvL van personen met (Z)EVMB. Professionele verzorgers zijn verantwoordelijk voor de dagelijkse verzorging van personen met (Z)EVMB in instellingen en dagcentra. Bij het omschrijven van goede en slechte KvL vonden wij dezelfde elementen als in de studie over de opvattingen van artsen. Bij goede KvL werden voornamelijk de emotionele aspecten omschreven en bij slechte KvL werden voornamelijk de fysieke aspecten omschreven. De professionele verzorgers gaven ook aan dat de mogelijkheid van de persoon met (Z)EVMB om zijn omgeving te beïnvloeden een aspect van goede KvL is. Wanneer zij gevraagd werden hoe zij KvL beoordelen gaven de professionele verzorgers aan dat zij KvL “aanvoelen”, testen (uitproberen) en KvL van personen met (Z)EVMB bespreken met collega's. Een aantal deelnemers omschreef de relatie tussen de KvL van personen met (Z)EVMB en de professionele verzorgers, en benadrukte nogmaals hoe afhankelijk personen met (Z)EVMB zijn van de professionele verzorgers. Deze relatie tussen KvL van personen met (Z)EVMB en de professionele verzorger kan op verschillende manieren omschreven worden. In epistemische zin betekent dit dat professionele verzorgers over kennis beschikken van KvL van personen met (Z)EVMB. In instrumentele zin kent en gebruikt de professionele verzorger instrumenten/methodes om KvL in personen met (Z)EVMB te verbeteren. In constitutieve zin is de professionele verzorger de bron van KvL voor de persoon met (Z)EVMB. Onze bevindingen wijzen erop dat al deze omschrijvingen van toepassing zijn op de relatie tussen de professionele verzorger en KvL van personen met (Z)EVMB. Het feit dat zij een bron van KvL zijn kan echter als een enorme verantwoordelijkheid ervaren worden door professionele verzorgers en kan de emotionele reactie van sommigen tijdens deze interviews verklaren. Deze professionele verzorgers kampen met name met emoties van falen en machteloosheid als zij er niet in slagen om de behoeften en wensen van personen met (Z)EVMB te beoordelen en hieraan te voldoen. Onze bevindingen wijzen erop dat het werken met personen met (Z)EVMB een emotionele uitdaging kan vormen

voor de professionele verzorgers, vooral als zij niet weten hoe ze goede KvL moeten verwezenlijken voor deze personen.

Hoofdstuk 5

In dit onderzoek vroegen wij ouders in focusgroepen wat er nodig is om KvL te beoordelen en derhalve wie KvL van hun kind met (Z)EVMB kan beoordelen. Wij constateerden dat alle ouders eerst zichzelf noemden als degenen die het meest geschikt zijn om de KvL van hun kind te beoordelen. Daarna noemden zij broers en zussen van het kind, familie van de ouders en professionele verzorgers, vaak bij naam genoemd. De personen die de ouders geschikt vinden om KvL te beoordelen zijn degenen die een relatie hebben met zowel de ouders als hun kind. De relatie met hun kind is niet alleen gebaseerd op tijd en frequentie (kwantiteit), maar ook bijvoorbeeld op wederkerigheid tussen de verzorger en het kind (kwaliteit). In de relatie tussen de beoordelaar en de ouders bleek vertrouwen het meest belangrijke aspect. Als artsen geen relatie hadden met het kind en/of de ouders, dan beschouwden de ouders deze artsen onvoldoende in staat om KvL van hun kind te beoordelen. Als artsen toch hun mening gaven over KvL, dan werd dit soms door de ouders ervaren als een oordeel. Ouders vermeldden echter wel dat de artsen een belangrijke rol kunnen spelen als ervaren medische evenknie van de ouders. Een van de uitkomsten van dit onderzoek was dat verder onderzoek naar de rollen van ouders en artsen (rekening houdende met de wettelijke eindverantwoordelijkheid van artsen) in het gezamenlijke besluitvormingsproces - inclusief het delen van meningen over KvL van het kind van de ouders - noodzakelijk is.

Hoofdstuk 6

In de algemene discussie werden de twee voornaamste conclusies gepresenteerd. De eerste conclusie is dat langdurige relaties van goede kwaliteit een voorwaarde zijn voor goede KvL en voor het beoordelen van KvL van personen met (Z)EVMB. Wij benoemden een paar gevolgen van onze eerste conclusie in de praktijk.

Ten eerste heeft dit effect op het beoordelen van KvL. Dit impliceert dat alleen personen die de persoon met (Z)EVMB zeer goed kennen hun KvL kunnen beoordelen.

Ten tweede is het zeer belangrijk dat professionele verzorgers de mogelijkheid hebben om deze relatie op te bouwen met personen met (Z)EVMB. Wij weten echter weinig over de persoonlijkheidskenmerken die nodig zijn om een relatie op te bouwen met personen met (Z)EVMB. Dit dient nader onderzocht te worden in toekomstige onderzoeken.

