

It's All Just Suffering: The Experience of Pain in Cystic Fibrosis

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I declare that this thesis is my own work and has not been submitted for the award of a higher degree elsewhere.

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Abstract

Life expectancy in cystic fibrosis (CF) has greatly improved, but as people live longer, they experience increasing symptoms. Literature indicates pain in CF is common and underreported, affecting quality of life, mental health and adherence to treatment. Pain is best explored in the context of the whole person, yet previous studies are almost exclusively quantitative.

The aim of this research was to gain understanding of the experience of pain, within the context of the whole person with CF. Within a social constructivist framework, narrative methodology focused on the complex and unique experiences of individuals. Data were gathered through interviews with nine individuals and analysed using a method of narrative analysis to present findings in stanzas, a poetic form that preserves the narrator's voice. Findings were interpreted through the lens of total pain theory.

Three key narratives contributed to understanding pain: emerging awareness, social legitimacy and invisibility. Emerging awareness was the gradually increasing understanding of what it means to have CF. Social legitimacy was critical in the experience of total pain, appearing as participants struggled to conform to societal expectations. Total pain was ever-present in these stories, and when participants were isolated or marginalised and lost social legitimacy, they felt invisible.

Total pain and suffering in CF are best understood as a singular experience happening to a whole person, encapsulating their very being. This deeper understanding highlights an opportunity to explore pain in the context of the whole person, to inform clinical practice and future research.

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Abbreviations

BPI: Brief Pain Inventory

CF: cystic fibrosis

CFRD: cystic fibrosis related–diabetes

CFTR: cystic fibrosis transmembrane conductance regulator

COPD: chronic obstructive pulmonary disease

FEV1: forced expiratory volume in one second. FEV1 is a standard unit of measurement used in CF to determine severity of disease and guide treatment decisions. A pulmonary function test is used to measure the amount of air someone can forcefully blow out of their lungs in one second. This value is the FEV1, reported in litres and as a percentage of what would be expected in a healthy person of similar gender, height, weight and race.

FHMREC: Faculty of Health and Medicine Research Ethics Committee

HADS: Hospital Anxiety and Depression Scale

HRPP: Human Research Protections Program

IASP: International Association for the Study of Pain

IV: intravenous

MPI: Multidimensional Pain Inventory

NSAIDs: Non-steroidal anti-inflammatory drugs

PICC: peripherally inserted central catheter

PRISMA: Preferred Reporting Items for Systematic Reviews and Meta-Analysis

PTSD: Post-Traumatic Stress Disorder

RA: rheumatoid arthritis

RCT: randomised controlled trial

RN: registered nurse

SSI: Supplemental Security Income

Chapter 1: Introduction

1.1 Personal reflections

I have spent most of my career as a registered nurse (RN) working in the areas of paediatrics, palliative care, and for the past 14 years, cystic fibrosis (CF). Working with pain has been a constant, but never has it felt more complex than pain in CF. In my clinical work, I found myself frustrated to hear some clinicians did not believe pain was an issue in CF, while people with CF told me they did not report pain because they believed nothing could be done. One of my core beliefs is that if I am not working to address an issue, I do not have the right to complain about it. I therefore sought others with similar concerns, and we formed a working group with a broad interest in palliative care in CF. To date, we have authored several publications and presented on a range of topics around CF and palliative care, including pain, advance care planning, barriers to palliative care, and education for palliative care in CF (Allgood, Kapnadak, Dellon, Goggin, & Lechtzin, 2018; Dellon et al., 2016; Dellon et al., 2018; Dellon, Hobler, Georgiopoulos, Chen, et al., 2019; Dellon, Hobler, Georgiopoulos, Goggin, et al., 2019; Goggin, 2017; Goggin & Cohen, 2016). The Cystic Fibrosis Foundation has supported many of these efforts.

As I continued this work, it became clear that pain in CF was controversial. In discussions following sessions about palliative care in CF, the subject of pain inevitably arose. Much disagreement seemed centred around opioids. The phrases “drug seeking” and “not really in pain” were common. Some viewed opioids as absolutely contraindicated in people listed for transplant, while others believed pain medication should rarely, if at all, be

prescribed in CF. Still others relayed scenarios where they felt people with CF had been prescribed opioids for symptoms other than physical pain. These anecdotes and concerns are not unique to the CF community, as similar concerns have been reported in the pain management literature (Carvalho et al., 2018). During these discussions, clinicians expressed emotion at a level not usually seen at professional conferences, yet clearly longed for more information about pain and wanted to help people with CF. I understood this. I still recall people with CF who died following an overdose or struggled with addiction. Even many years later, thoughts of what could have been done differently are still with me.

Around this time, having started the PhD program at Lancaster and begun thinking about research topics, a Facebook post from a young woman with CF caught my attention: “Does anyone else out there with CF have pain every single day?” (paraphrased). After reading the responses from people with CF talking about their pain, I knew this would be my research topic.

A quick search of PubMed around this time yielded few studies, and most reported results from brief surveys or pain assessments. What was missing from the data, at first glance, was the people. Missing was the voice of those who matter most in the experience of pain in CF. Missing was their experience, their point of view, their stories.

Some colleagues cautioned about embarking on a study in an area so controversial and complicated. I borrow this very caution as my own rationale for studying pain in CF, but poet Rich (1986, p. 111) may have expressed it best when she wrote:

attends to physical, emotional, social and spiritual pain, described further in Chapter Four (Clark, 2018a; Saunders, 1965). The research took place in the US, at a large CF program within an academic medical centre between 2015 and 2019. Interviews occurred between September 2016 and June 2017.

1.3 Structure of the thesis

Chapter Two provides background information for the research, beginning with an overview of expected illness trajectory, incidence and life expectancy in CF. Following this overview are descriptions of the usual model of care delivery, symptoms and treatment, and palliative care in CF. Chapter Two concludes with an introduction to pain theory.

Chapter Three provides a summary of previous research about pain in CF. A description of how the literature search was conducted and an overview of findings are presented with key themes identified. The themes were (a) pain characteristics, (b) disease severity, (c) quality of life, (d) management of pain and (e) coping. Key to the literature review findings was that most research on pain in CF has been quantitative, and a gap exists in knowledge derived from a patient perspective.

Chapter Four describes the methodology and methods employed in this study. A description is given of the research paradigm, rationale for qualitative research, narrative approach, and methods. Gee's style of data analysis is introduced along with an explanation of how it was applied in this study (Gee, 1991). Ethical considerations are also discussed.

Chapter Five presents the thesis findings in the form of stanzas, a poetic style informed by Gee (1991). Three narratives that seemed essential to understanding the experience of pain in CF are presented: emerging awareness of what it means to have CF, social legitimacy and invisibility.

In Chapter Six, an overview of total pain theory, the theoretical lens employed in this study, is presented. A brief history of the development of total pain theory by Saunders (2006c) is followed by a critique of the theory and contemporary perspectives. Finally, thesis findings are discussed in relation to the literature and existing knowledge. The breadth and depth of pain and suffering in CF is viewed through the lens of total pain. Highlighted in the discussion are the relationships brought to light in this study between total pain, the gradual recognition of what having CF means to a person, social legitimacy and invisibility.

Chapter Seven concludes the thesis with a summary of the study and reiteration of the contribution to knowledge. The conclusion also describes implications for clinical practice, limitations, and recommendations for future research.

Chapter 2: Background

This chapter will provide an overview of cystic fibrosis, including symptoms, treatment burden, and worsening illness. An introduction to the model of care for CF in the US and internationally is then presented, followed by a description of palliative care in CF. The chapter concludes with an overview of pain theory in the past fifty years.

2.1 Cystic fibrosis overview

CF is a life-limiting, multisystem, genetic disease affecting an estimated 30,000 people in the US and 70,000 worldwide (Cystic Fibrosis Foundation, n.d.-a). CF is most recognised for its effect on the respiratory system, characterised by chronic pulmonary infection, usually leading to respiratory failure and death (Chen et al., 2018; Philip et al., 2008). Once known as a paediatric disease due to short life expectancy, CF is now a chronic disease, with adults comprising more than half of those with CF in the US (53.5%) and the UK (56.2%) (Cystic Fibrosis Foundation, 2018; Cystic Fibrosis Trust, 2018).

Clinical care in CF is complicated and usually delivered within accredited care centres in the US (Mogayzel, Dunitz, Marrow, & Hazle, 2014). Established by the Cystic Fibrosis Foundation in 1961, these care centres provide specialised care by multidisciplinary teams of physicians, nurses, social workers, dietitians and respiratory therapists (Mogayzel et al., 2014). Some teams also have pharmacists, psychologists, psychiatrists, physical therapists, child life specialists, or other specialists. The Cystic Fibrosis Foundation provides funding and accreditation to 130 care centres across the US and

recommends that people with CF are seen at least quarterly at the centres (Cystic Fibrosis Foundation, n.d.-b). Accreditation of CF centres provides some level of standardization across centres; however, variation may exist related to differences in location, institutional or health systems. For example, although individuals living farther away from their centre may receive consultative care there, much of their day to day care is provided by a local primary physician. Additionally, people with CF without health insurance may not have access to an accredited care centre, especially if the institution does not provide robust charity care or the person does not have documented status as a US citizen or resident. Others may have insurance that only allows one visit annually to an accredited care centre and requires other care to be provided within their network.

Accredited care centres are part of a comprehensive network of care, support, research and quality improvement efforts of the Cystic Fibrosis Foundation. Another integral component of this network is the CF Foundation Patient Registry, established in 1966 to compile information on health status and outcomes of those with CF who have consented to participate (Mogayzel et al., 2014). Collecting and analysing information from the Patient Registry has made significant contributions in creation of clinical care guidelines, research and quality improvement.

Since the 1961 establishment of accredited care centres in the US, people with CF are living longer, with a median predicted life expectancy of 46.2 years for someone born in 2017 (Cystic Fibrosis Foundation, 2018). Unfortunately, this organized model of multidisciplinary CF care provided through specialized care centres, although common in

higher income countries such as the US, is not consistent globally (Bell et al., 2020). In addition, although many countries now have national patient registries (Australia, Belgium, Brazil, France, Germany, Ireland, Italy, Netherlands, New Zealand, UK, US), other resource poor countries do not, across significantly large areas of the world (Africa, Asia, and the Middle East) (Bell et al., 2020). Adding to the disparity in CF care is a lack of access to medications for both routine CF treatment and to treat infection; lack of access to specialists; and challenges related to economic and political systems. These issues, in combination with the relatively low number of individuals reported to have CF, likely contribute to the disparity in CF care and outcomes among countries (Bell et al., 2020; Seyed Bashir et al., 2017). Alarming, in some countries (Brazil, Soviet Union, El Salvador, India, Bulgaria) life expectancy in CF is only half (or less) than that of the US and UK (Bell et al., 2020; Cystic Fibrosis Worldwide, n.d.).

Even in the US and UK, where infants with CF today are *predicted* to live longer, the reality is that CF remains life-limiting and people are still dying quite young, with a median age of death of approximately 31 years in 2017 (Cystic Fibrosis Foundation, 2018; Cystic Fibrosis Trust, 2018).

2.2 Symptoms and treatment burden

CF causes abnormally thick and sticky mucous that affects the respiratory, gastrointestinal, endocrine, reproductive and other body systems (Davis, 2006). The usual illness course in CF consists of repeated lung infections, known as pulmonary exacerbations, that lead to lung damage, eventual respiratory failure and death (Davis,

2006; Yankaskas, Marshall, Sufian, Simon, & Rodman, 2004). Most people with CF have pancreatic malabsorption (inability to absorb fats and protein), leading to weight loss and sometimes CF-related diabetes (CFRD) (Yankaskas et al., 2004). As they live longer into adulthood, people with CF experience increased complications and comorbidities (Simmons & Plant, 2015).

Treatment of CF is primarily symptom-driven, aiming to improve symptoms and prevent or treat infection (Davis, 2006; Yankaskas et al., 2004). A primary goal of the daily treatment regimen is to help loosen and cough up the sticky mucous, often using a vest-like device. Treatment usually includes medications to treat respiratory symptoms and infection, pancreatic enzymes to improve gastrointestinal symptoms and weight, and sometimes other medications to treat complications such as diabetes. The treatment regimen is often reported to be time-consuming and burdensome, requiring two to four hours of therapy daily (Sawicki, Sellers, & Robinson, 2009). Although there is no cure at this time, improved therapies, possibility of lung transplant, and recent development of CF transmembrane conductance regulator (CFTR) modulator medications addressing the underlying cause of CF all offer hope to some people living with CF (Dellon et al., 2018; Edmondson & Davies, 2016).

As knowledge about CF has increased, infection control has become a significant component of treatment. Lungs in CF are usually colonised with various organisms, and people with CF may risk exposing each other to dangerous bacteria or other pathogens. Recently published guidelines recommend that people with CF avoid close proximity to

others with CF, recommending they stay a minimum of six feet apart at all times (Saiman et al., 2014). An unfortunate consequence of the guidelines is the potential for decreased interaction and social support between people with CF (Flewelling, Sellers, Sawicki, Robinson, & Dill, 2019).

In addition to physical symptoms, people with CF experience a high rate of depression and anxiety (Quittner et al., 2016). Mental healthcare in CF has been emphasised through recent requirements for anxiety and depression screening and efforts to facilitate social support for people with CF and their families.

2.3 Growing older and sicker

As people with CF live longer, they transition between childhood and adolescence to adulthood. This transition occurs within the context of expected growth and development but also advancing illness. Rather than a clear delineation between states, there may be movement between states such as stages of growth and development or states of health and illness, sometimes referred to as liminality (Bruce et al., 2014; Lowton & Gabe, 2003). A serious illness such as CF, may disrupt or at least complicate the usual state of identity formation that occurs across the life trajectory (Williams, Corlett, Dowell, Coyle, & Mukhopadhyay, 2009). Referred to by Bury (1982) as biographical disruption, this theory describes how people respond to change and uncertainty and construct identity in chronic illness.

2.4 Palliative care in cystic fibrosis

Despite innovations in treatment, CF is still a serious, life-limiting illness, and palliative care is applicable across the continuum from diagnosis to death. Although evidence on palliative care in CF is limited, the body of knowledge is growing (Chen et al., 2018).

In the US, a large, multi-centre retrospective study examining end of life care patterns for adults with CF who died between 2011-2013, found people most often died in the hospital, often in intensive care (Chen et al., 2018). Palliative care services were utilized in 50% of patients, but this was inconsistent among US centres with some reporting that none of their patients received palliative care or hospice and others reporting all patients over this time received services (Chen et al., 2018; Dellon et al., 2016). Information about timing of services was not reported and thus it is possible services may have been received only in the hours to days leading up to death.

Dying in the hospital with limited utilization of palliative care services is not unique to the US. In a systematic review examining utilization of palliative care for adults with CF, with included studies reporting on deaths in the US, UK, Australia, and Canada, Marmor et al. (2019) and colleagues reported that 62%-100% died in the hospital, with one-third dying in intensive care units. Even in a study at a single CF centre in the US that integrated a specialist palliative care nurse and physician within the CF care team, with the palliative nurse and physician attending CF clinics and team meetings over a three-year-period, only 7% of patients died at home (Stephen J. Bourke et al., 2016). Although Marmor et al.

(2019) reported that existing evidence supports that specialty palliative care is beneficial in CF, findings also indicated poor access to specialty palliative care.

Challenges cited about palliative care in CF include unpredictable illness trajectory, difficulty with prognostication, lack of available palliative care services, and difficulties arising in the simultaneous pursuit of transplant and other types of aggressive care with preparing for possible end of life (Chen et al., 2018; Dellon, Hobler, Georgiopoulos, Chen, et al., 2019; Sands et al., 2011). These perspectives on challenges or perceived barriers to palliative care in CF appear to be held not only in the US, but also in those who care for people with CF from Australia, Canada, and UK (S. J. Bourke et al., 2009; Chapman, Landy, Lyon, Haworth, & Bilton, 2005; Marmor et al., 2019). One of the greatest barriers to palliative care in CF may be rooted in misunderstanding palliative care as relating only to death, rather than as a model of care relevant across the entire continuum of illness (Aslakson et al., 2017; Lowton, 2002a). Robinson (2009) has suggested that CF physicians and care teams may not refer to palliative care due to their opinion that the care is best provided by the CF team as they have often provided care to an individual and family for many years. Other potential challenges in access to specialty palliative care in CF stem from issues with models of palliative care involvement at different CF centres or availability of palliative care.

There are no accreditation requirements or standards for integration of specialist palliative care within CF care in the US, however, in a 2004 consensus guideline for adult care, the Cystic Fibrosis Foundation acknowledged that palliative care may be useful in

symptom management and more recently has provided funding for research to improve the delivery of palliative care in CF (Cystic Fibrosis Foundation, 2019; Linnemann et al., 2016; Yankaskas et al., 2004). Furthermore, a committee for the Cystic Fibrosis Foundation has recently developed and submitted specific guidelines for palliative care in CF and these are pending publication. In the European Cystic Fibrosis Society Guidelines, Castellani et al. (2018, p. 167) similarly call for “attention to communication, symptom control and a multi-disciplinary approach to care, including expertise in palliative care”.

Even in well-resourced countries, access to specialist palliative care is likely affected at an institutional level due to differences in size, scope and availability of palliative care. For example, some CF centres may be located in an institution that only provides inpatient palliative care. For those that have outpatient palliative care, access may be limited by diagnosis, for example cancer. Other limitations may exist due to the makeup of the palliative care team with some teams only including a nurse and/or physician and others having social work, spiritual care, psychiatry, volunteers, child life and other specialists.

Marmor et al. (2019) suggests that integration of palliative care within CF care is a challenge in the UK, Australia, and Canada, all well-resourced settings with organized systems of care for CF informed by national patient registries. Unfortunately, in countries without an organized network for CF care and without patient registries, little is known about CF care in general and even less is known about specialist palliative care delivery in CF. This is an area that would benefit from further research.

As part of a broader effort to improve palliative research and clinical care in CF, a working group of stakeholders, including experts in palliative care, CF and quality improvement, as well as people with CF and their caregivers, recently participated in a modified Delphi process to develop a definition of palliative care specific to CF. The definition created was:

Palliative care focuses on reducing physical and emotional symptoms and improving quality of life for people with CF throughout their lives. Palliative care occurs alongside usual treatments and is individualised according to the unique goals, hopes and values of each person with CF (Dellon et al., 2018, p. 420).

The definition was created for use as a tool to introduce the idea of palliative care to those living or working within the CF community. Additionally, the authors hoped the definition would support future research and clinical care in CF (Dellon et al., 2018). Addressing pain clearly falls within this definition and studying pain in CF will provide information for CF team members who report they feel unprepared for symptom management at end of life, and would like more education about palliative care in CF (Goggin & Cohen, 2016).

Notably missing in the CF definition is suffering, a common concept in discussions about pain and palliative care and its relief a core part of the World Health Organization's definition of palliative care (World Health Organization, 2019). Suffering encompasses the whole person, comprised not only of physical pain, but all elements of a person's life (E. Cassell, 1982; E. J. Cassell, 1991). Dellon et al. (2018, p. 419) noted in their study that during the Delphi process considerable difference of opinion existed among CF stakeholders in whether the word suffering would be perceived negatively or cause alarm or whether it was the only word "sufficiently broad to address the physical, emotional, and spiritual aspects of the illness experience" and the consensus definition did not

include it. Looking to literature around suffering in CF, a search of titles for “suffering” and “cystic fibrosis” through Lancaster Library OneSearch resulted in only 48 articles with all using suffering as a verb (i.e. suffering from cystic fibrosis) rather than a noun with suffering viewed as an experience.

2.5 Pain theory in the past fifty years

Pain is an ongoing subject of debate and theory in medicine and healthcare. Throughout the 19th and early 20th centuries, theories of pain tended to be based around physiological pain, and significant advances were made in the understanding of nociception and perception of pain (Moayedi & Davis, 2013). For example, the Specificity Theory of pain described sensory receptors receptive to specific stimuli and although there are hints of specificity in writings as early as the third century, it was tested and emerged as a developed theory in the 19th century through the work of von Frey, Sherrington and other researchers (Moayedi & Davis, 2013). Moayedi and Davis (2013) assert that despite the important contributions made through Specificity Theory and other earlier theories, they did not adequately consider the complexity of pain.

As theories continue to evolve since the mid-20th century, pain has been increasingly described as complicated and multidimensional (Moayedi & Davis, 2013). Definitions have moved away from descriptions centred on physiology of perception and response to pain, towards recognition of pain as a complex phenomenon involving emotions and cognition (Loeser, 2005). In their widely accepted definition of pain, the International Association for the Study of Pain defines pain as “an unpleasant sensory and emotional

experience associated with actual or potential tissue damage, or described in terms of such damage” (Merskey, Bogduk, & International Association for the Study of Pain. Task Force on Taxonomy., 1994, p. 210). This move away from a “Cartesian” view of pain as a dualistic function of mind and body (that saw body and mind as separate) is seen in pain theories introduced throughout the last half of the 20th century.

Conceived by Melzack and Wall (1965), the gate control theory of pain was one of the first theories to move away from a dualistic “Cartesian” view. Still taught in many schools of medicine, this theory proposed that the spinal cord acted as a gate to the brain by either letting pain in or keeping it out (Melzack & Wall, 1965). When later broadening this theory, Melzack and Casey (1968, p. 434) described pain as multidimensional, consisting of three components: sensory (sensation, intensity, quality), motivational (unpleasant experience), and cognitive (previous experience, cultural influences, context of experience), illustrated in Figure 1. This strengthened the existing model through integrating emotional aspects of pain and emphasising interaction between the components (Olson, 2013).

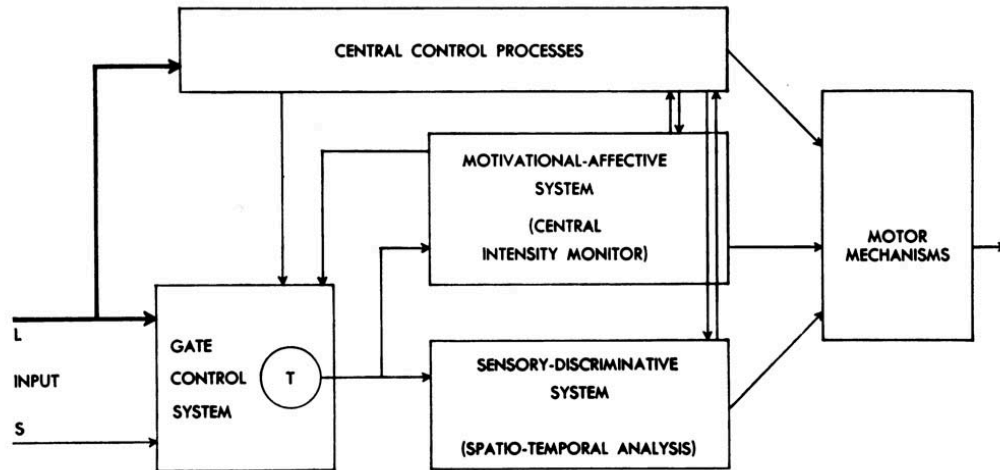


Figure 1. Melzack and Casey's model of pain. Reprinted from "Sensory, motivational, and central control determinants of pain: a new conceptual model" by Melzack and Casey (1968, p. 427).

More an advocate than a theorist, Margo McCaffery, an RN and pioneer in the field of pain management, nevertheless holds an important role in the field of pain. McCaffery described pain in her 1968 UCLA class syllabus as "whatever the experiencing patient says it is, and exists whenever he says it does" (Pasero, 2018, p. 89). This understanding valued the subjective experience of the person in pain, viewing treatment of pain as an ethical imperative, and eventually became a core concept of pain treatment in the US (McCaffery & Ferrell, 1997).

Building on models that envisioned pain as a complex experience, but recognising like McCaffery that pain is highly subjective, Loeser (1982) developed a nested or linear model of pain. Loeser (1982) described pain as consisting of four components: nociception (the ability to feel pain), pain, suffering, and pain behaviour. In his model, Loeser (2005) saw pain behaviour as the only component observable by others and all other components as

subjective, known only to the one experiencing it, as depicted in the nested model below (Figure 2).

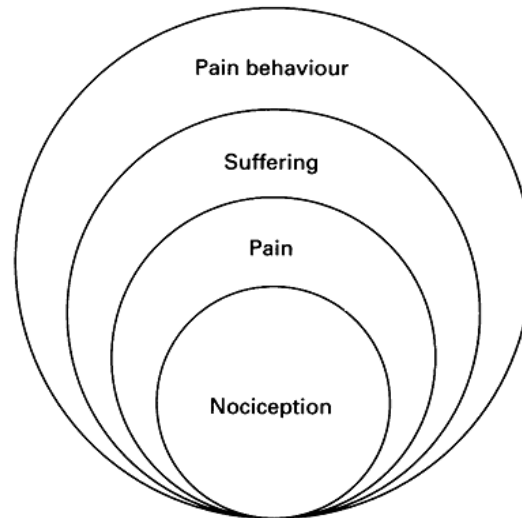


Figure 2. Loeser's model of pain. Developed from "Pain, Suffering, and the Brain: a Narrative of Meanings" in Narrative, Pain and Suffering by Loeser (2005).

Emphasis on the subjectivity of pain and recognition of the importance of each layer of the experience, including recognition that much of pain is invisible to all but the person experiencing it, provides a broader perspective of pain than the theories focused on the physical aspects of pain alone.

This chapter has provided an overview of CF, a multisystem genetic disease that despite increasing life expectancy is still a life limiting illness with many people dying in young adulthood from respiratory failure. A description of care delivery in CF was described as an organized network of care delivered by multidisciplinary teams for those in well-

resourced countries with less known about CF care in other areas of the world. An overview was provided of the worsening illness trajectory in CF and the treatment regimen and related burden usually beginning well before symptoms appear, with limited access to palliative care. In preparation to embark on this study of pain in CF, pain theory over the past 50 years was reviewed with Loeser's nested model initially favoured for its' inclusion of nociception, pain, suffering and pain behaviour and emphasis on the subjective nature of pain. Chapter Three will review the literature on pain in CF.

Chapter 3: Literature review

This chapter will introduce the concepts of pain in CF and describe the aim and methods of the review. The review findings will then be presented and discussed.

As people with CF live longer, they experience an increased symptom burden resulting from disease progression, CF-related complications and comorbidities (Sawicki, Sellers, & Robinson, 2008). Pain is a common phenomenon in this increased symptom burden of advancing CF (Festini, Ballarin, Codamo, Doro, & Loganes, 2004; Hayes et al., 2011; Kelemen et al., 2012). Pain has been associated with negative outcomes in CF, such as decreased quality of life, decreased treatment adherence, increased pulmonary exacerbations, and decreased survival (Abbott et al., 2009; Hayes et al., 2011; Kelemen et al., 2012). Despite these adverse effects, Festini et al. (2004) stated that pain may be underreported, with less than half of CF patients in their study reporting pain to their physician.

3.1 Aim

This review aimed to determine how the experience of pain for people with CF is described and examined in the literature. Secondary questions were a) what is known about types of pain in CF and its assessment, and b) what is known about the association of pain in CF with other symptoms or outcomes, and c) what is known from the perspective of the person with CF.

3.2 Methods

A systematic search of the literature was undertaken followed by a narrative synthesis of the findings to gain an understanding of what was already known about pain in CF. Aveyard and Bradbury-Jones (2019) report literature reviews usually lean towards either the highly structured 'systematic review' (such as developed by Cochrane or the Campbell Collaboration) or the more loosely structured 'narrative review'. Aveyard and Bradbury-Jones (2019) also note that literature reviews are increasingly described by a myriad of different terms and thus a clear description of the chosen approach is important. As more diverse evidence has been recognized as valuable and literature reviews have moved beyond assessment of intervention alone, approaches to undertaking literature reviews have evolved to encompass this heterogeneity (Aveyard, 2014; Aveyard & Bradbury-Jones, 2019). Approaches to literature reviews include systematic reviews, narrative reviews, scoping reviews, rapid reviews, and realist reviews (Aveyard, 2014; Popay et al., 2006; University of York Centre for Reviews and Dissemination, 2009).

Systematic reviews identify, critique, and synthesize empirical evidence to answer a specific question, often related to an intervention and whether it is effective (Aveyard, 2014; Cronin, Ryan, & Coughlan, 2008). Known to be rigorous and detailed, systematic reviews explicitly outline the methods followed to search, appraise, and analyse empirical evidence. Systematic reviews are often highly regarded, especially those published by reputable groups such as Cochrane and Campbell. A narrative review also brings together a body of literature, appraising and synthesizing the evidence but may not be as

transparent about the method of searches as is a systematic review (Aveyard, 2014; Cronin et al., 2008). Narrative reviews are sometimes criticized as having little rigour, without clearly defined methods, and positioned as a polar opposite to the systematic review. Although this perspective of a systematic review at one end of the quality spectrum with the narrative review at the opposite is correct at times, Aveyard and Bradbury-Jones (2019) found examples of less than rigorous reviews labelled as ‘systematic’ and high quality, rigorous reviews labelled as ‘narrative’ in their focused mapping review and synthesis, concluding that terms for literature reviews are poorly differentiated in practice and sometimes used interchangeably.

A scoping review is used to assess the size and types of evidence available in a specific area, often in the planning stages for a future study. Like systematic reviews, scoping reviews involve extensive searching for evidence but differ in that evidence is not synthesized (Aveyard & Bradbury-Jones, 2019; University of York Centre for Reviews and Dissemination, 2009). Rapid reviews are generally undertaken within a brief period (such as 90 days) and similar to a scoping review, are often used to inform a future study or policymakers when information is needed within a short time frame. With the limited time involved the breadth of evidence in a rapid review may be sacrificed but unlike the scoping review, evidence is synthesized (Aveyard & Bradbury-Jones, 2019). The realist review, like many systematic reviews, is focused on an intervention, but differs in looking beyond the question of whether an intervention works, seeking to answer why and in what context it works (Aveyard & Bradbury-Jones, 2019).

Within literature reviews, evidence from multiple studies is summarized, synthesized and presented in varying ways such as meta-analysis, meta-synthesis and narrative synthesis. Meta-analysis is a statistical method to combine quantitative data with similar properties to allow for greater statistical power and precision than can be achieved through the individual studies, while meta-synthesis is a non-statistical method to synthesize findings from qualitative studies (Cronin et al., 2008). Meta-analysis and meta-synthesis may be used separately or in parallel. In narrative synthesis, findings are presented in a narrative style, commonly used when statistical methods for pooling data are not feasible or suitable, often due to variability in study designs (Ryan & Cochrane Consumers and Communication Review Group, 2013; University of York Centre for Reviews and Dissemination, 2009). Synthesising data using a narrative style allows the main themes from heterogeneous studies to be identified and brought together (Mays, Pope, & Popay, 2005). Narrative synthesis is differentiated from the term narrative review, described earlier in this section as a term often used to describe a literature review that is not rigorous in approach (Popay et al., 2006).

A literature review, conducted from a constructivist perspective, values multiple research methodologies, including qualitative methods and smaller case studies. Evidence from diverse perspectives is important, and in health care evidence from the perspective of the person experiencing the phenomenon is especially important to enhance understanding of the individual's experience, and to inform clinical practice, health policy and research. The constructivist researcher values these diverse perspectives and through the literature review seeks to understand a phenomenon rather than to quantify or explain it.

A systematic approach employing narrative synthesis was chosen for this study. This approach aimed to help ensure the review was rigorous and transparent, with broad scope and depth; prioritizing critique, analysis, and synthesis of the evidence, whereas a scoping or rapid review was thought not appropriate as it risked sacrificing depth to facilitate rapid information. Although primarily quantitative, the heterogeneity of the literature meant that meta-analysis was not feasible. Narrative synthesis, using words and text to bring evidence together was selected as the best method to explain what was known about pain in CF. The process for narrative synthesis outlined by Popay et al. (2006) includes four primary elements: 1) theory development of how the intervention works, 2) preliminary synthesis development, 3) exploring relationships in the data, and 4) assessing the robustness or quality of the synthesis. A modified version of this process was used, excluding theory development as the review is exploratory and evidence is not intervention based. The guidelines for Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) format were followed in this review (Moher, Liberati, Tetzlaff, & Altman, 2009).

3.2.1 Database searches

The databases of AMED, CINAHL, PsycINFO, SCOPUS, Web of Science, and PubMed were searched in September 2015, without restriction on publication date. These databases were selected as they include a range of academic disciplines and research designs, and include journals that are known to publish articles about CF. An electronic “hand-search”

of the *Journal of Cystic Fibrosis and Pediatric Pulmonology* was performed. In addition, reference lists for all included articles were searched.

In July 2019, the search was updated to identify highly relevant articles published since the initial search. The search terms and databases were unchanged from the original search; however, the inclusion and exclusion criteria were tightened to include only adolescent and adult participants and qualitative research. One highly relevant article was located through this search update and has been integrated within the narrative synthesis.

