TREATMENT ADHERENCE IN **CYSTIC FIBROSIS** PATIENTS

Final Design Dissertation

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An investigation into methods of improving treatment adherence within adolescent and young adult Cystic Fibrosis patients

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ABSTRACT

The following report aims to identify and provide solutions to the barriers which may be preventing CF patients from adhering to their treatment plans. These treatments are administered via a variety of different delivery methods and treatment can consume hours of a patient's day.

After reviewing the current academic literature related to treatment adherence in CF patients, several major themes were identified. Clinic appointment attendance and it's importance in maintaining proper treatment plan adherence was identified as a research gap. These themes along with the identified research gap were used to justify the design and execution of the primary research methods, which were semi-structured interviews and an online questionnaire. These research methods were conducted with Australian CF patients.

The main findings centered around the importance of patients attending clinic appointments. These appointments provide an opportunity for consultation between patients and their specialist health care team. At each appointment patients undergo three procedures; testing their lung function performance via an instrument called a spirometer, having their airways examined by a respiratory specialist via the use of a stethoscope, and have consultations with several specialists in regards to their health and treatment plans. When patients don't attend these appointments, they run the risk of leaving chest infections go undetected, resulting in potential long term lung damage and ultimately a steep decline in health and quality of life.

After analyzing the interviews and online surveys it became clear that patients would be more likely to attend clinic appointments if they could be conducted remotely via video call. The inconvenience of travelling to and from hospital along with risk of cross infection between patients were cited as the main factors behind low clinic appointment attendance.

By developing and designing a product which allows patients to examine and monitor their respiratory condition from home, CF patients will have access to safer and more convenient methods of clinic appointment. delivery. As suggested by the primary research which was conducted, this will increase appointment attendance, in turn improving the treatment adherence rates of CF patients nation-wide.

INTRODUCTION

Cystic Fibrosis ('CF') is a genetic disease that predominantly affects the respiratory and digestive system. It is one of the most common life-threatening recessive conditions effecting Caucasians, with approximately 70,000 patients worldwide (Allen, Panitch, Rubenstein. 2010). The rate of disease varies drastically between different ethnicities. On average, CF affects .03% of the Caucasian population, .006% of the African-American population and .003% of the Asian-American population. Even between different Caucasian populations the occurrence of CF varies; for example the incidence of CF births in Ireland is .05%, whilst it is .01% in Sweden (Allen et. al, 2010). If both parents are carriers of the CF gene, there is a 25% chance of the child having CF (Figure 1). There is currently no cure for the disease, however recent leaps and bounds in medical technology and understanding over the last 40 years have allowed the average lifespan of CF patients to double (Alma et. al, 2019)(Figure 1).

Currently, CF patients are expected to live into at least their 40's, provided that treatment is started early and adhered to. Whilst this treatment plan can become time consuming, it must be followed strictly to maintain a patient's health. The routine includes prescribed medications, both oral and inhaled, as well as physical therapy and special dietary requirements. Patients can spend anywhere from 2 to 4 hours per day on treatments alone (Sawicki et. al, 2009). It has been reported that rates of patient adherence to these treatment plans range anywhere from 30% to 70% (Kettler et. Al, 2001), and that the consequences of poor adherence are 'significant' (Abbott et. al, 2001). Without proper treatment, patients can expect increased morbidity and mortality, reduced quality of life, and greater health care costs (Modi et. al, 2006). To ensure the highest possible quality of life, it is imperative that CF patients adhere as closely as possible to the treatment plan set out by their healthcare team.

The aim of this project is to identify the reasons behind why CF patients do not adequately adhere to their treatment plans and to provide insights on innovative methods that facilitate adherence. There are a multitude of factors contributing to low treatment adherence, and it is important for both the physical and mental health of CF patients and caregivers that these factors are identified, understood and addressed.

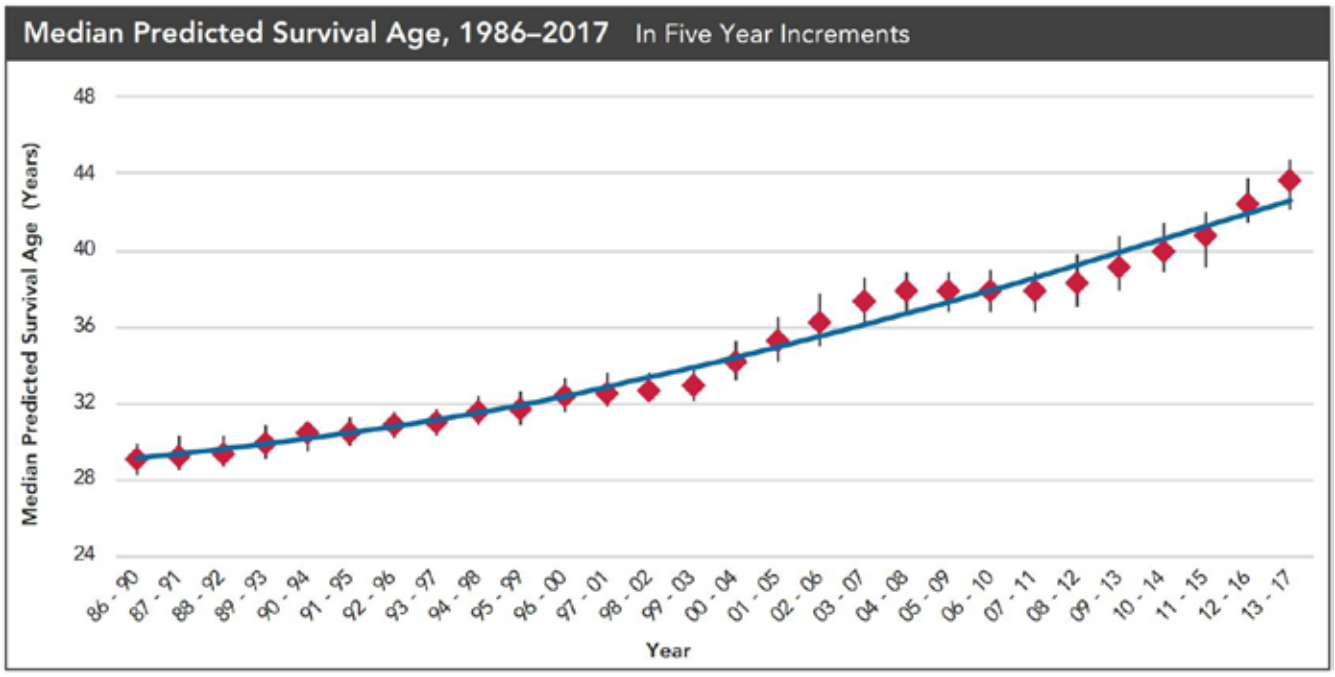

Figure 1: Median predicted survival age (CF Foundation, 2017)

LITERATURE REVIEW

LIMITATIONS

Sourcing accurate figures regarding treatment adherence allows physicians and health care teams to determine the impact of specific treatments on health status and, in health care systems where medication isn't publicly subsidized (US), to weigh the cost/benefit ratio for costly medication (Modi et. al, 2006). However, calculating the rates of adherence to any chronic disease treatment plan is difficult due to the absence of objective and accurate methods of recording medication ingestion (Allen et. al, 2011).

Each method of monitoring treatment adherence is associated with its own advantages and disadvantages. Self-reporting methods such as surveys and phone diaries, whilst easily accessible and affordable, are often compromised by factors such as social desirability bias and recall bias (Daniels et. al, 2011; Modi et. al, 2006). Pharmacy refill statements are relatively simple to obtain, yet only provide a history of how often medication is received from the pharmacy, and no confirmation of whether the medication is correctly ingested. Moreover, most patients also usually only restock once every 2 to 3 months, thus a window of at least 12 to 18 months is required to source enough data to inform adequate research (Eakin, 2011). Electronic devices such as Medication Event Monitoring System ('MEMS') medication bottle caps are used to detect when medication bottles are opened, and so provide the most accurate data relating to medication dispensing. Unfortunately, as with most electronic devices, they are expensive to deploy on a widespread basis and so may not be suitable for clinical use (Modi et. al, 2006)(Eakin, 2011).

The first study to deploy all four of these monitoring methods was conducted in 2006. Researchers concluded that whilst a multi-method approach resulted in more accurate figures, the accuracy of the results was still constrained by limitations in electronic measuring (Modi et. al, 2006). Technology has markedly improved in the ensuing years, which ought to have resulted in an improvement in the effectiveness of electronic measuring devices and subsequently the accuracy of such results. However, there has been no publicly available study employing all four of the previously mentioned methods since the initial study was conducted. As a result, factors such as social desirability and recall bias may still inflate data relating to treatment adherence rates.

BACKGROUND

Cystic Fibrosis was first distinguished from Celiac disease as a separate clinical condition in 1938 (Davis, 2005). At that time, infants with CF rarely lived longer than 6 months as there was very little understanding regarding the cause of the disease and how it might be managed. Physicians were aware that the primary cause of death was lung infection; however, it wasn't until the discovery of the sweat electrolyte defect, in 1959, that CF was recognized as not just a lung disorder (Davis, 2005). In the years since, continuous research into the causes of CF has led to medical and therapeutical breakthroughs, resulting in a decidedly higher average life expectancy and overall improvement in the quality of life of CF patients (Balli, 2018). Major advances include the development of Creon in 1985 (Huhn et. al, 2010), an enzyme replacement medication that provides proteins that can't be properly produced by the pancreas.

Recent research has revealed that CF is caused by mutations in the Cystic Fibrosis Transmembrane conductance Regulator ('CTFR') gene (Solomon et. al, 2015). This fault blocks channels within the cell walls from releasing and transporting chloride ions out of cells into mucus around the body. When chloride ions cannot leave the cell properly through CTFR, water is retained in the cell because of osmosis and mucus remains thicker than it should be. This is a major cause of problems for CF patients (Eckdahl, 2016), as this sticky mucus can be found throughout the body. This discovery has enabled targeted intervention at the cellular level via oral medication, giving patients access to state of the art medical treatment that aims to repair the faulty channels through the cell wall. This method of treatment has profoundly altered the course of the disease and drastically improved the outlook for CF patients (Solomon et. al, 2015).

In addition to oral medication treatment, CF patients must undergo rigorous physical therapy in order to clear the build up of mucus within their airways (Esther et. al, 2019). Physical aerobic activity is the most effective method of airway clearance and all CF patients are encouraged to exercise daily. For young children and those who are unable to partake in aerobic activity, a method of therapy known as Positive Expiratory Pressure ('PEP') provides accessible and relatively reliable clearance of the airways and lungs. The treatment involves exhaling through a mask which provides varying levels of resistance, thus opening the airways and forcing air in behind the trapped mucus (CFF, 2019). This loosens the sticky mucus lining the airways, which can then be expelled through controlled coughing (Allen et.al, 2010).

CF patients are educated about their disease from a young age and are encouraged to be as diligent as possible in engaging with their treatment plans. However, unfortunately research demonstrates that treatment adherence is suboptimal across the board (Eakin et. al, 2011). Patient commitment to treatment decreases as the perceived burden of the condition intensifies resulting in a decline in health, with some forms of treatments being neglected more than others (Narayana et. al, 2017). Adolescents and young adults are particularly at risk of reduced treatment adherence (Dzuiban et. al, 2010).

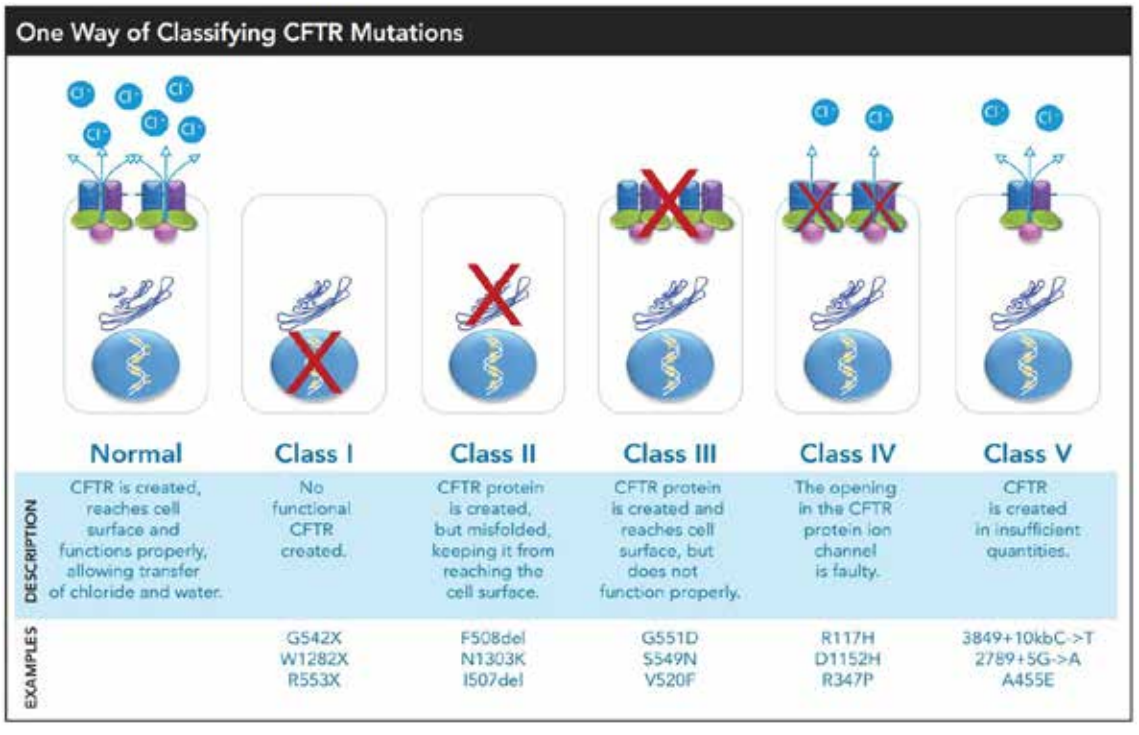

Figure 2: CTFR faulty cell wall functions (CF Foundation, 2017)

TREATMENT ADHERENCE

Over the course of the day CF patients take up to 40 tablets (Narayanan et. al, 2017), spend up to an hour and a half on physical therapy (Cff, 2019; Muther et. al, 2018), and are required to receive nebulized treatments which may take up to another 20 minutes to complete (Daniels, 2013). While adhering to this treatment plan is crucial for the longevity and quality of a CF patient's life, elements are often neglected or missed altogether (Narayanan et. al,2017; Eakin et. al, 2011). Treatment adherence rates in CF patients has been reported to be as low as 30% depending on the patient demographic and the type of treatment (Zindani et. al, 2016). In order to develop a solution that encourages the user to complete their treatments, it is important to understand the particular treatments being missed and the driving factors behind the lack of adherence.

The high number of tablets that CF patients must take per day is in part due to pancreatic enzyme replacement medication. They allow the intestines to absorb fat, protein and carbohydrates and are required with every meal (Barker et. al, 2016). Before this medication was available, children and infants with CF consistently suffered from malnutrition, which was the most common cause of premature death (HuiChuan et. al, 2008). Since this discovery, CF children have been able to absorb most of the necessary nutrients from their food and malnutrition is no longer such a prevalent issue. Nutritional status still has a significant effect of the progression of pulmonary disease and survival in patients with CF, however, so it is important that the pancreatic enzyme medication is adhered to strictly (HuiChuan et al, 2008). Studies have shown that adherence to nutritional medications is as low as 30% in some cases (Kuhn et. al, 2010). Adherence rates to this medication are particularly low in children with poor home environments (DeLambo et. al, 2004), which can have long term effects on nutritional status and BMI as they develop into adolescence. This is of particular concern as an unhealthy BMI has been cited as a contributor to poor lung condition (Alvarez et. al, 2015) (Figure 3).

Previous studies have reported that treatment adherence to physiotherapy such as PEP mask therapy ranges from 40% to 53% (Abbott et. al, 2001; Dziuban et. al, 2009). Another study reported that physiotherapy and airway clearance is often the most commonly missed form of

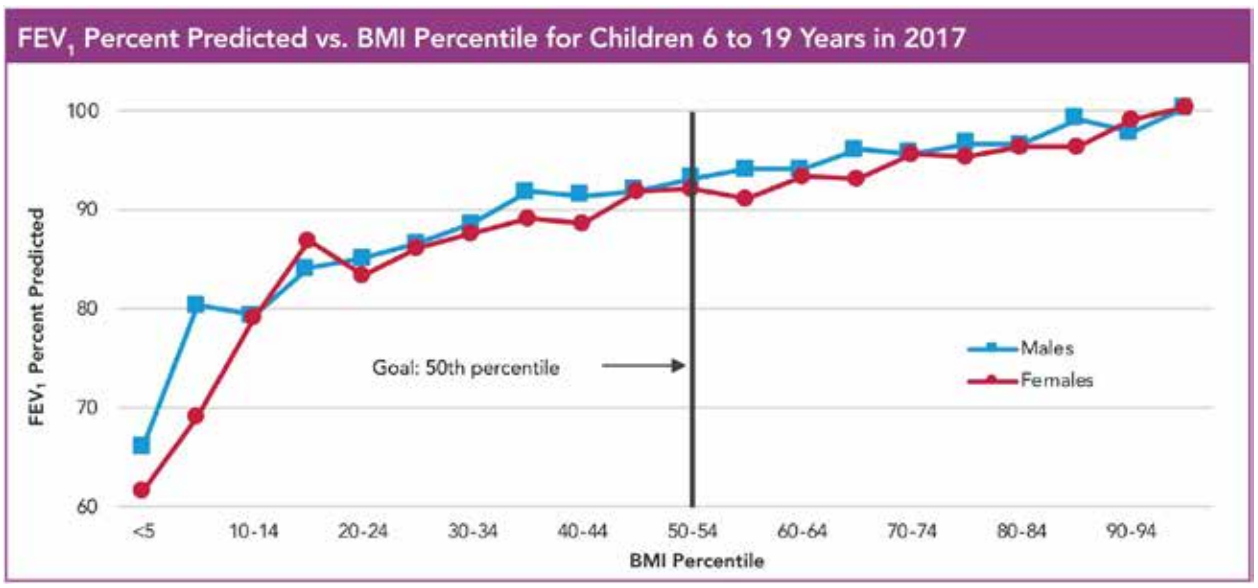

Figure 3: Lung Function vs. BMI (CF Foundation, 2017)

treatment as, because it is not pharmacological, there may be a perception that it is less essential to maintaining health (Dziuban et al, 2009). However, this is a damaging perception as the build up of mucus, rather than bacterial infection, appears to the initiating cause of lung damage (Esther et. al, 2019). As the build up of mucus within the airways provides an environment that harbors bacterial growth, airway clearance appears to be one of the most important treatments for CF patients (Esther et. al, 2019; Dzuiban, 2009). This is supported by data demonstrating that those with high physiotherapy adherence rates tend to perform better during lung function tests (Abbott, 2001). Related studies show that rates of adherence to other treatments such as inhaled antibiotics, oral medication and nebulized treatments range from 31-53%, 53-79%, and 41-72%, respectively (Eakin et. al, 2011; Eakin et. al, 2013). Each of these treatments contribute to the overall health and wellbeing of the patient, and together form a cohesive treatment plan aimed at reducing the progression of CF.

TREATMENT ADHERENCE BARRIERS

Multiple studies have established that the main factors behind low treatment adherence in CF patients are the volume of prescribed treatments, poor memory, time restrictions, CF related anxiety and depression, poor communication and home environments (Dziuban et. al, 2009; Bregenballe et. al, 2010) (Figure 4).

2.1 Volume of medication

CF treatment regimes are complex and time-consuming and vary slightly from patient to patient (Narayanan, 2016). While recent discoveries have led to medications that can significantly extend a patient's lifespan, increasing the amount of daily medications may add to the perceived burden experienced by CF patients (Muther et. al, 2018). The number and frequency of medications and treatments typically increases as the disease progresses, further contributing to this burden (Zindani et. al, 2006). A 2011 Danish study focused upon

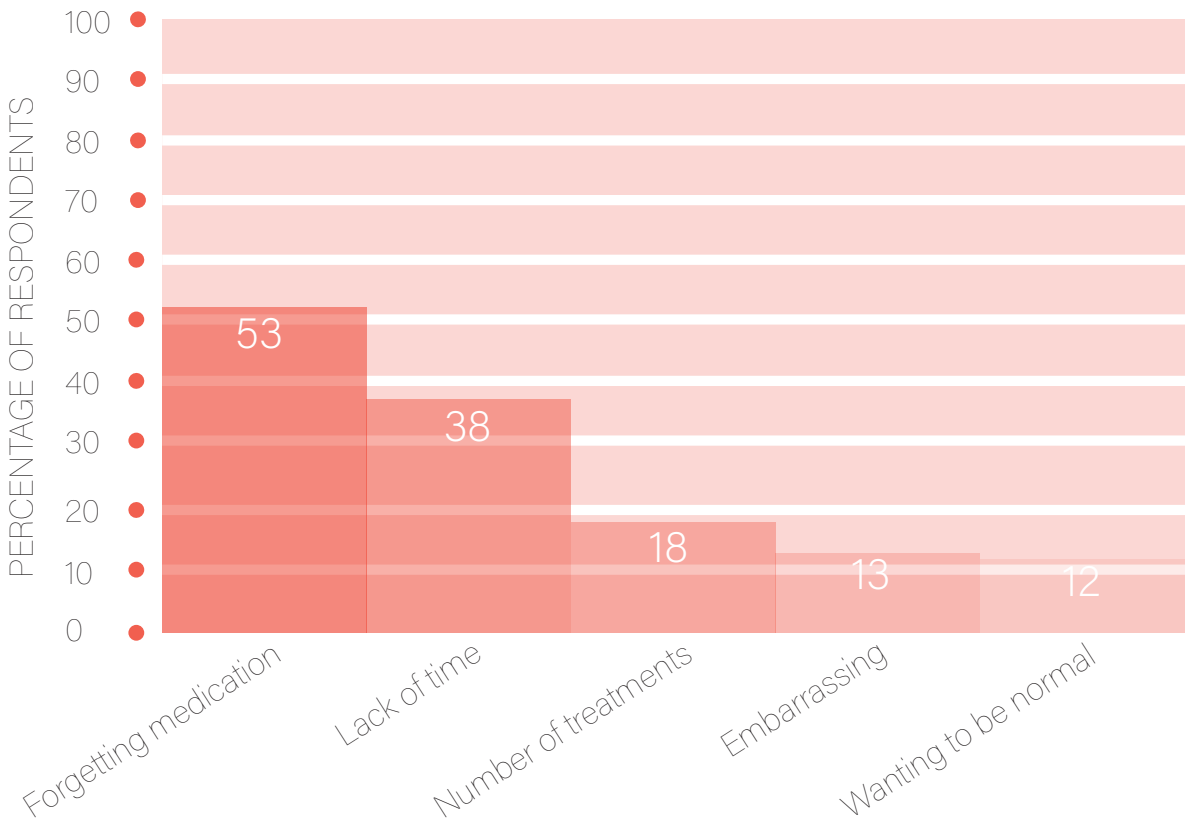

Figure 4: Barriers to treatment adherence (adapted from Dziuban, 2017)

barriers faced by adolescent and young adult CF patients, finding that 60% of patients and 62% of parents reported experiencing difficulties in relation to their treatment adherence (Bregnballe et. al, 2011). The study identified that poor adherence was particularly prevalent during adolescence, when the responsibility of managing treatments is handed over from the parent to the child. This, in combination with median FEV1 (Forced Expiratory Volume) levels of patients over 18 is on average 30% lower than those under 18 (VandanBranden et. al, 2011)(Figure 5), demonstrates that CF patients transitioning into adulthood are most at risk for accelerated lung function decline and increased risk of lung exacerbation (Figure 6). FEV1 is the volume of air expelled from the lungs within the first second of a lung function test.