Ten derde heeft de nadruk op een langdurige relatie van goede kwaliteit consequenties voor instellingen. Het is belangrijk dat instellingen de benodigde randvoorwaarden voor professionals creëren, zodat zij deze langdurige relatie met

personen met (Z)EVMB op kunnen bouwen. Verder bestaan er nog steeds vragen over wanneer een relatie tussen professionele verzorgers en personen met (Z)EVMB en zijn/haar ouders goed is en wanneer deze te hecht wordt.

Tot slot heeft de noodzaak voor een langdurige relatie van goede kwaliteit voor het beoordelen van KvL in medische besluitvorming ook gevolgen voor artsen. De meeste artsen hebben immers geen langdurige relatie van goede kwaliteit met de persoon met (Z)EVMB en zouden derhalve niet in aanmerking komen om de KvL van personen met (Z)EVMB te beoordelen. Wij denken dat artsen terughoudend moeten zijn in het bespreken van hun visie op de KvL van het kind met de ouders. Het verdient aanbeveling alleen de mening over KvL van het kind te geven als de ouders hier om vragen en als ouders hier niet om vragen, dan eerst de ouders te vragen of zij willen dat de arts zich hierover uitspreekt of niet.

Onze tweede conclusie is dat de beoordeling van KvL hoogstpersoonlijk en individueel is, omdat deze gerelateerd moet worden aan de voorgeschiedenis van een persoon met (Z)EVMB, die zich niet hierover kan uiten. Wij bevestigden in dit onderzoek dat het gebruik van slechte KvL een zeer beladen onderwerp is. Op basis van onze resultaten denken wij dat gegeneraliseerde uitspraken over KvL van de hele groep personen met (Z)EVMB (zoals zij in het verleden en nog steeds worden aangemerkt in maatschappelijke discussies) geen bestaansrecht hebben en bestreden moeten worden. Verder zijn we van mening dat beslissingen die gebaseerd zijn op slechte KvL alleen gemaakt kunnen worden op basis van de beoordeling door mensen die een liefdevolle relatie hebben met mensen met (Z)EVMB.

Appendix 1

Appendix, deel 1

A tool to support parents to think about and describe the QoL of their child

The tool we developed during this study can be helpful for parents in decisions on medical treatment and “give a voice” to the QoL of their child. Mirjam de Vos, in her book on children who are unable to make decisions, devoted a chapter to QoL [1]. This chapter highlights the importance for parents to describe the QoL of their child in medical decisions concerning their child. In their conversation with physicians about medical decisions, this tool can help parents to weigh the different medical choices and their consequences for the QoL of their child. This can empower parents in these conversations with physicians. Empowerment of parents is important in coping with the disabilities of their child [2] [3]. In literature, participation in decision-making is mentioned as one of the aspects of empowerment of parents of children with disabilities [4]. Therefore, we believe that our tool helps parents to make (the right) decisions for their child. Another tool that has been developed to empower and support parents is a digital tool to help parents of children with physical disabilities to formulate questions and find information [5]. Their tool and ours complement each other and they empower parents of children with PIMD.

Development of the tool

The tool was based on our findings and consists of emotional, physical and relational elements, autonomy of persons with PIMD and the temporal dimensions of QoL. The latter refer to changes in QoL that may give rise to concerns about QoL and the finding that QoL is always linked to the history and future expectations of a person with PIMD. As the interviews also suggested that the relationship with parents is essential for QoL of children with PIMD, we also introduced questions on these matters. From the focus groups with parents, we learned that parents hold the opinion that they know best who is able to assess the QoL of their child. Therefore, we decided to let parents choose for themselves if and with whom they want to discuss the QoL of their child and the answers to the questions raised in the tool.

A first draft of the tool was presented to parents in a specialised day-care centre for persons with PIMD and parents were asked to give comments (Omega, 22 may 2019). This led to minor revisions of the tool. This tool may be subject to modifications based on further studies. Furthermore, it is necessary in future studies to explore whether this tool is useful in practice, in which our main criterion would

be the degree to which it supports parents. As this tool in its current state may already be helpful for parents, it will be published on several websites.

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Appendix 2

Appendix, deel 2

Aandachtspunten kwaliteit van leven

Hulpmiddel voor ouders bij het nadenken over kwaliteit van leven van hun kind met (zeer) ernstige verstandelijke en meervoudige beperkingen.

