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RESEARCH PAPER



# “Me and ‘that’ machine”: the lived experiences of people with neuromuscular disorders using non-invasive ventilation

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## ABSTRACT

**Purpose:** Neuromuscular disorders (NMD) encompasses a wide range of conditions, with respiratory weakness a common feature. Respiratory care can involve non-invasive ventilation (NIV) resulting in fewer hospital admissions, a lower mortality rate and improved quality of life. The aim of this study was to explore the ‘lived experience’ of NIV by people with NMD.

**Methods:** Interpretive Phenomenological Analysis (IPA) with semi-structured, face to face interviews with 11 people with NMD, using bi-level positive airway pressure for NIV for more than 12 months.

**Results:** Three themes were interpreted: (i) Alive, with a life; (ii) Me and ‘that’ machine; and (iii) Precariousness of this life. NIV enabled hope, independence and the opportunity to explore previously perceived unattainable life experiences. Yet, participants felt dependent on the machine. Furthermore, practical considerations and fear of NIV failure created a sense of precariousness to life and a reframing of personal identity.

**Conclusion:** The findings highlight the broad ranging positive and negative effects that may occur for people with NMD when using this important therapy. Ongoing non-judgemental support and empathy are required from health professionals as the use of NIV challenged concepts such as ‘living life well’ for people with NMD.

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Lived experience; neuromuscular disorders; non-invasive ventilation; qualitative; interpretive phenomenological analysis; identity

## ► IMPLICATIONS FOR REHABILITATION

- Neuromuscular disorders may result in respiratory weakness requiring non-invasive ventilation (NIV).
- When prescribed early, NIV can result in fewer hospital admissions, a lower mortality rate and improved quality of life.
- The relationship of people with NMD with their NIV machine is complex and impacts on and requires adjustment to their identity.
- NIV users acknowledged that NIV provided hope but simultaneously recognised the precariousness of NIV on their life.
- In order to better support people with NMD healthcare professionals need to better understand how the physical, psychological and social implications of NIV affect an individual's life.

## Introduction

Neuromuscular disorders (NMD) encompasses a range of conditions which vary in aetiology and include both acquired and inherited. The global prevalence of NMD is estimated to be 1% of the world's population [1]. Associated thoracic muscle weakness and chronic respiratory failure often occur with NMD [2]. Respiratory failure symptoms typically develop during sleep when the drive to breathe is lower and the upper airway is more collapsible [3]. This results in nocturnal hypoventilation. However, with disease progression, respiratory failure may also occur during the daytime. Eventually daytime hypercapnia, morning headache, orthopnoea, and dyspnoea ensues [3]. Respiratory failure is the major cause of mortality and reduced quality of life (QoL) in people with NMD who have respiratory complications [4], accounting

for 80% of mortality in people with specific neuromuscular disorders [5].

Home non-invasive ventilation (NIV), typically bi-level positive pressure ventilation (Bi-PAP) is a critical area of respiratory management for chronic respiratory failure, that can be delivered during sleep (*via* nasal or oronasal interface) and in the daytime (*via* mouthpiece or nasal prongs), for people with some specific types of NMD [6,7]. As clinicians and health systems have gained experience with NIV [8], home NIV rates have increased. For example, an increase from 6.6 per 100,000 people to 12.9 per 100,000 between 2005 and 2015 in Europe [9] and Canada [10,11]. In Australia and New Zealand in 2013 the prevalence of NIV prescription was 9.8 and 12.0 per 100,000, respectively [12]. Of these NIV prescriptions 33% and 20% were for people with NMD in

Australia and New Zealand, respectively [12] and the prevalence continues to increase [13].

NIV, when introduced at the onset of symptoms of nocturnal hypoventilation [2,3], in conjunction with other factors such as therapeutic pharmacology, surgery, nutrition, and management of respiratory infections [14], has resulted in improved life expectancy for people with the neuromuscular disorders of Duchenne muscular dystrophy, motor neurone disease, and spinal muscular atrophy—type 1 [15–20]. Specifically, Eagle et al. [16] reported an 8 year increase in life expectancy when comparing a multidisciplinary approach and NIV compared to multidisciplinary care without NIV in people with Duchenne muscular dystrophy.

A growing body of ‘improved quality of life’ evidence indicates that the role of NIV is not only for palliative care, but also for symptom management in people with NMD who experience respiratory distress [10,21]. For example, nocturnal NIV treatment alleviates symptoms of respiratory insufficiency which include: daytime hypersomnolence, morning headache, and daytime hypercapnia [22]. Furthermore, early initiation of NIV reduces or delays the need for tracheostomy mechanical ventilation and its associated complications of institutionalisation, respiratory tract infection, fistulisation, and reduced speaking and swallowing capacity [23,24]. Consequently, NIV is currently recommended in the early stages of respiratory decline in people with NMD of Duchenne muscular dystrophy and Spinal muscular atrophy [2].

