A TICKING TIME BOMB: THE LIVED EXPERIENCE OF ADULT INDIVIDUALS DIAGNOSED WITH AMYOTROPHIC LATERAL SCLEROSIS

By

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ABSTRACT

Background: Amyotrophic Lateral Sclerosis (ALS) is a fatal neurodegenerative disease with involvement of the upper and lower motor neurons in the brain and spinal cord. It is characterised by progressive muscle atrophy and to date it is incurable. The aetiology of the disease remains unclear. Patients with ALS face many challenges due to the debilitating nature of the illness and mainly depend on supportive and palliative care. There is a paucity of literature on the lived experiences of individuals diagnosed with ALS, both from a local and international perspective.

Aim: The aim of this study was to explore the lived experience of individuals diagnosed with ALS. The objectives of this dissertation were to investigate the impact of living with a neurodegenerative disease and how they strive to cope.

Design: A qualitative phenomenological study following the principles and guidelines for an Interpretative Phenomenological Analysis (IPA).

Setting: Semi-structured audio-recorded interviews carried out at Dar Bjorn, a residential community home for individuals with ALS and other neurological disorders and the participants' residences respectively.

Participants: Five adult participants diagnosed with ALS were involved in this study.

Method: To recruit participants, purposive sampling was used. The data was transcribed verbatim and analysed using Interpretative Phenomenological Analysis.

Results: Two super-ordinate themes emerged from this study, namely, 'A ticking time-bomb' and 'Striving to stay afloat'. The super-ordinate theme, 'A ticking time-bomb' described the biopsychosocial impact ALS sufferers have to contend with on a daily basis. The second super-ordinate theme, 'Striving to stay afloat', describes the coping strategies adopted by the participants to come to terms with a fatal and neurodegenerative disease. Such strategies include focusing on spirituality, facing ALS with courage and living the present.

Conclusion: ALS is a rare neurological disorder that is not only fatal but also debilitating in nature and will ultimately result in a loss of independence and autonomy. A diagnosis of ALS is daunting and had a profound physical and emotional impact on the participants. This study goes into depth about how the participants strived to cope with a disease that is known for its rapid progression and devastating symptoms that ultimately renders them completely reliant on their caregivers. Additionally, the findings revealed the strength, courage and resilience that the participants showed despite being diagnosed with ALS.

Keywords: Amyotrophic Lateral Sclerosis, quality of life, lived experience, distress, burden

DEDICATION

To

My dearest sister and my brother-in-law Steve. Their strength and resilience in the face of adversity gave me the courage to see this dissertation to the end.

"However difficult life may seem, there is always something you can do and succeed at. It matters that you don't just give up"(Stephen Hawking, 2017)

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LIST OF ABBREVIATIONS

ACP	Advanced Care Planning
ALS	ALS: Amyotrophic Lateral Sclerosis
ALSAQ:	ALS Assessment Questionnaire-40 items
ALSFRS-R	ALS Functioning Rating Scale
BiPAP	Bilevel Positive Airway Pressure
CASP	Critical Appraisal Skills Programme
CDC	Centre for Disease Control
CEO	Chief Executive Officer
COREQ	Consolidated Criteria for Reporting Qualitative Studies
DPO	Data Protection Officer
EMG	Electromyography
FALS	Familial Amyotrophic Lateral Sclerosis
FREC	Faculty Research Ethics Committee
FTD	Frontotemporal dementia
FUS	Fused-in sarcoma
HADS	Hospital Anxiety and Depression Scale
ICTRP	International Clinical Trials Registry
IPA	Interpretative Phenomenological Analysis
JALS	Juvenile Amyotrophic Lateral Sclerosis
KALSA	Korean ALS Association
LMS	Langer Mindfulness Scale
MDH	Mater Dei Hospital
MeSH	Medical Subject Headings Browser
MND	Motor Neuron Disease
MRI	Magnetic Resonance Imaging
MS	Multiple Sclerosis
NGO	Non-governmental Organisation
PEO	Population Exposure Outcome
PRISMA	Preferred reporting items for Systematic Reviews and Meta-Analyses
QOL	Quality of life
SA-ALSFRS	Self-administered ALS Functional Rating Scale
SAMOC	Sir Anthony Mamo Oncology Centre
SODI	Superoxide dismutase 1
SPB	Self-perceived Burden
SPBS	Self-perceived Burden Scale
TARDBP	Transactive response DNA binding protein
UM	University of Malta
UREC	University Research Ethics Committee
WHO	World Health Organization
ZBI	Zarit Burden Interview Scale

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Chapter 1: Introduction

1.1: Introduction

Idiopathic in nature, amyotrophic lateral sclerosis (ALS) presents as a heterogenous, debilitating and fatal neurodegenerative disorder which significantly affects the quality of life (QOL) of both the patient and the individuals entrusted with their care (Bento-Abreu, 2010). The heterogeneity of ALS is notorious for its rapid degeneration of upper motor neurons, which extend from the cortex to the brainstem/spinal cord and lower motor neurons, which extend from the brainstem/spinal cord to muscle ((Hardiman, 2017). For further clarification, 'amyotrophy' refers to atrophy of fibres within the muscle, which denervates due to degeneration of their corresponding anterior horn cells which leads to weakness in the affected muscles, spasticity and involuntary rapid muscle twitches known as visible fasciculations (Rowland, 2001). 'Lateral sclerosis' refers to the hardening of the anterior and lateral corticospinal tracts whereby motor neurons in these areas degenerate and are replaced by a process called gliosis (Schneider, 2001).

This research study will explore the lived experiences of individuals diagnosed with ALS, both the familial and sporadic types.

1.2: Background of The Study

ALS is considered relatively rare, with an incidence rate of approximately 1-9 per 100,000. However, the socioeconomic consequences are considerably high (Schepelmann, 2009) and according to Johnston et al. (2006), 1/400-1/700 individuals are estimated to develop ALS over a lifetime. Furthermore, an interesting study by Arthur et al. (2016) addressed an estimation of individuals being diagnosed with ALS across the globe from 2015-2040. The results showed that in the coming 25 years, the cases of ALS will increase by 69%, the primary reason being population ageing, especially amongst developing countries. Additionally, the age group with the highest predominance of ALS cases was between 60 and 69 (Arthur, 2016). Research on the epidemiology of ALS shows that in Europe, the incidence of ALS is higher and, at present, is estimated to be approximately 2.6 per 100,000 (Mehta, 2022). Moreover, research from the Centre for Disease Control (CDC) indicates that Caucasian individuals are more likely to be diagnosed with ALS than other demographic groups and both the prevalence and the incidence slightly tilt towards men rather than women, the M: F ratio being approximately 1.5:1 (CDC, 2022).

ALS presents in two forms, sporadic, found in approximately 90% of patients and familial, which affects the remaining 10% (Van Damme, 2009). According to a study by Bento-Abreu et al. (2010), genetic mutations supposedly found in familial ALS (fALS) were also found in several individuals diagnosed with the sporadic form. However, researchers are still unclear whether this is a case of new mutations or whether ALS was not the sporadic type. Such a phenomenon can occur due to cases of non-paternity, vague and incomplete family histories or 'incomplete penetrance'.

For the most part, the inherent form of ALS presents itself with an autosomal dominant pattern of inheritance (Kirby, 2016). However, in rare instances, ALS can also be inherited in an x-linked dominant pattern (Valdmanis, 2009). Although the causation of ALS remains obscure, studies show that fALS and the number of sporadic cases are determined by single gene mutations (Al-Chalabi, 2017). For example, a study by Nguyen (2018) showed that there are over 25 genes known to be linked to ALS. However, four genes account for approximately 70% of all fALS cases and 10% of sporadic ones (Al-Chalabi, 2017), namely, C9orf72, superoxide dismutase 1 (SOD1), TARDBP and fused in sarcoma (FUS).

The survival rate of ALS, which is approximately 3-5 years from disease onset, varies amongst patients (Bento-Abreu, 2010). ALS causes progressive muscular paralysis, leading to loss of life due to 'neuromuscular respiratory failure' (Talbot, 2017) and bulbar dysfunction (Bento-Abreu, 2010). Furthermore, a recent study by Vahsen et al. (2021) noted a specific heterogeneity regarding disease onset, progression rate and pattern. Additionally, when this clinical heterogeneity is combined with the existing overlap between ALS and FTD, it is evident that the pathophysiology of ALS is guided and altered by complex biological factors (Vahsen, 2021).

According to Shellikeri et al. (2017), the disease onset of ALS may differ from one patient to another. There are two ways in which ALS may present itself. In the 'spinal-onset' form, patients may exhibit muscle weakness in their limbs, whereas those with 'bulbar-onset' disease may notice symptoms such as difficulties in speech and swallowing (Hardiman, 2017).

From a biological perspective, ALS is considered more than a motor neuron disease (MND) since there is clear evidence of various 'neuronal systems'. In addition, research shows that

approximately 50% of patients exhibit cognitive and/or behavioural impairment, while 13-15% present with 'concomitant behavioural variant FTD' (Phukan, 2007).

Individuals diagnosed with ALS go through a rollercoaster of emotions, as will be evident during the findings of this study. These patients are faced with a fatal disease that has a short lifespan, rapid neurodegeneration, and no foreseeable cure. Moreover, the individual will lose his/her autonomy and, at some point, will experience an inability to perform even the most basic activities of daily living. Thus their QOL will slowly start to diminish (Brown, 2017). Social marginalisation occurs when the patient becomes increasingly dependent on his/her caregiver (Galvin, 2016). This happens mainly due to a loss of communication. The atrophy of muscles will lead to spasticity that will inadvertently affect the patient's dexterity and gait (Wijesekera, 2009).

Furthermore, patients diagnosed with 'bulbar onset' ALS initially present with incoherent speech and difficulty swallowing. However, symptoms of weakened limbs may also appear together with bulbar symptoms (Leigh, 2009). In addition, patients may experience cognitive and/or behavioural impairments, such as impulsivity and feelings of apathy (Miller, 2009). In bulbar ALS, considered the most devastating form of ALS, the survival rate is 1-2 years from diagnosis, whereas in spinal onset ALS, death may occur in 3-5 years (Goldstein, 2002). Furthermore, in ALS, paralysis progresses rapidly, and patients succumb to the disease due to respiratory failure (Wijesekera, 2009).

To arrive at a diagnosis of ALS, a medical history, magnetic resonance imaging (MRI) and a muscle biopsy are carried out. These are combined with an electromyography (EMG), a clinical test that detects neuromuscular abnormalities. In addition, diseases which 'mimic' ALS must be ruled out (Wijesekera, 2009).

A diagnosis of ALS will understandably affect the well-being of the patient and his/her caregiver in the most profound ways. Furthermore, individuals diagnosed with ALS can be offered support, palliative care and management by a multidisciplinary team (Bede, 2011). Non-invasive ventilation may also prove helpful since it extends survival and improves the patient's QOL. On another note, Riluzole, used to treat ALS and other MNDs, has been known to be the only medication to date that delays ventilator dependence and possibly prolong life by 2-3 months (Miller, 2012).

1.3: The Local Scenario

At the University of Malta (UM), Professor Ruben J. Cauchi, an associate professor of neurogenetics, and his team of researchers lead the MND laboratory dedicated to placing forth a better understanding of ALS and MND. Currently, ALS is incurable (Cauchi, 2022). However, for over ten years, Prof. Cauchi and his team have been dedicated to studying the genetics of ALS and its mutations partly responsible for an individual developing ALS. In addition, they aim to search for therapeutic targets to treat the disease while supporting individuals with ALS and their families.

Furthermore, Prof. Cauchi and his researchers set up a 'biobank' repository whereby blood samples and data are collected from local patients and stored. By analysing blood samples from the biobank, the ALS/MND team at UM can understand the context of ALS in the Maltese islands. The team also collaborates closely with the patients' neurologists to advise and inform them of any promising clinical trials that may benefit their patients.

A study by Farrugia Wismayer et al. (2021) was conducted on 38 patients with ALS and 45 controls regarding environmental factors that may increase the risk of developing ALS. Results from this population-based case-control study showed that manual workers with a history of physical exertion may be more at risk of developing ALS. In addition, individuals of this particular type of occupation also had an increased risk of developing bulbar-onset ALS. Interestingly, research on other populations revealed that military professionals risk developing ALS more than their civilian counterparts.

In August 2015, Mr Bjorn Formosa, a 28-year-old businessman diagnosed with ALS, decided to spearhead a campaign in an effort to provide a residential home that provides around-the-clock care for individuals diagnosed with ALS and other neurological conditions. Mr Formosa, the founder of ALS Malta, established Dar Bjorn, a registered voluntary organisation which accommodates 13 patients and is situated in Qormi. It was officially opened on 1 November 2017 and began admitting its first patients on 27 November 2017. Increased demand for such services led Mr Formosa to open a second and larger home, ALS Malta Respite Home.

Vowing to dedicate the time he has left to raising awareness of the severe challenges faced by individuals diagnosed with ALS and their caregivers, Mr Formosa believes that his organisation, ALS Malta, has been instrumental in offering sufferers a sense of community whilst also helping to support the research required in the field of ALS and MND related

disorders. As Mr Formosa (2020) quoted on Dar Bjorn, 'It is about providing some sort of normality and giving everyone the same opportunities'.

A dearth of literature on ALS from every perspective exists from an international point of view. However, local studies on the lived experience of ALS patients were non-existent. The author found only one dissertation at Master's level whereby the researcher investigated the 'suffering and the request for assisted suicide by ALS patients' (Said, 2018).

1.4: The Present Study

During the literature review in Chapter Two, research on ALS from the past decade consisted of quantitative and qualitative studies. However, most articles, which were medical and primarily addressed ALS's genetics, were quantitative. However, research on the actual lived experience of patients with ALS was scarce and focused more on the burdens experienced by their primary caregivers. Moreover, quantitative studies delved into the technicalities from a medical point of view. They did not offer in-depth exploration and personal analysis of an individual with a fatal neurodegenerative disorder. This insight on a deeper level could only be approached via qualitative research methods since the individuals diagnosed with ALS could elaborate more on how their lives have changed since their diagnosis. That unique, deeply spiritual and emotional moment whereby the researcher allows her patients to voice their thoughts, fears, hopes and opinions on living with a terminal illness which leads to progressive paralysis and ultimately robs them of their independence, including the ability to speak, eat, drink and breathe. Billington (2006) states that the account of the 'insider' is considered an invaluable evaluation of existing care plans and change.

This research study explores the lived experience of individuals diagnosed with ALS. An in-depth account of the individuals' life experiences of coping with this neurodegenerative disorder was required. Thus, an Interpretative Phenomenological Analysis (IPA) was considered the best option (Smith et al., 2009). Data collection took place in the form of face-to-face semi-structured interviews carried out at Dar Bjorn and the individuals' residences respectively. According to Smith et al. (2009), semi-structured interviews are the epitome of placing the patient at the centre of the research whilst better understanding his/her lived experience.

Consequently, this dissertation aimed to collect data about the lived experience of patients diagnosed with ALS. The objectives of this study are listed in the table below:

Table1. 1. Objectives

➢ To understand from the patients' perspective, the impact a progressive, debilitating and neurodegenerative disorder has on the biopsychosocial aspect of their lives.
➢ To investigate the coping strategies of the patients

1.5: The Significance of The Study

The author of this study opted for a qualitative approach because it was the best option for her to research, at depth, the lived experiences of patients diagnosed with ALS. Furthermore, it was the evident paucity in local studies that target the lived experiences of individuals with ALS that encouraged her to address this area of research. In fact, it is the only local study that targets the biopsychosocial impact of ALS on patients. This research, thus, will allow patients with ALS to be able to voice their concerns about what it is like to be diagnosed with a fatal illness that is not only debilitating but rapidly progressive.

As a nurse specialised in Mental Health, the author understands that a diagnosis of ALS can have serious repercussions from a biopsychosocial aspect on both the patient and his immediate family/primary caregivers. Thus, by means of this research, it is her aspiration that the challenges and grievances of this particular cohort of patients are recognised and that the broad spectrum of healthcare professionals entrusted with their care will be provided with a deeper insight on how to provide the specialised care ALS requires.

1.6: The Dissertation Structure

A brief description of the structure of this dissertation is described in the table below:

Table1. 2: The Dissertation Structure

Chapter	Description
Chapter 1	➢ Introduction
Chapter 2	➢ Literature Review
Chapter 3	➢ Description and rationale of IPA
Chapter 4	➢ Analysis of data from the semi-structured interviews
Chapter 5	➢ Discussion of Findings
Chapter 6	➢ Strengths and Limitations

Chapter 2: Literature Review

2.1: Introduction

This chapter addresses the theoretical framework and critically reviews the literature. The aim is to understand the lived experiences of individuals diagnosed with ALS comprehensively. An intricate and detailed search of various electronic databases was carried out, with the researcher specifying how relevant literature was identified and extracted. The Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines (Moher et al.) were applied to ensure a systematic description of every step in the literature review process. Furthermore, this chapter will discuss and critique the research that emerged from the search strategy concerning the experience of being diagnosed with ALS.

2.2 The PEO Method

The aim and objective of this review was to explore the lived experience of individuals diagnosed with ALS, both sporadic and familial, hence the research question proposed by the author, 'The lived experience of individuals diagnosed with ALS. An Interpretative Phenomenological Analysis. The population, exposure and outcome (PEO) method (Khan, 2003) was used to determine which studies should be included and excluded in this review and is listed in the table below:

Table 2. 1 The PEO Method

Population	**Exposure**	**Outcome**
Male and/or female adults (aged 18 years and above) diagnosed with ALS both of the sporadic and familial type.	A diagnosis of ALS both of the sporadic and familial type.	Biopsychosocial impact, well-being, hope, quality of life, burden, dealing with a debilitating and terminal disorder, lived experience, resilience, advanced care planning.

2.3 Inclusion And Exclusion Criteria

Introducing inclusion and exclusion criteria were crucial in narrowing down and refining the search. According to Aveyard (2010), such criteria "give vital information about the scope and relevance". Furthermore, the criteria chosen for this review are tallied with evidence-based research on the PEO method. Most of the criteria were inserted using the database filter tools. However, this varied from one database to another. Table 2.2 briefly describes the inclusion and exclusion criteria used for each element of the PEO method:

Table 2. 2: Inclusion and Exclusion criteria applied during the search strategy

Inclusion Criteria	Exclusion Criteria
➢ Male and female adults aged 18 years and above diagnosed with ALS both of the sporadic and familial type. ➢ Individuals not at end-of-life ➢ Articles in English Language ➢ Articles published within the time-range of 2012-2022 ➢ Peer-reviewed articles ➢ Full-text articles	➢ Participants aged less than 18 years (in this case the disorder is known as Juvenile ALS). ➢ Participants that are at end-of-life. ➢ Articles that addressed neurological disorders that mimic ALS (such as myasthenia gravis, poliomyelitis, progressive bulbar palsy and motor neuron disease with other malignancies). ➢ Articles that discussed Multiple Sclerosis. ➢ Studies that addressed end-of-life care. ➢ Studies that did not relate to lived experiences quality of life and coping strategies as an outcome. ➢ Articles written prior to 2012 ➢ Articles which did not include full-text.

2.2: The Search Strategy

A tentative search was carried out on Google Scholar to explore what literature could be deemed relevant to the study. This freely accessible search engine indexes the full text of scholarly literature across various publishing formats. The search yielded a vast amount of

literature, both relevant to this study and irrelevant. However, due to time constraints, going through every entry would have proved impossible. In place of this, the researcher made use of the UM library whereby a detailed and methodical literature search was carried out on eight electronic databases, namely, Wiley Online Library, Taylor and Francis Online, Web of Science, Cinahl Complete, Nursing and Allied Health Database, Medline Proquest, Oxford Academic Journals and Pubmed. This comprehensive literature search was carried out to prevent the introduction of search bias and to increase the yield of relevant articles (Randolph, 2009). The researcher carried out a basic search of 'ALS in Malta' on Hydi to discover pertinent literature on ALS from a local perspective. The search yielded 14 articles. However, none of these articles could be used for this study since they did not address the lived experience of individuals diagnosed with ALS per se.

Consequently, the only studies found were conducted by Professor R. Cauchi, an associate Professor in Neurogenetics and a team of researchers at the Centre for Molecular Medicine and Biobanking at UM. These articles mainly addressed ALS and MND from a genetic and occupational perspective. Furthermore, while searching for local studies on ALS, a dissertation titled 'Suffering and the request for assisted suicide by ALS patients' (Said, 2018) came up. Therefore, the researcher reviewed the reference list and found 13 articles relevant to her study on the lived experience of individuals diagnosed with ALS.

Due to an evident paucity of literature that directly addresses the lived experiences of patients with ALS and in order to ensure a detailed and thorough search of evidence-based literature, various journals that specialise in neurological sciences, neurodegenerative disorders, neurology, palliative care and ALS/MND were explored (refer to table 2.4). However, most articles in these journals had already been encountered by the researcher in her chosen electronic databases. Moreover, apart from searching for literature consistent with the research question, the researcher needed to become well-versed in MNDs and the complex genetics and pathophysiology of amyotrophic lateral sclerosis. This was accomplished by finding separate articles and book chapters on ALS/MND.

During the first stage of the search strategy, the general search on the topic of interest is vast. Therefore, using the advanced search tab and introducing specific keywords with Boolean operators such as 'AND', 'NOT' or 'OR' were crucial in narrowing down or broadening the search. Furthermore, due to the medical nature of the topic, the author used the Medical Subject Headings browser (MeSH), a comprehensive online library that serves as a thesaurus for

medical terminology—a method known as truncation was utilised, whereby diverse variations of the stem are picked up. Finally, to further refine the search, filters were used. The filters included 'peer-reviewed articles', 'full-text', 'English language' and a custom range timeframe between 2012-2022 or 'the last ten years'.

Furthermore, the following keywords were used by the author during the entire search strategy: "amyotrophic lateral sclerosis", "ALS" "quality of life", "lived experience*", "QOL", "distress", "burden", "phenomenology". These keywords were identified after meticulous research and background reading of relevant articles. Overall, 1,986 relevant studies were found by searching eight electronic databases. A summarised table displaying the electronic databases used, the applied filters, keywords and results of the search is listed below:

Table 2. 2: The search strategies and databases used

Database	**Access Date**	**Search Strategy**	**Articles found**
Wiley online Library	October 23, 2022	Search Limits: Published dates 1/1/2012-31/12/2022 and English Language Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience* AND Distress AND Burden AND spirituality	**91 results**
Taylor and Francis Online	October 23, 2022	Search Limits: Published dates 1/1/2012-31/12/2022 and English Language Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience* AND	**102 results**

		Distress AND Burden AND spirituality	
Web of Science	October 23, 2022	Search Limits: Published dates 1/1/2012-31/12/2022 and English Language Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience* AND Burden AND Distress AND phenomenology	**975 results**
Cinahl Complete	October 23, 2022	Search Limits: Published dates 1/1/2012-31/12/2022, English Language, Full-text, peer-reviewed, all adults, male and female, humans Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience* AND Burden AND distress AND phenomenology	**351 results**
Nursing and Allied Health Database	October 23, 2022	Search Limits: Published dates 1/1/2012-31/12/2022, English Language, Full-text, peer-reviewed, adults, male and female	**162 results**

		Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND lived experience* AND Burden AND Distress AND Phenomenology	
Medline Proquest	October 24, 2022	Search Limits: Published dates 1/1/2012-31/12/2022, English Language, peer-viewed, all adults, males and females and humans Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience* AND Burden AND Distress AND phenomenology	**106 results**
Oxford Academic Journals	October 24, 2022	Search Limits: Published dates 1/1/2012-31/12/2022 and English Language Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience* AND Burden	**43 results**
Plos	October 24, 2022	Search limits: published dates 1/1/2012-31/12/2022 and English Language Amyotrophic lateral sclerosis OR ALS AND "quality of life" OR QOL AND Lived experience*	**156 results**

		AND Burden AND distress	

Apart from a meticulous search strategy covering electronic databases and specialised journals, the author also reviewed the reference lists of the articles extracted during the screening process. As a result, two articles were found using this method.

