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## A fine balance and a shared learning journey: Exploring healthcare engagement through the experiences of youth with neuromuscular disorders

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**Title:**

A fine balance and a shared learning journey: exploring healthcare engagement through the experiences of youth with neuromuscular disorders

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## Abstract

**BACKGROUND:** Youth with Neuromuscular Disorders (NMD) who are wheelchair users can now survive well into adulthood if their multisystem comorbidities are prudently managed. Uptake of health behaviors may optimize their health outcomes.

**OBJECTIVE:** To explore youths' perceptions of health, health behaviors and healthcare engagement.

**METHODS:** This qualitative study purposefully recruited 11 youth with NMD from a concurrent, population-based study for variability of age, gender, type of NMD and their ratings of motivation and engagement. Interview data were analyzed and synthesized by thematic content.

**RESULTS:** Participants perceived healthcare engagement as being given tools (knowledge and responsibility) and using them to maintain their finely balanced health. Nested in adequate social, emotional and physical support, they took responsibility for creatively integrating health behaviors they felt were informed by credible knowledge, gained primarily through personal experience.

**CONCLUSION:** Cognizant of their compromised health, youth with NMD in this study were motivated to maintain their physical health. Limited NMD condition specific knowledge challenged youths' uptake of health behaviors. They valued a learning partnership with their healthcare professionals. By embracing the youth's experience based knowledge and through facilitating supportive relationships, healthcare professionals co-construct youth's healthcare engagement that may optimize health behaviors and outcomes.

### **Keywords**

Adolescents / youth / young people

Neuromuscular Disorders

Healthcare engagement

Perceptions / experiences

Health education

### **Glossary**

CMD – Congenital Muscular Dystrophy

DMD – Duchenne Muscular Dystrophy

HCP – Healthcare Professionals

ICF – International Classification of Functioning, Disability and Health

IPA – Interpretative Phenomenological Analysis

NMD – Neuromuscular Disorders

NIV – Non Invasive Ventilation

QoL – Quality of Life

SMA – Spinal Muscular Atrophy

## Body of Text

### 1. Background

Neuromuscular Disorders (NMD) are genetically acquired, non-curable, rare diseases causing severe muscle weakness. Spinal Muscular Atrophy Type II (SMA II) and Duchenne Muscular Dystrophy (DMD) are examples of more prevalent NMD that necessitate wheelchair dependence in early childhood or adolescence (Katirji, Kaminski, & Ruff, 2013). Co-morbidities such as scoliosis and respiratory insufficiency relate to progressive muscle weakness and compromise physical health and life expectancy (Wang et al., 2012; Wang et al., 2007). Timely and appropriate health management can limit the onset and severity of body systems co-morbidities (Strehle, 2009). Best practice health management includes the use of steroids in DMD, spinal fusion and non-invasive ventilation (NIV) (Bach & Martinez, 2011; Bushby et al., 2010a, 2010b; Hull et al., 2012).

Youth with chronic disorders who are transitioning into the adult healthcare system are expected to develop autonomy in their healthcare, including making informed and independent choices and thereafter self-managing complex treatments (Gruman et al., 2010; Suris & Akre, 2015). Youth living with complex health conditions such as NMD are at particular risk of preventable illnesses that may lead to suboptimal health outcomes if self-management in early adulthood is not realized (Maslow & Chung, 2013; Sawyer & Ambresin, 2014) and their self-management may be hampered by social attitudes towards their uncertain lifespan (Gibson, Zitzelsberger, & McKeever, 2009; van Huijzen & van Staa, 2013). Current research informing transition from youth to adult healthcare services seeks to understand socio-ecological factors that impact on young peoples' development of self-management skills (Javalkar et al., 2016; Johnson, McMorris, Mapellentz, & Scal, 2015; Schwartz, Tuchman, Hobbie, & Ginsberg, 2011).

Uptake and adherence to health management behaviors are particularly challenging for adolescents (Sawyer, Drew, Yeo, & Britto, 2007). Adherence is frequently less than when they were

younger and more reliant on their parents' problem-solving and coping abilities (Shakkottai, Kidwell, Townsend, & Nasr, 2015). Even though adolescents with chronic conditions can be knowledgeable about their health, their individual journeys of identity formation and developing autonomy influence their choices to take on and maintain health behaviors, particularly if these are perceived as counter to their desire to appear and feel 'normal' (Bucks et al., 2009). For adolescents with NMD the introduction of respiratory support such as NIV is crucial for survival and its uptake can be problematic (Amin, Al-Saleh, & Narang, 2016). Limited information sharing is one potential factor (Nierse, Abma, Horemans, & Van Engelen, 2013; van Huijzen & van Staa, 2013) but little is known about how these adolescents acquire an understanding of their condition and other factors that may influence their uptake and maintenance of life-sustaining health behaviors.

The young persons' developing partnership with their adult healthcare professionals (HCP) can facilitate the learning of self-management skills (Johnson, McMorris, et al., 2015; van Staa & Sattoe, 2014). Studies involving youth with NMD have used the term engagement when referring to notions of shared decision making (Hull et al., 2012) and self-management (Nierse et al., 2013). Bright, Kayes, Worrall, and McPherson (2015) describe healthcare engagement as a co-constructed process and a state of connection between the HCP and patient. However, this conceptual review excluded child and adolescent services. No study sourced to date has explored healthcare engagement as a socio-ecological factor in the successful transition of youth living with rare disease.

This qualitative study sought to understand more deeply the experiences and perceptions of youth with NMD around health, health behaviors and healthcare engagement.

## 2. Methods

An Interpretative Phenomenological Analysis (IPA) approach was used for this qualitative study (Smith & Osborn, 2008). The study was approved by the Human Research Ethics Committee of the

University of Notre Dame Australia (REF 010144F). Youth and their parents (if youth aged under 18yo) were provided with full study information prior to providing consent.

### *2.1 Participants*

Youth aged 14-21 years were purposively sampled from a concurrent national study of wellbeing in youth with NMD who are wheelchair users ([www.mylifemyvoice.org](http://www.mylifemyvoice.org)), for variability of age, gender, type of NMD, severity of co-morbidities and ratings of motivation and engagement. Sampling continued until data saturation was reached, whereby no new themes emerged from two successive interviews. Thirteen youth were contacted by the first author (VT). Two declined to be interviewed; one 21-year-old was experiencing family instability and one 19-year-old did not give reasons. The 11 participants' characteristics are described in Table 1.