The search was run a third and final time in January 2020 using additional search terms for cystic fibrosis (1. exp Cystic Fibrosis/ 2. cystic fibrosis.tw. 3. fibrocystic near disease near pancreas.tw. 4. mucoviscidos\$.tw.5. (cystic\$ adj10 fibros\$).tw) and for pain (1. pain 2. chronic pain 3. discomfort). Like the 2019 search update, a tightened inclusion and exclusion criteria included only publications reporting qualitative research in adolescent and adults. No further articles meeting the tightened search criteria were found.

3.2.2 Database search terms

A combination of MeSH and other search terms was used. For example, in PubMed, the following search terms were used in the titles and abstracts: ("Cystic Fibrosis"[Mesh]) AND (((("Pain Measurement"[Mesh]) OR "Pain Management" [Mesh]) OR "Pain"[Mesh]) Filters: Humans. In databases that did not incorporate MeSH terms, the words “cystic fibrosis” and “pain” were used. Subject librarians from Lancaster University and

University of California San Diego were consulted at various stages of the search. See Appendix A: Database summary for a list of databases and search results.

3.2.3 Eligibility criteria

Inclusion criteria for the narrative synthesis were a) studies with primary focus on pain in CF b) studies in people with CF of all ages c) studies published in English, and d) empirical studies reporting primary research. Articles were assessed for relevance through screening titles and abstracts using established eligibility criteria (see Appendix D: Inclusion and Exclusion Criteria). When suitability for inclusion was uncertain, a review of the article's full text was performed. Any uncertainties were resolved by discussion with a second reviewer (AB or SGB).

3.2.4 Data extraction

Following guidelines recommended by Popay et al. (2006), a preliminary synthesis was performed followed by exploring relationships, before assessing the robustness of the synthesis. The preliminary synthesis consisted of extracting descriptive characteristics of the studies using a data extraction tool designed for use in reviews containing disparate data (Hawker, Payne, Kerr, Hardey, & Powell, 2002). Extracted data was then summarised in the data extraction table in Appendix B.

Following extraction of participant and study characteristics, emerging themes were identified. An effort to manage existing knowledge and preconceptions related to insider status was an important part of the review process. An early tendency to pay more

attention to some aspects of findings than others, usually biomedical, was ameliorated through consciously staying in researcher role not clinician role as a way of reducing potential clinical bias. Cross checking in which AB and/or SGB reviewed an article with color-coded themes addressed any uncertainty or overlooked emerging themes. After identification of themes, narrative synthesis moved towards examining the relationships within and between studies.

3.2.5 Quality appraisal

A quality appraisal tool by Hawker et al. (2002) was used to evaluate and assess the quality of the data in this review. This tool takes a broad approach to quality assessment that allows comparison of heterogeneous studies of both quantitative and qualitative design. See Table 1 for an example of quality appraisal of an article using the tool adapted from Hawker et al. (2002).

Table 1

Example of Quality Appraisal of Epker, Maddrey, and Rosenblatt (1999), adapted from Hawker et al. (2002).

Category	Score	Category	Score
Abstract/Aims	2	Ethics/Bias	1
Introduction/Aims	4	Findings/Results	3
Method/Data	3	Transferability/ Generalisability	2
Sampling	2	Implications/ Usefulness	3
Data Analysis	3		

Total Score	23
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Note: Scores range from very poor (1 point) to good (4 points) per category.

Total available score ranges from 9-36.

Appraisals using the Hawker tool resulted in ratings from 21–31 (total possible 9–36). The median score for the included studies was 27.5. In 11 of the 14 articles, ratings for ethics and bias were “poor” or “very poor”, indicating only a brief mention (or no mention) of potential ethical issues, ethical approval, or relationship between participants and researchers. Articles were rated highest in the area of results, indicating the findings were clearly stated and directly related to results, but in some cases would have benefitted from additional explanation. See Appendix B for total quality appraisal scores listed in the Data Extraction Table and Appendix C for the Hawker tool.

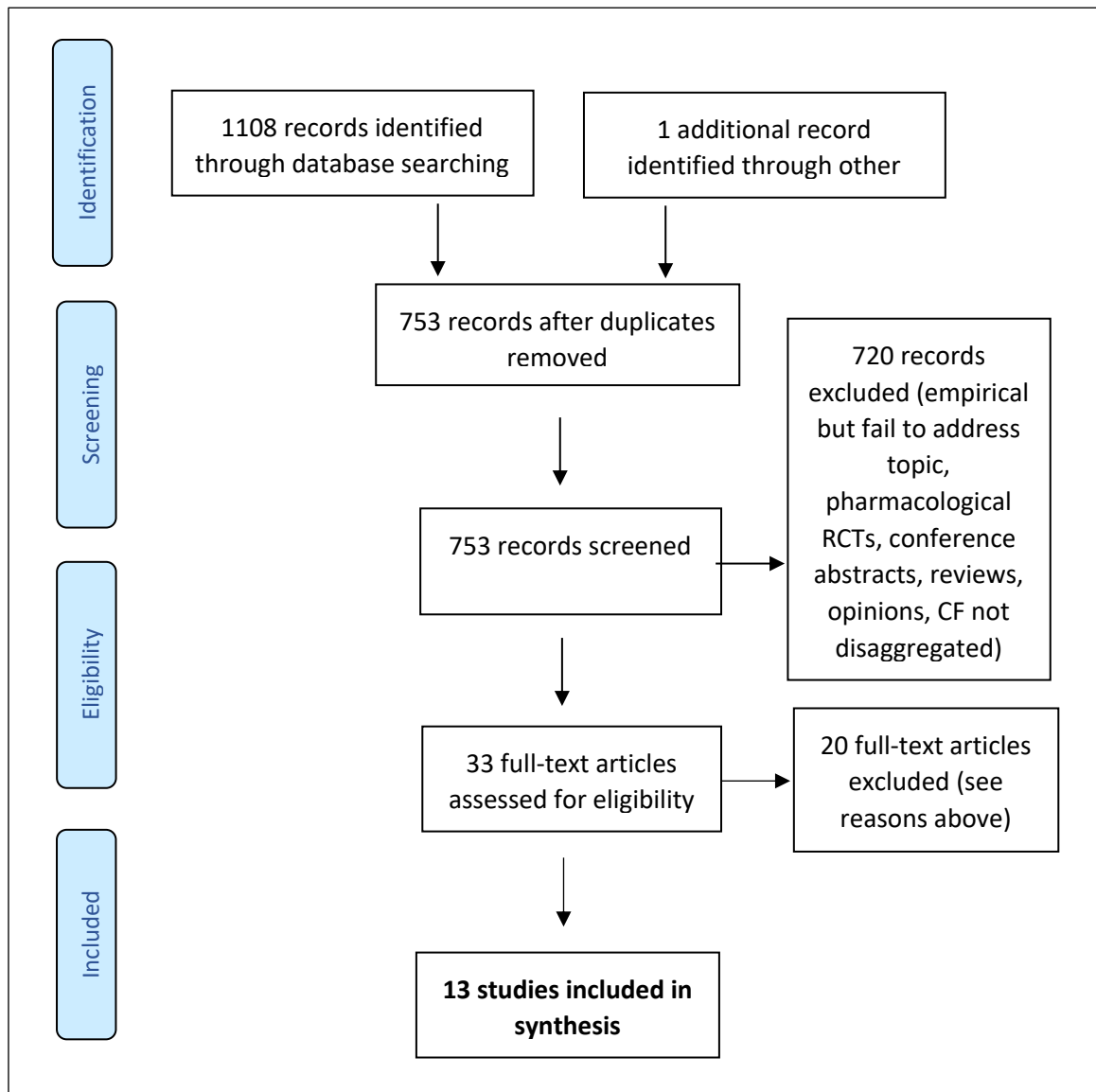


Figure 3. Retrieval Strategy Adapted from PRISMA Flow Diagram (Moher et al., 2009).

3.3 Literature review findings

In total, 1108 potentially relevant articles were identified, from which 13 of these articles fulfilled the eligibility criteria and were synthesised and appraised (see Figure 3). An

additional highly relevant article published in 2018 and identified in the search update was included in the synthesis, thus 14 articles are included in the synthesis.

3.3.1 Study characteristics

Publication dates ranged from 1990–2018. Of the 14 studies, most were conducted in the US (n=9), followed by France (n=2), Australia (n=1), Canada (n=1), and Italy (n=1). Sample sizes ranged from 10–239. The study populations included adults (n= 6), children and adolescents (n=3), combined populations (n=4), and adolescents (n=1).

Most participants were female (53%; n=598), with male participants comprising 47% (n=533). In studies of adults only (or combined studies that separated mean age of adult participants), the mean age ranged from 23–31.1 years. The mean age in the studies of children only (or combined studies that separated mean age for children) ranged from 10.2–13.6 years. The adolescent-only study had a mean age of 15.8 years. Stenekes et al. (2009) included children, adolescents and adults, and reported a mean age of 19.9 years (range 7–60 years). Two studies did not report mean ages (Allgood, Kozachik, et al., 2018; Hubbard, Broome, & Antia, 2005).

All but one of the identified studies were quantitative but were considerably disparate in data collection tools and study populations, including broad range of ages. Quantitative research designs included 13 observational studies (nine retrospective, three prospective and one chart review). Twenty-four different tools were used in the included studies. Only four tools were used in three or more studies, limiting statistical comparison of results

across studies. A single qualitative study, identified during the search update, employed a semi-structured interview design.

Pain theories, further described in Chapter Four, were almost absent across included studies. Epker et al. (1999) mentioned a “biopsychosocial perspective” but did not actually employ this as a theoretical lens. Similarly, Kelemen et al. (2012) also made brief mention of pain as a “multidimensional phenomenon” in support of including a measurement of pain catastrophising. No other studies, including the single qualitative study, employed pain theory, at least not in a manner transparent to the reader.

The concept of suffering was also absent from 10 of 14 studies with no mention of the word suffering in any context. Hubbard et al. (2005) and Kelemen et al. (2012) both used as a verb to indicate suffering from pain. Only Festini et al. (2004) and Epker et al. (1999) used suffering as a noun, referring to it as an experience. When suffering was mentioned, it was in background or discussion and not part of study findings.

3.3.2 Thematic analysis

The results of each study were coded according to common findings, and from these codes five themes were developed: pain characteristics, disease severity, quality of life, management of pain, and coping. Notation was made of differences in themes in relation to age or other participant characteristics.

3.3.3 Pain characteristics

The most frequent reported locations for pain in adults were the head (n=4/8), chest (n=2/8) and back (n=2/8) (Festini et al., 2004; Flume, Ciolino, Gray, & Lester, 2009; Hayes et al., 2011; Kelemen et al., 2012; Ravilly, Robinson, Suresh, Wohl, & Berde, 1996; Sermet-Gaudelus et al., 2009). The most frequent location of pain in children and adolescents was the abdomen (n=3/3) (Blackwell & Quittner, 2014; Koh, Harrison, Palermo, Turner, & McGraw, 2005; Sermet-Gaudelus et al., 2009). One study with a combined population of adolescents and adults reported the chest as the most frequent location of pain; another that combined children and adults reported the head as the most frequent site (Hubbard et al., 2005; Stenekes et al., 2009). Three studies did not report pain location. Although frequency was not reported by Allgood, Kozachik, et al. (2018) in their qualitative study, locations were consistent with the other studies.

People with CF were most likely to report pain intensity in the moderate range (n=5) (Festini et al., 2004; Flume et al., 2009; Hayes et al., 2011; Hubbard et al., 2005; Sermet-Gaudelus et al., 2009). Others with CF reported mild pain intensity (n=4) (Blackwell & Quittner, 2014; Epker et al., 1999; Kelemen et al., 2012; Koh et al., 2005). Four studies either did not report intensity or only reported it as a correlation with other variables (Munck et al., 2012; Palermo, Harrison, & Koh, 2006; Ravilly et al., 1996; Stenekes et al., 2009). Participants in the qualitative study were included if they reported moderate to severe pain at least weekly for at least a month (Allgood, Kozachik, et al., 2018). When

examined by age group, adult participants reported higher pain intensity (moderate n=4; mild=2) than children and adolescents (moderate n=1; mild n=2).

Tools used to assess pain intensity were all based on self-report. The tools were the Multidimensional Pain Inventory (MPI), Brief Pain Inventory (BPI), Faces Pain Scale/Faces Pain Scale-Revised, and ad hoc scales developed by the researcher, with frequency of use listed in Table 2 below (Bieri, Reeve, Champion, Addicoat, & Ziegler, 1990; Hicks, von Baeyer, Spafford, van Korlaar, & Goodenough, 2001). The lack of consistency in tools used to measure pain intensity, and variation in how the questions were asked, made synthesis of severity challenging and limits interpretation of results. For example, Festini et al. (2004) asked participants only about the maximum degree of pain intensity, while Kelemen et al. (2012) inquired about both average and worst intensity.

Table 2

Pain measurement tools used in studies

Pain Measurement Tool	Frequency
Multidimensional Pain Inventory (MPI)	1
Brief Pain Inventory (BPI)	3
Faces Pain Scale & Faces Pain Scale-	3
Adhoc scale developed by researcher	5

The reported prevalence of pain ranged from 64% to 94.1% of adults in the past one to two months. Ravilly et al. (1996) reported 84% prevalence in the posthumous group. Prevalence in children and adolescents ranged from 59% to 89%.

Words used by adolescents to describe pain, selected from a list of terms, included “sore”, “aching”, “pounding”, “cramping”, and “stiff” (Blackwell & Quittner, 2014). Allgood, Kozachik, et al. (2018) reported similar words in adolescents, and also noted that adults tended to describe their pain more thoroughly rather than a single descriptive word. No other studies described words or language used to talk about pain.

Most studies did not differentiate between acute, chronic and procedural pain. Procedural pain was assessed in two studies, with participants reporting procedural pain as mild (Koh et al. (2005) and common (Sermet-Gaudelus et al. (2009). Treatment-related pain was prevalent in adolescents, with airway clearance therapy the most common cause (Blackwell and Quittner (2014). Chronic pain, defined as pain most of the time for six months or more, was reported in 27% of adults according to Hayes et al. (2011). The qualitative study differentiated pain related to CF versus other causes (Allgood, Kozachik, et al., 2018).

3.3.4 Disease severity

Disease severity was most often assessed through pulmonary function measured by FEV1 (forced expiratory volume in one second), a common measure of disease severity in CF. Other parameters used were clinician assessment and the Shwachman Rating of Illness

Severity (Shwachman & Kulczycki, 1958; Stollar, Adde, Cunha, Leone, & Rodrigues, 2011). Most studies reported FEV1 as a mean or median as depicted in Table 3. In studies using FEV1 as a proxy for disease severity, there was no association with pain (Flume et al., 2009; Hayes et al., 2011; Kelemen et al., 2012; Koh et al., 2005; Sermet-Gaudelus et al., 2009). Epker et al. (1999) examined the relationship between pain severity and Shwachman Rating of Illness Severity, determining that pain was only associated with history and general activity level, and not associated with pulmonary physical findings and cough, nutritional status or chest x-ray findings.

Table 3

Pulmonary function as measured by Forced Expiratory Volume in 1 second (FEV1) and population in included studies.

Author(s)	Population	Mean or Median FEV1 <40%	Mean or Median FEV1 >40%-69%	Mean or Median FEV1 >70%	Comments
Allgood, SJ et al.	Adolescents Adults				Mean/median not reported (range: 23.5%-102%)
Blackwell, L S and Quittner A	Adolescents			X	Mean 80%
Epker, J A, et al.	Adults				FEV1 not reported
Festini, FS, et al.	Adults		X		Median 56.71%
Flume, PA et al.	Adults		X		Mean 58.4%
Hayes, M et al.	Adults		X		Median 63.6%
Hubbard, PA et al.	Adults				FEV1 not reported
Kelemen, LA, et al.	Adults		X		60.5% (unknown whether mean or median)
Koh, JL et al.	Children			X	Mean 80%
Munck, AA et al.	Children			X	Mean 85%
Palermo, TM et al.	Children Adolescents			X	Mean 80.2%
Ravilly, S et al.	Adults		X		Mean 58%
Sermet-Gaudelus JP, et al.	Children Adolescents Adults		X	X	Mean 70% (children); Mean 50% (adults)
Stenekes, SM et al.	Children Adolescents Adults				Mean/median not reported 40% with FEV1>81%; 29% FEV1 61-80%; 22% FEV1 41-60%; 4% FEV1 <40; 5% UNK

3.3.5 Quality of life

In addition to examining potential physical correlates of pain, most studies also examined potential relationships between pain and psychological correlates or quality of life (Blackwell & Quittner, 2014; Epker et al., 1999; Festini et al., 2004; Hayes et al., 2011; Hubbard et al., 2005; Kelemen et al., 2012; Munck et al., 2012; Palermo et al., 2006; Sermet-Gaudelus et al., 2009; Stenekes et al., 2009).

One of the most common tools used to measure quality of life was the Cystic Fibrosis Questionnaire–Revised (CFQ-R). The CFQ-R is a validated disease-specific questionnaire designed to assess various domains of quality of life in CF, including physical symptoms and functioning, social functioning and treatment burden (Quittner, Buu, Messer, Modi, & Watrous, 2005). Four studies used the CFQ-R, and two used the previous version, CFQ. Pain was found to be significantly related to CFQ-R domains for physical functioning, social life, treatment aspects, emotional response, and school or work (Blackwell & Quittner, 2014; Hayes et al., 2011; Kelemen et al., 2012; Munck et al., 2012; Palermo et al., 2006).

There was some evidence of associations between sleep and pain. Flume et al. (2009) reported that the relationship of pain to impaired sleep quality was strongly correlated, and in another study 30.2% of those who experienced pain also reported sleeping disorders (Festini et al., 2004). Pain was also indicated as a cause of insomnia (Sermet-Gaudelus et al., 2009).

Depression and anxiety were significantly related to pain, as measured by the Hospital Anxiety and Depression Scale (HADS) and the CFQ-R (Blackwell & Quittner, 2014; Hayes et al., 2011). Higher reported symptoms of depression or anxiety were associated with higher pain intensity (Blackwell & Quittner, 2014). Pain catastrophising, defined as a set of exaggerated and negative cognitive and emotional symptoms during actual or anticipated painful stimuli, was found to be associated with pain (Hayes et al., 2011; Kelemen et al., 2012; Sullivan, Bishop, & Pivik, 1995).

Pain was associated with interference in daily activities, work and school (Festini et al., 2004; Hubbard et al., 2005; Kelemen et al., 2012; Stenekes et al., 2009). Most studies found an association between pain and CF treatment (Epker et al., 1999; Festini et al., 2004; Hayes et al., 2011; Hubbard et al., 2005; Kelemen et al., 2012; Koh et al., 2005; Palermo et al., 2006; Sermet-Gaudelus et al., 2009). Similar themes about pain negatively impacting education, work and social activities were reported in the qualitative study (Allgood, Kozachik, et al., 2018).

3.3.6 Management of pain

Only one study addressed medication use, reporting NSAIDS were most often used, while 10 of 78 participants received opioids for more than three months (Ravilly et al., 1996). Limited evidence on medication pain management may be due to exclusion of pharmacological RCTs.

Non-pharmacologic management of pain included rest, physical activity, massage, acupuncture and chiropractic (Festini et al., 2004; Hayes et al., 2011; Koh et al., 2005; Ravilly et al., 1996). In two studies, some participants said they did nothing to treat their pain, an interesting finding given that pain has also been identified as underreported in CF (Blackwell & Quittner, 2014; Festini et al., 2004).

Festini et al. (2004) reported that only 46.2% of participants talked to a physician about pain. In contrast, Allgood, Kozachik, et al. (2018) reported that all 10 interview participants indicated they spoke to their healthcare providers about pain, and most also felt able to talk about pain with friends, family, and school or work colleagues. Allgood, Kozachik, et al. (2018) reported adolescents felt supported during these discussions about pain, while adults felt stigmatised.

3.3.7 Coping

Hubbard et al. (2005) examined coping techniques in participants with CF and pain, finding most participants used active coping techniques such as problem-solving, acceptance and self-encouragement more than passive techniques such as minimising pain, self-isolation and behavioural disengagement. Coping type was not associated with the intensity or duration of pain (Hubbard et al., 2005). Interpretation of coping mechanisms varied across studies, with Blackwell and Quittner (2014) reporting most frequently used coping mechanisms as rest, relaxation and taking medication.

3.4 Discussion

The aim of this narrative synthesis was to identify what is known about pain in CF, and what is known about associations between pain and other symptoms or outcomes in CF. All included studies but one were quantitative. Although several studies did include quality of life measurements, most were quantitative and did not provide (or at least did not report) an opportunity for participants to provide information from their perspective and in their own words. The lack of experiential qualitative knowledge in the literature is reflective of a research environment that values quantitative evidence over qualitative. It also reflects the perception of pain as a singular symptom to be assessed and treated in isolation, rather than a complex phenomenon best suited to a qualitative approach to explore personal experience and increase understanding.

The quality of included articles was assessed as between poor and fair, indicating improved research design in this area would be beneficial. Studies were rated the lowest on ethics and bias, with some studies not mentioning ethics at all (n=4) and others with only a minimal mention (n=7). The quality of future studies can be improved through clearly stating information about ethical issues and how they have been, or will be, addressed. Consideration of the relationship between researchers and participants should also be clear, with the researcher demonstrating awareness of potential bias.

Despite some issues with quality, evidence does suggest that pain is a common issue in CF, with prevalence ranging from 59% to 94.1%. This finding is especially interesting given at least two studies revealing that participants with CF were not reporting pain to their

clinicians. If pain is prevalent in the CF population, then why is it not reported in some cases? This area should be further investigated, preferably in a qualitative study that allows more in-depth exploration.

Pain was reported to be significantly related to quality of life, including physical functioning, social life, treatment aspects, emotional response, and school or work, and these relationships should be further explored. As adults with CF live longer and experience the usual activities of life including education, careers and relationships, impact of pain on these developmental activities is important to study. Similarly, the impact of pain on other quality of life indicators, anxiety and depression should be further studied. With evidence suggesting pain impacts daily activities, especially usual CF treatment, this may create a cyclical effect with increased pain causing decreased adherence to routine therapies, which in turn may lead to increased pain and so on.

Pain management was not a primary focus in most studies, as they focused more on pain characteristics and impact on quality of life, at least in part because this review excluded pharmacological RCTs. Although also limited, more information was available about non-pharmacological management of pain with participants using rest, massage, acupuncture and chiropractic. Some participants reported doing nothing to treat their pain and this should be further explored.

Coping was also studied in relation to pain in CF. As most studies were quantitative, the possible coping mechanisms reported were chosen from an established list and may not have captured all of the possible coping strategies actually used.

3.5 Limitations

There are several limitations identified in this review. First, most studies had a small sample size, but this is at least in part due to low prevalence of CF. Studies were primarily retrospective, and participants may not have had adequate recall of their pain. In addition, studies performed in a CF clinic setting may have included more outpatients and therefore weighted the sample towards a healthier population, but this information was not consistently described.

Multiple tools are used to measure pain in CF, many of which are ad hoc tools that lack validation and allow little ability to reliably compare results across studies. Notably, the tool developed by Hawker et al. (2002) for quality appraisal has been criticised for lack of sensitivity, but its purpose in this review is to provide a general assessment of quality rather than to exclude studies.

Another limitation was lack of consistency in definitions for concepts including pain, functioning, quality of life, and disease severity. Finally, all but one study that met the inclusion criteria were of quantitative design and engagement with literature on pain theory was virtually non-existent. Despite these limitations, the review was systematic and thorough, enabling an adequate answer to the question of what is known about the experience of pain in CF.

3.6 Conclusion

This review has demonstrated that pain is an important issue in CF that may affect quality of life, mental health and adherence to treatment. Both pain and CF are recognised as complex phenomena, yet existing research about pain within CF is almost exclusively quantitative in design. Of the 14 included studies, 13 employed pain assessment tools and surveys and one was a retrospective chart review and thus did not include assessment or surveys. The tools demonstrated in these studies tended to only provide information on clinical aspects of pain as deemed important by the tool's developer or researcher. Only one pain-based study also sought the perspective of the person with CF; a significant gap in the evidence that provides rationale for undertaking this study.

The literature review findings suggest a gap in knowledge about pain in CF from the experiential perspective of the person. There is a case to explore these experiences and using a qualitative approach provide an opportunity for people with CF to share their personal stories about pain. Qualitative research is a valuable way to deepen understanding about the experience of pain in CF and provide a foundation of knowledge to inform future studies, both quantitative and qualitative. The existing literature on pain in CF was found to lack engagement with theory.

The rationale underpinning selection of a qualitative approach, characteristics of narrative methodology, and a detailed description of methods are presented next in Chapter Four.

Chapter 4: Methodology

In Chapter Three, the literature review revealed pain is a key issue in CF and may affect many aspects of quality of life yet seems underreported to CF clinicians. In reviewing the literature, existing research about pain in CF was found to be chiefly quantitative. Pain theorists within the last fifty years have incorporated multidimensional aspects of pain within their models to address complexity of the pain experience, but these models are underrepresented in current evidence about pain in CF. This gap in the literature confirmed the need to better understand the experience of pain in CF.

The aim of this research study is to explore the complex experience of pain in cystic fibrosis from the perspective of those living with it. Next, Chapter Five will describe the philosophical assumptions, methodology and methods for the study, along with the rationale for their selection.

4.1 Research paradigm

Research paradigms, described by (Guba & Lincoln, 1994, p. 107) as a “set of basic beliefs”, are closely connected to all phases of research and provide a foundation or framework for key decisions made by the researcher. Research paradigms attend to ontological positions (the nature of reality and its characteristics), epistemological positions (nature of knowledge), axiological positions (values), and methodological positions (how knowledge is obtained) (Creswell, 2013). The particular paradigm, and how it guides a researcher’s approach, should be transparent (Guba & Lincoln, 1994). The

aim of this study is to better understand the experience of pain in CF, and the research paradigm must therefore support exploration of these experiences.

Historically, paradigms in health research have been largely positivist (Pope & Mays, 2006; Saks & Allsop, 2013). In a positivist paradigm, reality is understood as an objective, single reality that can be measured and known (Guba & Lincoln, 1994; Saks & Allsop, 2013). Positivist researchers usually adopt quantitative methodologies such as RCTs or surveys, and look to identify relationships between variables such as cause and effect (Bryman, 2012). While valuable information is gained through positivism, this paradigm does not best recognise multiple, unique realities of experience and therefore another paradigm, social constructivism, will be considered.

In social constructivism, individuals are understood to learn and develop meaning through social activities, sometimes called social construction (Creswell, 2013; Crotty, 1998). The ontological perspective in social constructivism is that reality is relative – that rather than one true reality, there are multiple realities subject to interpretation (Guba & Lincoln, 1994). In other words, reality in social constructivism is socially negotiated. As individuals experience the world and interact within it these subjective realities continually evolve (Bryman, 2012; Moen, 2006). By adopting a social constructivism paradigm, evolving and varied perspectives are valued in understanding unique pain experiences in CF.

Epistemological assumptions within social constructivism hold knowledge as socially constructed, with a close relationship between the knower and the known (Crotty, 1998;

Guba & Lincoln, 1994). Knowledge stems from subjective experiences of participants and in this construction of knowledge, researcher and participant are inextricably connected. This connection occurs through interaction between researcher and participant; which stories the participant chooses to tell, and how the researcher subsequently reacts to these stories, ultimately create knowledge (Guba & Lincoln, 1994).

Where ontological and epistemological assumptions are concerned with reality and knowledge, axiological assumptions are related to values (Creswell, 2013). Social constructivists recognise that values influence almost all aspects of research. Researchers' values are expressed through which topics are chosen as worthy of research, whose voices are heard, what communications are encouraged or discouraged, and what data are chosen for the findings. Similarly, participants choose what information to communicate, what details are deemed more or less important, and what they choose to leave out entirely. Finally, editors and publishers decide what is worthy of publication, and readers choose what to read and pay more or less attention to.

Just as social constructivist researchers believe research paradigms should be transparent, they also strive to recognise their values and biases and clearly communicate them (Guba & Lincoln, 1994). Through communicating their own values and biases, they position themselves to show how their interpretation may be influenced by their unique background and experiences (Guba & Lincoln, 1994).

4.2 Qualitative rationale

Underlying assumptions related to ontology, epistemology and axiology suggest particular research approaches as more or less congruent with social constructivism. Through recognition of multiple realities as socially negotiated and knowledge as socially constructed, social constructivist researchers seek complexity of experience and rely on an individual's view of their experiences (Guba & Lincoln, 1994). In seeking this complexity, social constructivist researchers also examine the interaction and knowledge formed between researchers and participants. For this study, the selected approach must then provide a format for participants to describe their experiences in depth, facilitate understanding of these experiences and explore how they construct and make meaning with these experiences (Robson, 2011).

There are two primary research approaches, quantitative and qualitative. In a quantitative approach, data are usually expressed in numerical form and then categorised, ranked or analysed in relation to other variables using statistical analysis. In contrast, data in a qualitative approach are non-numerical in form, such as interview transcripts or observations.

A qualitative approach was chosen for this study to facilitate a more complex and deeper understanding of experience than would be gained through quantitative means (Braun & Clarke, 2013; Creswell, 2013). Having decided that a qualitative approach was ideally situated to answer the research question, determination of which specific qualitative methodology is most appropriate was considered next.

In a social constructivist qualitative approach, methodology should be interaction- and discussion-based (Guba & Lincoln, 1994). Several qualitative methodologies facilitate exploration of experiences and meet the criteria of being interaction- and discussion-based, including phenomenology, grounded theory, ethnography and narrative (Creswell, 2013). Phenomenology aims to describe the meaning of lived experience (phenomenon) shared by several individuals, focusing on shared aspects, or essence of the experience (Creswell, 2013, p. 76). Grounded theory methodology moves past description and application of current theory and aims to generate new theories (Creswell, 2013). A key feature of grounded theory is that the theory emerges or is grounded in the data. Although grounded theory may involve interviews, the focus is usually on factors that affect social processes in a particular phenomenon (Braun & Clarke, 2013). In ethnography, the researcher describes and interprets behaviour of a group that shares a culture, primarily through participant observations in a natural setting. Analysis in ethnography often provides interpretations of how groups work or function, language, values, behaviours, and institutional patterns (Hammersley & Atkinson, 2007). Narrative methodology recognises the importance of stories people construct and how they make meaning through these stories (C. K. Riessman, 2008). In a narrative approach, researchers aim to explore participants' unique experiences, how they tell stories of their experience, and their relationship with an experience over time.

With limited evidence available in the literature review, any of these qualitative methodologies could make a meaningful contribution to understanding the experience of pain in CF. However, not every qualitative methodology is ideally situated to meet the

aims of this particular study. After careful consideration, narrative methodology was identified as most congruent with the aim, epistemological, ontological and axiological assumptions of this study. With a focus on the unique and evolving experiences of individuals, narrative is particularly congruent with the ontological perspective of this study, social constructivism, in which reality is multiple and subjective, which has been discussed in section 5.1. The co-construction that occurs through narrative research further situates it in the epistemological perspective of knowledge as socially constructed. Finally, the axiological positioning of narrative with an emphasis on participants telling the stories they choose to tell in the way they choose to tell them, and the transparent positioning of the researcher further support the positioning of narrative within social constructivism.

4.3 Narrative research methodology

4.3.1 General description

Narrative research has origins in multiple disciplines including literature, history, linguistics, sociology and anthropology (Andrews, Squire, & Tamboukou, 2013). As a research methodology, narrative has been developed as a means of exploring experience over the past three decades, and is increasingly used in health and social research (Elliott, 2005; Margarete Sandelowski & Barroso, 2006). Narrative research has been reported as useful in nursing research around health and illness (Haydon & van Der Riet, 2017; Wang & Geale, 2015). Using narratives exploring an individual's experience with illness contributes to understanding and subsequently informs both health policy and clinical

care. The particular attention to relationship in the co-construction of narrative is equally important in partnerships in healthcare between patients and families with nurses and other clinicians and is at the core of conveying compassion and building trust. Additionally, narrative is reported as beneficial to illuminate patient experience in palliative care, whether to accompany quantitative methods or as a stand-alone method for qualitative research (Thomas et al., 2009). Individual nuances in the experience of patients and clinicians, and in the relationships and interactions that occur between them come forth through narrative. These nuances of experience further understanding, leading to insights in clinical care, and future directions for research, health policy, and education (Bingley, Thomas, Brown, Reeve, & Payne, 2008). In this study, narrative methodology was chosen to facilitate insight into the experience of pain in CF.

The term “narrative” is used in different ways and meaning may vary depending on who is speaking. It is therefore difficult to come to a consensus on a precise definition (Andrews et al., 2013; C. K. Riessman, 2008; Smith & Sparkes, 2009). Although narrative may be used as a synonym for story, narrative as a methodology encompasses more than just the stories or narratives themselves (C. K. Riessman, 2008). According to Clandinin and Connelly (2000, p. 18) “narrative is both the phenomenon and the method of the social sciences”. To gain insight into serious illness when facing end of life, Bingley et al. (2008) recommends that researchers not only engage with the narrative itself, but employ narrative methodology to do so. Although Bingley et al did not include CF illness narratives in their analysis, the end of life experiences described are echoed in CF (Bingley et al., 2008).