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Inleiding

Dit is een hulpmiddel voor ouders¹ van kinderen of volwassenen met (zeer) ernstige verstandelijke en meervoudige beperkingen ((Z)EVMB), die verantwoordelijk zijn voor beslissingen over hun kind. Dit hulpmiddel kan gebruikt worden bij het nadenken over de kwaliteit van leven van hun kind, bijvoorbeeld bij medische beslissingen waarbij kwaliteit van leven een rol speelt. Dit hulpmiddel is ontwikkeld binnen een onderzoek naar kwaliteit van leven van mensen met (zeer) ernstige verstandelijke en meervoudige beperkingen ((Z)EVMB)². In dit onderzoek hebben wij aan ouders, persoonlijk begeleiders en artsen gevraagd wat zij goede kwaliteit vinden en wanneer ze gaan twijfelen aan de kwaliteit van leven van een persoon met (Z)EVMB. Uit deze interviews zijn aandachtspunten naar voren gekomen die kunnen helpen om na te denken over kwaliteit van leven van het kind of volwassene.

Gebruik

Hoe?

Dit hulpmiddel kan u helpen om na te denken over kwaliteit van leven van uw kind. Er worden aandachtspunten genoemd die uit interviews met ouders, persoonlijk begeleiders en artsen naar voren gekomen zijn. Deze kunt u langslopen en kijken of een bepaald aandachtspunt voor uw kind van toepassing is en hoe dat er bij uw kind uitziet. Er kunnen ook aandachtspunten zijn die u niet van toepassing vindt voor uw kind. Zo loopt u alle aandachtspunten langs en vult u in wat voor uw kind van toepassing is en hoe dat eruitziet. Op deze manier kan het u helpen om de kwaliteit van leven van uw kind, waar u (intuïtief) een idee over heeft, onder woorden te brengen.

Waarvoor?

Er zijn verschillende situaties waarin nadenken over de kwaliteit van leven van uw kind nodig kan zijn. Het meest bekend zijn situaties waarin u moet nadenken over medische beslissingen³. U kunt dit hulpmiddel bijvoorbeeld gebruiken als u een gesprek moet aangaan met artsen over een toekomstige medische beslissing voor uw kind.

¹ Hier kan ook gelezen worden: mensen die verantwoordelijk zijn voor beslissingen over een persoon met (Z)EVMB.

² 'Kwaliteit van leven van mensen met (Z)EVMB'. Onderzoekers: Prof. Dr. D. Willems, Prof. Dr. J.B. van Goudoever, Dr. E. Olsman, Drs. A.M. Nieuwenhuijse. Onderdeel van het project: 'Kwaliteit van Leven en Gezondheid' (NWO, 2019).

³ Zie Dr. M. de Vos (2019): 'Als je kind niet zelf kan beslissen...'

Voorbeeld

U moet beslissen over het uitvoeren van een scolioseoperatie bij uw kind. U heeft bij een van de aandachtspunten aangegeven dat uw kind het meest geniet van omrollen en zo de omgeving ontdekt. Het kan dan een vraag zijn in het gesprek met de arts of uw kind nog zal kunnen omrollen na de operatie.

Ook kunt u bijvoorbeeld een rangorde aanbrengen in de verschillende aandachtspunten. Zo krijgt u inzicht wat u het allerbelangrijkste vindt voor uw kind. U kunt er ook voor kiezen om deze aandachtspunten door meerdere mensen in uw omgeving te laten invullen voor uw kind. Dat kan iemand uit uw gezin zijn, maar ook iemand die zorg verleent aan uw kind. Dit kan dan weer een aanknopingspunt zijn voor een gesprek over de kwaliteit van leven van uw kind of over de zorg aan uw kind.

Welke aandachtspunten?

In dit hulpmiddel benoemen wij verschillende aspecten van kwaliteit van leven, namelijk: emoties en gevoelens van uw kind, de fysieke/lichamelijke situatie van uw kind, de mate van zelfstandigheid en aspecten in de relationele sfeer.

Daarnaast vragen wij u te kijken of er iets veranderd is of gaat veranderen op dat gebied. Wij hebben namelijk in de interviews opgemerkt dat kwaliteit van leven niet “statisch” is. Als een kind bijvoorbeeld heeft kunnen lopen en dat door achteruitgang niet meer kan, dan zien sommige ouders dat als vermindering van de kwaliteit van leven. Dat kan anders zijn voor ouders van een kind dat nooit heeft kunnen lopen. Iets anders wat uit ons onderzoek bleek, is dat kwaliteit van leven van ouders en kwaliteit van leven van hun kind “door elkaar kunnen lopen”. Dat wil zeggen dat de kwaliteit van leven van uw kind, uw kwaliteit van leven kan beïnvloeden, en andersom. Sommige ouders gaven zelfs aan dat die onlosmakelijk met elkaar verbonden waren. Andere ouders zagen dit niet zo of niet zo sterk. Daarom hebben we ook een aandachtspunt toegevoegd waarin we vragen hoe het met u is.