2.3: The Grey Literature

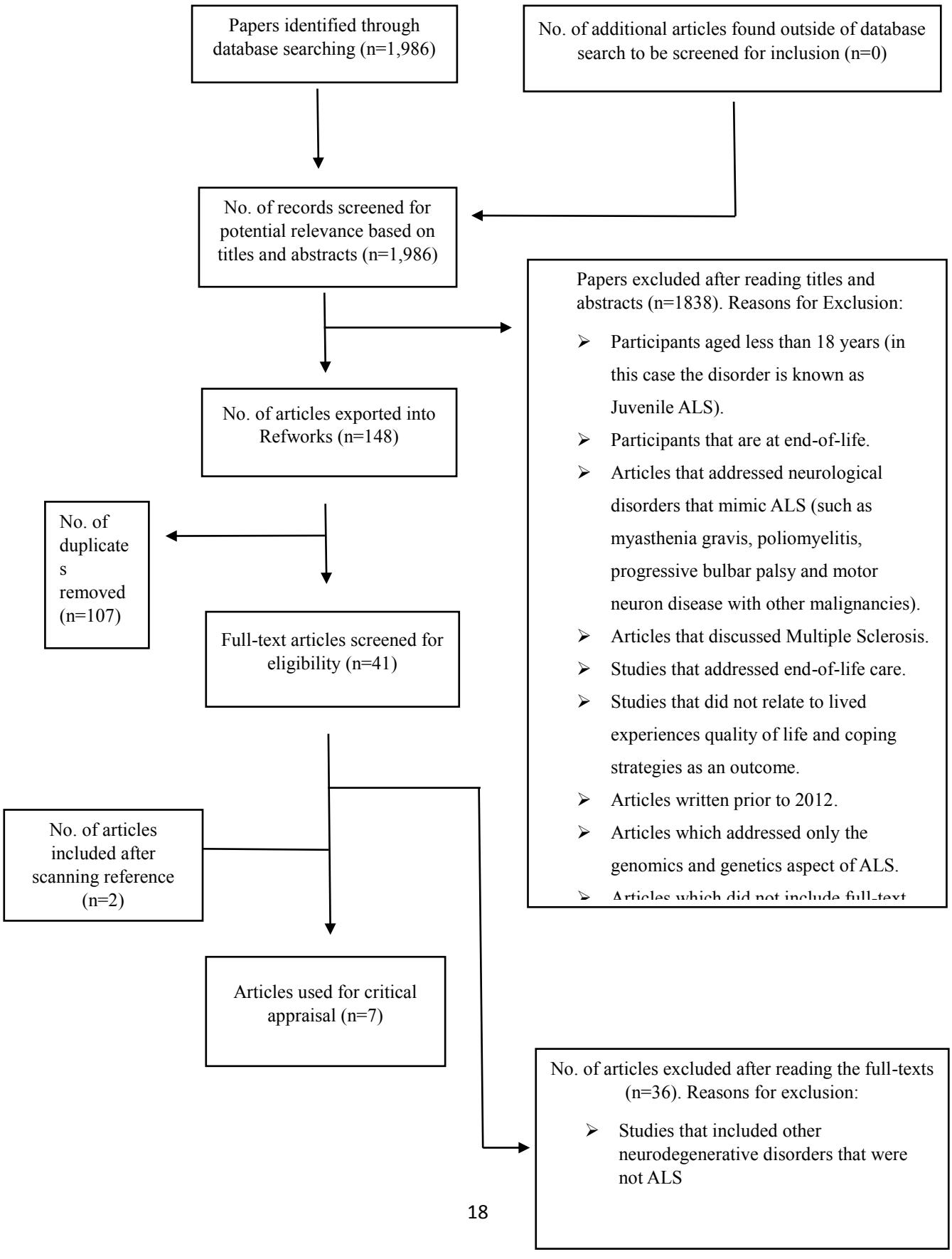
As per recommendation by Randolph (2009), a search for grey literature was carried out to reduce search bias and ensure sound and transparent literature retrieval. The Third International Congress (1997) defined *grey literature* as 'produced on all levels of government, academics, businesses and industries in electronic and printed formats not controlled by a commercial publisher (Auger, 1998). Sources for grey literature include non-governmental organisations, advocacy groups, university libraries and professional societies; thus, to ensure a sound and transparent literature retrieval, various databases such as open grey: eu, clinicaltrials.gov, and World Health Organisation (WHO) International Clinical Trials Registry (ICTR). A basic search used the key terms' amyotrophic lateral sclerosis' AND 'lived experience'. This search yielded no relevant articles that could be used in this review; however, Google's search engine revealed the ALS Association, a website dedicated to people living with ALS and their caregivers. This website contained several blogs and podcasts whereby individuals could share their experiences of what it is like to deal with being diagnosed with ALS. The author accessed these blogs and podcasts to gain more personal insight into the lived experience of such individuals.

2.4: Identification And Screening of Articles

The comprehensive search in the electronic databases identified 1,986 potentially relevant articles. As stated previously, the author adhered to the PRISMA guidelines (Moher, 2009); thus, the article's eligibility was based only on the title at this early stage. However,
abstracts were reviewed in papers that appeared equivocal and/or vague; after discarding irrelevant literature, the researcher chose 148 papers (including duplicates). Furthermore,

reading only the title and/or the article's abstract sometimes needed to be more accurate. Thus, the entire paper had to be scrutinised at this point. Reasons as to why papers were excluded are illustrated clearly in Figure 2.1. The domain ‘others' addressed the reasons for exclusion, namely, articles which concentrated solely on the genetic make-up of ALS, those on end-of-life, the burden on the caregiver and not the individual with ALS, the cost-effectiveness of having such an illness, the medications prescribed for ALS and so forth.

Figure 1: Flow diagram describing the screening process, modelled after Moher (2009)



As described in Figure 2.1 above, 148 extracted articles were exported into bibliographic software (RefWorks), and any duplicate papers were removed. They were using this method. Articles were excluded, and the full text of the remaining papers was retrieved. This was done by searching for google scholar or when that option was unavailable. The author searched for them on HyDi. One author was contacted directly via email for a highly relevant article only available in Korean. However, the author did not reply to the request. At this point, the author conducted a more intensive search for this article and finally found one published in English.
According to the inclusion and exclusion criteria, as previously stated, five studies were identified for critical appraisal. The data from these articles were further analysed and included in this literature review.
As stated beforehand, this chapter describes and analyses the critical appraisal of articles chosen for this dissertation in relevance to the PEO question. Critical appraisal can be defined as the systematic process of analysing research based on its trustworthiness and relevance (Burls, 2009). Furthermore, according to Melnyk (2014), a critical appraisal can be considered a crucial aspect towards evidence-based practice, and through its process, conclusions regarding the validity of data can be drawn and made viable for use in local practice (McLeod, 2004). To assist the author in carrying out a critical appraisal of each article's methodology, the Critical Appraisal Skills Programme (CASP) tool for qualitative research was used. The CASP tool is acknowledged as the most frequently used tool in health-related qualitative syntheses and is recommended for novice qualitative researchers (Long, 2020). Three major issues, namely rigour, credibility and relevance, are considered when applying the CASP tool to qualitative literature (CASP, 2018).

2.5: Review of Included Studies

The researcher extracted seven articles that were deemed relevant to her study. These investigated the lived experience of individuals diagnosed with the neurodegenerative disorder ALS. Furthermore, it was decided to group the articles into five themes: hope, self-perceived burden, psychological interventions, QOL and the illness journey.
While conducting the literature review, a dearth of studies whereby the issue of the self-perceived burden by the patient him/herself was noted since most of the articles explored the burden of caring for an individual with ALS from the caregiver's perspective. Furthermore, the in-depth exploration of the sufferer's lived experience indicated a paucity of literature. The role of the researcher in this dissertation is to investigate how ALS affects an individual's life

experience. The following section will describe an in-depth critical appraisal of seven articles that the author opted to group into themes. Table 2.5 below provides the salient points of the critical appraisals:

Table 2. 3: Data extracted from 7 articles used for critical appraisal

Author/s, Year of publication, Country	**Purpose**	**Methodology/Methods**	**Sample Characteristics**	**Main Findings**
Meng-Mei et al 2021 China	To explore the illness experience for people with ALS in China and the meaning they attach to their experiences.	Phenomenological Qualitative Study Colaizzi's method of data analysis was utilised. The 32-item checklist, Consolidated Criteria for Reporting Qualitative Research (COREQ) was used.	20 individuals with ALS were recruited from the Neurology Department of a tertiary hospital in Wuhan, China. Sample population was made up of 13 males and 7 females, the youngest patient being 28 years of age and the eldest 72. Participants had been diagnosed with ALS in the previous 6-60 months. Participants were subjected to an in-depth semi - structured interview	Three themes and eight sub-themes emerged from the study. The three themes addressed Life countdown, family self-help and reconstruction of life. The participants explained how the deterioration of their bodies as the disease progressed left a deep impact on their self-perception. The debilitating, physical effects of the illness caused them to become estranged from others. Denial of having a terminal illness also emerged with the participants hoping they had been misdiagnosed. Fear and hopelessness were most mentioned during the interviews since the patients associated ALS with death. Furthermore, family as a supportive role was crucial. In the case of these participants, the caregivers were either spouses or parents who provided them with great support. Acceptance of ALS was another concept the participants had to deal with and by confronting this, they could finally rediscover the meaning of life. Strongly influenced by their culture, the participants believed that thinking about their present life was more meaningful than worrying about death.

			with open-ended questions which investigated the individual's thoughts, emotions and situations of being diagnosed with the neurodegenerative disorder of ALS. Participants presented with a variety of functional states at time of interview.	
Hamama-Raz et al 2021 Israel	Understanding the meaning of hope among individuals with ALS	A qualitative thematic synthesis review which was based upon a phenomenological-hermeneutic perspective. Participants underwent in-depth, semi-structured interviews and a brief sociodemographic questionnaire.	A population sample of 12 individuals, the majority of which were males, were recruited via an Israelite non-profit organisation. Ages ranged between 42-79 and recruitment period was from January 2012-June 2015. Participants were Jewish and time	Based on data collected from the interviews, core themes were constructed using thematic data analysis. Two different themes regarding hope emerged. The first voice portrays hope as an obstacle that denies individuals with ALS selfhood and reduce one's free will. The second voice voiced defined hope as a source of empowerment. The majority of participants (n=7) viewed hope as an obstacle and stated that there is no hope in disease, and that hoping is considered a waste of time since to date, ALS is incurable. The rest of the participants (n=5), viewed hope as a resource and two participants compared hope to strength and power. With another participant explaining how hope is her 'spirit' and 'driving force'. Several metaphors emerged during this study, namely 'light' and 'tunnel', which can be translated as the individual, on diagnosis, feels as if he/she is trapped in a tunnel, however light is hope.

			of diagnosis ranged from 1-10 years and their present functional state was heterogenic.	
Marconi et al 2016 Italy	To investigate the experience of meditation for individuals with ALS and their caregivers.	A qualitative Analysis Analysis of data with a grounded theory approach	26 individuals with ALS and their caregivers were recruited ant the Neuromuscular Omnicentre (NEMO) in Milan, Italy. Meditation session were held once a week and duration of eight weeks. Time of diagnosis was between 1-2 years.	Two macro-areas were established, namely, Resources and limitations and eight main domains were identified. Results of the study revealed that both participants and their caregivers responded in a positive manner to meditation training. Improvements in anxiety and depression were recorded and sleep issues alleviated. Positive effects in QOL and mindfulness were also noted. The group setting was beneficial for both participants and caregivers. Who felt relieved of their burdens, understood and comforted. A necessity for studies on meditation and chronic disorder such as ALS were reported.
Pagnini et al 2015 Italy	Investigating mindfulness, physical impairment and psychological well-being in people with ALS.	Exploratory longitudinal study with online repeated measures. Mindfulness was evaluated with the LMS, physical impairment with the SA-ALSFRS, QOL with the MQOL-SIS, depression	197 individuals with ALS were recruited via the ALS National Registry and other ALS associations. Announcements were posted on ALS forums and mailing	SA-ALSFRS scores decreased with time however mindfulness reduced this decline resulting in a positive influence on the individual's physical functioning. SA-ALSFRS was 2.88% in participants who had high mindfulness whilst it was higher in participants with low mindfulness. Mindfulness also predicted a positive influence on QOL when evaluated by the MQOL-SIS. The same predictions resulted in depression and anxiety. The findings of this study suggest that psychological measures are not related to ALSFRS, which measures physical functioning.

		and anxiety with the HADS-A and HADS-D respectively. A mixed-effect model was used to evaluate the effect of mindfulness on patients' physical impairment. QOL, depression and anxiety. An online survey was completed designed with Qualtrics survey software. Data was collected twice, four months apart.	lists to individuals with ALS. Fewer females participated and the mean age at baseline was 58. 107 participants completed the first assessment with 102 completing the second survey after four months.	
Geng et al 2016 China	Investigation of patients' self-perceived burden, caregivers' burden and QOL for ALS patients.	A cross-sectional study Severity of ALS patients was measured by the ALSFRS-R, the QOL of patients was measured using the Chinese version of the WHOQOL-BREF, the SPB was measure by the Chinese version of the Self-perceived burden scale (SPBS) and finally, the caregivers' burden was measured using the Zarit-	81 patients with ALS were recruited from the Department of Neurology of West China Hospital of Sichuan University between March-September 2015. All patients were diagnosed with probable or definite ALS according to the revised criteria of El	The results of SPB showed that 27 patients felt little or no burden, 25 patients mild-to-moderate, 18 patients moderate to severe burden and 11 patients felt severe SPBS. Females scored higher than their male counterparts. Patients with mild physical impairment scored lower SPBS, whereas those with more severe impairment scored high SPBS. Patients who had better knowledge of ALS cored high SPBS whereas those who had poor knowledge scored lower. In caregiver burden, 29 caregivers bore little or no burden according to the mean ZBI score, 37 stated that they had mild to moderate burden, 13 had moderate to severe burden and two had severe burden.

		Burden Interview scale (ZBI). Face-to-face interviews and in addition to demographic information, participants/caregivers had to fill out self-rating questionnaires.	Escorial. Patients/caregivers were divided into two groups (Under 45 years of age and over 45). Under 45 were considered early onset and over 45 late onset. Mean age of patients was 52.7 and there were more males than females in this study. On the contrary, the majority of caregivers were females.	Patients with high ALSFRS-R scores were noted to have higher WHO-QOL-BREF scores than those with lower ALSFRS-R scores.
Kukulka et al., 2019 USA	The perspectives and implications for palliative care teams and stakeholders on the biopsychosocial and spiritual realties of living with ALS.	A mixed-method design with quantitative data supplementing qualitative data. The Biopsychosocial Model of Illness was utilised Semi-structured interviews were carried out.	14 Caucasian patients (7 males and 7 females) aged 50 and older and were patients living in a private residence in the community. 16 family members and 12 healthcare	Qualitative themes were organised into two categories. The first category pertained to the biopsychosocial needs of patients with ALS and their families, and the second category included the relationship between the spiritual and emotional aspects of patients' lives. The findings yielded that the most frequent need was additional caregiving support both at home and in the community. These included affordable care facilities, skilled personnel and practical needs at home. A need for more education on ALS was also identified in the study although the patients and caregivers did have knowledge of ALS in varying degrees.

		In addition, a 40-item ALS Assessment questionnaire (ALSAQ-40) was used to measure the patients' subjective well-being.	providers took part in the study.	Social support was another need that arose during the interviews. Patients stated that social interaction decreased which led to less contact with friends. 57% of patients reported that they experienced boredom and loneliness. Transportation and resources that led to the patients' independence were also mentioned since the patients who used large wheelchairs remained homebound. The impact of ALS on spiritual and emotional well-being varied. Nine reported that having ALS strengthened their spirituality whereas three reported no change and two felt confusion and anger at God. Patients who experienced positive effects stated that they attended religious services more regularly and had more interaction with their spiritual community. Lower report of helplessness and hopelessness were reported in these cases. In view of palliative care teams findings, a need for additional caregiving help, resources and biopsychosocial support was mentioned.
Oh et al., (2014) South Korea	A Journey of Suffering: The lived experiences of individuals diagnosed with ALS in South Korea	An ethnography method with semi-structured interviews, photo-elicitation, a survey and participant observation as a means to gain understanding into the illness experience of individuals diagnosed with ALS and their caregivers.	A population sample of 15 individuals (13 males and 1 female) aged between 38-64 years. Mean age of participants was 49.2 and the study was dominated by male participants. Patients with neurological disorders other than	This ethnography with photo-elicitation, semi-structured interviews, surveys and participant observation was carried out on 15 participants in Seoul, South Korea. The Journey of Suffering was the super-ordinate theme of which three themes emerged: Off the course, drifting and on a new boat. The three themes explore ALS from a cultural perspective and how the patients' lives were affected by this disease. The article also addressed the existential dimension of suffering. The first theme, off the course reflected how the actual journey of suffering began on the onset of symptoms and the diagnosis.

			ALS were excluded from the study. Patients had to be registered with the Korean Amyotrophic Lateral Sclerosis Association (KALSA).	The second theme, drifting, described the challenges faced and how their physical, emotional and social relationships changed. New challenges in regards to the rapid deterioration of symptoms was described. The third theme, 'on a new boat' described the participants' new found journey, now that they were diagnosed with ALS. They describe how new friendships within the circle of ALS were formed and how governmental support helped them to integrate their illness and suffering by using the resources offered.

2.5.1: The Meaning of Hope of Individuals Diagnosed With ALS

In the face of progressive neurodegenerative disorders, which will eventually lead to disability and death, this article by Hamama-Raz et al. (2021) addresses the issue of hope among individuals with ALS.

Using a phenomenological-hermeneutic perspective in this qualitative review, this study aimed to construe how people with ALS perceived their life experience of living with such a debilitating illness and how they made the necessary adaptations to cope (McLeod, 2001). The research design, which consisted of semi-structured interviews and a sociodemographic questionnaire based on an interview schedule, allowed the candidates to express their thoughts and emotions freely whilst taking on the form of a narrative rather than simply a set of questions that require answers (Roulston, 2010). How the interview schedule was prepared allowed the participants to depict their thoughts, reflections and behaviour on the meaning of hope when facing a terminal illness (King, 2011). However, the method for the researchers' data collection was not precise. A sample of 12 people, who were Hebrew speakers recruited via an Israelite non-profit organisation (NGO), participated in this study from 2012-2015 (Hamamas-Raz). For ease of reference and clarity, this study's authors inserted the participants' demographics and their functional state at the time of the interview in a table. Specific individuals, who had no speech difficulties at that time, gave diverse reasons why they did not wish to participate. These included not wanting to speak about their feelings, not wanting to be part of the study and outright denial whereby the participant believed he/she would be cured. With ages ranging from 42-79, the time of diagnosis varied between 1 to 10 years. The participants were at a different functional stages in their illness, with some still having the ability to walk; some paralysed in both arms and both legs and others paralysed in all four limbs and connected to bilevel-positive airway pressure machines (Norden, 2021). This study aimed to strengthen the voices of individuals with terminal illnesses since research shows that such people are insufficiently distinguished. Furthermore, the inclination to associate their explanations directly with the disease is rife (Peled, 2002). The participants' autonomy was ensured throughout the study, and their consent was given. This was done to avoid any unnecessary duress on behalf of the individuals since they had previously met their interviewer. At no point during the study were any theories discredited or supported (Shkedi, 2006).

Given the evident scarcity of research addressing the issue of hope in ALS patients and the disagreements encountered in the few studies that were carried out, this article proposed to gain further understanding as experienced by individuals with ALS and to encourage healthcare professionals to provide interventions from a psychosocial perspective and become the individual's advocate in promoting advanced care planning and the practice of patient-centred care (Hamama-Raz, 2021). General thematic data analysis identified core themes from
data collected during the interviews. Analysis of the content retrieved was conducted in three stages. During the initial stage, the authors repeatedly read interviews to form closeness and emotional intimacy with the participants (Kvale, 2009). During the first stage, it clearly emerged that the issue of hope was fundamental for the participants in order to deal with ALS. The second stage consisted of gathering and identifying the parts of the interview that directly included the participants' meaning of hope (Holloway, 2007). Special attention was given to repeated utterances which led to 'units of meaning' (Schreier, 2013). Furthermore, it was possible to identify, directly from the interviewees' words, how hope impacted the existential orientation of their future (Lincoln, 1985). An attempt by the authors to segregate any expressions that were dissimilar from hope and to integrate those that were connected to hope was carried out during the end of this second stage (Lincoln, 1985).

The third stage included the collection of 'units of meaning' to understand what hope meant to individuals diagnosed with ALS. By focusing, comparing and synthesising the action of data, the researchers formed themes that led to self-explanatory relationships of the participants towards themselves, with the hope being the dominant existential approach to coping with ALS (Van Manen, 2001). Two different themes emerged from this study which addressed the paradox of hope in people with ALS during the interviews. The first theme or voice was the description of hope as an obstacle. In contrast, the second theme described hope as a resource, and several participants compared it to a type of inner strength that allows for a continuation of spirituality and practicality within their life. Therefore, the interviews in this study were enriched with metaphors describing the two themes or voices of hope. Such metaphors include comparing hope with the strength to swim towards the hand that pulls one out from drowning, the light at the end of the tunnel, a mystical force, e.g., The idea that hope was viewed as an obstacle was not something the researchers

were expecting since hope is always perceived as the counteracting agent to despair, apathy, depression and any negative emotion (Rahimipour, 2015). Furthermore, the results of this study contradicted previous research that was carried out on other individuals with life-threatening diseases such as metastatic cancer and heart failure since hope in such situations revealed beneficial effects on quality of life and psychological and physical wellbeing (Farhadi, 2014). The researchers' possible reason for this surprising result may be due to the uniqueness of ALS. In other words, ALS has no particular stages; it is highly progressive with no possibility of remission and has devastating effects on different body areas with time. Therefore, the positive definition of hope may not be readily acceptable in the case of ALS patients.

The results of this study led to the theme of hope having a double-sided meaning. Recommendations for this small sampled study include inviting healthcare professionals to put aside any judgment they may harbour, given the patient's beliefs on hope. Moreover, healthcare providers should try and understand what hope means to people with ALS and proceed accordingly. For instance, if hope is viewed as an obstacle, the focus should be on guiding the individual towards empowerment and achieving realistic goals. On the other hand, if the individual's view of hope is that of a resource, then the healthcare provider should promote spirituality and help the person find meaning and purpose even in unrealistic circumstances.

Several limitations were noted during this study. First, the study was carried out on Jewish Israelites. Therefore, their culture may have introduced bias since the meaning of hope may not reflect well in their case. Additionally, the participants of this study were recruited via a support organisation. Therefore, caution should be taken regarding the results since other individuals with ALS did not belong to any organisation. Another limitation was the small study sample, whereby the findings should only be considered as a basis for further studies. The third limitation was the difference in the stage of the disease since findings may vary between individuals within 12 months of diagnosis and those requiring assistive communication.

The researchers suggested that any further studies should explore the meaning of hope from the perspective of other religions, cultures and terminal diseases. In summary, this qualitative review offered valuable insights into the double meaning of hope in individuals diagnosed with ALS.

2.5.2: The Illness Experience

This dissertation focuses on the lived experiences of individuals diagnosed with ALS; thus, articles discussing life journeys or experiences were chosen during the literature search. However, even after an in-depth search for such articles, more literature was needed. This confirmed that articles discussing lived experiences of individuals with ALS were needed.

In this phenomenological qualitative study by Yuan (2021), a thorough understanding of the experience of living with MND in Chinese individuals and the meaning behind their statements was carried out. Although many people living with ALS exist in China, very little is known about their illness journey and how they cope with such a devastating condition. Therefore, the aim and objectives of this research paper were specific and investigated the illness experience in a direct and organised manner.

A diagnosis of ALS is daunting, both on the individual per se and family members. In addition, it bears the burden of existential and biopsychosocial issues (Ozanne, 2013). This article addresses the challenges and hardships Chinese patients with ALS and their families face due to financial, social and economic issues (Gong, 2016). According to Stephens (2016), multidisciplinary clinics are the standard care for ALS in Western countries; however, in China, these are sparse, thus leading to limitations when caring for and treating ALS patients. A cross-cultural study revealed that cultural traditions significantly influenced attitudes towards death and decisions regarding therapy and life support (Andersen, 2018). The Chinese population harbours a philosophy and heritage that is unique to other countries, thus, the way ALS impacts their emotional and physical status differs (Thomas, 2018). This article, therefore, addresses the lived experience of Chinese patients from a diverse cultural perspective.

ALS is an incurable disease which progresses rapidly. Thus, treatment has to focus on improving the person's QOL (Prell, 2019). Psychological wellbeing, burden, anxiety, hopelessness and fear were all issues mentioned in this article. The majority of articles that addressed the psychological wellbeing of individuals with ALS were quantitative. However, these studies proved limiting since they were not designed to deal with ALS (Mistry, 2013). As previously stated, qualitative data on ALS is scarce; however, it has been recommended as the most appropriate design to assess mental health concerns in patients with ALS (Thomas, 2018). This article proved sufficient in

addressing the paucity of research on ALS that exists and persistently commented on the importance of tackling the lived experience from diverse cultural aspects.

This research study consisted of in-depth semi-structured interviews, and 24 individuals were invited to participate. Four of the invitees declined, leaving 20 individuals willing to participate. The participants were recruited from the Neurology Department in Wuhan, China. The recruitment period of this small participant sample, from February to July 2018, was relatively short. A purposive sampling method was carried out to ensure that sample diversity was accomplished. Determination of sample size was conducted after the saturation of data, and two more individuals were interviewed. If, during the interview, no new issues arose, then the recruitment would close. Individuals with respiratory insufficiency or verbal communication were not allowed to participate. All participants were asked if they could explain their experience after receiving a diagnosis of ALS, and each individual received a small remuneration. The interviews were audio-recorded with the participant's consent, and transcriptions were done within 24 hours. Interviews were held in the Chinese language but were later translated into English. Colaizzi's method was carried out to analyse data using seven steps: reading the transcript, identifying significant phrases and meanings, grouping themes, description, describing the phenomenon, and allowing the participants to verify what was discussed during the interview. As a guideline, the Consolidated Criteria for Reporting Qualitative Research (COREQ) was used (Wirihana, 2018). Ethical consideration was taken throughout the study.

The age of the 20 participants varied between 28-72 years, and during this phenomenological qualitative study, three themes and eight sub-themes came up. First, the participant's information and characteristics were gathered and placed in a table for ease of reference. A code was used to ensure anonymity. Two subthemes, namely 'body out of control' and 'inward suffering', emerged. The individuals recounted how being diagnosed with ALS led them to lose control of their bodies, which affected them psychologically and physically.

In the sub-theme inward suffering', denial of having ALS emerged consistently. One patient who had never heard of ALS could not understand why he had the disease and hoped he was misdiagnosed. On the other hand, the impact of solid cultural views appeared when one participant stated that he had been raised believing that 'evil has

its retribution', but he cannot fathom out what he had done wrong. Previous research yielded that 'personal faith' helps the individual fight the feeling of despair. Thus, healthcare professionals should be attentive not only to the patient's medical needs but also to a spiritual perspective (O'Brien, 2015). Words like 'fear', 'desperate' and 'hopeless' were frequently uttered. A state of ambivalence was also noted among the participants, who desired to be cured but still had suicidal ideations on the progression of the illness.

The family self-help theme emerged during all the participant's interviews, which the researchers stated may indicate an inadequacy in the 'Chinese Medical Insurance System' and a lack of medical facilities for individuals with ALS. Although a strong sense of cultural familism was noted among Chinese families, limitations in this family self-help model arose. These included more communication between the patient and caregiver, self-perceived burden and a lack of professional care knowledge (Ando, 2019).

In this study by Yuan (2021), the author noted how Chinese traditional cultures deeply impacted the participants' perspectives. For example, the interviewee's age significantly impacted their wellbeing. It is a custom in China for children to care for their elderly parents; thus, these participants did not feel burdened. Contrarily, the younger patients who had to be cared for by their elderly parents experienced more guilt because, to them, this was not in line with Chinese social tradition (Chen, 2016). According to Lin (2003), another Chinese cultural phenomenon is the avoidance of speaking about death since the Chinese belief in life and death is affected by 'Confucianism', which consists of offering life the utmost importance. The interviewees rarely mentioned death in this study; hence Confucianism was verified. Another recommendation of this qualitative study was for healthcare providers to offer and strengthen education on death in Chinese ALS patients. Their hesitation in talking about death led to an insufficient understanding of the significance of a 'good death'. The article went in-depth to help the reader understand the Chinese cultural system.

