

# **THE LIVED EXPERIENCE OF WOMEN WHO HAVE HEREDITARY ANGIOEDEMA**

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No part of the material in this thesis has been submitted for any degree or other qualification at any other institution by myself or, to the best of my knowledge and belief, by any other person.

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### **Abstract**

Hereditary Angiodema (HAE) is a condition which effects up to 1 in 50,000 people, there is a 60% pre dominance in women being diagnosed and women are also the more symptomatic of the sexes, as HAE can be triggered through a heightened hormonal activities, stress and females medication. The females shared their lived experience of HAE through online accounts and were recruited by an organisation for HAE sufferers. Aims for the research is to gain an understanding the effect HAE has on a females' livelihood and the effects medication and the healthcare systems due to HAE. The accounts were analysed by a descriptive phenomenological approach. The data revealed three themes; of a double edged sword, in which medication for HAE was has been found to be miraculous but there can be severe side effects, the second was restrictions in which there are events and activities females cannot do and females need to be wary of HAE triggers and finally the rarity of the disease as women are questioned about the legitimacy of their condition and there is a lack of professional awareness. The findings support previous literature of chronic illness and rare conditions.

## Introduction

Hereditary angioedema (HAE) affects between 1 in 10,000 and 1 in 50,000 people (Zuraw, 2008). Throughout an attack of HAE irregular functioning occurs due to an overproduction of the peptide bradykinin, increasing vascular permeability and leading to a large amount of fluid entering the site area and causing an attack of swelling and inflammation (Gower et al, 2011). The swelling occurs in mucous membrane of the epithelial tissue (Kaplan, & Greaves, 2005). Site areas are: the buttocks, arms, face, genital, feet and abdomen, upper airways and the tongue (Frank, 2005). In addition abdomen swelling causes symptoms of vomiting, diarrhoea or stomach pain due to swelling of the intestines, stomach, urethra and bladder (Frank, 2005). Tendencies and frequencies of attacks differ from person to person. The causes of attacks include: infections, stress, dental trauma, ACE inhibitor (treatment for high blood pressure and heart failure), trauma and heightened hormonal activity (Baker, 2012). In the main attacks start in puberty as this is the start of heightened hormonal activity; however attacks can occur during pre-pubescence (Bouillet, 2010). This makes the condition an episodic illness as it can occur during different hormonal activities.

A pre-dominance occurs in women, with 60% of females being diagnosed as opposed to 40% prevalence in men (Bork, Meng, Staubach & Hardt, 2006). This is due to the fact that women are more at risk of attacks as the female hormonal system has more episodes of heighten hormonal activity such as puberty, pregnancy, lactation, menopause excreta, making them the more symptomatic of the sexes (Bork, Meng, Staubach & Hardt, 2006) (Bouillet, 2010). For example past

literature reported pregnant women with HAE in the first and third trimester of pregnancy and during lactation increased the likelihood of an attack. (Chinniah & Katelaris, 2009). They also reported attacks were more frequent and severe after pregnancy (Chinniah & Katelaris, 2009). Medication can also exacerbate the frequency and severity of HAE attacks for example: the combined oral pill, hormone replacement therapy and physiological oestrogen replacement (Bouillet, 2010).

HAE is a rare condition but can have profound impact on a female's livelihood due to the attacks but the reality of living with HAE is unknown (Gomide, et al, 2013). HAE will impact aspects of life for women with regards to work, family, social and every aspect of livelihood (Gomide, et al, 2013). HAE is a hereditary condition so women may have children with HAE as there is a 50% chance of an offspring inheriting the condition from the parents (HAEA, 2008). However, approximately 20% of HAE cases are due to spontaneous gene mutation occurring at conception with no prior family history of HAE, so some women have no idea they have a condition or what is making them ill (HAEA, 2008).

The fact that the illness is episodic means attacks can occur spontaneously and very quickly (NHS, 2016). These can be debilitating, for example a foot swelling causes women with the Immobility problems meaning a woman cannot walk and thus causing inconvenience in her life (NHS, 2016). This inconvenience can affect her life economically, socially and professionally as they are immobile so it is difficult to perform day to day activities. The episodic nature of the illness also means over a particular episode there can be multiple and regular swellings for females (Hofman,

Relan & Hack, 2016). Research has found there can be up to five locations of swelling during one attack (Hofman, Relan & Hack, 2016). Swelling can also be extremely painful for women, causing vomiting and nausea from abdominal attacks (Ugochukwu, 2010). Typically attacks last 3 to 4 days but if attacks are untreated they can last up to 2 weeks, during which time the female remains in pain (Genetic Home Reference, 2009). Also the swelling of the throat and airways makes HAE a life threatening disease as swelling causes the airways to be locked which can cause suffocation (Dewald & Bork, 2006). However no literature has been completed into the effects of HAE on females lives but the impact can be major for example leading to immobility, with up to five attacks at once, severe pain which can last days and the possibility life threatening situations. Completing research would provide an understanding of what female's experience during their lives with HAE and what issues are presents.

HAE is a chronic illness and since there is little literature on HAE, theories of chronic illness and the literature of lived of experience can be applied. Previous literature of the lived experience of chronic illness involve the beliefs, perception, expectations, attitudes and experience placed on living with the illness (Charmaz, 1997). Each of these items is unique to the individual with chronic illness (Paterson, 2001). This is supported by schemas of chronic illness, which hold large amounts of information from aspects of our lives (Cherry, 2015) . Schemas can aid the individual to make sense and understand their condition through their experience, perceptions, beliefs and attitudes (Garro, 1997). So females with HAE will place different meaning on HAE from their experience, for example as HAE can be

triggered through dental trauma, they may hate going to the dentist as it could cause an attack placing more inconvenience on their life as they have had an attack (Baker, 2012).