*Insert Table 1a&b about here*

### *2.2 Data Collection*

The semi-structured interview guide was developed from definitions of engagement specific to healthcare (Bright et al., 2015) and high school education (Liem & Martin, 2012; Martin, 2007). Questions explored how youth felt about their health, health behaviors, motivators, supports, learning about health, and what healthcare engagement meant to them (see Appendix). Participants were asked specific probing questions to explore reasons for their perceptions (e.g. my support is pretty good). Probing questions to elicit deeper insights included: "How do you know this?"; "Can you describe this experience?"; and "Why do you think that?" (Epstein et al., 2016). The guide was piloted with two young persons (YP) living with NMD leading to minor adjustments. These data were included in the analysis.

All interviews were conducted by the first author (VT), who disclosed to interview participants her background and context as a PhD student, mother, and physiotherapist with clinical and



research experience with young people who are wheelchair users. Seven interviews were conducted face-to-face at the participants' home and four via Skype with participants living interstate. All participants were in a secluded environment away from parents and others, except for one who preferred to be interviewed with his mother present. She assisted by repeating his words when speech was too soft to be recorded. Interviews lasted between 24 to 80 minutes. Following each interview VT kept a research diary reflecting on the interview's context and how participants had shared their accounts. All interview data were audio recorded and transcribed verbatim by VT with names and other identifiable information removed.

### *2.3 Data Analysis and Trustworthiness*

Data were analyzed by thematic analysis: initial individual coding and subsequent identification, categorizing and synthesis of common themes (Braun & Clarke, 2006). First, transcripts were read and re-read, assigning initial descriptive codes not limited to participants' actual words, but also commenting on the use of language and ideas of meaning 'between the lines' (Smith & Osborn, 2008). Initial codes were organized into an a priori coding framework based on the International Classification of Functioning, Disability and Health (ICF) (World Health Organisation, 2007) and domain elements of the interview questions (including behaviors, motivation, support and learning) (see Appendix).

Initial coding of five randomly selected interviews was completed independently by two authors (VT and CB), who subsequently met and discussed the emerging themes. Research diary entries were included in discussions, acknowledging a co-construction of emerging themes. All participants were e-mailed their coded interview transcript for checking and five of the 11 provided feedback. The independent coding and member checking process facilitated further review and confirmed the authenticity, trustworthiness and credibility of the analysis.

The coding framework was entered into NVivo 11 software (QSR International Pty Ltd, Burlington, MA). Key concepts were mapped and quotes from each transcript re-read to merge similar codes and synthesize findings into themes and subthemes. An audit trail of key decisions regarding the thematic synthesis was kept and formed the basis of discussion with the team of authors. Any discrepancies regarding the coding and thematic analysis were resolved through discussion until consensus was reached.

### 3. Results

Participants described health care engagement as being involved in maintaining their health as best they could with their experiences, knowledge and support. These data synthesized into four themes: participants' accounts of health behaviors; motivation; and perceived trusted, supportive, nurturing relationships; which appeared scaffolded by their notions of valuing expectations of responsibility and the sharing of credible knowledge (Figure 1). Subthemes, categories and sample quotes within each theme are provided in Tables 2 - 5.

*insert Figure 1 about here*

#### 3.1 Theme 1: *Finely balanced health*

*'Finely balanced health'* emerged from participants' descriptions of what good health meant to them and how they integrated health behaviors into their daily lives. Participants valued good health, with mental and physical health considered equally important. Physical health was described as the absence of an acute episode of illness and maintenance of their current physical status. Participants' recognized that they were more vulnerable than typically developing peers both to the repercussions of acute illness as well as to health intervention errors. Examples of health interventions that had caused rapid deterioration in their physical health or negatively impacted their mental health included: being given an incorrect anesthetic that caused cardiac arrhythmia;

misdiagnosed cardiac failure resulting in fluid overload; spinal bracing that limited respiratory capacity and accelerated postural weakness; and NIV application during a sleep study without prior preparation that led to fear of the device. Participants reflected that such experiences had increased their vigilance to question whether recommended health behaviors were applicable to themselves.

Participants strived to maintain their normalcy, describing how acute illness and additional health visits “put the brakes on” their usual activities and participation. To this extent, they focused on health behaviors that they felt maintained their health, such as preventing contact with people who were unwell, early antibiotic use to manage respiratory illness, good diet, enjoyable physical activity and sufficient sleep. Participants described innovative ways to be physically active within daily routines, including wheelchair sports, independent or assisted muscle and joint stretches whilst doing academic work, novel strengthening exercises, and incidental assisted standing during transfers. Four participants utilized knowledge gained through a variety of sources to problem solve. For example, YP6 had modified a stretching and strengthening routine advised by his physiotherapist utilizing an upturned coffee table to align his legs for stretches, and modified a garden chair with felt sliders on which he could sit and push it backwards for strength. Similarly, YP3 and YP4 integrated their mothers’ Pilates’ exercise with exercises recommended by their physiotherapist. Through this exercise YP4 regained the ability to bring one hand to her nose, enabling her to itch and no longer needing to ask for help with this task. Five participants felt they could be doing more to be physically active describing barriers including physical access (for example, to swimming/hydrotherapy), lack of allied HCP follow up, lack of motivation, distraction by screen time and fatigue.

Minimizing fatigue and maximizing respiratory health emerged as health priorities, captured in YP8’s comment of “*my breath is everything, really*”. Strategies to manage fatigue, get sufficient sleep and use nocturnal NIV were interrelated. All but one participant had experienced use of NIV. Three admitted to using their NIV less than prescribed. One young person felt that consistent use may

further weaken his respiratory muscle strength, a fear echoed by two others. One participant discussed not wanting to use NIV unless he felt he needed it.

*Insert Table 2 about here*

### 3.2 Theme II: Motivation - A dynamic process

Motivation was identified as a key theme which captured participants' accounts of dynamic intrinsic processes leading to the development of feelings of autonomy over their choice of health behaviors, and included the following sub themes.

*'Acquiring knowledge'* comprised learning about their condition from a variety of sources including parents, peers and HCPs. Information was perceived credible coming from someone they trusted, whom they perceived had expertise in their condition or who 'knows what it is like'. Prompted to reflect upon anything else they would like to learn, participants expressed 'wanting to know what their future holds'.