Omgeving

Uit ons onderzoek is gebleken dat de omgeving grote invloed heeft op de kwaliteit van leven van mensen met (Z)EVMB. Dat komt omdat deze kinderen en volwassenen volledig afhankelijk zijn van anderen in alle aspecten van het dagelijks leven.

U kunt ervoor kiezen om ook aandacht aan te besteden aan de omgeving bij het in kaart brengen van de kwaliteit van leven van uw kind. De lijst met factoren, die van invloed kunnen zijn, is echter eindeloos. Ouders noemden bijvoorbeeld: faciliteiten in huis, vervoer van hun kind, hulpmiddelen van hun kind, medewerking van

leveranciers en gemeentes, de zorg die hun kind krijgt in dag- en woonvoorzieningen, tot aan de faciliteiten die de maatschappij biedt om voor hun kind te zorgen. Als laatste vragen wij dus of er aandachtspunten vanuit de omgeving zijn die grote invloed hebben op de kwaliteit van leven van uw kind.

Aandachtspunten kwaliteit van leven van het kind

1. Emoties/gevoelens van uw kind

<p>Positieve emoties <i>bijv. vreugde, plezier hebben, genieten van dingen, interesse tonen</i></p> <p>Hier kunt u bijvoorbeeld in kaart brengen waar uw kind het meest van geniet, of wat voor u het meest belangrijk is waar uw kind van geniet.</p>	
<p>Negatieve emoties <i>bijv. angst en paniek, somber zijn, boos, agressie, huilen</i></p> <p>Hier kunt u bijvoorbeeld aangeven wat u het meest “erg” vindt.</p>	
<p>Mate van beïnvloeding van deze gevoelens door anderen</p> <p>Hier kunt u aangeven of u makkelijk de positieve gevoelens kunt oproepen, of de negatieve gevoelens kunt verminderen.</p>	

<p>Veranderingen op het gebied van emoties/gevoelens bij uw kind</p> <p>Hieronder kunt u aangeven of de situatie veranderd is de (afgelopen) tijd. Ook kunt u kijken of u op dit gebied een verandering verwacht in de toekomst ten positieve dan wel ten negatieve.</p>

2. Fysieke/lichamelijke situatie van uw kind

<p>Algemene conditie van uw kind</p> <p>Hier kunt u aangeven of u de gezondheid van uw kind over het algemeen goed vindt of dat u vindt dat uw kind veel ziek is.</p>	
<p>Lichamelijke ongemakken <i>bijv. pijn, benauwdheid, moeheid, spierslape, temperatuurswisselingen, spierspanningen, epileptische aanvallen, automutilatie</i></p>	
<p>Slaappatroon <i>bijv. veel slapen of juist heel weinig</i></p>	
<p>Problemen in het eet- en drinkgedrag <i>bijv. weinig of veel eetlust, braken, verslikken, sondevoeding</i></p>	
<p>Problemen met de ontlasting <i>bijv. obstipatie, diarree</i></p>	

<p>Veranderingen op lichamelijk gebied bij uw kind</p> <p>Hieronder kunt u aangeven of de situatie veranderd is de (afgelopen) tijd. Ook kunt u kijken of u op dit gebied een verandering verwacht in de toekomst ten positieve dan wel ten negatieve.</p>

3. Mate van zelfstandigheid van uw kind

<p>Zichzelf duidelijk maken <i>bijv. aangeven of het iets wel/niet wil</i></p>	
<p>Keuzes maken tussen verschillende opties als deze geboden worden <i>bijv. broodbeleg of een speeltje</i></p>	
<p>Zelfstandig ergens naar toe bewegen (al dan niet met hulpmiddelen)</p>	
<p>In staat zijn om bepaalde dagelijkse activiteiten zelfstandig uit te voeren <i>bijv. met een vork eten</i></p>	

Veranderingen op het gebied van zelfstandigheid van uw kind

Hieronder kunt u aangeven of de situatie veranderd is de (afgelopen) tijd. Ook kunt u kijken of u op dit gebied een verandering verwacht in de toekomst ten positieve dan wel ten negatieve.

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4. Aspecten in de relationele sfeer

<p>Contact maken met anderen in positieve of negatieve zin (bijv. door te lachen)</p> <p>Hier kunt u ook aangeven of er veel mensen zijn in de omgeving van uw kind met wie uw kind contact maakt/heeft.</p>	
<p>Communicatie</p> <p>Hier kunt u aangeven of en op welke manier uw kind communiceert.</p>	
<p>Veel liefde rondom uw kind <i>bijv. is er naast een liefdevol gezin, ook een liefdevolle dagbesteding of logeeromgeving voor uw kind.</i></p>	
<p>Gevoel van wederkerigheid <i>bijv. in de relatie van uw kind met u of met anderen</i></p>	

<p>Veranderingen op het gebied van zelfstandigheid van uw kind</p> <p>Hieronder kunt u aangeven of de situatie veranderd is de (afgelopen) tijd. Ook kunt u kijken of u op dit gebied een verandering verwacht in de toekomst ten positieve dan wel ten negatieve.</p>

5. Kwaliteit van leven van de ouder/verzorger

Kunt u iets zeggen over uw eigen kwaliteit van leven? Hoe gaat het met u?
Heeft dit (hoe het met u gaat) invloed op de kwaliteit van leven van uw kind? Zo ja, op welke manier?