However schemas can be maladaptive and are difficult to modify if the new information contradicts the current understanding and belief about a particular illness, in which the information is either not processed or completely forgotten about (Leahy & McGinn, 2012). This is part of confirmation bias in which people are most attracted to information which affirms their current beliefs (Leahy & McGinn, 2012). With HAE being an episodic illness means with each new episode the condition can change for example the severity and site areas can differ (HAE UK, 2016). Something which challenges the schema since it would need to be modified with new information accompanying the new episode of HAE.

Although it is possible to create alternative schemas, in which they would be able to accept new episodes (Padesky, 1994). Those who can adapt schemas have found that acceptance of illness is higher (Leahy, 2015). This is supported in a four states of acceptance. The first was disavowal, there is no acceptance of the chronic illness and they feel there is a lack of control and balance of the illness (Ambrosio et al., 2015). The next is false normality, meaning no acceptance but the person feels they have some balance and partial control of the illness (Ambrosio et al., 2015). Afterwards is the new normal, a person has acceptance and feel they have control and balance within their lives (Ambrosio et al., 2015). However within chronic illness sometimes temporary distresses can occur such as a severe symptom of the

condition which causes disruption leading to a breakdown in acceptance, control and balance of the chronic illness (Ambrosio et al., 2015). However disruption only occurs for the false normality and new normality but the disruption state is only temporary and people can return their previous states of acceptance (Ambrosio et al., 2015). This is supported as those who adapt their lives and create a new normal state and as they adapt their schemas at the new episode on their life it is easier to accept chronic illness because when there is a breakdown they adapt their schemas. With HAE being an episodic illness, breakdowns would likely occur with possible changes in the severity and site areas of symptoms (HAE UK, 2016). However those diagnosed with chronic illness can find it hard to accept the condition due to the sense of grief which can accompany the condition (Kresser, 2011). The sense of grief can occur as people find they are unable to complete certain activities leading to a state of denial (Kresser, 2011) (Telford Kralik, & Koch, 2006). In the case of HAE this would be restrictions on physical activities in family life, occupational life and school which those suffering from the condition could undertake before the symptoms appeared (Bygum, 2014).

Acceptance of an illness has also been found to improve self-esteem even with the limitations chronic illness can bring (Nowicki & Ostrowska, 2008). Diagnosis has been found to lower self-esteem, something which has negative effects and has been linked to increased stress and an increase in the severity of symptoms (Juth, Smyth & Santuzzi, 2008). This can then impact on disease management, self-care and psychological wellbeing (Juth, Smyth & Santuzzi, 2008). This is supported as low self-esteem has been linked to greater stress and symptom severity of the

chronic illness (Maki, 2004) (Ireys, Gross, Werthamer-Larsson & Kolodner, 1994). Within HAE stress has been found to be a trigger (of HAE) so low self esteem could increase the triggers as stress has been linked to low self esteem in chronic illness (HAE Hope, 2016). However in a study it was found that there were similar levels of self esteem between those with chronic illness and those without so self esteem may not necessarily be due to the chronic illness (Schuttinga-Helder, Tuinstra, & Groothoff, 1996).

Diagnosis can affect self esteem but other factors after diagnosis contribute to chronic illness, especially if the condition is rare as the condition can be difficult to identify and there can be a lack of up to date research (Rare Disease UK, 2016). Also there is a lack of awareness for rare diseases as many health care specialist do not know about the condition or know very little, so on average it takes five years to receive a diagnosis (Stewart & Peers, 2013). Due to the lack of knowledge about rare diseases it can be common for those with rare diseases to receive misdiagnosis for their condition (Stewart & Peers, 2013). With up to 1 in 50,000 people suffering from HAE diagnosis could be an issue as it is rare, so many health care professionals may not know about HAE. Also due to the lack of a diagnosis the legitimacy of conditions can be questioned as well (Varul, 2001). If legitimacy of the condition is questioned there cannot be a transition to make the condition better for those who have it (Varul, 2001). Findings included that people can be questioned about the severity of the symptoms, the extent of the condition and what it is like to live with (Swoboda, 2006) (Richardson, 2005).

Diagnosis is also the starting point for stage models of chronic illness such as trajectory theory. The first stage is pre-trajectory, genetic factors or lifestyle choices place a person in risk of a chronic illness (Corbin, 1998). After this is trajectory onset, noticeable symptoms lead to a diagnosis and the patient understands implications of the condition (Corbin, 1998). Thirdly is the stable phase the condition is managed within limitations of the illness (Corbin, 1998). The next stage is instability, there is an inability to keep the condition under control and there is a difficulty in daily activities (Corbin, 1998). Following on is the acute stage, the condition develops complications (Corbin, 1998). The sixth phase is critical where there is a life threatening situation for the disease needing emergency treatment (Corbin, 1998). Then is the comeback stage, this involves living with the condition in the normal constraints of the disease (Corbin, 1998). HAE would follow this model from pre-diagnosis where females would show symptoms, then this would lead to a diagnosis of HAE. Once females receive a diagnosis females are provided with medication causing the condition to remain stable. However once there is a new episode of HAE this can cause an instability as the frequency and severity of symptoms can change and medication which may have previously been used may not work this time (All About HAE, 2016). However once the swelling reduces they can continue on with daily life (All About HAE, 2016).