*'Sense of self'* captured feelings about body awareness, body image and their physical abilities. Four participants described comparing themselves to peers with other NMDs to form an idea of their own health, as in severity of scoliosis, physical strength or accepting that NIV was part of living with NMD. YP6 recalled how being talked about in the hospital environment required him to reflect on his condition and accept that he needed help.

*'Understanding importance to self'* encapsulated participants' contemplations of whether health behaviors were worth integrating into their daily routines. A process leading up to understanding a health behavior's importance was described, whereby participants weighed up whether the information they had acquired was credible, and how this fit with their sense of self and personal priorities. They talked about 'questioning' as a normal process for teenagers, especially in relation to perceived new information. Participants said they were more likely to value health behaviors if they if they felt the information they were given was credible and familiar, they felt an immediate

physical benefit (for example, feeling refreshed as opposed to more fatigued after nocturnal NIV use) or saw negative outcomes (for example, turned in feet in older peers who had not worn foot splints). In order to decrease perceived fear of NIV, one participant suggested normalizing talk of and increasing familiarity with NIV by enabling younger children to play around with ventilators in practical workshop sessions.

*'Accepting and valuing importance to self'* captured acceptance that some things can be changed, and some cannot, though all felt comfortable with their current health routines. A hope for future technological advances or medication that may make things easier was voiced by two participants. They talked of a sense of control over their decisions, including accepting the repercussions when these conflicted with health care advice. A lack of condition specific knowledge motivated one young person with CMD to take the risk of not using his nocturnal NIV as prescribed, accepting responsibility if his respiratory status should change (Table 3 – sample quotes).

*'Being goal directed and proactive'* captured feelings of autonomy and readiness to take responsibility for oneself, with resolve to maintain physical function in apprehension of possible future change in function. Proactive health behaviors were described as seeking timely support if they perceived risk to their health and being diligent to continue with their current health behaviors: *"the more practice you do the better you will be whatever you are doing (YP6)"*. Two participants felt a sense of new focus on their health behaviors after recently receiving a specific subtype diagnosis (at age 16 and 20), contrasting with the uncertainty about their future felt by two of the three participants with undiagnosed CMD. Participants described diverse interests including music, writing a novel, and a career goal to become an astrophysicist. Barriers and enablers to their sense of empowerment linked with perceived support from their family, friends', teachers' and HCPs support.

*Insert Table 3 about here*

### 3.3 Theme III: Trusted, supportive, nurturing relationships

*'Trusted, Supportive, Nurturing Relationships'* captured extrinsic factors amongst family, friends and HCP teams, including the following subthemes.

*'Emotional and social support'* described feeling understood and adequately supported so there was no need to worry. Four young people described humor at home and with the HCP's as being helpful support to manage their health behaviors. Participants felt empowered to take responsibility within perceived nurturing healthcare environments. Longstanding, trusting, non-judgmental relationships helped the young person feel safe to openly discuss and ask questions, making it was easier to learn about and manage their health. Lack of continuity of HCPs and perceived lack of interest were described as a barrier. YP1 recalled occasions of not bothering to ask a 'rough carer' for help with putting on his splints, as he did not trust this carer's competence, and feeling uncomfortable to request a 'rushed carer' to slow down driving around corners. Trusted HCP's were described as persons they had known for a long time, did not stigmatize wheelchair users as intellectually impaired, made an effort to understand their individual condition, included the young person in conversation, listened to their ideas, laid out options and shared problem solving. Participants who had not personally experienced such communication suggested peer mentors to assist as advocates in the patient-HCP consultation.

*'Practical support'* included ready access to their HCPs, access to information about care and funding options, regular health check-ups, coordinated care and teamwork amongst their HCPs. The physical support of equipment was essential, although the visibility or bulk of equipment could be a barrier to participation in activities. The unsolicited help of school friends with tasks requiring physical strength was appreciated, such as picking up a dropped pencil, as well as practical and sometimes financial help from extended family that enabled their parents some time off. All participants described their parents' unquestioning, always-ready-to-assist practical support as most

valuable in supporting their overall health and wellbeing, comparing themselves as healthier than peers who did not have access to such family support.

*Insert Table 4 about here*

### 3.4 Theme IV: Expectations and shared knowledge

*'Expectations and shared knowledge'* emerged as the key theme that linked Theme II: *Motivation* with Theme III: *Perceptions of trusted, supportive, nurturing relationships*. Expectations of adult independence and the sharing of credible knowledge epitomized participants' perceptions of healthcare engagement.

*'Expectations'* captured that participants expected to take responsibility for their health. Older participants highlighted a gap in expectations and knowledge sharing between pediatric and adult services. They perceived that adult services expected young people to already understand possible repercussions of non-adherence or late uptake of NIV, whereas pediatric services had tended to withhold information that may instigate 'difficult conversations', such as conversation around life expectancy and adherence to NIV. High school education programs about the risk of accidental death due to drug use were perceived as more urgent and normalized than conversation of gradual death due to respiratory muscle weakness. Being part of a sporting team where older athletes with DMD had passed away had helped lessen the trauma and normalize conversation around NIV use and life expectancy.

*'The need for shared health education'* captured how participants dealt with the paucity of condition-specific knowledge upon which to base some of their decisions. Participants highlighted how their parents had helped them acquire knowledge, through seeking and deciphering health information and reinforcing important advice that had been given during health consultations. Participants valued shared problem solving, especially when they had experienced either benefit or detriment from taking on health advice. Social media and personal connections with others living

with the same or similar conditions, both locally and internationally were named as very helpful to gain information, especially when they perceived that their HCPs disease specific knowledge was limited.

*Insert Table 5 about here*

#### 4. Discussion:

For youth with NMD, healthcare engagement appeared to be a co-constructed process, whereby integration and uptake of health behaviors (Theme I) was balanced on dynamic processes leading to intrinsic motivation (Theme II) and influenced by feelings of adequate support (Theme III). Engagement was facilitated and 'scaffolded' through expectations of responsibility and shared, credible knowledge (Theme IV) (Figure 1). Knowledge was a recurring category in all themes where the type, content and context of information sharing influenced perceptions of healthcare engagement. These newly identified personal and environmental factors were broadly consistent with theories of motivation and engagement emerging from the field of educational psychology (Martin, 2007) and have implications for HCP's role as health educators.