The study had several limitations. First of all, the participants were only recruited from Wuhan. Thus, one would need to discover how individuals from other regions may express their experiences of being diagnosed with ALS. Secondly, participants were only interviewed once, which meant that their experience could not be followed over some time. Furthermore, additional research from various regions is required to gain

more insight into the lived experiences of individuals with ALS. Finally, another limitation was recorded: they may have influenced some of the participants their caregivers accompanied during their interview.

2.5.3: Psychological Interventions In ALS

A dearth of studies exists when it comes to investigating psychological interventions for individuals diagnosed with ALS. In a qualitative analysis study by Marconi et al. (2016), the effect of meditation on people with ALS and their caregivers was explored. In addition, the meditation training program was adjusted for ALS needs. According to Pagnini (2013), being diagnosed with ALS may lead to distressing psychological issues, which, in turn, may exacerbate the individual's QOL. Furthermore, caring for a family member with ALS is taxing since the challenges and stressful situations are continuous (Burke, 2015).

For this study, 26 individuals with ALS and 18 caregivers were recruited. However, the recruitment proved challenging since the researchers encountered difficulties due to psychological resistance towards the meditation program and logistical challenges involved in session attendance. The intervention was carried out at the Neuromuscular Omnicentre in Milan, Italy. The participants of the study were recruited from the same centre.

Meditation sessions were tweaked due to the physical limitations of ALS, and an emphasis on the cognitive and affective aspects was addressed instead. Certain exercises, such as 'Feldenkrais' that were retrieved from other approaches similar to meditation were included in the meditation sessions. The sessions emphasised accepting the physical limitations consistent with such a neurodegenerative disorder whilst honing in on resources and ability. Due to the challenging aspects of ALS, two trainers were involved in the sessions (Pagnini, 2014). The training protocol 'Mindfulness-based Stress Reduction' was applied (Kabat-Zinn, 1990). Sessions were held once a week and lasted eight weeks. A semi-structured interview was carried out with both participants and caregivers post-training. The interviews were audio-recorded and transcribed literally. A grounded theory approach was used to analyse data (Strauss, 1990). Any data collected was codified and divided into two macro-areas: resources and limitations. The aim of the study was brief but concise and clear throughout. Demographic information of participants and identified domains were

listed in table form, which allowed for ease of reference. Furthermore, the domains were listed under sub-titles, making it easier for the reader to identify each. Eight domains were extracted from the in-depth interviews.

For being an exploratory qualitative analysis, this study appeared promising and equivalent to studies that addressed other chronic neurogenerative disorders such as multiple sclerosis (Grossman, 2010). Consequently, this study is the first to report that meditation training could benefit individuals diagnosed with ALS and their caregivers. Two frequent issues encountered in ALS patients are anxiety and depression; however, after attending meditation training sessions, coping improvements were noted. At present, one of the aims of ALS research is to improve QOL (Simmons, 2005). Participants understood the value of the new skills taught, and many continued attending the sessions. They also recommended informing other individuals with ALS about such techniques.

The study results show that meditation training in individuals with ALS and their caregivers has vastly improved their QOL, sleep patterns, breathing and relationships within the family. Such results show the necessity of more studies on meditation and neurodegenerative disorders (Simpson, 2015).

Regardless of the evident positive results of this qualitative analysis, certain limitations were noted, leading to a hindrance in the clinical usability of the protocol per se. Logistical and practical issues limit any psychological resistance towards meditation and may prevent extended access. ALS is progressive and fatal. Thus, more studies and data on QOL and its influence on training are required to confirm or oppose the preliminary data results on the advantageous effects of meditation on ALS. No conflict of interest was reported in this study.

2.5.4: Psychological Wellbeing and QOL In ALS

A similar study to the one discussed on meditation above, which addresses the impact of mindfulness on the QOL and psychological wellbeing in people diagnosed with ALS, was carried out by Pagnini (2015) and his team of researchers. According to Keng (2011), studies on ALS have yet to be conducted. The researchers recruited 197 individuals with ALS from Departments in Psychology in Milan, Italy and Harvard

University, Cambridge, USA, respectively. Participants were assessed twice online, and four months were allowed before the second assessment. Data collection was in the form of an online survey that was designed with Qualtrics survey software, and the participants filled out questionnaires on demographics, how the illness was progressing and diverse physical and psychological consequences. Evaluations on each individual's physical impairment, QOL, anxiety, depression and 'trait mindfulness' were carried out. The inclusion criteria consisted only of a self-reported diagnosis of ALS. The effect of mindfulness on physical disability was carried out using the 'mixed-effects model'.

Since ALS is conventionally known as a 'mysterious' illness in terms of its pathogenesis and mechanisms and is considered genetically and biologically driven (Mitchell, 2007), this was challenged in the face of the researchers' central hypothesis. Given this, an exploratory longitudinal study was carried out to explore whether mindfulness may influence the progression of ALS using a 'mind-body framework'. However, regardless of a bid to take on a more biopsychosocial approach (Engel, 1977), which has sufficient evidence to prove that both concepts are correlated, medical science remains the dominating factor (Fava, 2010).

Mindfulness, an ancient Buddhist practice, is actively allowing oneself to be 'engaged' in the here and now rather than ruminating about past recollections (Langer, 2000). For this study, the Langer Mindfulness Scale (LMS), which consists of a 14-item questionnaire, was used. Three areas of mindfulness thinking were explored: novelty seeking, novelty producing and engagement. Various scales were used to carry out this study. Apart from the LMS, QOL was assessed using the McGill Quality of Life single-item scale (MQOL-SIS) and the hospital Anxiety and depression scale (HADS) by Zigmond (1983). Anxiety was rated using the HADS-A subscale, whereas, for depression, the HADS-D subscale was utilised. Such scales are formulated to detect the presence of anxiety and/or depression in medical patients (Snaith, 1983).

Furthermore, the self-administered ALS functional rating scale (SA-ALSFRS), which monitors disease progression in ALS, proved excellent for individuals with a physical impairment to be assessed online (Maier, 2012). In cases where multiple comparisons are required, as in this study, the application scores of the mixed-effects models competently addressed possible confounding effects and were preferred to the

Bonferroni method (Gelman, 2012). The study explained in concise but clear detail both the results and in which context the scales were used.

Out of the 197 participants, 102 went on to complete the second assessment after four months after the initial one. The mean age was 58, and fewer females participated. For ease of reference, data was placed in uncomplicated tables and graphs. At the start of the trial, scores assessing SA-ALSFRS showed a decrease over time; however, with the commencement of mindfulness, the trend was notably reduced. Additionally, a decrease in anxiety and depression symptoms was reduced, and a positive influence on QOL was recorded. Those individuals who scored high on LMS reported high mindfulness, unlike those with lower LMS scores.

This study had two aims. One was to see whether mindfulness influences ALS, and the second was to explore whether mindfulness affects QOL, depression and anxiety. The hypotheses corroborated the observations carried out by the researchers. Results of the study showed that individuals who practised mindfulness reported less physical impairment and minimal depression and anxiety symptoms. Furthermore, according to Carson (2006), individuals who practised mindfulness were better equipped to handle stress.

Recommendations of this study included utilising the results for more research and ensuring that healthcare professionals are aware of mindfulness's profound positive effect on individuals with ALS. The results clearly show that practising mindfulness may decelerate the progression of ALS, thus improving their psychological wellbeing. On the other hand, another recommendation that should have been investigated in this study was offering healthcare providers mindfulness sessions. This may change the trajectory of their thoughts about this fatal neurodegenerative disorder. The need for further studies on mindfulness and ALS are needed. Certain limitations were noted in this study. Data was self-reported by the participants, and since the mindfulness program was online, only a part of the ALS community was reached. In their search for participants for this study, the researchers were assisted by the National ALS Registry, which helped them reach an extensive sample. However, one must take into consideration that ALS is not a prevalent disorder. According to Nakamura (2012), the sample population was self-selected. Thus, recommendations included using small and random tests to support the results of this study further. To conclude, the data collected in this study revealed how psychologically constructed mindfulness can alter

the trajectory of ALS. Given the strong point of view on the genetic and biological perspective of the disorder, this longitudinal study sheds hope on the mind-body relationship framework. However, this must be strengthened by further studies since there is a definite paucity of literature that discusses psychological interventions in ALS.

2.5.5: Self-Perceived Burden In ALS

This cross-sectional study by Geng (2016) and his team of researchers from West China Hospital of Sichuan University addressed the self-perceived burden (SPB) in Chinese individuals diagnosed with ALS and how it affects their QOL and that of their caregivers. In addition, a clear explanation was given of the study's design.
The cause of ALS remains uncertain, and neurologists can only offer palliative treatment to lengthen life and improve QOL in their patients (Hwang, 2014). However, according to several studies, a decline in QOL in a cohort of ALS patients was noted, possibly caused due to depression and physical impairment (Korner, 2015). Furthermore, 'oropharyngeal dysphagia' (Paris, 2013), weight loss (Korner, 2013) and pain (Pizzimenti, 2013) all contributed to a decline in QOL.

Despite its rarity, 81 participants were recruited for this study between March 2015-September 2015. To be able to participate in this study, the participants had to be diagnosed with either probable or definite ALS, according to the 'revised criteria of El Escorial Scale'. An in-depth explanation of the differences was given to further guide the reader. Participants with any other neurological disorders or who presented with a diagnosis of severe psychiatric illnesses were excluded. Furthermore, caregivers who were paid for their services were also excluded from participating. Data were collected via face-to-face interviews at the Department of Neurology, and both carers and patients were interviewed apart and had to fill in self-rating questionnaires. To ensure that age did not influence the SPB or burden in the case of the caregiver, the researchers divided the 81 participants into two groups, namely, less than 45 years of age and over 45 (Yuan, 1997). These groups were also classified as 'young onset' and 'late onset' (Turner, 2012). The ALSFRS-R scale was used to gauge the severity of ALS.

Again the researchers explained the factors of ALSFRS-R in great detail, making it easier for the reader to follow. The ALSFRS-R scores were placed into three stages to assist in the progress of findings, mild, moderate and severe (Kimura, 2006). The QOL of each participant was calculated by the World Health Organisation Quality of Life-Bref (WHOQOL-BREF) questionnaire; however, for this study, the Chinese version was used (Hao, 2006). The SPB of the patients was evaluated with the Self-Perceived Burden Scale (SPBS). For the burden experienced by the caregiver, the Zarit-Burden Interview Scale (ZBI) was utilised (Zarit, 1980). Both of the latter scales were of the Chinese version. To analyse the statistical data, SPSS version 19.0 was used. Ethical considerations were followed throughout.

The characteristics of both the patients and their caregivers were listed in table form for clarification and ease of reference. The researchers then gathered the results of SPB, ZBI and QOL under separate sub-headings, making it easier for the reader to comprehend. In the case of SPB, results showed the majority of patients (n=27) had low/no SPB, 25 had mil-moderate SPB, 18 had moderate-severe and 11 revealed severe SPB. Interestingly, female patients showed a higher incidence of SPB than their male counterparts, and those with a higher academic status also showed higher SPB. The patients still in the mild stage of physical impairment showed less SPB than those in an advanced stage. To conclude, the patients more knowledgeable about ALS bore higher SPB scores than those who did not. The ZBI score results indicated that most caregivers experienced mild-moderate burden (n=37). This was followed closely by those who showed little or no burden in caring for their family member (n=29). Only two caregivers experienced severe burdens. The caregivers who reported high ZBI scores had to care for patients with low ALSFRS-R rates, pay for medical expenses themselves, and have patients totally or partially dependent on them.
The same can be said when assessing the results of QOL in patients. Again, those with high ALSFRS-R scores had a better QOL than their lower counterparts. Again, this was explicitly noted in the psychological and physical domains.

An interesting finding emerged from this cross-sectional study. Female participants had significantly higher SPB scores than their male counterparts. This result did not tally with two other studies from Western countries (Gauthier, 2007), shedding light on the fact that this may be due to the Chinese culture, whereby women are expected

to care for the majority of the household, including their in-laws and children under the 'Confucian concept of ethics' (Hashizume, 2000). Therefore, a recommendation for this study was for healthcare providers in China to pay particular attention when dealing with a diagnosis of ALS in a female patient (McCluskey, 2004). Another recommendation was to ensure medical insurance covers patients with ALS in China since financial worries were one of the reasons that SPB and QOL decreased in both patients and caregivers.

Several limitations were noted whilst conducting this study:

1. Selection bias may have been introduced since all participants were from one ALS centre, and those confined to their homes were not chosen.
2. A definite explanation cannot be concluded due to the cross-sectional study design.
3. The sample size was considered small since other subsets were investigated.

However, it was large enough for a statistical analysis to be carried out. To help the researchers verify this study, more longitudinal studies from multiple centres and larger population samples are required.

2.5.6: ALS From a Biopsychosocial And Spiritual Perspective

This qualitative, mixed-method study by Kukulka et al. (2019) aimed to explore the lived experience and needs of individuals with ALS and their families from a biopsychosocial and spiritual model of illness perspective. Another aim was to guide palliative care teams on how to knowingly treat patients with ALS, taking this model of illness into perspective.

A 'stratified purpose sampling' method was used to recruit patients, caregivers and healthcare providers (Creswell, 2013). Participants were associated with a single ALS association-certified clinic in the Midwestern United States and had to be over 18 years of age and, based on El Escorial criteria, must be diagnosed with either definite or probable ALS to be eligible for the study (Ludolph, 2015). Participants were subjected to a semi-structured interview whereby biopsychosocial and spiritual aspects of their lived experience since being diagnosed with ALS were discussed. Additionally, participants had to sit for a 40-item ALS Assessment Questionnaire known as ALSAQ-40 (Jenkinson, 2000). This questionnaire enabled the researchers to assess the participants' wellbeing. To analyse data, the Dedoose version 8.0.42 was used.

The results were presented clearly and concisely in three sections: the demographics, the quantitative results of the ALSAQ-40 questionnaire, and the themes that emerged from the qualitative part of the study. The sample population consisted of 14 individuals, seven men and seven women, in various stages of ALS and sixteen caregivers. The participants all resided in private residences in the community. For ease of reference, the researchers divided the qualitative results into two categories: biopsychosocial needs and information regarding spirituality and emotional aspects of the participants' lives. The results clearly showed that ALS is psychologically and physically daunting, and as it progresses, the individual tends to experience a loss of self-esteem and self-worth, loss of autonomy and despondency. Participants were asked about their feelings towards spirituality and their life meaning. The majority (n=9) stated that their diagnosis of ALS had strengthened their spirituality, whereas only two mentioned spirituality in a negative manner. Negative spirituality was explained as anger towards God, and positive spirituality included the more frequent attendance of religious services and an increase in prayer and self-reflection. To conclude, studies show that personal spirituality gives hope and reduces feelings of despair in individuals with ALS (O'Brien, 2015). On the other hand, 'religiousness' had a positive effect on caregiver QOL (Calvo, 2011).

This study's findings revealed various ways in which palliative care teams can support patients with ALS and their caregivers. Furthermore, another finding revealed the importance of early palliative care intervention in ALS, given the rapid and progressive degeneration of the disease.

The study revealed various strengths and limitations. Using a mixed-method design was one of the strengths since it combined the investigation of in-depth interviews with the measurement of patients' wellbeing and QOL using the quantitative aspect. Another strength was the introduction of different 'stakeholders', which led to a myriad of different views. Furthermore, as mentioned in other studies above, limitations included data collection from only one area whereby the Caucasian race was prominent. This could lead to generalisation in findings. Another limitation was the possibility that the participants may have replied to questions in a way deemed favourable by the research team. To conclude, it would prove beneficial if any future studies expanded their sample size and recruited from multiple clinics, thus allowing for a broader spectrum of perspectives.

2.5.7: A Journey of Suffering

An ethnography by Oh et al. (2014) addressed the suffering individuals with ALS go through in the socio-cultural context of South Korea, whereby the meaning of culture and the experience of illness was explored. The criteria for recruitment were a diagnosis of ALS as confirmed by a physician, had to be registered with the Korean Amyotrophic Lateral Sclerosis Association (KALSA), participant being able to communicate verbally or non-verbally, being over 18 years of age and being able to understand the reason for the study and must reside in Seoul in order to facilitate participant observation. Exclusion criteria included neurological conditions other than ALS or severe mental illness like schizophrenia. Data collection was in photo-elicitation, whereby the participants were asked to take photographs of their experienced with ALS. Consent forms were distributed and signed, and in the case of participants with difficulties in writing, an oral consent form was utilised. The participants' demographic data were gathered into tables for ease of reference and clarity, and the article, which was divided into subheadings, made it easier for the reader to follow each section. The meta-theme of this article was 'A Journey of Suffering', which then was divided into three themes: off the course, drifting and on a new boat. These themes offered insight into the patient's experience.

The strengths and recommendations of this article were not listed in a separate section but included in the conclusion, which was rather misleading for the reader. Limitations of the study were not included. Final results revealed the harrowing experience of individuals diagnosed with ALS and the emotional, physical and social toll it takes on them. Furthermore, the findings indicate implications for clinical practice and offer an invaluable source for healthcare professionals to develop more insight into the meaning of living with ALS. Recommendations of the study revealed a need for more programs and services to reduce the suffering ALS imposes on individuals. Moreover, the programs should emphasise psychological support and counselling to further help with the progress of the disease. ALS is incurable; therefore, therapeutic management is more so than medical treatment is required.

2.6: Conclusion

ALS is a fatal neurodegenerative disease involving both the brain's and spinal cord's upper and lower motor neurons. In a study by Geng (2016), ALS was described as an 'orphan disease due to its rarity. Thus, a dearth of literature was found across all situations in an individual diagnosed with ALS. The only abundant literature encountered was from a molecular and genetic perspective. The author of this dissertation attempted to group the studies she found relevant into themes, namely hope, burden, illness experience and psychological interventions that could be practised on both the patient and the caregiver to alleviate the physical and psychological toll ALS takes on these individuals. ALS was also viewed from a cultural perspective whereby one can take note of differences between, say, an Eastern culture and a Western one. All studies recommended further research on this incurable, neurodegenerative and fatal disease.

Chapter 3: Methodology

3.1: Introduction

This chapter aims to present the research question and provide the reader with an in-depth description of its methodology. The researcher opted for an IPA (Smith, 2009) as her chosen methodology for this research study since it is particularly sought after when dealing with complex and 'emotionally laden' topics. The heterogeneity of ALS is an ideal example of why this phenomenon should be adapted since it involves complex biopsychosocial aspects that are challenging to clinical and personal decisions (Pfohl, 2018). Furthermore, an IPA is a qualitative approach whereby an individual's lived experience can be intricately examined on its terms without relying on previous theoretical assumptions (Osborn, 2015).

Additionally, choosing IPA as a methodology for this study is advantageous since it allows the participant to describe their experience of being diagnosed with ALS. For this chapter, a description of data collection methods and data analysis will be included, together with a discussion of the ethical and methodological considerations of the study.

3.2: Aims, Objectives, and Research Question

In Chapter 1, the author described, in detail, the goal of this study which was the collection of data via a series of semi-structured interviews on the lived experiences of individuals diagnosed with ALS, both the sporadic and genetic types. Before collecting data, the researcher critically appraised the articles retrieved during the literature review. The research question for this dissertation was what is the lived experience of individuals diagnosed with ALS using an interpretative phenomenological approach? A list of the main objectives set by the researcher is listed in Table 3.1 below:

Table 3. 1: List of main objectives

➢ To understand, from a patient's perspective, the impact of living with the debilitating and terminal motor neurone disease of ALS.
➢ To explore how individuals strive to cope with living with ALS.

3.3: The Epistemological Position

The author chose an IPA approach as the most appropriate methodology for this study since this dissertation addresses the lived experiences of persons dealing with a fatal disease. According to Flanagan (2013), scientific methodology is considered the most effective tool for exploring new theories and carrying out empirical validation. As such, quantitative methodology's deductive process and hypothetically-driven nature are more effective when dealing with larger population samples where data collection can be quantifiable (Murray, 1998). Contrarily, a qualitative approach, which is inductive, does not focus on objectivity, statistical procedures and measuring of variables (Martin, 2021). In turn, it provides an in-depth comprehension of complex realities that cannot be quantifiable, allowing the researcher to be subjective whilst being able to correspond on a deeper level with the individual's life experience and focus on the dynamics at hand (Maxwell, 2013).

Furthermore, qualitative research yields abundant information, often in a setting void of manipulation. Furthermore, exploring new phenomena captures the person's innermost sentiments and thoughts by gaining insight into how these individuals view and experience the world (Given, 2008). Additionally, qualitative literature contributes to a rich, person-centred, profound data source that will not be perceived in a quantitative design (Merriam, 2009).

It may prove beneficial when using a qualitative design to inspect philosophical views, especially from an epistemological perspective, since knowledge is classified by purpose and design (Tennis, 2008). When taking on an epistemological perspective, it is pertinent for the researcher to become as familiar as possible with the participant, thus, allowing for knowledge to be known and acquired based on the individuals' personal views (Creswell, 2013). Furthermore, according to Creswell (2013), carrying out the research where the participants reside, and work leads to a higher understanding of what the person is narrating; additionally, the more time the researcher spends with the participant, the more they 'know what they know'. Guba (1988) explains that when an epistemological approach is used, an attempt to reduce the 'distance' or 'objective separateness' between the researcher and the participant undergoing the study is seen. Creswell (2014) defines the term epistemology as the study of how we know what we know about the reality we are researching and any methods used to understand this reality.

Due to this research study's sensitive and qualitative nature, the author opted for an interpretive/constructivist epistemological position that she felt enhanced her study and strengthened her bonding relationship with the participants involved. This paradigm emerged from the philosophy of Husserl's phenomenology, defined as hermeneutics, which is a study of interpretative understanding (Mertens, 2005). Taking on an interpretive/constructivist approach allows the researcher to understand 'the world of human experience' (Cohen, 1994), whilst Mertens (2005), on the other hand, states that 'reality is socially constructed'. According to Creswell (2003), such a researcher will define the findings by relying solely on the participant's lived experience whilst being aware of their research's impact on their background and experiences. Therefore, The data collection method for this study will give the author direct access to the participants' experience, thus allowing for an accurate representation of their narratives. According to Willig (2001), such an approach allows for various interpretations that may be applied to establish different versions of the experience.

3.4: Ration For Choosing IPA

IPA can be defined as a collective set of factors that ultimately lead to diverse perceptions of the individual's reality and a commitment to understanding their lived experience (Smith, 2009). This research study focused on the lived experience of individuals diagnosed with ALS.

The author's focal point for opting for an IPA as a rationale is concentrated on three main perspectives. First, as stated previously, adopting a qualitative research methodology enhances the researcher's exploratory and investigative nature of their studies while allowing them to exercise their interpersonal and subjective skills (Alase, 2017). Considered highly 'participant oriented', the mode of data collection for an IPA may include semi-structured interviews and focus groups whereby the participants can feel free to express themselves and their lived experience without fear of misrepresentation (Alase, 2017). The author chose semi-structured interviews for her data collection method since listening to the point of view of the individuals rather than simply observing would enrich and strengthen the study's findings. According to Bryman (2007), a possible disadvantage of this data collection method could be that

the participants may feel apprehensive and feel that an interview at such depth will invade their privacy.

Secondly, according to Lopez (2004), to apprehend the research participants' lived experiences on a deeper level, the researcher has to erase any previous knowledge on the topic of interest during data collection. However, the researcher is a psychiatric nurse and has never been exposed to or had the opportunity to work with individuals diagnosed with ALS. Therefore, before embarking on this study, she had first to become well-versed in MND with a particular emphasis on ALS and its psychological manifestations. In such instances, projecting a positive impression is required since it may transpire that the participants may consciously or subconsciously challenge the interview topic or its significance (Zuckerman, 1972).

Lastly, the type of methodology found in an IPA enables the researcher to perceive the participants' feedback on how they dealt with the diagnosis of ALS in an unrestricted and comprehensive manner whilst coming to terms with a physically incapacitating terminal illness. Furthermore, two major roles may be addressed when conducting interviews as a data collection method, namely exploration (Blumer, 1998) and by including the construction of a theory (Piore, 2006). In this research study, the researcher explores a field she is not overly familiar with. Thus, conducting semi-structured interviews may proffer situations whereby results may be both expected and unexpected (Piore, 2006). According to Blumer (1998), semi-structured and open-ended questions are a 'flexible pursuit of intimate contact with what is going on'.

3.5: Theoretical Underpinnings of IPA

Smith (2009) describes three theoretical underpinnings forming IPA: phenomenology, hermeneutics and idiography.

3.5.1: Phenomenology

Phenomenology can be defined as an approach from a philosophical and interpretive perspective which leads to the study of lived experiences (Parahoo, 1997). Furthermore, Thorne (1991) argues that from a phenomenological stance, the most fundamental human truth can only be accessed through inner subjectivity.

Husserl, a phenomenological philosopher, sought ways whereby an individual may correctly understand their lived experience of a particular phenomenon whilst identifying vital qualities of the said experience (Smith, 2009). Moreover, researchers using an IPA approach may produce a narrative of the interviewees' experiences free from bias and is a concept defined as phenomenological reduction (Koch, 1995). From a philosophical perspective, the phenomenological reduction is called 'bracketing'. According to Moustakas (1994), bracketing is a term used whereby an individual avoids passing judgement. However, when applied in practice, Creswell (2003) states that bracketing is a process where the researcher can put aside his/her preconceptions and concentrate on the participants' lived experiences. However, Dowling (2007) interpreted Husserl's definition of bracketing as 'completely abstaining' from any form of judgement during the research process. On the other hand, according to Heidegger (1926), holding back judgement could not be fully accomplished since human nature in itself is made up of
'unconscious subjectivity'. Creswell (1998) argues that because of this criticism, numerous researchers believe that using bracketing as a tool is an important part of the phenomenological research approach.