6. Aandachtspunten uit de omgeving

Zijn er aandachtspunten uit de omgeving die grote invloed hebben op de kwaliteit van leven van uw kind
<i>Bijv. faciliteiten in huis, vervoer van uw kind, hulpmiddelen van uw kind, medewerking van leveranciers en gemeentes, de zorg die uw kind krijgt in dag- en woonvoorzieningen etc.</i>

Dankwoord

Het is gelukt, het proefschrift is geschreven! Bloed, zweet en tranen: het leek er verdacht veel op, vooral dat laatste...

Het was niet gelukt zonder de hulp en ondersteuning van veel mensen, die me hebben geholpen met het onderzoek, met het schrijven, met die stomme computer of die mij hebben aangehoord, al die jaren. Mijn dank is groot!

Allereerst gaat mijn dank uit naar alle mensen die bereid waren zich te laten interviewen. De artsen, de persoonlijk begeleiders en de moeders. Het waren zeer inspirerende gesprekken die we hadden en ze gaven mij antwoorden, maar zorgden ook voor weer meer vragen. Ook de ouders in de focusgroepen wil ik heel hartelijk danken. Dank voor jullie openheid over dit tere onderwerp: de kwaliteit van leven van je kind. Jullie spraken over de mooie kant van het leven met jullie kind, maar ook over de moeilijke momenten. Gelukkig vonden jullie elkaar als ouders veelvuldig in deze gesprekken. De interviews en de focusgroepen waren de mooiste momenten van dit onderzoek.

Mijn promotoren en copromotor, mijn dank aan jullie is groot. Dick Willems, je deskundigheid, intelligentie, ondersteuning, kritische blik en geduld heb ik zeer gewaardeerd. Jouw vrije geest, altijd op zoek naar net een beetje anders, was voor mij soms onvoorspelbaar en daardoor voor mij niet altijd makkelijk. Ik ben meer van de structuur en controle. Toch heb jij mij hierdoor geleerd om nog beter te kijken en uitgedaagd om nieuwgierig te blijven naar de vragen over kwaliteit van leven van mensen met (Z)EVMB. Ook bleef je kritisch over mijn schrijven wat ertoe geleid heeft dat er nu een proefschrift ligt. Wij blijven voorlopig samenwerken binnen de onderzoeksprojecten die nu lopen en ook daarin ben ik dankbaar voor je altijd deskundige en verrassende inbreng. Hans van Goudoever, jouw niet aflatende steun en het geloof in mij dat ik dit onderzoeksproject en proefschrift tot een goed einde zou brengen, heeft mij heel erg gesteund. Ook als lid van de Raad van Toezicht van Omega steunde je mij en promoveerde mijn onderzoek zowel in de Raad van Toezicht als daarbuiten. Je kritische en genuanceerde blik als het ging om de betrokkenheid van artsen bij de kwaliteit van leven van mensen met (Z)EVMB heeft dit onderzoek verrijkt. Dank daarvoor. Erik Olsman, mijn copromotor, dank voor onze vele gesprekken over het onderzoek, maar vooral ook voor alle gezelligheid en de lol die we hadden. Heel erg belangrijk. Maar zeker ook voor al die tijd (echt heel veel) die jij in mij gestoken hebt, even snel wat op de computer doen voor me, samen onderzoek doen, al mijn stukken lezen, herlezen en becommentariëren met deskundig commentaar, me opbeuren als ik het niet meer zag zitten en mij altijd weer aan het lachen brengen. Mijn dank is groot!

Raoul Hennekam, Marc Benninga, Bea Maes, Hans Reinders en Sylvia Huisman, dank voor jullie bereidheid om zitting te nemen in de promotiecommissie en mijn proefschrift te beoordelen. Ook dank aan de leden van de projectgroep: Michael Echteld, Gertrud Gosenshuis en Eeke

Harting, die zo vaak met ons hebben meegedacht over het onderzoek en aan leden van de adviesgroep van dit onderzoeksproject: Jacqueline van Oosterom, Geraldine Raap, Johannes Verheijden, Jos Hiel, Riet Niezen, Carla Vlaskamp, Hans Reinders, Alice Padmos, Bert Bos en Mirjam de Vos.