Success with managing NIV depends on several factors including appropriate clinical case selection, funding, training of staff, the patient experience, and patient and caregiver education and support to manage application of NIV, and equipment cleaning and maintenance [8]. A review by Ngandu et al. [25], which included 32 published qualitative studies, synthesised evidence on experiences of NIV use by people with respiratory failure. Fear of technology, death, and pain; insufficient information exchange; decision making conundrums; and healthcare professional relationships were identified as the predominant issues that users experienced. However, only 11 of these studies included people with neuromuscular disorders, six of which focused specially on people with Amyotrophic lateral sclerosis, otherwise known as Motor Neuron Disease (MND). The remaining five papers were all conducted in Scandinavia and included a total of 33 people with other NMD (i.e., excluding MND). In addition, four qualitative studies, from Denmark, Japan, Canada and the U.S.A., not included in the review mentioned above, explored experiences of NIV and were published between 2004 and 2019. These four studies included a total of 36 people with NMD (excluding MND) on NIV, or whom had progressed from non-invasive to invasive ventilation, and discussed the anxiety provoked by transition to non-invasive or invasive ventilation, effects of ventilation on communication, ethics of ventilation, and quality of life [26–29]. This distinction of MND from other NMD disorders is relevant, as the age of onset, life course i.e., disease progression is typically different for MND [30], therefore the lived experience is likely to be different.

Other researchers have focussed on the perspective of family caregivers and relatives with children and adults who have NMD (including MND) on home ventilation [31–35]. They reported experiences of: responsibility, hope, sleep deprivation and care burden [31–35].

In New Zealand, one survey explored perceived benefits of NIV in people with a mix of conditions including obesity hypoventilation, sleep apnoea and NMD and found improved quality of life [36]. In addition, one qualitative study conducted in New Zealand

study found themes of confusion of service delivery, a need for greater information, and fear and apprehension with continuous positive airway pressure (CPAP) but its focus was people with obstructive sleep apnoea rather than those with a NMD [37].

It is important that healthcare professionals learn from the experiences of the people they work with so to improve patient care [38]. This is especially true for people with NMD who can require critical medical support and then subsequent rehabilitation as they recover from acute respiratory distress. A total of nine papers, exploring the lived experiences of 69 people with NMD (excluding MND) appear to have been published in the last 18 years. Furthermore, these studies span a time period over which non-invasive technology has changed and therefore clinical practice, service delivery and patient experiences might also have changed. There is therefore a paucity of past and recent qualitative research exploring the lived experience of NIV by people with NMD (excluding MND) internationally and in New Zealand.

## Methods

The aim of this study is to explore how people with NMD make sense of their experience of using NIV, that is their ‘lived experience.’

### Study design

Interpretive Phenomenological Analysis (IPA) underpins this study [39]. This methodology is flexible and holistic and offers the opportunity to gain insight into the personal lived experience of a phenomenon, the meaning of that experience to participants, and how participants make sense of that experience [40]. IPA is recommended when exploring experiences of significant health-related experiences, due to the subjective and contextual nature of these experiences and is underpinned by hermeneutic phenomenology with an inductive and idiographic analysis [41]. A key component of IPA is the acknowledgement of participants’ social and cultural context, thus ensuring its effective use with Māori and other indigenous populations [42]. Reporting in this manuscript has followed the Consolidated Criteria for Reporting Qualitative Research (COREQ) guide [43]. The study was approved by the University of Otago Human Ethics Committee (Health) (Ref # H18/005).

### Participants and recruitment

Eleven people over aged 16 years and over with NMD who had been using bi-level positive airway pressure (Bi-PAP) home NIV for at least 12 months were included. People with less than 12 months of NIV experience were not recruited as Ngandu et al. [25], had previously reported that prescription of NIV can be unexpected and stressful. We decided to interview participants who would have had some time to adjust to the therapy and living with NIV rather than the transition to NIV per se. Continuous positive airway pressure (CPAP) is not clinically considered NIV and therefore users of CPAP were not included. People with motor neurone disease, myasthenia gravis and multiple sclerosis were excluded, due to the different causes and disease progression of these disorders from other NMDs [30] and also the larger body of already existing literature about experience of NIV in these groups [25].

Participants were recruited *via* advertisement across disability non-governmental organisations in New Zealand (such as, the Muscular Dystrophy Association of New Zealand and the

Neurological Foundation). Participants were also recruited from the 16 centres providing NIV services across New Zealand (including public and private care). Advertising material such as flyers were placed either online or in the premises of these organisations.