##### *4.1 Theme I: Finely balanced health*

Participants appeared to have some sense of autonomy in their choice of health behaviors, gained through co-constructed healthcare engagement and captured in the theme '*Finely balanced health*'. For example, sense of autonomy appeared strengthened in those who had solved problems with their HCPs contributing to the maintenance of their physical status. Conversely, health interventions associated with deterioration in physical health diminished feelings of control, but increased their vigilance to become knowledgeable as to their health needs. Condition specific health literacy is a need that is also identified amongst European youth with NMD (Nierse et al., 2013; Sonneveld, Strating, van Staa, & Nieboer, 2013). A frustration with the lack of condition specific knowledge is shared by patients with other rare diseases (Daker-White et al., 2015).



Participants spoke of integrating health behaviors that would best maintain their physical status, but their choices were difficult when knowledge about which health behaviors would be most helpful was limited. Participants easily integrated health behaviors based on mainstream health messages and practiced by the whole family. Participants' focus on good diet, exercise and mental health appeared informed by current, public health messages based on accessible, robust evidence. In contrast, unfamiliarity with NIV and lack of accessible disease specific evidence of its benefits, as well as mixed messages from peers, challenged uptake of NIV in the three participants who were not adhering to prescription. Though recent guidelines support the physical benefits of NIV for NMD (Hull et al., 2012), studies suggest that NIV may have both positive (Rousseau, Pietra, Blaya, & Catala, 2011) and negative impacts on psychosocial wellbeing and quality of life (QoL) (Katz et al., 2010; Mah, Thannhauser, McNeil, & Dewey, 2008). Similar to adolescents with other life-limiting conditions (Sawicki, Heller, Demars, & Robinson, 2014), our participants' deliberations appeared multi-faceted. Their reflections of type and source of knowledge warrant a closer look at the psychosocial factors impacting their uptake of life-sustaining health behaviors during adolescence.

#### *4.2 Theme II: Motivation*

*Motivation* was a fluid process of developing intrinsic motivation closely influenced by the external factors of Theme III: Supportive Relationships and Theme IV: Expectations and Shared Knowledge. The dynamic processes of valuing and accepting knowledge, culminating in a sense of autonomy to embrace health behaviors were founded upon acquisition of basic knowledge. All participants spoke of learning with and from their parents, re-affirming the importance of parents' health literacy in the healthcare of children with developmental disability (Keim-Malpass, Letzkus, & Kennedy, 2015).

Similar to youth with other chronic conditions (Björquist, Nordmark, & Hallström, 2015; Sonneveld et al., 2013), participants expected to become more autonomous in their health

management. In relation to being able plan for the future, participants appreciated the efforts of specialist NMD doctors' to identify their individual diagnosis and the focus this gave. However, even with a specific diagnosis, their challenge to find condition specific information about long term management reflects that although research to date has significantly advanced successful rare disease diagnosis (Orphanet, 2014), uptake of guidelines is still slow (Landfeldt et al., 2015). Longitudinal research informing rare disease outcomes, facilitated through international collaborative efforts, is still its infancy (Bladen et al., 2013). It is not surprising then that participants felt that experience-based knowledge gained from others living with similar conditions, both in person and through social media, was at times more credible and influential on their healthcare behaviors than knowledge gained from HCPs.

Participants' preference for and uptake of experience based knowledge from same age peers is reinforced in studies describing knowledge acquisition at condition specific youth summer camps (McCarthy, 2015) and online adolescent peer support groups (Johnson, Fuchs, Horvath, & Scal, 2015). Online patient to doctor learning has shown successful outcomes for adults with rare conditions (Kauw et al., 2015; Vennik, Adams, Faber, & Putters, 2014). The health behaviors of adolescents with complex conditions utilizing online, condition-specific networking resources needs further study.

The perceptions amongst participants around "knowing enough" were particularly interesting when exploring themes pertaining to NIV use. Differences between those embracing NIV and those hesitating appeared to relate to their familiarity with the device and how they felt NIV fit with their sense of self. The two participants who struggled with uptake of NIV had negative impressions of the device, perceiving that NIV use went against their individual identities of a vocalist and an athlete (electric wheelchair sports), both of which intuitively portray images of respiratory strength and fitness. These sentiments are consistent with findings that uptake of health behaviors in adolescents with cystic fibrosis can be limited by a wish to maintain a sense of normality (Lang, Martin, Sharples,

& Crowe, 2013). What little is known about the body image and psychosocial wellbeing of adolescents with NMD who are NIV users may be gleaned from studies reporting psychosocial domains of health related QoL outcomes. Youth who are both wheelchair and NIV users since early childhood score higher in psychological wellbeing (Young et al., 2007) and body image (Vuillerot et al., 2010) than their more physically able peers with NMD. Similarly, participants in this present study living with severe but unchanging NMD, who were wheelchair and NIV users since early childhood, appeared to accept NIV use as part of their identity. Present findings support that more research is warranted to explore identity formation and its relationship to autonomous decision making around health behaviors in adolescents with NMD who become wheelchair users later in their childhood.

The ability to integrate health information and value it as personally important could be perceived as an empowering element of learning to self-manage their health. Both the aforementioned non-NIV users felt empowered that the choice of NIV uptake was theirs, and accepted responsibility for any repercussions of their decision on their own health. Risk of instantaneous versus gradual decline may be important discussion between HCPs and adolescents considering uptake of health behaviors which only manifest benefits in the long term: If an adolescent perceives they need to physically experience the benefits before being convinced of the value of a healthcare behavior, how is one still able to convince the individual when insidious deterioration is difficult to feel? How close to non-reversible physical deterioration does one allow the young person to get before making another concerted effort to understand whether their non-adherence is either an empowered choice, a sign of acceptance of failure or subsequent learned helplessness and ultimate disengagement?

The process of acceptance that some things can be changed, and others cannot, reflects a strategy of resilience identified in youths' with developmental disability (Cartwright, Fraser, Edmunds, Wilkinson, & Jacobs, 2015; Silverman, Verrall, Alschuler, Smith, & Ehde, 2016). Self-

awareness, self-reflection, goal orientation and acceptance were described by participants in this study as important elements to boost their sense of control. These strengths are affirmed to enable engagement within theories of motivation (Martin, 2007), as are participants' reflections that their positive attitude and mastery of their health behaviors relied on adequate emotional, social physical support.