During the data collection phase of research, researchers are expected to have experience and knowledge about their field of interest. However, it will prove helpful if the researchers remove any assumptions they may be experiencing at that particular moment (Smith, 2009). Furthermore, to ensure a deeper understanding of the participant's responses to the interview questions and to fully address any 'personal assumptions and goals', the author opted to keep a self-reflective journal throughout the research process, as recommended by Smith (2009). This journal aims to note the details of what the researcher felt and thought whilst examining the data collected and the rationale behind them. Reflective journals are becoming an increasingly important tool when applying qualitative research (Kelly, 2002). Additionally, Jasper (2005) states that keeping a reflective diary leads to a valid, transparent audit trail and enhances the researcher's critical thinking by developing conceptual skills.

3.5.2: Hermeneutics

Defined as the 'art of interpretation' (Abulad, 2007), hermeneutics is IPA's second major philosophical underpinning (Smith, 2009). Whilst going beyond the

participant's lived experience, an IPA approach aims to interpret the innermost and most profound meaning of the participants' narrative (Shinebourne, 2011). Furthermore, Guba (1985) argues that when the researcher uses a reflective journal to record his/her research process, the rigour with which the research is carried out will add to the study's trustworthiness. Additionally, reflective journalling helps the researcher provide an audit trail, thus, allowing for more transparency (Jasper, 2005).

Lopez (2004) explains that to arrive at the meaning, a process which involves both the researcher and the participant is required. The study's results do not solely rely on the researcher's interpretation of facts. Thus, a dual interpretation takes place. At the beginning of the interview, the participant will explain his/her lived experience. At this point, the researcher may intervene by asking more probing questions to help the participant elaborate further on his/her experiences. Finally, the researcher will offer his/her interpretation of the participant's account. Schleiermacher (1998) describes this type of dual interpretation as 'double hermeneutics' and states that it is more an art coupled with intuition rather than a rigid set of rules. Schleiermacher (1998) goes on to explain that an important aspect of the process of interpretation is not only to focus on the text but also to find the time to understand the individual writing the text. In other words, if a thorough and complete analysis has been conducted, the researcher may realize that he has acquired 'an understanding of the utterer better than he understands himself'.

The circular process of questioning the participant, retrieving meaning and questioning further is called the 'hermeneutic circle' (as cited in Smith et al., 2009). To conclude, Larkin (2006) states that this process will lead to an in-depth analytical review of the researched topic.

3.5.3: Idiography

The third major underpinning encountered whilst carrying out an IPA is idiography (Smith, 2009). Explained by Allport (1937) as a 'novel and somewhat daring' research method, idiography takes into account the intricacy and perplexity of how an individual thinks, perceives and behaves. Defined as a profound and comprehensive analysis of a single individual or case study, idiography emphasizes an individual's unique traits and behaviour whilst focusing on their lived experience (Larkin, 2006).

During an idiographic approach, the researcher concentrates on one participant at a time whilst investigating the case in a profoundly detailed, personal manner via a delicately conducted interview (Smith, 2009). Once all participants have been interviewed, the researcher will then cross-examine and compare the results of each interview in search of any similarities or differences across the accounts of all participants. When using an idiographic approach, the researcher will then be able to observe patterns emerging across the cases being analyzed (Smith, 2009). To conclude, an idiographic approach in IPA reveals 'within-person' patterns based solely on each person's uniqueness (Connor, 2009).

3.6: The Sample Selection

When conducting quantitative research, participants must be randomly selected to ensure that any potential influence on variables is removed (Creswell, 2009). In contrast, however, it is pertinent to select individuals who demonstrate knowledge and insight into the topic being researched (Kuper, 2008). Therefore, an important aspect to consider before embarking on a qualitative approach is the identification of appropriate candidates. This is brought about by carefully revising the research question and theoretical perspectives (Sargeant, 2012).

For this research study, the author opted for a purposive sampling technique, whereby participants are selected due to the characteristics or traits required. This particular sampling method, thus, is reliant on the researcher's acumen since it is crucial that the participants chosen can proffer information that is in-depth, meaningful and focused on the phenomenon being researched (Shinebourne, 2011).

The following section focuses on the sample eligibility criteria of the candidates chosen for this study.

3.6.1: The Sample Eligibility Criteria

This research study focused on the lived experience of individuals diagnosed with ALS, both sporadic and familial. Candidates chosen had to be male or female, and the age bracket of participants was 18 years and over. Given the extremely rare form of ALS known as Juvenile amyotrophic Lateral Sclerosis (JALS), children were not considered for this study. According to the Genetic and Rare Diseases Information

Centre (GARD), JALS is distinguished by specific variations in several genes. Before deciding on the sample eligibility criteria, the author researched JALS in Malta. According to Prof. Ruben J. Cauchi, the head of the University of Malta's (UM) MND Laboratory, JALS occurred in rare instances, with an extreme case of a child contracting the disease at the age of 18 months (as cited in an article in the Times of Malta, December 2022). Given this pertinent information, the author limited her studies to include only adults.

Furthermore, Smith (2009) argues that a steep variation in the participants' lived experience may lead to difficulty in interpretation whilst analyzing the data.

3.6.2: The Sample Size

IPA is idiographic; thus, the sample size is based on a small scale, which makes sense due to the careful time it takes to analyze each case (Smith, 2009). Furthermore, Smith (2009) advises researchers to opt for an IPA and start to select three to six candidates. The author initially had a sample size of six participants planned for this dissertation; however, one of the participants needed help to contact, and when the researcher finally settled on an appointment, his wife cancelled it a day before. Another appointment was supposedly set up, but the wife did not call back. The researcher did not pursue this participant further and respected the family's wishes that the participant seemed reluctant to be interviewed.

How the participants were recruited will be outlined in the next section.

3.7: The Recruitment Process

In order to invite patients to participate in this study, two intermediaries were approached by the author. The first intermediary was the President of the ALS Malta Foundation and the founder of 'Dar Bjorn', a residential community home that caters for individuals diagnosed with ALS, MS, and other neurological conditions. The second intermediary is a Consultant Neurologist who works at Mater Dei Hospital (MDH) within the Department of Neurology.

The two intermediaries identified the individuals that satisfied the eligibility criteria. The second intermediary forwarded the contact details of four patients under his care. Information letters were handed out to the participants by the researcher herself, who

described in detail the study's aim and that they would be subjected to a semi-structured interview of approximately an hour in duration. In the case of the residents at Dar Bjorn, the researcher went personally on-site. However, the other participants pertaining to the second intermediary were contacted by phone. During the phone call, the author introduced herself, discussed the aim of her study in layperson's terms and answered any queries the participants may have had. Once the participants agreed to participate in the study, a time and place that was convenient for them was set up. In this case, the researcher interviewed the comfort of their home. At the meeting, written consent to participate in the study was given to each individual and was duly signed.

3.8: Description of The Participants

Data collection started in November 2022 and continued until February 2023. At one point, the researcher had to refrain from continuing the interviews because she had to await approval and clearance from MDH since a second intermediary had to be introduced. The interviews were restarted in January 2023. A pseudonym was given to each patient to ensure anonymity and confidentiality. Specific information regarding the participants' demographics could not be included to protect the five participants' identities. These are listed in Table 3.1 below:

Table 3. 2: Patient demographics

Participant	**Age Range**	**Gender**	**Marital Status**	**Primary caregiver**
Elijah	50-70	Male	Married	Daughter
Isaac	70+	Male	Married	Spouse
Sophia	50-70	Female	Married	Spouse
Joshua	30-50	Male	Married	Spouse
Benjamin	50-70	Male	Married	Spouse

3.9: Data Collection

Before embarking on qualitative methodology, novice researchers must be aware of any limitations and biases they may have (Treece, 1986). This research study used semi-structured interviews as a data collection method. Known as one of the most common research tools (Crabtree, 2006), interviews can be defined as a method of data collection in which one person (the interviewer) asks questions of another person (Polit, 2006). The semi-structured interviewing format is most common among healthcare professionals (DiCicco-Bloom, 2006). An interview consisting of open-ended and direct questions should be a personal and intimate encounter between the interviewer and the interviewee since eliciting narratives and accounts will be divulged (Crabtree, 2006). Griffiths (2009) states that when compared to questionnaires, semi-structured interviews allow the researcher greater flexibility and a chance to delve into any issues that may arise whilst conducting the interview.

For this dissertation, an interview schedule was drawn up; however, according to Smith (2008), the researcher can abide by it in a more relaxed manner. Not following the schedule may make the participant feel more at ease while discussing their experiences. Consequently, the participants were informed that everything said during the interview would be in the strictest of confidence. Prior to initiating the dissertation, due to the sensitive nature of the topic, a decision was taken by the researcher to exclude focus groups from this study.

Two interviews were conducted at Dar Bjorn in Qormi, and the other three participants, due to issues of immobility and/or paralysis, preferred to have the interview carried out in the comfort of their own homes. Moreover, their surroundings made them feel more at ease whilst being interviewed.

All the interviews were audio-recorded. This ensured ease of reference when the researcher had to analyze her data. The recordings were transcribed verbatim in the language used during the interview. During one interview, the author did initially try to write everything down verbatim since the patient was exhibiting articulatory imprecision and a decreased speech rate; however, by listening carefully and gently asking the participant to repeat the word or phrase when she could not understand him, the interview was continued. Another disadvantage of writing everything down verbatim during the interview would be the possibility of failing to include certain aspects of the participants' narrative (Smith, 2008). Furthermore, the researcher

enlisted the help of a professional Maltese translator to ensure that the excerpts taken from the transcribed data in the Maltese language were translated adequately and professionally.

3.9.1: Construction of The Interview Schedule

Described as 'a conversation with a purpose', Smith (2009) explains how the conversation that arises from a semi-structured interview is structured to allow the participants to express their narratives in a way they deem comfortable.

A researcher opting for an IPA will present an interview schedule before commencing their research. This schedule is prepared beforehand with questions that the researcher may wish to ask and in the manner he/she may think is appropriate for the participant. However, this does not necessarily mean that the interview will follow this schedule flawlessly and as previously planned. Nevertheless, an interview schedule offers the researcher a way whereby he/she can navigate the topics at hand and deal with any sensitive issues that may emerge during the interview (Smith, 2008).

The interview schedule drawn up by the author for this study consisted of eight questions drafted in both Maltese and English versions. Prior to commencing the interview, the participant was asked which language he/she would prefer to converse in. This placed the individual at ease and allowed them to express themselves effortlessly. Since the initial stage of an interview may be daunting and elicit feelings of apprehension and uncertainty, it is essential that prior to commencing, a rapport is formed between the researcher and the participant (Crabtree, 2006). The first question on the schedule focused on an 'ice-breaker', which was purposely planned to get to know the participant better whilst gently guiding him/her into the conversation. Once this was done, the author felt she could become more analytical in her questioning. In fact, Smith (2009) emphasized the importance of such a question since it helps the individual to feel comfortable, as previously stated. The focal point of the rest of the seven questions was the participants' initial reaction to being diagnosed with a debilitatingly progressive and terminal disease, the impact such a diagnosis had on their immediate family and friends, their coping strategies whilst dealing with the rapid progression of ALS and the QOL post-diagnosis. The author made use of various prompts, such as 'How did that make you feel?' and 'Can you please explain your emotions at the precise time of diagnosis?' during her interview. The prompts enabled

the researcher to delve deeper into the lived experience of her participants whilst ensuring that she was not 'leading' the conversation. The interview schedule was then terminated by asking the participant if there were anything else he/she would like to share with the researcher. Smith (2009) states that behind a reliable and well-prepared IPA analysis, a good quality interview and deep engagement with the client is, therefore, necessary.

Smith et al. (2009) place forth a crucial point whereby they draw attention to how the novice researcher that has never carried out an IPA may have had no training in qualitative methods, thus leading to anxiety and stress. Therefore, constructing an interview schedule beforehand may help prepare the researcher better.

The interviews were deemed to take 45-60 minutes per participant; however, due to the sensitive nature of the study and the functional physical state of each participant at the time of the interview, the researcher could only carry out the proposed time frame once. She gently reminded the participants that the moment they felt tired, in pain or any discomfort, whether physical or emotional, they could inform her, and she would terminate the interview. The participants did not ask to terminate; however, when the researcher saw that either they were struggling to talk and, in one case, the participant even removed his BiPap device to be able to speak clearer, she used her professional judgement and gently steered the interview to conclude. Furthermore, the researcher still managed to gather sufficient invaluable data from her participants.

A table consisting of a checklist of points that may prove helpful as an explanation guide prior to conducting an interview is inserted below:

Table 3. 3: Guide to conducting an interview

of points to explain prior to carrying Checklist out an interview (adapted from Rose, 1994)
➢ Purpose of interview
➢ Clarification of topic
➢ Format of interview
➢ Duration of interview
➢ Ensure confidentiality
➢ Explain the purpose of the audio-recorder and who will be listening to the recording.
➢ Assure participants that they may ask to clarify any questions asked

➢ Assure participant that he/she can decline to answer a question at any time
➢ State that there will be an opportunity to ask questions during the interview

3.10: Ethical Considerations

It is pertinent that when carrying out research, ethical issues are meticulously followed. The ethical issues involved in this study focused on confidentiality, first and foremost, and protection from harm. The subsequent sections will describe ethical issues during different stages of this study.

3.10.1: The Recruitment

A challenge for researchers everywhere is establishing an equilibrium between the requirement of research participants and refraining from the possibility of coercion. This may inadvertently affect the integrity of a sample population and jeopardise legality issues and/or respect for the individuals participating in the study (Groth, 2013). Furthermore, as healthcare professionals, we must ensure that patients included in clinical research are treated with beneficence (Groth, 2013).

The participants for this research study were recruited from Dar Bjorn, a residential home for individuals diagnosed with neurological disorders and from the Neurology Department at MDH. Since the author is a psychiatric nurse employed within the state psychiatric hospital, there was no prior affiliation with any patients. According to Whiting (2010), when the researcher does not have direct contact with the participants, they will not be constrained or feel obliged to participate in the study. An intermediary approached the potential participants, not the researcher herself.

3.10.2: The Information Letter

Potential participants were each provided with an information letter which included detailed information about the study, the potential risks and/or harm involved and the participants' rights and what was expected of them if they opted not to participate.

Courtesy of transparency and self-determination was particularly emphasised during the penning of the information letter.

3.10.3: The First Meeting

On meeting the participants for the first time, the author introduced herself and explained, in brief, the reason why she decided to opt for ALS as a field of interest. The nature of the study was then explained in a manner that would prove straightforward for the individual about to undergo the interview. Ensuring that the interview was solely about them and their lived experience and that there could be no right or wrong answers immediately reassured the participants and placed them at ease. Furthermore, an explanation was given about the study being completely voluntary and that if the participant felt uncomfortable or wished to discontinue at any point during the interview, they could do so without explaining. Consequently, it was clearly explained to the participants that deciding not to participate or withdraw from the study would have no adverse effects on their care. Finally, after everything was clarified by the author and the client was ready to proceed, the consent form was signed and read by the participants.

3.10.4: Confidentiality

Polit et al. (2006) define confidentiality as the 'protection of study participants such that individual identities are not linked to information provided and are never publicly divulged'. The author emphasised that anything said during the interview would remain confidential. Every participant participating in a research study has the right to 'privacy, anonymity and confidentiality' as Burns (2005) states. Since the interviews in this study were face-to-face, true anonymity, whereby the participant's identity cannot be associated even with the researcher, could not be affirmed in this case (Burns, 2005). Transcripts and any data collected during the interviews were kept under lock and key throughout the research period and were utterly destroyed upon completion of the study. Levine (1981) states that any information the participant passes on to the researcher must never be shared with any other individual unless consent for this has been given. For data protection, the participants' identities were guaranteed by using pseudonyms. To further protect the confidentiality of everyone

involved, the researcher of this study was the only person who had access to the data collected during the research process.

3.10.5: Consequences and Minimisation of Harm

A research interview may be the only opportunity a participant may have to openly discuss his/her lived experience, no holds barred (Carpenter, 1999). Although it is not intentional, an interview may arouse deep emotional sensations and be distressing for the individual (Streubert, 1999). Carpenter (1999) advises that if this occurs, the participant should be allowed time at the end of the interview to recollect his/her thoughts, 'debrief' and seek help if required. Additionally, it is a common occurrence that the interviewee, to avoid feelings of nervousness, may persistently ask the researcher whether he/she has done well and offered sufficient information (Carpenter, 1999). This phenomenon, in fact, was encountered by the researcher in this present study.

This research study dealt with patients who were not only diagnosed with an incurable and fatal illness but also one which is physically and emotionally distressing. The patients understood only too well the rapid changes their bodies were undergoing daily. Thus, the author had to deal with high sensitivity and vulnerability during the interviews. The information letter clearly stated that if, at any time, the participant felt any anxiety or psychological distress whatsoever, the interview would be stopped, and the patient would be provided with professional counselling from a counsellor or a psychologist that had agreed to offer his/her services
free of charge.

3.10.6: Ethical Approval

The Department of Health (2001) states that the well-being of the individual participating in the study must be our first concern as researchers. Ethical procedures must be strictly followed when patients' vulnerability is at stake. This includes obtaining informed consent and receiving approval from the Local Research Ethics Committee to proceed with the study. In ethics, the two main areas of significant concern are confidentiality and the possible consequences of a research-related interview (Beauchamp, 2001).

Ethical approval for this study was obtained from the Faculty of Research Ethics Committee (FREC) and the University Research Ethics Board (UREC). Additionally, the researcher had to seek approval from MDH's Chief Executive Officer (CEO), the Data Protection Officer (DPO), the Chairman of Neurology, the MDH Legal Team, the Psychiatric Services and a treating Consultant from the Neurology Department, who acted as the second intermediary of this study.

3.10.7: Roles and Boundaries

The relationship built between the participant and the researcher during the research period is arguably the most intimate and essential part of the study since it will inevitably have a significant effect on the study outcome (Thurairajah, 2006). The levels of intimacy that are allowed without breaking boundaries depend upon the school of thought, whereby some researchers believe that objectivity is a must during the participant-researcher relationship since this prevents 'contamination of the field' (Thurairajah, 2006). However, such objectivity has been criticised, and its practicality questioned (Harding, 1993). Moreover, studies show that when there is a divide between researcher and participant whereby the researcher holds back from revealing anything about him/herself, this may negatively impact the depth and richness of any data collected (Harding, 1993).

Rebar (2011) argues that researchers must differentiate between their professional role and that of a researcher when conducting a study. In this case, the author is a psychiatric nurse by profession. However, she takes on the role of a researcher for the duration of her research. The author could not use her professional expertise as a nurse to address any difficulties her participants may have enquired about during their interview. Instead, she advised them to inform the healthcare professionals following their medical care. As stated previously, the fact that the researcher was employed in an entirely different hospital and had never encountered or worked with the participants prior to the study helped reduce any lack of clarity or uncertainty related to her role.

Once the data collection phase was accomplished, the next step in the author's research process involved data analysis. This is outlined in the next section.

3.11: Data Analysis

An IPA is a systematic process based on a thematic analysis of data collected (Smith et al., 2009). The process is steered by maintaining openness and willingness to scrutinise and extract the meanings of the participants' experiences. The process of data analysis involves six steps which are described below.

Step 1: Reading and re-reading:

The interview transcripts were read during the first step whilst listening to the audio recordings. This was done to familiarise the researcher with the participants" experiences (Smith et al., 2009). Whilst listening to the recordings, any pauses accentuates, repetitions or emotions expressed by the participant were jotted down. Such gestures are paramount when interpreting data (Rapley, 2001). The transcripts were read and re-read multiple times to ensure that the researcher grasped the context and actively engaged with the data (Smith et al.).

Step 2: Initial noting:

During the initial noting, any significant statements were highlighted and exploratory comments were written in the right-hand margin of the transcript. The initial noting revealed three types of comments: descriptive, conceptual and linguistic. Examples of the comments are listed in Table 3.5 below:

Table 3. 4: The three types of comments during initial noting

Type of comment	Definition	Example
Descriptive	The description of content of what and the manner by which the participant discussed it. Such comments included acronyms, figurative speech, emotional responses and assumptions	*"Imma nsomma, pero jekk jiena nara perezempju, it's too much now, u nirrifjuta l-maskla u nirrifjuta li niekol, qisni qieghed nitfi l-magna wkoll" (Benjamin, p.4, lines 103-106).* *"But anyhow, if I see, for instance that it is getting too much now and I refuse to wear the mask* [respirator] *and I*

		refuse to eat, then it is like I am turning off the switch as well" The participant here is talking about the subject of euthanasia. That he does not necessarily have to accept to be euthanised. He can reach the same aim of death by refusing to become respirator-dependent and refusing nutrition.
Conceptual	The identification of abstract concepts which guide the researcher to understand the meaning of the participants' experiences.	*"Xi kultant nigi fi zmien li nkun iebes" (Elijah, p.15, lines 434-435)* *"Sometimes a time comes when it feels hard* [his situation]*"* This statement could be interpreted in two ways. One that the participant is feeling that his diagnosis of ALS is too difficult to accept and bear or secondly, the participant is speaking about the symptoms of muscle stiffness caused by ALS. Thus, the transcript has to be read and re-read very carefully, taking into consideration every contextual meaning.
Linguistic	Linguistic components within the context of IPA are of extreme importance when reflecting and attempting to understand the participants' discourse and experience. Examples of linguistic components include: repetition,	*"Le, le, le, ghandi hija mhu kapaci xejn. Xejn, xejn, xejn" (Elijah, p.13, lines 367-368)* *"No, no, no, my brother is not capable at all. Not at all, not at all, not at all"* The repetition in this case reveals the disappointment and sadness of the participant towards his brother who

	metaphors, personification and digressions.	could not accept a diagnosis of ALS on his sibling.

Step 3: Developing emergent themes:

The analysis of exploratory comments to recognise emergent themes was initiated by organising the data and fragmenting the original transcript of the participants' recordings into sub-themes. The excerpts that fit into each sub-theme were highlighted with a different colour for the researcher's ease of reference and clarity. Furthermore, grouping sub-themes led to an emergent theme (Smith et al., 2009). Table 3.6 below illustrates the emergence of a theme from the sub-themes identified from a participant's transcript:

Table 3. 5: Developing a theme from a sub-theme

Theme	Sub-theme and excerpts
The Thief of Independence	Loss of independence and autonomy: *"The most difficult thing to accept probably is not being any more able to eat. I mean I love eating and not feeding myself traditionally it's kind of very difficult."* (Joshua, p.2-3, lines 55-59) *"Well, it's taken away my independence. I cannot shower myself or dress myself anymore"* (Isaac, p.5, lines 128-130)

Step 4: Searching for connections across emerging themes

Step 4 involves searching for connections between the theme and then go on to develop the super-ordinate themes. The transcripts were scrutinised for common factors and similarities which captured the participants' worries, concerns and interests of living with ALS.

Step 5: Moving on to the next case

After analysing one interview transcript, the researcher moved on to the next one and repeated the four steps described above. As per recommendation by Smith et al.,

(2009), the researcher gathered the ideas that emanated from the first analysed transcript whilst working on the second and so forth.

Step 6: Searching for patterns across each transcript

Once all five transcripts were analysed, the two super-ordinate themes and two emergent themes were identified and listed in two separate table for ease of reference and clarity. These two tables are illustrated in the next Chapter.

3.12: Establishing Trustworthiness

Qualitative research is becoming noticeably appreciated and recognised, therefore, to be accepted as trustworthy, data analysis has to be carried out in a meticulous and rigorous manner whilst being consistent and precise (Nowell, 2017).

Trustworthiness or accuracy of a study can be defined as the confidence in data analysis and the methodology used to ascertain the quality of a study (Polit, 2014). Moreover, according to Lincoln and Guba (1985), the concept of trustworthiness helps researchers convince readers that the research carried out is worthy and credible. To further clarify the concept of trustworthiness, Lincoln and Guba (1985) introduced the criteria of credibility, transferability and dependability. The three general criteria, namely credibility, transferability and dependability in relation to this research study will be discussed in table 3.3 below:

Table 3. 6: General Criteria

Criterion	Description
Credibility	Credibility, considered a particularly important criterion in qualitative research, can be defined as the 'confidence in the truth of the study' and its interpretations (Polit, 2014). In other words, credibility addresses how accurate the researcher's interpretation of the participant's narrative is (Tobin, 2004). Lincoln and Guba (1985) recommend certain methods to promote credibility. These include peer debriefing and reflective journalling, amongst others. For this study, the author opted for reflective journalling during her research period and also attended regular debriefing sessions with her academic supervisor to ensure that her interpretations were of high standard, logical and credible.

Transferability	Transferability refers to the in-depth description of the context, location and setting in which the research study occurred (Harrington, 1998). Moreover, researchers are required to provide a description of their study which is vivid and appeals to its readers (Amankwaa, 2016). To ensure that she abided by this criterion, the researcher attempted to describe, in great detail, the context. Similar to reliability in quantitative research, dependability refers to the stability of data over a period of time (Polit, 2014). It also refers to the detailed notes, referred to as 'process logs' taken by the researcher. These notes include clear documentation of all decisions and activities that occurred during the study (Koch, 1998). The decision trail carried out by the researcher of this study is clearly documented throughout this chapter. Additionally, dependability also refers to whether the choice of opting for an IPA was appropriate for this particular research study. Since the IPA methodology is known for allowing participants to express their lived experiences openly, it was decided by the researcher that opting for an IPA will in fact prove beneficial for this study since in-depth exploration of individuals with ALS was required.
Dependability	Similar to reliability in quantitative research, dependability refers to the stability of data over a period of time (Polit, 2014). It also refers to the detailed notes, referred to as 'process logs' taken by the researcher. These notes include clear documentation of all decisions and activities that occurred during the study (Koch, 1998). The decision trail carried out by the researcher of this study is clearly documented throughout this chapter. Additionally, dependability also refers to whether the choice of opting for an IPA was appropriate for this particular research study. Since the IPA methodology is known for allowing participants to express their lived experiences openly, it was decided by the researcher that opting for an IPA will in fact prove beneficial for this study since in-depth exploration of individuals with ALS was required.