In perhaps the simplest description, narrative is defined as a way to understand experience (Smith & Sparkes, 2009). C. K. Riessman (2008, p. 6) further describes narrative methodology as having three important distinctions: “the practice of storytelling (the narrative impulse – a universal way of knowing and communicating...); narrative data (the empirical materials, or objects for scrutiny); and narrative analysis (the systematic study of narrative data)”. This comprehensive understanding described by C. K. Riessman (2008) informs the present study, along the continuum from planning to data collection and analysis.

4.3.2 Importance of stories

A key assumption in narrative is that of story as the basic unit through which experience is communicated and a fundamental way that people make sense and meaning of these experiences (Elliott, 2005; Mishler, 1986; C. K. Riessman, 2008; M. Sandelowski, 1991). In the words of Polkinghorne (1988, p. 1), narrative is “the primary scheme by which human existence is rendered meaningful”. Narrative researchers seek to explore experiences and their meanings through the rich data found within stories (Clandinin & Connelly, 2000). Mishler (1986) drew attention to the importance of listening to people’s stories when conducting qualitative interviews, taking active efforts to encourage rather than silence these stories. Narrative researchers assert that facilitating these stories through narrative approaches makes sense since people are naturally storytellers (Charon et al., 2016; Kleinman, 1988; Mishler, 1986; Moen, 2006; Polkinghorne, 1988; C. K. Riessman, 2008).

4.3.3 What counts as narrative?

Narrative stories may be constructed from different types of data including interviews, observations, written documents, and visual data such as paintings or photographs (Andrews et al., 2013; C. K. Riessman, 2008). Stories may encompass a single experience or phenomenon, or an entire life (Elliott, 2005). Although narratives can be constructed from different sources, much narrative research uses data constructed through interviews, as is the case in this study (C. K. Riessman, 2008).

4.3.4 Co-construction of narratives

A key concept in narrative methodology is that of narratives as co-constructed (A. W. Frank, 2010; Mishler, 1986; C. K. Riessman, 2008). Narratives are created through interaction when narrative researchers co-construct and analyse the stories individuals tell about their experiences. In this study, narratives are co-constructed through the research interview. A narrative research interview facilitates storytelling, something C. K. Riessman (2008, p. 23) called a “narrative occasion”. A narrative interview has a conversational style, allowing the participant to freely tell their story with fewer interruptions than seen in other styles of interviewing or everyday conversation (C. K. Riessman, 2008). The goal in a narrative interview, through this conversational style, is to construct a detailed account rather than the more familiar brief response seen in other types of qualitative interviews (C. K. Riessman, 2008). In practice narrative interviews require the same level of preparation and adherence to high research and ethical standards (Beuthin, 2014; Pederson, 2013).

Narratives do not literally represent reality but rather present constructions of an individual's experience within a social context or community (Crotty, 1998). This perspective of narrative as co-constructed is aligned with underlying assumptions of social constructivism, where meaning is constructed through a person's interaction with the world (Crotty, 1998). Co-construction highlights the dialogical nature of narrative, regarding the participant as an important aspect of the co-constructed narrative (A. W. Frank, 2010). In creating a narrative, both researcher and participant are working with the story, exploring and creating it together. The personal account of illness experience and how it has affected one's life is known as an illness narrative (Kleinman, 1988). Illness narratives have become an influential area of study over the past several decades (Charmaz, 1999; A. Frank, 2009; A. W. Frank, 1998; Mishler, 1995; Smith & Sparkes, 2005). According to Cardillo (2010), one can gain a greater awareness and understanding by focusing closely to the illness narratives of those who have grown up with chronic conditions, such as CF.

4.3.5 Why narrative?

A key consideration in selecting a methodology is looking at what the specific approach does. In other words, what does narrative help discover about pain in CF that other methodologies may not? Narrative methods allow researchers to discern how participants understand their lives across the life course, accounting for temporality of experience (Clandinin & Connelly, 2000; C. K. Riessman, 2008). This aspect of time, to hear through stories how an individual reflects on their past, considers their present and

anticipates the future, is a valuable characteristic of narrative. People make meaning through storytelling, and these stories can help shed light on how meaning may evolve over time or differ even within the same conversation (Clandinin & Connelly, 2000). Sometimes, narratives may provide insight into why some stories are silenced and others are heard, identifying opportunities for improvements in care (Andrews et al., 2013).

Although they use varying approaches, narrative researchers share in common an interest in improving understanding of the illness experience through the stories that people tell. For example, Ahlsen, Mengshoel, and Solbrække (2012a, p. 317), in their study of men with chronic pain, used narrative methodology based on typologies such as “being comforted” and “being connected”, similar to the quest, chaos and restitution narratives depicted by A. W. Frank (2000). Through narrative, they learned that men found meaning beyond physical recovery at the rehabilitation clinic, men spoke of finding social connection and rebuilding the self (Ahlsen et al., 2012a). In another study on chronic pain, Ahlsen, Mengshoel, and Solbrække (2012b) employed narrative methodology with attention to both content and structure. This approach, also with men who are receiving rehabilitative care, facilitated recognition that below the surface in accounts on their objective experience with pain, the men shared vulnerability in their suffering and struggles with losing control (Ahlsen et al., 2012b). Dysvik, Sommerseth, and Jacobsen (2011) applied a traditional thematic analysis to a case study about a woman with chronic pain but did not use narrative methodology. While the case study provided details of experience a narrative approach may have yielded additional understanding such as the social context and temporal aspects of experience. In young adults with CF, Oddleifson

and Sawicki (2017) used thematic narrative analysis, including attention to the context in which stories were told. This shed light on the social world of the young adult with CF and the connections between this social world, adherence to the CF treatment regimen, and sense of self (Oddleifson & Sawicki, 2017). Moola and Faulkner (2014) analysed illness narratives of two children with CF. Their approach includes robust narrative methodology attending to content, structure, dialogic and performance aspects of narrative methodology. Through this approach they reveal the complexity of the child's experience with CF, illuminating how life-threatening illness, a child's developmental stage, social and cultural background may influence the stories they tell (Moola & Faulkner, 2014). Using narrative in a form that leans towards the traditional qualitative thematic analysis, Bruce et al. (2014) explored stories of those with life-threatening illness, uncovering nuances of invisibility in life threatening illness but attending to content only. Gatti et al. (2018) used a different style of narrative analysis, a quantitative form that counted words and phrases, as well as exploring metaphors, a less common method in narrative. Wicks, Berger, and Camic (2019) used an extensive narrative methodology attending to structure, content, literary elements and social context in her study of chronic illness in adolescents, enabling the relationship between adolescent development, physical and emotional health to come into focus.

Carr, Loeser, and Morris (2005); Lipman (2011); Rajagopal (2011) are all researchers who have recommended narrative as a good approach to the study of pain, given that it is exploring experience of pain over time in people with CF. As they have a lifelong life limiting illness it seems especially relevant to explore their experience over their lives.

With experience being so subjective “pain remains elusive, difficult to grasp, and hard to address” and narrative offers a way to better understand pain, through the stories of those that are experiencing it (Carr et al., 2005, p. 3; Lipman, 2011). These stories about pain facilitate rich description and exploration of these experiences. Through listening to stories participants choose to tell, how they tell them, and the context of telling, a deep understanding of complicated experience can be gained. This provides insight into how pain should be treated and studied (Rajagopal, 2011). Narratives may also encourage others to act. In the case of the researcher-clinician, it is recognised that participants may view the interview occasion as an opportunity to be heard, to ensure that their unique perspectives about pain are heard, and in some cases may hope the researcher takes some kind of action. Through sharing their experiences, participants may also encourage others with similar experiences to also speak out, thereby creating movement or even social action.

4.3.6 Empowerment through narrative

With so little known about pain from the perspective of individuals with CF, and indications that pain is underreported in CF, empowering individuals to share their stories is critical. Narrative interviewing begins with the researcher empowering the participant to tell their own story in their own way (C. K. Riessman, 2008). This results in a shift of power from researcher to participant as the researcher gives up some degree of control, encouraging participants to provide extended accounts of experiences (Elliott, 2005; C. K. Riessman, 2008). By the researcher listening carefully and avoiding interruption,

participants are empowered to tell their own stories, whereas doing otherwise may inadvertently train the participant to discuss only topics the researcher finds relevant or only provide brief responses (C. K. Riessman, 2008). The researcher inevitably does exert control through their leading the research enquiry, even in narrative research, by deciding which aspects of a story are included, and how it is presented and interpreted. It has been noted, for example by Esin, Fathi, and Squire (2014), that it is important to be aware of the differential in power between researcher and participants, particularly in a constructionist approach to narrative. This attention to power helps the researcher and reader to see how the power relationship influences which stories are told and which ones may be suppressed. The interest in the power relationship is not only concerned with that between participant and researcher, but also with each party in the context of their other unique positions, cultural and social environments (Esin et al., 2014). There are power differentials that occur in all types of qualitative work and narrative is no exception. But it is in the way that the narrative researcher stays alert to the power in the relationship and its influence on co-construction that is important in honouring the story.

4.3.7 Narrative analysis

Narrative studies differ from other qualitative studies in how narrative accounts are treated analytically. In contrast to other forms of qualitative analysis which fragment data into themes, narrative research treats the extended narrations as units (C. K. Riessman, 2008). By keeping narratives relatively intact, researchers honour the individual's unique

voice and way of telling their story, allowing characteristics such as sequence and structure to remain visible (C. K. Riessman, 2008).

Just as there are different perspectives about what is accepted as narrative, researchers may align with particular approaches to analysis. Mishler (1995) published a useful framework for understanding these different approaches to narrative analysis. In his framework, Mishler (1995) described different approaches to narrative as focused on content (events and experiences described), structure (how the story is told), or performance (context of how a story is produced). In addition to these approaches, C. K. Riessman (2008) adds visual analysis, which examines how people communicate with images. Selection of a particular approach to narrative analysis can have a significant impact on presentation of data and findings, and their interpretation, demonstrated through re-presenting narratives by Poindexter (2002). Through detailed descriptions of using five different narrative transcription methods, Poindexter (2002) provided an insightful illustration of how different methods can influence interpretation and meaning. Transcripts of the same narrative in different styles focused on 1) content informed by Catherine Kohler Riessman (1993), 2) structural elements of the stories by Labov and Waletzky (1967), 3) linguistic approach attending to emphasis, pitch, pauses and presenting the story in the form of idea units, lines, and stanzas by Gee (1991), 4) an iteration combining Gee (1991) with Mishler (1995) emphasizing co-construction and 5) a poetic style informed by Gee (1991), but with more liberty taken to reduce to a poetic style. Each of these methods of transcription led to different nuances of analysis, for

example, Poindexter (2002) found that using Gee (1991) approach conveyed emotion in a way that others did not.

Most narrative researchers use thematic or content analysis or structural analysis (C. K. Riessman, 2008). Rather than a clear delineation between these approaches, C. K. Riessman (2008) describes the boundaries as blurred, with many researchers modifying the approach or combining different approaches together. This combined approach focusing on both content and structure is adopted in this study to examine the stories about pain in CF. A thematic or content analysis informed by C. K. Riessman (2008) and structural analysis based on discourse analysis by Gee (1991) was undertaken to closely examine how the stories are told. Looking closely at how the stories are told strengthens the findings as new insights may be found that were not visible when focusing on content alone (C. K. Riessman, 2008).

In the approach to structural narrative analysis and content informed by Gee (1991), interpretation begins with the units that make up speech. Units of speech include idea units, lines, stanzas, strophes, and parts, outlined in Table 4 (Gee, 1991). Stanzas, purported by Gee (1991) as a universal component of language, are made up of a group of lines about a single topic, relatively short and spoken at about the same pace. When a narrative interview is represented in lines and stanzas it may appear as a poetic-like form, communicating the essence of the story. Identification of these units of speech and subsequent application of this method requires close attention to both structure and mechanics of how a story is told, including pauses, utterances, emphasis, and intonation

or pitch (Gee, 1991). The close attention required is achieved through listening to the recorded interview multiple times during and following transcription. Poindexter (2002), whose comparison of five different narrative methods was described earlier in this section, found that Gee's method facilitated attention to pace and pattern of speech, word usage and emphasis on different words, resulting in identification of meaning that was not realized through other approaches.

Table 4

Structure of narrative as described by Gee (1991).

Unit	Description
Idea units	A single idea, single pitch glide, often noted by a preceding pause. The basic unit of speech.
Lines	A line is made up of several idea units, appears similar to a sentence, often identified through pitch and pauses.
Stanzas	Several lines about a single topic, often four lines in length, relatively short, spoken at same pace.
Strophes	Comprised of two related stanzas.
Parts	Larger units made up of stanzas and strophes which when combined make up the whole story or narrative.

4.4 Methods

This section outlines in detail the specifics of how this study was conducted.

4.4.1 Participant selection

This study was conducted at an adult CF centre in the US accredited by the Cystic Fibrosis Foundation. Approximately 260 people with CF receive their care at this centre, with ages ranging from 18-72. The centre has an active research program consisting primarily of pharmaceutical trials.

Purposive or criterion-based sampling was used for recruitment in this study. This sampling strategy involved selecting participants who could provide “insight and in-depth understanding” of the phenomenon to be studied (Patton & Patton, 2002, p. 230). Purposive sampling is commonly used in qualitative research to ensure that the participants represent the criteria of interest in the study and to attempt to include diverse representation (Creswell, 2013; Ritchie & Lewis, 2003). Purposive sampling also common in narrative studies, for example, (Pooler, Richman-Eisenstat, & Kalluri, 2018; Sheilds et al., 2015; Wicks et al., 2019).

The target population was adults with CF who endorsed experience with pain. Since previous studies indicated many with CF underreport pain, it was decided not to limit the study to individuals with a diagnosis of pain in the medical record. Instead, potential participants were eligible if they self-identified as having experience with pain in CF as described on the study information sheet.

Potential participants were identified through members of the CF multidisciplinary team (nurse coordinator, research coordinator, and physician) who served as gatekeepers. The gatekeepers provided a participant information sheet (Appendix H) to potential participants during scheduled CF clinic visits, and if they were interested, requested their preferred method of contact (telephone and/or email).

Participants were contacted to explain the study's purpose and what participation included, including potential risks and the ability to withdraw. When possible, they were given a copy of the informed consent (Appendix G) before the interview. A total of 23 people with CF were invited to participate, and 16 expressed interest. Reasons stated for not wishing to participate (when given) were reported by gatekeepers to be primarily related to the time required for the interview. Of the 16 participants who expressed interest, seven did not participate for reasons that included interview cancelled with no reason stated (1), interview cancelled due to hospital admission (1), participant died before interview (1), participant changed mind (1), interview cancelled due to wildfire (1), or lost to follow-up (2). A total of nine participants were interviewed.

The primary barrier to recruitment was reliance on gatekeepers to provide study information sheets, and notes placed on the clinic packets were helpful reminders. Diversity in gender, age, and disease severity was an aim, but notably in this size and type of in-depth qualitative study, a representative sample is not possible. Originally the aim was to recruit 10-20 participants, however, only 9 were recruited. Relatively small numbers of participants is comparable with other narrative studies as the focus in

narrative research is on rich and unique experiences communicated through individual participant stories (Braun & Clarke, 2013; C. K. Riessman, 2008). Because of the depth of data collection and depth of the method it is not unusual to find smaller sample sizes of 4-6 participants, for example Haydon, Browne, and van Der Riet (2018). The flexibility of narrative methodology to be applied to various sample sizes was illustrated by Elliott (2005) in her book about narrative in social research. Narrative approaches and their focus on individual participant stories (or cases) can be especially time intensive in comparison to other qualitative methods (Thomas et al., 2009). This focus on individual stories does not indicate disinterest in the sociological, rather it can illuminate the individual within society (Elliott, 2005).

4.4.2 Setting

Participants selected where to have the interview. Four preferred to meet in their homes and five chose to meet in an office building conference room at the university campus. Attention was paid to ensuring the participant's comfort, including physical environment and privacy in both locations. In seven interviews, only the participant and the researcher were present during the interview. In two interviews, family members were present or nearby at the participant's request.

4.4.3 Data collection

Primary data collection consisted of unstructured or in-depth interviews, conducted in a conversational manner (C. K. Riessman, 2008; Rubin & Rubin, 2012). Participants were

invited to explore their stories with pain and had some control in the direction and pace of the interview (Offredy & Vickers, 2010) .

An interview guide (see Appendix E) was developed with a list of topics, and examples of open-ended question styles, prompts and probes, but this guide was used infrequently and usually at the end of the interview to see if any topic areas had not already been covered. Topics included in the guide were past and current experiences with pain, quality of life, reporting pain to others, treatment, self-management, fears and concerns, and types of pain. Interview duration ranged between one hour and one hour 20 minutes. All interviews were audio recorded with participant permission. Emotional responses, silence, reflections and other details not easily identified in later reviews of the transcript were noted in the field journal.

I began each interview by explaining that I was interested in the participant's experience with pain in CF. I explained that the style of interview was unstructured and explained briefly what that meant. Initial questions and probes invited participants to "tell me about yourself" to learn background information that may not be related to pain and to begin to establish comfort in the interview.

Following this initial question, I asked the participant to tell the story about their experience with pain in CF, although this question did not immediately follow the initial prompt in all interviews. I tried to allow the stories to flow in as natural a style as possible, and minimised interruptions except to ask for clarification or further description. At the end of each interview, I reviewed the interview guide and asked additional questions if

needed. Participants were asked if there was anything they were surprised we did not talk about or would like to add to our discussion. I offered each participant the opportunity to contact me if they would like to add additional information to their interview, but none did so.

4.4.4 Ethical considerations

This study was approved through the Human Research Protections Program (HRPP) at University of California San Diego and through the Faculty of Health and Medicine Research Ethics Committee (FHMREC) at Lancaster University. Annual research updates and renewals were submitted as required. See Appendix F for complete ethics approval documentation.

It is worth noting in regards to ethical considerations my position as an insider, in that I conducted research within a population that I also work with in the role of nurse manager (Hammersley & Atkinson, 2007). Recognising the possibility that people might feel pressure to participate given my role in the CF program, I took several steps to minimise this possibility. First, I used gatekeepers for initial contact with potential participants. Next, when contacting potential participants who had expressed interest, I directly stated that the decision to participate or not would not affect clinical care in any way and emphasised that I was inviting them to participate if they *wanted* to. I further stated that anonymity and confidentiality would be maintained in accordance with the informed consent and emphasised that this included the CF team as well. When talking in person I avoided wearing a lab coat or business suit to decrease any perception of power that

might be reinforced through this attire commonly associated with clinicians or executive leaders.

4.5 Data analysis

A staged narrative analysis approach was undertaken to analyse the participant narratives. The first stage of analysis was to transcribe the interviews. This initial transcription of the interview was done verbatim with notation of pauses, sighs, emphasis placed on words or syllables, or any verbal utterances such as *um*, *ah*, *hmm* and similar. Also notated were any emotions such as laughing, crying or nervousness. Including these details in the transcript was important for providing vital information needed for Gee's (1991) method of structural and content analysis, about which further details are included below. I transcribed all of the interviews myself, and although it required a large amount of time, it enabled me to become very familiar and begin to immerse myself in the data.

During transcription, all identifiers were removed such as names of family members or CF care team members. Participants were assigned an alias from the list of most common baby names in the US in 1990, starting with the most common name (BabyCenter, n.d.). Following initial transcription, the audio recording was played again while reading the transcript, and any errors or omissions were corrected.

In the next stage of analysis, I began to interact with the data through close reading and notation of areas of interest in the transcripts. Using paper copies of transcripts, I annotated interesting sections of text by highlighting or making notes in the margin. I also

used the process of mind-mapping to engage further with the data and begin to identify relationships to Saunders (1965) theory of total pain. See Appendix I for an example of this initial stage of analysis on paper transcripts and for an example of mind-mapping.

I then used an approach to structural narrative analysis and content informed by Gee (1991), where interpretation begins with the units that make up speech. Transcribing that included these details and close repeated listening to the recording helped to identify these units of discourse within the transcript, specifically lines and stanzas. As Gee (1991) did in his research, each stanza was given a title using the participant's own words. Identification of these stanzas was a subjective process and it could be said that another researcher working with the same material might identify different stanzas. In this way, application of structural analysis is a highly interpretive process and demonstrates the co-construction of knowledge inherent in qualitative analysis. An example that clearly demonstrates this co-construction in a full text excerpt and the associated stanza is included in Appendix J.

4.6 Public involvement in research design

As recommended by Hammersley and Atkinson (2007), I aimed to empower participants and other individuals with CF through involvement in the research planning process, providing some degree of control in the interview process itself, and participation in providing feedback on transcripts and analysis. In CF and other pulmonary diseases, patient and public involvement has been encouraged in both research and clinical care (Chalmers et al., 2017; Cystic Fibrosis Foundation, n.d.-c; Cystic Fibrosis Trust, n.d.).

Through Community Voice, the Cystic Fibrosis Foundation actively seeks patient and family involvement, expressing that because each person's experience is different hearing from as many people as possible is critical to identifying needs and planning present and future initiatives (Cystic Fibrosis Foundation, n.d.-c). During the early planning phase, three individuals with CF volunteered to provide feedback on the planned study. These individuals reviewed the interview guide and provided feedback. Two of these individuals also participated in a brief pilot interview which helped to develop interview skills and comfort using the interview guide and audio recorder.

During the pilot interviews, a need to provide tissues, wastebasket and water was identified (due to coughing up sputum). Also, during pilot interviews, there was an opportunity to practice listening more, talking less, and become aware of the impulse to want to speak up to solve problems. Conducting pilot interviews aided in the transition from the role of clinician to that of clinician-researcher for this study.

The unstructured interview style provided participants some control over the interview process, particularly which stories received more attention than others. Each participant was offered an opportunity to review their interview transcripts but only one did so. This individual commented that they were surprised by the depth of pain they were holding in and expressed gratitude for the opportunity to talk about it. Similarly, participants were provided an opportunity to view the interview segments that were formatted into stanzas. Five participants elected to review stanzas, and none requested changes. Rather, all five expressed satisfaction with the transformation to stanzas and felt representations

were accurate. Several commented on the “power” or “essence” present in this poetic form, and one participant tearfully asked if he could have a copy to share with his wife as “this is my heart in raw form”.

4.7 Conclusion

This chapter has outlined a research paradigm concerned with how individuals learn and develop meaning through social activities or social constructivism. In this paradigm, there are multiple realities subject to individual interpretation (Guba & Lincoln, 1994). Adopting social constructivism demonstrates value for varied perspectives in striving for a deeper understanding of unique pain experiences in CF and narrative research methodology is ideally situated for this study. Findings, analysed thematically as informed by C. K. Riessman (2008) and structurally as informed by Gee (1991), are presented in Chapter Five.

Chapter 5: Findings

This chapter presents the study findings. First, a summary of participant socio-demographics, data collection and analysis are presented. Next, the three key narratives and associated sub-stories identified during analysis are described. The narrative of emerging awareness of having CF is presented through the sub-stories of diagnosis, disease progression and limited life expectancy. The social legitimacy narrative is presented through the sub-stories of pushing through and independence. Finally, the narrative of invisibility is presented through the sub-stories of feeling misunderstood, choosing to be invisible, and becoming visible.

5.1 Sociodemographics, data collection and analysis

Ten individuals agreed to participate in the study, of whom nine were interviewed; one died before the interview could occur. Participants were aged 26–44 years, and all Caucasian but one. Gender distribution was six females and three males. The ethnic distribution of participants was consistent with the reported ethnic distribution nationally (Cystic Fibrosis Foundation, 2018). One third of participants had advanced lung disease with an FEV1 of $\leq 40\%$. Participant demographics including age, gender, ethnicity, lung function, marital status, and lung transplant status are listed in Table 5.

Table 5

Demographics

Age	Median 29.5 (26–44)
Gender	Female: 6 Male: 3
Ethnicity	Caucasian: 8 Latino: 1 Other: None
Lung function	FEV1 ≤40%: 3 FEV1 >40%: 6
Marital status	Married: 5 In a relationship: 1 Divorced and in relationship: 1 Single: 2
Lung transplant status	Received transplant: 2 Listed or undergoing evaluation: 1 Not applicable: 6

Four interviews were conducted in participants’ homes and five in a university office conference room. Interviews, lasting between 55 minutes and 75 minutes on average, were conducted between October 2016 and June 2017.

Interviews started with an invitation for participants to share a little about themselves, usually phrased: “How would you describe yourself and your CF to someone who doesn’t know you, a kind of [participant name] 101?” This prompt was given to help participants feel comfortable as the interview began. Initially an attempt to ease into the interview, these initial stories became a significant element of the findings.

After sharing about themselves, participants were asked to describe their experiences with pain. Some participants immediately asked if emotional pain was included, while

others simply assumed it was. All participants shared different types of pain, and most labelled their experiences as physical or emotional pain. Although many stories included elements of social and spiritual pain as well, these were not overtly identified by category, and often labelled as emotional pain, if labelled at all. When they described their experience with pain in CF, participants shared a narrative journey of their lives, incorporating their experiences with pain into these stories.

A narrative approach to analysis, conducted according to the description of methods in Chapter Five facilitated identification of overarching narratives and sub-stories. Inspired by Gee (1991), analysis focused on both content and structure. Through analysis, three key narratives were identified: emerging awareness of having CF, social legitimacy, and invisibility. They are presented here in the form of stanzas. These narratives, each composed of several storylines, expressed the essence of participants' all-encompassing experience with pain.

5.2 Emerging awareness of having cystic fibrosis

Throughout their interviews, participants shared how CF has increasingly affected their lives. As CF progressively worsened, participants experienced an emerging awareness of what having CF means. As they became sicker, participants were more aware of how CF was affecting them and expressed different types of pain. Pain was a constant presence and affected every sphere of their lives.

While increasing awareness occurred gradually, some memories stood out more than others, and it is these recollections that facilitated the emerging awareness narrative. For discussion purposes, the emerging awareness narrative is divided into three sub-stories: diagnosis and early years, disease progression, and limited life expectancy; however, these actually comprise more of a single, narrative journey through participants' lives. Participants shared this narrative as a chronological retelling of their experiences with pain from diagnosis and early years to the present, a style consistent with temporal structuring of illness narratives and other forms of life stories (C. K. Riessman, 2008).

5.2.1 Diagnosis and early years living with cystic fibrosis

All participants began with stories about diagnosis. In the first four interviews, these were elicited by an initial prompt requesting they talk a little about themselves and their CF, then adding an inquiry about diagnosis and disease course. This part of the initial prompt was later dropped to decrease interviewer influence on what they chose to share, but participants began with diagnosis stories regardless of which prompt was given.

For most participants, these stories were brief and factual in nature, coming from those diagnosed as infants or young children. For these participants, CF had always been a part of their lives, and information about diagnosis was relayed through parents rather than personally experienced. When diagnosed in infancy or early childhood they did not have the intellectual capacity or maturity at the time to understand the implications of diagnosis. However, participants diagnosed later (Amanda and Matthew) relayed more details about the impact of diagnosis.

Box 1

Stanza 2: They did a sweat test on me

5. I would cough in the mornings
6. I would cough so much I would vomit right when I woke up
7. and then throughout the day I'd be a little bit better
8. so, they did a sweat test on me

Stanza 3: I was in denial

9. I had never even heard of the disease before, so I was in denial
10. because I thought for years
11. I thought that they had did something wrong with the test
12. and that I had some other something.

Stanza 4: I thought eventually they would say we made a mistake

13. I thought eventually they would say we made a mistake
14. here's an antibiotic you're going to be better
15. I'm sorry to put you through this
16. that didn't happen

(Amanda)

Box 2

Stanza 2: Before I got diagnosed it didn't really affect me that much

1. Before I got diagnosed, it didn't really affect me that much
2. I was doing everything a normal 16-year-old was doing
3. I was playing soccer, I was playing sports at school,
4. like a normal 16-year-old

Stanza 3: But after I got diagnosed it did affect me emotionally

5. But after I got diagnosed it did affect me emotionally
6. knowing that I was sick
7. and that there wasn't a cure for it
8. it affected me a little bit

9. knowing that there wasn't a cure for it.

(Matthew)

These participants who were older at diagnosis did not describe themselves as “sick”, despite having symptoms. Instead, they described themselves as normal, with diagnosis presenting a tremendous disruption as they wondered what it meant and if perhaps it was a mistake. Figuring prominently in their stories was how diagnosis affected them emotionally. Their stories expressed pain around diagnosis that was not present in stories of participants who always knew they had CF. Although they mentioned physical symptoms, the dominant type of pain in these stories was emotional. Emotional pain was seen through denial of the diagnosis and in recognition that there was no cure.

For every participant, and throughout their interviews, laughter and sometimes whispering accompanied expression of their most painful experiences. Diagnosis stories seemed well rehearsed as if told many times before, especially for those diagnosed as infants. In contrast, the pain expressed in these stories seemed raw, less rehearsed than the factual information. As participants continued their narratives of emerging awareness, they moved from stories centred on diagnosis and early years to stories about getting sicker.

5.2.2 Disease progression: Harder to stay healthy

Similar to the overarching narrative of emerging awareness, disease progression stories were told chronologically, beginning with describing how it became harder to stay

healthy. Participants recalled different milestones as indicators of disease progression, such as Brittany in Box 3, who viewed her first hospitalisation as a turning point.

Box 3

Stanza 1: It just got harder and harder for me to stay healthy

1. When I turned 17, I had my first hospitalisation, my first IV, my PICC line
2. and after that it just got harder and harder for me to stay healthy
3. so ever since then the progression is just
4. it just gets worse

(Brittany)

As these stories progressed, realisation of how CF was affecting participants became increasingly evident, and they worked harder to maintain the same level of health. Every participant told these stories of working harder, whether seen through increased time and intensity required daily to clear their lungs, needing more medications, or more frequent and longer hospital admissions. As they worked harder to stay healthy, awareness of what having CF means, and how it was affecting their lives, became more apparent. Every participant referred to the treatment burden caused by hours of therapy daily, seen through Christopher in Box 4 when asked about his experience with pain.

Box 4

Stanza 6: It's very slow repetitive

1. I would consider almost everything I've experienced with cystic fibrosis
2. It's very, slow, repetitive
3. kind of like how a drop of water on your forehead isn't uncomfortable
4. until the 100,000th drop that lands on your forehead.

Stanza 8: It's just the frustration of having the same, slightly discomfort feeling

8. So, it's not something where you can take it in five seconds
9. and feel it and go yeah that's painful – you know
10. it's just the frustration of having the same, slightly discomfort feeling
11. for such a long period of time.

Stanza 9: Just the constant beat down that you kind of feel

12. The most prevalent thing just being just the consistency of it
13. the consistency of the discomfort
14. the consistency of the time management and the lifestyle
15. just the constant beat down that you kind of feel for having all those things at one time.

Stanza 10: It's the most unfair thing in the world

16. and then you feel like you're doing everything right
17. and then you start to get sick
18. and you're like what in the world?
19. you just feel like it's the most unfair thing in the world

(Christopher)

Although all participants described treatment burden, notable in Christopher's interview is the way that his visual style of speech and use of metaphors illuminates the experience. Repetition of "consistency" provides a rhythmic depiction to reinforce the "constant beatdown". Participants viewed increasing treatment burden as an important component of their experience with pain. Even the language Christopher chose ("constant beatdown") was reminiscent of pain.

While at first participants could avoid getting sicker by increasing therapies, eventually this no longer worked. As in their first hospitalisations, participants recalled not responding as well to treatment as a milestone or marker of disease progression.

Christopher's story in Box 5 represents this turning point when participants began not responding as well as before.

Box 5

Stanza 3: I got on the IV and just a couple days later I started to feel this lift

10. What I remember from last time being on IVs
11. not this time – the time before which I believe was 6 years ago
12. I remember I got on the IV and just a couple days later I started to feel this lift
13. and about 4 days later I felt like Superman
14. I felt like I could do anything.

Stanza 4: I feel like Superman – is this what normal people feel like?

15. I remember specifically I was on a run with my wife on a trail
16. and we're running and she's having trouble keeping up with me
17. which is not usually the case
18. cause I'm usually either with her or a little bit behind her
19. and I remember running and turning around going I feel like Superman
20. is this what normal people feel like – this is crazy!

Stanza 5: But this time... it was a little more slow going

21. But this time, which I just recently got off IVs
22. it was a little more slow going than I thought it was going to be
23. I think it was about 6 or 7 days until I started feeling good
24. and then at the peak I didn't feel quite as good as I did last time.

(Christopher)

Like Christopher, most participants used metaphors associated with power, such as Superman, battle and war; they compared CF to a lengthy battle, one that is both frustrating and exhausting. This frustration and sense of overwhelm is echoed in all participant stories. In these stories about increasing difficulty of staying healthy,

participants still responded to treatments, albeit not as well as before. Within these stories is an initial glimpse of what having CF means, but participants became even more aware as they grew even sicker. Described in these stories, pain had a gradual, insidious presence.