2.2 Poor memory

Forgetting to take medication doses throughout the day is a common contributor to low treatment adherence amongst CF patients (Dziuban et. al,2009; Bregenballe et. al, 2011). As some patients are required to take over 40 tablets daily, it is understandable that doses may be

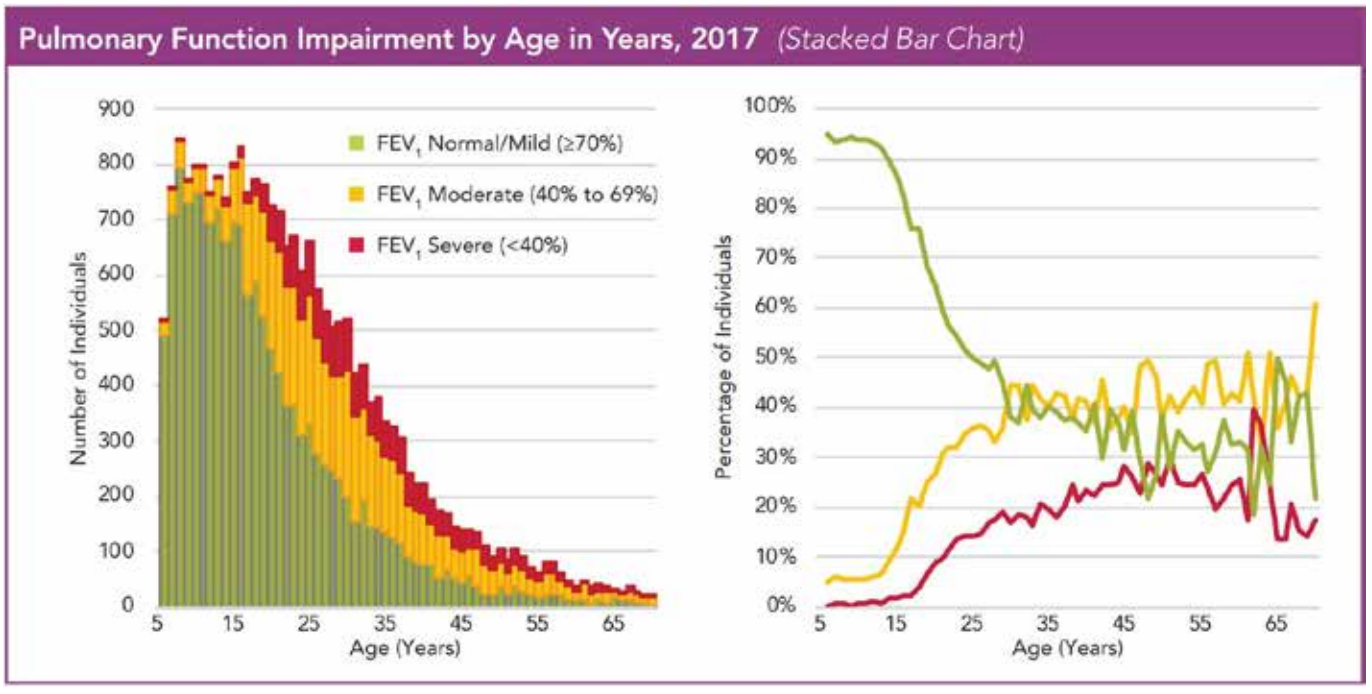

Figure 5: FEV1 scores vs age (CF Foundation, 2017)

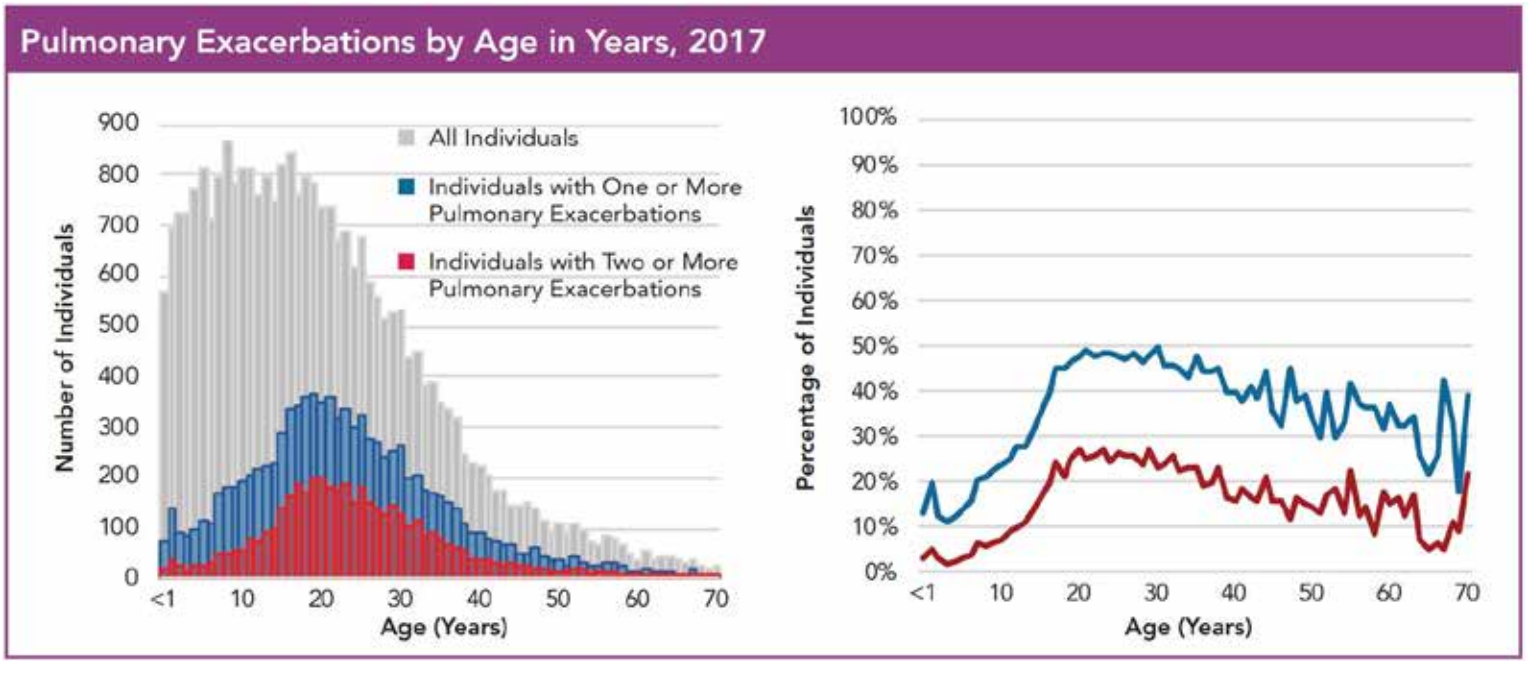

Figure 6 : Pulmonary exacerbations vs. age (CF Foundation, 2017)

occasionally missed. However a recent report has uncovered that in comparison to healthy control groups, CF patients perform considerably worse in attention and memory tests (Piasecki et. al, 2017). The researchers hypothesized that this may be due to CF patients being at a greater risk of fat-soluble vitamin deficiency, which has been linked to cognitive function (Koscic et. al, 2004). CF patients are prescribed vitamin A, B, D, E and K tablets from a young age, and the evidence from this study suggests that failure to take these tablets regularly may have adverse effects on memory function later in their life. In term, this has an impact on their memory and thus their ability to properly adhere to treatment plans, accelerating the development of pulmonary issues (Smith et. al, 2015). Thus, this research suggests that poor adherence to certain parts of the treatment plan at an early stage may be the driving factor behind missed doses of medication later on, rather than the volume of tablets.

2.3 Time restrictions

The lack of spare time to fit in one's treatments has also been cited as a factor behind decreased treatment adherence (Abbott et. al, 2011). As previously stated, daily treatments and therapies can take up a considerable amount of time daily (Smith et. al, 2015). More treatments are required as the disease progresses, resulting in an extended amount of time that needs to be dedicated to treatment per day.

Studies into treatment adherence barriers within CF children found that treatment adherence is highest during periods of structured routine. This research specifically revealed that adherence was higher during the school week than during the weekend and holiday periods (Ball, 2015; Bragenballe et. al, 2011). This suggests a need for families, as well as adult CF patients, to structure their daily routine in order to maintain optimal adherence to CF related therapies (Ball, 2015).

If patients are able to maintain high treatment adherence levels throughout adolescence and young adulthood, they may slow or even temporarily prevent the progression of pulmonary damage. Thus, consistent adherence to treatment plans may render additional treatments unnecessary, resulting in more spare time to dedicate to existing treatments.

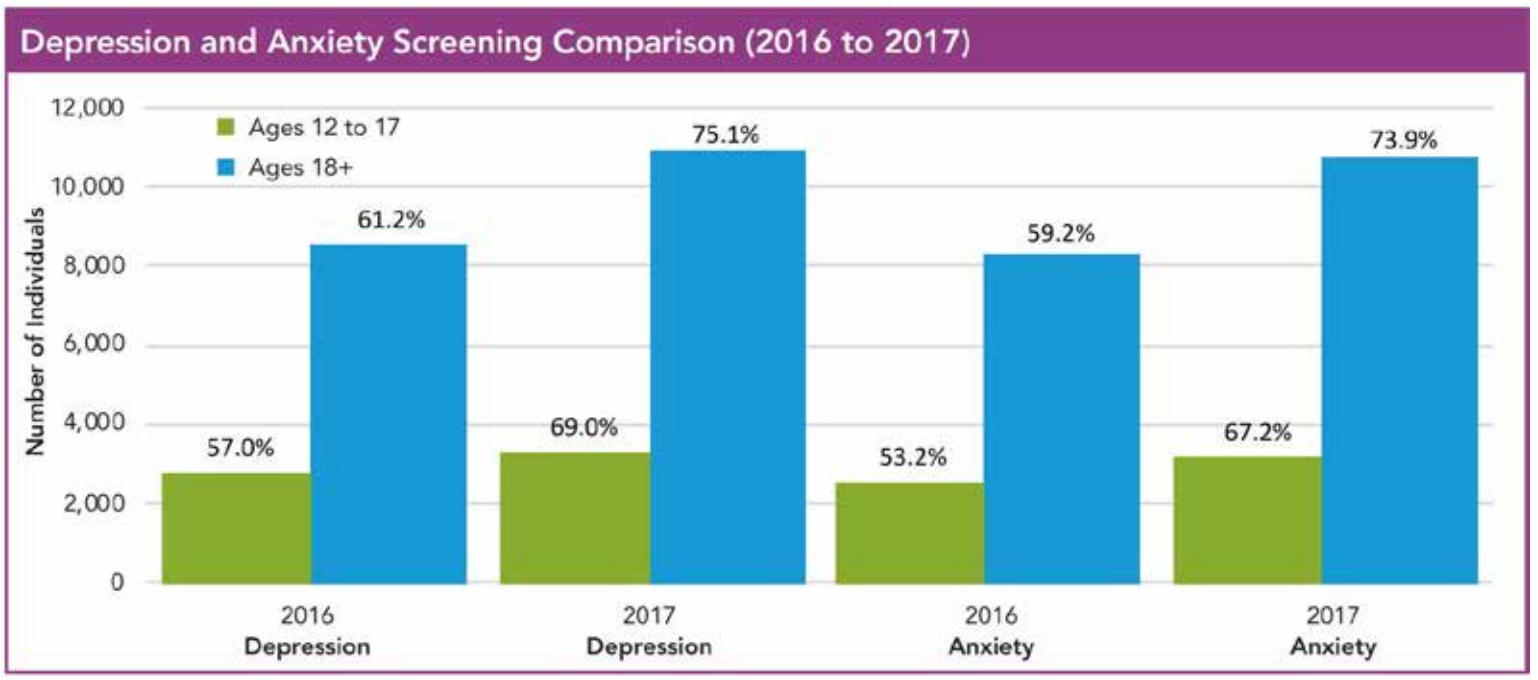

Figure 7 : Percentage of CF patients who experience depression and anxiety (CF Foundation, 2017)

2.4 Depression and anxiety

Unwillingness to take medication in public relates to root causes such as mental stress, anxiety and depression (Smith et. al, 2015). Research shows that adolescent CF patients cite revealing their condition to peers as a core concern (Abbot et. al, 2001). CF patients also report embarrassment arising from coughing in public (Whitworth, 2018). Factors such as these add to the perceived burden and embarrassment faced by those suffering from CF. CF patients are 2-3 times more likely to develop depressive symptoms when compared to wider community samples (Smith et. al, 2015; Muther et. al, 2018) (Figure 7) . This perceived burden is heightened throughout adolescence, stemming from factors such as becoming independent from parents, the natural progression of their CF, and being exposed to risky behaviour such as drinking and drug taking (Muther et al, 2018). This burden plays a large role in low treatment adherence (Bregnballe et. al, 2011; Smith et. al, 2015; Muther et. al, 2018). As a CF patient's condition worsens and more treatments are added to their treatment plan (Muther et. al, 2018), feelings of anxiety and stress are magnified. This cycle of condition related anxiety and stress can cause serious damage to a patient's physical and mental health (Figure 8).

2.5 Poor communication

Patients who are less knowledgeable about their disease and condition are less likely to adhere to their treatment plan (Modi et. al, 2006). Adherence problems are intensified by varying levels of understanding of the severity of illness and expectations for treatment amongst parents, children and physicians (Zindani et. al, 2018). Another study revealed that when patients perceive themselves to be healthy, there is a decline in completed treatments and regular communication with their health care team (Dziuban et. al, 2010).

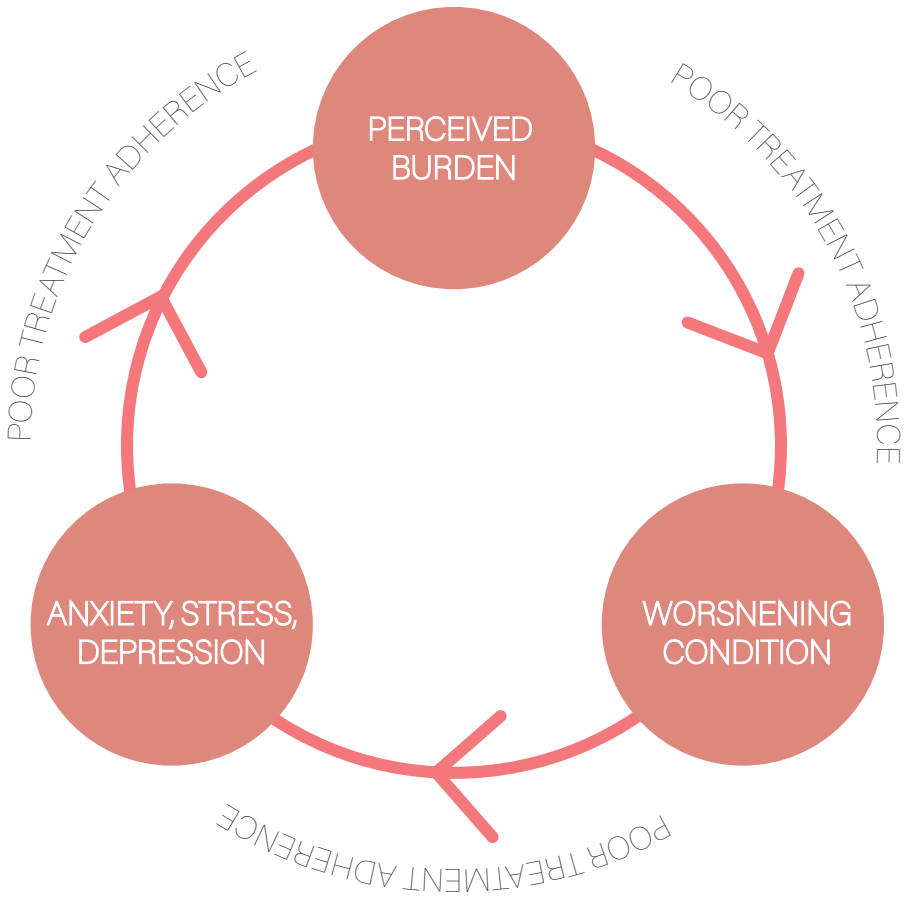

Figure 8 : Perceived treatment burden cycle
(Author's own illustration)

This research highlights the importance of patients being correctly informed on their condition, as reduced treatment adherence amid a chest infection can have long term effects on lung health and lung function decline (Muther et. al, 2018). The most effective way for CF patients to remain informed is to maintain regular communication with their health care team, and to have regular lung function tests approximately every 2 to 3 months (Muther et. al, 2018).

Quarterly clinic visits form the foundation of any CF treatment plan (CFF, 2020). These checkups provide CF patients with an opportunity to receive feedback on their condition through various tests and consultations with specialists. Non-attendance at clinic appointments is a common problem among adolescent and adult CF patients and may result in incomplete monitoring and exacerbate poor adherence to treatment (Walker et. al, 2008). Figure ttend their scheduled clinic visits (CF Foundation, 2020; VandenBranden et. al, 2011). Clinic visits include lung function tests and emotional wellbeing consultations, which are critical as these aspects of CF are rarely monitored outside hospital walls. Adjustments to treatment plans are also commonly made as a result of clinic visits, incorporating new therapies and adapting to changes in a patient's condition. Barriers to attending clinic visits include anxiety in relation to declining health and increased attention from their health care team, caused by poor treatment adherence.

Decrease in lung function is partly due to the natural progression of CF lung disease, and is also likely due to poor treatment adherence along with low clinic appointment attendance rates. If adolescent CF patients, who are particularly at risk of accelerated lung function decline in comparison to other age groups (Eakin et. al, 2017), can navigate this period with increased adherence to their treatments and frequent communication with their health care team, the quality and duration of their life is likely to improve.

2.6 Home environment

Most CF cases are diagnosed at birth through a heel-prick test and a sweat chloride test (Allen et. al, 2010). Thus, parents of CF patients face uncertainty regarding their child's health and future from the very beginning of their child's life. This uncertainty can often lead to anxiety and depression amongst parents of patients (Barker et. al, 2016), with depressive symptoms being reported by 37% of mothers and 31% of fathers of CF patients (Barker et. al, 2016).

As children with CF grow up, their family environment plays a significant role in their disease management (DeLambo et. al, 2004). CF patients are heavily reliant upon their parents to administer their medication at a young age and studies have shown that children with parents presenting depressive symptoms are less likely to adhere to treatment plans (Barker et. al, 2016; Smith et. al, 2016). Families who have more structured daily routines and are more emotionally expressive tend to report higher treatment adherence as opposed to families who are less so (DeLambo et. al, 2004; Bregnballe et. al, 2011). Other environmental factors also have a significant impact upon the health of children with CF, for example patients who are exposed to second hand smoke are more likely to experience respiratory infections (Loman et. al, 2020). This research emphasizes the importance of a supportive and communicative family environment for CF patients.

2.7 Conclusions

The reasons behind low treatment adherence are numerous and complex, yet a few key factors have been identified. After thoroughly reviewing the current literature discussing treatment adherence within CF patients, the following conclusions can be drawn:

- The mental health of CF patients and their care givers plays a crucial role in their treatment adherence
- At the same time, their physical health and condition plays a crucial role in their mental wellbeing
- Staying dedicated to their treatment plan from a young age has long term benefits to their mental and physical health
- A positive family environment plays an important role in forming this dedication as well as laying the foundation forstrong treatment adherence habits
- While all prescribed treatment contributes to a patients' medical condition, physiotherapy (aerobic exercise and PEP mask therapy) is one of the most influential factors in maintaining physical health
- Sustaining communication with healthcare teams and being aware of the state of one's condition improves long term health and treatment adherence.
- Adolescent and young adult CF patients are particularly at risk of accelerated lung function decline
- Forgetfulness, lack of time and CF related anxiety/depression are listed as the major factors behind low treatment adherence

2.8 Research Gap

Treatment adherence is one of the main factors in the physical condition of a CF patient, which in turn contributes greatly to their mental health. Through reviewing the available literature, it has been uncovered that mental health plays a significant role in the physical condition and quality of life of CF patients. During clinic appointments, CF patients discuss their condition with specialists such as doctors, physioterapists and dieticians. When patients fail to attend clinic appointments, they miss important opportunities to communicate with their healthcare team and receive feedback on the status of their condition. If the patient is harboring a lung infection which goes untreated due to clinic absence, sustained lung damage can occur.

Occupational therapists are also accessible during these appointment, providing a method to release mental strain and anxiety. By increasing communication and the frequency of their clinic appointment attendance, CF patients may perceive their condition to be less burdensome and be more likely to adhere to treatment.

Whilst a large body of research has been conducted regarding treatment adherence rates and methods, very minimal information is available regarding the available channels of communication and clinic attendance rates.

By increasing knowledge in this field, solutions may be developed which encourage CF patients to communicate with their health care team more often, ultimately improving their quality of life.

RESEARCH DESIGN

METHODOLOGY

3.1 Why qualitative research?

In order to understand the complex reasonings behind treatment adherence and levels of communication between patients and healthcare teams, qualitative research methods were employed. Qualitative research is more flexible and interpretive than quantitative research. By conducting qualitative research, deeper knowledge into the reasoning and behaviour behind the topic of treatment adherence could be obtained. Qualitative research is multimethod in focus, involving an interpretive, naturalistic approach to it's subject matter (McLeod, 2019). Conceptually, qualitative research is concerned with understanding human behaviour from the user's perspective. It is very much a user-centric form of research. Methodologically, qualitative data is typically collected through participant observations and interviews. While quantitative research is useful for collecting facts regarding social phenomena, it fails to understand the personal nuances and behaviour behind such phenomena, and would not yield the desired information. The pros and cons of qualitative research are listed below (Table 1).

Table 1: Benefits and limitations of qualitative research

Qualitative Research	
Benefits	Limitations
· Researches can gain insights into the subtleties and complexities of a user's journey that other scientific, quantitative methods may miss · Qualitative data allows for ambiguities and contradictions within the data, which reflect social reality (Denscombe, 2010) · In the case of semi-structured interviews, researchers can follow paths and points of information they may have otherwise not considered, resulting in valuable, user centric data.	· Depending on the method employed, qualitative research can be more labor intensive and time consuming for the researcher · Qualitative data collection may be limited by the willingness of the respondent to answer questions truthfully and at length · Due to time and costs involved, qualitative designs generally do not draw from large sample sets · Difficult to apply conventional standards of reliability and validity due to the subjective subject matter – each subject is different

3.2 Outline of research approach

The aim of this research project was to investigate whether low clinic attendance and infrequent communication between patients and health care teams has an impact on their treatment adherence and mental health. Qualitative research was able to provide a deeper insight into these factors.

An anonymous survey was used to gather information on CF patient's attitudes and behaviour in relation to their treatment adherence, levels of communication, mental health and clinic appointment attendance. These were identified as potentially sensitive topic for some CF patient, so consideration was taken in the formulation of questions. All respondents were notified that they would remain anonymous throughout the research and analysis process, encouraging the patients to answer questions honestly. The author has also used his position as a CF patient to reassure respondents that the research being conducting is in both of their best interests, and that at the end of this project a product or service would be developed which will ultimately improve the quality of life of all CF patients.

Once the initial responses to this survey were received, more in-depth video call or phone interviews were be conducted with CF patients. This allowed researchers to delve deeper into points of interest or trends that appeared in response to the survey questions.