While IPA methodology looks to ensure participants are homogenous around a phenomenon [39], purposeful sampling was used for the identification of information rich cases within this possible cohort of people [45] and to ensure Māori were specifically included in this research as per the principles of the Treaty of Waitangi, which ideally underpin all New Zealand research [45]. In addition, as most of the New Zealand population predominantly lives on two islands, we wished to ensure participant from both islands were recruited. Informed consent was gained from all participants prior to data collection commencing.

### Data collection

Data were collected via semi-structured interviews either in the homes of participants ( $n = 10$ ) or over a secure audio-conference system ( $n = 1$ ). Semi-structured interviews are an effective data-collection method for gaining in-depth information because it is obtained in an entirely open and flexible way. Semi-structured interviews facilitate participants to express their experiences in their own words [46]. Table 1 includes key domains and sample questions that guided the interviews. The interview schedule was iteratively created by four of the authors (TI, MJ, BJ, and MP) from background reading and consultation with the Muscular Dystrophy Association of New Zealand. These four researchers have health backgrounds in medicine, psychology, nursing and physiotherapy. Three have lived experience of disability, including one with NMD. Combined they have over 40 years of research experience.

All participants were given the opportunity to have friends/family or other support people present at the interviews, with five participants having their caregivers (formal paid caregivers and family members) present in the interview. The caregivers

contributed to the discussion through aiding recall and providing clarification if participants had difficulty communicating a point. Participants were offered reasonable accommodations throughout their participation in the study, including the offer to take breaks from the interview if they became fatigued. Interviews were also able to occur over multiple days if fatigue was a factor, although no participants took up this option. The interviewer (MJ) was an academic with experience in undertaking qualitative interviews. All interviews were recorded with an Olympus digital voice recorder and transcribed anonymously verbatim by an independent transcriber.

### Analysis

Transcripts were analysed using IPA methodology which is a highly iterative process [39]. Four researchers (MJ, MP, ER and KS) immersed themselves in the data by continuously re-reading the transcripts. Following this, each transcript was coded independently with initial thoughts and ideas recorded in the left margin of the page and quotes highlighted, and potential concepts in the right margin of the page. Once all transcripts were coded, the underlying concepts were categorised into provisional themes on a Microsoft Excel sheet. Over several weeks the provisional themes were considered and reflected upon by re-reading of the transcripts, further listening to the audio-files, and use of reflective interview memos until the initial provisional themes had reduced to five themes and these were sequentially ordered to tell a narrative story.

In addition to this, TI, BJ and HS independently read and coded a subset of the transcripts. Then provisional themes, with linked codes and quotes, were discussed with the whole research team until a consensus was reached on initial themes. The relationship between the themes was discussed and four key themes were considered, with two provisional themes being condensed into one [40,47].

Over the subsequent weeks the codes, with exemplary quotes from the initial excel sheet, were transferred into a new Excel

Table 1. Semi-structured interview key domains and sample questions.

Key domain	Sample Questions
Diagnosis	When? How old? Progression of the disease
What led to the need for NIV	Who decided there was a need? Symptoms Early prescription Change in prescription over time Current NIV and usage Experience of adjusting to NIV Professional or social support
Quality of life and function	Comparison between now and then? <ul style="list-style-type: none"> <li>• Mobility</li> <li>• Breathlessness</li> <li>• Pain</li> <li>• Independence</li> <li>• Sleep Quality</li> </ul>
Obtaining NIV	Access Wait Education Health and safety of the NIV <ul style="list-style-type: none"> <li>• Failure</li> <li>• Power</li> <li>• Natural disaster</li> <li>• Maintenance</li> </ul>
Role of the caregiver in supporting NIV uptake and use	Role of the caregiver in helping you decide to trial breathing support Role of the caregiver in the technical management of the breathing apparatus Any other support?

sheet, the transcripts were re-read again, and audio interview related to specific theme concepts listened to and the relationship between themes further considered. The team met a further two times to review the themes, refining the definition of each theme, and using memos and thoughts from the interviews and diaries to reflect upon our interpretation of the data. During this time, the four themes reduced to three. These themes reflected similarities and differences in experiences across the participants, along with the researchers' own interpretations of participants' experiences [47]. The researchers' interpretation, undoubtedly influenced by the researchers' own life experiences and making of knowledge, alongside the interpretation of the participants, is acknowledged in IPA. This is termed a double hermeneutic [41,47].

In addition, participants were sent a summary description of the three themes. While this is not usual IPA methodology, we considered this aspect important to understand if the research team had described the phenomenon in a way recognisable to the participants [41]. All included participants were happy with the overall summary, proposed theme names and content within the three themes. One participant specifically requested that the negative effect of NIV on intimate relationships be included to highlight to health professionals that people with NMD have the same needs as other people [48]. No other specific clarifications were requested.