5.2.3 Disease progression: Getting sicker

As CF progressed, emerging awareness figured prominently in participants' stories. Participants described getting sicker and sicker, regardless of their efforts to prevent or treat their symptoms. They spent more time in the hospital and pain took a more pronounced role in their narratives. Participants such as Samantha, who had previously been in denial about CF, described a breaking down of denial as they became increasingly aware of the impact CF was having on their lives.

Box 6

Stanza 1: Emotionally that was rough

1. So, there was a really hard learning curve for me when I became an adult
2. I mean if 18 can be considered an adult
3. So, I mean that was – I mean emotionally that was rough – or painful
4. really learning the depth of what the illness did.

(Samantha)

Notably, Samantha's depiction of emerging awareness was told in the past tense as she was clearly recalling a past event. Other participants changed to the present tense in these stories. For these participants, they were still in the midst of these experiences, as

when Brittany referred to pain as a reminder that she is sick. Participants viewed both the treatment regimen and their physical symptoms as reminders of CF.

Box 7

Stanza 1: Like a chink in the armour

1. It's like a milestone (pain) – isn't that great
2. A milestone in my cystic fibrosis progression
3. like a chink in the armour
4. it's a tangible reminder of everything else

Stanza 2: You know coughing is exhausting

5. and coughing – just the coughing
6. you know coughing is exhausting
7. you wake up and you feel like you have to cough
8. and automatically you're reminded

Stanza 3: It's a reminder that you like you have this thing

9. when you are in class and you need to cough
10. and you already coughed five times
11. it's a reminder that you like
12. you have this thing.

(Brittany)

As her story unfolds, Brittany repeatedly returns to physical reminders of illness. Referencing the progressive nature of CF, pain is described as a milestone, a symbol that she is getting worse.

For most participants (seven), as they delved deeper into this emerging awareness narrative, pain became more apparent, notably expressions of emotional and spiritual pain. Pain was not portrayed as separate and distinct, but was woven throughout, and in

a way inseparable from the greater experience of CF. Participants struggled to talk about pain as a distinct entity, and yet pain was present throughout the entire interview.

This depiction of pain as enmeshed in the life experience is seen in Brittany's continued narrative of emerging awareness as she moved out of her parents' house, then stopped going to college and moved back in as her health declined.

Box 8

Stanza 2: So now I live at home with my mom and dad

5. So now I live at home with my mom and dad.
6. I moved out a year and a half ago
7. which was a big thing for me
8. because when I was 18, I always thought I would be out

Stanza 3: This has been very hard for me the past few years

9. I always wanted to be very independent
10. I graduated school – I got my diploma early
11. I wanted to work, and I just wanted to – you know
12. so, this has been very hard for me the past few years

Stanza 4: This is not going the way that I thought it would go

13. Because I feel like my whole life has just like been completely different
14. As to what I thought I would be – obviously
15. it's just like – whoa – wait a minute –
16. this is not going the way that I thought it would go –

Stanza 5: I just never imagined it this way

17. I think about it all the time – I can't believe
18. that this is how things have gone for me
19. cause I just never imagined it this way.

Stanza 6: I was like thinking about emotional pain

20. I was like thinking about emotional pain –
21. and I don't know if I have a separate issue with
22. I probably have a separate issue with dealing with everything but

Stanza 7: All this stuff you get used to – your body copes

23. Back to where I was talking about in the beginning
24. how I feel like my life is so different from how I ever expected it to be
25. has probably been like –
26. all this stuff you get used to – your body copes

Stanza 8: These types of things are short

27. The pleurisy is one thing –
28. if I had to deal with that all the time then it starts to really –
29. that would be like – interfering – you know –
30. these types of things are short –

Stanza 9: What I feel like CF has limited me to do has been the hardest thing

31. I don't feel like they interfere with my life completely – yet –
32. my physical pain –
33. But dealing with what I feel like CF has limited me to do
34. has been the hardest thing and the most painful for me.

(Brittany)

As she talks, Brittany relates the loss of developmental milestones, the loss of independence, and the loss of the life she was anticipating or hoping for.

As participants became sicker, stories around developmental milestones were part of every narrative and each included pain. As they became sicker, their relationships to pain appeared to change. With only one exception, participants were more affected by emotional pain than physical pain in these earlier recollections of disease progression.

That is not to say they were unaffected by physical pain, but physical pain did not appear to cause as much distress as emotional pain.

In Box 8, Brittany is used to physical pain, but in her statement “I don’t feel like they interfere with my life completely yet” she anticipates increased physical pain in the future. She views physical pain as transient, but emotional pain as more permanent. In line 15, her use of the word “whoa” reflects her desire to slow the progression of her CF, a sentiment echoed by other participants. However, instead of slowing down, most participants told stories of getting much worse.

5.2.4 Disease progression: Advanced illness

Stories told about advanced stages of CF were markedly different than earlier stories. Not only were participants needing more therapy, but it was no longer working. For these four participants, CF was now so severe that even weeks in the hospital did not help. These stories were marked by resignation that nothing could be done and a sense of hopelessness. While other participants with advanced disease were still living it, Amanda recalled the months prior to having received a lung transplant.

Box 9

Stanza 4: The next morning when you wake up everything’s back in your lungs again

1. The treatments that you would do from sunup to sundown
3. and then the next morning when you wake up –
4. everything’s back in your lungs again –
5. to start over from zero –
6. it was like that – no matter what –

(Amanda)

Even when Amanda spent three or four weeks in the hospital, she still did not get better.

Box 10

Stanza 1: Getting discharged feeling the same exact way

1. As I've gotten older it would really suck
2. when you'd go in for a tune up and you'd be prepared to
3. okay I'm gonna fight whatever's going on
4. and I'm gonna feel so good when I get out of the hospital
5. and then getting discharged and feeling the same exact way.

(Amanda)

Amanda tried to remain optimistic, but even this was not enough as she was discharged three or four weeks later feeling the same as at admission. This disappointment of expecting one outcome and experiencing another was reminiscent of earlier stories, when participants became sick despite adhering to therapies. This was yet another bitter disappointment for them.

Pain was approaching a crescendo in these stories of advanced illness. Pain of all types was expressed, but physical pain was most present in advanced illness. Participants didn't just talk about physical pain more often but described it as more severe and seeming to permeate all aspects of their lives.

Participants rarely talked about pain in isolation of their overall experience with CF. As seen in Box 11 as Amanda described late stage CF, participants linked physical pain with

emotional pain. Where physical pain was mentioned, other types of pain were described or implied within close proximity. Comparison of the hospital to prison might also describe the lack of escape from CF.

Box 11

Stanza 2: Refracturing old ribs just from the coughing

5. With all the rib fractures before transplant
6. I was getting maybe 3 – possibly 4 a year
7. and refracturing old ribs – from just the coughing
8. that was really painful too

Stanza 3: You have to cough to get the stuff up

9. You have to cough to get the stuff up
10. and when you're breathing
11. it would hurt to take a deep breath.

Stanza 4: In the hospital the pain is more intense

12. And it seems like a lot of times
13. like when you're in the hospital
14. it seems like the pain is more intense
15. because you're stuck
16. it's like prison – and you can't go.

(Amanda)

Participants referred to “emotional pain” as the most painful, as seen in Amanda’s story below. When not overtly labelling pain as emotional, participants described it as one of the most painful parts of their experience. Emotional pain included fear, anxiety, uncertainty, and multiple losses such as health, relationships, independence and death.

Box 12

Stanza 1: More emotional pain than anything

1. I think with CF
2. You have a lot of
3. Like more emotional pain than anything
4. Cause I've had a lot of emotional pain because of this disease.

(Amanda)

Participants expressed resignation that nothing could be done, a sense of helplessness that was not as prominent in the earlier narratives. This is not to say only participants with advanced illness expressed resignation or helplessness; others expressed similar feelings, but the degree of resignation and helplessness in advanced illness was greater.

Similar to most others, Ashley appeared exhausted, with frequent sighs and sagging shoulders, as though the weight of the world were physically upon her as she shared her resignation that nothing could be done.

Box 13

Stanza 1: There's not anything they can do

1. I pretty much literally laid in bed and suffered
2. I mean – there's not really much you can do
3. there's not anything they can do

Stanza 2: With CF you just kinda roll with the punches

4. a lot of times with CF you just kinda roll with the punches
5. and take it as it comes you know
6. I mean you just gotta look at it as well that's my life.

(Ashley)

In telling these stories of progression, most participants were in the midst of the experience. That is, they were not only recalling events from the past and retelling them but were still experiencing them. These stories were less rehearsed, and participants were more emotional when sharing them. Pain was present, but not necessarily neatly labelled, and they began to recognise their limited life expectancy. These stories of limited life expectancy were most concentrated in stories about progression of CF.

5.2.5 Limited life expectancy

As the narratives of emerging awareness were shared chronologically, most evolved towards awareness of limited life expectancy or death. This was acutely emphasised when one person died before their interview, and another died after the interview, before thesis submission.

Life expectancy was the only topic related to lung function, in that the two participants with the highest lung function (Michael and Christopher) were the only ones who did not mention limited life expectancy. The only exception was Amanda, who had high lung function, but had previously been close to death before transplant.

Some participants only spoke of limited life expectancy in regard to others, while others talked about themselves. Several knew someone with CF who had died. Being a genetic disease, some had experienced a sibling's death from CF; others spoke of friends from the hospital or clinic who had died.

Ashley and Amanda were older than other participants and talked about being the only ones left of their original CF friend groups. As children, they were not discouraged from being around others with CF as happens today due to infection concerns, and they seemed to have more friends with CF than younger participants did. These social relationships were clearly meaningful and their loss painful. As seen in others, when describing their most painful moments, both laughed when quietly saying they were the only ones left. Amanda's story is a good example of this social pain present in all participants' stories.

Box 15

Stanza 3: When I first met people it was being in the hospital

10. You know – I didn't really know a lot of CF-ers in my early years
11. when I first met people it was being in the hospital
12. I remember there was like eight of us together one time
13. and so we sat out in the hall – and it was just like a big group of us

Stanza 4: Most of them have died

14. Well – it kind of sucks
15. but most of them have died
16. I'm trying to think of – well – one is still around – he just got married
17. I don't really talk to him all that much.

Stanza 5: I haven't really gotten close to anybody

18. but it's like everyone has died and you know honestly
19. I haven't really gotten close to anybody
20. I don't know any new CF-ers here.

(Amanda)

As Amanda concludes this part of her story, she admits she has not “really gotten close to anybody”, appearing to rationalise her reluctance to get close to others now.

While some participants limited their discussion of death to stories about friends with CF who had died, others reflected on their own mortality. In Brittany’s interview there was no gradual build up or subtle mention; rather, the listener and reader are thrust into the chaos of her story from the first moments. While this story was also told in a chronological form, it differed in that she introduced awareness of her limited life expectancy earlier in her narrative.

Box 16

Stanza 1: I remember having this like realisation moment when I was like 10

1. I remember having this like – realisation moment –
2. you know what I mean –
3. when I was like – maybe I was like 10 or something.
4. I don’t know.

Stanza 2: I asked them “could I die from this”?

5. I think I asked –
6. oh actually my parents told me this recently
7. and I completely forgot about this –
8. I had no idea this happened –
9. but I guess I asked them –
10. could I die from this –

Stanza 3: When do you tell your kid?

11. I don’t think they ever told me – like – straight out –
12. because it’s kind of like
13. when do you tell your kid –
14. especially because I wasn’t getting medication –
15. I was taking antibiotics – like oral antibiotics –
16. but I had never gotten a PICC line until I was 17.

Stanza 4: What does this mean?

- 17. So – I wasn't ever hospitalised when I was really young –
- 18. so I didn't really – I don't know –
- 19. I was just kinda like –
- 20. okay I have an illness and I take medication for it –
- 21. but like what does this mean?

Stanza 5: Does this mean that it could kill me eventually?

- 22. Does this mean that it's gonna get harder?
- 23. Does this mean that it could kill me eventually?
- 24. and I guess I asked them one day and they had to tell me –
- 25. this will kill you one day.

Stanza 6: I do remember when it just kind of clicked

- 26. They remember it perfectly –
- 27. I couldn't remember it –
- 28. I don't know why –
- 29. I was like "Oh I don't remember asking you that"
- 30. but I do remember when it just kind of clicked –
- 31. like – this feeling afterwards.

(Brittany)

Brittany, at about ten years old, was already searching for meaning, something she seems to still be doing years later. Throughout her interview, she seemed to be inquiring as much about existential meaning as she was about the potential of dying itself. Her questioning "but like what does this mean" was as relevant on the day of the interview as it was for her ten-year-old self. Throughout her interview she described a search for answers about the future. This existential pain implied by others was explicit in Brittany's story.

Limited life expectancy was more prominent in the stories of people who had more advanced disease and no participant demonstrated this throughout their story more than Matthew.

Matthew confronted pain as his CF worsened and he became increasingly aware of his limited life expectancy. This is demonstrated in Box 17 where, faced with worsening CF, he and his wife rushed to get married and have children. Their rush was evident as Matthew recalled this time, noticeably out of breath as he talked faster and faster, his pace of speech matching his description.

Box 17

Stanza 1: I try not to think about CF

1. We tried to like – just live a normal life
2. I try not to think about CF
3. Sometimes the disease
4. because when we had plans – like – we had plans together.

Stanza 2: Then I started getting the disease more

5. like I'm gonna finish my studies
6. and then after that we would get married and then have a kid
7. and then I started getting the disease more.

Stanza 3: We wanted to rush everything

8. We were more aggressive
9. Yeah we were more aggressive,
10. so we wanted to rush everything.

Stanza 4: We didn't know how much time we had

11. get married now – we want to have a daughter now
12. We wanted to do everything at that time
13. because we didn't know how much time we had.

(Matthew)

Other participants also expressed this desire to live life fully now, recognising the future as uncertain. This is evident in Jessica's narrative below.

Box 18

Stanza 1: Let's do what we can while we can

1. This past summer – my daughter was out of school
2. and it was let's fill up –
3. let's do what we can –
4. while we can kind of thing –

Stanza 2: We don't know how I'm going to feel

5. like let's go to Legoland – let's go to the Waterpark –
6. let's you know – go play with our friends – let's go to the beach –
7. let's do this stuff while I'm capable of doing it
8. because we don't know how I'm going to feel
9. when I wake up in the morning – and you know –

(Jessica)

As Jessica talked, her voice was excited, higher pitched, and she spoke quickly, at an almost frenzied pace, indicative of trying to fit it all in, to live fully while she can. As she continued, the pace slowed, and she took a deep breath as she appeared to reflect on the future.

Although all were aware of their limited life expectancy, only Matthew expressed certainty around what this meant. Having already received a lung transplant and been

informed he was unable to receive another one, he was oxygen-dependent and experiencing rejection of his transplanted lungs. Matthew was sure that he did not have long to live, but even this proved uncertain as he was still living almost two years after his interview.

These stories of diagnosis, disease progression and limited life expectancy comprised the emerging awareness narrative. In a chronological style, participants shared their experiences of pain in CF, covering the continuum from diagnosis to advanced illness and awareness of their limited life expectancy. As participants told these stories along their trajectory of CF, pain of all sorts became increasingly evident. Diagnosis and early disease progression stories contained mostly emotional pain. As CF became worse, physical and social pain became more evident, and existential or spiritual pain started to surface. As participants shared these experiences of pain in CF, another narrative that emerged was social legitimacy.

5.3 Social legitimacy

As participants told stories about becoming sicker and increasingly aware of the threat of CF, another prominent narrative arose. In this narrative, participants told of their struggle to maintain social legitimacy.

Social legitimacy is defined as a “generalised perception or assumption that the action of an entity is desirable, proper or appropriate within some socially constructed system of norms, values, beliefs and definitions” (Suchman, 1995, p. 574). Tyler (2006, p. 378)

further explained that “people who internalise social norms and values become self-regulating, taking on the obligations and responsibilities associated with those norms and values as aspects of their own motivation”.

These cultural beliefs then inform the expectations people use to judge themselves and others as legitimate or not. In this quest to maintain social legitimacy, participants described how they “pushed through” the pain in CF. They strived to meet social expectations such as independence, developmental milestones and productivity, and in doing so sometimes ignored their symptoms, trying to avoid the hospital at almost all costs.

Unlike emerging awareness, social legitimacy was not told in a chronological way. Rather than having a beginning (diagnosis and early years), middle (disease progression) and end (recognition of CF as life-limiting), these stories were less organised. While the emerging awareness narrative was dominated by expressions of physical and emotional pain, the pain seen in social legitimacy touched more on social and spiritual pain.

5.3.1 Pushing through

The concept of pushing through was identified in every participant’s story, and although it seems simple it is actually a complex experience. In its simple interpretation, pushing through related to such actions as completing a planned activity, such as when Jessica continued riding her bike despite severe joint pain in Box 19. Yet, beyond this simple understanding, participant’s stories described a more complex scenario of pushing

through in response to illness. Pushing through in this case occurred as participants strived to meet expectations of themselves and society. Stories of pushing through as a response to illness comprised the bulk of the social legitimacy narrative.

Pushing through happened in different contexts, but the stories were all a familiar style. In them, participants faced an obstacle of some sort such as pain and pushed through to meet a certain goal. In Box 19, Jessica pushes through when she develops joint pain on a day that she and her husband had planned to go to the park with their daughter. In this scenario, Jessica confronts physical pain as she pushes through to go on the bike ride despite the pain.

Box 19

Stanza 1: I woke up that morning and was hurting

1. We planned on taking our bikes to (the) bay
2. I woke up that morning and was hurting and I said
3. I'm just gonna get through it
4. We are going to go and have fun
5. It's gonna be a good day...

Stanza 2: I was just pushing through it

6. We got down there
7. My hands were hurting so bad
8. But we started riding
9. I have hand brakes
10. So I was just pushing through it...

Stanza 3: My hands started hurting so bad

11. My hands started hurting so bad
12. I couldn't stop my bike
13. Cause I had hand brakes
14. So I had to keep stopping with my feet.

Stanza 4: It was just too much

15. It was just the worst
16. That I think that I've ever felt
17. and I think it's because I tried to push through it
18. It was just too much.

Stanza 5: I still couldn't move the next day

19. So I came home and I laid down in bed
20. My husband had to take over
21. and then I called my grandma
22. Because I still couldn't move the next day.

(Jessica)

After pushing through, Jessica ends up in a very unwell state, unable to complete the ride and bedridden the following day. This scenario of feeling worse after pushing through was seen when other participants described similar instances of being bedridden after pushing through.

Participants also pushed through in response to emotional pain. Returning to Jessica, she talks about pushing through following the death of her brother (from CF) and the more recent death of her mother (from cancer).

Box 20

Stanza 1: You just have to push through

1. Losing my brother 10 years ago –
2. losing my mom this past year –
3. you just have to push through –
4. you just have to –

Stanza 2: They help me get through all this stuff

5. I just – like I said I have a really good support system
6. and I have a wonderful husband
7. and a really good kid and they just –
8. they help me get through all this stuff –

Stanza 3: CF has been the least of my concerns

9. so – I feel like – yeah – mostly – I mean –
10. I feel like in these past few years
11. my CF has been the least of my concerns

Stanza 4: Usually CF is my number one concern

12. Which is weird for me –
13. because usually my CF is my number one concern –
14. but I feel like these past few years it's been my mom's health
15. and my arthritis and just pushing through all of that stuff
16. that has been my bigger concern.

(Jessica)

Interestingly, although arthritis is a known complication of CF, Jessica regards pain from arthritis as unrelated to CF, as did Michael. Throughout her narrative she sees her roles as mother, wife and daughter as high priorities. Striving to be successful in their multiple roles, participants ignored CF at times, prioritising other activities over treatment.

Some participants were reluctant to ask for help when sick and described scenarios when they pushed through symptoms and delayed admission to the hospital. Brittany illustrates this when she needs a ride to the hospital but is reluctant to inconvenience anyone.

Box 21

Stanza 3: I waited until the next morning

10. I was texting my um cousin who's a nurse
11. and she was like I have a break
12. and if you need me to come get you I will –
13. so I waited until the next morning

Stanza 4: I've actually experienced pain with CF my whole life

14. Then my dad brought me in and that was my most – like
15. That's what I thought of when we were talking about this
16. But then I was thinking about it a little more
17. and I was like I think
18. I've actually experienced pain with CF my whole life
19. but I think I've become very kind of like numb to it.

(Brittany)

As Brittany reflects on her reluctance to ask for help, she realises she has experienced pain with CF her whole life. In her words, though, she has “become very kind of like numb to it”. It is not clear if Brittany is describing her physical pain during this particular episode or if she is talking about a broader experience of pain. However, stepping back and reflecting on Brittany's story in its entirety, it seems she is experiencing all-encompassing pain. Through the course of telling her story, Brittany appears to develop a greater awareness of her pain.

Amanda offers further insight into the lengths people with CF will go to delay admission, describing a friend who waited a week with severe pain before going to the hospital.

Box 22

Stanza 1: Finally, when she went to the hospital she had a collapsed lung

1. I had a friend and she didn't go here – she passed in 06 –

2. she had severe pain for a week and then finally, when she went to the hospital
3. she had had a collapsed lung for a week

Stanza 2: Some people just keep pushing it and pushing it

4. some people just keep pushing it and pushing it
5. thinking I'll feel better – I'll feel better –
6. but I think it's because they don't want to go back in –
7. they'll do anything they can to stay out –

(Amanda)

In pushing through to delay admission, participants were responding to their illness in a way that felt legitimate, displaying what they perceived as strength. In most of these stories, participants were pushing through specific scenarios or symptoms, but Ashley sums it up in a more general sense, explaining that everyone with CF pushes on.

Box 23

Stanza 1: We've been doing it all our lives it's all we know

1. I think everyone with CF has to push on
2. otherwise what is the point
3. there would be nothing to live for
4. We've been doing it all our lives
5. it's all we know.

(Ashley)

These stories of pushing through provide an initial glimpse of social legitimacy. Comments about pushing through seemed to relate to cultural expectations of an illness response,

in which the most appropriate response to illness is to “push through”. A related concept that arose in the narratives of social legitimacy was that of independence.

5.3.2 Productivity and independence

Stories of productivity, defined as participating in activities of value to the self or society, were included in the narrative of social legitimacy for all participants (Hay, 2010). This was not surprising given the moral value placed on productivity in the US (Charmaz, 1991). This value of productivity is pervasive throughout the US, including in those who are ill. Some stories about productivity seem closely related to pushing through but are different in an important way. The stories of pushing through are focused on the act itself, while the stories of productivity are focused on the outcome, with an emphasis on independence and perceived cultural expectations.

While all participants valued being productive, exactly what this meant to each participant varied. For Amanda, this meant being independent in caring for herself, but her desire to be independent extended beyond this, as she also desired financial independence. The port she refers to in the opening line is an implantable vascular access device used to deliver intravenous (IV) medications for those who need frequent infusions, such as those with cancer or other illnesses including CF.

Box 24

Stanza 1: I just wanted to be independent

1. I used to access my own ports
2. because I didn't want to have to rely on going to the hospital

3. what if I'm too sick to drive and I need – I just wanted to be independent
4. not financially – I can never do that.

Stanza 2: I'd always do it myself

5. but – when it came to all my meds and doing everything I needed to do
6. even with my IVs – I never got any help from anybody – ever
7. I did everything – I'd wake up no matter how late I had to infuse and how early
8. I'd always do it myself.

Stanza 3: I couldn't control the disease so I could control that

9. Like a sense of power – like something I could control
10. because I couldn't control the disease – so I could control that
11. and I wanted to learn how to do everything I could to help myself
12. I just didn't want to rely on anybody – I couldn't.

Stanza 4: I'm still on the system and I hate it

13. I don't like the fact that this disease is really expensive – It's very expensive
14. and if you know your future spouse has good insurance that's fabulous
15. but if he doesn't or you can't get insured you have to stay on the system
16. Which I'm still on the system and I hate it – even though they pay for everything.

Stanza 5: It feels like I don't contribute to society

17. that's the thing is that I don't make much money every month
18. but the medical is fantastic – it's fantastic – but it's humiliating
19. I'm on Medicare and MediCal – well isn't Medicare for old people?
20. It feels like I don't contribute to society.

(Amanda)

Participants such as Amanda who did not view themselves as independent and productive members of society expressed anxiety in these social legitimacy narratives. Social pain was present throughout, with participants expressing embarrassment, humiliation, and an overall lack of self-worth.

Christopher was embarrassed when perceived as “feeble” in social situations. The cultural perception of a man as strong and protective was at the forefront, and clearly informed his perception of social legitimacy, or lack thereof.

Box 25

Stanza 3: Feel like you’re being perceived as feeble

9. Yeah – the social thing and also just the – the feeling of
10. of being with your wife out some place and you have these awful spasms
11. that make you feel like you’re being perceived as feeble
12. I guess you would say – you just
13. you don’t feel like – Superman – anymore.

Stanza 4: That’s kinda the Superman – the protector and all that stuff

14. Yeah – that’s kinda the Superman
15. like nothing can stop me
16. when you feel like you’re the man
17. you’re the one that – stands up for
18. the protector and all that stuff.

Stanza 5: Then you’re bent over coughing into a bucket

19. and then you’re bent over coughing into a bucket
20. you don’t really like that person
21. it makes me feel great when I’m able to be that person
22. that’s a feeling that makes me feel really good
23. when I’m able to be the protector and be the everything and all that stuff.

(Christopher)

Christopher’s role identity is challenged when he falls short of his perception of what a man and a husband should be. In his mind, he should be “Superman”, “the protector”, and “the everything”. This is a high expectation for anyone, but especially for someone with CF, and he expressed surprise when realising during the interview the pain this was

causing him. He remarked that at the beginning of his interview he thought it would only last ten minutes, but he was now starting to recognise the extent of his pain. This expression of surprise at the depth of pain was not limited to Christopher, as three other participants made similar remarks after the interview concluded.

Also linked with productivity was achieving developmental milestones, closely related to the stories of independence and role identity. All participants spoke in some way about what they were supposed to be doing. For Christopher, this was centred around being a man and husband. For Brittany, it was a desire to be independent. Where Brittany's narrative differed from others was the depth of pain she expressed and the situation of her narrative in the present.

Box 26

Stanza 1: Having to stay at my parents' house

1. Having to stay at my parents' house for so long
2. not hard because I love them and they are like totally okay with me staying there
3. but like just as a personal goal and feeling successful
4. or feeling like I'm an adult.

Stanza 2: Being there has been really hard

5. being there – yeah – milestones that we work towards
6. being there has been really hard
7. painful for me like emotionally to deal with
8. and I'm wondering like how the rest of my life is going to go.

Stanza 3: I get paralysed by anxiety

9. I get paralysed with like anxiety
10. like thinking about my body issues – my physical state
11. and trying to address it and manage it
12. and then still think about everything else that I need to be doing

13. and creating a life and happiness and things that make me happy
14. and – doing those things when I'm like consumed by this like – problem
15. that kind of looms over me.

Stanza 4: My brain cannot tackle these things that I need to be tackling right now

16. Do people talk about this paralysed feeling?
17. Maybe they don't use that word but I use that word
18. it's like – oh my god – where do I start?
19. Oh my gosh I can't tackle
20. my brain cannot tackle these things that I need to be tackling right now.

(Brittany)

Brittany overtly labels her experience as painful emotionally. Social pain is evident in her account of feeling paralysed and unable to meet the milestones she perceives are expected of her. In parallel is her expression of existential pain.

The concept of existential pain is often used interchangeably with the concept of spiritual pain (this practice is not without critics, but addressing this controversy is beyond the scope of this thesis). Although multiple definitions of existential and spiritual issues are found in the literature, common themes exist among their definitions (Boston, Bruce, & Schreiber, 2011). In this thesis, existential and spiritual issues are understood through the search for meaning or purpose in life, questions about what happens after death, feeling connected to others, hope and hopelessness, and fear (Boston et al., 2011). All participants struggled with at least some aspects of existential pain, but for Brittany this experience seemed to consume her.

These stories of pushing through and independence reflect the cultural expectation, especially in the US, that individuals should contribute to society. Aspiring to independence is not necessarily negative but may not always be possible, especially as CF progresses and individuals get sicker. Less straightforward than the narratives of emerging awareness, these social legitimacy narratives elucidate more nuanced experiences with pain, especially the experiences of social and spiritual or existential pain. Closely linked, but distinct from the quest for social legitimacy, is the experience of invisibility.

5.4 Invisibility

Along with the narratives of disease progression and social legitimacy, all participants told narratives of invisibility. The phenomenon of invisible illness or invisibility has been described as an illness without a clear or visible outward sign, and has been closely linked with social legitimacy (Dow, Roche, & Ziebland, 2012; Hay, 2010). Pain, a highly subjective experience, and usually with no outward sign of illness or disability, is often included when discussing invisible illness. A close review of these narratives reveals a greater complexity surrounding invisibility than is suggested in the straightforward definition of an illness without a visible sign. In these narratives, invisibility is present when people feel misunderstood, is sometimes a choice, and can be disruptive when the invisible becomes visible.

5.4.1 Feeling misunderstood

Several participants spoke about feeling misunderstood, or that people do not understand the extent to which CF affects them and others. Other participants felt that even friends and family did not understand the unpredictability of CF. For instance, Michael had recently stopped working full-time at a grocery store because of attendance issues. He had near-normal lung function, but had severe gastrointestinal symptoms of CF.

Box 27

Stanza 1: The hard part of people understanding the pain with the CF

1. The hard part of people understanding the pain with the CF stuff –
2. cause they think – the CF they think lungs
3. they don't think the other stuff

Stanza 2: They look at CF lungs not CF everything else

4. which is part of a stressful part
5. with you trying to look at help for whatever like that –
6. they don't – they look at CF lungs –
7. not CF everything else so –
8. and that's stressful

(Michael)

Michael's words suggest that he feels invisible and misunderstood when others regard CF as only a lung disease. He appeared to think he would receive more empathy and accommodation if his symptoms were instead related to his lungs.

Feeling misunderstood for non–lung related symptoms was also expressed by Ashley, Amanda, Samantha, and Brittany. Symptoms described as misunderstood by these participants included abdominal pain, joint pain and fatigue. In addition to the physical symptoms, these participants also experienced social pain due to the conflicts that arose as a result of feeling misunderstood by others around them.

Lack of understanding when others looked at CF as primarily affecting the lungs was not limited to co-workers, friends and family, as participants also described similar experiences in CF clinic.

Box 28

Stanza 1: The focus has always been lung function

1. You know what
2. at the clinics it's very
3. the focus has always been lung function
4. Or am I gaining weight

Stanza 2: Lung function and weight

5. lung function and weight
6. these are the two questions that I focus on
7. what is my weight – has my weight changed, good or bad appetite
8. lung function – can I exercise

Stanza 3: Is there something else I need to be talking about?

9. that's what I focus on
10. So, I guess when I go to clinic
11. I'm not really thinking – like
12. is this something that I need to be talking about
13. you know what I mean?

(Brittany)

From Brittany's perspective, the focus of CF clinic was on lung function and weight, and these were therefore her primary focus as well. With this focus on only lungs and weight, Brittany did not report other symptoms that arose, including joint pain. Although joint pain did not appear to have a significant adverse effect on Brittany's quality of life, others (Jessica, Michael, Ashley, and Samantha) described severe joint pain, and yet only Jessica had discussed it during CF clinic. Interestingly, although Jessica reported joint pain and was receiving care from a rheumatologist, even she did not relate her joint pain to CF, although it is a known complication. When participants did not report joint pain, it remained invisible even to their healthcare team.

A common theme in discussions about invisibility is illness without outward visible symptoms. With a disease like cancer, people often lose their hair or have some other visible sign that they are ill. This is often not the case with CF, as many people do not have any outward signs of illness until advanced stages when oxygen is required. Pain is also often associated with invisible illness. Visible signs of illness communicate to others in society that the person is ill, and accommodation is warranted, but when visible signs of illness are not present, others may judge the person negatively. In Box 30, Amanda laments the loss of friends due to invisibility in CF.

Box 29

Stanza 1: I've lost a lot of friends over it

1. The thing with CF is you can feel fine one day
2. and then the next day be totally done
3. Just feel like you can't do anything –
4. I've lost a lot of friends over it

5. Cause they don't understand

Stanza 2: "Well you looked fine yesterday"

6. Because before you're wearing oxygen you look so normal

7. I've had so many say "Well you looked fine yesterday – you were fine"

8. That used to happen quite a bit –

9. So, you find out who your really true friends are.

(Amanda)

Amanda was not the only one to lose friends or feel judged by others because they did not look "sick". Only one participant (Matthew) did not describe such a scenario, perhaps because he had severe lung disease, was oxygen-dependent, and used a wheelchair due to severe shortness of breath. Matthew's illness was therefore visible and those around him may have excused him from expectations or responsibilities.

5.4.2 Invisibility as a choice

Another perspective is that of invisibility as a choice. Rather than thinking about invisibility from the perspective of others, from this view the person with CF is making a choice to be invisible.