3.13: The Reflective Account

Being a nurse specialising in Mental Health, one may ponder why I chose to carry out my research study on patients with ALS since my profession deals with acute cases of mental illness daily and not neurological disorders. The reason why I decided to base my research study on ALS was twofold. The first reason was that a member of my immediate family suffers from a debilitating neurological disorder, but not ALS. The second was the yearning I have always had to extend my knowledge of neuropsychiatry and neurogenetics. Admittingly, this research study did not come without difficulty. The cases I encountered were harrowing; however, the manner in which the participants faced their diagnosis of ALS with such strength, resilience, and courage left me speechless at the best of times. Being a novice researcher, I found the interviews challenging at times, and although I had practised independently at home, I still felt an amount of trepidation and anxiety during my first interview, and nothing could prepare me for the havoc ALS wreaks on the individual. However, my interviewing skills improved with each one I carried out. The heart-wrenching interviews sometimes left their toll on me; however, with the guidance and expertise of my academic supervisor and keeping a reflective journal, I overcame these difficult moments. I can declare that I have emerged as a different person from the one who first started this academic journey. The participants instilled in me a deep sense of courage and spirituality, and without knowing, I now look at life from a totally different perspective. One must keep in mind that this cohort of participants were given a fatal diagnosis and a disease that leaves a trail of destruction in its path and renders them locked in their own bodies as if frozen in time.

As a psychiatric nurse, I did have a degree of understanding regarding the difficulties patients with ALS go through from a psychosocial aspect. However, having experience in the field of research may be seen as a paradoxical influence. According to Bishop et al. (2011), the study may be enhanced in such cases because it may be easier for the researcher to build a rapport with the patient. I did build a rapport with my participants, and despite their poor prognosis, they did their utmost to share with me enough valuable data to complete this study.

Due to the sensitive nature of their illness, it was not easy to continuously prompt them for more information. Only one interview lasted between the speculated time-frame of 45-60 minutes. Some participants had breathing difficulties and tired quickly; others were in discomfort and/or pain. Therefore I decided not to cause any unnecessary distress and reminded them to inform me if they became tired or felt they did not wish to reply to a specific question. One participant was adamant that he did not wish to be given any information about the symptoms or the prognosis of ALS because he did not wish to know what would happen to him later. *"Yes, I do not want to know too many details if you do not mind, cause I do know, but I am not too keen to find out what is going to happen to me if you do not mind"* (Isaac, p.2-3, lines 54-58).
Keeping a reflective journal helped keep my thoughts on track and organised. I used my active listening skills and did my utmost to go for each interview with an open mind and a non-judgmental approach whilst curbing any preconceptions I may have had from previous experiences.

3.14: Conclusion

To conclude, and as explained in preceding sections, an IPA involves the in-depth exploration of an individual's lived experience and relation to a particular phenomenon. In this research study, the author aimed to understand and appreciate the lived experience of adults diagnosed with ALS, both sporadic and genetic types. Thus, IPA was her chosen and preferred methodology.

Chapter 4: Results

4.1: Introduction

This chapter describes the researcher's overall findings focusing on the lived experience of individuals diagnosed with ALS. Additionally, the author analysed her data via those described by Smith et al. (2009). Several themes emerged from the participants' narratives. Thus, the researcher will present the themes and superordinate themes together with her interpretations relevant to her study.

Smith et al. (2009) recommended that when carrying out an IPA, emergent themes taken from the excerpts of semi-structured interview transcripts are to be identified. The chosen excerpts are then typed in an italic font and enclosed with quotation marks. For interviews carried out in English, only the English version is required; however, for any interview conducted in Maltese, the excerpts have to be presented in both the Maltese version and the corresponding English translation. If part of an excerpt has to be omitted, then this is indicated by inserting an ellipsis ("…"). On the other hand, in academic writing, square brackets are used when the researcher has to include words within an excerpt that are not part of the original quote, as stated by the participant. According to the data protection act, the researcher used pseudonyms to protect the participants' identities. Five pseudonyms relating to participants were: Joshua, Isaac, Elijah, Benjamin and Sophia.

While conducting the interviews, the researcher noted specific linguistic components that her participants used but were oblivious to. It is recommended that the interviewer takes note of the individual's use of language whilst going through the transcripts since, according to Smith, Larkin and Flowers (2009), this adds a clearer perspective behind their stated words.

4.2: The Linguistic Component

Four different types of linguistic components were observed from the transcriptions, namely, repetition, metaphors, digression and personification. The researcher noted that repetitions were predominant among the four types of linguistic components. There are various types of repetition in linguistics; however, in this case, the type observed by the researcher is known as pali logia. It is an ambiguous form of speech primarily used to show intense emotion whereby the orator is noted to emphatically

repeat the same word or phrase consecutively with no intervening words (Tarpinian, 2020). A clear example of this mode of repetition is seen in the following excerpt, whereby Elijah became emotional when describing how his brother was incapable of accepting that he, Elijah, had been diagnosed with ALS:

"Heqq…ma haduhiex nahseb tajba u? Le, le, le. Ghandi hija mhu kapaci xejn. Xejn, xejn, xejn. Iwaqqa lili hazin imbaghad." (Elijah, p.13, lines 366-368)
"Heqq…I do not think they took it very well. No, no, no. I have a brother who is not capable at all. Nothing, nothing, nothing. He makes me feel bad, then."

A linguistic component that stood out during analyses of the transcripts was a digression. Digression is moving away from the main subject in speech or writing and initiating a discussion on an unrelated topic (Nordquist, 2019). A clear example was when Sophia explained how, due to her decreased dexterity in her fingers, she could not tend to her infant grandson anymore because she could not do basic things such as feeding him or changing his nappy. She suddenly switched the conversation to how she considered her children a godsend. This digression could mean that Sophia felt guilty for causing them the grief of having a mother who cannot help her grandchildren due to her being diagnosed with ALS. The following excerpt highlights the apparent digression:

"I used to ehm…babysit, but as soon as they learned I have ALS, they stopped it for me. Because they did not want me to… I cannot handle certain things with a baby, you know? Ehm…well, the youngest is two now…you know. Ehm…you know. Changing nappies and feeding him. Cause of my fingers. Exactly…so I had that problem. So I didn't have the…I mean, they're a Godsend, my kids! I am not saying that because they're my kids, but they are." (Sophia, p.3, lines 68-80).

The third linguistic component encountered was personification. *Personification* is defined as a statement of something which is not human, however, it is noted to be given human traits (Melion, 2016). Melion (2016) adds that although personifications are commonly encountered in everyday language, the 'pictorial effects' are not given enough academic attention, thus leading to the use of personification as a means of communication that is ultimately being taken for granted.

The excerpt from Elijah's transcript below shows a clear example of personification whereby the participant strongly emphasises how being diagnosed with ALS took away his enjoyment of eating and drinking. This, leading him to have to rely on others to feed him:

"Ehh…mill-ikel. Kont niekol wahdi. F'Mejju ehh…perezempju f'Mejju hrigna u ghajjejt niekol. L-imgharfa ghajjietni." (Elijah, p. 12, lines 340-343)
"Ehh…from the food. I used to be able to eat on my own. …for instance, we went out in May, and I became tired of eating. The spoon tired me."

Another linguistic component noted was the use of metaphors by the participants. According to Merriam-Webster (2016), a metaphor can be defined as a 'figure of speech' whereby rhetorically, literally refers to one object or idea by mentioning another, thus suggesting an analogous relationship. Additionally, metaphors are a helpful tool that helps researchers interpret and analyse their data whilst illuminating the lived experience being narrated (Patton, 1990).
During transcription and whilst carrying out the interviews, the researcher did notice that, at times, the participants used metaphors. An example indicating a metaphor extracted from Benjamin's transcription is shown below, whereby the patient discussed his opinions on euthanasia and the devastating effect such a decision would have on loved ones. However, on the other hand, he states that refusing to wear his 'mask' and refusing to eat will eventually lead to his death anyhow:

"Pero, jekk jiena nara perezempju, it's too much now u nirrifjuta l-maskla [respirator] *u nirrifjuta li niekol, qisni qieghed nitfi l-magna* [life-support machine or death] *wkoll."* (Benjamin, p.4, lines 103-106)
"Although, If I see, for example, it is too much [to bear] *now and I refuse the mask* [respirator]*, and I refuse to eat, then it is like I am turning off the machine too* [life-support machine or death]."

4.3 Superordinate and emergent themes

In the following tables (i.e., Tables 4.1, 4.2 and 4.2.1), the researcher presents two superordinate themes, their emergent themes and sub-themes, which further explain the lived experience of individuals diagnosed with ALS. Thematic excerpts taken from the participant's transcripts are also presented.

Table 4. 1: Superordinate themes and emergent themes describing the overall experience of individuals diagnosed with ALS.

Thematic Excerpts	**Super-ordinate Themes**	**Emergent Themes**
"You can't do the normal things that normal people would do with their normal able body. So yes, I miss doing normal stuff like walking and running and all the stuff that people do with their muscles" (Joshua, lines 39-43, p.2)	**A ticking time bomb**	➢ *The thief of independence* ➢ *A rollercoaster of emotions* ➢ *The financial blow*
"I know it's extraordinary really and I'm rather glad. I mean, I'm nearly 73 now so I am lucky. My father got it when he was in his 50's" (Isaac, lines 49-51, p. 2).	**Striving to stay afloat**	➢ *Adapting to the situation* ➢ *Be it done unto me according to Your Word* ➢ *Support for a fatal diagnosis* ➢ *Facing ALS with humour* ➢ *Living the present*

The following table (Table 4.2) provides details regarding the emergent themes and sub themes.

Table 4. 2: Emergent themes and sub-themes

A ticking time bomb	
Emergent Themes	**Sub-themes**
The thief of independence	➢ Rapid progression and debilitation of ALS ➢ Loss of independence ➢ Becoming a burden
A rollercoaster of emotions	➢ Suppression of emotions ➢ Fear of developing depression ➢ Denial of ALS being fatal ➢ Anger at being treated differently ➢ Upset at no longer being considered normal ➢ Frustration at a dysfunctional body ➢ Fear of ALS being passed on in the family
The financial blow	➢ Loss of employment ➢ Increased financial burden on family ➢ Expenses of commodities and necessities

Table 4. 3: Emergent themes and sub-themes

Striving to stay afloat	
Emergent themes	**Sub-themes**
1. Adapting to the situation	➢ Finding peace and serenity ➢ Achieving great things ➢ Compensating for the loss in quality of life ➢ Gratitude at being diagnosed later in life ➢ Finding resilience
2. Be it done unto me according to Your Word	➢ Finding solace in spiritual faith ➢ Sorrow at not being able to participate in religious activities ➢ Acceptance of the diagnosis
3. Support from various quarters	➢ Support from treating clinical neurologists and healthcare providers ➢ Support from UM ALS/MND research team ➢ Support from Mr. Bjorn Formosa, founder of Dar Bjorn ➢ Support from family, colleagues and friends ➢ Support from the family pet
4. Facing ALS with humour	➢ Humour as a coping mechanism ➢ Lifting the morale
5. Living the Present	➢ Facing ALS with courage ➢ Living day by day

4.3: A Ticking Time Bomb

The researcher opted for the super-ordinate theme *of A ticking time bomb* because it was a common factor she noted amongst all five participants whilst carrying out the interviews. It clearly describes the disbelief and shock of the participants being told, after a battery of tests, that not only have they been diagnosed with a rare and fatal disease, but rapidly progressive and physically debilitating. The initial signs and symptoms the participants exhibited were vague and sudden. Thus, the participants

could not correlate to any particular disorder, least of all MND. All five participants recounted how they were not immediately aware that something was amiss, and three explained that they thought the sudden falls they were experiencing were because they had tripped over something while they were working or walking.

The first super-ordinate theme, *A ticking time bomb,* describes how Joshua, one of the five participants, mentioned that he went from being a sportive, healthy and non-disabled individual to one where rapid muscle deterioration started to take place, leading to him becoming paralysed in all four limbs in a short period. Consequently, Sophia, the only female participant, stated that her initial symptoms were weak fingers and that it never crossed her mind to consult this issue with her family doctor because she thought it would resolve on its own accord. Only when she started to lose weight rapidly and the weakness in her fingers worsened did she seek medical help. Apart from Sophia, Benjamin also reported unexplained weight loss, prompting the author to research this phenomenon further. According to Bouteloup (2009), possible reasons for severe weight loss in patients with ALS tend to be heterogeneous. However, it may also be due to cachexia, malnutrition, anorexia and hypermetabolism (Bouteloup, 2009).

Furthermore, the diagnosis was harder to bear because they realised that they would become completely paralysed and incapable of eating on breathing on their own. At a late stage, they will eventually have to be assisted by a ventilator to breathe. This will render them utterly dependent on their caregivers. Furthermore, this super-ordinate theme also honed in on the impact a diagnosis of ALS had on the participants and the myriad of emotions they go through daily.

4.3.1: The Thief of Independence

This particular theme depicts the life experiences as explained by the participants when the progressing symptoms of ALS caused them to lose their independence. Of the five, only Sophia had just been newly diagnosed and could still walk unaided. The participants go into great detail about how simple things that everyone takes for granted and bring them joy, such as walking, eating, playing a sport, fastening up a shirt, going to the bathroom, or cooking, have suddenly been snatched away from them, thus leaving them to feel robbed of their independence and dignity. The excerpt below, taken from Isaac's transcript, clearly indicates this:

"Well, it has taken away my independence. I cannot shower myself or dress anymore. Ehm… we have got a very nice Philippine helper who comes in three times a week, if possible, to help me." (Isaac, p.5, lines 128-132).

Another participant, Sophia, who stated that prior to being diagnosed with ALS, she led a very healthy and active lifestyle, explained how ALS robbed her of the opportunity of helping out with her grandchildren.

"I used to take care of their kids because they are both…. Two of them are married, and they both have two kids. I used to, ehm…babysit, but as soon as they got to know I have ALS, they stopped it for me. Because they didn't want me to…cause I couldn't handle certain things." (Sophia, p.3, lines 66-70)

Shock and disbelief at being diagnosed with a rare neurological disorder were discussed in depth throughout the interviews. However, One participant, who had the familial form of ALS and refused to know anything about the outcome he may be facing, was not surprised at his diagnosis. Both Joshua and Sophia could not understand how such a rare disease afflicted them, especially since they both took care of their bodies and exercised regularly:

"I was shocked at first. I could not believe that I got such a disease because ALS affects only 4 in every 100,000 people, eh? (Joshua, p.1, lines 14-16
"I couldn't believe that I have ALS, to be honest. I couldn't believe how it came on me, you know, 'cause I wasn't the type that…ehm…I always used to eat healthy. Like I said before, Exercise, I used to really take care of myself, you know." (Sophia, p.1, lines 16-22).

Another main concern which stood out during the interviews was the rapid progression of the disease and the physical changes that ensued. Sophia, who was in the early stages of ALS, admitted that the most challenging part of the diagnosis was seeing her body change so radically. During the interview, it was evident that Sophia had always taken great pride in keeping herself healthy and fit. Being the only female participant in the study, the fact that she seemed so worried about her appearance tallied with the

fact that women may be affected by a diagnosis of ALS in a different way than that of a male counterpart from a physical perspective:

"I get my days when I'll be sad. You know, especially looking at my body. You know. That upsets me a lot. I think that's the worst part, actually, not even my fingers. It's my body…ehm…You know, cause, come on, from 65-66 [kilograms], *then you go down to 48. It's not a joke, you know."* ((Sophia, p.7, lines 203-209).

Joshua, Benjamin, Isaac and Elijah, on the other hand, stated that the physical debilitation of ALS affected them personally in a myriad of ways:

"U qbadtu jien [zimmer frame]*, u bdejt nimxi bih mill-ewwel, hekk. Ghidt, il-allu kemm hu komdu!. Il-persuna li hadni u din* [daughter] *baqghu jharsu lejja! Imma hassejtni komdu fih, Biss imbaghad kienet qaltli* [sales representative] *'dan temporanju'. Ifhem, heqq, ipprova ifhem, hadtha naqra hekk. U hekk ghidt, ghidt daqshekk fast din?* [ALS]" (Elijah, p.9, lines 241-248).	*"And I grabbed it* [zimmer frame], *and I started walking with it immediately. I said 'il-allu' how comfortable it is! The person that took me and this one* [daughter] *just stared at me. I found it comfortable. But then she* [sales representative] *told me 'this is only temporary'. Try and understand; I took it a little bad. And that is what I said. I said is it* [ALS] *that fast?"*

At specific points, the researcher noticed that most participants were either in denial or did not wish to mention that ALS is a fatal disease. Thought suppression, which is a possibility in this case, maybe a coping mechanism in order not to feel anxiety. However, fears and concerns of being a burden to their loved ones did emerge. All five participants were married.

During the interview, fears and concerns of becoming a burden and dependent on their loved ones did emerge. Elijah admitted that apart from being diagnosed with ALS, he also had the burden of worrying about his wife, who had mental health issues and was incapable of caring for him. It was evident in the way he repeated certain words and

his demeanour during the interview that this was a very emotional and sensitive topic for him to talk about:

"Le, mhix tajba xejn [wife]. *Xejn, xejn, xejn, le xejn. Mhux kapaci xejn xejn. Ijja, le mhix kapaci. Jigifieri jiena, barra ha nghidlek, barra din li ghandi* [ALS], *ghandi taghha wkoll* [mental health issues]." (Elijah, p.17, lines 86-89).	*"No, she* [wife] *is not good at all. Nothing, nothing, nothing. No, nothing. She is not capable at all, at all. Yes, no, she is not capable. So me, let me tell you, apart from having this* [ALS], *I also have hers* [mental health issues]."

"He (husband) *stepped down* (from his position at work) *because of my situation, so I am anxious that he feels he can go out and, you know, do things. He doesn't leave me in the evenings, obviously, but he does go to the gym during the day. He just goes for a walk in the afternoon. That's absolutely fine."* (Isaac, p.7, lines 183-189).

4.3.2: A Rollercoaster of Emotions

The first thing that came to light during the five interviews was the incredible strength and courage the five participants portrayed in view of their dire circumstances and, at times, even appeared in denial. So much so that at one point, the researcher started to ponder whether they had come to terms with the fact that they were slowly dying. One participant, Sophia, who had just been recently diagnosed, stated, "I am *really, truly, fighting this. I'm not gonna give up, you know. Even if I'm 80*" (Sophia, p.5, lines 122-124). Sophia seemed to be oblivious to the fact that ALS is rapidly progressive and terminal. Furthermore, it was noted that out of the five, only Benjamin and Joshua outrightly mentioned that ALS is a fatal condition and that they were slowly dying. Out of the five, only Benjamin broached the subject of euthanasia. He seemed to believe that outrightly requesting to be euthanised would be a difficult decision to make and would disrupt the individual's loved ones and cause unnecessary grief. Additionally, Benjamin also stated that if the time came when he felt that he could not continue suffering, he could resort to refusing his ventilation and refusing any type of nutrition. To him, this method is just another way to end his life without going into the

complex and sensitive ethical repercussions of euthanasia. This showed the researcher that both Benjamin and Joshua were aware that ALS was terminal:

"*Imbaghad ser joffrulek a wide range of options biex wiehed jittermina hajjtu. Imma nsomma. Pero jekk jiena nara, perezempju, it's too much now u nirrifjuta l-maskla* [respirator] *u nirrifjuta li niekol, qisni qieghed nitfi l-magna* [life-support machine/death] jiena wkoll (Benjamin, p.4, lines 101-106).	*"Then you are going to offer a wide range of options so that one can terminate his own life. But anyway. Although, if I, for example, see that it is getting too much now and I refuse to wear the mask* [respirator] *and I refuse to eat, then it is as if I am turning off the switch too* [life-support machine/death].

"I don't care much about what is happening to my body. Even though what is happening is quite obvious. I mean my body is dying bit by bit and I'm totally getting paralysed more and more every day." (Joshua, p.2, lines 48-52)

Out of the five participants, Elijah was the only one that mentioned a fear of falling into a depressive disorder. Throughout the interview, Elijah showed courage, faith, strength and, at times, even humour. However, he did mention that he immediately pulled himself together during intense sadness for fear of falling into a depressive state. This showed that the fear Elijah had of becoming depressed was most probable due to seeing his wife suffer from a similar disorder:

"Imbaghad, f'dak il-hin ikklikkjali mohhi. Ghidt isma, ghidt jiena rrid noqghod attent hawnhekk. Ghidt ghax issa ma rridx nigi…li nigi, x'jismu, nigi hazin u nispicca niehu l-pirmli [antidepressants]. *Ghax hassejtni hekk, li ha…eh…depression."* (Elijah, p.7-8, lines 196-202)	*"Then, at that moment, it dawned on me. I said listen; I have to be careful here. Because I do not want to end up feeling so bad that I will have to take pills* [antidepressants]. *Because I felt as if…eh…depression."*

Considering how devastating a diagnosis of ALS can be, the researcher was surprised that an apparent suppression of emotions emerged from two participants. Joshua explained how he warded off any emotional feelings by concentrating on other things and keeping himself occupied and busy so as not to expose himself to what he called terrible thoughts:

"Most of the time, I concentrated on doing other things, so I don't have time to think about other emotions." (Joshua, p.1, lines 19-21).

Benjamin, on the other hand, admitted that, as a person, he was very 'cold' and that talking about his emotions was something he was not willing to do. In a previous excerpt, Benjamin stated that he does not wish to disclose his concerns about the possible heredity of ALS in front of his wife. The researcher could not distinguish whether this suppression of emotions was not to worry his wife, if it was a personality trait or whether he wanted to appear strong in front of his family and not be seen as making a fuss:

"*Difficli biex tispjega l-emotions hux? Ehe…ghidli inti? Toqghod tibki? Jiena bhala persina cold hafna. Bhala persuna jiena cold fir-reaction tieghi.* (Benjamin, p.2, lines 42-47).	*"It is difficult to explain emotions, isn't it? Ehe… Do you tell me? Do you start to cry? As a person, I am very cold. As a person, I am very cold in my reaction."*

Elijah and Sophia did not have a problem admitting that their diagnosis of ALS became so hard to bear that they cried and allowed themselves to express their emotions. Furthermore, Isaac did not mention any emotions at all. He portrayed a cheerful demeanour throughout the interview, so much so that to the researcher, it seemed as if he was unwilling to admit that he was suffering from a rare and fatal neurodegenerative disorder that was about to change his life.

Resilience in the face of a devastating and fatal neurological disorder was seen across all five participants. They all found different ways to deal with their diagnosis but showed courage, strength and resilience. Joshua discovered that keeping his mind occupied, breathing with the aid of a ventilator and dealing with paralysis still allowed him to compensate for the reduced quality of life:

"There is probably nothing worse that could happen to a person, but in a certain way, there are some things to do if you use your brain and you put your mind to it. You can still do things and compensate for the big loss in quality of life." (Joshua, p.3, lines 70-73).

Being told they had ALS after enduring so many medical tests was a daunting experience for all five participants. Not once did Elijah mention the word ALS during the interview, and on being told of the diagnosis, Elijah said:

> *"Qalli* [treating neurologist]*, li gejt bl-M…* [MND]*…dan" "He [neurologist] told me that I have been diagnosed with M* [MND]*…this".* (Elijah, p.7, line 184).

In another interview, Isaac clarified that he does not want the researcher to go into great detail about how ALS will affect him physically. In light of his father dying from ALS, this statement seemed to show a sense of denial and a refusal to accept that he would be suffering the same fate. His wishes were respected, and the researcher ensured that she did not mention anything about ALS being a terminal disease:

*"Yes, I do not want to know too many details, if you do not mind, because I do know, but I am not really keen to find out what's going to happen to me if you don't mind. I have a rough idea, but it's much better trying to think positively and…ok?" (*Isaac, p.2-3, lines 54-59).

At one point during the interview, Isaac became paradoxical when he stated, "I don't really want people around" (Isaac, line 82, p.3). Throughout the interview, he emphasised the importance of having supportive friends; however, on the other hand, he stated that he did not want people around him, especially those who showed pity towards him or made fun of him for being diagnosed with ALS:

"I just want to be treated as I have always been treated. I don't suddenly want to be this peculiar person sitting in the corner who everyone jokes about. I won't put up with that." (Isaac, p.4, lines 86-90).

The aspect of what normality or being normal meant to them did emerge, especially during Joshua's interview. Although he was very knowledgeable about ALS and knew what to expect on prognosis, it was evident that Joshua felt saddened about losing everything he considered 'normal' before he was diagnosed with ALS:
"Before the diagnosis, I had a normal life. I was a person doing his things. Enjoying sports and doing my business, and then suddenly this condition came out ok?" (Joshua, p.1, lines 10-13).

On the other hand, Sophia emphasised how frustrated she became on realising that something as simple as doing up the fastening of her necklace was now impossible. To her, it was difficult to accept that something so 'normal' to any other person was no longer an option for her:

"You can't clip up a necklace or, you know, or anything like that, and it is frustrating. Believe me. I try as I said, but I can never do it." (Sophia, p.6, 152-154).

A common issue across all five participants was the frustration at realising that their bodies could no longer function as they used to. This was due to various reasons. Three of the participants had led active and healthy lifestyles before their diagnosis, whereas Elijah and Benjamin were dedicated to their profession. Due to the debilitating nature of ALS, they could no longer engage in their lifestyle activities and ultimately had to give up their jobs. Furthermore, Benjamin had concerns about being a risk to himself and others. Thus the decision to end his employment was final:

Tfixkilt wire, u ksirt il-hip bone. Imbaghad fis-sena recovery li kelli, ehm…s-sintomi aktar avvanzaw u ma ergajtx dhalt ghax xoghol. Jigifieri kont due li nidhol November, 2019, pero ma kontx tajjeb biex immur ghax-xoghol. Kont ta' Periklu ghalija u ghal haddiehor." (Benjamin, p.1, lines 12-18.)	*"I fell over a wire, and I fractured my hip bone. Then, in the year of recovery that I had, ehm…the symptoms progressed a lot and I did not return to work. I was due to return in November 2019, but I was not well enough to go to work. I was a danger to myself and others."*

"You can't do the normal things that normal people would do with their normal able body. So yes, I miss doing normal stuff like walking and running and all the stuff that people do with their muscles." (Joshua, p.2, lines 39-43).