Participants are de-identified however pseudonyms have been used following the participant feedback process. Participants requested the use of pseudonyms rather than impersonal identifiers such as 'Participant 1' to emphasize that the experiences and feelings presented occurred to 'real' people.

## Results

The study sample comprised of 11 participants with NMD (see Table 2. for participants' demographic information). One participant had been on NIV for 2 years, four for between 5 and 7 years, three for 12 to 13 years and three for 20 to 27 years. These six latter participants had all been using NIV for close to half of their life; three of these participants were in their twenties and three over the age of 50 years. In addition, one participant started with invasive ventilation and then weaned back to NIV. Interviews were between 40 and 80 min in length, with an average interview time of 60 min. Upon the request of participants, three caregivers were present for and contributed to the participant interviews by providing clarification when participants' verbal communication was difficult to hear clearly.

Three themes were interpreted: (i) Alive, with a life; (ii) Me and 'that' machine; and (iii) Precariousness of this life. A symbiotic relationship was formed with the NIV machine; one, which enabled new opportunities but one that also created dependence and thus necessitated an adjustment of identity. Exemplary quotes from participants for each theme are presented with in text or in Table 3. These themes are discussed in depth below.

### Alive, with a life

Participants acknowledged that NIV had not only forestalled death, that is they were alive, but had allowed life to be lived. The appreciation for being alive, albeit an adjusted life following NIV prescription, was expressed by all participants. There was a perception that NIV kept them alive. This was especially so for four of the study participants, when NIV was initially prescribed at a time when critical care was required for respiratory failure. NIV

Table 2. Summary of participant demographic information.

Demographic variable	N =
Gender	
Female	6
Male	5
Age range (20–64)	
20–29	4
30–39	2
40–49	1
50–59	2
60–69	2
Ethnicity*	
New Zealand European	9
Māori	2
Chinese	1
Neuromuscular disorder	
Muscular dystrophies (Congenital myopathy, Duchenne muscular dystrophy, Emery-Dreifuss muscular dystrophy, Limb-Girdle muscular dystrophies, Myotonic)	8
Metabolic disorders (Pompe, Mucopolysaccharoidosis MPS IVA)	3
Duration of NIV use	
2–27 years	
Hours of NIV per day	
Nocturnal only	3
Nocturnal and intermittent day use	7
Dependent (> 15 h per day)	1
Current stable relationship	
Yes	4
No	7
Residence	
Home	11
Residential care	0
DHB location	
North Island	7
South Island	4

DHB: District Health Board (Geographical areas of health service delivery in New Zealand); NIV: Non-invasive ventilation.

\*Participants could identify with more than one ethnicity.

was also perceived to keep them alive since, "I probably wouldn't be alive at 28 with the muscle weakness, if I didn't have that machine" (Daniel; <40 years old + >15 years of NIV). These participants particularly appreciated the lifesaving effects of NIV, even if they had not had the opportunity for informed consent at the time.

At first, participants found that the machine with its noise, the tubing and the pressure from the masks or nasal prongs was awkward. For others, the machine was frightening as they felt claustrophobic. However, the joy of being alive outweighed these experiences, "You've gotta relax and trust it, let it take over... I think I might have just fallen asleep and it just got easier... and I woke up. Alive! (laughs)" (Ava; ≥40 years old + >15 years of NIV). Persevering with the machine was also made easier when positive effects on activities of daily living (ADL), such as sleep, decreased fatigue and participation in activities meaningful to each participant were noticed, "Definitely helped my quality of life, I mean I can't sleep, [and] I can't really sleep without it" (Daniel; <40 years old + >15 years of NIV).

Participants recognised that NIV had not only extended their lives beyond expected years but had allowed participants to regain aspects of life thought lost and provided space for reflection about what was important to them. The extended time imbued hope and was frequently seen as an opportunity to fulfil dreams that had been hitherto perceived as unattainable. International travel became viable, "I have been lucky enough to travel the world and live to tell the stories." (Hayley; <40 years old + ≤15 years of NIV), and tertiary study, jobs and careers became reality.

Table 3. Summary of superordinate themes and definition, subordinate themes and representative quotes from participants.