Dit onderzoeksproject heeft niet kunnen plaatsvinden zonder de medewerking van Omega. Sowieso het besluit in 2014 van de Raad van Toezicht, met Gert Grift als voorzitter in die tijd, om mij te detacheren naar het AMC zodat ik de tijd kreeg om dit onderzoek te doen. Verder dank ik de leden van de Raad van Toezicht, Nienke Sondern, Frank Nivard, Justin Lintjer, Marinus Knulst, Yvette Paludanus, Angelina Bakker, Mark Companjen en de huidige voorzitter Hedwig Slot voor hun meebeleven van dit project. Ook de stichting Vrienden van Omega, Fred Plukker, Wim van Minnen, Marc Benninga, Justin Lintjer, Marinus Knulst en Gert Grift, dank ik voor hun oprechte belangstelling naar de voortgang van dit onderzoek. En vooral Leendert Krol, eerste voorzitter van de Raad van Toezicht, alhoewel al sinds 2007 overleden, jij bent het geweest die me altijd heeft voorgehouden dat je je hele leven moet blijven leren en ontwikkelen.

Maar ook alle (emotionele) steun die ik heb mogen ontvangen. Van het MT, eerst Hanny Hoeijmakers en Nicolien Blokhuis, later Ellen van Breemen, Ria Weitkamp, Heleen Vissers en recent ook Helga Balk en Sebastian Kampa. En niet te vergeten de steun van Gerard Nijssen, die altijd voor- en meedenkt en mijn werk ontlast waar hij kan. Maar ook de steun van de Ouderraad en belangstelling van de OR en medewerkers van Omega is erg belangrijk geweest. Een speciaal dank aan het groepje vaders, Marc van Gemert, Robert Weijs en Henk Siemonsma die vooral in het begin, met mij en mijn collega Hanneke Vrielink, meedachten over de kwaliteit van hun leven van mensen met (Z)EVMB, en dan vooral van hun dochters. De ouders van Omega dank ik voor hun vertrouwen in Omega en in mij. Mijn ervaringen met jullie en met jullie kinderen zijn een verrijking van mijn leven en hebben gemaakt dat ik het pad van onderzoek ben opgegaan.

Vanuit Omega werd ik gedetacheerd naar het AMC en ook daar werd ik aan alle kanten ondersteund en begeleid. Het secretariaat, eerst Alice Karsten en later Annelies van der Geest die altijd alle vragen konden beantwoorden, de leden van filosofie van de zorg die meedachten en kritisch reflecteerden op onderdelen van het onderzoek, de leesclubjes over Foucault en Nussbaum, de mensen in de lessen in 'empirical ethics' en basis-ethiek, Maartje Hoogsteyns (mijn voormalig kamergenoot) en Kasper Kruithof met wie ik nu intensief samenwerk in de vervolgonderzoeken, allemaal heel veel dank.

Het schrijven van een proefschrift is zowaar geen sinecure, vooral niet voor een digibeet zoals ik. Zonder de hulp van Caroline Nieuwenhuijse-Ward (en Bas) en Sandra Valkema (en Henk Siemonsma) was dit proefschrift er niet gekomen. Dank voor het helpen en voor jullie geduld.

Henk en Nancy, dank dat Britt haar levensloop als rode draad door dit proefschrift mag lopen.

Dank Carla van Burik (en Hans van Goudoever dat ik haar mocht “lenen”) voor alle hulp bij het organiseren van het symposium.

Mijn vrienden en vriendinnen, dank voor alle steun en het aanhoren van mijn wel-en-wee op onderzoeksgebied tijdens de vele etentjes en zeiltochtjes in de afgelopen jaren.

Familie, nichten en neven, tantes, achternichtjes, wij houden van feestjes en bijeenkomsten, dat er nog maar vele mogen volgen, om te beginnen met de promotiedag.

Mijn broers, (schoon)zus(sen) Peter en Jolande, Kitty en Haitze, Gilbert, Leo en Karolien, Karin, ik ben erg dankbaar en blij dat jullie er zijn!

Mijn ouders, ik geloof zeker dat ze trots zouden zijn geweest op hun jongste dochter. Zij hebben mij alle mogelijkheden gegeven in mijn leven om mij te kunnen ontplooien en hebben mij daarin altijd gesteund. Ze hebben ons opgevoed met de waarden: hard werken, goed voor andere mensen zorgen, een goed mens zijn en vooral gewoon blijven en ‘niet-naast-je-schoenen-gaan-lopen’. Mede door hen ben ik geworden die ik ben.

Gilbert, mijn broer, wij schelen nog geen jaar. Mama noemde ons altijd ‘haar tweeling’ als ik jarig was. Dan waren we weer twee weken even oud totdat jij jarig was. Wij waren onafscheidelijk in onze jeugd en nog altijd hebben we een hechte band. Ik ben dan ook blij dat jij mijn paranimf bent.