Invisibility as a choice is demonstrated clearly, albeit sadly, at the end of Amanda's narrative about the death of her friends with CF (Box 15). As discussed previously in section 5.3.5 about mortality, she says, "It's like everyone has died and you know honestly I haven't really gotten close to anybody – I don't know any new CF-ers here."

In this situation, Amanda is making a choice to remain invisible to others with CF. While they may know that she has CF, she seems to be hiding her inner self. This is even more interesting considering that she does a lot of work in public awareness of CF and lung transplant, so is publicly visible but at the same time choosing to be invisible.

Other participants never formed relationships with others who have CF. Some expressed they did not feel a need to get to know others with CF (Christopher and Michael), while others avoided the pain, they believed would accompany these relationships. In Box 30, Samantha describes her reactions as she became aware of what CF meant in her life.

Box 30

Stanza 1: I mean it was hard – I cried a lot

1. I mean it was hard – I cried a lot
2. I went through like a pretty depressed phase
3. I hung out at home a lot
4. I quit my job
5. I stopped going to school.

Stanza 2: I don't have relationships with any other CF patients

6. I don't have any relationships with any other CF patients
7. I've never tried to be part of like a community – with the CF patients
8. because we can't be around each other
9. and then everything else is just like
10. well you could do online relationships
11. and that was just never appealing.

Stanza 3: Maybe end up with a very tragic result

12. Of course there's always the knowledge
13. They're dealing with something very serious and terminal
14. So it also feels weird to start up that kind of relationship
15. Knowing that you're both gonna deal with a lot of the same things
16. and maybe end up with a very tragic result.

(Samantha)

As the reality of having CF set in, Samantha withdrew from virtually all social relationships, quit both school and work and made herself invisible.

Participants tried to protect themselves by not forming relationships and avoiding being a witness to others' decline. This ensured they were invisible not only to others, but also themselves.

Several participants talked about choosing whether to reveal their CF to friends or not. Continuing with Amanda's narrative around invisibility, she talks about being diagnosed with CF during adolescence. In describing her adolescence, the pain of not fitting in is evident. She decided in adolescence not to reveal her diagnosis, although from her narrative it is clear that there were indeed visible signs of her CF, signs that others attributed to smoking which seemed preferable to Amanda over CF.

Box 31

Stanza 1: Teens were really awkward

1. Teens were really awkward
2. Because it was high school
3. I always made a big scene with the coughing
4. people thought I was a smoker

Stanza 2: My stepmother made me feel ashamed of it

5. I was almost embarrassed of it
6. My stepmother made me feel ashamed of it

7. I didn't really tell any of my friends about it

Stanza 3: I was just really embarrassed by it

8. I didn't want to tell any guys about it because I used to think

9. I don't want them to know when they're kissing me that I spit up this really ugly stuff

10. it's so gross and I was just really embarrassed by it

11. In my 20s I was more accepting of it.

(Amanda)

The decision of whether or not to reveal their diagnosis was a difficult one for most participants. Although as an adult Amanda now reveals her diagnosis, she keeps others with CF at a distance in an effort to avoid pain in the event of their death.

In another nuance of invisibility, some participants rationalised not talking about their pain because others had it worse.

Box 32

Stanza 2: That's how I get through things

1. Deep breath in – you just gotta focus

2. and you gotta know that it's not gonna be like that forever –

3. That's how I get through things –

4. and I always think that somebody else has it far worse than I do –

5. that's what I did before the transplant –

Stanza 3: There are so many people far worse than I am

6. I'm like there are so many people far worse than I am –

7. and then I look at all of my friends that have died –

8. look at their circumstances and just think –

9. I'm pretty lucky – pretty lucky

(Amanda)

This reluctance to express pain because others have it worse may be mediated by appreciation for all participants do have, or may be due to a cultural expectation to be tough and not complain. Regardless of the reasoning behind not expressing pain, it results in invisibility where the pain may not be recognised, understood or treated. This recognition that others have it worse recurred in several narratives.

Some made a choice to remain invisible by choosing not to talk about their pain. Michael equated talking about pain to a reminder of CF.

Box 33

Stanza 1: I'm tired of doing this all the time

1. Growing up I used to have no problems telling people what's going on
2. but I'm tired of doing this all the time
3. The CF, the pain, everything
4. I had no problem talking about it because
5. it didn't bother me as much as it does now

Stanza 2: I'm just tired of talking about it

6. Now it drains me a little bit here
7. and then the pain and everything
8. and I'm just like tired of talking about it
9. to remind me about everything going on.

Stanza 3: It's out of mind and I don't deal with it

10. Out of thought – out of mind
11. or whatever you know
12. instead out of sight out of mind
13. it's out of mind I don't deal with it.

(Michael)

As participants continually became sicker, having CF was increasingly overwhelming. Not talking about it was yet another way they remained invisible, even to themselves. At times it was simply more than they could deal with.

5.4.3 When the invisible becomes visible

Sometimes participants who previously chose to be invisible became visible. Visibility emerges then as a somewhat fluid state, changing in different situations. In some circumstances invisibility can be beneficial. For example, disclosing a diagnosis of CF may hinder employment or social relationships. However, efforts to remain invisible might also be harmful, as seen in Jessica's narrative in Box 34.

Box 34

Stanza 4: I don't know why I let it go so long

1. I feel like there was one pneumonia that I had that I didn't even know I had –
2. I think my daughter was still a baby –
3. maybe one – and I don't know why –
4. I don't know why I let it go so long but I was in pain – and I had a fever

Stanza 5: Feeling like I was drowning

5. I started waking up in the middle of the night
6. feeling like I was drowning –
7. like I – literally could not take – I could not breathe.

Stanza 6: I don't know why I waited so long

8. And I'd get out of bed and I'd go sit in the bathroom
9. and I would just be like struggling to breathe –
10. and I don't know why I waited so long –
11. I don't know why I didn't go in sooner

Stanza 7: By the time that I went in I had pneumonia

12. but by the time that I went in I had pneumonia
13. and I think that was the time I had the pleural effusion –
14. and my fevers were really – really high

Stanza 8: That was the most pain that I can remember

15. so it was just – that was the most pain that I can remember being in with pneumonia.
16. And I think it lasted probably about a
17. week – a good week – that I couldn't sleep –
18. and I was you know – I was in the hospital for a week and a half or two weeks I think.

Stanza 9: Why did I wait so long?

19. I look back on it and I'm like –
20. why did I wait so long –
21. how did I not know –
22. that there was something wrong –

Stanza 10: I can't imagine ever letting it get to that point again

23. I remember laying on the floor at my house
24. with the fan on and an ice pack on my forehead because I was so hot –
25. and I don't know why I waited so long
26. Cause I can't imagine ever letting it get to that point again

Stanza 11: I had a little one and I didn't want to admit that something was wrong

27. I had a little one and I –
28. I didn't want to admit that
29. something was wrong
30. and that I'd have to go in –

(Jessica)

Jessica was not the only participant with narratives about delaying or avoiding the hospital. This was also described by Ashley, Amanda and Brittany, but Jessica's story illustrated her thoughts and feelings around this in a way that is more informative than other narratives. It might be that Jessica wanted to be invisible to the hospital; it is not

entirely clear in her narrative. Her selection of words at line 36 is important. She said, “I didn’t want to have to be in the hospital.” This is different than not wanting to go to the hospital. She does not want to *have* to go. Perhaps her real meaning is that she does not want to have CF and is attempting to make the illness invisible. The meaning may not be clear, but the pain she feels as she lies on the bathroom floor, feeling as if she is drowning, is tangible.

In a slightly different perspective, Matthew talks about his struggle to express emotional pain in Box 34. When pain is kept inside, the person may be invisible, with those around them not knowing the extent of their suffering.

Box 35

Stanza 1: I didn’t used to cry

1. I think it’s more like emotional pain – like for me
2. Like – she (wife) taught me like it’s okay to cry
3. cause I didn’t used to cry
4. I used to just like keep everything inside of me
5. and only show machismo.

(Matthew)

Unlike other participants, Matthew’s narrative did not contain many elements of invisibility. The exact reason for this is not clear, but it may be because Matthew was near the end of his life. As previously described, he had been told that he could not receive a second lung transplant and was facing death. Matthew seemed to take pride in having come to terms with his prognosis, showing his emotion and talking openly with his family

about dying. Connected to oxygen continuously and needing a wheelchair, he was clearly not invisible.

When participants began talking about invisibility it was similar to an iceberg. Beneath the whole of the experience of having CF was this great iceberg of suffering that others didn't see. Clinic staff may have seen the tip of the iceberg, and friends and family just a bit more, but the world around them, their workplaces and schools, likely saw even less. To the outside world, their lives were largely invisible, but in telling their experiences for this research, the depth of their pain and suffering became more transparent. If one story seemed to most succinctly represent participant experiences with pain in CF, it was Brittany's retelling of a conversation she had with a co-worker.

Box 36

Stanza 4: Someone asked me like how does CF make you feel?

- 13. Someone asked me
- 14. a dorky guy at my work asked me
- 15. he was like what do you feel about
- 16. like how does CF make you feel?

Stanza 5: Do you really want to know?

- 17. and I was like okay do you really want to know?
- 18. really want to know –
- 19. I could go there do you really want to know?
- 20. and he was like yeah tell me

Stanza 6: It's just suffering, I can't explain it any other way

- 21. and I was like
- 22. it's just a lot of suffering
- 23. it's all just suffering
- 24. I can't explain it any other way.

5.5 Conclusion

This chapter has explored pain in CF through participant narratives of emerging awareness of having CF, social legitimacy and invisibility. These narratives included physical, emotional, social and spiritual (existential) pain.

In the narrative of emerging awareness, participants began by relaying stories of pain related to their diagnosis of CF. Following in chronological order were stories of disease progression and worsening symptoms, including pain. These stories of disease progression dominated the emerging awareness narrative and seemed to have great significance to the participants' experiences. As CF progressed and reality of what this meant set in, pain became more evident, especially spiritual or existential pain. Pain of all types was seen through this emerging awareness narrative, along the continuum of disease, from diagnosis, through early stages of disease progression, to advanced and more severe disease.

The narrative of social legitimacy was seen in stories of pushing through and independence. Stories of pushing through revealed the response to illness when cultural expectations encourage one to keep going at all costs. Closely related were stories demonstrating the cultural value of productivity, of being productive whether for the self

or society. Dominating the social legitimacy narrative were experiences of social and spiritual (existential) pain.

Finally, the narrative of invisibility permeated all interviews. When CF was invisible to others, participants felt misunderstood. Sometimes they chose to remain invisible, both to the outside world and even to themselves.

In Chapter Six, the participant narratives of emerging awareness, social legitimacy and invisibility are discussed in relation to existing knowledge through the lens of Saunders' theory of total pain.

Chapter 6: Discussion

In the findings, reported in Chapter Five, participants communicated their experience of pain through three key narratives: emerging awareness, social legitimacy and invisibility. Emerging awareness was a gradual unveiling of what having CF meant to participants. Multiple types of pain were present along the entire continuum as participants developed this awareness and struggled to be independent and maintain or achieve social legitimacy, sometimes sharing stories of feeling invisible.

This chapter begins with reflection on developing an understanding of pain as multidimensional and situated within the whole experience of living with CF. Next, a pain theory that will best facilitate understanding of study findings was sought and selection of total pain as a theoretical lens described. Findings are then discussed in relation to the literature presented on cystic fibrosis and pain in Chapters Two and Three, the broader literature around pain, living with CF and other chronic illnesses and palliative care.

Through the research process, an initial understanding developed of pain as multifaceted and important in participants' experience of living with CF. As seen in the findings, participants described their pain as primarily "emotional". It is not that participants did not talk about physical pain; indeed, they did, and it was evident that physical pain had always been a part of their lives. However, they described pain that was multidimensional, nuanced and complex.

Further illuminated in the setting of CF was that pain is integral to the lived experience of CF and cannot be extracted or carved out of the whole experience of living (Good, Brodwin, Good, & Kleinman, 1994). The interviews provided an opportunity for participants to share their experience with pain and when they did, they shared their overall experience of living with CF. However, these stories about pain were shared within the greater experience of living with a chronic and life-limiting illness, supporting the argument for studying pain in context of the overall life trajectory rather than in isolation (Good et al., 1994).

6.1 Theory to illuminate pain in CF

With pain in CF recognized as a multidimensional, complex experience occurring within the context of living with a chronic illness, a theory was needed that could illuminate and promote understanding. In exploring the theoretical bases of pain literature described in Chapter Two, none seemed sufficiently focused on the multiple facets of pain as experienced within the context of the whole person living with CF. Returning to the pain literature, the biopsychosocial model is now considered.

6.1.1 Biopsychosocial model

The biopsychosocial model of health and illness (Figure 4) by Engel (1977) has also been applied to the pain experience, encompassing physical, emotional and social elements of pain (Gatchel, Peng, Peters, Fuchs, & Turk, 2007). Engel (1977) argued that the biomedical model of health and illness alone is insufficient and proposed that psychological and social

factors should also be considered. With a perspective of pain as not strictly physical or biomedical, but rather a complex experience involving many contributing aspects, the biopsychosocial model has been increasingly adopted in medicine (Bervers, Watts, Kishino, & Gatchel, 2016; Gatchel et al., 2007). Of note, while acknowledging aspects of pain beyond the physical, Engel still saw pain as situated in the physician's realm, and advocated the biopsychosocial model to be used by physicians with psychiatrists addressing the non-physical aspects of pain (Engel, 1977).

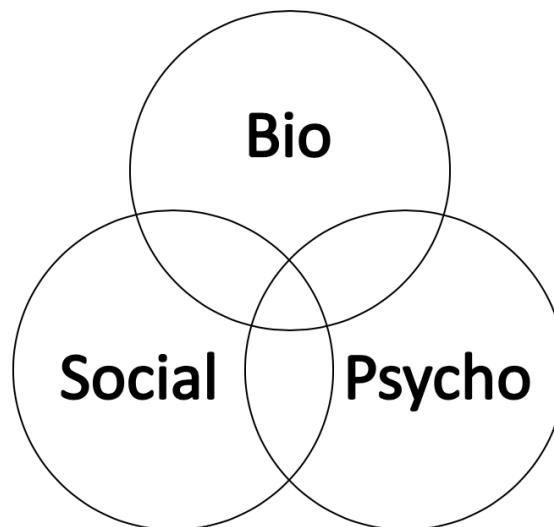


Figure 4. Venn diagram illustrating Engel (1977) theory for biopsychosocial model.

While the biopsychosocial model may come close, a theory with more emphasis on the person as a whole was desired. This endeavour led towards the palliative care literature and the theory of total pain by Saunders (1965).

6.1.2 Total pain

Introduced in the 1960s and evolving over the following decades, the concept of total pain includes physical, emotional, social and spiritual components (Clark, 2018a;

Saunders, 1965). In this model, the elements of total pain are interactive, as depicted visually in Figure 5. Saunders' theory of total pain greatly influenced the field of palliative care, which still strives to address physical, emotional, social and spiritual needs (Clark, 2018a).

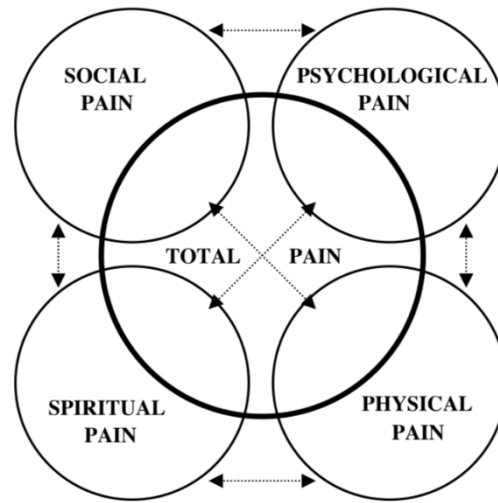


Figure 5. An interactive model of total pain. Reprinted from 'Understanding of the concept of "total pain": A prerequisite for pain control' by Mehta and Chan (2008, p. 30).

6.1.3 Development of total pain theory

Although Saunders introduced the concept of total pain in the 1960s, there is evidence of its beginnings in her earlier writings (Clark, 2018a). Conceptualised through her unique combination of experiences as a nurse, social worker (almoner), physician and scholar, total pain was a turn away from the previous notion of pain as chiefly physical (Clark, 2018a; Graham & Clark, 2008). For Saunders, total pain was comprised of not only physical pain, but emotional, social and spiritual pain as well. In developing the theory of total pain, Saunders spent extensive time with patients, listening and talking with them,

recording conversations and drawing on these narratives as she developed her theory. Saunders (2006b) valued these stories and their context within an individual's life and found it extraordinary that patients did not expect a physician to be interested in their stories of pain. In contrast, Saunders saw these stories as key to understanding multiple dimensions of suffering, and would likely support the narrative medicine movement today (Clark, 2018a).

6.1.4 Domains of total pain

The theory of total pain, having developed gradually through Saunders' work, continued to evolve over her lifetime (Clark, 2018a). Recognising that this division into domains may seem somewhat artificial, Saunders (1995, p. 45) nevertheless felt the domains would "enlighten us as we endeavour to understand the suffering of each patient and to help both them and their families". In other words, total pain provides a lens to view each individual, taking into consideration the many dimensions of their experience with pain and suffering, without which the clinician (or researcher) may miss an important aspect of their pain experience. Along with the physical, the other domains of total pain are emotional, social and spiritual.

Physical pain is often the primary focus of pain assessment and treatment. Saunders' perspective of physical pain was strongly influenced by the cancer population that made up most of her patient base. Types of physical pain included pain related to cancer itself, as well as pain related to debilitation (such as pressure ulcers and constipation), and pain related to treatment (such as surgery and radiation-induced esophagitis). Alongside

development of total pain theory, Saunders strived to improve treatment of physical pain, advocating for medication to be given routinely to prevent rather than only treat pain (Clark, 2018a). Additionally, Saunders viewed physical pain as inextricably linked to other domains of pain (Saunders et al., 1995).

Emotional pain, also referred to as mental pain and psychological pain by Saunders and others, encompasses a broad range of experiences (Clark, 2018a; Krawczyk & Richards, 2018; Saunders et al., 1995; Seymour, Clark, & Winslow, 2005). Depression, anxiety, uncertainty and even anger are all part of the manifestation of emotional pain. Fears about the future and worry about what will happen to friends and loved ones contribute to emotional pain. Isolation, encompassing the domains of both emotional and social pain, may cause some of the most severe suffering (Saunders, 2006b).

Saunders, following a period of questioning and self-proclaimed atheism, was a devout Christian and this undoubtedly influenced her to include spiritual pain as she developed total pain theory (Clark, 2018a). Despite her own personal views, however, Saunders recognised the importance of supporting the ill in their own beliefs rather than pushing them to adopt Christianity. Within total pain, spiritual pain may or may not have a religious dimension, depending on the beliefs of the ill person.

The search for meaning or a feeling of meaninglessness may be expressions of spiritual pain, as may regret and despair. Hope is often associated with spirituality, and when absent may cause deep suffering. Saunders felt that spiritual pain may cause the deepest suffering of all domains (Saunders, 2006b). She saw listening as critical in addressing

spiritual pain, even when the practitioner has no answers or the right words to say (Saunders et al., 1995).

Saunders' perspective of social pain primarily concerned the family of the ill person (Seale, 1998). In the context of family, social pain may include financial burdens experienced by a person and their family, or feelings of the ill person not wanting to be a burden to others or the real burdens of caregiving on the family. Saunders also saw grief as a form of social pain, beginning around diagnosis and continuing throughout the course of illness and into bereavement following death (Saunders et al., 1995). In the years since Saunders developed total pain theory, the notion of social pain is sometimes expanded to include not only the family, but the expanded social network of friends, work, community and society, discussed further in 4.2.3 Critique of total pain theory (Goebel et al., 2009; Gunaratnam, 2012; Krawczyk & Richards, 2018).

A sometimes overlooked aspect of social pain, is pain in clinicians who care for people with serious illness. Saunders and Baines (1983) contended that focus on total pain, on the whole person, also benefits clinicians. Pain was seen as an expected part of work with people who are dying, and Saunders advocated for both formal and informal support of medical staff, especially those new to the work (Saunders, 2006c; Saunders et al., 1995). A life outside of work was considered essential to longevity in the field, as was finding meaning in caring for the seriously ill and dying. In the years since total pain emerged through Saunders writing, it has become a key idea in the field of palliative care.

6.1.5 Critique of total pain theory

Enduring as a central focus in palliative care, total pain has become a foundational concept for many in hospice and palliative care (Krawczyk & Richards, 2018; McCaffery & Ferrell, 1997; Saunders, 2001). Despite being a central focus in palliative care, there is little research about the theory. Furthermore, total pain has primarily been applied in case studies or as a general recommendation, rather than the subject of study itself (Krawczyk, Wood, & Clark, 2018). This may be, at least in part, due to confusion around definitions of total pain, what it is and what it does (Krawczyk & Richards, 2018). Turning to how total pain has been applied and discussed in the literature will provide at least some information to begin to answer these questions.

As Saunders was developing her theory of total pain, she was aware of potential pitfalls in operationalising such a broad concept. She recognised the ideal of seeing the whole person but remained concerned that critical parts of suffering might be missed without a framework. Her vision of total pain was as much a philosophy, a way to see someone in deep suffering, as it was a guide for assessment and treatment. Her choice of the words 'total pain' in describing the theory is not without critics. Proudfoot (1976) criticised Saunders for using the word pain to describe mental, social and spiritual suffering, concerned that it implied these feelings should be avoided or medicated (Saunders, 2006c, p. 148). Saunders et al. (1995, p. 45) addressed this herself when she argued that these types of pain "should usually be faced rather than merely blotted out", cautioning

that use of the word pain should not imply that all domains of total pain require treatment with medication.

Strang, Strang, Hultborn, and Arnér (2004) similarly reported that it was suggested that total pain is not congruent with the widely accepted International Association for the Study of Pain (IASP) definition, relating concern about possible confusion regarding whether it refers to physical pain or to other types of suffering. Yet, Krawczyk et al. (2018) see the definition as congruent with total pain, perhaps because it is enveloped within “emotional experience” of the definition (Merskey et al., 1994, p. 210). Saunders (2006c) herself relates that her choice of the word *pain* in describing these facets of suffering was an intentional decision to guide the clinician’s gaze to these sometimes overlooked but important parts of the dying experience. Krawczyk et al. (2018) add to the rationale for use of the word *pain* when they remind readers that the word pain is already used figuratively by many people when they speak of illness and is therefore appropriate in the context of total pain.

There are multiple ways of understanding total pain, as seen in the numerous ways it has been conceptualized in the literature (Krawczyk & Richards, 2018). For example, Kendrick, Hudgell, Hellman, and Weaver (2019) applied total pain in a case study of a boy with Juvenile Huntington’s Disease. Their application had a predominantly biomedical focus, however in social pain they extended the theory to encompass the healthcare system and community, identifying a gap in available respite providers that prompted legislative advocacy (Kendrick et al., 2019). Warlow and Hain (2018) examine the literature and their

own anecdotal experiences to consider total pain in children with neurological impairment. Also emphasising the biomedical, they use the term “psychosocial” but social pain itself was unexplored. In contrast, Warlow and Hain (2018) thoughtfully attend to the domain of spiritual pain in children with neurological impairment, with an emphasis on attachment and relationship. Goebel et al. (2009) explored total pain and its application in heart failure, with more emphasis on the history of total pain and its situation amongst other pain theory. These authors demonstrate total pain applied in a straight-forward but limited manner, with symptoms delineated within the associated domains, but without critical review or engagement with the theory itself.

Although often applied simply, some researchers are engaging with total pain more critically such as Gunaratnam (2012), a sociologist who researches and teaches in the areas of race, ethnicity, and migration. Gunaratnam’s work with total pain helps expand the theory into the broader context of community, society and culture. Social pain in this broader context may be challenging for people to express, thus total pain theory may serve to facilitate recognition of an otherwise invisible phenomenon (Gunaratnam, 2012).

A prolific researcher on Saunders and total pain theory, Clark (1999) has considered the theory from multiple social and philosophical perspectives and continues to interrogate the theory today. In one of his many writings on total pain, Clark (1999, p. 734) presents the idea of total pain as a “paradox” that can both help and harm, as an extension of the “clinical gaze”, promoting a position of power. In this sense, total pain not only extends the realm of pain into the domains of the emotional, social and spiritual but also extends

the reach of the clinician's treatment into these areas. This essentially places the clinician in a position to both create and solve a problem, giving power to the clinician rather than the person in pain, in other words total pain is given by the physician, albeit unintentionally. Addressing this paradox, Clark claims, comes in viewing total pain as "lived" instead of "inscribed" (Clark, 1999, p. 734). In this shift of perspective, total pain allows one to embrace the position of another, to facilitate understanding of a complex experience, which Clark (1999) asserts was Saunders intention with the theory.

Krawczyk and Richards (2018) call attention to some of the challenges surrounding total pain. For example, the delineation of experience into the domains of total pain may artificially compartmentalize these aspects of experience. In a multidisciplinary team, this may serve to divide focus into these domains and actually diminish focus on the whole person. Saunders, with her training as a nurse, social worker and physician, and her wartime experience with the dying, was highly unusual (Clark, 2018a). Having such a unique combination of training and experience in a single person gave her a clinician's gaze unlikely matched by others who may not look past what they see as their specialty area. Krawczyk and Richards (2018) surmise that some of this confusion may stem from the original application of total pain theory in cancer at end of life and later expansion to the entire continuum of experience in serious illness, an expansion that not all health systems can support.

There are several examples of this expansion of total pain in other diseases, although similar to oncology it is most often applied in case studies (Goebel et al., 2009; Klepping,

2012; Mehta & Chan, 2008; Middleton-Green, 2008; Phenwan, 2018; Rao, 2016; Satija, Singh, Kashyap, & Bhatnagar, 2014; Warlow & Hain, 2018). Reflecting on the evolution of palliative care, including total pain, Saunders herself viewed the focus on primarily cancer as a failure, but acknowledged limitations as she added: “But how do we balance need, skills and resources?” (Saunders, 2001, p. 432). Since the time that Saunders wrote that article, answers to this question have been at least in part found in a delivery model recognising primary palliative care as the domain of all clinicians, with specialists delivering specialty palliative care in more complex cases.

Also contributing to lack of uptake may be the disparity between Saunders’ ideal and the realities of modern healthcare systems (Krawczyk & Richards, 2018; Krawczyk et al., 2018). The healthcare system during Saunders’ time and the fragmented healthcare system today are arguably quite different. With increasing health care costs and increased demands of electronic health records, clinicians today are expected to see increasing numbers of people in less time (Irving et al., 2017). Medical care in the US has become more complex, and people are living longer with more complications and comorbidities (Krawczyk & Richards, 2018; McPhail, 2016).

Further developing the evidence base, Krawczyk then joins with Clark as they look to further understanding and utilization of total pain. Krawczyk et al. (2018) consider that lack of understanding of the definition and operationalization in total pain may be limiting formal application of the theory. To further current understanding, Krawczyk et al. (2018,

p. 9) consider total pain theory in the context of a “bio-ecological” perspective. Krawczyk et al. (2018, p. 9) explain that:

“In this framing, who we are—the combined physical, emotional, and social aspects of being human—emerges as an embodied consequence of existing within, and being able to act on, dynamic individual (micro), community (meso), and structural (macro) environments that together make up a complex and always changing ecosystem”.

In this positioning of total pain all domains are equal in importance, rather than the often predominance of the biomedical, and total pain is seen not just as an experience that happens in or to an individual patient, but as an experience that occurs within a complex system that includes patients, clinicians, health systems, and communities, relationships between all of these entities, as well as all of the many values and priorities inherent in each one and the system as a whole (Krawczyk et al., 2018).

There is something aspirational about total pain as a guiding concept in palliative care, seeking understanding of the whole person, within their complex social networks. Total pain guides people to look beyond the patient and see a whole person, it facilitates a style of care, and indeed a style of research, with the person at the centre. With the person at the centre, connected not only to their care team, but also within social networks, care and research move beyond doing *for* and are better situated as simply part of the social world.

Although any of the pain theories described here and in Chapter Two would likely move the almost exclusively quantitative body of research forward, the way Saunders conceptualised total pain seems most appropriate for this study. Total pain theory, with its inclusion of spiritual, emotional and social pain (in addition to physical pain), offers an opportunity to conceptualise these other dimensions of pain. With total pain selected as the theoretical lens for this study, now the findings are discussed in relation to the literature on pain and living with CF. This discussion begins with the continual evolution of emerging awareness, with emphasis on the temporal experience in CF. This is followed by a discussion on total pain and suffering. Next, the discussion explores social legitimacy and invisibility in relation to the literature.

6.2 Continual evolution of emerging awareness

Findings reported in Chapter Six highlight that participants underwent continual change as they both lived with illness and searched for meaning, echoing previous research in chronic illness (Charmaz, 1983; A. W. Frank, 2002). This continual evolution has been reported in both chronic and life-threatening illness, as well as illnesses that fall somewhere in between (Bruce et al., 2014).

For participants, not only was their experience with CF changing as they became sicker, they were changing in relation to it. This is at least in part because the illness experience itself is considered to be socially constructed, reflected in the way people come to understand and navigate daily life in the context of their illness (Conrad & Barker, 2010).

These changes created a sense of movement or progression, and the significance of what it means to have CF continually evolved.

6.2.1 Experiencing time differently

As the meaning of having CF unfolded, participants seemed to experience time differently, a phenomenon consistent with previous studies of people who are ill (Gergel, 2013; Good et al., 1994). Participants experienced a shift from the linear sense of time (past, present and future), to a more pendulum-like experience of time, as described in chronic illness research (Charmaz, 1991; Marini & Stebnicki, 2018). Participants seemed to simultaneously live in the memory of the past by remembering a time they felt healthier, or even remembering trauma or pain, while at the same time coping with present physical and emotional stressors, all in the setting of anticipating future events and uncertainty. This is reminiscent of Bruner (2002, p. 93), who said, “Through narrative, we construct, reconstruct, in some ways reinvent yesterday and tomorrow. Memory and imagination fuse in the process.” Heaton (2015) characterizes a similar depiction of people with CF looking back and forth between their past, present, and future selves as “temporal comparison” of the self, a term introduced by Albert (1977). In a “temporal comparison” the individual seeks to make sense of the experience. These examples illustrate the ability of narrative to demonstrate how meaning changes over time. As they looked back, participants recalled milestones that seemed to mark these past events in their memories.

6.2.2 Milestones as markers of change

Signalling progression from diagnosis to advanced disease, participants described milestones in their illness, resonant with Charmaz (1991) description of “time markers” or “turning points”. These milestones created signs along a road map of participants’ lives. Milestones provide more than a marker of time; they help to create meaning as people reflect on past events to understand the present (Charmaz, 1991). Milestones or turning points described by participants signalled transition or movement along the continuum of illness. Some milestones were discrete events such as diagnosis of CF, first hospital admission or lung transplant, with a clear change in the experience that followed. Regardless of what the event was, the meaning to participants seemed to evolve, demonstrating the changing meaning over time (Charmaz, 1991).

Some milestones, rather than being a distinct event, occurred over a less defined period, as when participants were getting sicker, not responding to treatment, or developing new symptoms. Although Jessup and Parkinson (2010, p. 356) did not use the terms milestones or turning points, their description of “watersheds” leading to a “stark moment of the actual reality of having CF” is similar, and their emphasis on fear caused by these moments was echoed in participant narratives. Palser, Rayner, Leighton, and Smyth (2016) also echo the significance of milestones in their qualitative study of the first pseudomonas infection in CF. Palser et al. (2016, p. 4) in their descriptions of the first infection with pseudomonas (and other ‘firsts’, such as needing oxygen) as a “turning point in people’s relationship with CF” and marking a transition from one stage to

another. Similar transitions have been discussed by others researchers in CF as well (Dellon, Robinson, & Klick, 2011; Schmid-Mohler, Caress, Spirig, Benden, & Yorke, 2019). These milestones signalled a significant event, transition or turning point, and were often accompanied by fear or pain. In this study on pain in CF, laughter was especially prominent around milestones when participants were discussing their deepest pain, thus illuminating the key role that milestones may play in identifying total pain.

Milestones also supplied form and direction to the flow of emerging awareness. Participants looked back, reflecting on these milestones, and forward, anticipating future changes, becoming increasingly aware of what CF meant in their lives. Meaning shifted as they were continually in a different location along the trajectory of illness. That is, as Cassell stated, “Meanings also flow. Like the value or importance invested within them, meanings are not static, even if they often remain stable for long periods.” (E. Cassell, 2005, p. 309).

6.2.3 Between sickness and health

Although participants generally shared stories in a chronological form, their location along the narrative at any given point had an element of flexibility, a liminal quality. This phenomenon of liminality was present throughout the full spectrum of emerging awareness. In early illness, participants had CF but did not necessarily view themselves as sick, even though they still required hours of treatment every day.