Superordinate themes	Subordinate themes	Quotes
Alive, with a life: the recognition that NIV has enabled life to continue and the opportunity to re-evaluate plans and to have hope	Alive	<p>"To be fair without it, I probably wouldn't be alive!" (Jacob; <math>\geq 40</math> years old + <math>&gt; 15</math> years of NIV)</p> <p>"I can't live without it" (Jessica; <math>\geq 40</math> years old + <math>&gt; 15</math> years of NIV)</p> <p>"Definitely helped my quality of life ... I can't really sleep without it" (Daniel; <math>&lt; 40</math> years old + <math>&gt; 15</math> years of NIV)</p> <p>"It [BiPAP] is great, you know, it makes everything better" (Hayley; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"I find it really hard enough to breathe when there is not [a machine]" (Leah; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p>
	Fear, anxiety, apprehension	<p>"I woke up with this horrible mask on my face, and it was awful, and I thought, oh this is going to be terrible, but it wasn't ... just sort of claustrophobic I think, but it turned out not to be like that at all" (Claire; <math>\geq 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"... just physically putting the mask on Chris and I was so worried that it was going to leak and that sort of thing." Chris' mother</p> <p>"You've gotta relax and trust it, let it take over ... I think I might have just fallen asleep and it just got easier ... and I woke up alive (laughs)" (Ava; <math>\geq 40</math> years old + <math>&gt; 15</math> years of NIV)</p>
	Opportunities	<p>"I am quite happy with it, it keeps me alive and I can still do what I like to do" (Chris; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"And the more people said, ah stay (here, you can't travel. The more determined I was to go). I went. So, I have been lucky enough to travel the world, ah, and live to tell the stories." (Jessica; <math>\geq 40</math> years old + <math>&gt; 15</math> years of NIV)</p>
Me and 'that' Machine: the relationship and adjusted identity NIV users develop with the machine, along with their family and carers.	Adjusted identity	<p>"I kind of loved that thing, machine, I hate sleeping without it" (Hayley; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"It's my buddy, comes with me everywhere" (Hayley; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"It's also grieving and change of identity, giving up stuff but then finding a way to doing other stuff, still creating a life through process of adjustment and giving up some things, trying to rebuild and create something out of it." (Ava; <math>\geq 40</math> years old + <math>&gt; 15</math> years of NIV)</p> <p>I guess my lifestyle's probably reduced in capacity so I'm not putting the same strain on it [the lungs]." (Jacob; <math>\geq 40</math> years old + <math>&gt; 15</math> years of NIV)</p>
	Autonomy	<p>"I do feel I am a lot more informed and they (healthcare professionals) give me all the options and stuff [now that I'm older]" (Hayley; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"I was certainly left with the impression from his death speech, that invasive ventilation would never be offered and that concerned me, because it seemed to me that the day might come (for me) when that would be a more sensible thing (than what I am currently using) ... I think I should be aware of whether I have any other options" (Sophia; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p>
Precariousness of this life: the fragility of this new life and the associated emotional impact of NIV use.	Concerns with equipment failure	<p>"if I am lying down in my bed, and the power goes out and nobody comes to help me, I am going to die." (Sophia; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"If the power does go off and my breathing is impaired, and the mouth [mask] is on it is quite dangerous" (Daniel; <math>&lt; 40</math> years old + <math>&gt; 15</math> years of NIV)</p> <p>"I raised the fact that I was so dependent on it [with hospital staff] and what will I do if there's a power cut and the reply was 'You'll have to ring someone.'" So I sort of thought hmm, I would've thought there was a model that had a battery back-up but they didn't seem to be too terribly concerned, even though I was." (Noah; <math>\geq 40</math> years old + <math>\leq 15</math> years of NIV)</p> <p>"I guess the other issue with care is I believe that being on BiPAP would be a serious complication if mum were to become unavailable ... Because I have heard of young people often ... without other serious issues being in rest homes because somebody has decided that a BiPAP or a CPAP machine requires nursing oversight ... And so, I worry about that." (Sophia; <math>&lt; 40</math> years old + <math>\leq 15</math> years of NIV)</p>
	Trust	<p>"We have had that machine serviced and they stuffed up ... the settings ... It just wasn't, well it just wasn't working at all ... and I thought this doesn't feel right, this isn't good, so I didn't wear it, can't breathe through it ..." (Claire; <math>\geq 40</math> years old + <math>\leq 15</math> years of NIV)</p>

NB: All names used are pseudonyms used to protect anonymity.

I am quite happy with it. It keeps me alive and I can still do what I like to do ... still able to use my computer and still able to see [people] and go out. I think I have really got used to it. (Chris;  $< 40$  years old +  $\leq 15$  years of NIV)

### 'Me and 'that' machine'

Balanced against opportunity was recognition of the caveats created by NIV dependence. While NIV provided freedom and independence it also prevented spontaneity as, regardless of the level

of dependence on NIV, participants reflected upon restrictions created by daily use. These restrictions were not aspects discussed with the participants by their healthcare professionals prior to prescription of NIV but did seriously curtail many aspects of life. Travel, for example, had its difficulties, as participants considered where they might be able to plug in their machine, or whom to call if something went wrong. Any unplanned activities were therefore not practical, and this had social and personal implications. For example, the logistical planning required before sex heightened self-consciousness and that meant relationships were

unlikely to be casual or spontaneous; a factor that changed the nature of intimate relationships.