The theory presents some beneficial information about the stages of living with chronic illness such as diagnosis and acceptance, but the final stages of chronic illnesses do not always follow these steps (Burton, 2000). The final stages are the downward stage leading to rapid decline of the physical self and increase in disability

and lack of control of the symptoms and the final stage is dying (Corbin, 1998). However the decline and death of a person are not always caused by their chronic illness. Although HAE can be life threatening when the throat swells it is not a degenerative disease so over time it does not cause a decline in health (All About HAE, 2016).

Trajectory theory has limitations as HAE does not cause a decline in a females health. Trajectory theory also determines one episode not episodes and HAE is an episodic illness (Burton- Jeangros, Cullati, Sacker & Blane, 2015). The trajectory approach to health was challenged by the shifting perspective theory (Paterson, 2001). This theory suggests living with chronic illness is a process of continuous change from being well to being ill and as illness changes so should the perspectives (Paterson, 2004). With different episodes there are different situations such as new symptoms or more severe symptoms (Hummel, 2011). As these shifts occur the condition should be examined and interventions put in place, in accordance with the new shift (Paterson, 2004). As HAE is an episodic illness there can be shifts in the frequency and severity of symptoms with HAE during each episode. In addition what was a possible intervention during one episode may not be able to be used in the next, for instance in the case of pregnancy where the foetus can be harmed by medication used in one episode (Gorman, 2008).

One way to help during new episodes is to successfully self-manage. This can be accomplished through making decisions about care backed up with thorough

information, successfully completing activities to manage the condition and finally by gaining necessary skills for managing psychosocial functioning, in doing this self-management is completed successfully (Clark, Becker, & Janz, 1991). One demonstration of this is with the home therapy where women are provided information about the condition. With women then gain training so they gain the skills to manage the condition by stopping attacks developing any further (Markovchick, Pons, Bakes & Buchanan, 2016). Also part of some self-management tasks are illness specific activities such as responding to symptoms and using specialised medication, managing diets and dealing with acute episodes (Van Houtum, Rijken, Heijmans & Groenewegen, 2015). In particular, specific tasks are used such as the self-management is used to stop the progression of an attack and the quicker there is a response to an attack the lower the progression of swelling (Markovchick, Pons, Bakes & Buchanan, 2016).

Overall it is hard to predict if previous research relating to chronic illness can be applied to HAE due to the lack of research. However by conducting this research it will enable insight into the lives of women with HAE. To investigate this phenomena qualitative methods provide the most beneficial approach, as it collects detailed views and perceptions from participants, which can then be used to develop a complex picture about the phenomena being study (Creswell, & Clark, 2004). Previous studies about lived experience of chronic illness are Taylor, Gibson & Franck, 2008, Ingadóttir, 2006 and Fox, S., & Chesla, 2008. Most of these studies use phenomenological methods as this provides participants the ability to share a first person point of view about their experience (Smith, 2013). These studies found

several commonalities amongst their data. Due to the condition, qualitative and phenomenological methods would provide the most insight as the females can share their personal experience of the condition and the impacts this had on their lives which will provide commonalities occurring in the females lives. In which the aims of this research is to investigate the experiential affect of HAE in different aspects of women's lives such as their personal life, social life, family life and professional life. The research also investigates the experience of attacks in women's lives, the experience of the healthcare system and medication in women's lives. However women were also invited in the study to discuss anything else which they thought was relevant to their experience which may not fit within the stated aims of the study.

## Method

### Design

The design of the study was a qualitative design using self report measures. Using a qualitative method allowed an investigation of the experience of illness (Waters, 2011). A self report measure was used as this allowed participants to write their personal experience from their perspective (Baker, Pistrang & Elliott, 2005). The method used were personal accounts which were completed through an online study which provided ease at collecting data, also accounts allowed the participants to organise their experience into meaningful expressions (Fraser,2004). This method of research allowed the participant to reflect on the reality and search the challenges and beliefs they face in their experience (Fraser,2004). This provided insights and interpretations of the life course and experience of the world around the participant (Bornat & Bytheway, 2012). Accounts were an unobtrusive measure as participants were asked two questions which were extremely broad, meaning there was no interference in the flow of the accounts they provide, as the flow was only interrupted as they moved onto the second question (Trochim, 2006). This is due to the participants being experts as they are the ones who have experienced the phenomena (Knapik, 2006). Phenomenology was beneficial for exploring illness as this method was used for analysing the main theme and will find reoccurring commonalities in the female's accounts of their lived experience (Smith, Flowers & Osborn, 2002).

## **Sampling Strategy**

The criterion to take part was females, over the age of 18 and with a HAE diagnoses. Participant's age ranged between 20 – 65 years old from several different areas of England. Ten participants were recruited as more than ten participants would cause complexity in the analysis and too few participants would cause an inability to detect themes (DePaulo, 2000). Women with any type of HAE were invited to take part. Participants who took part had type 1 and type 2 HAE but even though type 3 was not included they still share the same symptoms which contribute to a similar experience (Genetics Home Reference, 2009). The length of times participants had been diagnosed with HAE varied, some were diagnosed as a child while others were not diagnosed until early 30's, however most received a diagnosis mid teens to early 20's. Participants were recruited through HAE UK who sent the monthly newsletter (see appendix 1) also an email was sent to members (see appendix 2) and advert was also posted on their website (see appendix 3) and on their Facebook group.