When recalling earlier phases of illness, “healthy” became a relative term. Participants referred to themselves as healthy, while at the same time recalling the burden of treatments and symptoms. Participants were in a fluid state, moving between sick and not sick, with the definition of “sick” seen in retrospect of how they felt previously compared to how they feel now. What was considered sick one day may be health later. Along this continuum, participants moved from hope for a normal state of health, into the recognition that they would never have this. This notion of health echoes Lowton and Gabe (2003) in their work in adults with CF, where perception of health was found to be a relative and dynamic state, related to the participant’s status of illness and treatments, as well as the general context in which they live their lives. Similarly, Huyard (2008, p. 543) in her study of adults with CF, described a transformation from the perception of oneself as healthy to that of ill as an “illness career” in her study of adults with CF. She found that participants viewed their life in two distinct phases. The first phase was relatively straightforward and in this phase, participants did not consider themselves to be ill. The second phase was more convoluted, and it was in this phase that participants started viewing themselves as ill. As Huyard (2008, p. 539) describes, recognizing self as ill is a learning process that occurs over a period of time and awareness “generally emerges with a sudden worsening of the condition”. Milestones described in 6.2.2 echo this description of the shift that occurs with an abrupt change in condition, and the corresponding shift in recognition of self as ill is seen in Box 6 as Samantha begins “learning the depth of what the illness did”.

Badlan (2006) noted a similar ambiguity between being healthy and unhealthy in her study of young people with CF. In her study, she found that the CF treatment regimen seemed to influence the perception of self in young people, noting that this process can be painful and may affect acceptance of their illness (Badlan, 2006). Badlan (2006) emphasized the importance of CF team members understanding the temporality of the experience of being sick and recognizing that people with CF usually spend time in several other roles in addition to a sick role. This supports the goal of viewing a person in their wholeness, a goal supported in this study through use of total pain as a lens to consider the experience of pain in CF.

6.2.4 Death always looming

For participants, death was always in the background, lurking even when not the focus of the story. Increasing realisation of living less well meant that death became less hypothetical and more real. Participants were no longer able to keep it in the background as they became increasingly short of breath and spent more time in the hospital. There was a delicate balance of living life fully now, with increasing awareness of dying from CF.

It was this particular liminal space, between living and dying, where emerging awareness led. At its core, emerging awareness was not about becoming aware of what it means to have CF in regard to medications or facts about the disease, but a journey towards recognising that death is looming, that it is getting closer with each milestone. This recognition of mortality in CF. Uncertainty and fear were prominent along the continuum as participants became increasingly aware that they would eventually die from CF.

All participants talked about having outlived their life expectancy. People with CF often grow up with the thought that they are always at or close to the end of their life, perhaps first told they may live to about 12 years, later extended to 20 and still later to 30 and so forth, with death always looming not far off in the distance (Schubert & Murphy, 2005). As seen in this and other studies, for example Badlan (2006) and Jessup and Parkinson (2010), deaths of friends or even siblings from CF becomes a somewhat common occurrence in life and participants expressed a great deal of emotional and social pain with these experiences. This experience, with death always looming near but continually moving just a bit more into the future, constitutes a particular type of uncertainty.

At times, uncertainty presented in more practical ways, such as when participants felt they were unable to make plans for vacations or even dinner for fear that they would get sick and have to cancel. This is characterised as just one of many losses that occur in serious illness (A. W. Frank, 1995, 2002). Participants also shared other concerns about uncertainty within illness including uncertainty about diagnosis, severity of illness, treatment, and implications for life and future plans (Mishel, 1988, 1990). These were seen in narratives around diagnosis, treatment no longer working as well, school and work, and worrying about romantic rejection.

The deepest fears and uncertainties were seen as participants faced the reality of getting sicker and dying, and struggled to find meaning, as noted in Chapter Six when they whispered and laughed when relaying these concerns. Other researchers have suggested that uncertainty and unpredictability diminish the construction of meaning in the illness

experience (Charmaz, 1991; Marini & Stebnicki, 2018; Mishel, 1988, 1990). In a study examining symptom burden and unmet existential needs, adults with CF reported needs for additional support around fears of worsening CF, uncertainty, and fears of death, elements of emotional and spiritual pain (Trandel et al., 2019). In people with more severe disease, Trandel et al. (2019) found even more reported unmet existential needs.

In the face of uncertainty, participants were challenged to revise expectations, goals for the future, and even identity. Although uncertainty has often been presented as an experience to be feared, it may also evolve over time, leading to recognition of uncertainty as an expected state and even to personal growth, such as when participants talked about uncertainty being normal or living fully while they still could (Mishel, 1990).

6.2.5 Biographical disruption in cystic fibrosis

Echoes of the evolution of emerging awareness can be seen in the body of research on biographical disruption, yet there are differences in the experience in CF. In his work on biographical disruption in rheumatoid arthritis (RA), Bury (1982) aimed to speak with people early, identifying potential participants as they were referred to a rheumatologist. In both RA and CF, symptoms are often gradual in onset. However, CF diagnosis usually occurs in the first two years of life, and most people with CF (even children as seen in Brittany's narrative) are aware of their terminal diagnosis. Furthermore, although symptoms might increase gradually, the treatment regimen, or "lifestyle" as Christopher called it, is perceived as a burden by participants even in the apparent absence of progression in early disease. In CF, then, the experience of biographical disruption is likely

different than in RA. Indeed, Williams et al. (2009) also found biographical disruption in CF to be different than when described in RA. In interviews with young people diagnosed with CF, they described the experience to be more of a “continual biographical revision” as opposed to distinct disruption, appearing to overlap with the description in this study (section 7.1.1) of experiencing time differently, with participants continually reflecting on their past and anticipating their future (Williams et al., 2009, p. 1453). Although there is not space in this discussion for a thorough examination of biographical disruption in the context of CF, there was a glimpse in the narratives when Ashley said, “We’ve been doing it all our lives, it’s all we know.”

Considering emerging awareness as a whole, participants were clearly still in the midst of this continuum. Even as participants recalled past events, their relationships to these events had evolved over time. This evolution occurred in the setting of changing meaning, worsening illness, and changing cognition as they became adults and had increasing capacity for understanding. All stories of emerging awareness involved this sense of movement, change, and an ever-increasing presence of total pain and suffering.

Total pain, as a theory, makes sense of emerging awareness by providing the context to view this narrative within the wider experience of living with cystic fibrosis. Without the lens of total pain, the emerging awareness narrative may have been overlooked as unimportant and simply the timeline upon which pain is described, rather than an integral and illuminating component of the experience of pain in CF.

Figure 6 illustrates a conceptual model for the continual evolution of emerging awareness. It shows the gradual increase in awareness as participants began to recognise what having CF meant in their lives. Bidirectional movement, often pendulum-like, is displayed along the continuum. Milestones represent events that signify change and are depicted by signs along the evolution of awareness. The figure shows death as present, lurking in the background of participants' stories as they became increasingly aware of its presence.

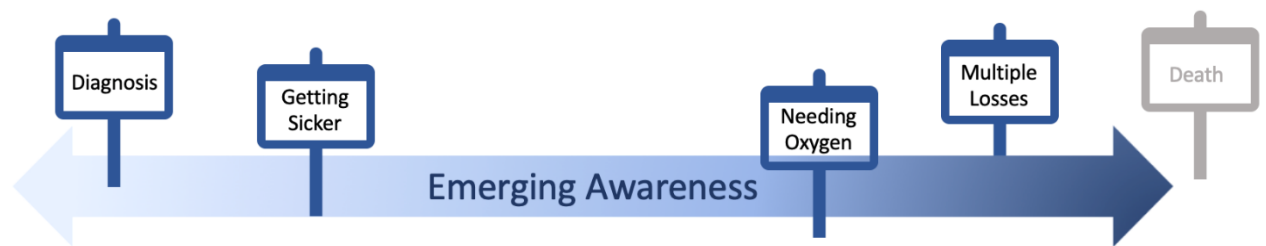


Figure 6. Emerging awareness in cystic fibrosis.

6.3 Total pain in cystic fibrosis: “It’s all just suffering”

As seen in the literature review, presented in Chapter Two, studies of pain in CF have been centred on physical pain, which is common (Havermans, Colpaert, De Boeck, Dupont, & Abbott, 2013). Locations of physical pain in the findings were consistent with the literature review. However, while all narratives included discussion of physical pain, their focus was on other types of pain, most often dubbed “emotional pain” by participants. Participants interpreted pain in CF broadly, albeit reluctantly, asking hesitantly if emotional pain was included – in the words of one participant, if emotional pain “counted”.

The quantitative nature of previous studies on pain in CF provided little more than a glimpse of the “emotional pain” described by participants, seen in the studies that examined potential relationships between pain and psychological correlates for quality of life using the CFQ-R (Blackwell & Quittner, 2014; Hayes et al., 2011; Kelemen et al., 2012; Munck et al., 2012; Palermo et al., 2006). In this narrative study, participants shared much more detailed and rich descriptions about their experience with pain in CF.

6.3.1 Emotional pain more than anything

Although participants usually began with describing physical pain, they quickly moved on to descriptions of emotional pain, which then encompassed much of their pain experience. The stories of physical pain served as a kind of entry point into discussion of emotional pain, and this in turn revealed a deeper and far more complex experience than previously understood. Participants referred to any pain that was not physical as emotional pain but did not label it more specifically. Amanda said it best: “It’s emotional pain more than anything.”

Participant descriptions of their experiences with emotional pain were more diverse than the label emotional pain would suggest. Emotional pain for participants included pain with loss of friendship, loss of jobs, death of friends and family, grief associated with loss of physical abilities with increased severity of CF, loss of an imagined future self or meaning, fear and uncertainty, to name just a few of the scenarios described as emotional pain. Although participants did not label their experiences beyond “emotional pain”, a language and framework are found in the general pain and palliative care literature.

As noted in the literature review, historical definitions of pain have been fundamentally about physical pain. While some definitions included the psychological response and social context of physical pain, they fell short of acknowledging emotional pain as pain in and of itself. Saunders, in recognising physical, emotional, social and spiritual pain, bridges this gap in understanding. Participant narratives well represent Saunders' concept of total pain as affecting physical, emotional, social and spiritual realms, allowing pain to be considered in different ways. If pain is understood as an experience other than just physical, total pain then supplies a framework and language to communicate about it.

Although different domains of pain in Saunders' theory provide a framework to talk and think about pain, the findings in this thesis suggest that separation into distinct domains may not recognise the importance of the relationship between domains of total pain as experienced in a whole person living with chronic illness. In other words, despite the theory's usefulness, the concept of total pain may risk reductionism. As described in Chapter Four, Saunders recognised this risk of reductionism and in her earlier writing described the risk of overlooking the whole person when focusing on separate domains. Heeding Saunders' caution to listen to the individual's story and their experience as they describe it may improve the risk of reductionism.

In initial consideration and analysis of data for this study, pain was indeed visualised in distinct categories, envisioned as more or less separate experiences. The findings, however, show that total pain is much more complex than just a distinct category depending on which part of the body or mind is affected. In CF, total pain is part of

everyday living with emphasis across the domains changing in relation to time and context. Pain cannot be carved out of the overall experience of living with CF. Total pain is a complex of the whole being in pain, a totality that is much more nuanced than just being in different domains on different days.

Understanding pain in CF requires considering how it infuses every part of people's lives, their relationships and their whole being. Total pain theory furthers understanding of the experience of pain in CF, but if viewed only within distinct categories damage to the entire person may not be fully appreciated. To deepen this understanding, an exploration follows of the concept of suffering as part of the CF pain experience.

6.3.2 Suffering

Despite controversy, as outlined by A. W. Frank (2001), about whether suffering can truly be studied at all, and with minimal representation in the literature on pain in CF, it is relevant in this thesis to include a discussion about suffering, as most participants referred to their own experience as suffering. The use of narrative appeared to facilitate the identification of suffering in CF as a constant experience throughout people's lives, Narrative interviews, for instance, can allow participants the space to share rich accounts of experience that may span their everyday lives and express their suffering, even described as giving voice to suffering (Kleinman, 1988; Loeser, 2005).

Participants spoke of suffering in the context of living with CF, supporting A. W. Frank (2004) that like pain, suffering is best explored in the context of the illness experience.

Similarly, Cassell stated that “suffering is experienced by persons, not merely by bodies, and has its source in challenges that threaten the intactness of the person as a complex social and psychologic entity” (1982, p. 639).

As participants shared stories about their illness, they were not merely sharing facts about their illness or treatment but revealing what it was like to suffer. Even when not overtly labelling it, participants expressed suffering through sadness or a sense of unfairness. Sometimes suffering was expressed in a pained expression or body language, and made clear through metaphors, a powerful use of language common in those who suffer (J. Bourke, 2014; Good et al., 1994). Christopher’s metaphor of pain in CF as “kind of like how a drop of water on your forehead isn’t uncomfortable until the 100,000th drop that lands on your forehead” brings the listener closer to his experience. Suffering is also found in the participants’ search for meaning or in asking the same, unanswerable questions, illustrated by Brittany as she wonders what CF “means for my life” (Charmaz, 1983). Although all participants were searching for meaning, Brittany returned to this question of meaning repeatedly in her interview and had a sense of desperation not seen in the other stories.

Suffering, closely linked with loss in the literature, is seen in the narratives as participants describe numerous losses of control, self, health, friends, future dreams or plans, and life as they know it (Ferrell, 2008). Through their stories, participants communicated that much more is going on under the surface than shared with others in their lives, including family and friends, and even more than they themselves may appreciate. Participants

who seemed well from outside appearances and described themselves initially as doing well revealed that they were in fact, often in their own words, suffering. For instance, participants described pain in all its forms, from becoming aware of what having CF meant in their lives, to striving to be independent and feel successful, to feeling invisible and misunderstood, getting sicker, and wondering when their life would end, while life as they knew it was already changing. None of these experiences occurred in isolation; they converged and affected the whole being. When viewed as a whole, it was evident that their experience was all-encompassing. Suffering seemed indistinguishable from the experience of illness and total pain, supporting the notion that it is best understood in the experience of the whole person (Ferrell, 2008; Good et al., 1994).

This understanding of suffering in CF as the expression of total pain, where the two are in essence one experience, is a key contribution to knowledge about the all-encompassing nature of pain in CF. Illustrating the experience of total pain and suffering in CF, Figure 7 builds on the earlier depiction of the continual evolution of emerging awareness. Total pain and suffering encompass the domains of total pain described by Saunders (2006a), and are shown as a singular experience occurring within a whole person living with CF, alongside the continuum of emerging awareness.

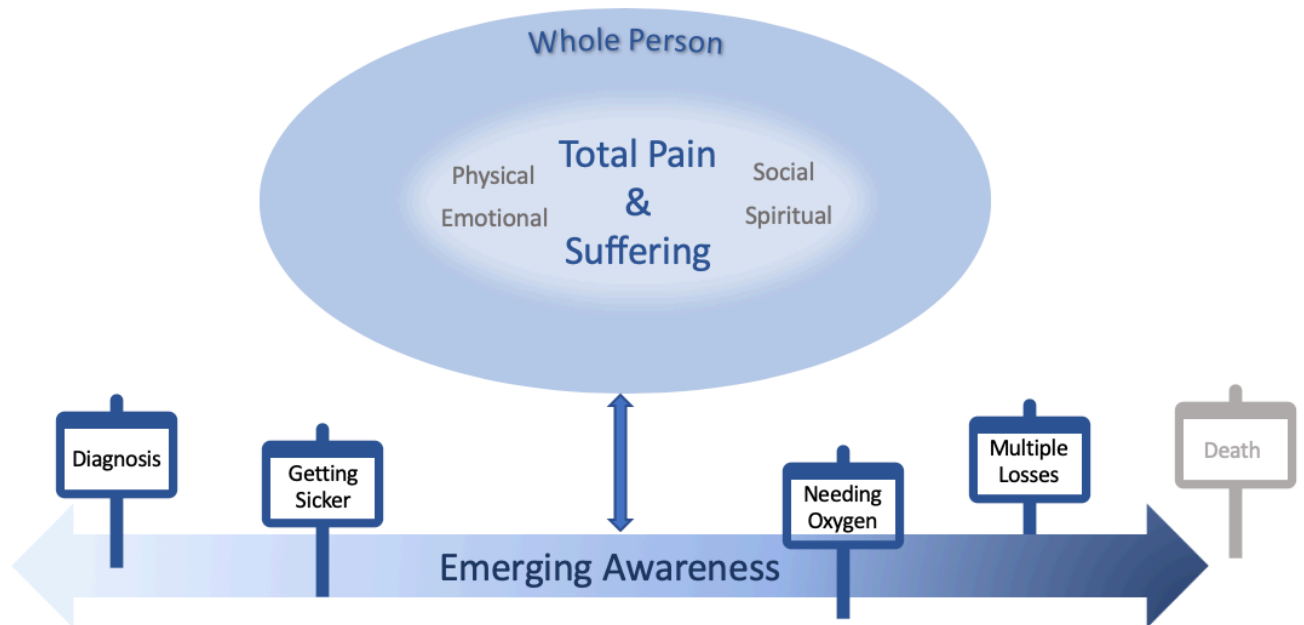


Figure 7. Total pain and suffering in the context of the whole person living with cystic fibrosis.

6.3.3 Finding meaning in suffering

Much has been written about finding meaning and purpose in suffering (Charmaz, 1983, 1999, 2008; Frankl, 2004; Kleinman, 1988). Some may find suffering to have positive consequences, such as valuing life more, striving to live more fully or finding deeper meaning in life. The participants in this study spoke of doing things while they still could, such as Matthew getting married and he and his wife having a child sooner than planned. Therefore, to say that participants are suffering does not lessen the meaning some may find through the experience.

Suffering was present in all participant narratives, but that is not to say that the participants were complaining or had a victim mindset. Indeed, while some participants

seemed to experience spiritual pain as they searched for meaning in their experience, suffering for others may have had some positive aspects. Some researchers have described narratives of suffering as integral in coming to terms with and understanding a changed or changing life (Charmaz, 1999). Suffering may lead to a more evolved self (Kearney, 2000). In CF and other life-threatening chronic illnesses, a search for meaning may lead to transcendence as people navigate their way through the experience of worsening illness, disability and death. When Canda (2001) interviewed 16 people with CF many of them described a heightened interest in meaning attributed to awareness of their own mortality. However, he noted that this path of transcendence was not an easy one, stating that participants “described times of great physical discomfort, emotional stress, intellectual quandary, and social relational impediment” (Canda, 2001, p. 122). The focus of his research is clearly centred on the spiritual, but this sentence gives a hint of a potentially broader experience not unlike total pain. Returning to the experience of Brittany and her desperate search for meaning, perhaps she is on a path towards transcendence but at the time of the interview she was, in her own words, suffering.

The scope of this study does not support an in-depth evaluation of the benefits of suffering in CF, but further investigation by those interested in exploring this phenomenon may be warranted. Although there may be some benefits in suffering, for many, pain and suffering in the context of chronic, life-limiting illness can seriously threaten social legitimacy.

6.4 Social legitimacy

Thus far in discussing the experience of pain in CF, this thesis has explored the emerging awareness of what it is like to have CF, and the experience of total pain and suffering. It has drawn from the literature on chronic illness, pain, suffering and palliative care to help make sense of the participant narratives in the context of existing knowledge.

In examining the narratives, the depth of suffering expressed by some participants was astonishing, but not entirely unexpected. The close link of social legitimacy to the experience of living in CF, however, was not expected yet affected all participants. It was in looking through the lens of total pain, the wholeness that included emerging awareness of what it means to live with CF, pain and suffering, that the critical role of social legitimacy was identified. Indeed, the place of social legitimacy in the experience of pain in CF is a key contribution of knowledge emerging from this thesis. For participants, after a lifetime of emerging awareness of what it means to have CF, as pain in all its facets led to suffering, nowhere was the deeper impact of living with CF more obvious than when they felt that they had no social legitimacy as people.

Social legitimacy first presents in the participant narratives as a response to illness, a coping mechanism. As people emerge into awareness of this chronic, life-limiting disease, this is part of how they deal with it, by striving to achieve or maintain social legitimacy. This was clear in all participants in their aim towards independence. It was also a driving force behind striving to achieve or maintain a desired social role, seen when participants “pushed through” to attend a social event when in pain or sick, or when they dedicated

significant effort to maintain employment despite worsening illness. Similar to the participants in this study, Saldana, Pomeranz, and Young (2018) found that people with CF may continue to work even with quite severe disease. Other studies in CF reported severe lung disease is not in and of itself an indicator of work disability, finding that flexible work and other accommodations, along with psychological and educational factors are more closely associated with work status (Burker, Sedway, & Carone, 2004; Hogg, Braithwaite, Bailey, Kotsimbos, & Wilson, 2007). Good et al. (1994) even described work as a respite from pain.

Social roles for participants mirrored those seen in the general public, such as friend, spouse or partner, parent, child, caregiver, employee and volunteer. Pain, suffering, chronic illness and disability have been studied in relation to social roles, strategies used to maintain functioning, and disruption that results from lost ability to maintain these roles (Good et al., 1994).

6.4.1 Sociocultural conditioning

The striving for social legitimacy seen in participant narratives reflects points described in the literature on sociocultural conditioning. In western society, value is placed on hard work, productivity, accomplishment, financial success and independence (Charmaz, 1983; Hay, 2010; Marini & Stebnicki, 2018). This perception of social legitimacy may be difficult to reconcile with an illness such as CF, and despite understanding CF as life-threatening, progressive and unpredictable, society often reinforces these expectations of hard work and productivity. Even when those around them are understanding and accommodating,

people with CF may still place similar expectations on themselves, illustrating self-regulation of societal norms.

Western societies often place high value on this model of pushing through illness, including their healthcare systems (Hay, 2010). For participants in this study, their society, including family, friends and even clinicians, reinforced this style of coping through praise and admiration, often elevating the person to hero status. For example, Ashley is talking about this very admiration when she describes her experience pushing through in CF, and how this has inspired her friends and family. This is so ingrained in the expected way of coping with illness that participants did not identify with any other way of living with CF, best demonstrated by Ashley's remark: "I think everyone with CF has to push on... it's all we know".

This social model is also seen through using language of battle and war when talking about people who are ill. They are encouraged to fight, illness is framed as a battle or a war, and they are said to have "lost" their battle when they die. Especially prevalent when talking about cancer, this language of battle is also common in CF and is present throughout the narratives.

6.4.2 Trying to be independent

The desire to be independent was also influenced by the high value placed on independence in western society (Hay, 2010). This was especially prominent in Amanda's case as she said that she wanted to be independent, take care of herself and be financially

self-supporting, but because financial independence was not possible given her CF, she was “on the system” and felt humiliated. Given the diagnosis and associated medical expenses and disability, people with CF can be caught in a cycle of receiving assistance through Supplemental Security Income (SSI) and then being subject to the limitations on employment required to maintain their coverage, an experience not unique to CF in the US (Marini & Stebnicki, 2018). The social cost of being trapped in this system manifests in social pain as seen in feelings of shame and worthlessness, illustrated so clearly in Amanda’s story.

As they became increasingly aware of what it meant to have CF, participants continually strived for social legitimacy, pushing through to be “normal” and independent. Present even in children, D’Auria, Christian, and Richardson (1997, p. 106) described participants ages six to twelve years trying to “keeping up” with their peers. This is a part of usual growth and development, but in CF it is interrupted at best and may be impossible in the face of a burdensome treatment regimen and worsening illness. Some with CF report that they are often faced with skipping daily treatments so that they do not miss social events (Gjengedal, Rustoen, Wahl, & Hanesta, 2003). Striving for social legitimacy, pretending not to be sick, and trying to be normal and independent is often impossible in the face of worsening illness. Even basic social outings may be skipped due to sheer exhaustion and embarrassment related to social judgement (Cordeiro, Jesus, Tavares, Oliveira, & Merighi, 2018). Striving for social legitimacy may at times be beneficial, but can be physically exhausting and unsustainable, and when unable to keep up the effort people may lose their sense of identity and self-worth, thus experiencing social pain (Hay, 2010).

6.4.3 Working harder

As these participants with CF became sicker, they explained how they had to work harder to maintain earlier levels of function. Inevitably there came a time when regardless of increased effort they could no longer keep up.

As in the earlier discussion about liminality, participants were situated somewhere along the continuum between the societal role expectations of the healthy and of the sick. Society expects those considered healthy to conform to expectations discussed earlier. Those in a sick role, excused from these societal expectations, may have other obligations now placed on them. These obligations require the sick person to want to get better as soon as possible and to seek and comply with medical advice (Hay, 2010; Larkin, 2011). However, what happens when a person does not fit neatly into either role?

Narratives for participants clearly caught in the middle of these two roles had an element of chaos that was not as prominent in the others. This is seen throughout the entire narrative for Brittany as she struggles with maintaining social legitimacy, exclaiming repeatedly that she does not know how to do all the things she is supposed to be doing in her life. The chaos depicted in Brittany's narrative may, at least in part, stem from the pattern of constant change related to illness, leading to starting and stopping school, moving out and back in to her parents' house, a pattern described by Saldana et al. (2018, p. 431) in participants with more advanced CF as "disruption and interruption". As participants became more dependent on others as illness progressed, they viewed this as negative and expressed embarrassment about it. Although adults, like Brittany, may

express embarrassment or frustration about being somewhat dependent on others for help with therapies and activities of daily living, assistance from others is common in adults with CF, especially assistance from parents (Edwards & Boxall, 2012; Lowton, 2002b; McGuffie, Sellers, Sawicki, & Robinson, 2008).

6.4.4 Losing social legitimacy

As participants were moving away from the healthy role and towards the sick role, they experienced numerous losses, from social role, identity and independence, to hopes for a life previously known or hoped for. As these losses began to mount, they contributed to total pain and suffering and decreased social legitimacy. Some participants, in response to these changes, also described becoming more socially isolated (Charmaz, 1983). Friends stopped inviting them to social events when they repeatedly said no or had to cancel at the last minute, as shown in Amanda and Ashley's stories. Participants sometimes lost friendships over these events. Other times, as seen in several participants' stories, the increased focus and energy required to deal with worsening CF meant they no longer had the energy for keeping social relationships and became even more isolated. Some were marginalised on the fringe of society, becoming increasingly isolated as illness progressed, and barely taking part in social processes, such as when Amanda described doing treatments from sunup to sundown. Isolation, marginalisation and lack of social legitimacy can lead to invisibility.

6.4.5 Gradual descent towards invisibility

As illustrated in Chapter Six, stories depicting invisibility were prominent in the findings. Saunders (2006b) spoke of invisibility as a source of great suffering, and indeed as these stories repeatedly surfaced, the depth of participants' social pain, suffering and loss of social legitimacy started to appear. This finding of invisibility led to identification of social legitimacy. The extent of total pain and suffering beneath the surface is key to understanding the experience of total pain in CF.

Participant narratives were filled with reports of feeling misunderstood and judged by friends, family, colleagues and even the CF team. The phenomenon of feeling misunderstood and feeling the need to remind or prove to others that they had a chronic illness is not unique to CF. Researchers studying invisibility in chronic illness have written about people with invisible symptoms who are unable to be productive feeling devalued, misunderstood and judged by others about the legitimacy of their illness (Charmaz, 2002, 2008; Hay, 2010; Larkin, 2011; Marini & Stebnicki, 2018). Similar experiences have been reported in other chronic lung diseases, including COPD and pulmonary hypertension (Fraser, Kee, & Minick, 2006; Gysels & Higginson, 2008; Yorke, Armstrong, & Bundock, 2014).

Invisibility can be problematic in those who are unable to conform to societal expectations, but has also been described as beneficial when people want to hide or minimise the effects of illness (Charmaz, 2008; Hay, 2010). There are several possible explanations about why someone with CF might choose to be invisible. Some participants

explained that they keep suffering out of view of others, sometimes to protect them or in response to an earlier loss of moral status. This concern about being stigmatised or not wanting to be labelled as sick and treated differently also led participants to choose to be invisible by not disclosing their diagnosis of CF. Broekema and Weber (2017) found that participants carefully weighed the risks and benefits of disclosure after previously experiencing negative reactions in friends. Lowton (2004) also described a careful consideration of risks and benefits in disclosing a CF diagnosis, adding that fear surrounding disclosure was related to the level of risk. Less risk was present in casual encounters with the greatest risk present in discussions with potential partners or employers, as seen in this thesis, with Samantha's fear of disclosing to boyfriends. Even children struggle with whether to tell others about their diagnosis and fear the negative social consequences of disclosure (D'Auria et al., 1997). At times this hiding or silence can become a habit and perpetuate invisibility, as when participants sometimes isolated themselves when unable to fulfil social roles in the way they wanted (Charmaz, 2002). This was seen in participants who chose to stay home more often and no longer spent time with friends or quit their jobs when they felt devalued.

Similarly, participants avoided seeking treatment for worsening symptoms, thereby being invisible to their clinical team, although this may have been more of a way to exert some degree of control over their illness than an adherence or avoidance issue.

In this study's findings, recall Jessica lying on the floor, unable to breathe and feeling that she was drowning, but refusing to go to the hospital. In her story, she was unsure why

she did not go to the hospital and wondered aloud if this was because she had a baby and did not want to leave her. But her statement about not wanting to *have* to go suggested a feeling of loss of control over her illness, magnified in the setting of being a new parent. Fear and existential threat around loss of control has been previously described in pulmonary exacerbations in CF, chronic obstructive pulmonary disease (COPD), and asthma and also compared to the experience of Post-Traumatic Stress Disorder (PTSD) (Schmid-Mohler et al., 2019). In an ethnographic study in CF, Maynard (2010) viewed situations like Jessica's as a pursuit to be normal while living with CF, measured through comparison with individuals perceived as healthy. This constant comparison of self to others and trying to be normal led people to try and make CF invisible, sometimes leading to a "dysfunctional interpretation of the true risks confronting them" (Maynard, 2010, p. 196). Seeing this as set in the greater social context of what a mother should be, Maynard (2010) sees this as more limited by society's ableist narrative of what being a mother should be, rather than a limitation born of CF itself.

Participants sometimes felt clinicians reinforced invisibility by conditioning people in what is proper to discuss in their clinical visits, albeit unintentionally. This was clear when all participants commented that clinic visits are all about lungs and weight. Despite the longstanding presence of a multidisciplinary care model in CF, people still felt that teams focused on just lungs and weight. With so much focus on these aspects, pain and suffering often remain invisible. Participants adjusted to what they thought was expected of them. In this way, the healthcare community encourages people to view illness, in this case CF, from a biomedical perspective and this has contributed to invisibility.

Viewing the social legitimacy and invisibility narratives through the lens of total pain illuminated participants experiences, highlighting connections between the narratives and how they contribute to the experience of pain in CF. In Figure 8, social legitimacy and invisibility are now added to the conceptual model of total pain and suffering in CF. This illustrates how total pain and suffering occur not only in the whole person living with CF, but also within the expectation of social legitimacy. Invisibility is depicted within the individual as a consequence to loss of social legitimacy and may be present throughout the life course.

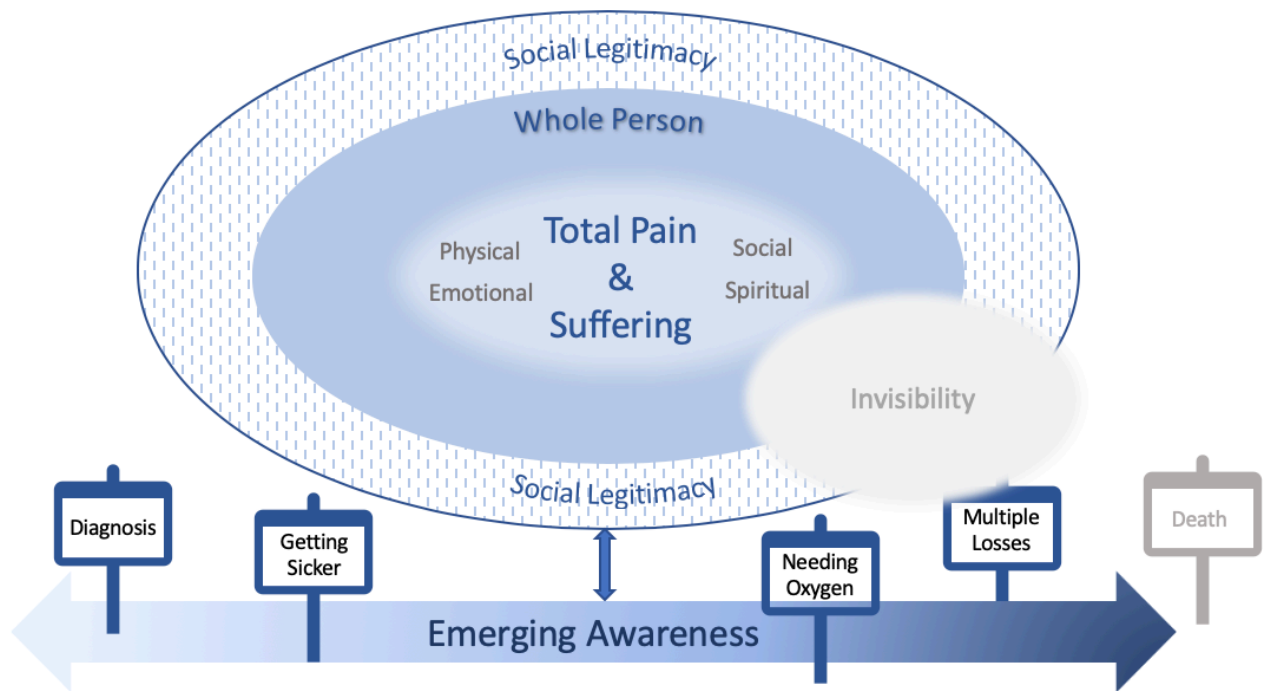


Figure 8. Social legitimacy, invisibility and their relationship to total pain in cystic fibrosis.