#### *4.3 Theme III: Trusted, supportive, nurturing relationships*

*Trusted, Supportive, Nurturing Relationships'* was a key theme that enabled and empowered participants. The value of supportive relationships to adolescent health outcomes (Cartwright et al., 2015; Molcho, Gabhainn, & Kelleher, 2007) and coping with NMD (Lamb & Peden, 2008) has been previously reported. An important finding was the value of trusted, personal relationships to youth's learning and uptake of health behaviors. Participants' preference for gaining credible information from their peers, rather than HCPs, has not been explored in literature exploring development of self-management in youth with NMD. A proposed socio-ecological model of adolescents' readiness for transition includes parents and HCPs, but omits recognition of the value of learning from peers (Schwartz et al., 2011).

Participants' valued longstanding relationships and familiarity with their HCPs as previously reported (van Staa, Jedeloo, & van der Stege, 2011), particularly when communication included an affective, emotional relationship (the 'feeling understood' relationship) beyond just knowledge sharing (Kauw et al., 2015). The importance of personal connectedness in the HCP-adolescent relationships is confirmed by Johnson, McMorris, et al. (2015), who showed that youth's feelings of relatedness to their HCP and their HCPs respectful, empathetic communication with youth with mobility limitations correlated with their sense of health-related autonomy.

#### 4.4 Theme IV: Expectations and shared knowledge

Expectations of adult independence and the sharing of credible knowledge through open communication appeared crucial elements to healthcare engagement. Present findings suggest that youth with NMD experience healthcare engagement similarly to adults, as a co-constructed process to develop a healthcare partnership (Bright et al., 2015).

In contrast to qualitative research with younger persons with NMD, when expectations for the future appeared less achievable (Gibson et al., 2009), participants in this study expressed motivation to prepare for and learn about their future. Participants displayed high levels of self-belief in their goal orientation to achieve diverse personal goals across education, occupation and participation outside the home. These goals appeared realistic, given participants' high academic achievements. Both Vuillerot et al. (2010) and Young et al. (2007) report higher scores in domains of school performance in youth who were wheelchair and NIV users than for youth who were more physically able. Youth with NMD who are intellectually able likely prioritize intellectual achievement when their disability precludes them from physical achievement in preparation for a gainful career in adulthood.

A few participants in this study were open to the difficult conversations about life expectancy, though whether their HCPs had initiated this recommended conversation (Bushby et al., 2010b; Hull et al., 2012) was less obvious. Late initiation of conversation around life expectancy can lead to cycles of anxiety and depression amongst all the family (Samson et al., 2009). Freeman et al. (2015)'s review of a transition resource concluded that instead of a 'special' conversation, adult goals should be a continuous dialogue throughout adolescence. It is known that HCPs may feel inadequately prepared to broach difficult conversations with youth, including conversation about life expectancy, sexuality and reproductive health (Peter, Forke, Ginsburg, & Schwarz, 2009), especially with youth who are wheelchair users (Seburg, McMorris, Garwick, & Scal, 2015).

Our data challenges HCPs to review their role in knowledge sharing with patients with rare disease (Budych, Helms, & Schultz, 2012). HCPs who view adolescents with NMD as experts in their own condition, elicit their questions and concerns, and who provide feedback appear more likely to optimize youth's health literacy and self-management skills.

### *Strengths and Limitations*

Recruitment of young people from across Australia and use of multiple technologies to facilitate interstate interviewing enabled a capturing of a maximum variety of points of view, confirmed through data reaching thematic saturation. This study's primary limitations are twofold. Firstly, the opt-in sampling strategy of the MyLifeMyVoice study likely limited the response to youth who felt confident and empowered to participate in research (Liamputtong, 2013), although their varied experiences provide sufficient depth to extrapolate personal barriers and lack of support experienced by those who are disengaged. Secondly, the inclusion of youth experiencing different NMD disease trajectories limits generalizability to individual diagnostic groups. The decision to group by impairment and functional impact of condition (muscle weakness and wheelchair use) is justified by the finding that youth with rarer conditions, such as undiagnosed CMD, valued learning from peers living with relatively more common DMD. Importantly, the inclusion of youth with different disease trajectories enabled deeper insight into factors around NIV uptake during adolescence that would not have been possible in a sample with less varied experience.

### *Conclusions:*

The perspective of youth with NMD involved in this qualitative study deepens understanding that healthcare engagement is a dynamic, multifactorial process enabled through shared knowledge, expectations, intrinsic motivation and adequate support. By taking time to be a sounding board for these open minded and self-reflective young people, with creative ideas about their health, HCPs

shared teaching and learning may avoid “errors” that take more time to fix and may enable safer “trials” if their behaviors or non-adherence pose a known risk to their health. The value of personal relationships with peers with similar health experiences to enhance knowledge acquisition and uptake of health behaviors remains underexplored. The experiences of youth with NMD in this study have implications for HCPs’ role as health educators that may extend to inform healthcare engagement with youth living with other complex conditions and uncertain lifespan.

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### **Declaration of Interest**

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## Appendix

## Interview Schedule

1. Tell me about your health. **What does health mean** to you?

(Prompt: What your health is good, what sorts of things are going on for you? When things are not so good, what is going on?)

2. What do you feel **might influence** your health?

(Prompt: What helps/motivates you to be as healthy as possible?)

3. Can you describe to me **what you do** to stay healthy/well?

(Prompt: What sorts of things do you do every day to stay healthy/well? Is there anything else you think you could or should be doing? What has been your experience of being given responsibility for your health?)

4. Tell me about the support YOU FEEL YOU **GET** to be as healthy as possible.

(Prompt: From your health care providers? From your friends and family?)

5. Tell me about the support YOU FEEL YOU **NEED** to be as healthy as possible.

(Prompt: From your health care providers? From your friends and family?)

6. Tell me how you **learn** about your health.

(Prompt: What do you want to learn about your health? How do you find information? How do you want to learn about your health? Who do you want to learn from? Why?)

7. What does health care engagement mean to you?

8. Is there anything else you would like to talk about?

PROMPTS: "How do you KNOW this?"

"Can you describe a specific experience?"