A partner, that's a more bit of an adjustment ... when you're blowing air on them when, you know [having sex] and I know for some of my friends, they struggle with the fact that it is a bit Darth Vadery ... not very romantic. (Ava;  $\geq 40$  years old +  $>15$  years of NIV)

Participants also reported many physical side-effects, were frequently frustrated by pressure sores from ill-fitting masks, shifting of teeth, bloating, gastrointestinal upset and an inability to communicate if they wore a mask. For example, Sophia found the mask particularly painful as decreasing muscle tone made it difficult for the mask to fit securely leading to pressure sores, "I was never told about the risk of pressure sores [from the mask]... And the information about pressure sores should be given." (Sophia;  $<40$  years old +  $\leq 15$  years of NIV) and Noah found increased water in the lungs distressing, "Moisture that's in the humidifier is going into my lungs... [I] can have some mornings where I'll wake up and I'm just full of, like coughing up water for the whole day" (Noah;  $\geq 40$  years old +  $\leq 15$  years of NIV). Thus, there was a discordance between the participants' gratitude for being alive, thanks to NIV, and the many unanticipated consequences of a life lived with long term NIV use.

They never told me about the teeth! The risk of a mask that is meant to sit between the knob of your chin and the bottom of your lip, knocking your back teeth out of place ... I mean really painful for the dentist to try and get behind it ... So, I had to have that out under general anaesthetic. Now I have a mouth guard, like a thin little boxers mouth guard. (Sophia;  $<40$  years old +  $\leq 15$  years of NIV).

Participants were still adjusting to the constant presence of NIV as a new facet of their identity while grieving the loss of their old 'self.' The adjusted 'self' was intrinsically linked to their NIV machine; an obligate symbiotic relationship where participants acknowledged grudgingly that they were inseparable due to their machine dependence. NIV provided the breath of life therefore maintenance and care of the NIV machine was paramount. This was recognised by Chris ( $<40$  years old +  $\leq 15$  years of NIV), "I knew that I had to have it [the machine], or I wouldn't make it, I'd die." Yet at the same time, NIV was a constant reminder of neuromuscular disorder progression and likely endpoint. This made coming to terms with this new aspect of their identity challenging and it negatively affected some of the participants' self-image, self-esteem, self-efficacy and confidence in their ability to participate in life.

It's also grieving and change of identity, giving up stuff but then finding a way to doing other stuff. Still creating a life through process of adjustment ... and giving up some things, trying to rebuild and create something out of it ... It takes away some of the spontaneity on life. It's a continual adjustment process physically, emotionally, psychologically, socially ... you need support. (Ava;  $\geq 40$  years old +  $>15$  years of NIV).

Jessica also described this emotional journey of adjustment as something bigger than just her, as it affected her whole family (whānau) and caregivers. Everyone needed to plan more or make practical choices regarding participation. Therefore, NIV influenced how family and caregivers led their own lives. Additionally, the physiological changes the machine represented were psychologically distressing, "The emotional journey it hasn't just affected me, it has affected my whole whānau" (Jessica;  $\geq 40$  years old +  $>15$  years of NIV).

Conversely, other participants highlighted the importance of now being able to move forward because they were using NIV. They explored new ways of achieving tasks and of rebuilding themselves with their adjusted identity. They viewed their use of

NIV more positively as a partnership, upon which they have an emotional and physical dependence, "I kind of love that thing, machine ... It's my buddy" (Hayley;  $<40$  years old +  $\leq 15$  years of NIV). For these participants, their identity, entwined as it was with NIV, provided increased autonomy and a resultant decreased dependence on others. Increased autonomy and decreased dependence was also reported by carers and family.

Once stable on NIV, some participants took the opportunity to reflect on their experiences within the health system and their prescription and use of NIV. They commonly expressed difficulties with sub-optimal consent processes, a lack of person-centred care and barriers with service delivery. However, because NIV helped to decrease fatigue and 'brain fog,' participants found they had more time to research their condition, share their experiential knowledge with health professionals and collaboratively problem solve issues. They became more actively involved in their care, able to research alternative options and advocate for themselves, "When I don't like what they are saying, I can choose for myself" (Leah;  $<40$  years old +  $\leq 15$  years of NIV).

### **Precariousness of this life**

Participants felt vulnerable; a consequence of dependency on their NIV machine. If the machine were to stop, participants expressed their uncertainty as to their life's continuation. Therefore, participants were continuously aware of the precariousness of their life because of NIV use. The need for a reliable electricity source highlighted this. For example, in the event of a power outage, and if the NIV machine had no backup battery, then death was a possible consequence. This potential occurrence weighed on participants thoughts with Sophia describing the likely outcomes if such an event was to occur.