Two of the five participants did not have children, so any concerns about ALS being passed on genetically were not mentioned. Furthermore, with Isaac being the only participant with the familial form of ALS, the others, except Benjamin, were not concerned about this issue since they understood clearly that their case was sporadic. At one point, though, Elijah's daughter, who was present during the interview with her father's consent, did show concern about herself developing ALS and had personally asked a medical professional regarding this matter. During the interview, her father put her mind at rest. Benjamin was the only participant concerned about ALS being passed on to his children, even though it was confirmed that he had the sporadic form. Whereby Benjamin showed great faith in science, this issue was something that worried him greatly, even more so than the deterioration of his own body:

"Pero l-iktar haga li hi ta' concern ghalija kienet li jekk hemmx eredita'. Ghax ma ridtx nghidlek quddiem il-mara. Imma dik hija l-ikbar concern tieghi. Jekk tergax [ALS] *titfacca fil-familja."* (Benjamin, p.3, lines 79-83).	*"Though the thing that is most concerning is that it may be hereditary because I do not want to tell you in front of my wife. But that is my greatest concern. That it* [ALS] *will reappear in the family*."

4.3.3: The Financial Blow

Out of the five participants, only Elijah commented on the possible financial repercussions a disease like ALS can have on a family. He stated how he was fortunate enough to keep his business running, even though he could no longer work himself, and thus, his family was not affected by a loss of income. He explained the importance of certain commodities and necessities, such as comfortable wheelchairs and pillows, which are expensive to purchase. Having a terraced house, Elijah soon became unable to climb the stairs. Thus, he decided to transform his garage into a bedroom for himself. Since he was financially stable, he could see to it that construction works could be carried out there and then and could finish the project in three months. Elijah

also pointed out how it is already difficult to accept a diagnosis of ALS, let alone if one is financially unstable. The researcher noticed that Elijah became emotional when mentioning how other individuals with ALS who could not afford specific necessities would suffer even more than he is:

"Ghandek ohra li jiena ma hassejthiex [finanzjarjament] *ghax kont nahdem ghar-rasi. Jigifieri, tista taffetwak* [ALS] *bhala finazni. Jiena ma affetwatniex ghax dan kollu xoghli. Dawn l-affarijiet kollha gholjin. Issa jiena ma bghatejtx daqshekk ghax ifhimni, affordjat li nimxi step by step. Ehe…miskin min ma jkollux* [flus] *ha jbati hafna izjed. Hafna izjed"* (Elijah, p.12, lines 313-321).	*"And another thing, I did not feel it* [the financial blow] *because I was self-employed. So it* [ALS] *can affect you financially. It did not affect me because this is all my work. These things are all expensive. Now I did not suffer financially that much because I could afford to do things step by step. Ehe…the poor soul who does not have the financial means will suffer a lot more. A lot more."*

In summary, all five participants described in detail the initial shock and disbelief of being diagnosed with ALS, the frustration of losing their independence, their fear of being a burden to their loved ones, the emotional distress at seeing their bodies slowly deteriorating and the impact such a debilitating and terminal neurological disease leaves on the individual and their loved ones.

4.4: Striving To Stay Afloat

Striving to stay afloat is the second super-ordinate theme extracted from a participant's transcript who metaphorically describes
that being diagnosed with ALS is a *ticking time bomb* even though he is facing it with tremendous courage and strength. The other four participants also admitted that diagnosing ALS is difficult to apprehend. However, they all described their coping mechanisms and striving to overcome the emotional rollercoaster of dealing with a fatal, neurodegenerative and physically disabling disease. All five transcripts showed a strong sense of gratitude in the face of a terminal disease.

4.4.1: Adapting To The Situation

Despite the physical and psychological suffering ALS causes to whoever is afflicted by it, the participants still managed to find peace and serenity within themselves. Sophia, for instance, commented that although at times a sense of sadness does come over her, especially when she sees how much weight she is losing, she is still a happy person at heart:

"I can't say I'm not a happy person because I'm still happy ok?" (Sophia, p.7, lines 201-202).

Although the participants required assistance from their caregivers, their sense of selflessness was reflected clearly by thoughts of others' safety, the help they gave to various charities and support to individuals diagnosed with other neurological disorders such as multiple sclerosis (MS) and Huntington's disease (HD). Furthermore, each participant managed to find their way of finding peace amid such a progressive illness, even though, at times, they did admit to the voyage ahead being a tough one:

"I lie on the bed, I read, I watch television, I do crosswords and things like that so, you know, that's fine." (Isaac, p.7, lines 189-191).

"Le, le. M'ghandiex l-ebda rabja. Inhossni ghandi serena hafna eh! Hafna eh! U paci." (Elijah, p.15, lines 419-421).	*"No, no. I do not feel any anger. I feel very serene, eh! A lot, eh! And at peace."*

A deep sense of gratitude that varied in reasons was evident throughout the interviews of all participants. Furthermore, the researcher noted that one participant was grateful that he did not have cancer and did not have to undergo chemotherapy. However, his illness was fatal and very progressive. The same participant, the only one with familial ALS, stated that he was grateful that he did not yet succumb to ALS as early as his father did. Additionally, two other participants were grateful that they were diagnosed later in life and not at a young age, thus, giving the impression that at least they got to

enjoy life as much as possible. The following excerpts below show the participants' gratitude from different perspectives:

"I mean, I suppose it's frustrating not being able to do things you've done all your life without thinking about it. But, you know, there are worse things, frankly in the world. I, you know, I see friends who have cancer and go through chemotherapy, and they don't always get better, so I have a lot to be grateful for." (Isaac, p.8, lines 220-224)

"I'm rather glad, I mean, I'm, I'm nearly 73 now, so I am lucky. My father got it when he was in his 50's." (Isaac, lines 47-51, p.2).

"Issa tghidli ma kontx taf ghalxiex diehel? Imma x'naghmel? Tghid ghallinqas ma ghandiex 20 sena, imma ghandi kwazi 60. Tghid hekk. Heqq." (Benjamin, p.5, lines 137-144).	*"You may tell me, didn't you realise what you are getting into? But what can you do? At least I am not 20 years old, but I am nearly 60. You say that. Heqq."*

4.4.2: Be It Done Unto Me According To Your Word

Elijah was the only participant who showed great spiritual and religious faith despite his dire situation. He spoke at length about how he found solace in facing his crippling disease by placing his faith in the Holy Mary and Jesus Christ. Furthermore, he explained how he became frustrated and could not accept specific discourse when his close friends felt sad at how, out of all people, he had been the one to be diagnosed with ALS. Elijah also described his one sorrow ever since being diagnosed with ALS, that he can no longer participate in his beloved village feast. The following excerpts will show how Elijah used to hope and religious faith as a coping mechanism and how he felt thankful for all his blessings:

"Jiena ha nghidlek, jiena tkellimt ma qassisin u ha nghidlek ta, jiena ma nitlobx biex infieq. Le. Jiena nirringrazzjah [Alla]. *Stajt hadtha*	*"Let me tell you, I spoke to priests and let me tell you, I do not pray to be healed. No. I thank Him* [God]. *I could have taken it against her* [Holy Mary]. *But no."*

kontriha [Holy Mary]. *Imma le."* (Elijah, p.15, lines 413-416).

Jiena nhobb il-Madonna u ghidt isma, il-Lunzjata [Patron Holy Mary of the local village feast] *x'qalet? 'Ha jkun minni skont kellmtek'. Daqshekk kienet easy ghalija (...). Imbaghad qalli* [treating neurologist] *li harget l-ALS. Biss imma eh, hassejt li l-Bambin hadli haga u tani hafna affarijiet ohra imma eh?"* (Elijah, p.8, lines 206-226).

"I love the Holy Mary, and I said, listen, what did the Lunzjata [Patron Holy Mary of the Village feast] *say? 'Be it unto me according to Your Word'. That's how easy it was for me (...). Then he* [neurologist] *told me that I have ALS. But eh, I felt that Jesus Christ took something away from me but gave me so much more in return though, eh?"*

Contrary to Elijah's evident religious solid faith, Benjamin became very frustrated and bitter during the interview towards the Catholic Church and faith in general. However, this occurred after he was diagnosed. He claimed that religions are only a human necessity to ensure hope in a dying person. Furthermore, he derided individuals who placed their faith in their religion and called them 'weak' and 'gullible'. Benjamin also showed resentment towards anyone within the catholic hierarchy who dared to judge him:

"Ehm, ricenti jiena kont bniedem kredenti hafna. Up to qisu 2013. Pero kredenti bhala kattoliku. Imbaghad bdejt qisni nara t-tfal ma jimpurtahom xejn u hafna quddies artificjali u bdejt nirrejalizza li 'its all made up'. (...). Jigifieri r-religjonijiet gew neccessita' tal-bniedem bhala assigurazzjoni u tama ta; meta jmut." (Benjamin, p.4-5, lines 114-122).

"Ehm, recently, I was an individual who harboured great religious faith. Up to 2013, I would say. I was a Catholic believer. But then I started seeing the children not caring about anything and a lot of artificial church masses, and I started to realise that it is all made up (...). So religions became a necessity of humans to assure them hope when they die."

“Issa min ikun weak joqghod jibza u jemmen f’kollox, min ikun accetta l-affarijiet. Issa, issa li hemm hemm u jiena had mhu ha jigi jghidli li int ghamilt dnub u ghall-infern. Nghidlu mur ixxejjer ghax jiena dawn l-affarijiet ma tiggudikaniex fuq hekk.” (Benjamin, p.5, lines 123-128).

"Now, there are those who are weak, fearful and gullible and those who accepted the situation. Now, what there is, there is, and no one is going to tell me that I sinned and am sentenced to hell. I will tell him to go and get lost because you cannot judge me on such things."

Accepting their diagnosis was another sub-theme that emerged strongly throughout the interviews. Although ALS can be considered a 'ticking time-bomb', they still accepted the illness with courage and a will to 'fight'. Metaphorically speaking, Elijah described how the best medicine one can take for a diagnosis that is so dire is, by far, acceptance:

“Nahseb li l-aqwa medicina hi li meta taccettaha [ALS]. *Taccetaha eh? Trid u ma tridx. Ikollok taccettaha. Issa jekk ma taccettahiex ha tbati izjed. Meta accettajtha jiena hassejtni hafna ahjar* (Elijah, p.16, lines 448-453).

"I think the best medicine is when you accept it [ALS]. *You accept it, eh? Whether you like it or not. You have to accept it. Now, if you do not accept it, you are going to suffer more. When I accepted it, I felt so much better."*

During the interview, it was evident that Sophie's fulcrum of support and protective factor was her family. Whereas at certain moments, the researcher wondered if she was in denial of her situation, Sophie spoke lovingly about her four grandchildren, her children and her husband and mentioned that she 'fights' and accepts her situation for them.

"Well listen, ehm, I have a family, ok? And I have four grandkids, ok? So, yes, I am fighting it [ALS] *for them, I guess. You know what I mean?"* (Sophie, p. 4, lines 98-101).

Benjamin, who throughout his interview seemed despondent, also stated that he accepted that being diagnosed with ALS was something he had no control over and refused to make a big deal or fuss about it:

"Ma ppanikkjajtx daqshekk le. Ma ppanikkjajtx. I mean mhux ma kontx naf, imma accettajt." (Benjamin, p.2, lines 36-38).

"I did not panic that much, no. I did not panic. I mean, it is not as if I did not know, but I accepted."

4.4.3: Facing ALS With Humour

For individuals facing death from a debilitating disease that at end-of-life will render them unable to breathe unaided, the emergence of the sub-theme of finding time to joke about certain situations in their life was unexpected. For Elijah joking about himself and the situations he finds himself in was a coping mechanism and made him feel positive. Furthermore, Benjamin also felt better at joking about a situation that, although was not at all funny, made him feel better:

"Ghax jiena bqajt niccajta. Le, inwaqqa lili ghac-cajt signifiers. Perezempju, das-sajf konna, kien hemm, ehm…midhla hekk u kien hemm wiehed gab flixkun inbid minn dak tal-ghasir. U qallu dan, qallu 'ticcajtax mieghu' ta qallu 'ghax jaqtalek saqajk'.Ghidtlu mela jiena xejn ma nhossu! Ghidtlu ghalija tajjeb. Ghidtlu ghax jiena ma nhossux dak! U harsu lejja hekk jigifieri." (Elijah, p.17, lines 476-485).

"Because I continued to joke. No, I joke about myself; I meant. For example, this summer, we were at a gathering, and one guy brought a bottle of homemade wine. And he said, 'Do not joke around with this because you will not feel your legs'. I told him, 'Well, I will not feel it (the wine); for me, it is good then because I do not feel anything'. They just stared at me."

"Ehm…I joke about it kultant. Ehe…fis-sens x'taghmel u?" (Benjamin, p.2, lines 50-51)

Ehm…I joke about it sometimes. Ehe…I mean, what can you do?"

4.4.4: Support In The Face of A Devastating Diagnosis

While conducting her interviews, the researcher noticed that all five participants had a robust support system from various entities. All participants had families, friends and colleagues who supported them in every aspect, encouraging them to deal with their diagnosis much better. Only one participant felt saddened at not having the full support of his wife and brother, who could not come to terms with his diagnosis. However, his daughter, who has to keep up with her studies at University whilst being his primary caregiver, compensates for his wife and brother:

"Le, xorta hi [tifla] *taghti hafna hin. Ifhimni, inti, x'jismu, inti, le tinduna. Tinduna. Tghidli inti mintiex qieghed sewwa. Ghax hi li taghmel, ha nghidlek, taghmlu ghax missierha, imma taghmlu ghax qed tara b'ghajnejha u ghax mill-qalb. Meta tkun professjonali hemm differenza."* (Elijah, p.18, lines 494-500).

"No. She [daughter]*still gives a lot of time. Understand, she notices. She tells me that I am not feeling right. Because what she does, she does it because I am her father but also because she sees with her own eyes, and she does it from the heart. When you are a professional, there is a difference."*

"I mean, I'm dealing with it. I do. have a very strong support (family). The most important thing I have a very strong support." (Sophie, p.5, lines 115-117).

"It is literally happening to someone close to you, but then like everything in life, everyone gets used to it and tries to help you out in the very possible way. That is what happened" (Joshua, p.2, lines 28-31).

Apart from the support from their loved ones, all the participants mentioned receiving impeccable care from their treating neurologist and healthcare professionals at MDH. Being told that they have ALS was difficult for every one of them, but knowing they have full support from MDH made the burden easier to carry:

"Eh…(treating neurologist) *is fantastic. Very supportive. Ehm…I am very pleased that he is my person."* (Isaac, p.8, lines 203-205).

"Then I went to see (treating neurologist) *and ehm… that's where it all started (…). But I Had told him about my fingers, and I think, yes. But as soon as I told him that I had very weak fingers and what have you, il-bravu li hu… yes*." (Sophie, p.2, lines 34-49).

At the time of writing, two participants were residents of Dar Bjorn. This residential home caters to people with ALS, MS and other neurological conditions, whereas the other three still live in their own homes. Most of the participants showed deep gratitude towards Mr Bjorn Formosa, the founder of ALS Malta, for their constant ongoing support and for his relentless work in building residential homes that catered for their specific needs:

"I have met Bjorn Formosa, eh… who's a wonderful person. I admire the work he has done (…). I, I, there's people from ALS Malta who come and see me. They're very kind and helpful and supportive, and if we need any help or information, not just me but Matthew as well, we can ring them. We know who they are, they know who we are and that, that is great." (Isaac, p.7-8, lines 193-203).

Apart from support from his family, friends and healthcare professionals, Elijah also mentioned how he finds comfort and solace in his daughter's dog, a tiny chihuahua. His voice breaking with emotion, he explained how the dog, who was only a little puppy at the time, somehow sensed that he was unwell because she used to cuddle up next to him and constantly lick his forearms and toes:

"Ghandi kelba ta' din [tifla], *jigifieri tiskanta fil-bidu. Fil-bidu kienet kienet tilghaqli, kienet puppy eh? Tilghaqhom eh?*[swaba tas-saqajn u idejn] *Hafna. Fuq idejja imbaghad. Waqfet issa. Tmur*	*"I have a dog of her* [daughter]. *You will be amazed. In the beginning, she used to lick me. She was a puppy, eh? She used to lick them* [toes], e*h? A lot. On my arms, then. She has stopped doing it*

fuq is-swaba mill-ewwel. Qisha kienet taf!" (Elijah, p.18, lines 504-509).	*now. She goes directly to the toes. It is like she knew!"*

To summarise, the super-ordinate theme, 'A ticking time bomb'taken directly from one participant's transcript, describes the day-to-day challenges individuals diagnosed with ALS face and the sufferance of seeing their bodies deteriorate progressively. Emergent themes discussed the support received from family, friends and healthcare professionals, using religious and spiritual faith as a coping mechanism, acceptance of a terminal illness, the importance of humour and positivity, and how they adapt to their situation.

4.4.5: Living The Present

Whilst conducting the interviews, one aspect that emerged constantly was the courage and positivity that emanated from the participants, even though, as Joshua commented, *"there is probably nothing worse that could happen to a person." (Joshua, lines 70-71 p.3)*. Joshua continued to explain that although being diagnosed with ALS is difficult to accept, if one concentrates on keeping occupied and tries to fill his/her days with enjoyable activities and/or work, then great things can still be achieved:

"It may be difficult, but if you put your head down to it, you can manage to achieve great things. Even though the condition is so difficult." (Joshua, p.3, lines 75-79).

Elijah, who had spent a long time going from one healthcare professional to another in search of answers and seeking help for his sudden, debilitating symptoms, faced his diagnosis with great courage and strength:

"Anke shabi, min jitkellem mieghi. Hekk jghiduli. Eh…kif jiena ghandi dan il-kuragg." (Elijah, p.11, lines 308-310).	*"Even my friends, those who speak to me, that is what they tell me. Eh…How I find this courage."*

On the other hand, Sophie explained how she would not go down without a fight. Still, in the early stages of ALS, she is adamant that she wants to live in the present and face each day with courage whilst enjoying life with her family as much as possible:

"So that's it like I said. I'm living day by day, and I make the most of it. I make the most of life. We only live once. That's it. I make the most of it." (Sophie, p.8, lines 209-213).

Sophia's only symptoms during the interview were weak fingers and reduced dexterity since her fine motor skills had been significantly impacted. She was aware of this issue. However, she decided to do her utmost not to give up fighting:

"I do not cook anymore. He (husband) does the cooking. But you know what? I am going to say it out straight. Ehm…I am fighting it, ok? I'm not going to let this…ehm…you know. Take me back, so I am fighting it. You know? I am doing as much as I can and what I can." (Sophia, lines 86-93, p.4).

The five participants revealed what made them happy and enjoy life, even after being diagnosed with ALS. For instance, both Sophia and Isaac loved to travel and go out to restaurants, whereas Elijah loved to spend time in his mother's boathouse near the sea:

"Eh…But I said to myself, well, I cannot do anything about it, so I've just got to get on with life and make the best of it frankly and eh…enjoy every day. Which I do. Yes, yes, we travel a lot." (Isaac, p.3, lines 62-66).

"Yes, I'm coping with it. And the quality of life, we travel, we go out to dinner, we…as normal. As normal. Why not? I mean, you know what I mean? As normal. I'm living the present." (Sophie, p.5, lines 131-137).

Four participants did their utmost to remain cheerful and occupied, and busy. Isaac and Joshua remained very active while Elijah continued to run his business. Sophia, a house-wife, filled her days with her family and did her best to remain as active as possible:

"I fill up my days, and I work, and I work a lot, and I try to produce as many things as I can." (Joshua, p.2, lines 45-47)

"Yes, well, I mean I am quite a strong person, eh? I am still going to my charitable things [voluntary organisations]*"* (Isaac, p.6, lines 145-146).

4.5: Conclusion

This chapter presented two super-ordinate themes: *A Ticking Time Bomb* and *Striving to stay afloat.* Moreover, an arduous voyage emerged from the data collected by the researcher from semi-structured interviews of patients diagnosed with ALS. The first super-ordinate theme, a ticking time bomb described the impact ALS had on the participants. The second super-ordinate theme, striving to stay afloat, described how the participants coped and dealt with knowing they had been diagnosed with not only a fatal illness but one which would rob them of their independence in the cruellest and most debilitating ways.

Chapter 5: Discussion

5.1: Introduction

Chapter five discusses the findings of this research study which focuses on the lived experiences of five individuals diagnosed with the neurodegenerative disorder ALS. Both the implications and the findings that originate from the study will be described in the light of existing literature and within the context of two frameworks, namely, the Common Sense Model (CSM) (Leventhal, 1980) and the Person-Centred Practice Framework (McCormack, 2017). Moreover, keeping in line with the guidelines of the IPA methodology (Smith et al., 2009), the researcher's interpretation of the study will be considered, thus, leading to the amalgamation of the research findings, extant literature and the researcher's interpretation. Likewise, the qualitative nature of this study may prove helpful in providing an in-depth account that may strengthen and guide healthcare professionals into further understanding the consequences of this severe disease.

As highlighted in the literature review of this study, an evident paucity of literature on the lived experience of individuals diagnosed with ALS exists, and such a phenomenon has rarely been investigated. The researcher did encounter an abundance of literature on ALS. However, these were either quantitative, addressing the molecular genetics of ALS or concentrated mainly on the QOL and the impact of ALS on the patient and their primary caregiver. The dearth of literature is even more evident from a local perspective, whereby the only studies encountered by the researcher addressed ALS's genetic and occupational aspects. The researcher also went through the dissertation database of the UM library in search of studies that focused on ALS. However, only one was found which addressed the suffering and the request for assisted suicide by ALS patients (Said, 2018). Hence, exploring the lived experience of individuals diagnosed with ALS may provide further insight and awareness of such a progressive, degenerating, terminal neurological disorder's impact on the afflicted individual. Moreover, utilizing her study on this particular cohort of patients, the researcher is targeting one of the vital person-centred processes; understanding the narratives or voices of persons that encompass their concerns, emotions and challenges of living with ALS.

Table 5.1 compares findings retrieved from the literature as presented in Chapter Two of this study with the findings of the present study extracted from Chapter 4. For ease

of reference and to facilitate comparability, findings extracted from the retrieved literature will be aligned opposite to similar findings obtained from this study. However, when data was extracted in the international OR local study, only that will be listed under the relevant column.

Table 5. 1: Comparison of findings presented in the literature review with findings of the present study emerging from the superordinate theme 'A tough voyage'.

Findings extracted from the literature review	**Findings obtained from the present study**
The impact of being diagnosed with ALS	The following themes emerged under the super-ordinate theme **'A ticking time bomb'**
As ALS progressed, the participants were aware of changes in their body. Seeing themselves become weaker, thinner, walking in a strange manner and suffering from sialorrhea left a profound impact on the way they perceived their self-image (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	All participants were distressed about the rapid progression of ALS and felt disheartened at the loss of limb function and the way their bodies were deteriorating, thus experiencing reduced feelings of self-worth.
Enjoying healthy lifestyles, the participants described, how at onset of ALS all they experienced were minor health issues which had made them go to their healthcare provider (Oh, Schepp &McGrath, 2014).	The participants recounted how initially, their symptoms seemed minor and at no point did they associate them with something as serious as ALS or any other neurological disorder. The minor health issues prompted them to seek medical advice.

When their physical function started to decline and they lost the ability to move, communicate, wash themselves and lead an independent life, the participants experienced a loss of autonomy (Kukulka, Washington, Govindarajan & Mehr,).	All the participants also felt frustrated at the inability of doing the most basic of things such as feeding themselves, tending to their personal hygiene, becoming paralysed and losing their independence.
Some participants started to isolate themselves on purpose from others out of shame and embarrassment as ALS progressed, limiting their functionality (Kukulka, Washington, Govindarajan & Mehr, 2019).	
An association of ALS with death and dying was noted, however, participants showed a sense of ambivalence in the fact that while they wished to be cured, they still experienced suicidal ideations as the disease progressed (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	One participant broached the subject of euthanasia during the interview but, was not ready to choose that option for himself out of respect for his family. However he still mentioned that if the situation ever became too much for him to bear, he can resort to refusing nutrition and refusing to be placed on a ventilator, thus leading him to his demise.
Whilst their muscle weakness and stiffness became worse, some of the participants stated that they may as well watch themselves wither away and die (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	
It was difficult for the individuals to accept a diagnosis of a rare disease like ALS, at times even convincing themselves that they	Two of the participants could not fathom out how they were diagnosed with a rare neurological disease like ALS since they

may have been misdiagnosed (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	both took great care of their bodies and worked out regularly.
Having to be cared for by their primary caregivers caused feelings of guilt and fears that they are becoming a burden (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	Most of the participants were concerned about becoming a burden to their loved ones in different ways. One participant worried about his spouse having to give up a professional career to care for him, another felt guilty because his daughter had to juggle her studies at UM whilst caring for him and another worried about becoming a burden to both her husband and her children.
Participants opted to keep their concerns and sufferance to themselves rather than place added worries onto their caregivers (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	One participant, who although was diagnosed with the sporadic form of ALS was worried that, at some point, the disease will reappear in his family. He felt he could not discuss this with his wife for fear of worrying her unnecessarily.
Some participants went into denial and found it difficult to accept a diagnosis of ALS (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	One female participant portrayed a sense of denial when during the interview she commented that she was willing to fight ALS even if she reaches 80 years of age. Being in her 50's, living till the age of 80 with a diagnosis of ALS is not possible due to its rapid progression and fatal outcome. One participant did not mention the word ALS during the interview, referring it to 'this' or 'that', whereas another participant did not want to receive any information on the symptoms and/or prognosis of ALS. The reason here could be considered

	thought suppression, whereby allowing themselves to think about ALS may provoke feeling of anxiety and stress.
Participants became frustrated and unhappy as the disease progressed because of their physical decline and reduced QOL, although they fully accepted that ALS is progressive. Feelings of bitterness, anger and sadness are among the emotions felt when they realised that they cannot do the same things they did before (Oh, Schepp &McGrath, 2014).	All the participants, at some point were affected negatively, when they could no longer enjoy the normal daily activities that one takes for granted, such as going shopping, playing a sport, attending mass or going to work. They felt frustration at a body that no longer functions. Three of the participants who were at various stages of paralysis knew that they will soon be rendered completely paralysed. One participant, who placed his faith in science felt hopeful that a cure for ALS will one day be found. Another participant placed his hope and faith in The Holy Mary and God.
In the face of a disease that to-date has no cure, they considered hope as an unnecessary obstacle since they knew that being diagnosed with ALS was a certain death sentence, so being filled with hope was like lying to oneself (Hamama-Raz, Norden & Buchbinder, 2021). Participants wanted to make the best of the life they had left however they avoided to discuss death. Although this may show optimism and may have proved beneficial in some aspects, it may also be due to repression of thoughts in order to not provoke anxiety. Furthermore, not	

broaching the subject of death may have lacked understanding and significance of what a 'good death' actually means (Oh, Schepp & McGrath, 2014)	
	Only two participants mentioned death as something inevitable in ALS and seemed prepared and informed about what to expect at the latter stages.
The financial burden was not that significant as long as the total family income was high or the patients were covered by a health insurance. Different cultural beliefs, especially in developing countries also viewed financial burdens from different perspectives (Geng, Ou, Miao, Zhao, Wei, Chen, Liang, Shang & Yang, 2016).	Only one participant mentioned the financial burden ALS may have on a family if they happened to be financially unstable. Malta cannot be compared to the findings in the source of the literature found, since healthcare in our country is free, however the participant did emphasise that certain necessities and commodities, such as comfortable wheelchairs and restructuring the residence to make it more accessible are highly expensive. He also mentioned that being diagnosed with ALS will eventually lead to loss of employment so that in itself will place a financial burden on the family.
It reflected clearly how different cultures dealt and reacted with a diagnosis of ALS in different ways. South Korean and Chinese cultures do not emphasise on autonomy. The family hierarchy, whereby the father automatically takes on the role of head of family, is thrown into disarray and confusion if the father is diagnosed with a disease such as ALS that will make him reliant on his	

primary care-giver (Oh, Schepp &McGrath, 2014).	One participant showed a fear of becoming depressed and reliant on anti-depressant medication. During the interview, he had admitted that apart from having to deal with his own illness, he also had to deal with his wife's mental health issues too, thus placing a double burden on him.