Due to the current situation involving COVID-19 in-person interviews and observations were not a possibility. The range and depth of questions within the surveys reflected this limitation and aimed to provide a deeper level of insight into respondents' behaviour and habits.

3.3 Research methods used

Triangulation was used in order to gather thorough, reliable data on the topic of CF patient treatment adherence.

By employing more than one method of data collection, triangulation allows the limitations from each method to be transcended by comparing findings from different perspectives (Williams, 2005).

In the case of this study, semi-structued interviews and online questionnaires were used as the two methods of data collection. The trends and points of information which appeared within the online questionnaires could be expanded on in greater detail through conducting semi-structured interviews. Employing both methods also resulted in a larger sample of data to draw conclusions from.

3.4 Participants

The participants involved in this study were Australian CF patients. Invitations regarding survey participation were extended to international Cystic Fibrosis organisations yet no response was received.

The ideal sample group were young adult CF patients aged between 18-25. However, in order to gather as much data as possible, results were accepted from patients outside this age bracket. Subsequently most of the respondents fell within this group anyway, with only a few patients aged over 25.

CF patients who were under 18 were required to get their parents and/or caretakers to respond in their place due to ethics approval requirements.

3.4 How research was depolyed

Contact was made with Cystic Fibrosis Australia (CFA) and their database of CF patients throughout the country was used to circulate the surveys. An identical online survey was sent to as many CF patients within the age bracket of 16-25 as possible. This survey gathered information regarding the CF patients' habits and attitudes around treatment adherence, clinic appointment attendance and communication between themselves and their CF care team.

3.5 Variables and limitations

It was understood that some CF patients will be more dedicated to their treatment and clinic attendance than others. These variances provided valuable information on habits across the CF population. The health of different CF patients will in turn vary as well. It was also understood that different CF clinics may have different routines and processes regarding patient clinic visits and communication.

The distance between where patients live and how far their nearest clinic was may also have an impact on their attendance rates. These variables could uncover whether remotely located patients had lower lung function. A patients mental wellbeing may have also contributed to their willingness to answer questions fully and truthfully.

RESULTS

ANALYSIS

4.1 Interview Analysis

Responses from the semi-structured interviews were transcribed and coded. Common topics that came up throughout the interviews were identified and grouped into themes. These themes uncovered the respondent's opinions and beliefs on areas of interest in relation to the research topic. Themes that appeared more often were considered to be of greater relevance to the research question in comparison to themes which appeared less often. The rate at which they were mentioned was used to justify the conclusions that have been drawn throughout the discussion section of this report.

Examples of how this process was executed can be found in Table 2 (Appendix page 50), while the specific topics of conversation and the frequency at which they were mentioned throughout the interviews can be found in Figure 9.

4.2 Survey Analysis

Some questions on the online survey used ordinal measurements, such as those relating to treatment adherence rates, while others employed nominal and interval/ratio measurements, such as which treatments were prescribed to patients and their opinions on varying topics, respectively.

Likert scale answers were plotted on variability graphs displaying the mean response along with standard deviation (Figure 15). Ordinal and nominal responses were plotted on clustered bar graphs which illustrated the data in an effective and easy to read manner (Figure 12). By illustrating the data in these forms, comparisons could be made, trends could identified, and conclusions drawn.

RESULTS

4.3 Interview Results

After conducting semi-structured interviews with CF patients, several key themes arose. The analysis codes which were used to identify these themes along with how frequently they were mentioned are illustrated in Figure 9. These themes were formed by grouping together relevant topics which appeared throughout the interviews.

The most common topic of discussion across all interviews was that of a structured daily routine around treatment adherence habits. Interviewees discussed the importance of routine in their daily lives and how it contributed to their health and lifestyle. Interviewees who were not as dedicated to their treatment plans reported placing less importance on routines. This is evident in P2's (Tamika?) response to whether they had any routines in place around treatment adherence:

'Nope.'

The second most common topic of discussion centered around treatment habits and treatment plans. Interviewees discussed the details of their treatment plans, including which medications they were on, and provided insight into the completion of treatments and the circumstances around when these treatments are changed. P3 explained:

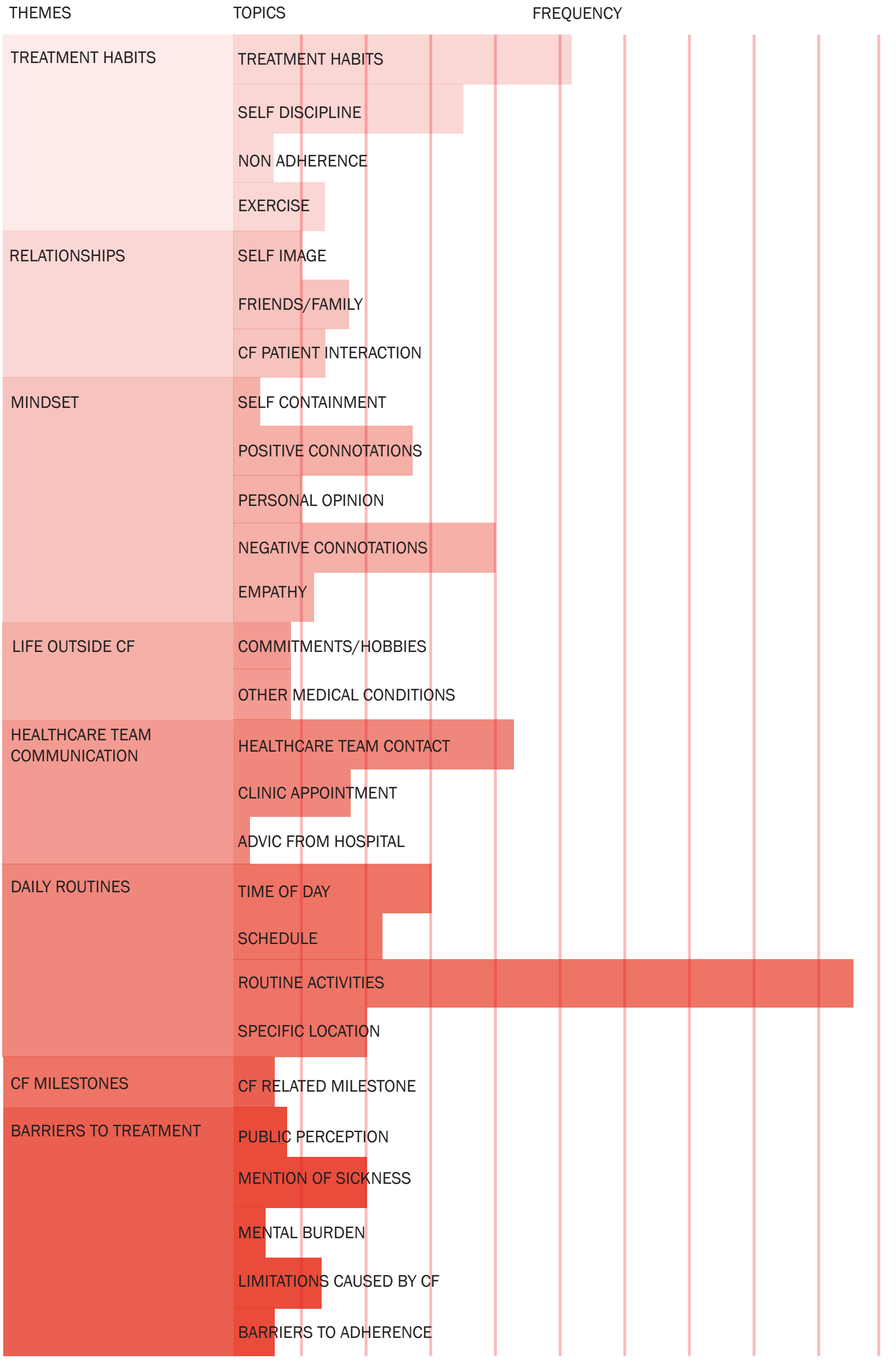

Figure 9: Frequency of coded interview responses

'Usually I have 2 months on, one month off for some treatments but at the moment due to COVID I'm constantly on medication so I stay well.'

When asked about which treatment they find the most difficult to complete, P3's explanation provided an insight into the challenges CF patients face in completing their treatments:

'I'd say probably the Hypertonic saline because it takes quite a while and I have to get a bit motivated to do it.'

Attending 'telehealth' clinic appointments - remote appointments via videocall instead of being in physical attendance - was another topic that frequently arose. Interviewees spoke of their preference of this method:

'It was interesting – it felt like I was in a normal clinic appointment. They set up a video and put you in a room and people come and go. I felt like it took a little longer than a regular clinic appointment, but it worked and it was effective. I have another one next week.'

While the majority of interviewees preferred Telehealth over regular clinic appointments, P3 provided some insight into the drawbacks of the system. Her statement highlights the apparent importance of familiarity between patients and their doctors:

'It [the telehealth appointment] was kind of like 50/50 – my first one was with a doctor I hadn't heard of before – I'm talking to a complete stranger, no video etc.'

When asked about their communication between themselves and other individuals with CF, interviewees gave a mixed response. Some believed communicating with other CF patients was a beneficial process which provided them with some reassurance of their condition (P4);

*'Yeah usually when other people have an issue they might go 'hey have you gone through this? We have b**** sessions about treatments. It's nice to have people who understand what you're going through.'*

while others found that hearing about other CF patient's experiences and struggles resulted in feelings of anxiety about their own condition. This mindset was described as 'self containment' within the coding process (P3):

'No, I don't [speak with other CF patients]. I have once before but it sort of freaks me out knowing what I could go through. I rather stay in my own little world.'

Several responses mentioned the link between health decline and an inability to lead a 'normal' lifestyle or the lifestyle they usually lead when healthy. Respondee's spoke of wanting to keep up with friends and family but being unable to due to their condition. P4 highlights this in her response:

'I think it's probably like wanting to keep up with people but in my head I know I can't keep up with people – trying to be normal but knowing I can't be.'

they also spoke of the public perception of CF and how this has an impact on their own mindset;

'I guess because looking from the outside CF people look like everyone else but the struggles you go through at home that no one sees is where the understanding of people is not quite there – sometimes it is tough and although my family understand, there's only a certain level of understanding they can have and don't fully get it.'

Finally, a question regarding whether patients sometimes forget they've taken

medication shortly after consuming it resulted in novel responses, as can be seen in P1's answer:

'Oh all the time! I guess when you're out at someone place and you're eating and think 'oh **** did I take my CREON' and then you ask your friends/family and they say 'yes you have'. Sometimes at home I forget if I've had them and then take extra just in case.'

4.4 Survey Results

The online questionnaire research aimed to explore treatment adherence to several specific treatments: PEP (Positive Expiratory Pressure) chest clearance therapy, nebulized treatments named Pulmozyne and Hypertonic Saline, and several medications in tablet form called ABDEK, CREON and Azithromycin. Whilst certain respondents reported being prescribed various other medications, the rate of prescription to these medications was too low to fall within the scope of the study. The treatments and the number of participants taking these treatment can be found in Figure 10.

4.4.1 Chest Therapy

86%(12) of respondents reported PEP as part of their treatment plan (Figure 10). Over half (64.3%) of those respondents reported missing their PEP airway clearance treatment at least once per week, with 38% missing treatment at least 3-5 times per week, and 28.6% consistently missing their PEP treatments (Figure 11). 64.3% of respondents reported that being unable to fit their treatment into their daily schedule was a factor behind missing treatments, while 42.9% of respondents stated the reason behind not completing PEP treatments was due to forgetfulness (Figure 12).

4.4.2 Nebulized Treatments

78.6%(11) of respondents reported being prescribed the nebulized medication Pulmozyne as part of their treatment plan (Figure 10). 66.7% of respondent reported to missing their nebulized Pulmozyne treatment at least once a week, with 41.7 % of these respondents always missing their Pulmozyne treatment (Figure 11). Similarly to the PEP responses, the most common reasons behind missed treatments are fitting them into their daily schedule (75%) and for-

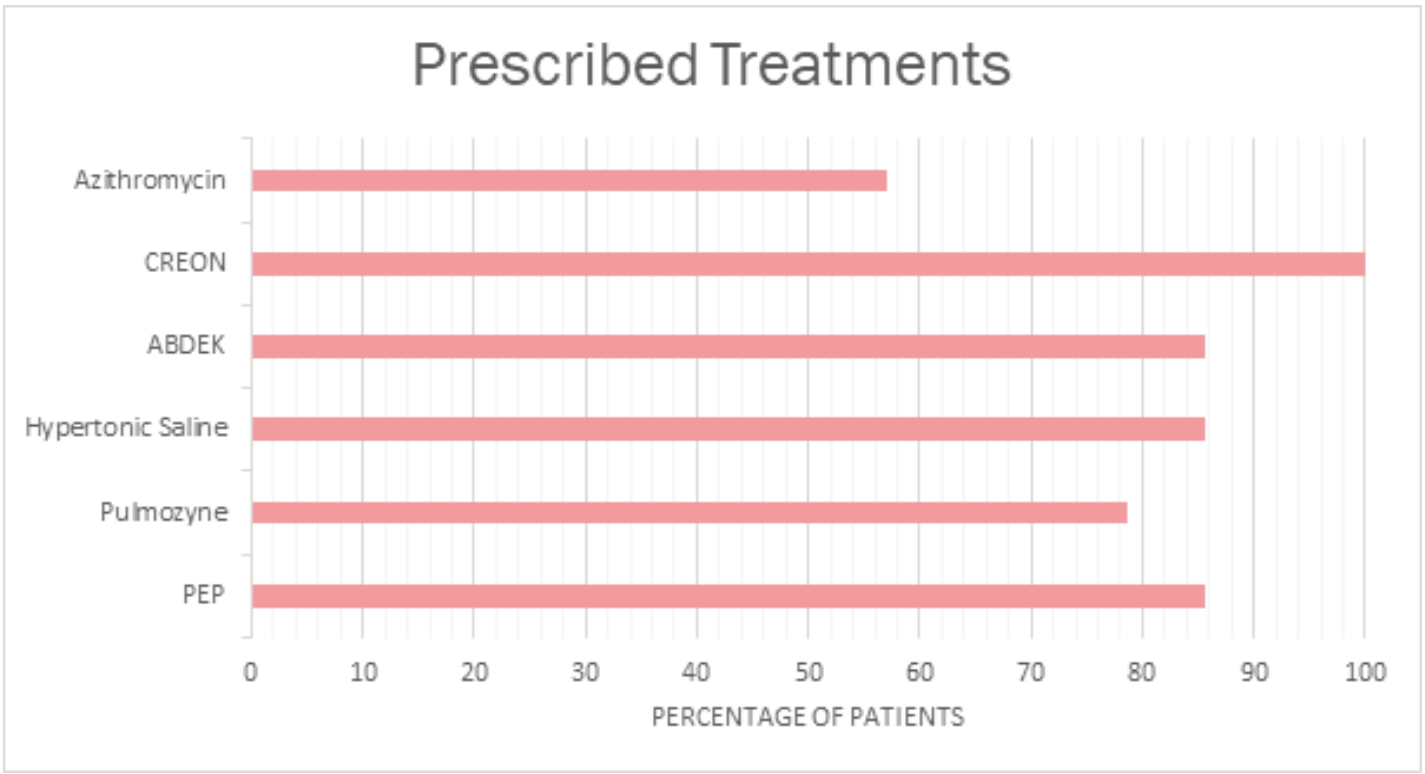

Figure 10 : Perceived treatment burden cycle

getting to complete the treatment (50%) (Figure 12).

Another nebulized medication commonly prescribed to CF patients is Hypertonic Saline. 71.4 %(10) of respondents were prescribed this as part of their treatment plan (Figure 10). 75% of respondents reported to missing their Hypertonic Saline treatment at least once a week, with half of respondents missing at least 3-5 treatments per week (Figure 11). Again the main reasons behind missed treatments were attributed to being unable to fit them in amongst their daily schedule (75%) and forgetting to complete the treatment (50%) (Figure 12).

4.4.3 Tablet Medication

ABDEK, a high concentration multi vitamin tablet, was prescribed to 85.7% of respondents (Figure 10). 53.9% of these respondents reported to not taking their ABDEK tablets at least once a week, with 46.2 % missing the treatment at least 3-5 times per week (Figure 12). The most common reason behind missing their ABDEK tablet treatment was forgetting to take the tablets (45.5%). The second most common answer was missing ABDEK treatment due to the lack of any noticeable effects (27.3%) (Figure 13).

100% (14) of all respondents were prescribed CREON, a pancreatic enzyme replacement medication (Figure 10). 50% of respondents reported to not taking their CREON tablets once a week, with 42.9% of respondents missing their CREON tablets at least 3-5 times per week. Interestingly, 28.6% of patients reported always missing their CREON tablets, being the treatment with one of the highest rates of complete non adherence (Figure 11). Forgetting to complete the treatment was the most common reason (58.3%). The second most common reason was a reluctance to complete the treatment around friends/family or in public (25%)(Figure 12).

57%(8) of respondents were prescribed Azithromycin, an antibiotic in tablet form (Figure 10). 50% of respondents reported missing their Azithromycin tablets at least 3-5 times per week (Figure 11). As this medication is

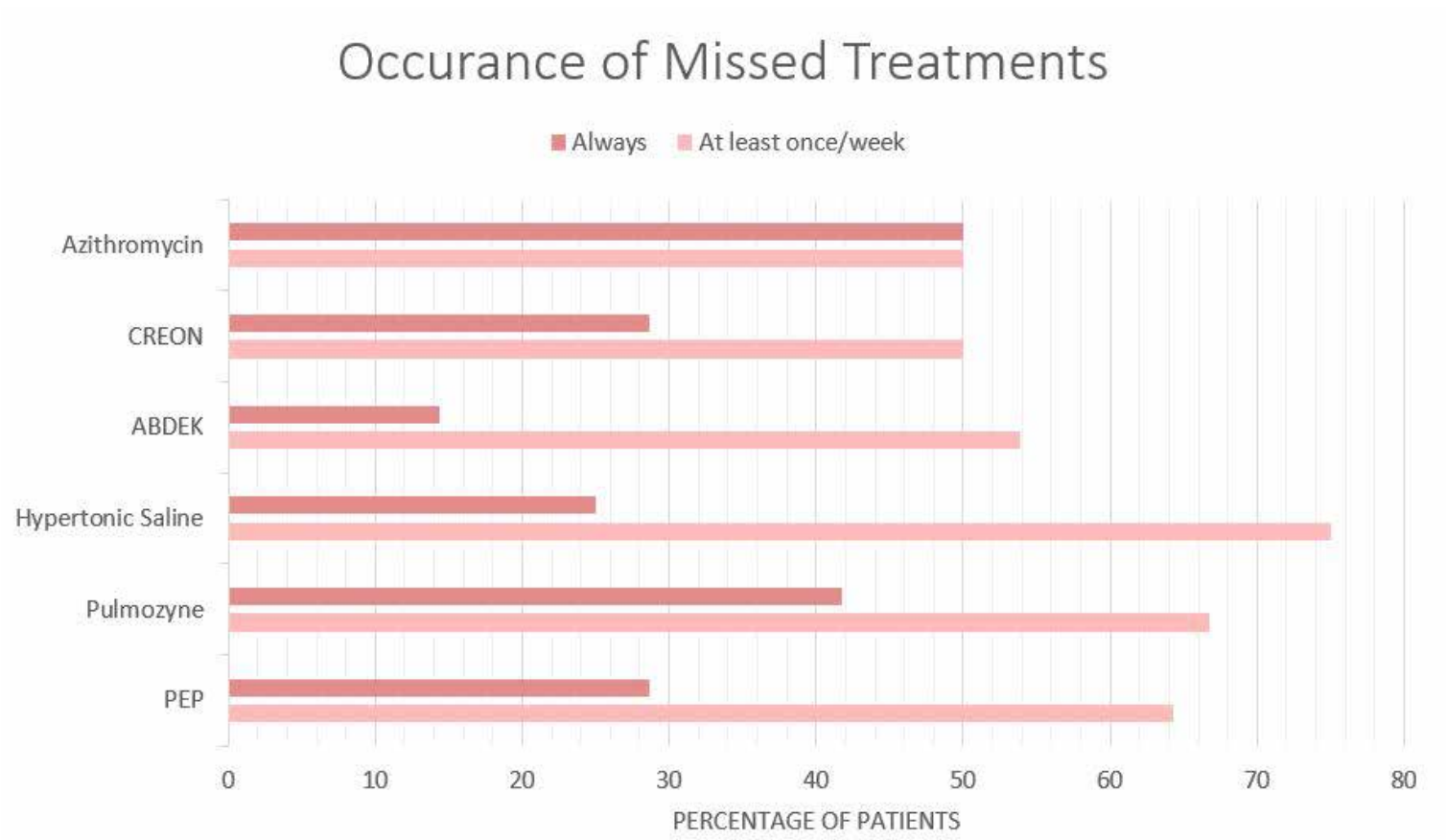

Figure 11 : Frequency of miss treatments

prescribed to be taken 3 times per week, these responses are considered to fit within the 'Always missing' category. The most common factors behind missed treatments included forgetting to complete treatments (62.5%) and the treatment having no noticeable effects (37.5%) (Figure 12).

4.4.4 Pharmacy Ordering

92.9% of respondents state that they ensure their medication is restocked before running out of their current medication rounds. Only 7% of respondents contact their pharmacy via email, with another 7 % visiting their pharmacy during monthly clinic visits. The remainder of respondents contact their pharmacy over the phone (Figure 13). Although 64.3 % of respondents agreed that they found the process of ordering medication easy and convenient, 85.7% of respondents stated that they would prefer to use an online platform to check script levels and order medication, with 7.1% feeling indifferent (Figure 14).