If I am lying down in my bed, and the power goes out and nobody comes to help me I am going to die. The choices are I die [in my bed] or I take the mask off and attempt to roll out of bed. [I] possibly break something on the way down and quite possible land lying [face] down and just as stuck as I was before and die. People do die. If you Google it, people have died. (Sophia;  $<40$  years old +  $\leq 15$  years of NIV).

This concept extended to other experiences such as damage to the machine when travelling or dependence on carers' ability to operate the machine. The unpredictability and uncontrollability of these potential events heightened the sense of fragility of participants lives. For example, Hayley described one particular ordeal where she nearly drowned in her own vomit, as she was unable to independently remove the face mask.

There [have] been occasions where I vomited in a [full face mask] and it is like, 'Oh crap' ... [the machine] is still going so it pushes the vomit in and out and up the nose and I was drowning ... I tried to scream. (Hayley;  $<40$  years old +  $\leq 15$  years of NIV).

The following quote highlights how helpless the participants felt when problems arose but also an element of trust that either support will be available when required and that the machine will work as it is supposed to. However, trust could be strained by a machine being returned from servicing with incorrect settings "We have had that machine serviced and they stuffed up... the settings ... so I didn't wear it, couldn't breathe through it." (Claire;  $\geq 40$  years old +  $\leq 15$  years of NIV), or receiving an older machine that was outdated as compared to their previous one. This added to the anxiety and fear associated with NIV use. Regardless, participants needed to continue to use the machine and trust it would work as intended.

We just have to go to sleep and trust it's gonna be alright, yeah especially over winter. If something has happened, the power's gone

out ... That's a real issue. It is always a concern. (Ava;  $\geq 40$  years old +  $>15$  years of NIV).

## Discussion

The aim of this study was to explore the lived experiences of people with NMD who use NIV. The experience was difficult, complex and emotional. The NIV machine was perceived to have saved the participants' life when critically unwell, and once medically stable ongoing use of NIV provided choice around opportunities previously considered unattainable. However, participants were dependent on the NIV machine and it was challenging adjusting their identity to one that encompassed one of 'me and *that* machine.' Furthermore, many perceived their life as precarious in nature due to dependency on the machine and the persistent and pervasive fear of the equipment failing (due to power disruption, lack of backup power source or an adequate technical maintenance programme).

Previous literature has reported that living with a terminal diagnosis can result in ongoing feelings of hopelessness [49]. While only a few of the participants in our study had a terminal diagnosis, we found that all participants perceived that the introduction of NIV into their care plan provided an opportunity to re-evaluate their life plan and goals. This was highlighted in an overarching theme that NIV restored hope for people living with NMD. Participants prospered through improvements in symptom control which enabled night-time independence and ability to complete ADLs. Both machine portability and symptom control improved travel tolerance, which enabled participants to partake in recreational activities and provided participants with opportunities to network with others about their condition. There was hope that life with choices could be had.

Hope is an essential component to manage and adjust to illness [50] and is positively associated with outcomes in neurological conditions [51]. Hope can be described as an intrinsic phenomenon whereby an individual can control their existence and is pursued through goals yet to be achieved [34,50]. By this definition, NIV imbued hope within participants; in the sense that life can have meaning, despite an actual or perceived imminence of death. Other studies have described how hope is facilitated through healthcare professionals in ventilated patients [34,52,53]. However, no studies have specifically examined how NIV imparts hope in people with NMD until now.

In the current study it became apparent that there was a complex and emotive relationship that existed between participants and their NIV machine, despite all but one participant having used NIV for more than 5 years. This suggests that despite the duration of NIV use the relationship between people with NMD and their machine was still psychologically difficult for most participants. While NIV enabled life, improved independence and provided hope, participants simultaneously acknowledged their reliance on the NIV machine curtailed potential life opportunities (for example, NIV created challenges with intimate relationships). To date there has been limited research describing the impact of NIV on the sex lives of people with NMD. Furthermore, all participants noted that without NIV they would have died or there would be no life to live because their health would deteriorate. If their NIV machine stopped or broke they perceived that they could still die. While these perceptions can not be individually medically validated, it emphasises the participants dependence on NIV. When considering control or power in the human to machine relationship the balance was perceived by the participants in our study to lay very much in favour of the machine. In

some instances, participants described their NIV machine with human-like characteristics and reflected upon their need to accommodate this by adjusting their own identity to one of 'me and *that* machine.'