## **Materials**

The data was collected through accounts written by the women (see appendix 4). The data set (the account) asked the participants to complete the account and consisted of two components, the first was about how living with the condition has impacted their lives. The questions were left broad and open as research has found that people tend to provide more description and detailed with broad questions (Wyse, 2014). Also to cover all aspect of HAE would have included a high number of individual questions which would be off putting, also these questions were closed

which would not have provided depth and detail to the answers provided by participants (Griffiths, 2016). The second component was about the attacks as these are main symptoms within HAE and the experience of attacks within a woman's life could cause an impact on a females experience of HAE. The materials also include a participant information sheet, this provided information about the research (see appendix 5). A consent form which abounded by BPS (British Psychological Society) code of ethics, where consent is needed for participants to engage in research (see appendix 6). Also a debrief was provided to participants so if they wished to withdraw their data they were able to (see appendix 7).

## **Procedure**

Participants were recruited through various methods of HAE UK and if they wanted to participate were asked to email the researcher. Once participants had contacted the researcher they were forwarded the documents of the participant information sheet, the consent form, the questions and the debrief sheet. When participants had completed their accounts they emailed their accounts and the consent form to the researcher. Participants completed the accounts within two to three weeks of initially being emailed the research materials.

## **Method of Analysis**

The method of analysis used was Giorgi's phenomenological method. The researcher performed phenomenological reduction which previous preconceived

ideas, prejudice, discrimination and schemas about them the data is removed from the conscious (Giorgi, 2012). By following Giorgi's method then phenomenological reduction should be completed and no preconceived notions are attached to the data. Also special sensitivity needed to be considered within this data as the personal information was shared by participants (Giorgi, 2012). The reduction was completed prior to the analysis phases. To overcome the issues of phenomenological reduction an outside source was used with no prior understanding of HAE read over the analysis of the analysed data to ensure no notions were applied to the data (Rolls & Relf, 2006)

At the start the researcher must read the whole transcript in order to get an understanding of the data, the researcher then went back to the beginning of the data and rereads and every time there was a new transition in meaning of the data it was marked, large descriptions are difficult to retain easily so marking new transitions makes it easier to notice themes (Giorgi, 2012). Afterwards the data was changed into meaningful expressions from the words in the data, allowing the data to be made explicit for the phenomena being studied, then the expressions have been gathered they are reviewed and the initial structure of the expression was started (Giorgi, 2012). Finally the initial structure was then used to help and clarify the raw data and interpret the research (Giorgi, 2012).

## Analysis

Three main themes found are the double-edged sword, rarity of the condition and the restrictions caused by the condition.

### Theme 1 Double of Edged Sword

For this theme women reported the medication was beneficial as it stopped their symptoms of HAE but the medication came with some severe side effects for the women which can cause more issues for the females

*“I spent all of my 20’s and 30’s mostly unaffected by HAE, I went back to college, moved away to study dance, graduated, found a job which involved traveling over the country, had nights out, partied, went on holidays, etc. etc. All the typical things you do. I rarely gave HAE a second thought. I took a few danazol every other day, increased the dose slightly if I felt rundown or had a cold. Very occasionally I would get swollen feet, if I forgot to take them, but by working from home it meant that I rarely had to mention it at work.”*

C1, lines 40-45

*“Many months later, and on my second appeal, I was finally granted the funding and was taught to self infuse. This was such a life changing time; I finally had the freedom and confidence to travel away from*

*home and could plan a holiday with my family. Although HAE does have a big impact on my day to day activities, I am determined to not let it stop me doing the things I enjoy and leading a full and active life.”*

R2, lines 62- 66

Medication for R2 has been “life changing” as the females have suffered with HAE symptoms without anyway of stopping them and the only thing which could be done was to wait until they stopped. However they were then prescribed danazol and hormone therapy, they have overcome all the attacks and the issues which came with HAE, enabling these women do all the things they wanted to do, such as with C1 who went on holiday and got a job which she could not do before. The medication was that beneficial as C1 said that she did not even have to give HAE another thought as her medication was controlling the condition now. Also due to how beneficial the medication is they can now have a life which is not controlled by HAE and is actually almost free from the condition, which gives the females a normal life and can build their confidence as they are not confined by the condition and can live like other females who are not confined by the symptoms of HAE. However some of the medication which has been prescribed to women has severe side effects.

*The side effects of the high doses of anabolic steroids that I was given when I was first diagnosed 27 years ago, have left me with hair loss and my scalp shows through. I was given wigs but have not got the courage to wear them. Increased body and facial hair, polycystic*

*ovaries that have robbed me of having children, weight gain, mood swings, high cholesterol and feeling like a freak. People look and point at me in shops like NEXT. Going into the changing rooms leaves me in tears as I see the body fat, the scalp showing my hair loss and body hair that is on most of my body. Children have been known to ask if I am a man. I have no self esteem.”*

F1, lines 6-12

*“I immediately became pregnant but didn’t worry as I was very healthy and in a happy and stable marriage. When I told the consultant at my next visit he said that I couldn’t carry the baby to term because the Danazol would damage the fetus and so I had to have a termination at 18 weeks, via an a very painful and prolonged induced live birth. I was not told this would happen and had no family there to support me, they thought I was having an operation and would come back later. Obviously I was extremely distressed and upset, particularly at the hospital where I was treated with disgust and I couldn’t understand why. Both the nursing staff and other patients ignored me and I gave birth alone in a side ward, and was left for 30 minutes before a nurse came to take the baby away. This is something I will never forget and I will never forgive the doctor for not informing me of this risk. I had a sterilization at the same time so this horrible event wouldn’t be repeated.”*