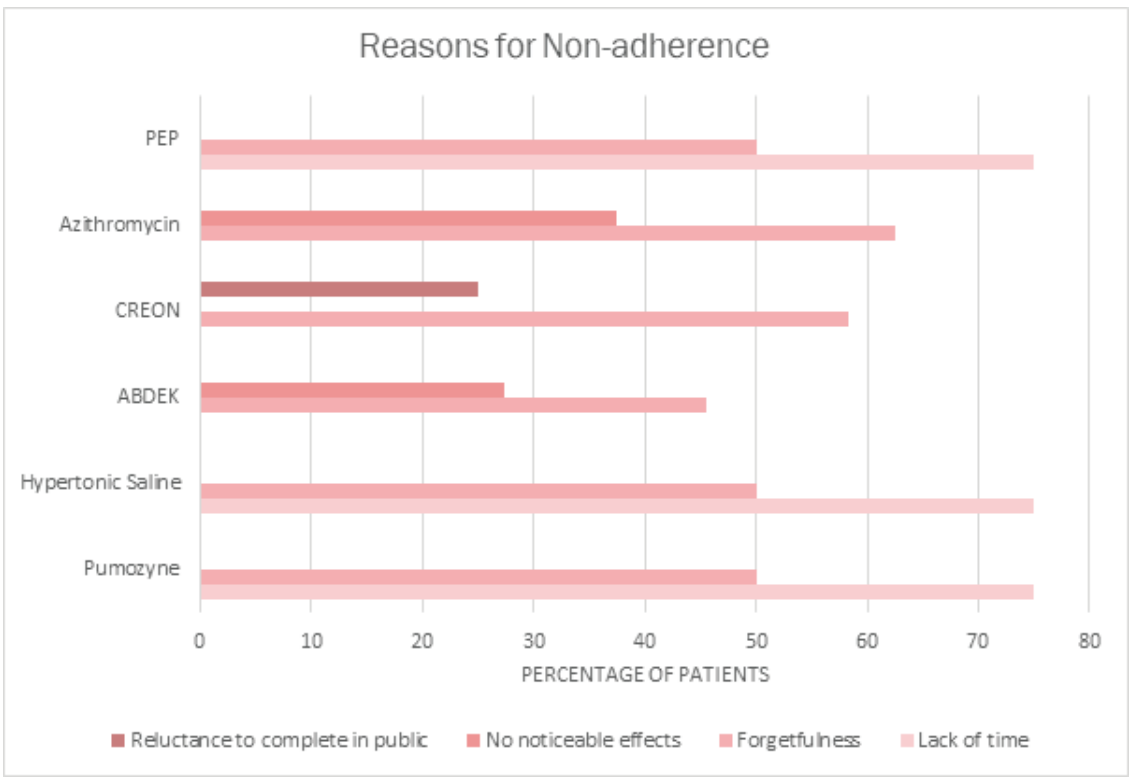

Figure 12 : Reasons behind missed treatments

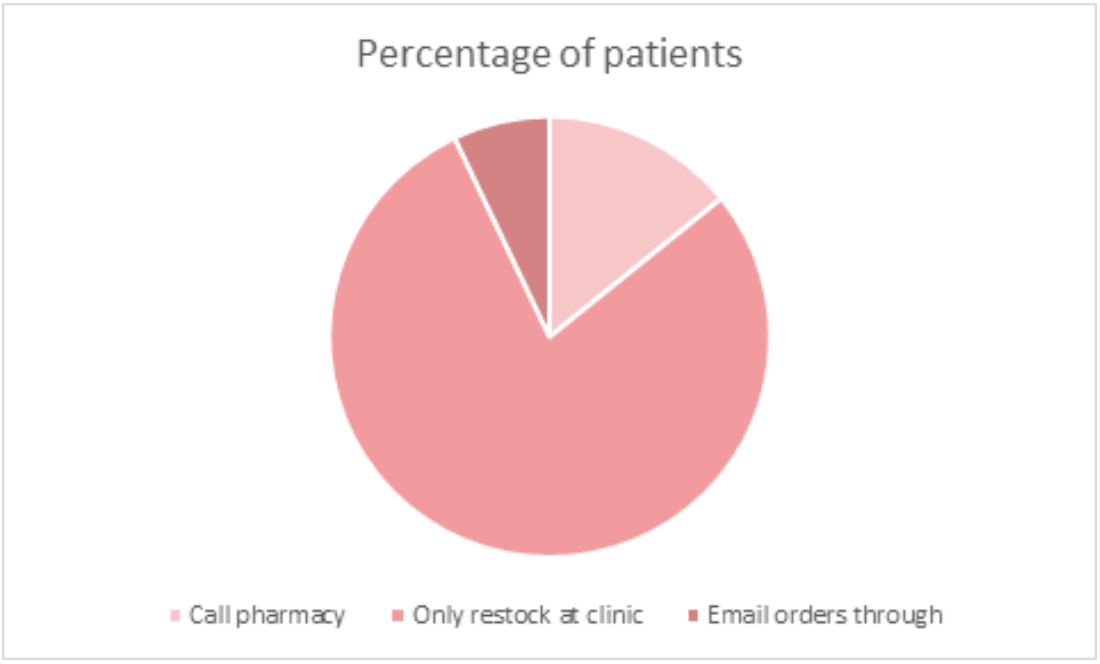

Figure 13 : Medication ordering methods

4.4.5 Treatment Adherence Attitudes

64.3% of respondents agreed that they are more likely to adhere to their treatment plans when they are feeling unwell, as opposed to the 57.2 % who are more likely to miss their treatment when they are feeling well (Figure 15).

78.5% of respondents agreed that they are more likely to complete treatments that have noticeable effects (Figure 16).
64.3% of respondents stated that they sometimes forget whether they have taken medication or not.

4.4.5 Clinic Appointment Attitudes

There was a relatively even distribution of responses rating the importance of attending clinic appointments. 42.9% of respondents rated attending clinic appointments as important, with 28.6% believing they are essential. On the other hand, 42.9% respondents rated clinic appointment attendance as not important, with 14.3% believing they are a waste of time.

Patients were, on average, more likely to adhere to treatment after attending clinic appointments (Figure 17).

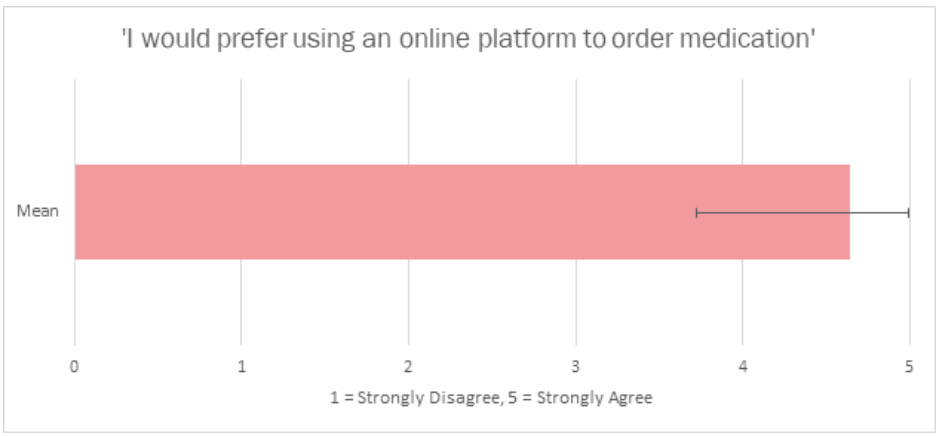

Figure 14 : Online medication ordering opinions

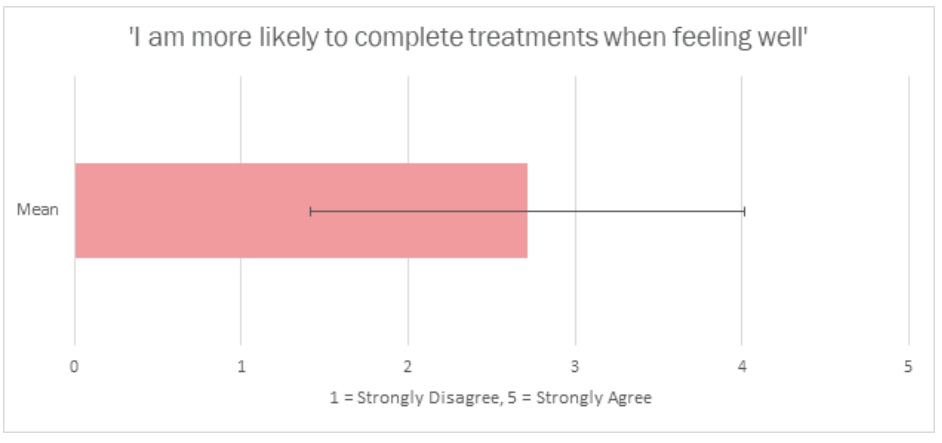

Figure 15 : opinions on treatment adherence when well

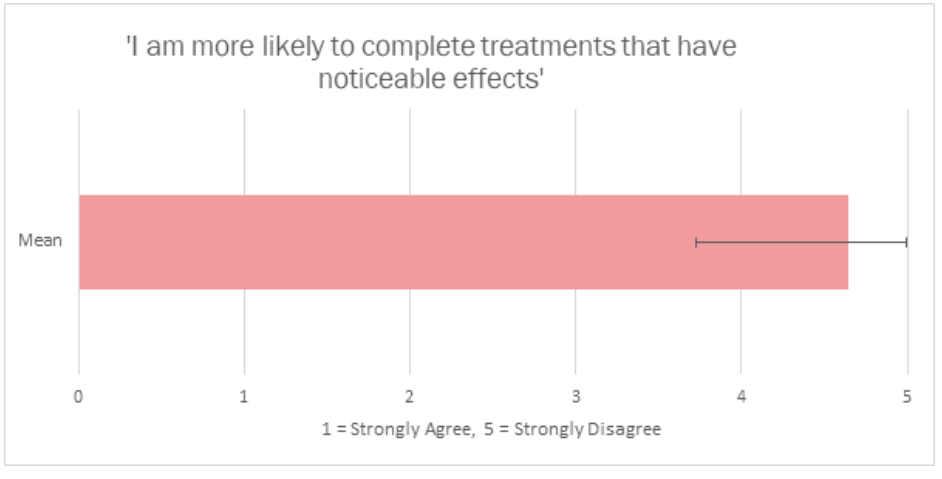

Figure 16 : Opinion on treatments with noticeable effects

Respondents were asked to state which 3 aspects of clinic appointments they believed to be the most important. The top 3 most common responses were the testing of lung function (92.9%), consulting with doctors (92.9%), and giving sputum and blood tests for analysis (64.3%) (Figure 18).

Recording low lung function results and coming into contact with other CF patients were the 2 most common responses to causes of anxiety in relation to clinic appointments, with figures being 71.4% and 42.9% respectively. The 3rd most common answer was a tie between travelling to and from hospital (28.6%) and being within a hospital environment during clinic appointments (28.6%).

50% of respondents either strongly agreed or agreed with the statement 'I feel more optimistic about my condition after attending clinic appointments'. Another 35.7% responded neutrally to this statement, while 14.3% disagreed. Similarly, 57.1% agreed with feeling more encouraged to complete their treatments after attending clinic appointments. Only 1 respondent (7.1%) disagreed with this statement, while 35.7% responded neutrally. Only 28.6% of respondents agreed with the statement 'I am able

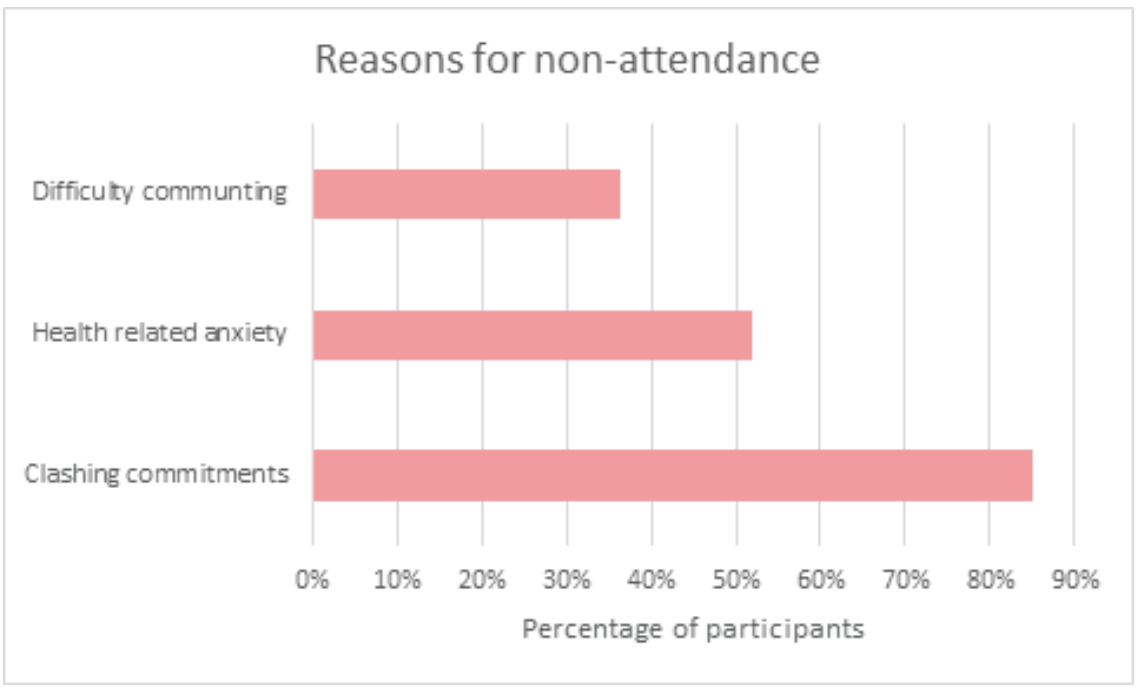

Figure 17 : Reasons for non-attendance at clinic

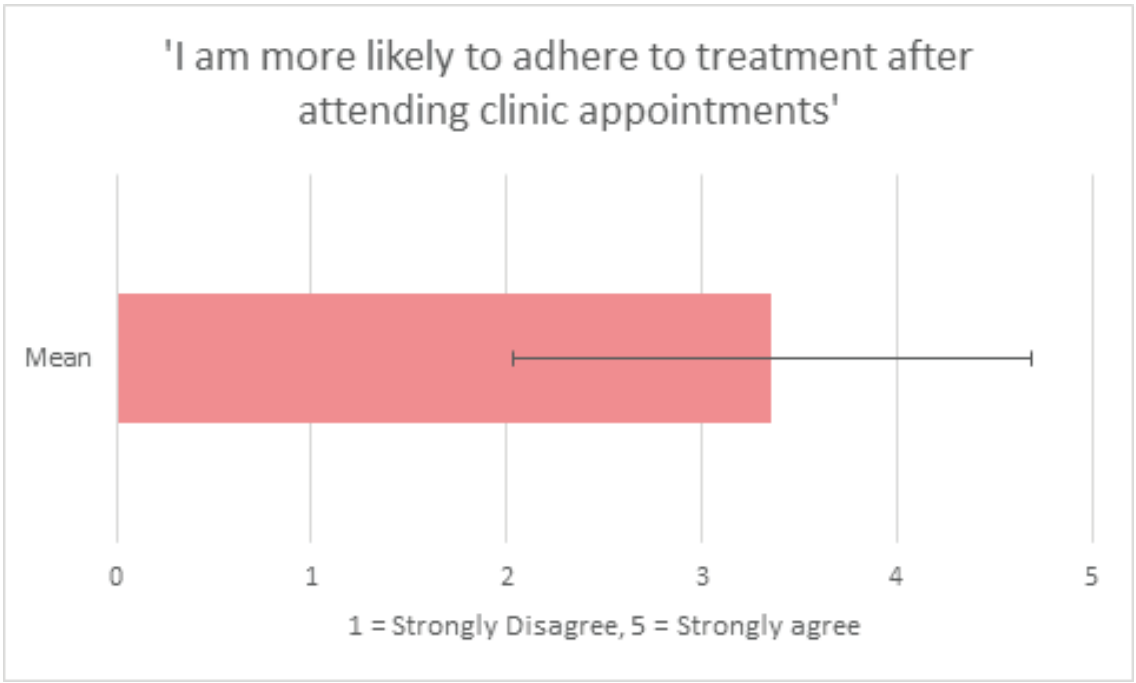

Figure 18 : Participant opinions on treatment adherence post clinic

4.4.5 Short Answer Response

Respondents were asked to provide a short answer response to the question 'If you could change one aspect of the clinic appointment process, what would it be?'. The full list of responses to this question can be found in the appendix (Figure X). Several trends were identified within the responses of this short answer question. The most common complaint, with 5 mentions out of the 13 respondents, centred around the length and timing of clinic appointments. Respondents stated that some appointments can take several hours to complete, with a significant amount of this time being spent waiting to see different specialists. Respondents suggested speeding up the process would result in greater levels of appointment attendance.

The next most common complaint, being mentioned by 4 of the 13 respondents, related to the risk of cross infection between CF patients while attending clinic appointments. The proximity between patients and the use of common equipment across multiple patients was stated as the most acute cause for concern, as individual CF patients may have tolerance to certain bacteria which others do not.

The application of a 'Telehealth' system for remote clinic appointment attendance was mentioned 3 times. The final complaint, mentioned twice, questioned the need to speak with every health care member at each clinic appointment.

The next section of the report will discuss these findings and identify areas for potential innovation.

opposed to the 57.2 % who are more likely to miss their treatment when they are feeling well (Figure 16).

78.5% of respondents agreed that they are more likely to complete treatments that have noticeable effects (Figure 17).
64.3% of respondents stated that they some-

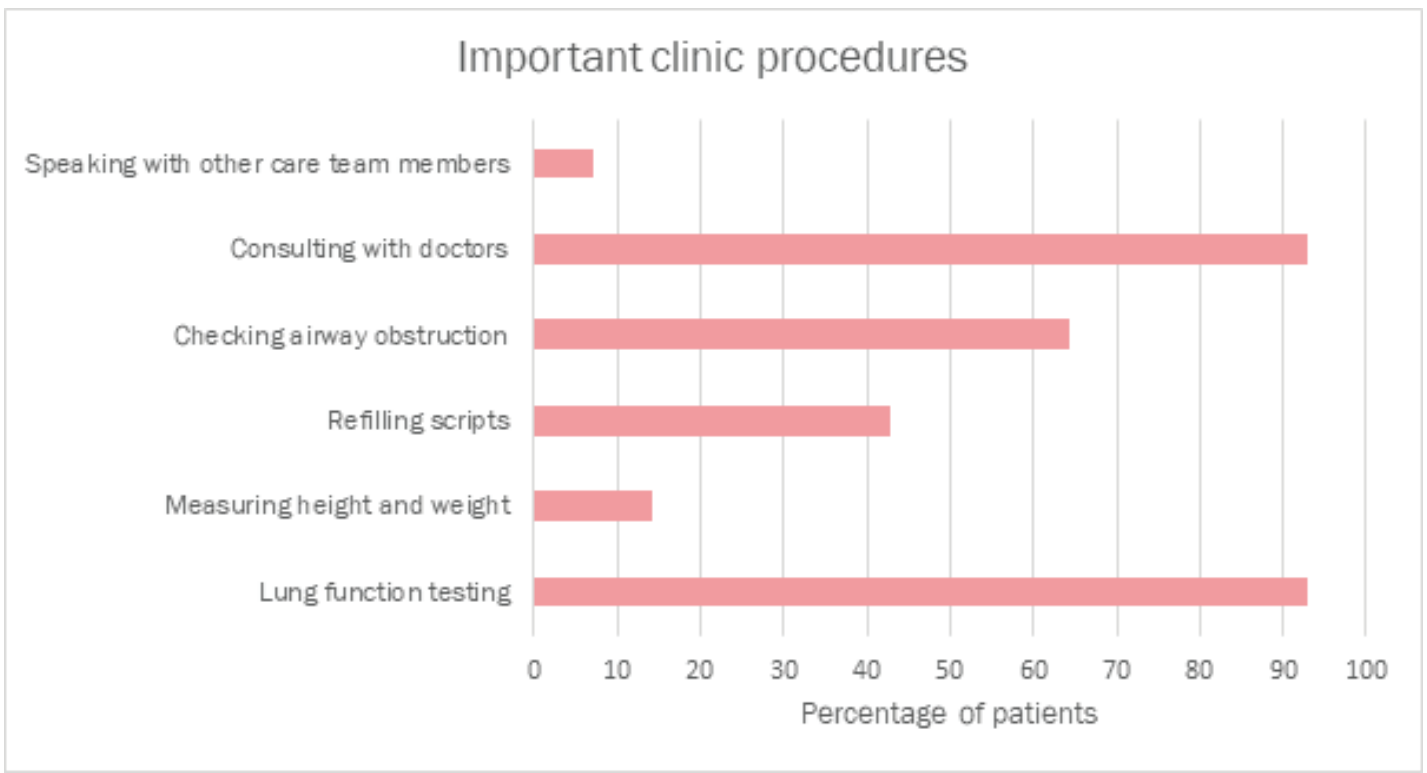

Figure 19 : Important clinic appointment procedures

DISCUSSION

DISCUSSION

Conducting primary research into CF patient treatment adherence has gathered information that both supports the existing literature as well as fills in some of the identified research gap. By analyzing and discussing these results, potential pain points surrounding CF patient treatment adherence can be identified. Once these problematic areas have been identified, proposals can be provided which aim to solve such issues.

5.1 Existing Framework

Dealing with CF is a long term commitment and requires organization and strong time management skills. Balancing CF with other life commitments is comparable to work/life balance, therefore some of the existing theoretical frameworks surrounding work/life balance can be used to justify the actions and consequences of CF patients and their treatment adherence.

The 'positive psychology' movement led by Seligman shifts the emphasis away from what is wrong with people, to what is right; focusing on strengths, not weaknesses (Morganson et, al, 2014). Seligman also laid the foundation for the theoretical framework known as 'learned helplessness' which explains how humans and animals can learn to become helpless and feel they have lost control over what happens to them. By encouraging CF patients from a young age to practice 'positive psychology' and limiting their exposure to environments that facilitate 'learned helplessness', a positive mindset may be instilled which manifests itself through a desire to adhere to treatment.

An example of this presented itself in P1's response to communicating with other CF patients:

'The doctors got me onto some chat rooms for people with CF but I found the people on those chatrooms were people that didn't share the same mindset as me – they were online just to dwell on the fact that they've got CF.... I believe those people made me question my positive mindset – mum had the same experience with other CF parents who would always focus on the negatives.'

As Scott & Barnes (2011) explain in relation to 'positive psychology'; '"putting on a happy face" won't necessarily make you feel happier, but putting in a little bit of effort likely will' (Scott & Barnes, 2011). This can be related back to CF treatment adherence and the importance of routine; ignoring is much less effective than making positive steps towards instilling routine and completing treatment (Figure 20).
Ipatients and healthcare teams, qualitative research methods were employed. Qualitative research is more flexible and interpretive than quantitative research. By conducting qualitative research, deeper knowledge into the reasoning and behaviour

5.2 Importance of Routine

The results of the interviews support Abbott's (2011), Ball's (2015) and Bragenballe's (2011) existing research on the importance of time management and maintaining a routine around completing daily treatments. Those participants who reported having set routines around treatment adherence appeared to have a more positive outlook on their condition. This could be attributed to the fact that their mindset was a direct result of their health, which may have been a direct result of their treatment adherence routines. An example of this can be seen in P1's statement:

'I found myself doing physio and nebs was something I'd do in the afternoon because that's when I'd get the most downtime after uni and before sport. All my tablets are taken in the morning/night when they need to be taken. The bulk of treatments are done in the afternoon...It's crap but if you've been dealt this hand of cards you've got to deal with it the best way possible.'

Which is in stark contrast to P2's response when asked about the existence of treatment routine:

'Nope... Probably my augmentin – the one I have when im really sick. Other than that I don't take my antibiotics anymore.'

The survey results show that the rate of non-adherence to chest therapy nebulized treatments, which generally take much longer than other forms of treatments such as consuming medication tablets, was higher across the response population (Figure 11). This is supported by the data within Figure 13, showing a lack of time as the main contributor to low adherence.