Josje, mijn vriendin. Jij laat mij in de spiegel kijken (letterlijk en figuurlijk, ‘die trui kan echt niet meer’), jij geeft advies (gevraagd en ongevraagd zoals dat heet) en je leeft met me mee in lief en leed. We zien en spreken elkaar veel te weinig, mede door corona. Ik hoop op betere tijden en nog heel veel jaren om met je te praten, wandelen, eten, naar de kapper te gaan, te huilen en te lachen. Ik ben heel blij dat jij mij bij staat op deze promotiedag en mijn paranimf bent.

Hans, waar zal ik beginnen? Het is niet helemaal gegaan zoals we hadden bedacht. Jij zou met pensioen gaan en ik zou minder gaan werken. Toen kwam het onderzoek langszij (om maar in de scheepstermen te blijven), en ik ging alleen maar meer werken. Dankzij jou kreeg ik (nog net) genoeg buitenlucht en beweging, want je nam me mee fietsen, wandelen en varen. Ik heb enorme bewondering voor je geduld en daarvoor heel veel dank. Partners van promovendi hebben het niet makkelijk! Lieve Hans, er komen betere tijden, met nog meer buitenlucht, uitjes, bootjes, hotelletjes, harinkjes bij de lunch en vooral genieten van het leven en elkaar!

About the author

Marga Nieuwenhuijse was born on May 25, 1959 in Bennekom. After her secondary education (VWO) at the Christelijk Streek Lyceum in Ede, she started studying “pedagogiek” at the Free University in Amsterdam. She completed her studies in 1984 and majored in “speciale pedagogiek” with Han Nakken as her tutor. During her studies, she cared for persons with physical disabilities in several institutions.

After her studies, she became manager in “Amstelrade” an institution for persons with physical disabilities. In 1989, she became principal of Omega in Amsterdam, a specialised day-care centre for children with profound intellectual and multiple disabilities. In 1998, she became managing director of Omega. Day-care centre Omega has also been offering day treatment for adults since 2009 and has grown into an expert centre for persons with profound intellectual and multiple disabilities.

From 2003 until 2005, she studied towards a Master of Business Administration (Health) at the Erasmus University in Rotterdam and she has chaired Platform EMG since 2008. In 2014, she started her PhD at the AMC in Amsterdam and combined this with her work at Omega. Marga lives with her husband Hans in Vinkeveen.



Name PhD student: Marga Nieuwenhuijse
PhD period: June 2014 - January 2022
Name PhD supervisor: Prof. Dr. Dick Willems, Prof. Dr. Hans van Goudoever
Co-supervisor: Dr. Erik Olsman

1. PhD training

General courses	Year	Workload (Hours/ECTS)
PubMed (e-learning course)	2014	0.1
Endnote	2014	0,1
Scientific Writing in English for Publication	2015	1.5
Oral presentation in English	2017	0.8
Qualitative Health Research	2014	1.9

Workshops	Year	Workload (Hours/ECTS)
Research meeting 'Philosophy/Ethics of care',	2014-2021	Monthly
Workshop 'Basics of normative ethics'	2019/2020	0.4
Workshop 'Make ethics happen'	2019	0.4
Reading group on Ethics, AMC/Amsterdam UMC	2017, 2019	0.4

Presentations	Year	Workload (Hours/ECTS)
Workshop op het jaarlijkse congres Landelijk Platform EMG 14 maart 2016	2016	0.4
Dutch Academy of Childhood Disability; Klimmendaal Arnhem oktober 2016	2016	0.2
Gedragsdeskundigen Landelijk Platform EMG 14 november 2016	2016	0.4
Met een panel van ouders van Dagbehandelingscentrum Omega de concept checklist KvL besproken en feedback gekregen 22 mei 2019	2019	0.4
Ouderavond op Dagbehandelingscentrum Omega in Amsterdam om ouders te informeren over het onderzoek en de resultaten 1 oktober 2018	2018	0.4
Kennisplein, Platform EMG en Vilans; EMB in het vizier: "Kwaliteit van Leven van mensen met (Z)EVMB. Vanuit het perspectief van persoonlijk begeleiders 4 februari 2019	2019	0.4
Diverse presentaties medewerkers Omega	2016-2019	0.4
Slotconferentie NWO-programma Kwaliteit van Leven en Gezondheid 6 juni 2019	2019	0.4