L1, lines 13- 23

*This didn't come without side effects though, and being a woman, I did find it difficult at times when I had more facial and body hair than all of my friends, my voice was deeper and I had very good muscle tone. My symptoms over the years have changed frequently."*

R1, lines 28-31

The side effects of the medication can be drastic for women as due to the external effects such excess body and facial hair, hair loss and even a deeper voice, which can then cause women to become upset, lose confidence and lose self-esteem because they feel different from other women around them who do not have HAE. As F1 said the side effects of the steroids she was given made her feel like a "freak" and as she looked so different children thought she looked like a man and due to this she has lost all her confidence. However the internal effects can be even more drastic as women can have fertility issues with the medication, for example developing polycystic ovaries or the medication being so strong it can cause damage to a foetus. L1 found this such a harrowing effect she got sterilized so it would never then happen again, she will never forgive the doctor for not telling her about the risk of danazol to foetus. Women already have fertility issues with HAE due to the increase in hormonal activity causing more attacks and the female uterus can swell up during pregnancies causing issues for the foetus and the medication can cause even more, meaning it is very difficult for women to naturally conceive and carry the foetus to full term. As F1 says her "polycystic ovaries that have robbed me of having children", she also said the thing she missed the most was not being able to have

children. Although the medication has been seen to been seen as “life changing “ and even a “god send” because women can then have a life like others without HAE as they no longer have symptoms of the condition, so they are able to do things they never could be before. However the medication can also cause issues such as lack of confidence due to external side effects and it can rob women of fertility issues, making them different again from other women who do not have HAE, causing a double edged sword, women would have to weigh up is the side effects worth the relief from HAE.

### **Theme two Restrictions**

For the women daily life can be interrupted due to the spontaneity of attacks and restricted what the females are able to do in daily life. Also attacks can be triggered through stress and dental trauma that women need to be careful they do not trigger attacks.

*“HAE became something that we considered all the time, weather is was questions about where we would store my meds when we went on holiday (camping, B&B’s hotels etc) whether we could commit to certain events, either a toddler group or out with friends. Or if it was a good idea to go out, play in the park, meet friends. I found it didn’t matter what I did or didn’t do. What I ate or didn’t eat I would have abdominal attacks, swelling in a hand or foot, but it would always end up as an abdominal attack, It was impossible for us to spend time*

*apart. What would I do without my husband to treat me? If I was away with the children who would look after them if I was ill? We did feel very restricted, and frustrated. My husband was my carer! That felt ridiculous.”*

C1, lines 70-78

*“However, this really affected my social life as a child as I wasn’t able to keep many friends, as by the time I was back they had all moved around friendship groups etc. I remember feeling very alone”*

N1, lines 6-7

*“I started working as a PA around 20 years old, I loved my job but my health was awful I had so much time off work. I couldn’t really progress because my health stopped me from doing that”*

R2, lines 31-32

HAE can cause restrictions to daily life as attacks are so spontaneous it is difficult to detect when an attack is going to happen, also the length and severity of an attack can vary so this makes it difficult to determine how long attacks can cause restrictions. When these attacks occur there is not a lot the females are able to do other than wait them out. Meaning for women when the attack does occur it will just stop them in their tracks no matter what they were doing, which would also make planning events difficult due to the spontaneity. For example N1 did not have a social

life as she was unable to keep friends due to her attacks making her feel alone as she didn't have many friends. This would cause feelings of frustrations and loneliness because they would be housebound and it would be extremely difficult to do anything or go anywhere because of the attacks. This applied to R2 as she was unable to progress in her job that's she loved because her HAE restricted her because of her attacks. However attacks can also cause restrictions for females about what they can and cannot do as they have to account for attacks or the possibility of an attack occurring.

*"I have mentioned that stress is a trigger for HAE but unbelievably, good stress is also a trigger. We can't get excited about things like family events as that can trigger a swell! The number of times I have made plans to visit family or friends and had to cancel at the last moment as a swell has been triggered. We have to learn to stay calm."*

F1, lines 47 – 50.

*"My first attack was when I was 8 years old. I visiting the dentist for a routine checkup not realising a day later. I woke up looking like the elephant man. My parents and I always wondered why and how what happened that day. After a week later my face seemed to go down without any medication was in bed hiding could hardly eat or drink. I was so upset and so worried what happened"*

## R2, lines 3-7

Attacks can cause restrictions for women but females are also restricted so they do not trigger an attack either which can cause more inconvenience as they have to factor in what can cause triggers. For example F1 demonstrates how stress but particularly any type of stress can cause restrictions in her life because if she gets stressed about an event can trigger an attack and then she is unable to attend. If potential triggers are not consider the HAE attacks can be severe for example what happened to R2 due to dental trauma as she had not received a diagnosis of HAE. The effects of the swelling were so severe due to her face she hid in her bed that it looked that bad. Also she was unable to do simple tasks such as eat or drink the swelling was so severe. She and her family were so upset and worried about what happened as they had no idea what had caused her swelling Due to HAE there can be a lot of restrictions in a females life that they are unable to do because attacks can be so spontaneous, severe and would stop a female for a few days while she recovers from her HAE attack leaving the female house bound which would restrict what she is able to do. Also HAE always needs to be considered as there can be restrictions to what females can do as the HAE can be triggered by the situation or event, which would restrict the female even more if she had an attack.