Typically PEP and nebulized treatments are conducted consecutively, resulting in a block of treatment than can last up to 40 minutes

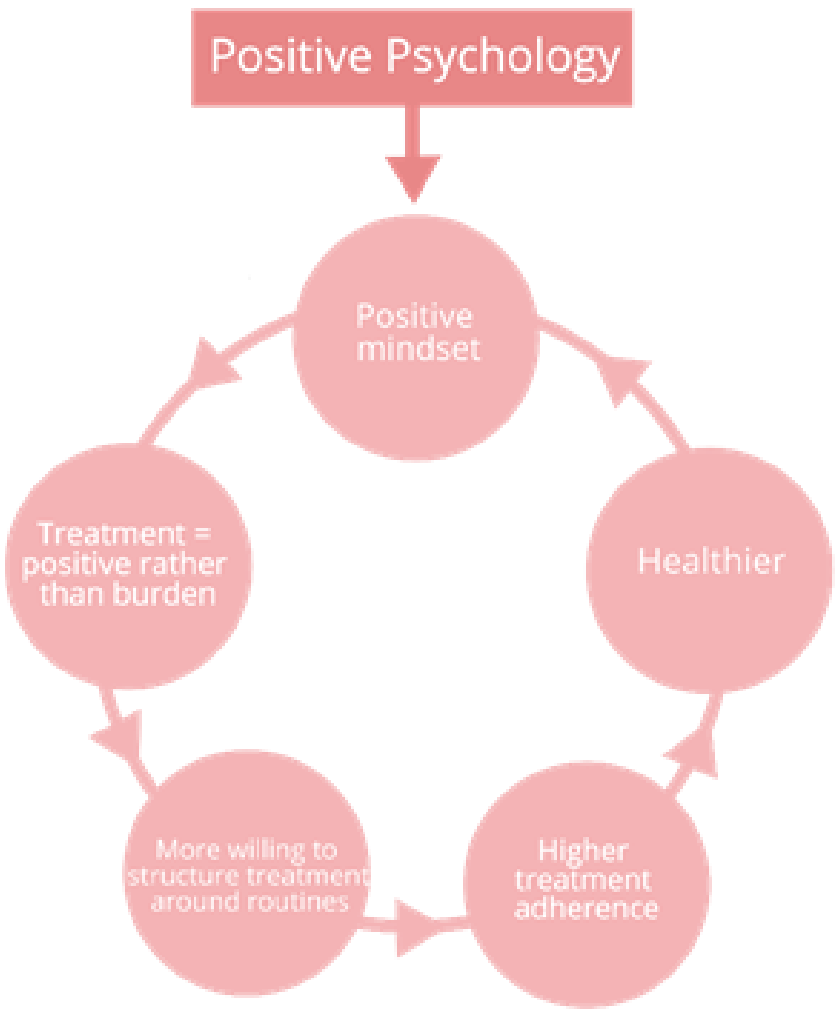

Figure 20 : Outcomes of 'positive psychology' (Author's illustration)

(Narayanan, 2016). This is a significant period of time to set aside for treatment each day and relates back to importance of time management and routine. An example of patients avoiding these treatments due to their time constraints appeared in P3's response to which treatment she found most difficult to stay on top of:

'I'd say probably the Hypertonic saline because it takes quite a while and I have to get a bit motivated to do it.'

While the results show that nebulised therapy has one of the lowest adherence rates, it is considered to be one of the most important treatments on a CF patient's treatment plan (Abbott. Et al, 2011). The evidence suggests that reducing the time it takes to complete chest therapy treatments may result in a higher rate of adherence, and thus an improvement in CF patient wellbeing. Adjustments could be made to the equipment (Figure 21, Figure 22) and/or processes that are used to deliver the treatment in order to shorten treatment completion times.

5.3 Online Platform

To order medication patients must manually call their pharmacy to check which scipts are available and to order their medication. This process can be time consuming and relies on the punctuality/accountability of each indivudual pharmacy. While the majority of patients currently call their pharmacy to check script levels and order medication (Figure 14), rurvey results show that patients would prefer to use an online platform (Figure 15). By integrating this process into an online system, other services like delivery of medication and auto script refilling could be included, streamlining the process of ordering and receiving medication.

Building an online platform could also establish channels of communication between CF patients. Interview responses suggested that while family members are important pillars of CF patient support networks, they can't fully relate to the struggles of living with CF. The ability to speak with other CF patients about their experiences provides significant value, as P3 stated during her interview response:

Figure 21 : Pari-Sprint Nebuliser (Pari, 2020)

*'Yeah usually when other people have an issue they might go 'hey have you gone through this? We have b**** sessions about treatments. It's nice to have people who understand what you're going through.'*

Due to the unprectable nature of CF, anxiety amonsgt CF patients around the progression of the disease and life expectancy is common. By establishing these communication channels between CF patients, they are given the opportunity to speak with older CF patients who can provide information and reassurance to those younger patients who may be scared of what the future holds. P1 explained during her interview how this experience helped her:

'There is an older CF lady who I have been in contact with who does a lot of CF advocacy stuff and I've been talking to her about how her day to day life is and stuff like that – keeping it real and not dwelling on it. I found that to be quite a positive thing.'

Along with medication ordering and CF patient communication, an online 'hub' could be used as an administration tool for health care teams to organise and remind patients when their next telehealth clinic appointment is. CF related news such as medical advancements and community events could also be displayed to patients, providing positive news to help patients practice 'positive psychology'.

5.4 Telehealth Clinic Appointments

The majority of answers in response to opinions on telehealth clinic appointments and online medication ordering services were positive (Figure 23). While telehealth appointments do currently exist, they are rarely conducted. All of the interview respondents had only recently attended their first telehealth clinic appointment due to the restrictions brought on by COVID-19. Interview and survey responses suggest that CF patients prefer these remote style appointments over traditional physical clinic appointments which are typically held at

Figure 22 : PEP chest therapy mask (Bronchiectasis,2020)

major hospitals. P3's response to the telehealth appointment outlined her preference:

'It was kind of good because I didn't have to worry about getting dressed, leaving at a certain time'

This information was not available within the current literature surrounding CF and is a valuable contributor to both the identified research gap as well as the improvement of clinic appointment attendance rates. As is previously stated, clinic appointments are an essential aspect of monitoring and treating CF (Narayanan, 2016). One of the main obstacles preventing the implementation of widespread telehealth clinic appointments is the access to the necessary fequipment used during clinic appointments.

Patients were asked to record the 3 most important aspects of attending clinic appointments. The results are found below in Figure 24.

The results of the survey seen in Figure 24 show that the 3 most important aspects of clinic appointments are:

1. Consulting with doctors

Consultation with doctors occurs after patients have recorded their lung function test. Usually the patient and their doctor discuss how their treatment plan is going and whether any adjustments need to be made due to changes in health or patient wishes. These consultations can be effectively conducted over remote connection.

2. Testing lung function

Lung function testing requires the use of a spirometer – an instrument which records how much air can be inhaled, and the rate at which and amount of air that is exhaled. Key spirometer measurements include: 'Forced Vital Capacity' (FVC) – the largest amount of air a patient can forcefully exhale after breathing in as deeply as possible – and 'Forced Expiratory Volume (FEV) – the

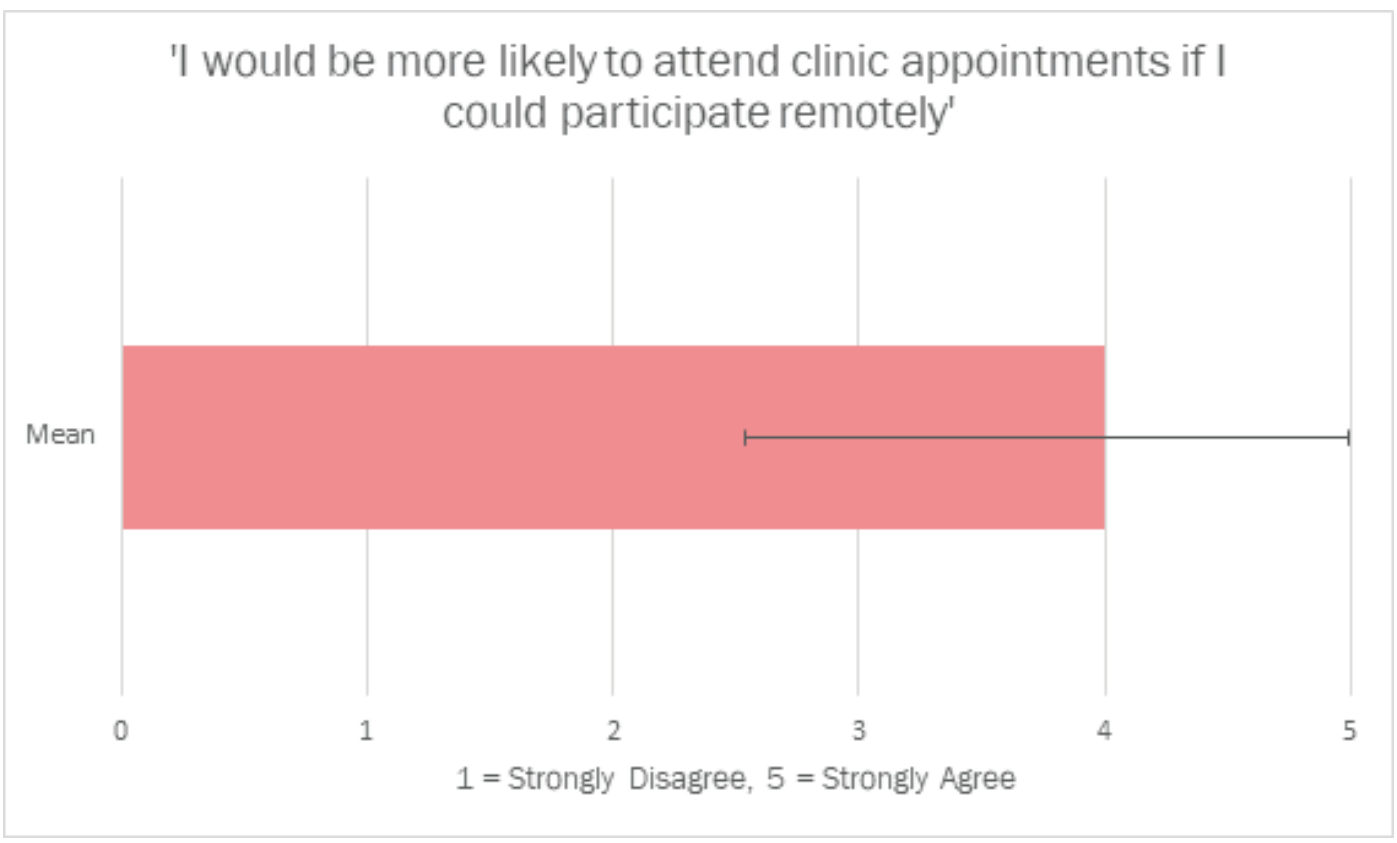

Figure 23 : Likert scale response

volume of air a patient can expel from their lungs in one second (MayoClinic, 2020). Lower FEV readings indicate more significant obstruction. While the CF clinics have the most accurate measuring equipment, there are portable alternatives which patients can use from home to measure their lung function. These portable spirometers are rarely used and can be expensive due to their lack of relevance, however with increased demand an opportunity exists to develop and design a cheaper, more accurate alternative to the products currently on the market. This would allow patients to provide doctors with their lung function numbers from home.

3. Checking airway obstruction

Doctors will often listen to a patient's chest via a stethoscope to check for any blockages ,or 'pops and crackles', within the lungs and small airways. While not an essential procedure, it is useful to identify any build ups of mucus that may require extra chest therapy or a round of antibiotics to clear. Doctors are specially trained to be able to identify certain pops and crackles when listening to a patient's chest through a stethoscope, which means patients would not be able to recreate this procedure themselves. However, an opportunity exists for innovation in this area; perhaps something like an electronic stethoscope which can listen to a patient's chest and identify any causes for concern.

By identifying these 3 essential clinic procedures and providing alternative solutions which can be conducted from home, the practicality of regular telehealth clinic appointments may be drastically increased. The survey research show that on average patients are more likely to complete treatments after attending clinic appointments (Figure 19). This could be attributed to either a response to a decrease in lung function, or an effort to sustain good health. By increasing clinic appointment attendance rates via conducting telehealth appointments, patient treatment adherence should improve. Conducting telehealth appointments also eliminates the risk of cross infection between CF patients, which was stated by respondents to be one of the main causes for concern when attending clinic appointments.

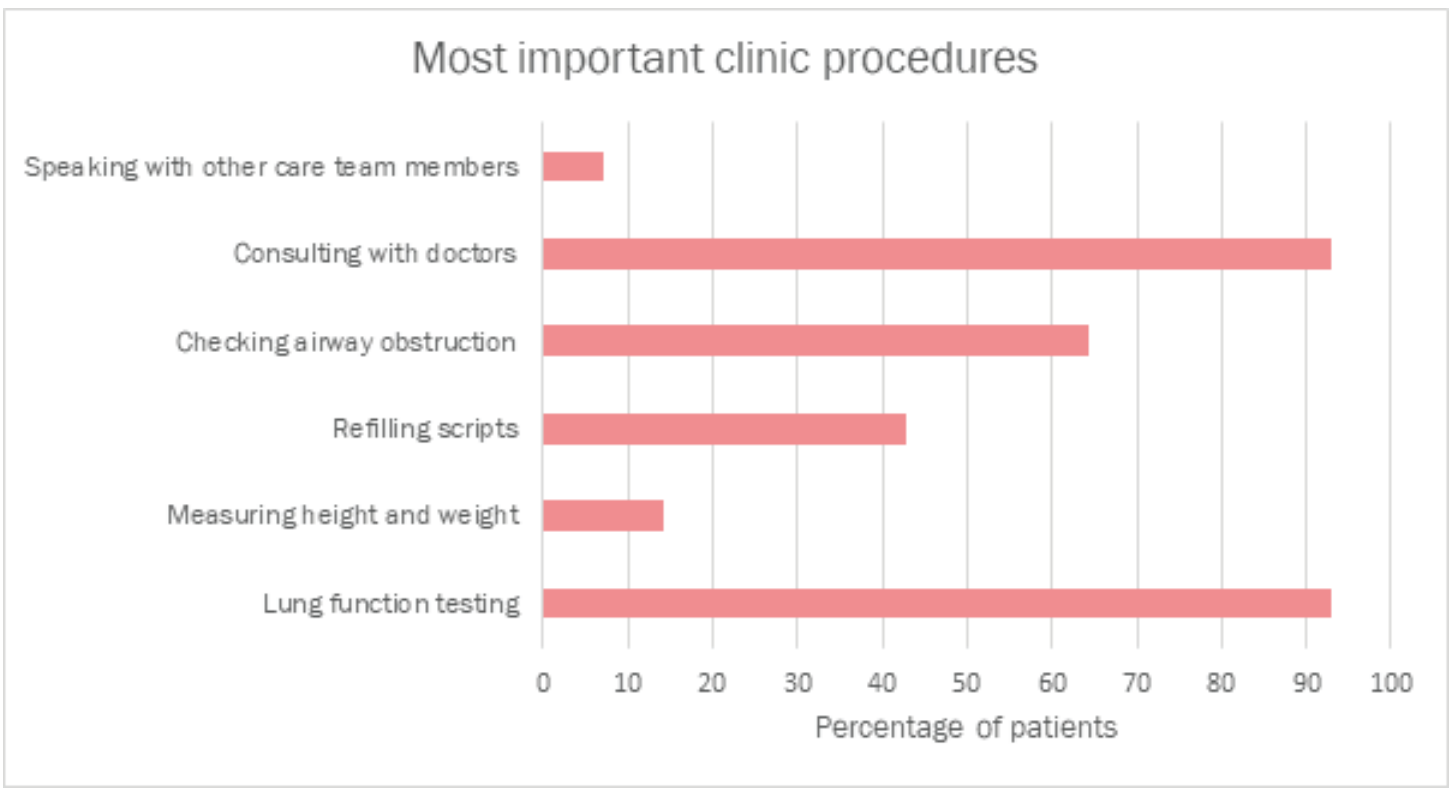

Figure 24 : Important clinic procedures

5.5 Poor Memory

It was identified through the literature review that poor memory may be both a cause and a result of low treatment adherence to certain medication. Survey results suggest that patients frequently miss treatments due to forgetfulness (Figure 13). This claim is supported by responses within the interviews, with P1 responding to a question regarding forgetting whether or not when she has taken medication:

'Oh all the time! Sometimes at home I forget if I've had them [CREON] and then take extra just in case.'

This evidence suggests patients forget to take medication completely as well as taking medication and then forgetting, taking more in the process. This presents two issues; one being patients often miss treatments altogether, resulting in non-adherence, while the second issue suggests patients may be consuming more medication than they're prescribed, resulting in potential health issues. While previous literature has identified forgetfulness as a contributor to low treatment adherence (Abbott et. al, 2011), the knowledge that forgetfulness may be causing patients to over-medicate is new information.

A solution to this problem may involve patients storing medication in some sort of dispenser which reminds them when treatments are due as well as informing them on which medications they have already taken.

Now that the results have been discussed and areas on, the next section of the report will focus on providing recommendations on potential design solutions.

RECOMMENDATIONS

The following design recommendations are made in response to the problem areas identified in the previous section of the report. An explanation, along with potential limitations, of each recommendation are included with a related page of sketches to illustrate the ideas discussed.

6.1 Online CF Hub

An online platform for CF patients (Figure 23) would act as a hub from which they could conduct various actions. Survey and interview results suggested that patients would prefer to check their script availability and order medication via an online service, rather than ordering over the phone as is traditionally done. The proposed online platform would have a personal medication monitoring feature, from which patients could order and track their medication stock.

The research responses also suggests that patients would benefit from communication between themselves and other CF patients. The online platform would facilitate these connections, allowing patients from across the globe to share their experiences with one another and promote positive psychology.

The platform would also provide communication channels between patients and health care teams. A calendar/reminder function would keep track of various patient appointments, including traditional clinic appointments as well as telehealth clinic appointments. By integrating towards a streamlined online service, patients could structure their appointment times around their own schedules, as this was stated to be one of the main contributors to low clinic appointment attendance (FIGURE X).

Accessibility to the online platform for all CF patients may prove as a limitation, however as technology become further integrated into our lives, the probability of CF patients having no internet access is very low.

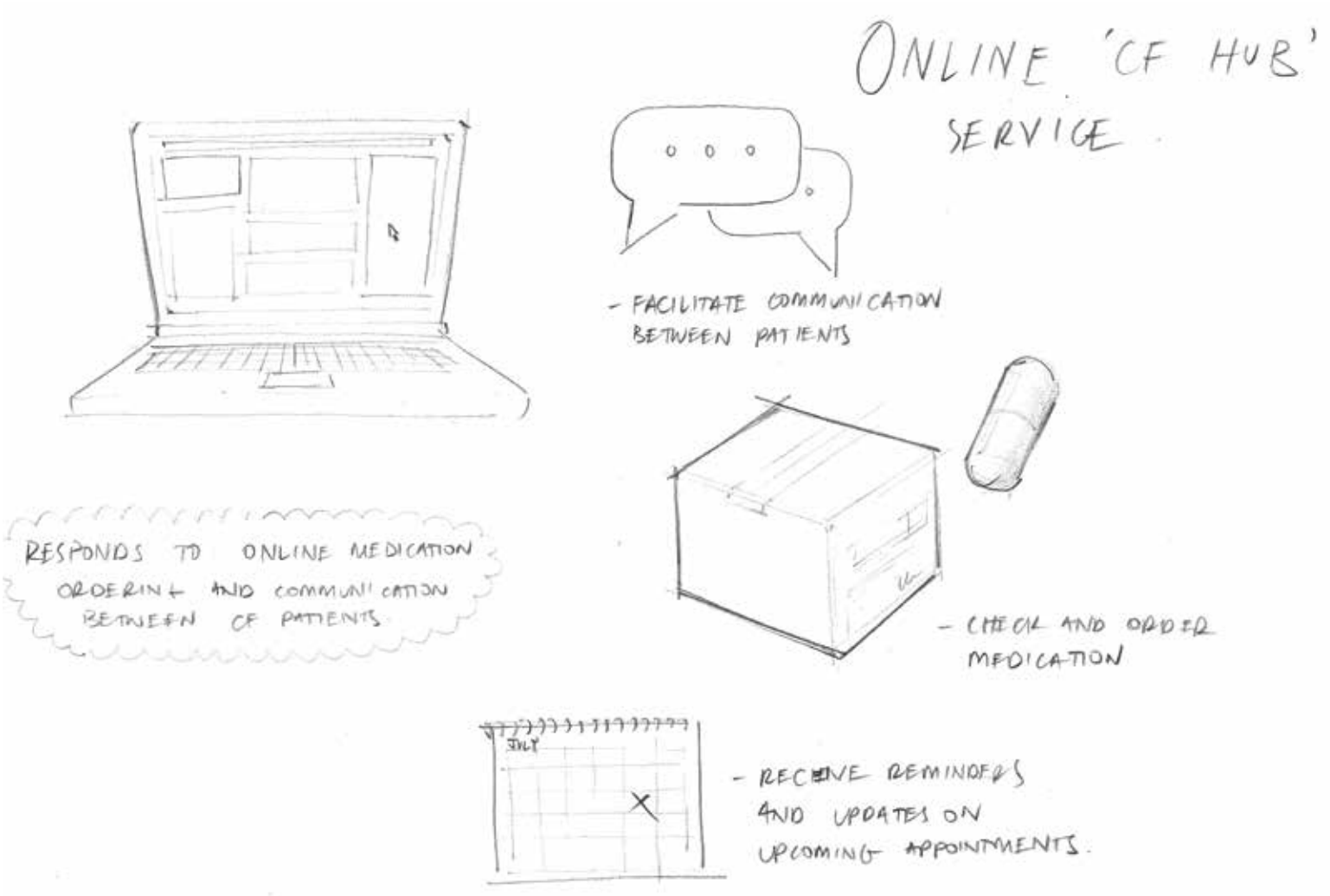

Figure 23: Online CF Hub proposal (Author's own illustration)

6.2 Personal Spirometer and Stethoscope

Survey results show that the majority of CF patients would rather attend telehealth clinic appointments over traditional clinic appointments. Telehealth appointments provide patients with a more convenient alternative to regular clinic appointments, while eliminating the risk of cross-infection between CF patients.

One of the major obstacles preventing widespread and regular telehealth clinic appointments is the availability of equipment required to monitor patient's health effectively. Without specific testing instruments, doctors are unable to make an informed evaluation of a CF patient's condition, resulting in the potential for mismanagement of treatment and treatment plans.

The most important procedures that occur during clinic appointments are consultation with doctors, lung function testing, and airway obstruction tests. While doctor consultations can occur efficiently over video call, the latter two procedures require specific equipment. By designing a user-friendly, affordable and portable alternative to the equipment used within clinic environments, CF patients will be able to comprehensively monitor their own condition and provide accurate updates to their doctors during telehealth consultations. Limitations of this proposal may include the price and availability of the technology required, however with some dedicated design and thought process a viable solution could be achieved.