Table 5. 2: Comparison of findings presented in the literature review with findings of the present study emerging from the super-ordinate theme 'Striving to keep afloat'

Findings extracted from the literature review	**Findings obtained from present study**
Coping with ALS	The following themes emerged under the super-ordinate theme **'Striving to stay afloat'**
Accepting a diagnosis of ALS was considered crucial for the participants well-being and encouraged them to embrace their value and meaning of life. Many took to comparing themselves with others who were in worse situations than they were thus allowing themselves to move from 'victimhood to survivors' (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	Two participants felt gratitude at being diagnosed at a later age, namely 73 and 60 years of age respectively. One of the participants, who was diagnosed with the familial form said he felt grateful about this since his father, who also had ALS succumbed to the disease when he was in his 50's.
Participants felt the need to remain occupied and helpful by devoting their time in helping others. One participant expressed his wish to donate organs as a way of continuation of life (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	The majority of participants, although two were predominantly home-bound, kept themselves constantly busy as a means of coping with their disease and ensuring that they get the most out of the days they have left. One participate involved himself in

	helping international charities, whilst two others worked from home. The female participant kept as active as possible and tried to help out around the house as much as possible.
Introducing mindfulness as a cognitive approach to individuals diagnosed with ALS, may help to slow down its progression and promote psychological well-being (Pagnini, Phillips, Bosma, Reece, & Langer, 2014)	
Participants described hope as a way to cope with their diagnosis and help them continue with their activities and keep up their spirits (Hamama-Raz, Norden & Buchbinder, 2021).	All participants showed courage and a will to keep on fighting. Keeping busy helped compensate for their reduced QOL.
One participant, who relies deeply on his religious faith, described hope as a ray of light that emerges from a tunnel, a metaphorical expression for describing ALS as the tunnel and the light as hope. Placing his faith in God, he believed that nothing in life is accidental (Hamama-Raz, Norden & Buchbinder, 2021).	One participant described how his strong religious faith is what gives him hope and strength to deal with his illness. He feels grateful that ALS has not taken away his blessings in life and that he feels peace and serenity.
Participants described being greatly supported by their families and children and how they strived to cope (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	Out of the five participants, three had children. The female participant, who also had grandchildren stated that the love for her whole family gives her the courage to fight. She also describes her sadness at no

Participants felt that living in the present and utilising the days left to the full helped them forget about their illness for a while (Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang, 2021).	longer being able to care for her young grandchildren. All the participants showed courage, strength and resilience and spent their days living each moment to the best of their ability.

The next section will briefly describe the Common Sense Model (Leventhal et al., 1980) and the Person-centred Practice Framework developed by McCance and McCormack (2017), which are the theoretical frameworks chosen to interpret the study findings.

5.2: The Common Sense Model

The Common Sense Model (CSM) will be additionally incorporated in this study to discuss the participants' psychological representations of their experiences dealing with ALS, a fatal and neurodegenerative disease. The CSM (Leventhal et al., 1980) is a psychological model that presents the health processes that help individuals cope with chronic illness. When people are confronted with a new health threat, they create patterns of beliefs and symbolic meanings about the situation at hand (Leventhal et al., 1980). They then try to comprehend what the threat is by forming cognitive representations (Leventhal et al., 1980). The threat in this case being a diagnosis of a fatal illness which is incurable. These cognitive representations may also be influenced by the experiences of other individuals who have also faced a similar threat (Petrie, 2006). Furthermore such symbolic images may be influenced both by culture and the media (Leventhal, 1996). Table 5.3 below describes the six domains of illness representations:

Table 5. 3: The six domains of illness representations

<u>Identity</u>	The label of the health threat (e.g., ALS) and the symptoms associated with it (e.g., fatigue and muscle weakness)

Cause	The individual tries to establish a reason for the health threat in a bid to find a meaning. The majority of the participants, with the exception of the patient who was diagnosed with the familial form of ALS, could not understand why such a rare illness was inflicted upon them.
Timeline	The perceived time-frame of how long the illness will last (e.g., chronic)
Consequence of the illness	The perception of how the illness is affecting the individual with ALS. The findings of this study show that the most common consequence was their loss of independence.
Curability/controllability	The notion that the threat can be either be cured or kept under control by themselves or others. In this case ALS is an incurable disease, thus the participants opted to remain active for as long as possible and deal with the symptoms when they appear. Furthermore, Petrie et al., argue that the perceived control over an illness depends on two types of control beliefs, namely, treatment control belief (whether or not the illness can be controlled by medication) and personal control beliefs (whether the patient can help him/herself contribute to controlling the illness).
Illness coherence	The knowledge and understanding the patient has about his/her illness. Moreover, the CSM (Leventhal et al., 1980) states that whenever there is a

	perceived health threat, the individual elicits an emotional response, hence, in an attempt to reduce this response, the individual resorts to coping mechanism. This aspect was clearly highlighted in this study when participants used humour directed at themselves and their paralysis as a means of relief in such dire circumstances.

Moreover, the CSM (Leventhal et al., 1980) states that whenever there is a perceived health threat, the individual elicits an emotional response; hence, to reduce this response, the individual resorts to a coping mechanism. This aspect was highlighted in this study when participants used humour directed at themselves and their paralysis to relieve such dire circumstances.

5.3: Person-Centred Practice Framework

The Person-Centred Practice Framework developed by McCance and McCormack (2017) will be incorporated with the CSM (Leventhal et al., 1980) in the discussion of this chapter. The researcher opted for this particular framework due to its significant relevance to humanistic caring practices and the complexity of person-centred care. Such a framework corresponds perfectly to this study since the dissertation focuses on individuals facing a fatal diagnosis of a rapidly progressive disease. Although a person-centred practice framework should be integrated into every situation, dealing with the severity of a condition such as ALS requires an even higher standard and competence of healthcare professionals since they successfully manage various contextual and attitudinal factors at the heart of a caring interaction. By applying the person-centred practice, healthcare professionals, in particular mental health and palliative nurses, may prove invaluable in cases where a diagnosis of ALS is given, thus having the potential to ensure that support and commitment are delivered to such individuals. Furthermore, for healthcare professionals to be better equipped to provide

person-centred care, a number of prerequisites, including competence, interpersonal skills and the belief that the care they provide will leave a positive impact on their patients.

5.4: The Lived Experience of Individuals Diagnosed With ALS

The lived experience of individuals diagnosed with ALS will be discussed in two introductory sections, namely the impact of being diagnosed with a terminal and progressive neurodegenerative disease and how the individuals strived to cope with their illness.

5.4.1: The Impact of Being Diagnosed With ALS

The five participants in this research study courageously accepted to be interviewed and openly discussed the intense emotions, incomprehensible shock and disbelief when, after a battery of medical tests and imaging, they were diagnosed with ALS. This rare disease to date is incurable. They discussed how life was suddenly turned upside down for them when from strong, healthy and able individuals, in a short matter of time, they had succumbed to a progressive paralysis that in one participant (Joshua) rendered him unable to breathe and eat on his own. A common factor noted amongst all participants was how, unbeknownst to them, the vague symptoms they were suddenly experiencing (reduced dexterity in the fingers, clumsiness, frequent unexplained falls, weight loss) were the onset of ALS, hence the shock when they were finally diagnosed. One participant (Isaac) recounted how it was only after one nasty fall where he injured himself that he decided to seek medical attention. Another participant (Sophia), the only one in the early stages of ALS, explained how the only symptoms she experienced were reduced dexterity in her fingers and unexplained weight loss. In an ethnographic study by Oh, Schepp & McGrath (2014) which discussed the experience of patients living with ALS from South Korea, their findings were consistent with the study by Yuan et al. (2021) and this research. In the ethnography by Oh (2014), the participants similarly explained how they had always

maintained a healthy and active lifestyle and only ever experienced minor health issues at the onset of ALS. Only when the symptoms started interfering with their abilities to perform basic tasks, such as experiencing a weak hand grip or repeatedly dropping dishes, did they seek help from their healthcare providers. In the study by Oh et al. (2014), the participants additionally stated that the rarity of ALS made it even more difficult for them to accept such a disease. They went on to explain how, although they understood the physical limitations and symptoms of ALS, they could only really comprehend the devastating trajectory of ALS when they met other patients that were in the advanced stage of the disease and were respirator-dependent. These participants also expressed a fear of death and an inability to speak.

Furthermore, a participant in the study carried out by Oh et al. (2014) explained that if she ever became respirator-dependent, she would simply kill herself by removing the respirator because her life would be meaningless. Additionally, a participant in the study by Yuan et al. (2021) explained how she frequently dreamt that she was cured but, on awakening, felt her 'stiff limbs'. She went on to say that, at times, she becomes so desperate that she wishes she could end it all. Such reasoning from the participants of the three studies showed a sense of ambivalence. On the one hand, they wish they could be cured; on the other, they wish to end their lives. This statement was completely consistent with what one participant (Benjamin) in this study stated. He was the only one out of five participants who brought up the subject of euthanasia. When asked if he would ever consider it, he declared that accepting to be euthanised would cause considerable grief for the patient's loved ones. In contrast, if he (Benjamin) ever felt that ALS was becoming too much to bear, he could refuse nutrition or remove his respirator. The thoughts of the two participants from different studies, one in South Korea and the other from Malta, showed how the suffering of being diagnosed with ALS affected everyone relatively in the same way, regardless of their background and/or culture. However, according to Lin (2003), a phenomenon of Chinese culture was the avoidance of speaking about death since the Chinese people's view on life and death is influenced by Confucianism, which was confirmed in their study. Confucianism places great importance on life but avoids the aspect of death (Chen, 2017). This lack in mentioning of death was also consistent in this present study. In the present study, only two participants (Benjamin and Joshua) showed the researcher that they knew that ALS was terminal and spoke outright about death.

Another participant (Isaac), who was diagnosed with the familial form of ALS, and was very knowledgeable about the prognosis, asked the researcher not to mention anything about the symptoms of ALS or what may happen to him further down the line because he did not wish to think about it. He explained how he knew what to expect because his father had succumbed to ALS. However, he would rather dismiss it in his case and live in the moment. The researcher respected his wishes and was careful not to delve into areas she knew would distress him further. One participant in the study by Oh et al. (2014) similarly stated that he knows that he will be experiencing the loss of a normal future. Thus, he prefers not to think about any future events. According to Bolmsjo (2001), this phenomenon, known as existential suffering, which includes loss, anxiety about the future and a wish to die, is commonly experienced in patients with life-threatening diseases. Moreover, although the patient may have accepted his illness and even learnt how to cope, the existential aspect of suffering may never end. The remaining two participants (Elijah and Sophia) never spoke about a fear of death. Contrarily, they kept on insisting that they would fight it, even at a later senior age, which made the researcher ponder whether they were in denial or if they lacked information about the illness since both were in their early fifties and that they would never live to see their senior years.

In the qualitative study on the illness experience of people diagnosed with ALS, carried out in Wuhan, China, by Yuan, Peng, Zeng, Wu, Chen, Zhang & Wang (2016), the participants were highly aware of the changes in their bodies after being given the diagnosis. They described the embarrassment of constant drooling (sialorrhea), their strange gait and how they became thinner, thus causing them to estrange themselves from others. Moreover, they stated how they felt like a freak and preferred to be alone. These findings were consistent with a study by Kukulka, Washington, Govindarajan & Mehr (2019), whereby the participants also found themselves steering away from social interaction due to embarrassment regarding their functional limitations. The study participants mentioned the frustration of seeing their bodies changing so rapidly, especially the female one (Sophia). She commented on how she led a very active lifestyle and took great pride in her body and appearance. She explained how it was not the weakened dexterity in her fingers that bothered her but seeing her body change due to the rapid weight loss she was experiencing. She admitted to being rather 'vain' about her physical appearance. Therefore, it saddened her to see her body waste away.

Interestingly, none mentioned they were ashamed to be seen in front of others. Contrarily, one participant (Elijah) stated that when the time came that he could no longer hold a spoon in his hand because it tired him, he felt no shame whatsoever in being fed in front of his colleagues at work or in front of strangers in a restaurant or even seen in his wheelchair. Likewise, another participant (Isaac) expressed his frustration at overhearing a house guest comment on his disability and adamantly stated that he wanted to be seen as the person he was before being diagnosed with ALS and would not tolerate being considered 'the particular person sitting in the corner'. Interestingly some of the participants in the study by Yuan et al. (2021) compared themselves to a 'rusty robot' that is slowly being destroyed by disease and pain. This comparison clearly explained how ALS manifests in muscle weakness and stiffness as common symptoms.

Whereas the participants in this present study never doubted their treating neurologists, the ones in the study by Yuan et al., at times, did believe they had been misdiagnosed since a diagnosis of ALS, apart from being rare, is very difficult to come to terms with and accept. The five participants in this study discussed how they had to learn to accept their diagnosis; otherwise, it would take an emotional toll on them, rendering them unable to cope. In both this study and the extant literature retrieved, the participants discussed how acceptance was the first crucial step they had to take to commence their challenging journey of meaning-making. In the study by Yuan et al., most patients appeared to take a 'downward comparison approach' to further accept their diagnosis by comparing themselves to individuals in a far more advanced stage of ALS than they were at that time. This indicated a shift from 'victimhood' to 'survivors'. Interestingly, this phenomenon was also encountered in this study. Two participants (Isaac and Benjamin) felt 'gratitude' that they were diagnosed with ALS at 73 and 60, respectively, and not at a younger age.

The five participants in this study mentioned one common factor: how ALS robbed them of their independence and how it rendered them dependent on their caregivers. One participant (Joshua) explained how from being a non-disabled man with a penchant for sport, he became confined to a bed and could only breathe via a ventilated tracheostomy. The female participant (Sophia) became emotional when she explained how she adored caring for her grandchildren. At the same time, their parents went to work but had to stop due to the weakened dexterity in her fingers. She could no longer

carry out tasks involving fine motor skills, such as fastening a nappy or pressing a button. Two other participants (Benjamin and Elijah) had to retire from a job they loved because they realised that they might be a danger to themselves and others due to their weakened limbs and frequent falls. Although the participants in this study seemed to have adapted well to a life turned upside down so suddenly, there were moments when they felt frustrated and angry at being dealt such an intricate hand. A participant (Elijah) became deeply emotional when he explained that the one thing that affected him the most was that he could no longer participate in his local village feast, where he used to carry the statue of his beloved Holy Mary every year. A distinct difference in cultures was noted in the other extant literature. In fact, in the study by Yuan et al. (2021), one participant, due to the Chinese culture, was brought up to believe that being virtuous will have its reward; however, if one is evil, then he/she will have to face retribution, became very confused at being diagnosed with ALS. It rendered her trying to fathom out what she had done wrong in life, and she could no longer understand why she was 'punished' in such a way.

Furthermore, the study by Yuan et al. (2021) revealed the strong presence of Chinese traditional culture yet again when some of the patients strongly believed that life is more meaningful than death and that it is wiser to enjoy life to the full without disappointment their loved ones than mull about death. This aspect of culture was surprisingly consistent with the beliefs in this present study since most participants believed in living and enjoying their lives as a whole, irrespective of their diagnosis. In the study by Oh et al. (2014), the patients in the study described that becoming dependent on others for things that people take for granted, such as eating, using the bathroom and washing, led to a distressing feeling of meaningless and some participants even meant a loss of the value of life and a loss of autonomy.

Another impact being diagnosed with ALS had on some participants in this study was their fear of becoming a burden to their caregivers. One participant (Isaac) was concerned because his partner had given up a professional career to care for him when he was diagnosed with ALS. Furthermore, another participant (Sophia) had to rely on her husband to do certain chores, such as cooking and washing the dishes, that she had always cared for before being diagnosed with ALS. Another participant (Elijah) felt distressed because his daughter, his primary caregiver, had to juggle being a student at University whilst taking care of him at the same time. He explained how it hurt him

to see how his wife and brother could not cope with his diagnosis and how, apart from his burden, he also had to support his wife because of her mental health issues. Subsequently, in the study by Oh et al. (2014), married participants stated that their relationships had been affected after their diagnosis. For instance, South Korean culture emphasises the family structure rather than the individual's autonomy. For instance, to keep the family hierarchy, the father's role as head of the family is significant; however, with a diagnosis of ALS, the father will have to become the one receiving care. A shift in the family order may then be the cause of disarray and confusion. This type of reasoning was not encountered in this present study. In the study by Yuan et al. (2021), which was also consistent with this study, one participant worried about his wife's mental health. However, he used to laugh constantly. He could feel his illness was taking a toll on her because she was his primary caregiver and always cared for him.

In this study, a participant (Benjamin), who worried constantly that ALS may reappear in his family, although he was diagnosed with the sporadic type, explained how he did not speak about this particular issue in front of his wife for fear of worrying her unnecessarily. This showed that although they were diagnosed with ALS, some participants still felt the guilt of possibly being a burden to their loved ones. Furthermore, a study by Geng et al. (2016) showed that female patients had higher self-perceived burdens than their male counterparts. This was inconsistent with two other studies from Western countries (Chio et al.,) which again emphasised that women from Eastern countries are responsible for all the household chores, including raising their children and caring for their in-laws. This is the 'Confucian concept of ethics (Hashizume et al.). Focusing on the biopsychosocial aspect of ALS, the study by Kukulka et al. (2019) stated that, consistent with other research (Bolmsjo, 2001), ALS takes a physical and psychological toll and reduces the individuals' self-worth. One participant in this study (Joshua) explained how he knows no worse fate than being diagnosed with ALS.

In this study, only one participant (Elijah) mentioned the financial impact that ALS might bring on the family. At some point, the individual will have to terminate his employment because of the rapid progression of paralysis. This participant stated that in his case, he was fortunate because he was self-employed and he could manage his business from home because he had employees working for him. However, he

emphasised that if one struggled financially, the individual would suffer a lot more because specific necessities and commodities, such as comfortable wheelchairs, walking frames and pillows, were very costly. Furthermore, adaptations to the house had to be made because he could no longer walk up the stairs. He stated that due to his financial stability, the structural changes to his residence could be done immediately and quickly.

Interestingly, in a study by Geng, Ou, Miao, Zhao, Wei, Chen, Liang, Shang and Yang (2016), patients stated that their caregivers reported lower caregiving burden if their family had a higher family income or health insurance covered their patient. Other studies show that the financial burden of ALS is higher because healthcare is not free. In Korea, the patient has to pay 44.8% of the medical fees, whereas the Korean government covers the remainder. Additionally, in South America, Riluzole, a costly medication used to slow down the progression of ALS, is not covered by a government subsidy. In contrast, Riluzole has not been included in healthcare insurance in China. Considering the low income of a Chinese individual, paying more than $700 per month to purchase Riluzole is very difficult. However, the Chinese culture states that if one family member has a severe and chronic illness such as ALS, the whole family will help pay for any medical bills incurred, even if they become penniless.

In the study by Geng et al. (2016), findings showed that the patient's QOL diminished as ALS progressed. These results are tallied with several established studies and indicate the necessity of practical nursing care to improve the patient's QOL (Ozanne et al.). These results were consistent with this current study, whereby the participants all reported a reduction in their QOL, although they tried to maintain normality as much as possible. A recent study by Londral et al. (2015) discovered that intervening at an early stage with an assistive communicative device positively impacted the patient's QOL. However, such devices are costly, thus increasing the burden on the family because they cannot afford one. Out of the five participants of this study, only one (Joshua) made use of such a device because he was the only one in the advanced stages of ALS.

5.5: Coping Strategies In Individuals Diagnosed With ALS

In the superordinate theme 'striving to keep afloat,' the participants discuss how despite being diagnosed with a terminal disease, they still ill-harboured a strong sense of serenity, peace and resilience. They admitted to moments of sadness and frustration. However, they found ways to resist feeling overwhelmed by incorporating coping strategies into their daily lives. Two participants (Joshua and Isaac) immersed themselves in voluntary work because they stated that helping others instilled a sense of happiness and peace. Another participant (Elijah) found solace and gratitude in still being able to run his business from home, whilst the only female participant (Sophia) tries to keep up a sense of normality by travelling and going out with her husband.

In the study by Yuan et al. (2021), some participants concentrated on their sense of self-worth to help them cope better. They involved themselves in activities, such as small chores around the house or continued to go to work whenever possible. Keeping themselves active enhanced their sense of value, and at the same time, they felt needed. These participants also felt a great need to help and encourage other patients. These findings tallied similarly to what the participants of this present study explained.

ALS is a severe neurodegenerative disorder with progressive decline and no remission (Rowland et al.,2011). A study by Hamama-Raz, Norden & Buchbinder (2021) discussed the double sides of hope and how the 'two voices' of hope can be different among people with ALS. For some patients, hope was seen as an obstacle and was perceived as an unnecessary emotion which reduces the individual's strength by placing their hope in an illness of which there is no cure by creating an illusion. Some study participants believed that being armed with knowledge about ALS and understanding the truth about its fatality and incurability is far better than hoping in vain because there is no hope for ALS. In contrast, other participants believed that hope offers them strength and helps them face each day with their disease whilst keeping up their spirits. Some referred to hope as 'light in a dark tunnel' and considered it a crucial resource that helped them cope. Surprisingly, the article by Hamama-Raz et al. (2021) on hope being seen as an obstacle proved to contradict a myriad of other articles that indicate how hope had beneficial effects on the well-being and QOL of individuals diagnosed with life-threatening conditions (Farhadi, 2014; Li, 2016; Rock, 2014). According to Hamama-Raz (2021), an explanation for such contradiction may be due to the unique characteristics of ALS and that it is very different from other life-threatening diseases such as metastatic cancer, AIDS and heart failure. In this study,

most participants faced each day with hope and strength. They did not pertain to the first voice of hope. For them, each day was a fight to live. They found that living in the present, emphasising normality and surrounding themselves with family, friends, and colleagues helped them avoid spiralling into despair. One participant (Benjamin), who trusts science, says he is hopeful that a cure or a way to halt the disease will be found. Another (Elijah) leans towards his spiritual faith as a coping strategy. His faith in the Holy Mary and Jesus Christ allows him to continue fighting and not give up. He states that he becomes distressed when people around him say that his fate at the hands of ALS is not fair. Does he believe that why should it not be him and not the contrary? He continues to explain that he finds solace in prayer. Another participant (Joshua) explained how although he is paralysed and can no longer feed himself, he understands that although as each day passes, his body is slowly dying, he can still achieve great things.

In a study by Marconi, Gragnano, Lunetta, Gatto, Fabiani, Tagliaferri, Rossi, Sansone & Pagnini (2016), meditation training was another method that patients with ALS and their caregivers found beneficial. They reported improvement in anxiety and depression, commonly found in ALS individuals (Palmieri, 2010). Furthermore, participants reported how practising breathing exercises and awareness of mindfulness led to reported an improvement in their sleep patterns. Additionally, mindfulness training led to a better understanding and acceptance of the diagnosis, being more mindful overall and improving their QOL (Pagnini et al., 2014). Moreover, the group setting where the training sessions occurred proved comforting for the patients and their caregivers. Individuals could sit and share tips and advice, and an overall feeling of well-being and burdens being lifted were felt. In fact, participants recommended that mindfulness techniques should be shared with other individuals who have ALS. Subsequently, An improvement in QOL is one of the main goals of research in ALS (Simmons, 2005). In this present study, there was no mention of the participants attending a mindfulness-based intervention. However, the female participant (Sophia) stated that she practised yoga daily before being diagnosed with ALS.

All five participants in this current study reported having a robust support system in the form of their treating neurologist, the healthcare providers at MDH, Dar Bjorn and the UM ALS/MND research team, their families, friends and colleagues. One participant (Elijah) even mentioned feeling at peace whenever his pet dog lay at his

feet. He explained how his dog seemed to sense that he was unwell and never left his side.

In the study by Yuan et al. (2021), participants explained how they found meaning in life by resetting their priority lists. They realised that spending time with their families was far more critical than constantly working and saving money. Another coping strategy was taking each day as it comes and living the moment whilst embracing every moment of happiness encountered. These findings were consistent with this present study whereby all five participants stated that living in the here and now helped them cope and, for a short while, liberate them from thinking about ALS and compensate for the loss in QOL.

5.6: Conclusion

The findings of this study have provided an in-depth understanding of the lived experience of individuals diagnosed with ALS and the biopsychosocial difficulties associated with its threatening characteristics.

Chapter 6: Conclusion

6.1: Introduction

This chapter summarises a research study exploring the lived experiences of individuals diagnosed with ALS. It will also present the strengths and limitations that the researcher encountered whilst conducting this study, together with a list of recommendations for clinical practice and any future research.