### **Theme Three Rarity of the Condition**

The final theme is rarity of the disease, in which the people in the females' lives can question females about the extent of their condition and the existence and health care professionals have a lack of awareness of the condition as they have never come across it before.

*“[Females husband] I has always been very supportive and helpful about my HAE attacks and it was he would insist I go to see my GP about the frequent attacks when I was always reluctant to go (I'm still the same, I will do anything to get out of seeing a doctor if possible) but the psychosomatic diagnosis shook him and he admits he began to doubt me. I couldn't believe that anyone could think I would put myself through all the misery of such frequent attacks and hours of pain and being physically sick, let alone someone who had seen me in the throes of such attacks so many times. He says now he didn't really doubt me but it did make him wonder.”*

E1, lines 178- 184

*“feeling as though a lot of people were judging me, teachers included for “skiving”, as they didn't understand nor wanted to understand my illness.”*

N1, lines 8 – 9.

*“This went on for years and years I use to think people thought I was making it up and especially my family. The swelling was visible”*

R2, line 11-13

Within the accounts normally there was an issue with legitimacy before females were diagnosed with HAE. Due to the lack of a diagnosis and after several tests and even GP's informing the women there was nothing wrong females legitimacy of the disease would have been questioned. For example people thought E1 purposely doing this to herself because she did not have a diagnosis, which can then effects the family opinion as they believe the doctors that the females is doing this to herself. Also as the disease has reoccurring symptoms some people would have thought this was psychosomatic or the female was making it up. One of the issues is how rare the disease so many doctors would not know about it or the fact that the condition actually did exist so it would not be tested as it is so rare. Due to the extent and the severity of the condition the females can feel like N1 does that they are being judged because they do not understand the condition. There was also an issue with the rarity of the condition with health professionals and their awareness of the condition.

*“When I went back to see the GP to find out the result of my hospital visit I was given some more pills. I took them but they were no more help than any other pills and then to my surprise I discovered that they*

*were tranquillisers! I returned to my GP to ask him why I'd been prescribed tranquillisers. He replied that it was because my attacks were psychosomatic. In shock I just said "What"? He simply said "It means you're doing it to yourself. It's all in your head". I truly couldn't believe that anyone could possibly think I would inflict those horrendous sickness attacks on myself but I could tell from the doctor's tone and delivery that he totally did believe this."*

E1, lines 98 -104.

*"I spent a lot of time in and out of hospital seeing different doctors and consultants only to be given different diagnosis's each time such as: Gastro-enteritis Irritable bowel syndrome. And even told it was psychological and I was making myself ill. Along with this I underwent various operations listed below before I had a confirmed diagnosis. At the age of 19 I had my appendix removed At the age of 21 I had an ectopic kidney re-routed (situated in my pelvic area)At the age of 23 I had to have my kidney removed as was banging against my pelvic bone as they had moved it during the previous operation"*

J2, lines 8-17.

*"The whole of my face swelled up, my eyes were completely shut and the doctor who visited me at home called an ambulance, he thought my husband had beaten me up! The ambulance crew also thought that I*

*had been beaten and didn't believe my story. No one had heard of HAE."*

L1, lines 54 – 57

The main issue with health professionals awareness is they have probably never come across HAE so they do not know about the condition, thus they do not know how to test for it. This is why some of the women have had misdiagnoses due to the condition and even had operations for conditions they were misdiagnosed with. This can be seen with J2 she received several different diagnoses and even underwent operations for conditions she did not have and then only received a diagnosis from a bowel consultant. Also even when HAE is diagnosed a lot of health care professionals have not come across the condition so they do not know what it is or what the symptoms are so they will make their own conclusions about illnesses based on the information they do already know about symptoms of conditions. Even when women do have a diagnosis they can have issues because doctors believe other factors have contributed to the swelling such as in L1 cases, they did not believe her HAE had caused her swelling but her husband had beat her. HAE is a rare disease as up to 1 in 50,000 people have the condition making the condition incredibly rare and not many people will know it exists. This is especially case pre-diagnosis as there is no explanation as to what is making the female ill which is why they are misdiagnosed and the legitimacy of the condition is sometimes challenged. Also once the condition is diagnosed still a lot of doctors do not know about the condition so they draw their own conclusion about attacks and symptoms of HAE.

## Discussion

The findings of the research has shown that the medication females have taken are beneficial for the women and can really help alleviate the symptoms they have and can help them live a life like women can without HAE and do things they were unable to do due to HAE. However as much as the medication is miraculous it can have severe implications due to side effect of the medication causing a double edge sword where the women need to weigh up is the medication worth the side effects it comes with. Another finding was HAE can cause restrictions in daily life due to the spontaneity of attacks can stop the female in her tracks due to an attack involving issue with school, professional life and the females social life; also attacks can be caused by triggered through stress and dental trauma so females have to be careful in their daily life that they do not trigger attacks as this cause inabilities within a females life. The final theme is the rarity of the condition, which the females legitimacy was questioned about whether they actually had the condition or if it was psychosomatic but this was due to a lack of diagnosis as the condition is so rare it can be difficult to diagnosis. Also with the rarity of the condition several health professional lack awareness of the condition as they have not come across HAE so they are unsure of the symptoms so they create their assumptions of what is causing the swelling and how to treat it. (See appendix 8 for theme tables with further quotes supporting these themes).