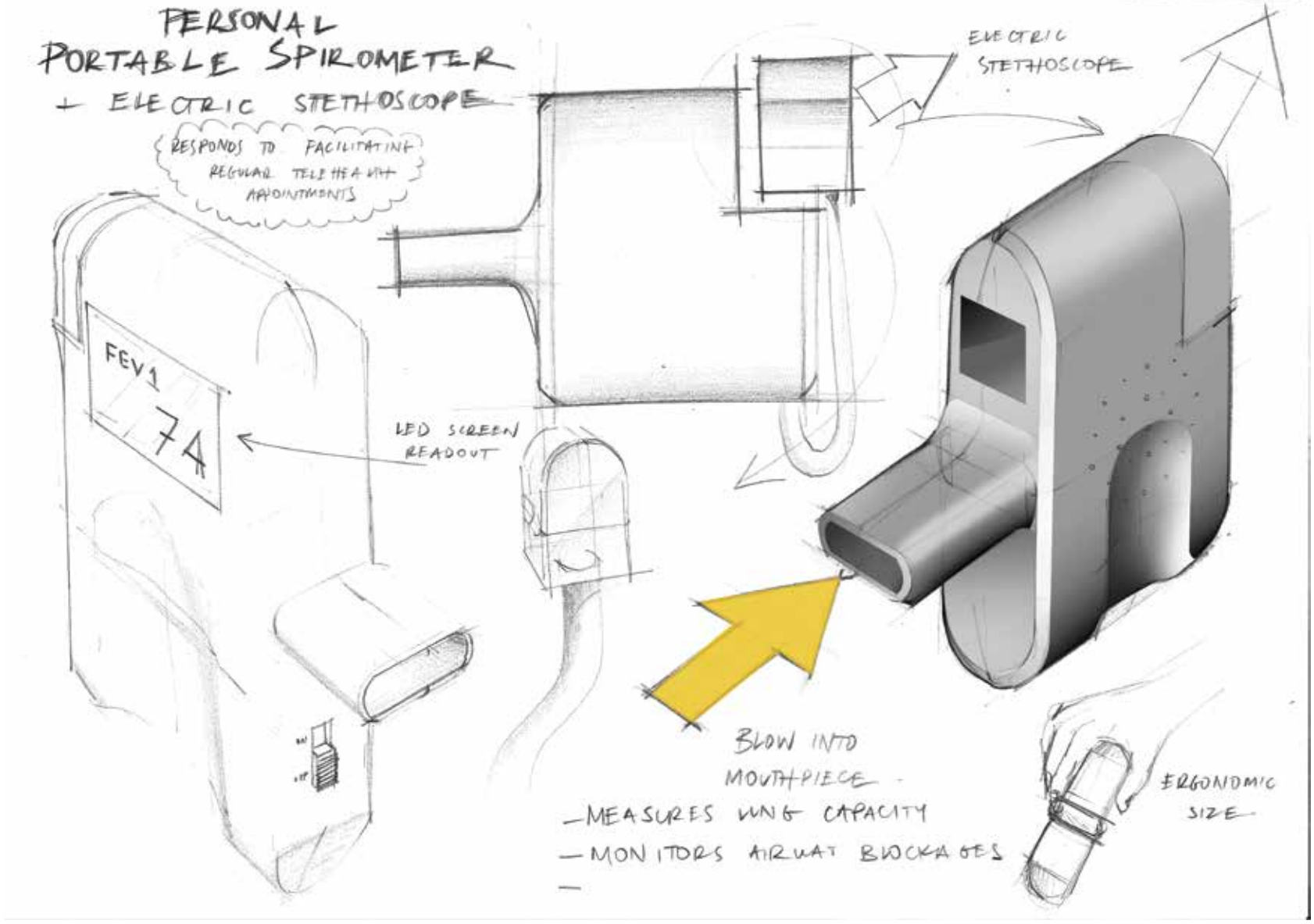

Figure 24: Spirometer/electronic stethoscope proposal (Author's own illustration)

6.3 PEP/Nebulizer Integration

A lack of time to fit one's treatments around their daily schedule is the main contributor to missed PEP and nebulizer. When completed one after another, as is recommended by doctors, PEP and nebulizer treatments can take upwards of 40+ minutes each day. By combining these two treatments into one, a significant amount of time can be saved (Figure 25). Reducing the time required to complete both treatments will result in patients being more inclined to adhere to these treatments.

In order to combine PEP and Nebulized treatments, a system must be designed which both treatments to be integrated into one functioning piece of equipment. Both treatments require patients to inhale and exhale. The similarities between how patients execute these treatments means they are able to be combined while still providing the benefits that they would if they were completed separately.

As PEP and nebulizer treatments are considered amongst the most important treatment on a CF patient's treatment plan, it is essential that they adhere to these treatments as often as possible. By reducing the time it takes to complete these treatments, patients will benefit from the increased adherence.

Limitations may include successfully integrating the two pieces of equipment, however a PEP mask already utilizes a two way valve which could be modified to allow the inhalation of nebulizer vapours.

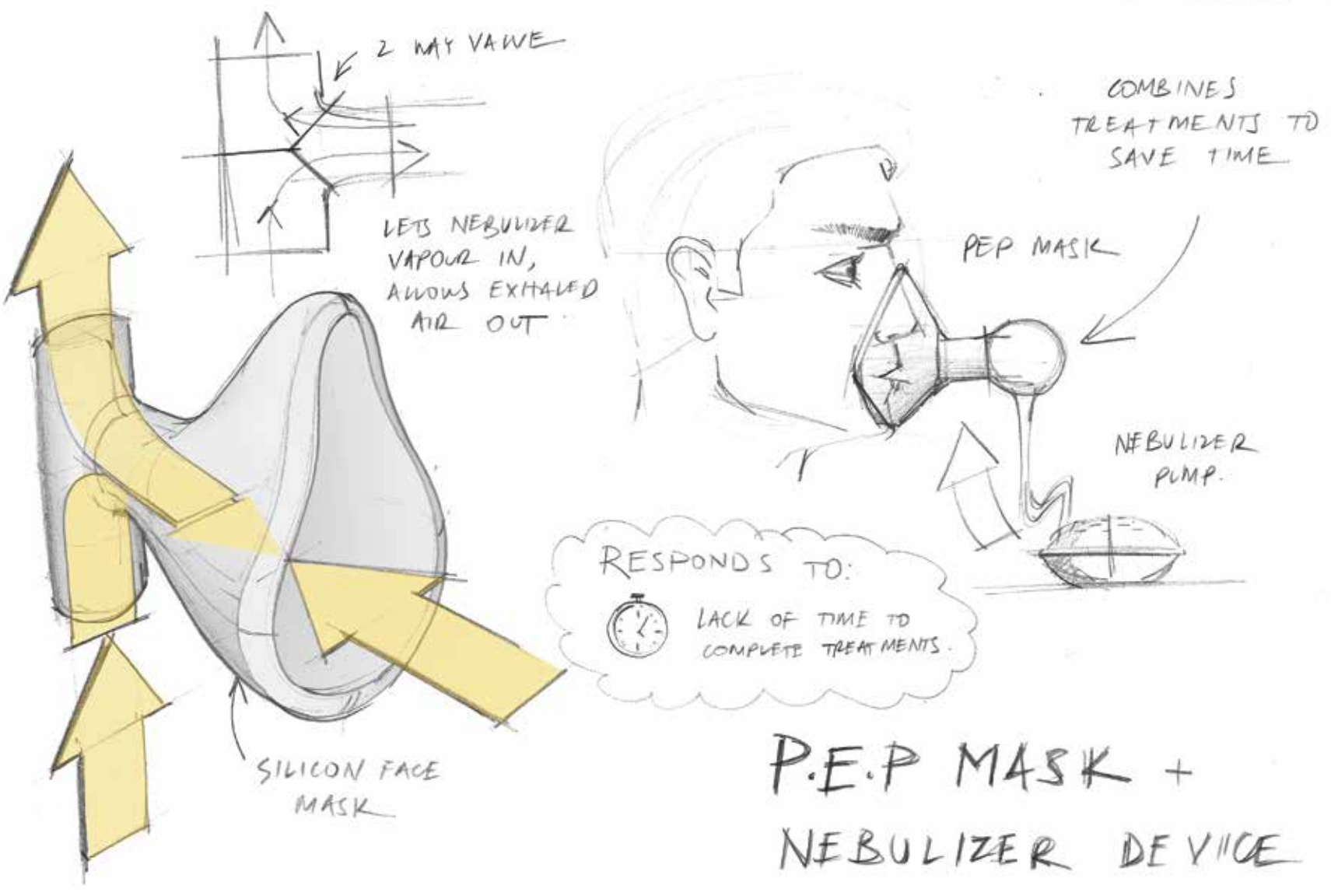

Figure 25: PEP mask/nebulizer combination (Author's own illustration)

6.4 Treatment Dispenser/Reminder

Interview and survey results suggest that forgetfulness is a major contributor to missed treatments, especially treatments which are delivered in tablet form. Having a portable piece of equipment that could dispense and remind users when their medications are due, would reduce the chances of patients forgetting their treatments, and thus increase treatment adherence (Figure 26).

Patients also reported that they would be more likely to complete treatments that have noticeable effects. Monitoring features could be included which keep track of how often treatments are taken/missed. This information could be used to provide an overview of a patient's health and treatment adherence. A log could be kept and projected against a model of the progression of a patient's condition. If patients could view a visual representation of the relationship between their treatment adherence and their lung function figures, perhaps they would be more likely to adhere to treatments they otherwise would have neglected due to a lack of noticeable effects.

Limitations of this proposal could include issues around keeping medication stored in a sterile environment. Another limitation may relate to accurately keeping track of when and how patients are consuming their medication.

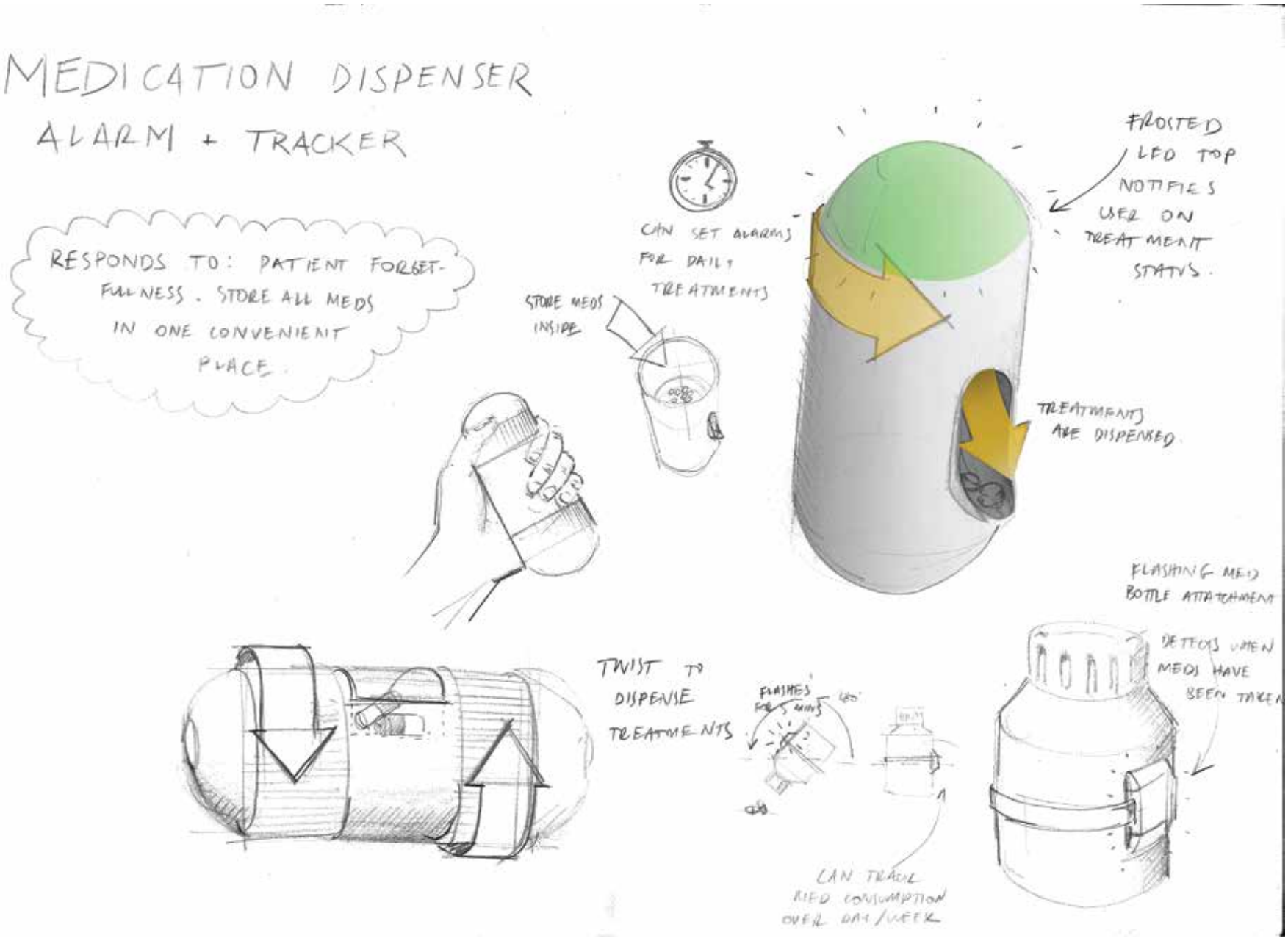

Figure 26: Medication dispenser/reminder (Author's own illustration)

DESIGN PROPOSAL

INTRODUCTION

Cystic Fibrosis is a chronic genetic lung disease that effects roughly 1 in 7000 Australians. Patients must adhere to lengthy treatment plans in order to keep well, often requiring 2+ hours of treatment a day along with 30+ tablets. Treatment adherence has been identified as an issue within the CF patient population, and the aim of this proposal is to provide a solution which ultimately increases treatment adherence.

An important aspect of monitoring CF is the attendance of clinic appointments. Patients report of increased treatment adherence post clinic appointment attendance. At these appointments patients measure their height and weight, lung function performance as well as having their chest listened to by a respiratory specialist via a stethoscope (Figure X). The results of these tests are examined by several specialists and adjustments to each patient's treatment plans are made. Attendance at these clinic appointments are crucial in monitoring a patient's health in order to catch any infections before they do sustained damage to the patient's lungs.

'Telehealth' appointments are clinic appointment conducted over video call. Telehealth appointments provide CF patients with a convenient and safer alternative to traditional clinic appointments which are typically held within hospitals where cross infection between CF patients can occur. In order to facilitate regular telehealth appointments, patients require the ability to detect airway blockages as well as test and monitor their lung function performance.

7.1 Design Intent

This design proposal aims to give CF patients the ability to monitor their own lung function figures and airway blockages from home. By providing patients with these abilities, patients can keep track of their condition from home and implementing widespread telehealth clinic appointments will become a realistic option.

7.2 Design Objectives

In order to achieve the aims which have been set out, the design must fulfill specific objectives:

1.Allow patients to independently measure their respiratory health.

2.Store this information for instant and future recall.

3.Have remote interaction capabilities to allow respiratory specialists to view test results

4.Ultimately improve CF patient treatment adherence rates

7.3 Design Justification

This proposal will benefit the lives of CF patients, their family/caretakers, as well CF health care team members. Gaps within the previous literature relating to clinic appointment attitudes have been investigated and valuable data has been collected. Responses to an online questionnaire (Figure X) suggest that on average patients are more likely to adhere to treatment after attending a clinic appointment.

This increase in treatment adherence could be attributed to a response to low lung function performance testing, or conversely an effort to sustain good health. Furthermore, interview and questionnaire results suggest that patients would be more inclined to attend telehealth (remote) clinic appointments via video call rather than traditional physical clinic appointments (Figure X).

Making clinic appointment attendance more convenient (Figure X) and reducing the time required for appointment attendance means patients are more likely to adhere to treatment (Figure X) along with facing less of a mental burden in relation to CF. A more positive outlook on their condition results in a happier patient, along with a happier household. Higher clinic attendance rates also make the job of healthcare team members easier, as they can effectively monitor a patient's health and make adjustments to their treatment plans accordingly.

7.4 Design Context

Respiratory monitoring equipment (lung function testing and airway blockage detection) such as what's being proposed within this proposal can be used year-round and not exclusively during tele health appointments. This gives patients the ability to constantly monitor their own condition from the comfort of their own home, enabling them to catch infections in their infancy and ensure optimal health.

7.5 Design Criteria

To achieve the objectives stated above, the design must adhere to a list of key design criteria. This list is a tentative outline of key design criteria, and it is expected to be altered and expanded upon as the project continues.

7.5.1 Function

Must accurately measure patient lung function parametrics (FEV1, FVC, predicted FEV1%)

Must accurately listen to and record 'pops and crackle's within patient's airways

Must be able to detect airway noises which may be cause for concern

7.5.2 Usability

Must be easy to use and simple to understand

Must have an easy to read user interface display

Must be able to be used by a range of different demographics

Should be easily cleaned

7.5.3 Ergonomics

Must be small enough to comfortably be held by single user

Must not be heavy and bulky

7.5.4 Form

Should be aesthetically pleasing

Should integrate modern design philosophies

7.5.5 Sustainability

Should be designed with sustainable practices in mind. This includes the use of sustainable materials and manufacturing methods.

Should integrate reusable and recycled parts where applicable

7.5.6 Communication

Should have the ability to send results to hospital servers for doctors to analyze

7.5.7 Technology

Must integrate spirometer technology to accurately measure FEV and FVC readings

Should integrate highly sensitive resonator and microphone to detect internal airway noises

Should be disconnect-able from mains power and become wireless while in use

7.5.8 Feedback

Must provide visual feedback on test results

Should provide haptic/audio feedback

7.5 Design Direction

Initial ideas for this proposal involve integrating a hand-held spirometer and an electronic stethoscope into one cohesive product. The spirometer will use existing tech to measure patient's lung functions performance, while the electronic stethoscope may require some further design thinking.

Initial thoughts on the stethoscope include using a highly sensitive microphone to pick up small noises within patient's airways which may be caused by airway blockages. In order to identify which noises are cause for concern, the stethoscope will be able to compare what it's currently picking up against a large database of noises that have been recording during patient consultations.

As the project develops further development and refinement will be made in relation to this design direction and initial design concept. While as a whole the package and some of the features of the end product may differ from what is presented in this design proposal, the ultimate end goal for this product is enabling CF patients with the ability to monitor their health from within their own home, providing an incentive to maintain high treatment adherence.

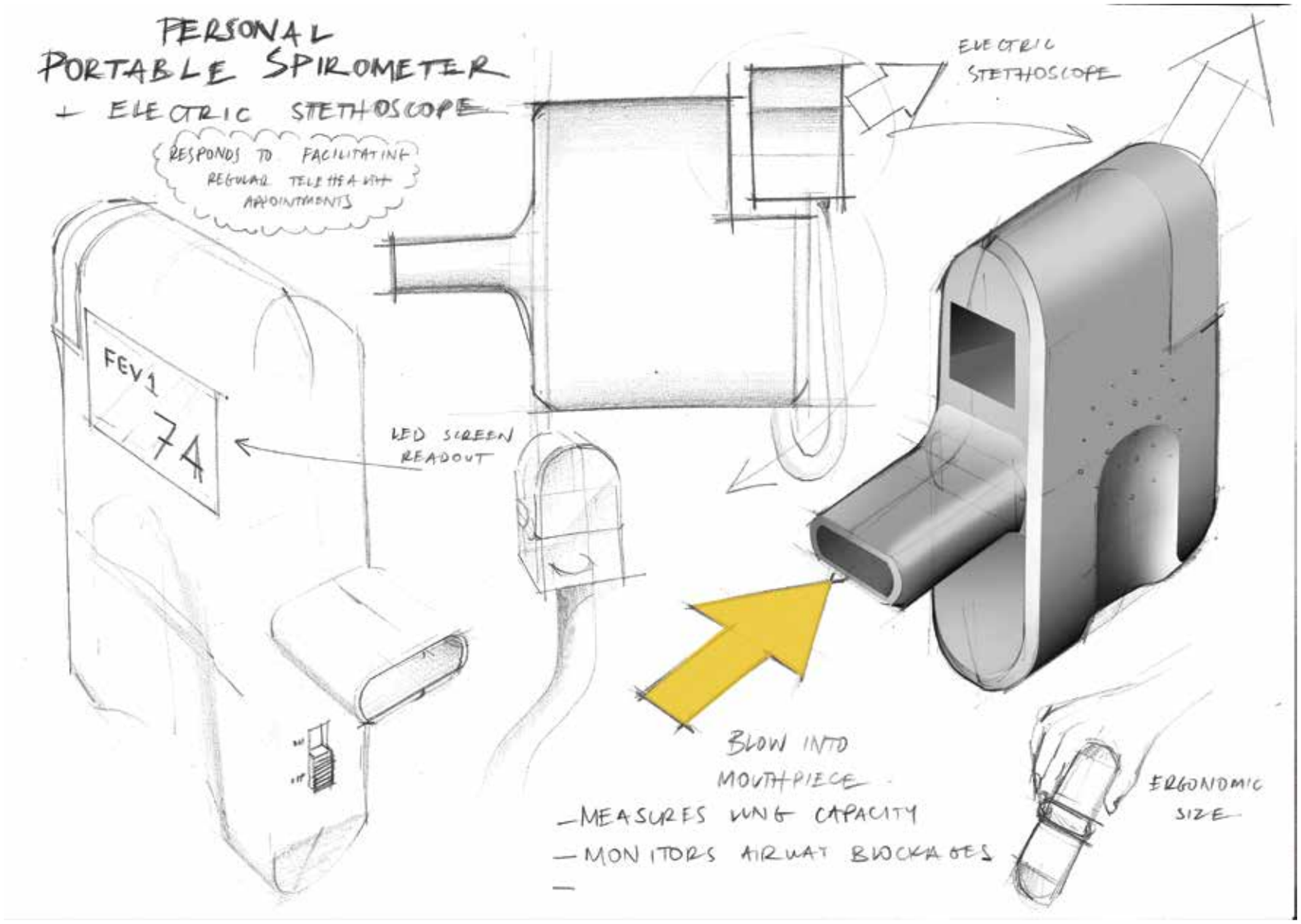

Figure 24: Spirometer/electronic stethoscope proposal (Author's own illustration)

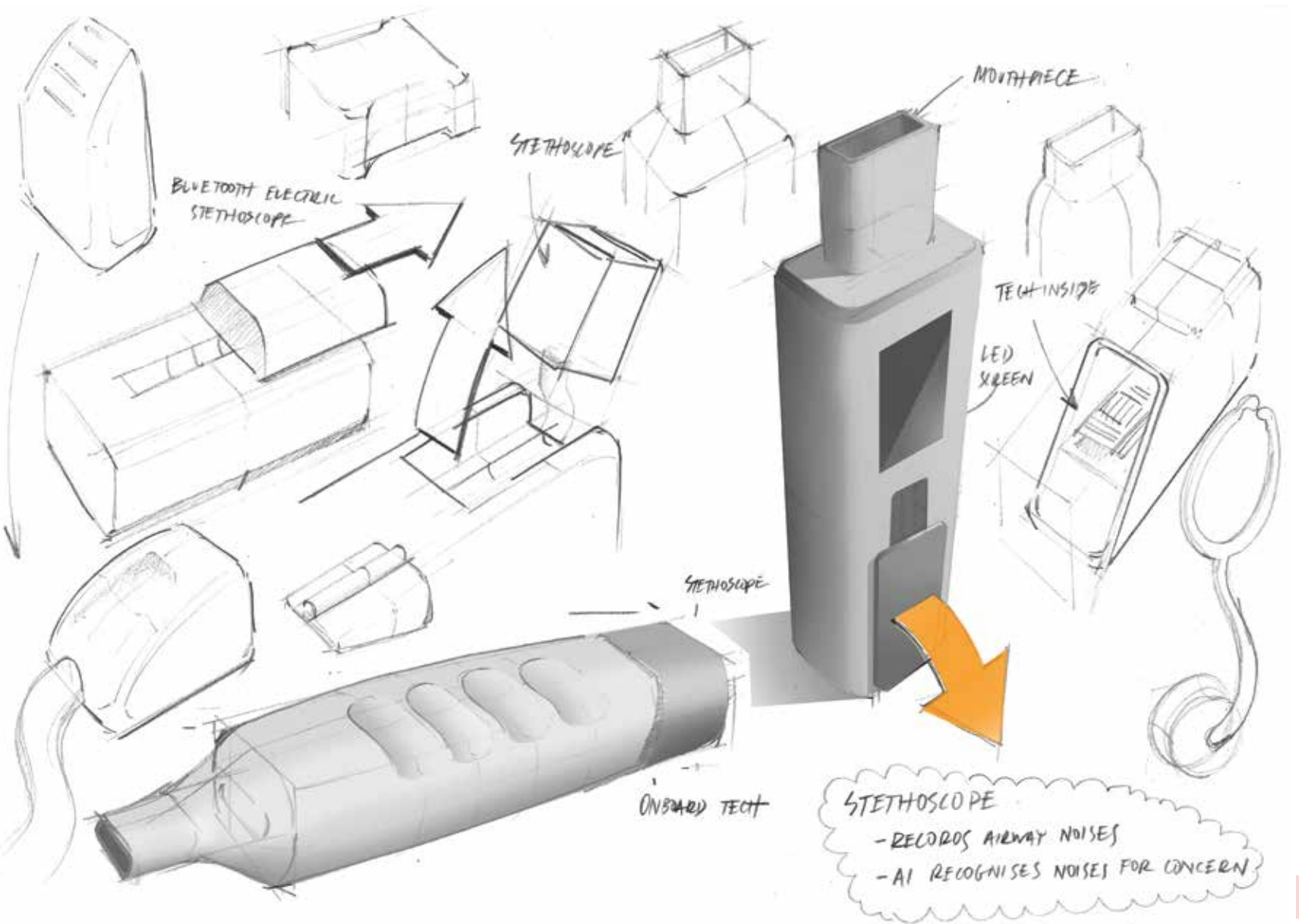

Figure 27: Spirometer/electronic stethoscope proposal (Author's own illustration)