6.2: Summary of The Research Study

A plethora of literature exists exploring the molecular genetics and pathophysiology of ALS and articles discussing the burden of ALS on primary caregivers. In contrast, the researcher noticed a paucity of literature directly addressing the lived experiences of individuals diagnosed with ALS, both from a local and an international perspective. Moreover, 14 local research studies were identified that targeted ALS from a genetic, situational and occupational point of view. Additionally, one local dissertation by Said (2018) was extracted that addressed the request for assisted suicide by ALS patients. Interestingly a study by Wyatt (2013) compared MND among Maltese in Malta and Australia and how it was not linked to poliomyelitis. This paucity of qualitative phenomenological research on the lived experiences of individuals with ALS prompted the researcher to address this evident gap, namely the experiences of persons having ALS within a Maltese context. Furthermore, the researcher's primary reason behind this study was to provide a voice for this particular cohort of patients whose sufferance, at times, goes unnoticed and who, until now, have never had the opportunity to describe what it means to be diagnosed with a fatal and debilitating illness. Consequently, the aims and objectives of this qualitative study were to delve deeper and gain insight into the lived experience of individuals diagnosed with ALS whilst exploring the impact such an illness has on their lives, both from a physical and psychological aspect.

The researcher decided that an IPA was a suitable approach for this study as she wanted to investigate her participants' experience living with ALS. To ensure that the conclusions she drew from her small sample size were valid, the researcher opted for a relatively homogenous sampling of five patients diagnosed with ALS. As a data collection method, semi-structured interviews were carried out in the participants' residences and Dar Bjorn. Furthermore, the researcher transcribed and analysed the

audio-recorded interviews as proposed by Smith et al. (2009). Subsequently, two super-ordinate themes emerged from the research carried out in this study.

The first super-ordinate theme, A ticking time bomb*,* describes how the initial symptoms experienced by the five participants were so vague that it did not occur to them that they may be suffering from MND, let alone ALS. The most frequent symptom encountered were frequent falls and instability that the participants blamed on clumsiness or tripping over something. Moreover, this super-ordinate theme addressed the impact of being diagnosed with a fatal and neurodegenerative condition and how, from strong, healthy and hardworking individuals, they became unable to walk, eat, wash or do simple tasks such as fastening up a button. All participants unanimously stated that ALS robbed them of their independence, rendering them reliant on their primary caregivers. The concern of ALS being passed on in the family, even though the participant was diagnosed with the sporadic form, emerged significantly, as the participant did not wish to discuss such issues in front of his wife for fear of worrying her.

Another aspect discussed during the first super-ordinate theme was the rollercoaster of emotions the participants went through on being told, after a battery of tests, that they had ALS. The financial impact ALS may have on the family since the afflicted individual will eventually have to leave their employment was another concerning issue.

One participant stated that although awareness of ALS has increased nowadays, people seem not to understand the devastation of having ALS, and all they see is a person in a wheelchair. Additionally, the exasperation of being treated differently because of their illness was addressed with another participant stating that he did not want to be considered 'that peculiar person sitting in the corner' and just be treated as the person he used to be before being diagnosed with ALS. Although a diagnosis of ALS was described as overwhelming for the five participants, they all showed strength, courage and resilience. Only one participant broached the sensitive topic of euthanasia and the possibility of refusing nourishment and a mechanical ventilator to help him breathe if he could no longer cope towards the end stages of ALS. Interestingly, most participants stated that engaging in things they usually would have done with their families before the diagnosis and living in the present gave them some sense of normality and hope.

Suppressing their emotions was another way the participants dealt with their diagnosis since thoughts about the illness trajectory were too distressing. Although the participants showed strength and resilience,
it was a front not to worry their families unnecessarily.
The second super-ordinate theme, 'Striving to stay afloat', was a participant's metaphoric description explaining the repercussions of ALS on him and his family. Ways to adapt to their situation by living in the present emerged. One participant resorted to his religious faith to find solace and inner peace. Furthermore, this super-ordinate theme also highlighted that humour could assist in coping with ALS. Additionally, a common factor amongst all participants was the considerable amount of support and care that they received from various entities such as their treating neurologist, the ALS/MND team at MDH, Mr Bjorn Formosa, the President of the ALS Malta Association, and last but not least their families, colleagues and even pets.

6.3: Strengths and Limitations

6.3.1: Strengths

Several strengths emerged from this study, which was the author's first attempt at conducting research:

- This is the first local study that directly explores an in-depth account of the lived experience of individuals diagnosed with ALS. This study also contributes to other international studies since there is a gap in the literature that addresses the patients' lived experiences rather than the caregiver.
- In line with the IPA methodology, the study had five relatively homogenous participants willing to participate and help the researcher carry out her study by providing sensitive information about their experience in being diagnosed with ALS. Semi-structured interviews, as Smith et al. (2009) recommended, were deemed appropriate since they provide a framework of questions that guide the novice researcher.
- By the end of the study, and with each interview, the researcher felt more confident and knowledgeable about conducting phenomenological research and interviews with patients with terminal illnesses. The information generated

can assist in formulating interventions sensitive to the needs and concerns of individuals with ALS.

- The researcher kept a reflective journal throughout the research project. Furthermore, the academic supervisor carried out independent mini-audits to ensure that any preconceptions did not influence the data analysis.

6.3.2: Limitations

- Being a novice researcher in the methodology of IPA is a limitation in its own right. The researcher had an opportunity to practice interviewing a previous study unit. However, she still felt apprehensive about her capabilities of conducting interviews with patients with a fatal and physically debilitating disease. However, the researcher became more confident and at ease with each interview.
- The participant's medical condition could have influenced the interview duration in two cases. Two of the patients used assistive breathing devices; however, although the interview conducted was shorter in duration, rich data was still obtained. Moreover, all the participants were
- Eager to voice their experiences with the researcher. Throughout the interview, the participant's health and well-being had to be kept in mind. Thus, the researcher gently reminded them that if, at any point, they felt too tired to continue, she would conclude the interview.
- The rarity of ALS and its destructive and debilitating nature led to a scarcity of patients willing to be interviewed and presented as a challenge. The researcher required the assistance of two intermediaries who recruited patients from Dar Bjorn and MDH.
- Translating verbatim from Maltese to English proved challenging for the researcher since certain Maltese metaphorical sayings and phrases that are native to our language may not always have a corresponding English version. To reduce this limitation, the researcher requested the help of a certified Maltese proofreader and translator to ensure the translation from Maltese to English remained faithful to the original excerpt. Additionally, three out of the

five participants preferred to converse in English. Hence no translation was required in that case.

- Three participants insisted that their primary caregiver was present during the interview. This may have led to the participant holding back specific sensitive details and/or emotions. However, the researcher could not confirm this.

6.4: The Learning Experience

Adopting Bloom's Learning Theory (Bloom et al., 1956), the researcher will describe her learning experience while carrying out this study.

She was conducting a qualitative study with IPA as its methodology proved to be a learning curve for the researcher. From barely knowing where to start and how to process everything, the researcher gradually gained more confidence in conducting phenomenological research effectively. Additionally, she increased her knowledge of how to conduct face-to-face interviews whilst establishing high professionalism and ensuring the ethical issues involved in such a study were adhered to. Subsequently, the researcher extended her knowledge of mental health nursing and neuropsychiatry by studying individuals diagnosed with ALS.

Apart from conducting in-depth research from the clinical aspect of ALS, the researcher also became knowledgeable from a linguistic perspective during her research study. Using her active listening skills and also focusing on the linguistic aspect used by the participants (including the use of metaphors, personifications, digressions and repetitions), the researcher was more appreciative when it came to analysing the narratives and life experiences of persons with ALS. Furthermore, given the sensitive nature of their illness, the researcher paid particular attention to any outbursts of emotions experienced during the interview.

Due to the distressing nature of the study and the predicament of her participants, the researcher used a reflexive journal throughout the research process. The researcher was dealing with participants diagnosed with a terminal illness, so the interviews were heartbreaking at the best of times. However, one particular interview was so distressful in content that the researcher felt overwhelmed with emotion and, at one point, thought she could not continue with the study. Nonetheless, she overcame this issue by jotting these thoughts down in her journal and seeking guidance from her academic

supervisor. She learnt the importance of reflecting on such preconceptions and beliefs personally and in interactions with others.

The researcher evaluated her performance at the end of each interview and was aware that her interviewing skills had significantly improved. She felt more confident using prompts to engage the participants in describing their experiences in more depth; however, she still prioritised her participants' psychological well-being and medical health throughout the interview. Subsequently, the research was perceived with an initial sense of trepidation and apprehension at writing the first chapters of this dissertation for fear of being on the wrong track; however, with time, she understood what was requested of her, and her critical skills improved.

The following sections delineate recommendations for clinical practice.

6.5: Recommendations For Improved QOL in ALS

To- date, ALS is incurable. However, medical interventions and technology can significantly improve the QOL for people with ALS by management of symptoms and proactive use of equipment which can make a positive difference in day-to-day living and possibly even prolong life. The section below will address such issues:

The findings of this study repeatedly showed the challenges patients with ALS face daily from the onset of symptoms to the point of diagnosis and after that. The patient's autonomy should be considered a priority in care planning. Treating such individuals is complex because clinical management involves biopsychosocial issues and physical disability. Hence, a need for multidisciplinary ALS clinics to assist patients during their illness is a way to ameliorate their quality QOL. For instance, a palliative nurse may prove instrumental in addressing issues that centre around the individual's well-being, providing physical comfort and emotional support to the patient and his/her family. Additionally, a psychiatric nurse may be a valuable part of the team since emotional lability, anxiety, and depression frequently surface in ALS patients.

- Research shows that patients that are referred to specialised ALS clinics have less and/or fewer hospital admissions (Chio, 2006), further emphasising the importance of such services.
- A prompt diagnosis, patient and caregiver involvement, and a care plan emphasised by evidence-based guidelines are required for effective clinical

management. More importantly, the narratives explained by the patient and primary caregiver should be considered.

- The researcher noted during her interviews that significant time had passed between the onset of the patient's symptoms and the final diagnosis, leading to concern and worry in most participants. A diagnosis of ALS should be given without unnecessary delays. Hence, a fast-tracking system within the healthcare system should be implemented for patients with ALS and other neurological disorders. Furthermore, the time that passed from the onset of symptoms to the actual diagnosis of the participants of this study was tallied with research by Millul (2005), who stated that the timeframe of diagnosis varies between 13-18 months.
- Although one participant was diagnosed with the sporadic form, both the participant and family members worried that ALS might reappear in his family. This led the researcher to believe that the participants lacked knowledge about the hereditary aspects of ALS. Thus, there is a need for the services of a genetic counsellor for both the patient and their families, irrespective of whether they have a sporadic or familial form.
- All five participants admitted to having a solid family support system and that their families played an essential role in their lives. This finding was consistent with a study by Yuan et al. (2021), who recommended that healthcare providers actively promote the family self-help model whereby the individual with ALS is encouraged to take on a supportive role in the family. The caregivers, in turn, will be given more support and offered professional care knowledge to alleviate their burden. Additionally, it is crucial to encourage patients and their families to speak about ALS and professionally broach the topic of death education.
- The participants all required assistance to carry out their active daily living skills (ADLS). Community healthcare systems, including occupational therapists and social workers, may be an invaluable source of assistance in situations whereby help with financial issues is required and offering the patient help in his/her active daily living skills (ADLs). These include managing basic physical needs and personal hygiene (Yuan et al., 2021).
- Although one participant was diagnosed with the sporadic form, both the participant and family members worried that ALS might reappear in his family.

This led the researcher to believe that the participants lacked knowledge about the hereditary aspects of ALS. Thus, there is a need for the services of a genetic counsellor for both the patient and their families, irrespective of whether they have a sporadic or familial form.

- At an advanced stage, patients with ALS will inevitably lose the ability to walk, talk, swallow and breathe. Thus an electronic computing device, known as an eye-gaze tracker that reads eye movements so that the patient can select letters by directing his/her gaze on them may help the patient regain some of his/her independence by communicating better. Although there is a local foundation that helps families acquire such technological devices, which are very expensive, there is a need for ALS patients to be given the option of owning one as soon as they become unable to speak and/or communicate.
- ALS is incurable and one of the most biologically and genetically driven diseases (Pagnini, 2015). The psychological impact after diagnosis is understandable, and frequently, depression, anxiety, and a sense of hopelessness ensue. The rapid progression of symptoms may also affect the patient's well-being and reduce QOL. Offering the participants sessions on practising mindfulness may prove beneficial since it is known to slow the disease's progression and improve psychological well-being (Langer, 1989).
- One participant described in detail how his beloved dog somehow sensed that he was ill and used to constantly lick the areas where he was becoming paralysed, such as his limbs. He also found solace in her presence. The possibility of providing animal-assisted therapy in residential homes may prove beneficial for patients with ALS.

6.6: Recommendations For Management and Policy

- ALS is a severe disease that affects both patients and their families. Thus, sensitive communication of the diagnosis is a necessity. Research shows that how a diagnosis is imparted to the individual will significantly impact the relationship between the healthcare professional and the patient. Thus, there is a need for a policy whereby a nurse navigator, similar to the ones at SAMOC, will be assigned to the patient from the moment of diagnosis and after that. The nurse navigator is an exceptional personal advocate to the patient who

guides him/her through the treatment process and assesses their psychosocial needs. The skillset of the nurse navigator should include the knowledge and attitude to provide effective person-centred care.

In addition to the above recommendation, having a policy containing guidelines for communicating such diagnoses and continuing care afterwards would be beneficial.

- Furthermore, several evidence-based guidelines should be incorporated into one internationally accepted guideline for ease of reference.

6.7: Recommendations For Future Research

- Further research addresses the limitations encountered during this study, namely, a need for more international and local studies that concentrate more on the patients' lived experience on receiving the diagnosis and towards the advanced stages of ALS.
- There exists a plethora of research on advanced care planning (ACP); however, from a local perspective, the researcher found a paucity of literature on ALS in general, not only on ACP. The articles the researcher found primarily addressed the molecular genetics and occupational point of view of ALS and were written by researchers from UM's ALS/MND laboratory. Therefore, there is a need for more awareness and, subsequently, local literature on ACP as an integrated palliative care approach. ACP is a professional communication process whereby the patient discusses his/her wishes, goals and preferences for treatment during a fatal illness (Seeber, 2019). Setting up such an approach for ALS patients will enhance their QOL and allow them autonomy over their decisions involving their care before the illness renders them unable to do so.

6.8: Conclusion

A diagnosis of ALS is a harrowing experience for patients and caregivers alike. As the research findings in this study show, the patients' lives are fraught with fear, trepidation, concern and, at times, even anger and frustration. ALS is not only a fatal condition whereby there is no cure to date, but also a rapidly progressive,

neurodegenerative and debilitating disease. So apart from the fact that the individuals know that they are going to die, they also have to contend with the pain, paralysis, loss of independence and a myriad of other symptoms that ALS brings with it. Seeing their bodies degenerate from day to day proved to be challenging, traumatic and life-changing for them. Apart from emotion liability being a symptom of ALS, the patients had to deal with the raw emotions of being diagnosed with a terminal diagnosis for which there is no cure. The patients, however, all spoke highly of the support they received from their healthcare providers, both at MDH and in the community. Moreover, knowing that there are options for residential community homes for people with ALS and other neurological disorders was crucial for them.

An IPA approach was the methodology most suited for a topic that addresses the challenges and struggles a patient diagnosed with ALS has to go through since it allows the researcher to explore, in-depth, the intimacy and rich understanding of a compassionate aspect in their lives.

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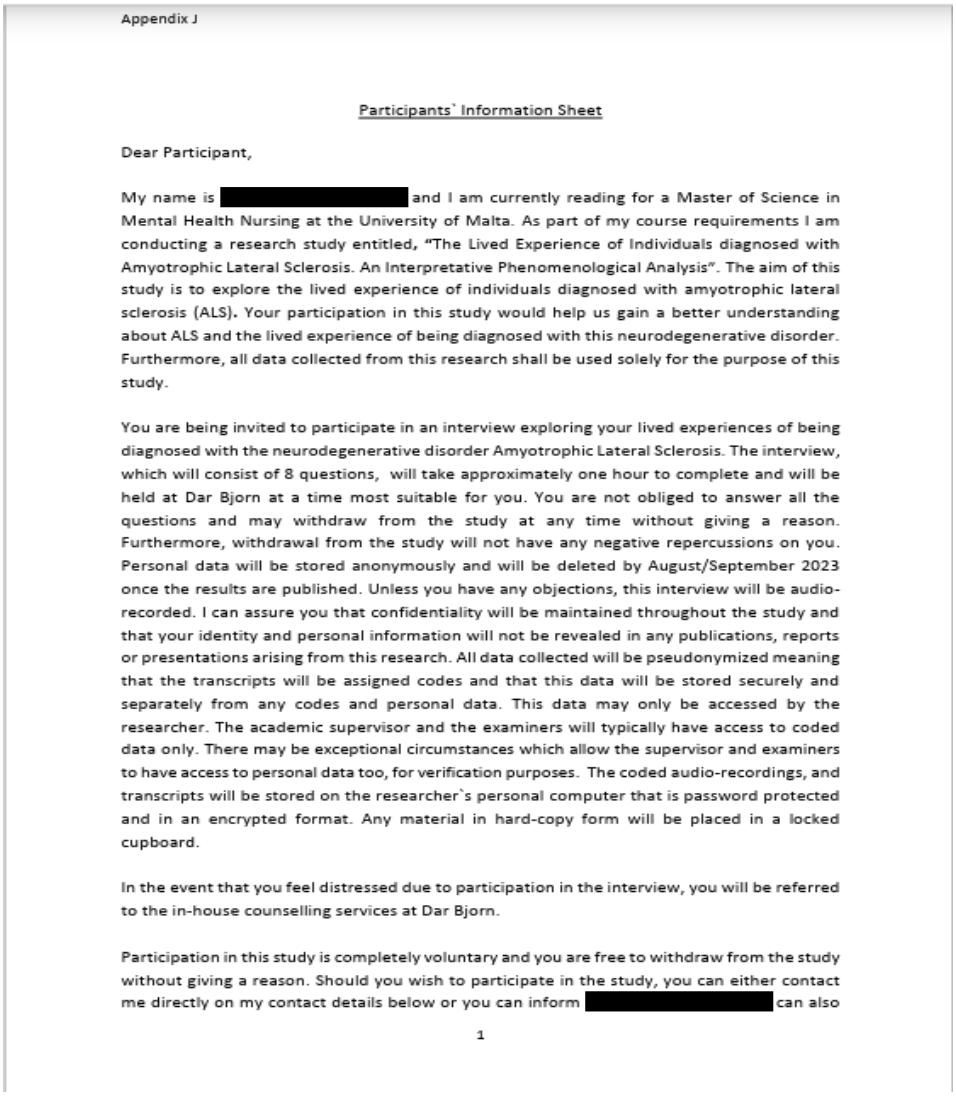
Appendices

Appendix A*:* Participants Information Letter



Appendix J

Participants` Information Sheet

Dear Participant,

My name is [redacted] and I am currently reading for a Master of Science in Mental Health Nursing at the University of Malta. As part of my course requirements I am conducting a research study entitled, "The Lived Experience of Individuals diagnosed with Amyotrophic Lateral Sclerosis. An Interpretative Phenomenological Analysis". The aim of this study is to explore the lived experience of individuals diagnosed with amyotrophic lateral sclerosis (ALS). Your participation in this study would help us gain a better understanding about ALS and the lived experience of being diagnosed with this neurodegenerative disorder. Furthermore, all data collected from this research shall be used solely for the purpose of this study.

You are being invited to participate in an interview exploring your lived experiences of being diagnosed with the neurodegenerative disorder Amyotrophic Lateral Sclerosis. The interview, which will consist of 8 questions, will take approximately one hour to complete and will be held at Dar Bjorn at a time most suitable for you. You are not obliged to answer all the questions and may withdraw from the study at any time without giving a reason. Furthermore, withdrawal from the study will not have any negative repercussions on you. Personal data will be stored anonymously and will be deleted by August/September 2023 once the results are published. Unless you have any objections, this interview will be audio-recorded. I can assure you that confidentiality will be maintained throughout the study and that your identity and personal information will not be revealed in any publications, reports or presentations arising from this research. All data collected will be pseudonymized meaning that the transcripts will be assigned codes and that this data will be stored securely and separately from any codes and personal data. This data may only be accessed by the researcher. The academic supervisor and the examiners will typically have access to coded data only. There may be exceptional circumstances which allow the supervisor and examiners to have access to personal data too, for verification purposes. The coded audio-recordings, and transcripts will be stored on the researcher`s personal computer that is password protected and in an encrypted format. Any material in hard-copy form will be placed in a locked cupboard.

In the event that you feel distressed due to participation in the interview, you will be referred to the in-house counselling services at Dar Bjorn.

Participation in this study is completely voluntary and you are free to withdraw from the study without giving a reason. Should you wish to participate in the study, you can either contact me directly on my contact details below or you can inform [redacted] can also

1

Appendix J

pass your contact details to me. A copy of the information sheet and consent form will be provided for future reference. As a participant, you have the right, under the General Data Protection Regulation (GDPR) and national legislation that implements and further specifies the relevant provisions of said regulation, to access, rectify and where applicable ask for the data concerning you to be erased. Once the study is completed and the results are published, the data will be retained in anonymous form. Any personal details will be destroyed.

This study has been approved by the Research Ethics Committee of the Faculty of Health Sciences at the University of Malta.

Thank you for your time and consideration. Should you have any questions or concerns do not hesitate to contact me on [redacted] by e-mail on [redacted]

Yours sincerely,

Appendix B: Approval of Intermediary

15 April 2022

ALS Malta

Dar Bjorn

308, Triq il-Kbira

3rd May, 2022

To whom it may concern,

I am [redacted] Chairperson of ALS Malta + and I am here accepting to also be the intermediary of [redacted] apart from accepting her to carry out the study at DAR Bjorn.

Regards,

Appendix C: Intermediary Number 1 Request Letter

15 April 2022

ALS Malta

Dar Bjorn

308, Triq il-Kbira

Qormi

Malta

Dear Mr. [redacted]

My name is [redacted] and I am a psychiatric nurse by profession working in an acute ward at Mount Carmel Hospital. I am presently reading for a Master of Science Degree in Mental Health Nursing at the University of Malta and as part of its fulfillment requirements, I will be conducting a research study. The proposed title is "The Lived Experiences of Individuals diagnosed with Amyotrophic Lateral Sclerosis. An Interpretative Phenomenological Analysis". Through this research study I intend to explore the lived experiences of clients diagnosed with Amyotrophic Lateral Sclerosis (ALS) residing at Dar Bjorn. This will be carried out by conducting in-depth audio recorded interviews to six to eight clients diagnosed with ALS, who are over 18 years of age, are male and/or female and are not receiving end-of-life care.

To fulfill research and ethical requisites, I would appreciate your help as an intermediary to approach and recruit six to eight clients diagnosed with ALS. I will be providing an information letter explaining the nature of the study and your role will be to hand out the information letter which will be offered in both an English and Maltese version. The clients will then be given sufficient time to decide whether they wish to participate in the study or not. Your role will also include providing me, as the researcher, with contact details of any participant who is willing to participate in the study unless the participant wishes to contact me themselves directly.

Whilst thanking you in advance, should you require further details regarding my study, please feel welcome to contact me on mob. [redacted] by e-mail on [redacted] or my supervisor Professor [redacted] by email on [redacted]

Kind Regards,

Appendix D: Intermediary Number 2 Request Letter

L-Università ta' Malta [redacted]

Permission to act as an intermediary

2 messages

[redacted] 28 August 2022 at 12:46

[redacted]

[redacted] and I am a psychiatric nurse by profession. I presently reading for a Masters in Mental Health Nursing and the topic I chose to research is the lived experience of individuals diagnosed with Amyotrophic Lateral Sclerosis, both in the familial form and sporadic cases. The study will be on both male and female adults over 18 years of age, however, they cannot be at the stage of end-of life.
I understand that my supervisor, [redacted], has already mentioned this to you and requested your kind assistance. In fact, you had kindly demonstrated a willingness to assist me in this. I still have to receive approval from FREC/UREC though, so at present, I cannot collect any data, however, I am kindly requesting your approval to act as an intermediary. Motor neuron disease is a branch of medicine that I hold very close to heart and although the masters I am reading for is in mental health, I still managed to incorporate the psychological aspect into a medical subject.

Attached please also find the intermediary information letter for your perusal.

I am in great appreciation of any help you can offer at this stage[redacted]

[redacted] 29 August 2022 at 08:58

[redacted]

Yes sure. Once you start recruiting patients let me know.

Regards,

[redacted]

Appendix E: Interview Schedule

Interview Schedule

English Version

Introduction

The aim of this interview schedule is to briefly provide an overview of the open-ended questions that will be discussed during the interview. The schedule will be available in both English and Maltese versions.

Questions:

1) Would you kindly tell me a little about yourself? (This question may be used as an ice-breaker to place the participant at ease).

2) Amyotrophic Lateral Sclerosis (ALS) is a progressive and debilitating neurodegenerative disorder. What was your reaction when you were told that you have ALS?

3) How did your family and friends deal with your being diagnosed with ALS?

4) Following the initial impact of your diagnosis, how has ALS affected your quality of life (QOL)?

5) Can you share with me how you strive to cope with such a diagnosis?

6) What has been the most difficult change to cope with since being diagnosed with ALS?

7) Try and think back to before you were diagnosed with ALS, has your definition of quality of life changed after the illness?

8) Is there anything else you would like to share with me before we end this interview?

Appendix F: FREC and UREC Approval

Application ID	Application Date	Project Title	Faculty	Status
FHS-2022-00093	05/06/2023	The Lived Experiences of Individuals diagnosed with Amyotrophic Lateral Sclerosis. An Interpretative Phenomenological Analysis.	Faculty of Health Sciences	Approved
FHS-2022-00091	11/05/2022	The Lived Experiences of Individuals diagnosed with Amyotrophic Lateral Sclerosis. An Interpretative Phenomenological Analysis.	Faculty of Health Sciences	Draft
FHS-2022-00081	01/05/2022	The Lived Experiences of Individuals diagnosed with Amyotrophic Lateral Sclerosis. An Interpretative Phenomenological Analysis.	Faculty of Health Sciences	Draft