The nature of invisibility is that it is not usually seen by those surrounding the person with CF in their social circle. This presents challenges in learning more about the experience, but given these interviews, people with CF are willing to share their stories when given

the opportunity. Although the study was not seeking knowledge about the experience of invisibility, it was overtly present in the narrative and seemed closely related to total pain and suffering. Invisibility seemed to both come out of and cause total pain and suffering, which calls for further investigation.

6.5 Conclusion

This chapter has discussed the experience of pain in CF, in context of the overall experience of living with illness. Literature in pain, palliative care, and living with chronic illness helped to make sense of the findings in relation to existing knowledge. Findings reinforced the assertion that researchers should study the experience of pain in relation to the overall experience of living, as when asked about pain, participants told their stories in this context. Focusing on the person as a whole has been a goal throughout the study, essential to both research and clinical practice in pain and palliative care. Total pain has helped this study to move towards accomplishing this goal.

Participants shared their stories along a narrative of emerging awareness where across their lives, they gradually became aware of what having CF meant. Findings suggested that participants lived in a liminal space of always being “in between”, existing between sick and not sick, health and illness, living and not living. Total pain was present in various forms across the entire spectrum of emerging awareness, but especially impactful to participants was the uncertainty and fear found in these liminal spaces.

The findings show that participants viewed the experience of pain in CF as more than just physical pain, with emotional pain dominating their narratives. Using the lens of total pain to help guide interpretation, all domains of total pain were present within the narratives. However, participants also spoke of suffering, so it was beneficial to turn to palliative care literature on suffering to further understanding.

After considering suffering in relation to total pain theory and as affecting the whole person, understanding of total pain and suffering in this study evolved to a view of total pain and suffering as a singular experience. Participants described suffering in CF, and through their stories suffering was like an iceberg, with only a glimpse visible above the surface and great depths of suffering below. This study supplied a narrative opportunity for participants to communicate this suffering.

This study argues that total pain in the context of living with CF threatens social legitimacy. Efforts of participants to maintain societal expectations often hide the pain and suffering from even those closest to them, including family, friends and clinical teams. Eventually, when pain and suffering persist, participants may lose social legitimacy and subsequently feel invisible. These findings highlight the importance of studying the experience of pain in CF in the context of the whole person living with illness.

Chapter 7: Conclusions and recommendations

This chapter presents a summary of the study and original contribution to knowledge. It also includes a discussion of study limitations as well as strengths. Implications for clinical practice and a self-reflection follows, and the chapter concludes with opportunities for future research.

7.1 Summary of the study

In this thesis, I sought to gain a deeper understanding of the experience of pain in CF. This narrative study, set within a social constructivist epistemological framework, enabled me to explore the experience of pain in CF through interviewing nine adults with CF. I analysed the interview transcripts using a method of narrative analysis by Gee (1991), which presents the findings in stanzas, as a poetic form that aims to preserve the voice of the narrator. Essential to research in pain and palliative care, focus on the person as a whole was maintained throughout the study. The findings were then interpreted through the lens of total pain theory. Through these findings I have come to understand total pain in CF as a complex of interrelated elements occurring within the whole person. It is through these narratives of emerging awareness, social legitimacy and invisibility that I came to appreciate the nuances and complexity of total pain in CF.

The emerging awareness narrative, occurring across participants' lives, was the gradual increasing awareness of what it means to have CF. A crescendo effect of increasing total pain and suffering occurred alongside the continuum of emerging awareness. This effect

was part of an overarching sense of movement or fluidity and continual change. Signals or key indicators described by participants along this narrative supported the notion of milestones described by Charmaz (1991). Discussion of these milestones was prominent in emerging awareness as participants referred back to key moments such as diagnosis, first hospitalisation, transplant, moving back home and other recollections of key moments. In sharing these milestones, participants told stories revealing their experiences with physical, emotional, social and spiritual pain, the domains of total pain. Descriptions of these milestones also revealed another key feature across participant stories, liminality, which was seen in the experience of living in between, between health and illness, living and dying. Liminality and milestones appeared to be closely related to uncertainty and fear, expressions of emotional and spiritual pain. Uncertainty and fear appeared to cause a great deal of distress for participants.

Total pain in CF permeated across all narratives. Despite participants not using the precise words “total pain”, total pain was at the centre of their stories. The typologies or domains of total pain (physical, social, emotional and spiritual), were instead labelled as either physical pain, emotional pain or suffering by participants. For participants, total pain was experienced by their whole person in the larger context of living with illness, rather than as a distinct experience. Taken as a whole, the experience of total pain and the experience of suffering were indistinguishable from one another. This understanding moves away from E. Cassell (1982) understanding of pain as happening to bodies and suffering as happening to persons, an understanding perhaps better placed in the context of physical pain than total pain. Instead, my understanding of total pain and suffering in CF sees them

as a singular experience happening to a whole person, encapsulating their very being. This understanding is one of the key contributions to knowledge arising out of this thesis.

In an unexpected finding and another key contribution to knowledge stemming from this study, social legitimacy held a critical role in the experience of total pain in CF. Social legitimacy appears in the stories as a struggle to maintain independence, as participants tried to meet societal expectations. As participants became sicker and were no longer able to maintain these social expectations, the changes they experienced such as in social role, job, relationships and independence were experienced as losses. Echoing previous knowledge, the stories reflected values in western society of hard work, productivity, accomplishment, financial success and independence, and participants shared how difficult these were to achieve or maintain when living with a serious illness (Hay, 2010). Total pain was ever-present in these stories, and when participants were isolated, marginalised and lost social legitimacy they felt invisible.

Nowhere was total pain and the impact of living with CF more obvious than when participants felt invisible. This is another key contribution to knowledge in this thesis. It was within these stories of invisibility that the true depth of pain and suffering was revealed. An iceberg was a useful analogy to visualise this phenomenon, as participants revealed in their stories that much more was going on under the surface than was known even to those closest to them. This recognition of the loss of social legitimacy related to total pain and suffering, and eventually leaving participants feeling invisible, is key to understanding the experience of pain in CF.

A narrative approach to this study provided an opportunity for participants to tell their individual stories. In this unstructured style of interview, participants chose which stories to tell and which to keep within. In other words, this was *their* story, told in *their* way, albeit ultimately a product of interaction between interviewer and participant. This approach helped maintain focus on the whole person while exploring the experience of pain in CF, striving to study pain not in isolation but in the context of living with CF.

7.2 Original contribution to knowledge

This thesis presents original empirical research into the experience of pain in CF, as examined through the lens of total pain. In seeking to examine pain in the context of the whole person living with CF, the thesis draws together knowledge in an original way from the fields of palliative care, chronic illness, pain, and sociology. Drawing from these varied perspectives enabled identification and exploration of the closely related concepts of pain, suffering, living with serious illness, social legitimacy and invisibility in a way that has not been previously described in CF.

This study offers unique insight into how pain is experienced along a continuum of emerging awareness, as people gradually become aware of what having CF means in their lives. Through examining total pain along this continuum, pain is visualised as an evolution, as a movement or process, rather than an event. This enhances understanding of total pain as a dynamic process a concept recently described by Clark (2018b).

A key contribution to knowledge is visualisation of total pain and suffering in CF as a singular experience. This is in direct contrast to the depiction of pain happening to bodies and suffering to persons, possibly because E. Cassell (1982) may have been speaking primarily of physical pain, as opposed to total pain. Through the narrative design, participants told the stories of pain they felt were important, and through these stories, revealed pain and suffering to be a singular experience. Telling these stories in their own words also illuminated their shared language, as all participants told stories of total pain, but the words used to describe various domains were almost always emotional pain and suffering.

Another important contribution of the thesis in understanding the concept of pain in CF is the revelation that when participants experience total pain, it threatens social legitimacy and leads to invisibility. Invisibility, both resulting from and causing pain, was where the depth of pain in CF was most evident. In examining the role of social legitimacy within the experience of pain, the concept of total pain was considered in relationship to societal dimensions rather than as only situated within the individual.

Demonstrating that total pain is applicable in CF, the study provides evidence that total pain is generalizable not only in CF but across other non-cancer illnesses as well. Finally, while total pain theory was developed more than five decades ago, this study shows that it is still as relevant today as when Saunders introduced it in the 1960's.

In summary, this study is a move forward in deepening understanding of pain in CF. Through adoption of total pain as a theoretical lens, discourse has been taken beyond

primarily physical pain, where there is a tendency to view pain as a symptom for clinicians to manage, to a broader understanding of pain as a phenomenon to engage with more existentially.

7.3 Implications for clinical practice

This thesis supports the importance for healthcare practitioners to see the patient as a whole person and in relation to their broader social world. Knowledge gained through this study leads to several practical recommendations that can be operationalised. Other recommendations require more of a philosophical shift in how clinicians practice and view the role of healthcare.

The close relationship of social legitimacy and invisibility with total pain and suffering suggests that paying attention when clinicians hear these may yield greater understanding of pain in CF. People may speak of challenges in their various roles such as parents, partners, friends, students, employees or even patients. Talk of embarrassment, judgement, feeling misunderstood, change or loss may be heard in conversations, as they were heard from participants in this study. These are often invitations to explore if attention is paid to hear them. Recognising stories of invisibility may be especially important, as it seemed associated with the deepest pain.

All participants spoke of milestones as important markers of change, and attention to these milestones may help busy clinicians recognise when a person might benefit from additional support. Attending to these milestones echoes previous recommendations to

recognise certain events as potential “triggers” for the need for palliative care in CF (Dellon et al., 2016). Similarly, laughter seemed to be paired with the most painful experiences and therefore may serve as an indicator for pain or a cue to explore further.

Providing opportunities for people with CF to share their stories may provide insight into the experience of pain. Listening to the stories people tell, acknowledging and honouring their meaning, and acting on these stories is the hallmark of narrative medicine, a practice advocated by Charon (2001). Particularly relevant to findings coming out of this thesis is the assertion of Charon (2017) that through narrative medicine clinicians are able to see suffering and, as understood in this thesis, total pain. Carvalho et al. (2018, p. 970; 972) argued, from an ethical perspective, that narrative medicine is not only a “logical pathway” for addressing pain, but an ethical imperative, part of a “comprehensive paradigm of pain management that recognises the complexity of pain as a ‘total’ phenomenon and experience”. A closer look at narrative medicine may provide interested clinicians support in ways to develop their narrative competence.

Embracing care of the whole person need not entail dissolution of the team approach, indeed the team approach is a valuable component of CF care. It is important to recognise, however, that the relationship between a person with CF (or any illness) and their doctor is often regarded as sacred. The challenge is to maintain focus on the whole person living with illness within the context of the team approach. Subtleties in approach matter. Referring to a social worker or psychologist at the first mention of emotional pain may unintentionally imply *this is not my department or I only care about your lungs*, even

though unintended. The ability to hear suffering, to sit with it, even if only for a few moments before bringing in another team member, shows that the person is seen as a whole, complex being.

To embrace these recommendations may mean a shift for some in how they view the key role of healthcare. This shift, away from an understanding of healthcare as only concerned with the physical, towards healthcare as concerned with the full experience of illness, means caring for the whole person, including the complexity of total pain and suffering (Kearney, 2000).

Accepting this broader view of healthcare requires accepting a role in addressing total pain and suffering. Participant Ashley, who “laid in bed and suffered” because “there’s not anything they can do”, may have been right if the only treatment considered was a pill, procedure, or whatever the care map says to do. However, in this broader view of healthcare there is more to be done, there is a place to listen, room for silence, and to simply be with a person, to accompany them. With much of the focus in healthcare on *fixing*, it may be difficult to appreciate the value in simply listening or *being with* someone in silence. It is less about what to do and more about how to do it, with what Frank called “generous medicine” (A. W. Frank, 2004; A. W. Frank, 2008). In the foreword for the book *Mortally Wounded* by Kearney (1996, pp. 11-12), Saunders described this as “the way care is given can reach the most hidden places”, and A. W. Frank (2005, p. 296) said this is “a way of doing things – a spirit of practice – and many of those things have to be done

anyway". With this in mind, the recent trend towards personalised medicine to treat disease may be just as fitting to treat the whole person living with serious illness.

7.4 Limitations of the study

There are several limitations to this study which should be acknowledged. Some critics of narrative research argue that the participant's voice can be lost in the act of retelling the story (Polkinghorne, 2007). While it should be acknowledged that the narrative is created in the interaction between participant and researcher, the decision to represent the stories in a poetic style aims to better preserve the essence of the participants' stories. Attention to body language, verbal utterances, silence, and expressions of emotion such as laughter also help to preserve the voice of the participant, as did cross-checking the transcripts and stanzas with participants as described in Chapter Five.

Another potential limitation is that all of the participants attended the same accredited CF centre and experiences from other centres may have been different. In recognition of this concern, recruitment included people who were new to the program and these efforts yielded two participants who had only recently transferred to the centre. These were also the only two participants who had not met me prior to the day of the interview. Strategies to address potential issues for "insider research" or power imbalances were described in Chapter Five.

There was also a possibility of bias, as the researcher has been working within the specialty of CF for more than ten years and has been integrally involved in efforts to

advance palliative care in CF. This experience and interest may have led the researcher to pay more attention to some parts of stories than others and may have affected the selection of which stories to include in the thesis narratives. Some participants were likely aware of the researcher's interest in these areas and this may have affected participant choices of which stories to tell and which to keep silent.

7.5 Future research

During the literature review phase of this study, almost all published studies about pain in CF were quantitative. While this study provides insight into the experience of pain in CF, more qualitative research is needed. The scope of the narratives that participants shared about their experiences with pain in CF and the nuances contained within these stories suggest that pain in CF is best explored through methods that focus on the whole person and can support this degree of complexity. Using other qualitative methods in addition to narrative may also provide further insight. A similar study to learn more about the experience of people with CF that regularly take pain medication (prescribed or illicit) would also be insightful.

Recognising the strong biomedical positioning of the field in CF, a mixed method study may be best readily received by CF specialists and findings from this study could be used to inform study development. I advocate, however, that mixed method research not replace qualitative study but rather may be considered in addition as an effort to build a comprehensive knowledge base on pain in CF.

As mentioned in Chapter Four, total pain seems to have been either operationalised in individual case studies or used as a recommendation or discussion point in a theoretical discussion rather than the research process itself. This study applied total pain theory on a slightly larger scale (nine participants) and application on a larger scale may be beneficial. Researchers will need to consider the challenge of applying total pain on a larger scale with the importance of studying in the whole person. Carrying forward with a contemporary view of total pain, future research should examine total pain from the perspective of a dynamic, evolving experience with dimensions beyond the individual (Clark, 2018b).

Invisibility held a powerful place in participant narratives and seemed to both come out of, and cause, suffering. The challenge with invisibility, however, is that it is by nature invisible. This study is an indication that if people with CF are provided an opportunity to share their experiences, and those around them value and listen to them, there is an opportunity through these stories to advance knowledge further in this area of research.

Finally, the findings in this study may cause some degree of concern for CF clinicians who understandably wonder how they can possibly address this magnitude of scope within an allotted visit, even in the context of multidisciplinary care. This is an important concern to anticipate and should be included in future research.

7.6 Concluding remarks

At the end of this thesis, I reflect again on the events that culminated in my research about pain in CF. Rounding out this string of events was someone asking if others with CF also have pain every day, so this project has been influenced by the voices of those who live with CF from the start and for these voices I am grateful.

Reflecting on my status as an insider, in these final words of the thesis, I was almost hyper aware of my positioning as an insider and took seriously the responsibility to conduct ethical research, as noted in Chapter Five (5.4.4). While I spent a considerable amount of time thinking about risks and cautions in conducting insider research, there are strengths as well. In many cases, participants themselves independently highlighted the strengths, both before and after the interviews. Importantly, they stated that because they had known me for long time, they trusted me and felt safe sharing their stories. This was demonstrated in the depth of sharing by participants, especially evident in the rich stories of their experiences. From a practical standpoint, insider status also helped facilitate contact with participants for cross checking findings.

I have been honoured that those I spoke with so generously shared their stories and time with me. I have been taken aback by how little I knew about their experiences before taking the time to listen to their stories and surprised by the emotional and at times tearful gratitude expressed to me simply because I asked about their pain. This unexpected gratitude of participants reminds me of Saunders, who said, "Often we are merely called on to show that we are trying to understand, even though we have few

answers to give and no comfort that can take away the pain we stay alongside” (Saunders, 1990, p. 2). I do hope that this research has provided a few more answers about pain in CF, although I suspect it may have raised as many questions as answers.

In closing, I cite Saunders (2000, p. 9) account of her patient Mrs Hinson, who captured the essence of total pain when she said to Saunders, “It seems that all of me is wrong”. The narratives participants shared through this study illuminate the experience of total pain in those living with CF. Like Mrs Hinson many years ago, Brittany captured the essence of total pain in CF when she exclaimed, “It’s all just suffering” as described in Chapter Six. In this study, total pain, captured so clearly by Mrs Hinson, seems as relevant in CF today as it was in terminal cancer 55 years ago. The challenge now is to move forward with the new knowledge from this study in a way that meaningfully informs clinical care and research in CF and other serious illness. As I look towards the next phase of my career, I see this as an important and exciting challenge.

Appendix A: Database Summary

Database	Search Terms	References	Comments
AMED	For AMED database, searched with AB "cystic fibrosis" AND AB pain.	6	
<u>CINAHL</u>	Using "CINAHL headings" searched for cystic fibrosis and pain as major concept.	12	
<u>PsycINFO</u>	For PsycINFO database, searched using AB "cystic fibrosis" AND AB pain.	23	
SCOPUS	"cystic fibrosis" AND pain in abstract.	411	
<u>Web of Science</u>	Used "cystic fibrosis" AND pain	509	Web of science does not have a thesaurus. Did not limit search to title as decision made that relevant articles might be missed; therefore, search is done by topic (search in title, abstract is not available in WOS).
<u>Pubmed</u>	Searched ("cystic fibrosis"[Mesh]) AND (((("Pain Measurement"[Mesh]) OR "pain Management"[Mesh]) OR "Pain"{Mesh] filters: Humans.	147	
<u>Total References</u>		Total References: 1108	

Appendix B: Data Extraction Table

Author(s) Year Country Title	Design	Participants	Data Collection/Outcome Measures	Key Findings	Limitations	Quality Score
Allgood, SJ et al. 2018 US Descriptions of the pain experience in adults and adolescents with cystic fibrosis	Qualitative Exploratory descriptive design	n=10 5 females; 5 males Population: adolescent and adult Age: no mean or median available, range 13-46 years FEV1: 23.5%-102% Site: single centre	Brief Pain Inventory (BPI) Semi-structured interviews	Differences in pain experience for adolescents and adults. Pain adversely affects all aspects of life. Adolescents report decreased socialization due to pain. Adults report more emotional pain than adolescents. While adolescents feel supported by clinical team, adults feel unsupported.	No mention of theory	31
Blackwell, L S and Quittner A L 2015 US Daily pain in adolescents with CF: Effects on adherence, psychological symptoms, and health-related quality of life	Quantitative Observational Prospective Retrospective Questionnaire s	n=95 59 females; 36 males Population: adolescent Age: mean 15.8 years FEV1: mean 80% Site: 6 centres	Initial visit followed by 6-day online diary Demographics and Medical characteristics <u>Prospective</u> Daily Pain Assessment Questionnaire for Cystic Fibrosis (DPAQ-CF) <u>Retrospective</u> Hospital Anxiety and Depression Scale (HADS) Medication Possession Ratio (MPR) Cystic Fibrosis Questionnaire Revised (CFQR-R)	74.5 % participants reported pain, average 2.1 out of 10 (mild). Pain associated with decreased adherence to CF treatment regimen, decreased HRQOL, and increased psychological distress.	Limitations average 4.34 diaries completed (out of 6) Small sample size HRQOL, CFQR-R, HADS only completed on day 1	30
Epker, J A, et al. 1999 US Pain and pain-related impairment in adults with cystic fibrosis	Quantitative Observational Retrospective Questionnaire s	n=75 37 females; 38 males Population: adults Age: mean 26.8 years FEV1: not reported Site: single centre	MPI (Multidimensional Pain Inventory) Shwachman index "Nurse perception scale"	65.7 % reported mild pain. Significant relationship between age and pain related interference, did not find significant relationship between disease severity and pain, nurses rated patients pain higher than patients rated themselves.	Only included outpatients so may not be as sick. Only included 2 nurses. No correlation with FEV1. Minimal demographic information provided.	23
Festini, FS, et al. 2004 Italy Prevalence of pain in adults with cystic fibrosis	Quantitative Observational Questionnaire Retrospective	n=239 125 females; 114 males Population: adults Age: mean 26.1 years Median FEV1: 56.71% Site: 15 centres	Demographics and medical information 6 item questionnaire developed for this study	Study reported high incidence of pain for intensity and frequency. 59.8 % report pain as adversely affecting life. Only 42.6% participants discussed with CF clinical team.	Tool not validated or pilot tested	27

Flume, PA et al. 2009 US Patient reported pain and impaired sleep quality in adult patients with cystic fibrosis	Quantitative Observational Retrospective Questionnaire	n=50 23 females; 27 males Population: adults Age: median 31.1 years FEV1: mean 58.4% Site: single centre	Brief Pain Inventory (BPI) Pittsburgh Sleep Quality Index Demographics	More than half participants reported recent pain and issues with sleep quality. Pain was associated with decreased sleep quality.		29
Hayes, M et al. 2011 US Pain is a common problem affecting clinical outcomes in adults with cystic fibrosis	Quantitative Observational Online survey Retrospective	n=83 47 females; 36 males Population: adults Age: median 29.3 years FEV1: median 63.6% Site: single centre	Brief Pain Inventory (BPI) CFQ-R Hospital Anxiety and Depression Scale Pain Catastrophizing Scale	82% participants reported pain in last month. Pain negatively impacted general activities, mood and work. Pain associated with decreased quality of life, depression and anxiety. Pain associated with increased pulmonary exacerbations and death.	Selection bias (61% response rate), single centre, recall bias, BPI not validated in CF.	29
Hubbard, PA et al. 2005 US Pain, coping, and disability in adolescents and adults with cystic fibrosis: A web-based study	Quantitative Observational Retrospective Questionnaire via web	n=18 12 females; 6 males Population: adults Age: 67% older than 23 years FEV1-not reported Site: all US centres	Demographics Pain Disability Index (PDI) Pain response inventory (adapted)	Pain reported to be common with about 50% participants reporting daily pain. Pain adversely affected recreational activities, work and social life. Common coping strategies were both active and accommodative.	Very low participation Posted info to all CF centres in US	24
Kelemen, LA, et al. 2012 Australia Pain impacts on quality of life and interferes with treatment in adults with cystic fibrosis	Quantitative Observational Prospective study with cohort of 33 patients with acute exacerbation	n=73 35 females; 42 males Population: adults Age: mean 30 years FEV1: 60.5% (unknown whether mean or median) Site: single centre	CFQ Brief Pain Inventory (BPI) Pain Catastrophizing Scale (PCS) Body outline Demographics	Pain was not associated with severity disease severity but did adversely affect treatment adherence. Mild pain reported by 89% stable participants.	Recall bias Only patients who attended CF clinic were eligible	27
Koh, JL et al. 2005 US Assessment of acute and chronic pain symptoms in children with cystic fibrosis	Quantitative Observational Retrospective Questionnaire s	n=46 24 females; 22 males Population: children Age: mean 12.9 years (8-17) FEV1: mean 80% Site: 3 centres	FACES pain intensity scale Self-report questionnaire (homegrown, not validated) McGrath tool (measures degree pain caused emotional upset) VAS for perceived functional limitation Body outline for location Demographics	46% of children reported pain at least weekly, usually mild. 11% reported moderate intensity of pain. Chest pain associated with increased disease severity. 70% children reported pain caused emotional upset.	Outpatient only Retrospective for last 4 weeks may not be reliable in younger children	29
Munck, AA et al. 2012 France Recurrent abdominal pain in children with cystic fibrosis: a pilot prospective	Quantitative Prospective Observational	n=8 2 females; 6 males Population: children Age: mean 13.6 years FEV1: mean 85% Site: single centre	Eland Pain location Faces Pain Scale-Revised (FPS-R) Mc Gill emotional status R-CMAS anxiety score Health-related quality of life (CF-QOL) A 28-day daily pain diary	Reported low prevalence (6%) of abdominal pain in children with CF.	2 of 8 participants withdrew from the study Descriptive results only due to low sample size	23

longitudinal evaluation of characteristics and management						
Palermo, TM et al. 2006 US Effect of disease-related pain on the health-related quality of life of children and adolescents with cystic fibrosis	Quantitative Observational Retrospective	n=46 24 females; 22 males Population: children and adolescents Age: mean 12.9 years (8.1-17.8) FEV1: mean 80.2% Site: single centre	CFQ-R Retrospective pain interview Faces pain scale Body outline Demographics	Pain reported to negatively impact quality of life in physical functioning, role functioning, CF treatment regimen, and physical symptoms.	Recall bias (especially in younger children) Small population Single centre	31
Ravilly, S et al. 1996 US Chronic pain in cystic fibrosis	Retrospective Chart review	n=78 (58 deceased, 23 alive) 25 females; 30 males (deceased) 11 females; 12 males (alive) Population: adults Age: mean 26 years (deceased); mean 23 years (alive) FEV1: mean 58% Site: single centre	No tool mentioned or referenced	Pain increased in last 6 months of life. Headaches and chest pain were most common. 53% received opioids for pain in last 6 months of life.	Only descriptive statistics used. Defined pain episode as event requiring medical intervention and study based on chart review so based on other study results with less than ½ reporting to MD this may limit results significantly. Usual limitations of chart review, inconsistent documentation	23
Sermet-Gaudelus JP, et al. 2009 France Pain in children and adults with cystic fibrosis: a comparative study	Quantitative Observational Retrospective	n=73 children; 110 adults 36 females; 37 males (children) 62 females; 48 males (adults) Population: children and adults Age: mean 10.2 years (children, 1-18); mean 28.5 years (adults, 18-52) FEV1: mean 70% (children); 50% (adults) Site: 2 centres	Ad hoc questionnaire Body outline	Children and adults experienced similar rate of pain but adults reported increased intensity and duration of pain than did children. Pain associated with decreased physical activity. Mean intensity of pain was moderate. Authors reported pain as common and undertreated.	Study is retrospective only, parent reported pain in children <8 years, QOL not fully explored,	28
Stenekes, SM et al. 2009 Canada Frequency and self-management of pain, dyspnea, and cough in cystic fibrosis	Quantitative Observational Retrospective	n=64 children; 59 adults 71 females; 52 males Population: children and adults Age: mean 19.9 years (7-60) FEV1: 40% with FEV1>81% 29% FEV1 61-80% 22% FEV1 41-60% 4% FEV1 <40 5% UNK Site: 3 centres	Demographics Questionnaire (ad hoc) Dalhousie Dyspnea Scale (DDS)	84% participants reported pain, head and abdomen most frequent locations of pain. 64% reported dyspnea, 83% cough.	Recall bias, response rate 64%,	21

Appendix C: Quality Appraisal Tool

Quality Appraisal Key (Hawker et al. 2002)

1. Abstract and title: Did they provide a clear description of the study?

Good: Structured abstract with full information and clear title.
Fair: Abstract with most of the information.
Poor: Inadequate abstract.
Very poor: No abstract.

2. Introduction and aims: Was there a good background and clear statement of the aims of the research?

Good: Full but concise background to discussion/study containing up-to-date literature review and highlighting gaps in knowledge.
Clear statement of aim and objectives including research questions.
Fair: Some background and literature review.
Research questions outlined.
Poor: Some background but no aim/objectives/questions, OR
Aims/objectives but inadequate background.
Very poor: No mention of aims/objectives.
No background or literature review.

3. Method and data: Is the method appropriate and clearly explained?

Good: Method is appropriate and described clearly (e.g., questionnaires included).
Clear details of the data collection and recording.
Fair: Method appropriate, description could be better. Data described.
Poor: Questionable whether method is appropriate. Method described inadequately.
Little description of data.
Very poor: No mention of method, AND/OR Method inappropriate, AND/OR No details of data.

4. Sampling: Was the sampling strategy appropriate to address the aims?

Good: Details (age/gender/race/context) of who was studied and how they were recruited. Why this group was targeted. The sample size was justified for the study. Response rates shown and explained.
Fair: Sample size justified. Most information given, but some missing.
Poor: Sampling mentioned but few descriptive details.
Very poor: No details of sample.

Quality Appraisal Key (Hawker et al. 2002)

5. Data analysis: Was the description of the data analysis sufficiently rigorous?

- Good: Clear description of how analysis was done.
Qualitative studies: Description of how themes derived/
respondent validation or triangulation.
Quantitative studies: Reasons for tests selected hypothesis driven/
numbers add up/statistical significance discussed.
- Fair: Qualitative: Descriptive discussion of analysis.
Quantitative.
- Poor: Minimal details about analysis.
- Very poor: No discussion of analysis.

6. Ethics and Bias: Have ethical issues been addressed, and has necessary ethics approval been gained? Has the relationship between researchers and participants been adequately considered?

- Good: Ethics: Where necessary issues of confidentiality, sensitivity, and consent were addressed.
Bias: Researcher was reflexive and/or aware of own bias.
- Fair: Lip service was paid to above (i.e., these issues were acknowledged).
- Poor: Brief mention of issues.
- Very poor: No mention of issues.

7. Results: Is there a clear statement of the findings?

- Good: Findings explicit, easy to understand, and in logical progression. Tables, if present, are explained in text. Results relate directly to aims. Sufficient data are presented to support findings.
- Fair: Findings mentioned but more explanation could be given. Data presented relate directly to results.
- Poor: Findings presented haphazardly, not explained, and do not progress logically from results.
- Very poor: Findings not mentioned or do not relate to aims.

8. Transferability or generalizability: Are the findings of this study transferable (generalizable) to a wider population?

- Good: Context and setting of the study is described sufficiently to allow comparison with other contexts and settings, plus high score in Question 4 (sampling).
- Fair: Some context and setting described, but more needed to replicate or compare the study with others, PLUS fair score or higher in Question 4.

Quality Appraisal Key (Hawker et al. 2002)

Poor: Minimal description of context/setting.
Very Poor: No description of context/setting.

9. Implications and usefulness: How important are these findings to policy and practice?

Good: Contributes something new and/or different in terms of understanding/insight or perspective.
Suggests ideas for further research
Suggests implications for policy and practice
Fair: Two of the above (state what is missing in the comments).
Poor: Only one of the above.
Very poor: None of the above.

Adapted from Hawker et al. 2002

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Appendix D: Inclusion and Exclusion Criteria (Literature Review)

Articles for Inclusion	Articles for Exclusion
Focus on participants of all ages with CF	Focus on multiple diagnoses where evidence specific to CF is not disaggregated
Focus on experience of pain in CF	Empirical studies that fail to address topic (pain in CF is mentioned but not the primary focus of the article, primary focus on specific intervention (i.e. acupuncture) or diagnostic criteria rather than the experience of pain), pharmacological randomised controlled trials (RCTs).
Published in English	Non-English studies
Empirical papers reporting primary research	Conference abstracts, opinion or editorial papers, reviews, letters.

Appendix E: Interview Guide

Interview Topic Guide for Unstructured Interviews

- **Researcher introduction of self and study**
- **Reminder re: confidentiality**
- **Reminder regarding right to decline question or end interview**
- **Thank you for participating**
- **Ask verbal permission to record and take notes**
- **Initial questions to establish comfort, help the participant to be more comfortable**
 - o Please tell me about yourself
 - o What has your day been like so far today?
 - o How would you describe your CF?
- **Possible topics to explore**
 - o Earliest pain experience
 - o Locations/types
 - o Frequency
 - o Addiction/Tolerance
 - o Pain disbelief
 - o Barriers to relief
 - o Beliefs about cause
 - o Effect on QOL
 - o Daily activities
 - o Reporting pain/seeking assistance/communicating about pain
 - o Complementary therapy
 - o Medications
 - o Exacerbations
 - o Adherence
 - o Guilt

- o Treatment regimen
- o Airway clearance therapy
- o Sleep
- o Self-management
- o Mood/Anxiety/Depression
- o CF team
- **Strategies for researcher**
 - o LISTEN
 - o Open-ended questions
 - o Ask questions about what they have said
 - o Summarize/Echo back, “What I hear you saying is...”
 - o Silence is okay
 - o Ask participant to “Tell me more” or “Then what happened?”
 - o Try to allow participant to tell their story in their own way

Appendix F: Ethics Approval Letters

150745



UNIVERSITY OF CALIFORNIA, SAN DIEGO HUMAN RESEARCH PROTECTIONS PROGRAM

TO: Dr. Jessica Goggin
RE: Project #150745
Experience of Pain in Cystic Fibrosis: A Narrative Study

Dear Dr. Goggin:

The above-referenced project was reviewed and approved by one of this institution's Institutional Review Boards in accordance with the requirements of the Code of Federal Regulations on the Protection of Human Subjects (45 CFR 46 and 21 CFR 50 and 56), including its relevant Subparts. This approval, based on the degree of risk, is for 365 days from the date of **IRB review and approval** unless otherwise stated in this letter. The regulations require that continuing review be conducted on or before the 1-year anniversary date of the IRB approval, even though the research activity may not begin until sometime after the IRB has given approval.