SEMESTER SCHEDULE

PHASE	WEEKS	TASKS
1	1 2	INITIAL CONCEPT IDEATION CONCEPT MODELLING
2	3 4 5	USER RESEARCH CONCEPT DEVELOPMENT PROTOTYPE DEVELOPMENT
3	6 7 8	USER TESTING CONCEPT REFINEMENT BRAND IDEATION
4	9 10 11	MANUFACTURING RESEARCH PROTOTYPE REFINEMENT USER TESTING REFINEMENT
5	12 13 14	CONCEPT FINALISATION RESEARCH & PRODUCT SYNTHESIS

DESIGN JUSTIFICATION

INTRODUCTION

In order to facilitate clinic appointments remotely via video call, a device had to be designed which allows CF patients to test their lung function parameters and lung health from home. By designing such a device patients would be more likely to attend these crucial clinic appointments, and according to the research outlined earlier in this report, as clinic appointment attendance increases, so too does treatment adherence. Providing CF patients with this ability, their levels of treatment adherence should hopefully increase across the board, and with that, an increase in their quality of life. This device is the *AUSPIRO (*Figure 28*).*

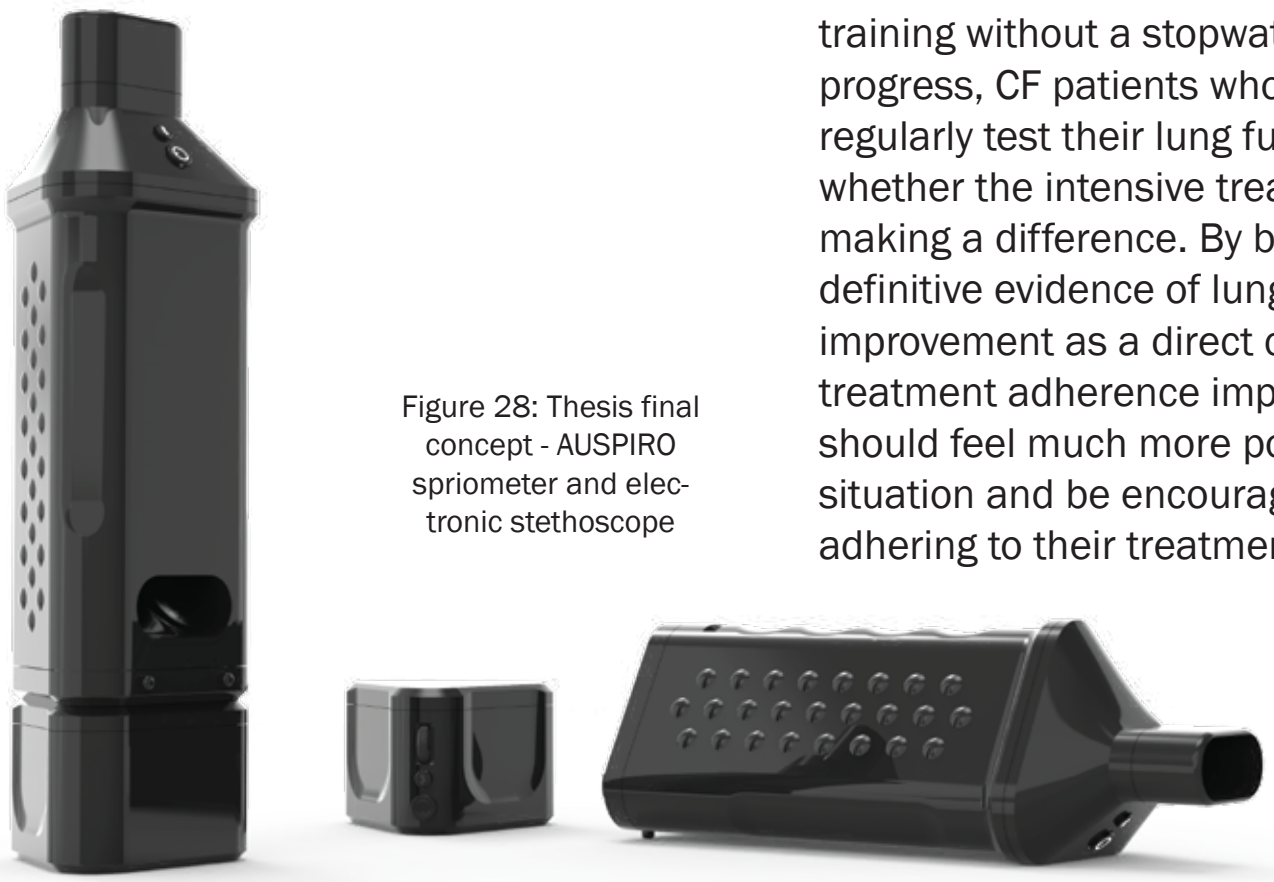

Figure 28: Thesis final concept - AUSPIRO spriometer and electronic stethoscope

FURTHER RESEARCH

In order to effectively and efficiently design a device for the young adult CF population, it was important to survey the current personal spirometry device market. By conducting research into potential competitors, I was able to gain an understanding of the price-points of competitor devices as well as the technology which is currently in use.

In addition to conducting research into existing products on the market, I also explored the idea of integrating an electronic stethoscope into the device. Upon researching the possibilities of this, I came to the conclusion that it was indeed possible to develop an electronic stethoscope capable of recording one's airway noises and uploading this recording to a database, where it would be analyzed by AI technologies and forwarded to the patient's specialist team for review.

Some further research was also conducted into the psychology of positive reinforcement and taking ownership of one's condition. Similar to a track runner completing lap training without a stopwatch to record their progress, CF patients who are unable to regularly test their lung function can't track whether the intensive treatment plans are making a difference. By being able to see definitive evidence of lung function improvement as a direct correlation to treatment adherence improvement, patients should feel much more positive about their situation and be encouraged to continue adhering to their treatment plan.

CONTEXT

The context of this project is based around how Cystic Fibrosis patients respond to clinic appointments and whether providing them with a device that allows them to test their lung function from home has an impact on their decisions around treatment adherence. In this context, Cystic Fibrosis patients are the main stakeholders, with the additional actors being the specialists within the patient's health care team. Family/friends of the CF patients may also be a part of this scenario, as an improvement in quality of life can have large run on effects to a home's general happiness and atmosphere.

The device was designed to be used outside of the typical hospital setting, whether that be within the patient's home, a friend or family members home, or outdoors. The ability to still attend clinic appointments remotely no matter the patient's location is what makes this device a useful tool in the journey of a Cystic Fibrosis patient.

DESIGN PROCESS

The basis of the design process of this project was the Double Diamond method. The design process of this project began with identifying the design opportunities related to treatment adherence through primary and secondary research. Once the user group had been researched and the problems and design opportunities were identified, the process could shift towards resolving the technical, ergonomic and aesthetic vaues of tyhe device.

Once the key functions of the device were known, the technological requirements and components needed to run the device were identified through secondary research.

Once the technical side of the device was resolved, the design process could move on to working through the form and ergonomic qualities of the *AUSPIRO*. Initial sketches and rough quick form models resulted in the decision to continue down the path of a

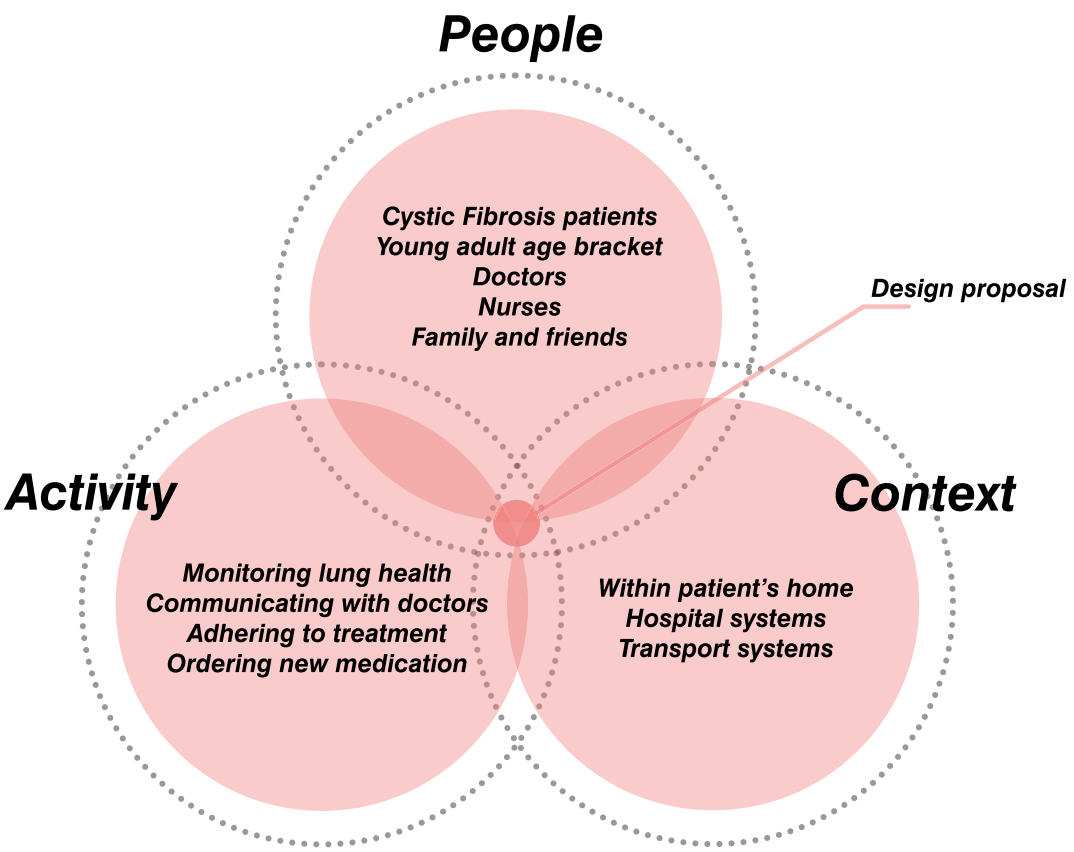

Figure 29: P.A.C

triangular form. User testing receiving feedback on the grip comfort of the device led to these decisions.

During the form ideation process 3D modelling tools were used to gain a better understanding of the necessary proportions required for the device. Taking advantage of the resources on offer, 3D printing allowed access to rapid and invaluable modelmaking. Using these models allowed for greater inspiration along the form resolution process, as well as providing a display during final presentations conveying the technical elements of the device.

DESIGN VALIDATION

The context of this project is based around how Cystic Fibrosis patients respond to clinic appointments and whether providing them with a device that allows them to test their lung function from home has an impact on their decisions around treatment adherence. In this context, Cystic Fibrosis patients are the main stakeholders, with the additional actors being the specialists within the patient's health care team. Family/friends of the CF patients may also be a part of this scenario, as an improvement in quality of life can have large run on effects to a home's general happiness and atmosphere.

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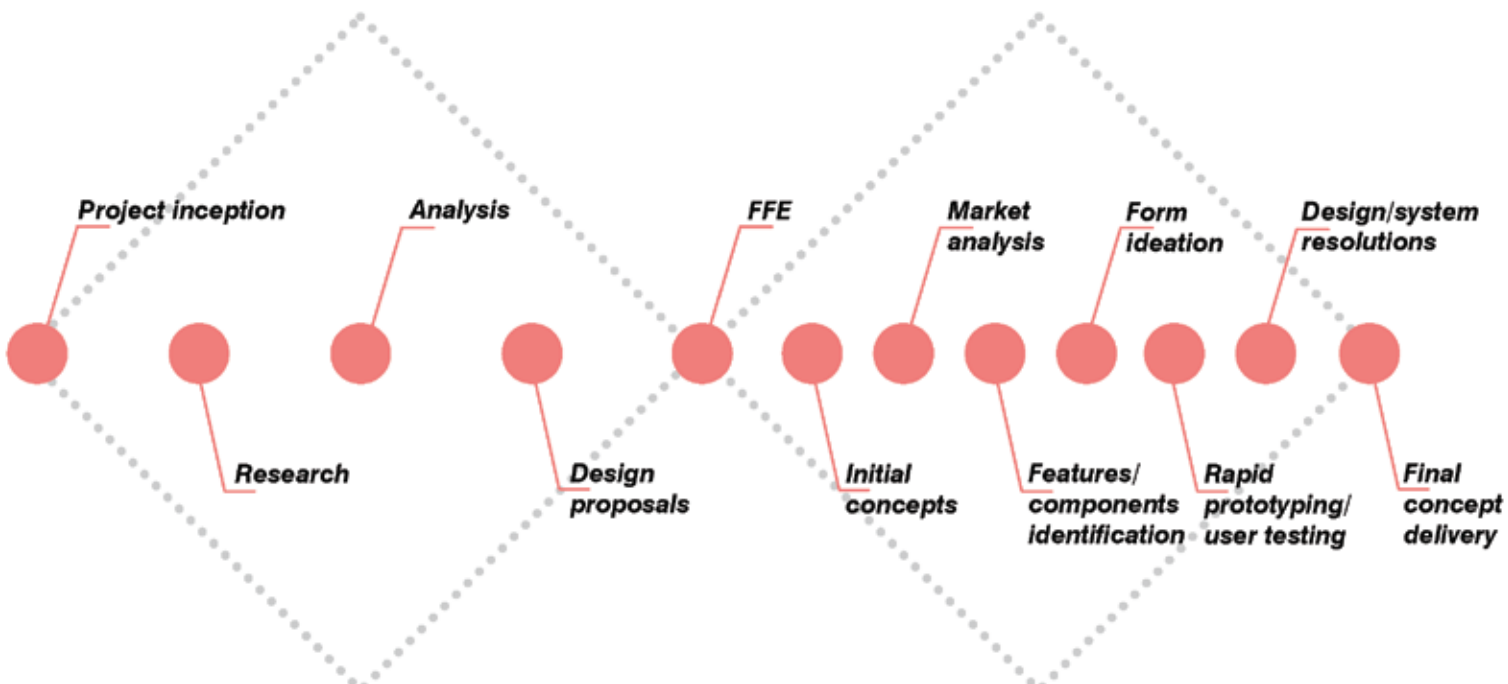

Figure 30: Project design process graphic visualisation

BUSINESS MODEL CANVAS

Table 1: *AUSPIRO* business model canvas

<table>
<tr>
<td rowspan="2">Key Partners

Manufacturers
Injection moulding
PCB

Suppliers
Microphone tech
Flowmeter tech
OEM parts

Distribution
Shipping companies local & international
App stores

Data
Server suppliers
Web domain</td>
<td>Key Activities

Technological R&D

App and server upkeep

Manufacturing and distributing AUSPIRO

Design support for CF community</td>
<td rowspan="2">Value Propositions

Phone Application
Communication
Lung function monitoring
Treatment & appointment reminders

Convenience

Health security

Ownership over CF

Happier life</td>
<td rowspan="2">Customer Relationships

Trust

Accountability

Speed

Accessibility</td>
<td rowspan="2">Customer Segments

Young adult CF patients

CF patients

Family/friends of patient

CF clinics

Respiratory specialists</td>
</tr>
<tr>
<td>Key Resources

Initial capital

Partnerships with CF clinics

Crowdfunding</td>
<td>Channels

Advertisements
Online
Medical magazines

Cystic Fibrosis Australia

Online store</td>
</tr>
<tr>
<td colspan="3">Cost Structure

Advertisements

Distribution

R&D

Manufacturing

Purchasing supplier components</td>
<td colspan="3">Revenue Streams

Customer purchase of product

Ownership schemes
Rent to buy
After-pay

App revenue</td>
</tr>
</table>

FINAL DESIGN DISCUSSION

In Chapter 7 a set of design criteria were listed. These criteria outlined the aims and objectives of the design process that would follow, and serve as an integral checklist of features that the final concept must satisfy. The main design criteria outlined included design aims on the functionality of the device, it's usability, ergonomics, form, and technology among others. Through several channels of validation, the final product of this semesters work adheres to the criteria outlined in chapter 7.

The AUSPIRO allows CF patients to test their FEV1 and FVC numbers from their own home. It is a lightweight, portable, simple to use consumer product aimed at Cystic Fibrosis patients. The user interface has been fully integrated onto a separate mobile phone app which, along with the crucial testing procedures, provides various other features such as treatment and appointment reminders as well as communication channels for patients and doctors.

Some of the initial design criteria was not met, while other points were added as the design process progressed, however overall the AUSPIRO meets the initial criteria in that it is a device which provides CF patients with the ability to test their lung function from. That information is then sent through to their specialist care team who are able to review that information during remote clinic appointments. The AUSPIRO eliminates the need for patients to travel to and from a hospital clinic, which for rural patients can require air travel, while releasing some of the pressure on hospital systems which are already under stress.

The integration of a solid, pheasible device and an intuitive, useful phone app into a singular product satisfies the objectives and aims of this design project. As well as the importance of clinic appointments, communication channels and treatment reminders were other topics that came up in conversation during the research stage.
The context of this project is based around how Cystic Fibrosis patients respond to clinic appointments and whether providing them with a device that allows them to test their lung function from home has an impact on their decisions around treatment adherence. In this context, Cystic Fibrosis patients are the main stakeholders, with the additional actors being the specialists within the patient's health care team.
Family/friends of the CF patients may also be a part of this scenario, as an improvement in quality of life can have large run on effects to a home's general happiness and atmosphere.

The device was designed to be used outside of the typical hospital setting, whether that be within the patient's home, a friend or family members home, or outdoors. The ability to still attend clinic appointments remotely no matter the patient's location is what makes this device a useful tool in the journey of a Cystic Fibrosis patient.

SUMMARY

The aim of this design project was to develop a respiratory device for Cystic Fibrosis patients to test their lung function from home. By having this ability, patients can attend clinic appointments from home via remote video call, which removes the stress of long commute times and the risk of cross infection between patients. By integrating modern technologies and manufacturing processes into a cohesive consumer product, the AUSPIRO satisfies the initial design criteria regarding design

features, ergonomics, functionality, and technology among other categories.

The AUSPIRO (figure 31, 32) has been developed as a tool for CF patients to monitor their lung function, and will hopefully encourage CF patients to adhere more strictly to their treatment plans. By having the ability to regularly test their lung function and see the relationship between high treatment adherence and increased lung function, patients can make informed decisions on their treatment adherence habits.

Figure 31: *AUSPIRO spirometer*

Figure 32: *AUSPIRO*

CONCLUSION

The aim of this research project was to identify treatment adherence rates within CF patients and the barriers which may be preventing CF patients from adhering to their treatment plans. Roughly 1 in 7000 Australians are born with CF and in order to remain healthy they must ensure continued diligence and dedication to their treatment. These treatments are administered via a variety of different delivery methods and treatment can consume hours of a patient's day.

Through reviewing the current academic knowledge surrounding CF patient treatment adherence, a range of potential causes and consequences of low treatment adherence were identified. While the knowledge within this field was considerable, there was little information available on the importance of clinic appointments, their attendance rates and the importance these appointments have in ensuring continued good health and treatment adherence.

After conducting two forms of primary research - online questionnaires and phone interviews - with CF patients across Australia, valuable information relating to both treatment plan adherence habits as well as attitudes around clinic appointments was gathered. These findings supported the previous literature on the topic while also providing an insight into the identified research gap. These results were analyzed and then presented and discussed.

The main findings centered around the importance of patients attending clinic appointments. These appointments provide an opportunity for consultation between patients and their specialist health care team. At each appointment patients undergo three important procedures; testing their lung function performance via an instrument called a spirometer, having their airways examined by a respiratory specialist via a stethoscope, and have consultations with several specialists in regards to their health and treatment plans. When patients don't attend these appointments, they run the risk of leaving chest infections to go untreated, resulting in potential long term lung damage and ultimately a steep decline in health and quality of life.

After analyzing the interviews and online surveys it became clear that patients would be more likely to attend clinic appointments if they could be conducted remotely via video call. The inconvenience of travelling to and from hospital along with risk of cross infection between patients were cited as the main factors behind low clinic appointment attendance.

In order to facilitate remote 'telehealth' clinic appointments, the various examinations undergone by patients while at clinic must be completed by the patients themselves within their own homes. Design proposals and recommendations were presented within the final stages of this report and outline what would be required of a product to facilitate home telehealth clinic appointments.

By developing and designing a product which allows patients to examine and monitor their respiratory condition from home, CF patients will have access to safer and more convenient methods of clinic appointment delivery. As suggested by the primary research which was conducted, this will increase appointment attendance, in turn improving the treatment adherence rates of CF patients nation-wide.

A design processed based on user testing and iteration via sketching and 3D modelling resulted in a validated end product which provides on the requirements outlined within the design brief.

ADVANTAGES OF PROPOSED DESIGN

The advantages of the AUSPIRO include increased functionality when considering the device as well as the app as a cohesive product. The paired app provides users with the ability to not only operate the AUSPIRO, but also make use of fitness goal tracking and reminder/calendar features regarding treatment and appointments.

In terms of it's technical features, the AUSPIRO features modern and accurate flowmeters that beat competitor products through lack of weight and moving parts, resulting in a more mechanically reliable device. No other product on the market integrates a modern, accurate spirometer with an electronic bluetooth stethoscope.

The AUSPIRO holds a unique and identifiable design language. The triangular form provides both aesthetic and structural value, while the magnetic connetion between the spirometer and stethoscope provides the user with a novel interaction between themselves and the product.

DESIGN IMPLICATIONS

The AUSPIRO will provide a unique option for consumers both aesthetically as well as functionality wise. The release of both the device as well as the app results in a product that spans multiple market places (physical medical devices and the app market), resulting in increased market exposure and subsequently an increased customer base. With an increase in sales comes extra revenue which can be spent on R&D of future products and future technology, resulting in more options for the consumer.

CF patients and family/friends of CF patients will be positively affected by the release of this device. It will act as a tool for these stakeholders to live a less stressful and interrupted life, while also providing a means from which patients can monitor and take responsibility of their own health. By removing the need for patients to travel to and from hospitals for their clinic appointments, there will be less of a burden across both hospital and transport systems. As Tskeleves (2017) explains, by freeing up resources within hospitals nurses and doctors are able to focus their attention and time towards higher risk patients, and as a result the whole system benefits.

As these technologies improve and remote clinic appointments become more common, the dependence on traditional hospital environments may decrease. Other devices may be developed which limit the necessity of patients visiting hospitals for treatment and appointments. Perhaps a user friendly alternative to traditional blood tests could be developed, allowing patients to run blood tests themselves from the comfort of their own home. Perhaps an innovative medicine delivery system could be developed which reduces the number of tablets a patient needs to take each day, resulting in a less intensive treatment plan which patients are more likely to adhere to.