The women have reported the medication to treat HAE can be miraculous, which is due to successful self management, women did mention if they missed a

danazol tablet that was an increased likelihood of swelling, other than this with medication they reported few symptoms and an ability to do things they were unable to do prior to taking the medication (see appendix 8 for the theme table). Successful self management is completed through making decisions about care, which some women have decided to complete training for home therapy and now take medication (Clark, Becker, & Janz, 1991). The second is completing activities to manage the condition and by taking the medication or using home therapy it can stop attacks from coming on or developing any further (Clark, Becker, & Janz, 1991). The final part is gaining the skills for psychosocial functioning women have skills because by taking the medication they are able to complete activities in their social life and due to the effectiveness of the medical there is no psychological impact as they do not consider HAE when planning events (Clark, Becker, & Janz, 1991). Self management also includes specific activities to respond to symptoms, women completed these activities correctly which is why there was such a positive perspective of the medication as they were taking it correctly so they are able to part take in normal activities they could do previously before the symptoms appeared (Van Houtum, Rijken, Heijmans & Groenewegen, 2015). Responding to symptoms was also found with restrictions due to attacks as some females reported being careful not to become too stressed and being wary when undergoing dental trauma as this can trigger an attack.

Also responding to the possible triggers and the beneficial medication would show some women who incorporate this within their lives have accepted their illness. Previous literature found the third stage of acceptance is the new normal, in which

those who adapt their lives to the condition feel balance and control, with the medication some women reported they had control of their medication and did not really think about HAE because the medication stopped their symptoms, thus accepting the disease (Ambrosio et al., 2015). This is further supported as first stage of disavowal as they lacked control and balance as HAE restricted their daily lives, in which they lacked control due to the spontaneity of the condition so females were unsure when attacks would occur, meaning some women would miss school, impacted their work and their social life (Ambrosio et al., 2015). This further supported by schema theory as those who are able to create alternative schemas, find it easier to accept changes within their live due to chronic illness, as their illness has now changed due to medication, they find it easier to accept the illness (Padesky, 1994) (Leahy, 2015).

Despite this, the fourth stage is disruption in which changes of the illness disrupt normality but this only last for a short amount of time and those with the condition return to stage of the new normal (Ambrosio et al., 2015). The disruption stage can occur due to the side effects of the medication causing more issue, such as weight gain, high cholesterol, extra facial hair and liver problems. However some females are unable to incorporate this into their new normal stage as the symptoms are so severe like infertility or the external side effects so they cannot acceptance them as it has severely lowered their self esteem, this is supported as it was found those who have lower self esteem find it difficult to accept their illness (Nowicki & Ostrowska, 2008). This also supported as acceptance of chronic illness can be difficult due to some people feel sense of grief as they are no longer able to do

things they once were, for example with HAE some of the women thought they were able to have children or without any issues before the side effects of the medication (Kresser, 2011) (Telford Kralik, & Koch, 2006). Also the side effects of the medication have been found to cause low self-esteem and low confidence due to increased facial hair, loss of hair and a deeper voice because they are different from other women who do not have HAE. Previous literature for chronic illness conditions has found the physical side effects of chronic illness can effect self-esteem and self-confidences (The British Thyroid Foundation, 2015). However other literature on self esteem does differs from the current literature on chronic illness as diagnoses has been found to lower self-esteem, yet this was not found (Nowicki & Ostrowska, 2008). Also self-management was not affected by low self-esteem (Juth, Smyth & Santuzzi, 2008).).

Also previous literature of the lived experience of chronic illness involves the expectations and experience of living with the illness (Paterson, 2001) (Charmaz, 1997). In which expectations and experience are placed on the illness as females have restrictions which intervene in daily life due to the spontaneity and length of an attack and women need to be cautious of possible triggers, interrupting daily life as women are unable to do activities or attend events. This is also supported, as it was found the reality of living with HAE interrupts all aspects of life especially with regards to professional and social life (Gomide, et al, 2013). This is especially shown as these findings reported there was an interruption of daily life with females as HAE attacks can be frequent, spontaneous and long lasting.

However with the regards to trajectory theory the first three stages of chronic illness are applicable to HAE, the first stage is genetic or life style places a person at risk of illness, HAE is a genetic mutation which causes the condition (Corbin, 1998). The next stage is the person notices they symptoms of the condition, in which the females will notice the swelling, the third is the person lives within the constraints of the condition (Corbin, 1998). This was found for HAE as females have the skills to use home therapy and take medication which can control HAE, enabling them to do activities then were unable to do before the medication but they do live within the constraints of HAE as they must consider triggers of HAE in their everyday life. Although the stages after this do not comply with the finding as following stages describe a degenerative condition which HAE is not (Corbin, 1998). A more applicable theory is the shifting perspective theory in which those with chronic illness are in a continuous shift between illness and being well (Paterson, 2004). As the shift occurs so should the interventions for the illness, for example if symptom severity increases the intervention should change (Paterson, 2004). The condition can shift to wellness due to the medication as the symptoms are relieved, however another shift can occur back to being ill because of the side effects.