This is the first study to explore these perceptions of control and identity in the relationship between people with NMD and their NIV machine. In this instance, the relationship reflects a physical and psychological interaction between the human body and NIV machine to augment the user's residual function [54]. While symbiosis usually refers to living organism, the concept of symbiosis between human-machine has been evident for over half a century [55] and is usually an obligate relationship, that is, with humans dependent on the machine for survival. To date, only a few studies have described the relationship developed between people with a chronic illness and the machines that keep them alive. Patients with kidney disease have described relationships with their dialyzer as being 'one' with the machine or being a machine themselves [56]. While these patients relished the independence provided by dialysis, they were acutely aware of their dependence on and the control the machine held over their life. Another study has described the symbiotic relationship patients with diabetes developed with their insulin pump, whereby participants were one with the machine and had subsequently created adjusted identities that incorporated the machine into their lives [57].

The participants with NMD in this study were not typically involved in the decision-making process of their NIV prescription. Indeed, four participants were initially prescribed NIV as a life-saving measure whilst critically ill in an intensive care unit and the remaining participants clearly remember a discussion with no other options. An interesting juxtaposition was therefore observed in the current study. NIV provided participants with time to learn about their condition and experience hope for their life and as such they gained independence and autonomy, but the initial NIV prescription did not necessarily respect their autonomy. Participants did not feel fully informed of alternative treatment options, nor was there any meaningful discussion regarding potential benefits and adverse effects. This finding identifies the need for people with NMD to be kept central to decision making so that they can take an active role in their own care. Furthermore, tolerance and accommodation of NIV is correlated with increased patient education in both condition and treatment options [25,58]. In New Zealand, individual decision making is an integral component of the patient-clinician relationship as it recognises and respects the person's right to autonomy [59]. "Respect for inherent dignity, individual autonomy including the freedom to make one's own choices, and independence of persons" are a key principle of Article 3 of the United Nations Convention on the Rights of Persons with Disabilities [60]. Our results suggest that healthcare professionals need to be reminded of the importance of upholding these principles of respect for dignity and autonomy. The incorporation of a health literacy approach as per the New Zealand Medical Association's policy briefing document [61] may enable healthcare professionals more readily meet patient expectations around knowledge sharing, autonomy and respect of decision making.

Although not a focus of this study, participants consistently identified issues with equipment maintenance and discussed issues, such as power failure, that could threaten their existence. The process for NIV service delivery, NIV machine maintenance and solutions for NIV failure is a topic of interest for future research.



### Strengths and limitations

There are several methodological strengths in this study. The interviews were structured around open-ended questions, supporting participants to answer questions more freely and elaborate where necessary. A strong commitment was made to ensure participants felt at ease during the interviewing process. This was established through the interviews taking place at the participants' own homes, the interviewer adopting an attitude of sensitivity and naive enquiry with regards to the topics being discussed and being aware of fatigue. A thorough in-depth inductive analysis occurred, whereby the data and its interpretation were iteratively considered from an existential, and idiographic perspective underpinned by a hermeneutic phenomenology [41,47]. Furthermore, the life experiences of the research team were integral to the data interpretation providing nuanced understanding of the cognitive dissonance presented by the participants about the NIV machine.

A limitation was the potential for self-selection bias, based on participants having volunteered to take part in the study; i.e., participants may have had their own personal reasons for wishing to participate in the study, for example, being very satisfied or unsatisfied with their experiences. No explicitly obvious differences were found between the experiences of men and women. However, this was not a specific objective of the research and would have required an alternative analytical approach. In addition, we were unable to specifically explore Māori lived experiences due to the number of Māori recruited in this study. Future research is necessary to do justice to the lived experiences of this specific group. However, IPA should then be undertaken with a Kaupapa Māori Research [62] methodology; whereby Māori are defining the process, completing the research with and about Māori, with the eventual outcome having impact Māori [42,63]. Consequently, while selection bias does not invalidate the results of the study, we can only assert that the data is an interpretation of the lived experiences of participants in this study.

### Conclusion

This study explored the lived experience of individuals with NMD using NIV. Results demonstrated a complex relationship with their machine; the independence and hope imbued by NIV was moderated by the caveats of a life dependent on NIV. Consequently, NIV prescription necessitated the challenge of adjusting to how users perceived themselves or one's identity. The physical, psychological and social implications of NIV prescription for people with NMD, particularly the complex relationship with their machine, need to be recognised by healthcare professionals so appropriate supports can be provided. Furthermore, the life given by NIV use was one of a precarious nature, due to both the vulnerability of participants and their dependence on their NIV machine. This research emphasises the psychological resilience required to use NIV while living with NMD and provides healthcare professionals with insight and guidance of where support and empathy are required.

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The authors have no conflicts of interest to declare. The principal investigator, co-investigators and direct members of their families have no commercial interest in the interventions being studied, nor did they have any financial relationship to the funder that may inappropriately influence his or her conduct in the study. Principal investigator and co-investigators will, in no way, be remunerated for their involvement in a way that may inappropriately influence his or her conduct in the study. No other researchers involved face other conflicts of interest.

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