The AUSPIRO is one of the first devices on the market to specifically target Cystic Fibrosis patients, and proves that there is a viable market segment here that can be designed further towards. While medical treatments may be improving as modern medicine and technology develops, Cystic

Fibrosis will continue to be diagnosed within the greater population, meaning there will always be a market for such devices. As the brand grows and gains a reputation of reliability and user centric design, the opportunity to develop more CF targeted product increases, and with it a chance to establish a strong market niche.

APPENDIX

SURVEY RESULTS

10

10.1 Short Answer Responses

If you could change one aspect of the clinic appointment process, what would it be?

Speed- usually in clinic for 2+ hours

Often I am sitting around waiting most of the time, I understand this is hard to change, but it's the only part of the process which disappoints me from time to time.

To not have to see the whole team when its really not necessary, I feel like they just need to tick their boxes that they seen 'X' amount of patients that day but this causes me to be there for several unnecessary hours

Cross infection

None. Very professional and supportive team.

waiting time between health care professionals

Telehealth when feeling well

I would prefer Telehealth services or earlier appointments

Nothing

Testing of height and weight in each clinic examination room instead of using one communal set of equipment that all patients use. This would help to reduce contact with surfaces that other CF patients would have touched as well as help to reduce cross-infection risks. Of course, the equipment in each CF clinic room would need to be thoroughly cleaned and wiped over following each patient use.

Telehealth

Limiting contact between patients and speeding the process up
Being in close proximity to other cf patients

Table 3: Treatment prescriptions and adherence results

Treatment	No. of patients	Frequency of missed treatments							
		Never	Rarely	Once/month	Once/fortnight	Once/week	3-5 times/week	Once/day	Always
PEP	14 (85.7%)	2 (14.3%)	-	1 (7.1%)	2 (14.3%)	2 (14.3%)	2 (14.3%)	2 (14.3%)	4 (28.3%)
Pulmozyne	12 (78.6%)	1 (8.3%)	2 (16.7%)	-	1 (8.3%)	1 (8.3%)	2 (16.7%)	2 (16.7%)	3 (25%)
Hypertonic Saline	12 (71.4%)	2 (16.7%)	1 (8.3%)	-	-	3 (25%)	3 (25%)	1 (8.3%)	2 (16.7%)
ABDEK	13 (85.7%)	4 (30.8%)	2 (15.4%)	-	-	1 (7.7%)	4 (30.8%)	1 (7.7%)	1 (7.7%)
CREON	14 (100%)	3 (21.4%)	4 (28.6%)	-	-	1 (7.1%)	2 (14.3%)	-	4 (28.6%)
Azithromycin	10 (57.1%)	3 (30%)	1 (10%)	1 (10%)	-	-	4 (40%)	-	1 (10%)

INTERVIEW RESPONSES

PATIENT #1
MAY 20TH

What hospital do you attend for clinic appointments?

I attend Prince Charles currently

What medication are you currently taking?

I do hypertonic, pulmozyne, creon, symdeko, abdek, salt, multi vits, aspirin, I've got a skin condition as well so I'm on medication for that. I'm also a diabetic so I take insulin.

Do you find any of your treatments/medications difficult to stay on top of?

I think probably doing the nebulisers. I still do percussion with the hypertonic so I use a combination of the PariSprint, Aerobeaker. I think that's what works best for me, the hospital said there's no studies which have shown that there's any big difference in just doing Pep and just doing Aerobeaker alone, but I feel that's what works best to fit in everything around my schedule with uni and work. I'm quite heavily into sport so I guess that's one of the bigger things to fit around.

Do you have a set routine for completing your treatments?

There's not really a set routine, I found in myself doing physio and nebs was something id do in the arvo because that's when id get the most downtime after uni and before sport. All my tablets are taken in the morning/night when they need to be taken. The bulk of treatments are done in the afternoon.

Touching on sport there, have you played sport for most of your life?

Yes, dad was quite heavily into sport and so was mum so when I was diagnosed they were very big believers that the more sport I do the healthier ill be in the long run. Always been super active and play a lot of touch football and referee 4 nights a week and all weekend. Its lots of running which keeps me healthy. When I am sick, not being able to do that shows that I do rely on sport and my body does need it.

Where do you store your medication?

The ones that need to be in the fridge stay in my bar fridge and then I have a medication cupboard

Do you ever take medication and then forget that you've taken it and have more?

Oh all the time! I guess when you're out at someone place and you're eating and think 'oh *** did I take my CREON' and then you ask your friends/family and they say 'yes you have'. Sometimes at home I forget if I've had them and then take extra just in case.

How long does it take to get to clinic and how do you get there?

I live in Warner, it probably takes about 30 minutes to get there depending on traffic. I go to separate hospitals for different conditions.

How long does the whole process take?

It varies a lot – at the childrens hospital things were done in a very regimented style. My transition from the childrens to the adults hospital hasn't been great for me – things are done at their own pace so I guess appointments start at 1 and I wont leave until 3/3:30, It varies. Annual reviews take the longest.

Have you attended any telehealth clinic appointments?

Yes I had one In April.

What are your opinions on those?

There were efficient, all the video conferencing didn't work so it was all phone calls. It was a phone call from each health care member over the course of 2 hours so you had to be there waiting. It wasn't great but I guess there's not much else they can do over the phone. I have only been in contact with the dietician since then. It's as much as they could do over the phone.

Have you been sent out the required equipment for telehealth clinic appointments?

No, they spoke about that but I haven't heard anything else about that.

Do you ever communicate with other CF patients?

When I was younger, there were quite a lot of other CF patients that you would speak to – not face to face. The doctors got me onto some chat rooms for people with CF but I found the people on those chatrooms were people that didn't share the same mindset as me – they were online just to dwell on the fact that they've got CF. It's crap but if you've been dealt this hand of cards you've got to deal with it the best way possible. I believe those people made me question my positive mindset – mum had the same experience with other CF parents who would always focus on the negatives. She felt because I was healthy and not in hospital much there was a barrier between herself and them.

There is an older CF lady who I have been in contact with who does a lot of CF advocacy stuff and I've been talking to her about how her day to day life is and stuff like that – keeping it real and not dwelling on it. I found that to be quite a positive thing.

Does it get tough not really knowing what's in store for you later down the track?

I guess because looking from the outside CF people look like everyone else but the struggles you go through at home that no one sees is where the understanding of people is not quite there – sometimes it is tough and although my family understand, there's only a certain level of understanding they can have and don't fully get it.

What is the most difficult part of dealing with CF?

I think the public image – I've never been embarrassed by CF. People misunderstanding the illness – without the physical signs of sickness people don't understand to the extent they really should – 'it's just a runny nose and it's just a cough'.

PATIENT #2
MAY 20TH

What medication are you currently taking?

I know I'd probably take 10ish tablets in the morning plus all the CREON.

Which hospital do you attend for treatments?

Prince Charles

Which of your treatments do you find the most difficult to complete?

Probably getting 2 physio sessions a day in, I usually just do 1. Pep with hypertonic is my main form of physio.

Do you have a set routine for completing your treatments?

Yes I make sure that I do it around 9 o'clock every morning.

Where do you store your medication?

In like a trolley thing in the lounge room. It's right next to the couch so when I wake up and go sit on the couch it's right there.

Do you ever set yourself reminders for your treatments e.g alarms

No not anymore.

How long does it take to get to clinic and how do you get there?

On average about 30 minutes, I usually drive in.

How long does the whole process take?

Anywhere from 15 minutes to 2 hours.

Have you attended any telehealth clinic appointments?

Yes just since COVID has been around.

What are your opinions on those?

It was kind of like 50/50 – my first one was with a doctor I hadn't heard of before. – im talking to a complete stranger, no video etc. It was kind of good because I didn't have to worry about getting dressed, leaving at a certain time. I still couldn't get out of the physio and dietician coming to see me though – sometimes I can get away without seeing everyone.

Have you been sent out the required equipment for telehealth clinic appointments?

I haven't received it yet but I've been told it's on the way

Do you ever communicate with other CF patients?

I've met one person through a mutual friend – the rest have been because I've met them at hospital.

Do you find speaking with other CF patients helps to alleviate any worries that you may have?

Yeah usually when other people have an issue they might go 'hey have you gone through this? We have bitch sessions about treatments. It's nice to have people who understand what you're going through.

What is the most difficult aspect of dealing with CF?

I think its probably like wanting to keep up with people but in my head I know I cant keep up with people – trying to be normal but knowing I cant be.

PATIENT #3
MAY 21ST

What medication are you currently taking?

They've got me on 4-5 different ones but I only take maybe 1 or 2 – just my insulin

Which hospital do you attend for treatments?

Brisbane Mater hospital

Do you find any of your treatments/medications difficult to stay on top of?

Probably my augmentin – the one I have when im really sick. Other than that I don't take my antibiotics anymore.

Do you have a set routine for completing your treatments?

Nope

Where do you store your medication?

The fridge.

How long does it take to get to clinic and how do you get there?

About 2 hours – my parents drive me

How long does the whole process take?

About an hour tops.

Have you attended any telehealth clinic appointments?

Yes my last one.

What are your opinions on those?

Yeah it was easy – a lot easier than coming into hospital

Do you ever communicate with any other CF patients?

No, there's one other person I know who has it but I just keep to my self usually.

What's the most difficult aspect of dealing with CF?

Probably other people's reaction to it.

PATIENT #4
MAY 22ND

Which hospital do you attend for treatments?

I attend the Prince Charles hospital

What medication are you currently taking?

I take around 30 tablets a day

Do you find any of your treatments/medications difficult to stay on top of?

I'd say probably the Hypertonic saline because it takes quite a while and I have to get a bit motivated to do it

Do you have a set routine for completing your treatments?

Yes – I get up and prepare my treatments, do hypertonic and colistine. Then at night time I just repeat that

Where do you store your medication?

Some of its in the fridge and the rest is in the pantry

Do you ever set yourself reminders for your treatments e.g alarms?

No I can pretty much always remember to do them

How long does it take to get to clinic and how do you get there?

About 40 minutes

How long does the whole process take?

About an hour its usually pretty good – a massive difference to the childrens hospital

Have you attended any telehealth clinic appointments?

Yes I have since COVID has began

What are your opinions on those?

It was interesting – it felt like I was in a normal clinic appointment. They set up a video and put you in a room and people come and go. I felt like it took a little longer than a regular clinic appointment, but it worked and it was effective. I have another one next week.

How have your habits changed during covid?

Usually I have 2 months on, one month off for some treatments but at the moment due to COVID I'm constantly on medication so I stay well.

Do you ever communicate with other CF patients?

No, I don't. I have once before but it sort of freaks me out knowing what I could go through. I rather stay in my own little world.

So you'd find it detrimental speaking to other CF patients?

Yes

What's the hardest part of dealing with CF?

I'd say the mental side of it - thinking of life expectancy and stuff like that.

Table 2: Benefits and limitations of qualitative research

Classification	Theme	Topic	Example
Mental	Treatment habits	Treatment habits	'I take around 30 tablets a day.'
		Self-discipline	'I make sure I do lots of running to keep fit.'
		Non-adherence	'Other than that I don't take my antibiotics anymore.'
		Exercise	'I'm quite heavily into sport.'
	Relationships	Self-image	'Trying to be normal but knowing I can't be.'
		Friends/family	'Sometimes it's tough and although my family understands, there's only a certain level of understanding there.'
		CF patient interaction	'It's nice speaking to other CF people because they know what you're going through.'
	Mindset	Self-containment	'I rather just stay in my own little world.'
		Positive connotations	'About an hour which is pretty good…a massive difference to the children's hospital.'
		Personal opinion	'I feel that's what works best for me to fit everything in.'
		Negative connotation	'My transition from the children's hospital to the adult's hasn't been great.'
		Empathy	'It wasn't great but I guess there's not much else they can do.'
Lifestyle	Life outside CF	Commitments/hobbies	'I referee 4 nights a week and all weekend.'
		Other medical conditions	'I'm also a diabetic so I take insulin.'
	Healthcare team communication	Healthcare team contact	'I have only been in contact with the dietician since then.'
		Clinic appointment	'It was interesting – it felt like a normal clinic appointment.'
		Advice from hospital	'The hospital said there's been no studies to prove otherwise.'
	Daily routines	Time of day	'Appointments start at 1 in the afternoon usually.'
		Schedule	'I feel that's what works best to fit everything in around my schedule.'
		Routine activities	'I get up and prepare my treatments for the day as soon as I wake up.'
		Specific location	'I attend Prince Charles hospital.'
Condition	CF Milestones	CF related milestones	'When I was diagnosed with CF…'
	Treatment barriers	Public perception	'Probably other people's reaction it.'
		Mention of sickness	'People think it's just a runny nose and a cough – it's more than that.'
		Mental burden	'I'd say the mental side of it – thinking of like expectancy and stuff like that.'
		Limitations caused by CF	'…trying to keep up with people but knowing that I can't.'
		Barriers to adherence	'I have to get motivated to do it.'

APPENDIX

REFERENCES

Abbott, J. Dodd, M. Gee, L. Webb, K. (2001) Ways of coping with cystic fibrosis: implications for treatment adherence. Disability and Rehabilitation. 23(8). 315-324

AHPA. (2020). Physiotherpay. Allied Health Professionals Australia. Retrieved from: https://ahpa.com.au/allied-health-professions/physiotherapy/

Allen, J, L. Panitch, H, B. Rubenstein R, C. (2010) Cystic Fibrosis. Lung Biology in Health Disease. 19(118) 352-353

Alma, L. Malay, B, T. Lutz, G. (2019) Growing Up With Cystic Fibrosis. Achievement, life satisfaction, and mental health. 21(10) 1829-1835

Alvarez, J. Ziegler, T. Millson, E. Stecenko, A. (2015) Body composition and lung function in cystic fibrosis: association with adiposity and normal weight obesity. Nutrition. 32(4). 447-452

Balli, F. (2018) Developing Digital Games to Address Airway Clearance Therapy in Children With Cystic Fibrosis: Participatory Design Process. JMIR Serious Games. 6(4)

Barker, D. Qutiiner, A. (2016) Parental Depression and Pancreatic Enzymes Adherence in Children With Cystic Fibrosis. Pediatrics. 137(2)

Bregenballe, V. Schiotz, P. Boisen, K. Pressler, T. Thatsum, M. (2010) Barriers to adherence to adolescents and young adults with cystic fibrosis: a questionnaire study in young patients and their parents. Patient Preference and Adherence. 5. 507-515

Broncheatitis (2020). Positive Expiratory Pressure Therapy. Retrieved from: https://bronchiectasis.com.au/physiotherapy/techniques/positive-expiratory-pressure-therapy

CFF. (2020) Positive Expiratory Pressure. Cystic Fibrosis Foundation. Retrieved from: https://www.cff.org/Life-With-CF/Treatments-and-Therapies/Airway-Clearance/Positive-Expiratory-Pressure/

Cowlard, J. (2017) The role of the cystic fibrosis nurse specialist. Nursing Times. Retrieved from https://www.nursingtimes.net/clinical-archive/respiratory-clinical-archive/the-role-of-the-cystic-fibrosis-nurse-specialist-21-03-2002/

Daniels, T. Goodacre, L. Sutton, C. Pollard, K. Conway, S. Peckham, D. (2011) Accurate Assessment of Adherence: Self-Report and Clinician Report vs Electronic Monitoring of Nebulizers. Chest. 140(2) 425-432

Daniels, T. Mills, N. Whitaker, P. (2013) Nebuliser systems for drug delivery in cystic fibrosis. Cochrane Database of Systematic Reviews. 4

Davis, P. (2005) Cystic Fibrosis Since 1938. American Journal of Respiratory and Critical Care Medicine. 173(5)

DeLambo, K. Levers-landis, C. Drotar, D. Quittner, A. (2004) Association of Observed Family Relationship Quality and Problem-Solving Skills with Treatment Adherence in Older Children and Adolescents with Cystic Fibrosis. Journal

of Pediatric Psychology. 29(5) 343-353

Dziuban, E. Saab-Abazeed, L. Chaundry, S. Streetman, D. Nasr, S. (2010) Identifying barriers to treatment adherence and related attitudinal patterns in adolescents with cystic fibrosis. Pediatric Pulmonology. 45(5) 450-458

Eakin, M. Bilderback, A. Boyle, M. Mogayzel, P. Kristin, A. Riekert. (2011) Longitudinal association between medication adherence and lung health in people with cystic fibrosis. Journal of Cystic Fibrosis. 10(4) 258-264

Eakin, M. Riekert, K. (2013) The impact of medication adherence on lung health outcomes in cystic fibrosis. Current opinion in pulmonary medicine. 19(6). 687-691

Eckdahl, T. (2016) Genetic diseases or conditions: cystic fibrosis, the salty kiss. Human diseases and conditions.

Esther, C. Muhlebach, M. Ehre, C. Hill, D. Wolfgang, M. Kesimer, M. Ramsey, K. Markovetz, M. Garbarine, I. Forest, M. Seim, I. Zorn, B. Morrison, C. Delion, M. Thelin, W. Villalon, D. Sabater, J. Turkovic, L. Ranganathan, S. Stick, S. Boucher, R. (2019) Mucus accumulation in the lungs precedes structural changes and infection in children with cystic fibrosis. Science translational medicine. 11(486)

HealthEngine. (2017). Respiratory & Pulmonology Physicians. Health Engine. Retrieved from: https://healthengine.com.au/info/respiratory-medicine

HuiChan, L. Shoff, S. (2008) Classification of malnutrition in cystic fibrosis: implications for evaluating and benchmarking clinical practice performance. The American Journal of Clinical Nutrition. 88(1) 161-166

Kettler, L, J. Sawyer, S, M. Winefield, H, R. Grenville, H, W. (2002) Determinants of adherence in adults with cystic fibrosis. Thorax. 57(5). 459-464

Koscik, R. Farrell, P. Korosok, M. (2004) Cognitive function of children with cystic fibrosis. Pediatrics. 113. 1549-1558

Kuhn, R. Gelrud, A. Munck, A. Caras, S. (2010) CREON (Pancrelipase Delayed-Release Capsules) for the Treatment of Exocrine Pancreatic Insufficiency. Advances in Therapy. 27(12) 895-916

Modi, A. Lim, C. Yu, N. Geller, D. Wagner, M. Quittner, L. (2006) A multi-method assessment of treatment adherence for children with cystic fibrosis. Journal of Cystic Fibrosis. 5(3) 177-185

Morganson, V. Litano, M. O'Neill, S. (2014) Promoting Work–Family Balance Through Positive Psychology: A Practical Review of the Literature. American Psychological Association. 4, 221-244. Retrieved from https://www.apa.org/pubs/journals/features/mgr-0000023.pdf

MayoClinic. (2020) Sprirometry. Retrieved from: https://www.mayoclinic.org/tests-procedures/spirometry/about/pac-20385201#:~:text=A%20spirometer%20is%20a%20diagnostic,a%20machine%20called%20a%20spirometer.

Muther, E. (2018) Overcoming psychosocial challenges in cystic fibrosis: Promoting resilience. Pediactric Pulmonology. 53(3).

Narayana, S. Mainz, J. Gala, S. Tabori, H.

Grossoehme, D. (2017) Adherence to therapies in cystic fibrosis: a targeted literature review. Expert Review of Respiratory Medicine. 11(2) 129-145

PariNeb. (2020) Pari-Neb Products. Retrieved from: https://www.pari.com/uk-en/products/pari-nebuliser/pari-lc-sprint-nebuliser-1/

Piasecki, B. Stanislawska-Kubiak, M. Strzelecki, W. Mojs, E. (2017) Attention and memory impairments in pediatric patients with cystic fibrosis and inflammatory bowel disease in comparison to healthy controls. Journal of investigative medicine. 65(7) 1062-1067

Peterson, C. (2008). What is positive psychology, and what is it not? Psychology Today. Retrieved from https://www.psychologytoday.com/us/blog/the-good-life/200805/what-is-positive-psychology-and-what-is-it-not

Sawicki, G, S. Sellers, D, E. Robinson, W, M. (2009) High treatment burden in adults with cystic fibrosis: challenges to disease self-management. Journal of Cystic Fibrosis. 8(2). 91-95

Smith, B. Georgiopoulos, A. (2016) Maintaining mental health and function for the long run in cystic fibrosis. Pediatric Pulmonogy. 51(44).

Solomon, G. Marshall, S. Ramsey, B. Rowe, S. (2015) Breakthrough therapies: Cystic Fibrosis (CF) potentiators and correctors. Pediatric Pulmonology. 50(40)

VandanBranden, S. McMullen, A. Schechter, M. David, J. Pasta, M. Michaelis, R. Konstan, M. (2011) Lung function decline from adolescence to young adulthood in cystic fibrosis. Pediatric Pulmonolgy. 47(2).

Walker, J. Edenborough, F. Wildman, M. Perveen, Z. Locke, Y. (2008) Improving clinic attendance. Journal of Cystic Fibrosis. 7(2) 98

Whitworth, J. (2018) My CF Cough Barges into Normal Experience. Cystic Fibrosis News Today. Retrieved from https://cysticfibrosisnewstoday.com/2018/07/11/cf-cough-barges-normal-moments/

Zindani, G. Streetman, D. Pharm, D. Samya, Z. (2006) Adherence to treatment in children and adolescent patients with cystic fibrosis. Journal of Adolescent Health. 38(1) 13-17
patients with cystic fibrosis and inflammatory bowel disease in comparison to healthy controls. Journal of investigative medicine. 65(7) 1062-1067

Sawicki, G, S. Sellers, D, E. Robinson, W, M. (2009) High treatment burden in adults with cystic fibrosis: challenges to disease self-management. Journal of Cystic Fibrosis. 8(2). 91-95

Scott, B. A., & Barnes, C. M. (2011). A multilevel field investigation of emotional labor, affect, work withdrawal, and gender. Academy of Management Journal, 54, 116-136. doi:10.5465/AMJ.2011.59215086

Smith, B. Georgiopoulos, A. (2016) Maintaining mental health and function for the long run in cystic fibrosis. Pediatric Pulmonogy. 51(44).

Solomon, G. Marshall, S. Ramsey, B. Rowe, S. (2015) Breakthrough therapies: Cystic Fibrosis (CF) potentiators and correctors. Pediatric Pulmonology. 50(40)

VandanBranden, S. McMullen, A. Schechter, M. David, J. Pasta, M. Michaelis, R. Konstan, M. (2011) Lung function decline from adolescence to young adulthood in cystic fibrosis. Pediatric Pulmonolgy. 47(2).

Walker, J. Edenborough, F. Wildman, M. Perveen, Z. Locke, Y. (2008) Improving clinic attendance. Journal of Cystic Fibrosis. 7(2) 98

Whitworth, J. (2018) My CF Cough Barges into Normal Experience. Cystic Fibrosis News Today. Retrieved from https://cysticfibrosisnewstoday.com/2018/07/11/cf-cough-barges-normal-moments/

Williams, GR. (2005) Illustrating triangulation in mixed-methods nursing research. Nurse Res. 12. 7-18. Retrieved from https://journals.rcni.com/nurse-researcher/illustrating-triangulation-in-mixedmethods-nursing-research-nr2005.04.12.4.7.c5955

Zindani, G. Streetman, D. Pharm, D. Samya, Z. (2006) Adherence to treatment in children and adolescent patients with cystic fibrosis. Journal of Adolescent Health. 38(1) 13-17
Pediatric Pulmonolgy. 47(2).

Walker, J. Edenborough, F. Wildman, M.