Figure 1

Emerging themes of engagement perceived by youth with NMD

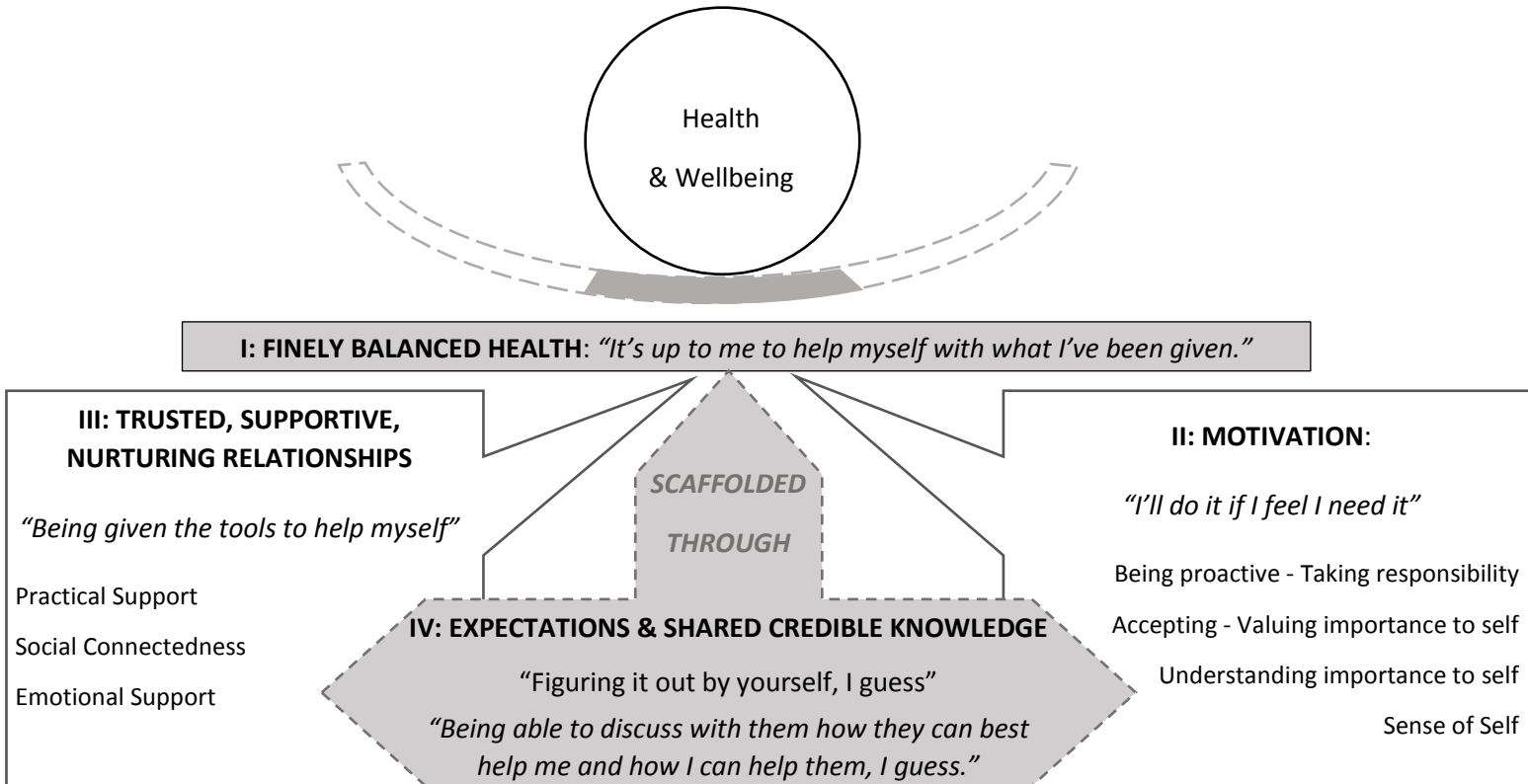


Table 1

Participant Characteristics (n=11) self-reported in the MyLifeMyVoice Questionnaire

Table 1 a. Demographics and Co-morbidities

DEMOGRAPHIC	n	PHYSICAL FUNCTION AND COMORBIDITIES	n
<b>Age and Health Service Context</b>		<b>Functional Ability</b>	
14-18yo (still in pediatric services)	6	<b>Transfer Ability -</b>	
18yo (transitioning to adult services)	2	Able to stand transfer with assistance & shift weight in sitting	4
21yo (transitioned to adult services)	3	Hoist for all transfers, able to shift weight in sitting	1
<b>Gender</b>		Hoist for all transfers, support required to maintain sitting	6
Male	7	<b>Upper Limb Function</b> – able to self-feed	6
Female	4	<b>Respiratory function supported by nocturnal NIV use**:</b>	
<b>Diagnosis</b>		Every night + additional occasional diurnal use	3
Duchenne Muscular Dystrophy	4	Every night	2
Congenital Muscular Dystrophy (non specific diagnosis)	3	Not every night	3
Ullrich Congenital Muscular Dystrophy	1	Trialed but not issued for home	2
Emery Dreyfuss Congenital Muscular Dystrophy	1	Not yet prescribed	1
Nemaline Rod Myopathy	2	<b>Respiratory Illnesses in past 12 months</b>	
<b>Academic achievement*</b>		None	7
Above average	7	1-3 managed at home	4
Average	3	<b>Cardiomyopathy</b>	2
<b>Location of Residence</b>		<b>Scoliosis</b>	
Metropolitan	8	No curve	1
Regional	3	Conservatively managed scoliosis	5
		Spinal fusion	5
		<b>Nutrition and bowel function</b>	
		Gastrostomy/Nasogastric tube feeding	1
		Assisted bowel voiding	1

\*Academic Achievement response choices – *Above average* “My marks in most subjects are higher than those of most other kids at my school.” *Average* “My marks in most subjects are about in the middle compared to other kids at my school.” Nil *below average* reported; \*\*NIV: Non-Invasive Ventilation, prescribed when sleep study shows extended periods of hypoxia and hypercapnia related to respiratory muscle weakness, usually prescribed nightly.