(Inter)national conferences	Year	Workload (Hours/ECTS)
Informal conference in AMC, met wetenschappers van Disability studies Nederland, wetenschappers AMC, Prof. Simo Vehmas (hoogleraar Disability Studies) uit Finland 23 maart 2016	2016	0.5
Presentaties op congres Disability studies in Amsterdam: "Belonging as important element of QoL in persons with PIMD: A qualitative study on physicians' perceptions" 13 November 2017	2017	0.5
IASSIDD (International Association for the Scientific Study of Intellectual and Developmental Disabilities) 6 Augustus 2019: "Quality of life of persons with Profound Intellectual and Multiple disabilities. The perspective of professional caregivers." IASSIDD 7 Augustus 2019: "Physicians perception on Quality of Life of persons with PIMD. A qualitative study." IASSIDD 7 augustus 2019: panel lid bij roundtable van de Special Interest Research Group "Ethics" IASSIDD 8 augustus 2019: panel lid bij roundtable van de Special Interest Research Group "Quality of Life" van de IASSIDD.	2019	1.5
IASSIDD 7 juli 2021 "A life worth sharing", understanding quality of life of persons with PIMD.	2021	0.5

2. Teaching

	Year	Workload (Hours/ECTS)
Tutoring, practica ethiek BA studenten geneeskunde	2014-2021	1,5
Tutoring, practica ethiek, Msc studenten geneeskunde	2014-2019	0,75
Debatteren Msc studenten geneeskunde	2018, 2020	1.0
Moreel beraad Msc studenten geneeskunde	2019	0.25

3. Parameters of Esteem

Grants	Year
ZonMW: Gewoon Bijzonder, Nationaal Programma Gehandicapt Subsidieronde / Subsidy round : Gewoon Bijzonder – EMB Projecttitel / Project title : Kwaliteit van leven en veroudering van mensen met EMB.	2018
ZonMW: Programma Langdurige Zorg en Ondersteuning Subsidieronde / Subsidy round : Praktijkgericht onderzoek naar de relatie tussen de cliënt, naasten en/of mantelzorgers en professionals in de langdurige zorg en ondersteuning Projecttitel / Project title : Tacit knowledge: een kwalitatieve studie naar het gebruik en de overdraagbaarheid van impliciete kennis in de zorg en ondersteuning voor mensen met (Z)EVMB	2020

4. Publications

Peer reviewed	Year
A. Nieuwenhuijse, D. Willems, J. van Goudoever, A. Echteld and E. Olsman, "Quality of life of persons with profound intellectual and multiple disabilities: A narrative literature review of concepts, assessment methods and assessors," <i>Journal of Intellectual & Developmental Disability</i> , vol. 44, no. 3, pp. 1-11, 2017.	2017
A. Nieuwenhuijse, D. Willems and E. Olsman, "Physicians' perceptions on Quality of Life of persons with profound intellectual and multiple disabilities: A qualitative study," <i>Journal of Intellectual & Developmental Disability</i> , vol. 45, no. 1, pp. 176-183, 2020.	2020
A. Nieuwenhuijse, D. Willems, J. van Goudoever en E. Olsman, "The perspectives of professional caregivers on quality of life of persons with profound intellectual and multiple disabilities: a qualitative study," <i>International Journal of Developmental Disabilities</i> , 2020, published online: DOI: 10.1080/20473869.2020.1737469	2020
A. Nieuwenhuijse, D. Willems, J. van Goudoever en E. Olsman, "The perspective of parents on the assessment of quality of life of their children with profound intellectual and multiple disabilities" submitted 2021	2021
E. Olsman, A. Nieuwenhuijse and D. Willems, "Witnessing Quality of Life of persons with profound intellectual and multiple disabilities. A practical-philosophical approach," <i>Health Care Analysis</i> , 2021.	2021
E. Olsman, A. Nieuwenhuijse and D. Willems, "'My son has lost dignity': Dignity of persons with profound intellectual and multiple disabilities. A conceptual study." submitted, 2021.	2021
Kasper Kruithof, Lisa IJzerman, Appolonia Nieuwenhuijse, Sylvia Huisman, Alice Schippers, Dick Willems & Erik Olsman: Siblings' and parents' perspectives on the future care for their family member with profound intellectual and multiple disabilities: A qualitative study, <i>Journal of Intellectual & Developmental Disability</i> , 2021, published online DOI: 10.3109/13668250.2021.1892261	2021
Kasper Kruithof, Erik Olsman, Appolonia Nieuwenhuijse & Dick Willems "I hope I'll outlive him": A qualitative study of parents' concerns about being outlived by their child with profound intellectual and multiple disabilities, <i>Journal of Intellectual & Developmental Disability</i> , 2021 Published online DOI: 10.3109/13668250.2021.1920377	2021
Kasper Kruithof, Erik Olsman, Appolonia Nieuwenhuijse & Dick Willems: "Parents' views on medical decisions related to life and death for their ageing child with profound intellectual and multiple disabilities: a qualitative study" submitted, 2021	2021

Dit proefschrift gaat over kwaliteit van leven van mensen met (zeer) ernstige verstandelijke en meervoudige beperkingen. Het leven van Britt Siemonsma, een meisje met (Z)EVMB, wordt als voorbeeld van een leven vol kwaliteit beschreven in dit proefschrift.