The IRB determined that this project presents no more than minimal risk to human subjects in that the probability and magnitude of harm or discomfort anticipated in the research are not greater in and of themselves than those ordinarily encountered in daily life or during the performance of routine physical or psychological examinations or tests.

Date of IRB review and approval: 5/28/2015

On behalf of the UCSD Institutional Review Boards,

A handwritten signature in black ink that reads "A Magit".

/mb

Anthony Magit, M.D.
Director
UCSD Human Research Protections Program
(858) 657-5100; hrpp@ucsd.edu

Note: IRB approval does not constitute funding or other institutional required approvals. Should your studies involve other review committees such as Office of Clinical Trials Administration (OCTA), Office of Coverage Analysis Administration (OCAA), Conflict of Interest (COI), Protocol Review Monitoring Committee (PRMC), and committees under Environmental Health & Safety (EH&S) such as Institutional Biosafety Committee (IBC), Human Exposure Committee (HERC), and RSSC (Radiation Safety and Surveillance Committee), it is the researchers responsibility to ensure that all approvals are in place prior to conducting research involving human subjects or their related specimens.

Approval release date: 7/1/2015

UCSD HUMAN RESEARCH PROTECTIONS PROGRAM

GENERAL APPROVAL INFORMATION

The information below does not encompass all human subjects protections requirements, however, is intended to highlight those of significance to ensure awareness by researchers engaged in research involving human subjects or their related specimens and data.

Approval Letters and Consent Documents

Unless otherwise stated, approval letters will be accompanied by stamped, approved consents. Should a study be closed to accrual and no consent released as a result, this information will be documented on the approval letter. Also, any waivers will be documented in the approval letter (such as waiver of documented consent or waiver of authorization for use of PHI).

The PI must ensure approval is in place from other appropriate review boards (such as Radiation Safety, Institutional Biosafety Committee, Conflict of Interest, ESCRO, etc.)

If other institutions are involved, the PI must ensure that IRB approvals (or other administrative approvals) from those sites are secured and forwarded for the study file. In addition, PI's must ensure that the clinical trial agreement, as applicable, or other funding (such as a grant) is appropriately in place prior to conducting any research activities. IRB approval does not constitute funding approval.

Duration of IRB approval

The IRB may grant approval up to 365 days. (See 45 CFR 46.109(d) (DHHS) and 21 CFR 56.109(d) (FDA)). However, for some studies the IRB may grant approval for a lesser period or a specific number of subjects to allow for more frequent monitoring. The approval letter or related documentation will indicate this information.

Because IRB review of research studies must be completed at least annually, investigators should plan ahead to meet required continuing review dates. **Please submit complete continuing review documentation at least 45 days prior to the expiration date to guard against a lapse in IRB approval**. The signed continuing review ~~documents~~ facepages and any other required hard copies must be received by the HRPP office before the continuing review process can begin.

As a courtesy, automated continuing review reminders can be set-up by PIs at various intervals (75 days, 45 days, 30 days, for example) on the website at <https://irb.ucsd.edu>. However, as these are automated electronic messages based on data entered, and the HRPP cannot anticipate which type of software programs (such as spam-blockers or anti-virus software) may block receipt of the messages, **PI's are required to not rely upon notification, but have internal mechanisms which track continuing review submission times**. Ultimately, it is the PI's responsibility to initiate a continuing review application, allowing sufficient time for the review and re-approval process to be completed before the current approval expires.

Continuing review is required even if no changes are made, or if the only study activity is participant follow-up, and even if the only study activity is data analysis.

What happens if there is a lapse in IRB approval?

If the IRB has not reviewed and approved a research study by the study expiration date, **all research activities must stop**. This includes the following:

All research-related interventions or interactions with currently enrolled subjects (unless the IRB finds that it is in the best interests of the individual subjects to continue participating in the research interventions or interactions;*) recruitment and informed consent procedures; and continued collection and/or analysis of data/information.

**Exception:* Research-related interventions or interactions with enrolled subjects may continue if the IRB determines that stopping the research would jeopardize the rights or welfare of current subjects. The IRB will decide which subjects should continue receiving the intervention during the lapse in approval. A request for such an exception must be submitted in writing to the attention of the IRB Chair by the Principal Investigator. If any project activity—even activity required for participant safety—occurs or continues after the expiration date, the investigator is out of compliance with both federal regulations and university policy. Retrospective approval for work done after the expiration date cannot be granted.

Amendment/revision to an IRB approved study

IRB approval is required before implementing any changes in the approved research plan, consent documents, recruitment materials, or other study-related documents. Please see Amendment Fact Sheet at <http://irb.ucsd.edu/amendmodchg.pdf> for submission guidance.

Adverse Event and Unanticipated Problems Reporting

All problems having to do with subject safety must be reported to the IRB within ten working days. All deaths, whether or not they are directly related to study procedures, must be reported. For adverse events, please utilize the form found at https://irb.ucsd.edu/UPR_biomedical.doc. For deviations and other reports, a cover letter and any supplemental information appropriate to the review should be provided. Please see IRB Guidelines for more information at <https://irb.ucsd.edu>.

Changes in financial Interest or Conflict of Interest (COI) disclosure

Any changes in the financial relationship between the study sponsor and any of the investigators on the study and/or any new potential conflicts of interest must be reported immediately to the Independent Review Committee via the Conflict of Interest Office. If these changes affect the conduct of the study or result in a change in the required wording of the approved consent form, then these changes must also be submitted as an amendment request.

Applicant: Jessica Goggin
Supervisors: Amanda Bingley and Sarah Brearley
Department: Health Research
FHMREC Reference: FHMREC15138

12 September 2016

Dear Jessica,

Re: Experience of Pain in Cystic Fibrosis: A Narrative Study

Thank you for submitting your research ethics application for the above project for review by the **Faculty of Health and Medicine Research Ethics Committee (FHMREC)**. The application was recommended for approval by FHMREC, and on behalf of the Chair of the Committee, I can confirm that approval has been granted for this research project.

As principal investigator your responsibilities include:

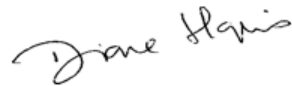
- ensuring that (where applicable) all the necessary legal and regulatory requirements in order to conduct the research are met, and the necessary licenses and approvals have been obtained;
- reporting any ethics-related issues that occur during the course of the research or arising from the research to the Research Ethics Officer at the email address below (e.g. unforeseen ethical issues, complaints about the conduct of the research, adverse reactions such as extreme distress);
- submitting details of proposed substantive amendments to the protocol to the Research Ethics Officer for approval.

Please contact me if you have any queries or require further information.

Tel:- 01542 592838

Email:- fhmresearchsupport@lancaster.ac.uk

Yours sincerely,



Dr Diane Hopkins
Research Integrity and Governance Officer, Secretary to FHMREC.

Appendix G: Informed Consent



#150745

University of California, San Diego
Consent to Act as a Research Subject

The Experience of Pain in Adults with Cystic Fibrosis: A Narrative Study

Who is conducting the study, why you have been asked to participate, how you were selected, and what is the approximate number of participants in the study?

Jessica Goggin is conducting a research study to find out more about pain in adults with cystic fibrosis. You have been asked to participate in this study because you have cystic fibrosis and self-identified as having experience with pain. There will be approximately 20 participants enrolled in this study.

Why is this study being done?

The purpose of this study is to explore and improve understanding about the experience of pain in adults with cystic fibrosis.

What will happen to you in this study and which procedures are standard of care and which are experimental?

If you agree to be in this study, the following will happen to you:

You will meet with the researcher, Jessica Goggin, in person or via an online connection such as Skype or Zoom (your preference). The interview location may be your home, another location mutually agreed upon by you and the researcher, or via an online connection. The interview will last about 1 to 1 ½ hours and will be audio-recorded. During the interview, the researcher will ask you about your experiences with pain in cystic fibrosis. You may decline to answer any question or stop the interview at any time.

How much time will each study procedure take, what is your total time commitment, and how long will the study last?

The interview will last approximately 1-1 ½ hours, which is your total time commitment.

The study is estimated to last approximately 2-3 years, from the planning phase to the final written reporting of results.

What risks are associated with this study?

Participation in this study may involve some added risks or discomforts. These include the following:

1. There is a risk of distress when telling your story about your experiences with pain in cystic fibrosis.
2. There is a risk of loss of confidentiality.

If you choose to participate in the interview via online connection such as Skype or Zoom, there is a risk of loss of confidentiality and confidentiality in online communications cannot be guaranteed. The researcher will take all reasonable measures to ensure confidentiality risks are addressed and you are encouraged to do the same, including:

1. Set up a new account via the selected online platform (Skype, Zoom, etc.) and delete the account when the interview is completed.
2. Connect to the online platform using a secure internet connection.
3. Close all other websites during the interview except for the selected online platform (Skype, Zoom, etc.)
4. Ensure use of a strong password for their online platform account (at least 8 characters, including upper and lower case and symbols).
5. Following interview, exit the online platform and shut down their computer.
6. Select a private location for the interview and minimize interruptions.

Because this is a research study, there may be some unknown risks that are currently unforeseeable. You will be informed of any significant new findings.

What are the alternatives to participating in this study?

The alternative to participation in this study is to not participate in this study.

What benefits can be reasonably expected?

There is no direct benefit in taking part in this study. The investigator(s), however, may learn more about the experience of pain in adults with cystic fibrosis.

Can you choose to not participate or withdraw from the study without penalty or loss of benefits?

Participation in research is entirely voluntary and you are free to withdraw at any time without giving any reason and without penalty or loss of benefits. However, once your data have been ~~anonymised~~ and incorporated into themes it might not be possible for it to be withdrawn, though every attempt will be made to extract your data up to the point of assessment of the thesis. If you wish to withdraw from the study please notify Jessica Goggin by telephone, in person during the interview, or by email.

Can you be withdrawn from the study without your consent?

You may be withdrawn from the study without your consent for the following reasons:

1. Your physician, Dr. Douglas Conrad believes that it is in your best medical interest.
2. You may also be withdrawn from the study if you do not follow the instructions given you by the study personnel.
- 3.

Will you be compensated for participating in this study?

There is no compensation for participating in this study.

Are there any costs associated with participating in this study?

There will be no cost to you for participating in this study

What if you are injured as a direct result of being in this study?

If you are injured as a direct result of participation in this research, the University of California will provide any medical care you need to treat those injuries. The University will not provide any other form of compensation to you if you are injured. You may call the Human Research

Protections Program Office at (858) 657-5100 for more information about this, to inquire about your rights as a research subject or to report research-related problems.

What about your confidentiality?

Research records will be kept confidential to the extent allowed by law. The following procedures are established to maintain your confidentiality:

1. A digital audio recorder will be used to record the interviews.
2. The interview will be transcribed by the researcher and all identifying data will be removed.
3. Hard copies of the transcript or other paper documents will be stored in a locked cabinet.
4. All study records maintained on the computer will be encrypted and stored on a password-protected computer.
5. At the end of the study, hard copies of the documents will be stored securely in a locked cabinet in a locked office for a maximum of 10 years. At the end of this period the documents will be destroyed.
6. Personal identifiers and contact details will be kept separately from ~~anonymised~~ data and stored securely in encrypted password protected files or hard copy in a locked cabinet. All personal identifiers will be destroyed once the thesis is assessed.

Research records may be reviewed by the UCSD Institutional Review Board.

Under California law, we must report information about known or reasonably suspected incidents of abuse or neglect of a child, dependent adult or elder including physical, sexual, emotional, and financial abuse or neglect. If any investigator has or is given such information, he or she may be required to report such information to the appropriate authorities.

Who can you call if you have questions?

Jessica Goggin has explained this study to you and answered your questions. If you have other questions or research-related problems, you may reach Jessica Goggin at 858-657-7073.

You may call the Human Research Protections Program Office at (858) 657-5100 to inquire about your rights as a research subject or to report research-related problems.

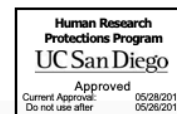
Your Signature and Consent

You have received a copy of this consent document and a copy of the “Experimental Subject’s Bill of Rights” to keep.

You agree to participate.

Subject’s signature

Date



VOLUNTEERS NEEDED FOR RESEARCH STUDY ON PAIN IN CYSTIC FIBROSIS

We are looking for volunteers to participate in an interview about pain in cystic fibrosis (CF). Participants must be at least 18 years of age, have CF, and have experience with pain (now or in the past). As a participant in this study, you will be asked to talk about your life history and experience of pain. The interview will take approximately 1-1 ½ hours for you complete.

If you are interested, please contact:

Jessica Goggin, MAS, RN

Principle Investigator

Telephone (858) 657-7073

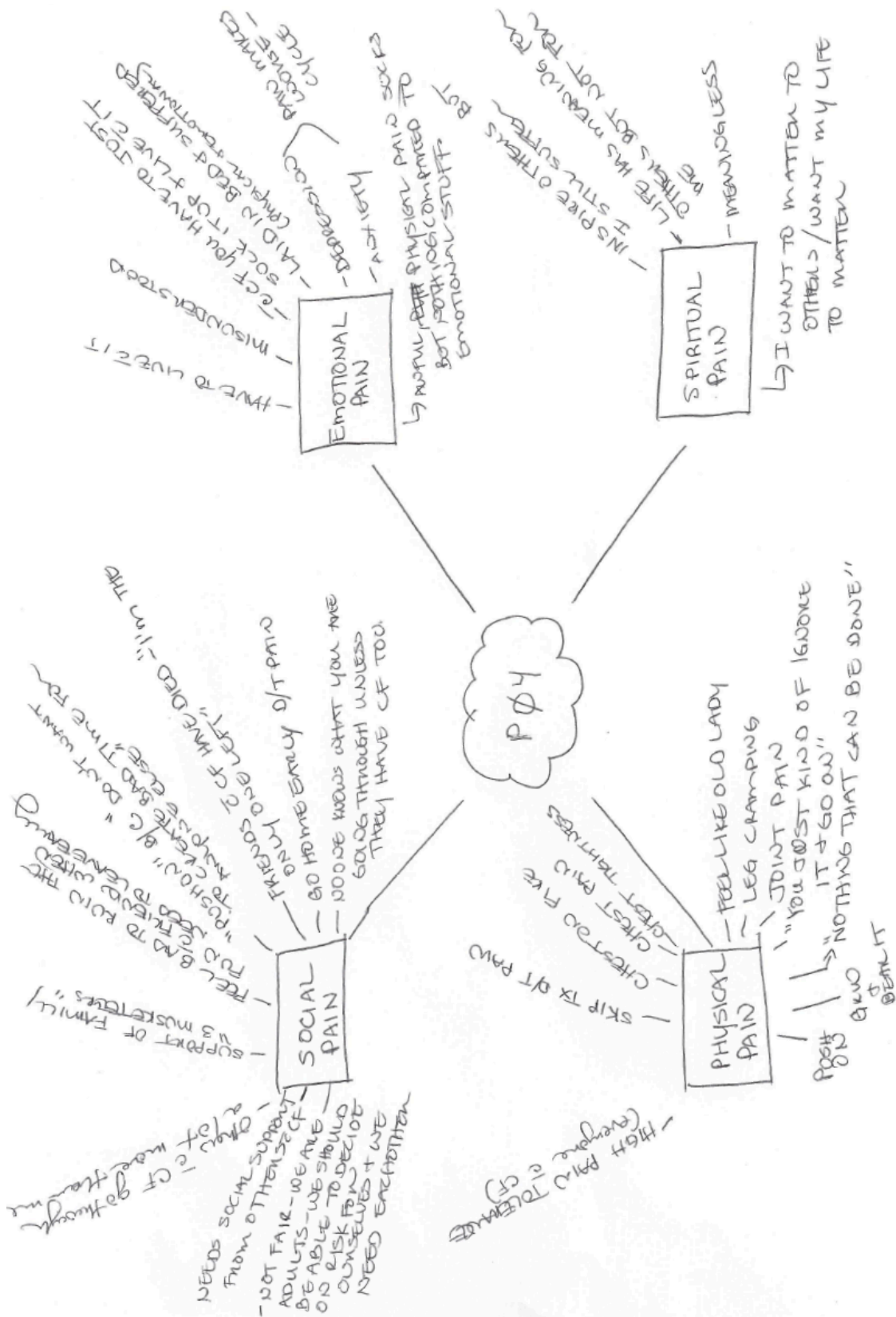
Email: jlkoggin@ucsd.edu

Thank you!

Appendix I: Example Initial Stage Analysis (Atlas.ti)

<p>JG Does your relationship help you in terms of coping and dealing with CF, is there any influence there at all?</p>	<p>P08 Yeah definitely-he-he helps me-I mean-I've gotten to the point where it's not hard for me to remember every little thing that I have to do so it's not like I have to be reminded-my routine is pretty on point-yeah-better than it's ever been-and I guess that like comes with age and learning it and doing it so much-but there's—I mean—the more I learn about people who have had CF-the more I learn that it's actually pretty common to just be so rebellious in your like uh young adult- and like teenage life-to act like you're not sick and you're fine and ignore it for a little while but uh-then you learn-laugh-that you better step up and fix it-so um-I've learned-I've been on a really good routine and I've just learned how to deal with me on a really regular basis-but um-there's little things like when I get sick anyway-you know-I'm doing everything I can and I still get a chest infection and even a cold you can't really prevent-um-and then that's gets me sick and he's just-um-he's really great with helping me and being there when I'm pissed off and upset and um-the last couple hospital stays I was in he was there-he stayed with me this whole two weeks-yeah-so he did that-one of them was here in (CURRENT TOWN) so he could have easily just went home and went back and forth and he slept in a stupid foldy chair-laugh-1 was-it was really great um-yeah-he's been amazing-helping me with it-mm hmm</p>	<p>13:15 P08 Yeah definitely-he-h...</p> <p>13:7 I've been on a...</p> <p>Emerging understanding of ha... unpredictability</p> <p>Pushing through</p> <p>emotional pain</p> <p>invisibility social pain</p>
<p>JG Is there anything I haven't asked you about that you think wow I'm really surprised she hasn't asked me this or anything that you would want to add or communicate about the experience with pain, with emotions, just having CF in general? Anything else that you feel you would like to add to what we've talked about?</p>	<p>P08 I guess uh-with like emotional pain-a lot of people deal with it in their own way-I recently started like mental health therapy-and I don't necessarily know what part of it is working but I feel like it has been great to sit in a room and just talk about it—with somebody who isn't my friend-who I don't have to feel like I'm burdening-with my fears-</p>	<p>13:8 JG Is there anything I haven't asked you about that you think wow I'm really surprised she hasn't asked me this or anything that you would want to add or communicate about the experience with pain, with emotions, just having CF in general? Anything else that you feel you would like to add to what we've talked about?</p> <p>13:16 G Is there anything I haven't asked you about that you think...</p> <p>13:17 G What is that look? ...</p> <p>13:18 P08 It's like this li</p> <p>social legitimacy social pain</p> <p>invisibility</p>
<p>JG Okay-so more of a neutral party?</p>	<p>P08 Yeah-exactly-because of course-(boyfriend) and I are so honest with each other-and I tell him like every little ache and pain-and I feel like that can-even the-event the not serious stuff-like it can-it can weight him down more than it needs to-and I feel like maybe having an outside source is a really good idea-laugh-for CF and emotional pain-um-it just-I mean I think it will help my relationship with (boyfriend) long term not wearing it as thin as quickly or um-so yeah-I guess dealing with the emotional pain-um-communicating about it-is probably one of the best outlets for it-that I have found so far-</p>	
<p>JG Along the same line if you were to give one piece of advice to someone else who has CF what would that piece of advice be</p>	<p>P08 To talk about it-um-cause I know that there was a long period of time where I didn't—not that I was ashamed of it or anything-or like too shy-I just-I didn't want to tell people-it just wasn't-it felt like it was none of their business-and I didn't want the look that people give me when they find out-</p>	
<p>JG What is that look?</p>	<p>P08 It's like this like oh I'm so sorry-like pity-kind-of—laughs and it's irritating-and so I didn't talk about it um-but I don't think that that was necessarily the right decision-overall-um-I think it's been better for me now that I've been more upfront-and I communicate more about it than I ever have-</p>	
<p>JG It sounds like it's not only the willingness to talk to other people but maybe acceptance within yourself also-</p>	<p>P08 Yeah-and I think that that acceptance probably wouldn't have come around if I wouldn't have started talking about it-</p>	

Appendix I: Initial Stage Analysis (Mind-mapping)



Total Pain
Physical
Emotional
Social
Spiritual

Appendix I: Initial Stage of Analysis Paper Transcripts

1 JG So, um, I know you took some notes in preparation? Yeah, that's completely
2 fine. What I'd like for you to start with-cause I you-you know not super well
3 but fairly well-because we've been knowing each other for a number of years
4 now-
5
6 P05 Yeah, a few years
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8 JG But, if I did not know you at all-if I was just any Joe Blow down the street,
9 what I want you to do is to describe a little bit about yourself, about your CF,
10 just kind of what your life is like currently or has been like-so just very
11 general. If someone wanted to know (P05) 101.
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13 P05 So, I was diagnosed with cystic fibrosis when I was 4 months old. Um-my
14 mom's always told me it was because I had failure to thrive-um-so-I didn't-I
15 feel like it never really registered-I guess I always knew-well of course I
16 always knew I had CF-but I didn't really ever know what it meant. Until-I-I
17 remember having like-this like-realization moment-you know what I mean-
18 when I was like-maybe I was like 10 or something. I don't know-and I think I
19 asked-oh actually my parents told me this recently and I completely forgot
20 about this-I had no idea this happened-but I guess I asked them-like-could I
21 die from this-and I don't think they ever told me-like--straight out-because
22 it's kind of like when do you tell your kid-especially because I wasn't getting
23 medication-I was taking antibiotics-like oral antibiotics-but I had never
24 gotten a PICC line until I was 17.
25
26 JG Oh, okay
27
28 P05 So-I wasn't ever hospitalized when I was really young-uh huh-so I didn't
29 really-I don't know-I was just kinda like-okay I have an illness and I take
30 medication for it-but like what does this MEAN-does this mean that it's gonna
31 get harder-does this mean that it could kill me eventually-and I guess I asked
32 them one day and they had to tell me---this will kill you one day (laughs).
33 Um-
34
35 JG I'm sure that was a memorable day for them?
36
37 P05 For them-yes-I mean-they remember it perfectly-I have-I couldn't remember
38 it-I don't know why-I was like "Oh I don't remember asking you that" laughs-
39 but um-I do remember when it just kind of clicked-like-this feeling
40 afterwards-I don't remember the specific like moment
41
42 JG The conversation?
43

TRANSCRIBE TO STANZA R GEE
"COULD I DIE FROM THIS?"

← Death

← Death

Looking to future

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P05 [Yeah-and then-but even after that-I-people would ask me-like-oh they saw me taking my enzymes-or-all my friends knew about my vest-and they would all try it on-laugh-and um-and then um-so I had to do all those things- everyone knew-but other than that I would just be kind of like-oh it's nothing-you know-it doesn't really-I felt like it didn't really affect my life a whole lot.]

JG Hmm

P05 Cause I just did what I had to do-I still had fun-and it wasn't getting to the point where it was like interfering with my life-

JG Okay

P05 Until I was like 18-or 17-

JG And how old are you now?

P05 I am 26. Um-yeah-I should probably mention that I'm 26-laugh-um-so-um-and-so when I turned 17 I had my first hospitalization-my first IV-my PICC line-and after that-it just got harder and harder for me to stay healthy—um-so ever since then it's just been kind of-the progression is just-you know how it is-it just-gets-worse-and um-so-yeah-so now I-I live at home with my mom and dad-laugh-

First admission

JG Have you always lived at home?

Dev. milestones moved back home

P05 No I moved out a year and a half ago which was a BIG thing for me-because when I was 18 I always thought I would be out-like-I always wanted to be very-very independent-I graduated school-I got my um diploma early

JG Mhmm-oh early-you got it early

P05 Yeah-well know-actually they gave me my diploma- I walked with 2008 but I finished all my courses-um-the summer before the 2008 school year started

JG Oh so like a year before

P05 Yeah-a year before-and I wanted to work-and I-I just wanted to-you know-you know-I-so---this has been very hard for me the past few years because-I feel like my whole life has just like---been completely different.

Life has Δ

JG Mmm

STANEA (GEE)

"THIS IS NOT GOING THOUGH I THOUGHT IT WOULD"

Appendix J: Example Interview Excerpt and Related Stanza

- JG Have you through the years-have you had support from other people with CF aside from transplant-so just in general-have you like social network wise-
- P06 You know-I didn't really know a lot of CF-ers in my early years--when I first met people-it was being in the hospital (lists names)--I remember there was like eight of us together one time--and so we sat out in the hall-and it was just like a big group of us--
- JG Unofficial support group
- P06 Yes! Yes! Yes-that was pretty cool-that was pretty cool-um-yeah-
- JG In more recent years do you still have any sort of friends with CF
- P06 Well-it kind of sucks--but most of them have died--yeah-(voice trails off)-I'm trying to think of-well-(male with CF) is still around-he just got married--I don't really talk to him all that much--but it's like everyone has died (whispers)-and you know honestly--it's ooh-I haven't really gotten close to anybody--I don't know any new CF-ers here--I mean like there is a (hospital) CF chat group-I know (female with CF)-we are friends-we've done some stuff together-(female with CF)-we went to (female with CF) thing (funeral)-we saw (CF MD) there-um-I don't really-I mean it's like they've all died (whispers)

Box 15

Stanza 3: When I first met people-it was being in the hospital

10. You know – I didn't really know a lot of CF-ers in my early years
11. when I first met people it was being in the hospital
12. I remember there was like eight of us together one time
13. and so we sat out in the hall – and it was just like a big group of us.

Stanza 4: Most of them have died

14. Well – it kind of sucks
15. but most of them have died
16. I'm trying to think of – well – one is still around – he just got married
17. I don't really talk to him all that much.

Stanza 5: I haven't really gotten close to anybody

18. but it's like everyone has died--and you know honestly
19. I haven't really gotten close to anybody
20. I don't know any new CF-ers here.

(Amanda)

Appendix K: Symposium Summary

Symposium Session Summaries

the quality of adolescent relationships. We chose to focus on adolescents, as developmentally they are likely to experience opportunities to disclose their CF independently as their general level of independence increases (6) and their ability to understand and effectively express disease processes to others develops.

We hypothesize that adolescents who have disclosed their diagnosis will have families with more favorable attitudes towards disclosure, greater social support, and less perceived stigma.

Adolescents between the ages of 12-18 were recruited for the current study. Inclusion criteria require confirmed diagnosis of CF with pancreatic insufficiency, current prescription of pancreatic enzyme replacement therapy (PERT), access to the internet, and proficiency in reading English. Exclusion criteria include diagnoses of developmental delay or intellectual disability, history of a lung transplant, or a diagnosis of CF-related diabetes. Participants meeting inclusion/exclusion criteria are mailed a recruitment letter and a flyer describing the study along with a return addressed, stamped "do not contact" postcard that they can return requesting not to be contacted. All families for whom a postcard is not received within 10 days of mailing are contacted by phone or at their next clinic visit by study personnel to explain more about the study, assess the family's interest in participation in the study, and schedule a study visit. Adolescent participants complete baseline self-report measures assessing disease disclosure behaviors (Cystic Fibrosis Disclosure Scale [CFDS]) (2), family attitudes towards sharing private information (Family Privacy Orientation [FPO]) (7), perceptions of stigma (Social Impact Scale [SIS]) (8), and relationship quality (Network of Relationships Inventory [NRI]) (9). Participants' caregivers also complete the FPO. Participants' demographic and health data (ie, body mass index and FEV1 percent predicted) are obtained via medical record review.

Recruitment is ongoing with an anticipated final sample size of 20 participants. In our presentation, we will

discuss patterns of disease disclosure behaviors among adolescents with CF as well as associations between disease disclosure behaviors and other psychosocial variables. Findings have implications for anticipatory guidance and recommendations provided to patients with CF and their families with regard to potential health implications of disclosure and access to emotional and practical social support.

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S07.3

INVISIBILITY AND THE EXPERIENCE OF PAIN IN CF

Jessica L. Goggin, MAS

Pulmonary, University of California San Diego, San Diego, CA, USA

Background: As treatment and clinical care have improved, people with cystic fibrosis (CF) are living longer and CF, although still life-limiting has emerged as a chronic disease (1). This improved life expectancy often occurs alongside an increased symptom burden resulting from disease progression and co-morbidities (2). One of the common symptoms in CF is pain.

In a literature review, pain in CF was found to be a common phenomenon (3). The location of pain in CF is most commonly reported to be head, chest, back, and

stomach (3). Pain has been associated with lower quality of life, decreased adherence, increased pulmonary exacerbations and decreased survival (4-6). Despite indications that pain in CF is common and associated with negative outcomes, only 50% of CF patients who endorse pain report it (3). The phenomenon of invisible illness or invisibility has been described as an illness without a clear or visible outward sign (7,8). Pain, a highly subjective experience, and usually with no outward sign of illness or disability is often included when

discussing invisible illnesses but is this relevant in the setting of CF?

Aim: Existing research on pain in CF is limited and is primarily quantitative in design, with little known about the experience of pain. The aim of this study was to learn about the experience of pain in adults with CF. Preliminary data from the interviews, in conjunction with existing research, was used to explore the experience of pain in CF as viewed through the phenomenon of invisible illness.

Method: A narrative research approach, a subgenre in qualitative research methodology, was undertaken for this study. In-depth, conversational style interviews were held with participants that endorsed experience with pain in CF.

Findings/Discussion: Preliminary data from in-depth interviews reveals that the concept of invisibility may be relevant within the experience of pain in CF. Evidence that contributes to the phenomenon of invisibility of pain in CF includes patients not reporting their pain to clinical teams, lack of clear explanation for pain, lack of understanding or acknowledgement of pain by others, cultural expectation to overcome, and resignation that nothing can be done for the pain.

One participant explained: *"I don't really ask for help because there's not a lot anyone can do."* Another said *"...the pain is always there-always in the back of my mind...it's always going to be there and affect me still no matter what I do."* When people with CF do not report their pain, CF teams may not recognize it and perhaps more troubling, may not appreciate the impact pain may have on quality of life in CF.

When a clear explanation for pain in CF is not identified, it may have a negative impact on psychological adjustment or at a minimum lead to discouragement as demonstrated by a young woman with CF: *"I guess the doctors-they don't know really why-there's not been a study about it or anything but they're finding that in some order of CF it's kinda common-that's what (CF physician) said-so um-but there's not really anything they can do about it."*

Lack of acknowledgement or understanding by health care professionals, friends, family or even strangers may contribute to the concept of invisibility: *"You can feel fine one day and then the next day be totally done...I've lost a lot of friends over it cause they don't understand...I've had so many say 'Well you looked fine yesterday-you were fine'"*. The person with CF may feel a continual need to explain or legitimize pain and disability that may in turn lead to social isolation.

Several participants expressed the desire to push through: a young woman in her thirties described: *"I generally do a pretty good job of putting a front up to where I'm fine-even though I'm not."* A young man in his thirties similarly described *"I just push through and*

do what I have to do." A cultural expectation to overcome, previously referred to as the "John Wayne" syndrome, may lead to decreased reporting of pain to the CF team as people ignore the symptoms of pain and focus on remaining productive. Furthermore, society, including family, friends and even clinicians reinforce this style of coping through praise and admiration, often elevating the person to a hero status (8).

Finally, pain in CF may remain invisible due to a generalized expectation of suffering. This may be preface by failure of efforts to effectively diagnose or treat pain in CF that eventually leads to resignation that nothing can be done. One young man, expressed: *"I pretty much literally laid in bed and suffered-so-I mean-there's not really much you can do-there's not anything they can do so-I mean you just-a lot of times with CF you just kinda roll with the punches and take it as it comes-you know-I mean-you just gotta look at it as well that's my life."*

Recommendations: Efforts to increase visibility about the experience of pain in CF should be targeted towards both research and clinical care. Assessment of pain in CF should be a routine component of CF clinical care but must move beyond a simple yes/no or numerical pain scale as pain is underreported in CF despite existing requirements for pain assessment. Obtaining narratives from those with CF, whether in a clinical or research context, is an important first step in improving understanding of this complex phenomenon.

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