Table 1 b. Motivation and Engagement Scale High School (MES-HS) Grades\*\*\*

RATINGS OF MOTIVATION AND ENGAGEMENT	n			
	scoring within each MES-HS Global Ratings Grade			
	A	B	C	D
	"This a strength"	"Good, and can grow into a strength"	"This needs some work"	This needs relatively more work"
+ve Thoughts – Boosters	2	5	2	1
+ve Behaviours – Boosters	4	1	3	2
-ve Thoughts – Mufflers	6	2	1	1
-ve Behaviours - Guzzlers	6	2	2	0

\*\*\* (Martin et al 2007, [www.lifelonglearning.com](http://www.lifelonglearning.com))

Table 2

## Theme I: FINELY BALANCED HEALTH - categories and sample quotes

Subthemes	Categories
<b>Good health = maintained physical and mental wellbeing</b>	<ul style="list-style-type: none"> <li>• Physical and mental health are equally important</li> <li>• Normalcy – living life the way you want without too much intervention</li> <li>• Lack of condition specific knowledge and erroneous health interventions can be dangerous</li> <li>• Being sick really puts the brakes on things</li> <li>• Breath is everything</li> </ul>
<p><b>Example quotes:</b></p> <p><i>YP: Just feeling good, and active, like being active...and being happy, I suppose. Mental and physical health.</i></p> <p><i>YP: I think that was the main thing when I was younger that would annoy me. You just have to wait ages at appointments and you'd always have to go to appointments and you think I'd rather be doing something else.</i></p> <p><i>YP: Well if our health is not so good we have to stay home. But if it's good we can go about our daily business without too much intervention.</i></p> <p><i>YP: Just ehm..... Being able to do anything I could do really..... Not necessarily a specific thing. Just be well enough to be able to do whatever I want.</i></p> <p><i>YP: Sometimes doctors know what should be done, but it isn't always the best case for us. For example with our scoliosis when it happens to a normal person the doctors put them in a back brace, so that's what they did to us. However in our case that made things worse because we were no longer using our muscles to stay upright.</i></p>	
<b>Doing enough to maintain and prevent</b>	<ul style="list-style-type: none"> <li>• Preventing illness through taking precautions: Isolation vs participation</li> <li>• Getting on top of things early</li> <li>• Optimizing immunity &amp; minimizing fatigue: Good diet, exercise, sleep</li> <li>• Tight routines vs flexibility</li> </ul>
<p><b>Example quotes:</b></p> <p><i>YP: I try to minimise contact with any ehm.. You know sick people and stuff like that. You know if my mates have got a cold I'll be like don't come around here . I don't want to get sick. you know...It sounds silly but.... With my condition ehm.. Getting a cold ...it can last like three weeks. I mean so ehm.... I'd just rather not get it in the first place.</i></p> <p><i>YP: I guess getting treatment when you're unwell. Getting treatment when you have got an issue. I mean leaving an issue unsolved is not gonna help anything.</i></p> <p><i>YP: Doing regular physio. Make sure we get enough sleep. Not too stressed.</i></p>	



Table 3

## Theme II: MOTIVATION - categories and sample quotes

Subthemes	Categories
<p><b>Acquiring knowledge</b></p> <p><i>Example quotes:</i>  YP: ...often its (learning) via my parents cos they both like..doctors and then ..also...when I go to the camps I'll catch up with people with muscular dystrophy and I can gain a bit of knowledge about other muscular dystrophies from watching them...every now and then they might talk about it.</p> <p>YP: I guess...ehm.. Because it is so new to me I haven't really ehm....like I feel like it was yesterday that I was diagnosed so.... I haven't really been in a rush to find out.....</p>	<ul style="list-style-type: none"> <li>• Experience-based knowledge</li> <li>• Knowing what the future holds</li> </ul>
<p><b>Sense of self</b></p> <p><i>Example quotes:</i>  YP: With me I... Stretching I know I need it when I can just feel that my legs are a bit achy... They just feel uncomfortable in the muscles. So that's how I know that I should stretch. Or...yeah...so its just getting to know your body I guess.</p> <p>YP: Well. I guess...I guess they (NIV) are more invasive. That's the way I see 'em. Leg splints are just orthotics. They just sitting on your legs. They're not like right your face.</p> <p>YP: Like, my respiratory function was very, VERY bad. And also I saw other guys like, wearing respirators and stuff during the day, so then that (using the BiPAP* every night) sort of clicked.</p>	<ul style="list-style-type: none"> <li>• Body awareness</li> <li>• Body image</li> <li>• Comparing self to others with similar conditions</li> </ul>
<p><b>Understanding importance to self</b></p> <p><i>Example quotes:</i>  YP: It's the way activities are done...Well I think when people are more familiar with the device..well.... It's normalised a lot more. That the way I see it anyway. If you're not familiar with something.... It tends to be human nature to question and be concerned about it.</p> <p>I: And who do you want to learn from?  YP: ehm... I have NO idea. Maybe just anyone that I.. Trust....if I've known them for ages. Like, mmm. like some of my doctors I've known them since I was very little. So.. I trust them more than somebody I just meet.</p>	<ul style="list-style-type: none"> <li>• Questioning is normal for teenagers</li> <li>• Taking credible advice from someone you trust</li> <li>• Believing that it is worth it - Feeling the benefits</li> </ul>
<p><b>Accepting and Valuing importance to self - bittersweet responsibility</b></p>	<ul style="list-style-type: none"> <li>• A sense of responsibility for self</li> <li>• Getting on with things vs hope to make things easier</li> <li>• Maturity – that's just the way it is</li> </ul>

**Example quotes:**

YP: *The doctor who prescribed me the BiPAP ehm....she told me that ehm... like I wouldn't become reliant on it but I said well...can I have a little bit more proof ...like that because I've seen it happen (with my sibling) and she can't really assure me that it wouldn't happen so ehm.. You know..eh... I don't want to take the risk because...ehm...you know later in life I may regret it... at least I couldn't blame it on the BiPAP if something went wrong or something...I would rather, you know, blame myself than blame a piece of technology.*

YP: *When you're at school and you know... You don't really want another thing for people to point out that's different about you when you're already rolling around in a chair you know. But you know, back then I didn't have... Well like I didn't think I had much control over you know... Equipment and that kind of thing... So ehm... Yeah these days, ehm...being an adult, I don't worry too much more about it I suppose.*

**Being goal directed and proactive**

- Planning for the future - Having a diagnosis helps
- Having a positive outlook
- Individual priorities and sense of purpose
- Feeling empowered

**Example quotes:**

YP: *Ehh...well...I've got like good friendships so I want to stay in touch with them...ehm...when I grow up I want to be a physicist so...I want to be able to study a lot so....to do that I have to be fit enough to be able to write fast enough ..eh...I also want to be strong because ehm..if I am weak I would have even less independence than what I do now.*

YP: *ehm...yeah.... So that's the main responsibilities I've had for my health. Ehm... And also, I guess, knowing when to ask for help is a responsibility. So that I'm never in situations I shouldn't be in.*

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\*BiPAP: Bi-level Positive Airway Pressure, a form of non-invasive ventilation (NIV)

Table 4

## Theme III: TRUSTED, SUPPORTIVE, NURTURING RELATIONSHIPS - categories and sample quotes

Subthemes	Categories
<b>Emotional &amp; Social Support</b>	<ul style="list-style-type: none"> <li>• When support is lacking, everything is harder</li> <li>• Parents' loving encouragement</li> <li>• Seen as an individual – non-judgemental relationships (disability stigma)</li> <li>• Established relationships with HCPs</li> <li>• Open communication</li> <li>• Humour</li> </ul>
<b>Example quotes:</b>	<p data-bbox="193 725 1398 936"><i>YP: You kinda feel like you can't do too much. So I think ehm... Just having them there to help and being encouraging and never kind of.. Yeah always be willing to do something if I want in terms of being healthy. Ehm... Yeah being willing to accept what I need. ... And so I guess... The more help you get the less hassle things are for you. The less down you feel about your health and.... You feel more positive about how you are going. So yeah... if you are not... If you feel a lot of things are quite hard for you... You can feel quite ehm... dispirited.</i></p> <p data-bbox="193 972 1398 1115"><i>YP: I guess...ehm.... Feeling like that they relate to you and understand ways that you think would help and ehm..... Feeling like ehm... They're actually properly thinking about you and not just going on what's standard procedure. Ehm.. Or what other people have told them. At least be influenced by... Us.</i></p> <p data-bbox="193 1128 1398 1234"><i>YP: (Doctors appointments) can get annoying sometimes. Especially if those people think, because we're in a wheelchair, that we have an intellectual problem, too. At least the doctors that know us talk to us.</i></p> <p data-bbox="193 1247 1398 1496"><i>YP: Luckily my schoolmates were...eh.... You know..eh... Very open. And they weren't going to judge. My condition is not anything eh... You know, new to them, so... If we had something organised or something and I say...ahh.... I can say I'm really not feeling good today, I don't have the strength or something, you know... Which I feel is really important because.... You know if you can't be honest with your friends and your family about that kind of thing then.... You know... Who can you be honest with? But ehm.... I learnt that very early on. Ehm... Yeah... But... It's like, you know, ... So, yeah. Friends and family are sort of go to support, you know what I mean.</i></p>
<b>Practical support:</b>	<ul style="list-style-type: none"> <li>• Access to information</li> <li>• Equipment</li> <li>• Easy access to services</li> <li>• Coordinated care</li> <li>• Physical help from many sources</li> </ul>
<b>Example quotes:</b>	<p data-bbox="193 1742 1398 1877"><i>YP: Because I can just... Sa if it's a health care provider I can just call them up and get an appointment. Especially my cardiologist. (...) We can go straight to him. Whereas ..yeah.... You don't have to.... He doesn't query anything. We can just call him up. I guess same with.... Other people as well.</i></p> <p data-bbox="193 1912 1398 2024"><i>YP: Mum's family has been really great. They've helped out a lot, ehm...like... My auntie will come up and look after me if mum and dad were on a break or...ehm.... Help financially with some things or.... Just like little things like that.</i></p>

Table 5

## Theme IV: EXPECTATIONS AND SHARED KNOWLEDGE - categories and sample quotes

Subthemes	Categories
<b>Expectations of responsibility</b>	<ul style="list-style-type: none"> <li>• Shared problem solving</li> <li>• Normalizing the tough conversations</li> <li>• Supported autonomy</li> </ul>
<b>Example quotes:</b>	
<p>YP: Like the doctors say this is..... Now that I'm older..... They sort of don't say to mum.... They sort of treat me..... And I need to say when I need something else or something. I think it's more, like, you're more independent instead of being told... Its sort of... Figuring it out by yourself I guess.</p> <p>I*:. How do you know this? Or what are you expecting?</p> <p>YP: ehm... Well... Because at (the Children's Hospital) I'm sort of still... I'm sort of treated like a child but not really... Yeah... So I think if I moved to (the adult hospitals) I'll sort of be... There will be more on me to... Like not really mum or dad or....</p> <p>YP: I think a lot of people wait for the impact. ... Sometimes you have to give the cold hard facts... but with this stuff it tends to be more touchy. I think the reason people are more comfortable talking about....say death...from motor vehicle accidents... And drugs is because it is instantaneous. It's an instant kind of death while with some of these other factors it's more of a gradual.... It's more of a gradual thing. So because it's not something that instantaneously hits, I guess it's not always seen to have to be... Talked about...urgently. ...Yeah I think it gets stigmatized. So people need to take stigma out of the topic... and I think if you go to...when I've been involved in sport I've seen the passing away of older athletes. And it's just...yeah...you kind of...it becomes a bit less traumatic. A bit more normalized. If that's a way to look at it.</p>	
<b>The need for shared health education</b>	<ul style="list-style-type: none"> <li>• Knowledge is limited – avoid error</li> <li>• Shared problem solving – safe trial</li> <li>• Credible experience based knowledge through trial and error</li> </ul>
<b>Example quotes:</b>	
<p>YP: It's (Facebook Group) kinda like first hand experience. Whereas doctors are kind of just reading about it on the Internet.</p> <p>YP:... 'cos I guess that can be more reassuring to know that they've been through it. And they'll be able to relate to you more than anyone else can.</p> <p>YP: The most I've learned is just from experience. As I've grown and stuff ehm.... And like the few doctors we've been to. Ehm.. Usually we tell the doctors more than they know, just 'cos we've experienced the condition for ourselves and because they don't know what it is. So ehm...yeah ehm... 'cos if there's one thing ehm...that there's too many of, is people talking about stuff they don't know about.</p> <p>YP: It kind of means two things to me....ehm... Like me engaging myself in my health and all that and taking care of myself with the ways I've been ehm... told or.... Ways that have been recommended to me....and all that....ehm.... And also about... Healthcare professionals, like physio and all these people helping me, to be as healthy as I can, so they're like the two things that stand out to me the most...they kinda tie in together.... So it's like them helping me... And then me kind of helping myself with what they've helped me with...kind of thing....yeah..its like they give me the tools to help myself...and ehm.... its then up to me to help myself with what I've been given.</p>	

I\* - Interviewer