Legitimacy has links to transformation to feeling well within chronic illness, as the recognition of conditions are crucial to transform the illness, for the transformation legitimacy needs to be from both health care professionals and those from around the person with the illness (Varul, 2001). The legitimacy of the females was mainly questioned prior to diagnosis which may be why there was no crucial

transformation such as the females did not know what was causing the swelling. However legitimacy for medical issues has been found in previous studies of chronic illness (Swoboda, 2006). People can be questioned about the extent, the severity of the symptoms and reality of the condition for those who have it (Richardson, 2005). Findings within this study found that females were questioned about the existence of their disease and the severity of the symptoms because it was difficult to diagnosis as the condition is so rare and without a diagnosis other people would perceive nothing was wrong with the females.

For professional health awareness there is a difficulty in the identification of rare diseases and there can be issues for those with the condition to get up to date research and information about the condition (Rare Disease UK, 2016). Care for those with rare conditions has also been found to be fragmented and poorly conducted due to communication issues between specialists (Rare Disease UK, 2016). It has also been found health professionals need help or a source to gain information about rare conditions (Stewart & Peers, 2013). Also because of the lack of awareness of rare diseases as many health care specialist do not know about the condition or know very little, so on average it takes five years to receive a diagnosis (Stewart & Peers, 2013). Due to the lack of knowledge about rare diseases it can be common for those with rare diseases to receive misdiagnosis for their condition (Stewart & Peers, 2013). These findings revealed HAE could provide difficulty of a diagnosis due to its rarity and commonly health professionals lacked awareness about HAE as they have never come across it. The women also reported

that because of its rarity they commonly received a misdiagnoses prior the HAE diagnosis.

As the literature is not gender specific, as the themes did not reveal issues caused by an increase of attacks during heightened hormonal activity it would be insightful in future research to see if the same findings are found with males with HAE.

Research could also be more generic, as it found that there is a lack of awareness by health professionals on rare diseases and there can be a lengthy wait before diagnosis, so researching into doctors' experience of rare conditions or could prove to be helpful for males and females with rare conditions. One thing which research has shown to have a large effect of chronic illness is lower self-esteem, although some of the research did not apply it was found that the physical side effects did lower self esteem so it would be interesting to investigate this further. However for further investigation into females experience of HAE, researching if they find the medication is worth the side effects.

As this research used self report measures there are issues such as certain negative emotions like denial are repressed into the unconscious in which people cannot report (Baker, Pistrang & Elliott, 2005). However as the questions were open and the participants are the experts as it is there experience, it is possible the participants would become in a state of flow, where their accounts were continuous

thought process as they are fully immersed so they will have reported these emotions which will not be trapped in the unconscious (Csikszentmihalyi, 1996). Trustworthiness can be hard to argue as for example as the consistency to other studies are difficult because no other literature has been conducted into lived experience of HAE, however it is transferable to other studies, such as with rare illness and other findings within chronic illness which are consistent with these results. Dependability is also difficult to prove due to the lack of studies of HAE but when comparing to chronic illness and rare diseases there are similar findings such as rarity of the condition, acceptance and the impact on life and how beneficial medication can be. The credibility can be argued though as when the question and the study was in the early development there was an input from Kristy Brosz, a social worker who has experience of working with people who have HAE, HAE UK and HAE Canada to ensure the questions were suitable and lacked bias before recruiting participants. All have been involved in working with researchers investigating HAE, their input included leaving the questions open so more detail and depth would be provided (Werthamer-Larsson & Kolodner, 1994).

However this research could provide helpful aid for women who suffer from HAE. The research can provide a realistic view of how HAE can impact on a females life, such as through the medication, the medication can provide a relief but does come with side effects which females need to consider before taking it as it can be life changing. Also it informs that HAE can really inhibit a female's life due to the pain and severity of attacks, stopping them from attending work or school and due to their condition they need to take time off, yet when informing others of the pain or swelling

they can be questioned about the existence of the disease. The main hope is that more research can be conducted for those with HAE to get a greater understanding of HAE life and then learn and develop tactics from this research to then help people and improve the experience of HAE as currently there is no research been published. Also due to the issues with doctor's awareness it also demonstrates the need for health care professionals to have source of information especially for rare conditions.

Overall the research has been a beneficial starting point for the experience of women who have lived with HAE and hopefully in the future more research will be developed to gain a greater understanding for health care professionals, people without HAE and those newly diagnosed about how difficult it can be living with the condition.

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## Appendices

### Appendix 1– Advert in HAE UK newsletter for the study

HAEi Newsletter - Issue 8 - December 2015

# HAE News from Around the Globe



United Kingdom [www.haeuk.org](http://www.haeuk.org)

### Status from the CEO

After the excitement of Patient Day, the next event in the calendar of the HAE UK CEO Laura Szutowicz was the UKPIN (UK Primary Immunology Network) meeting. Held in Belfast this year, it was hosted by Dr David Edgar who is chairman of UK PIN and although it is primarily an immunology meeting there was quite a considerable emphasis on HAE this year, with several posters that were on HAE related topics. Also, there was a very interesting presentation from Dr Ania Manson of Addenbrookes Hospital in Cambridge on gene therapy and HAE. But according to Laura Szutowicz the highlight of the meeting was the Shire symposium at which HAE UK member Faye Davenport had been asked to give a presentation. Ruby, her daughter, was diagnosed aged six and is now nine. Faye was diagnosed a little over a year ago. She spoke brilliantly on the impact the diagnosis has made on their lives, not just of her and is extraordinary and many of the clinicians were very moved by her presentation. As one of them said: 'We know about the condition, but we very rarely think of the impact on the person.'

Laura Szutowicz am now in to her sixth month of running HAE UK and she tells us that she has learnt so much and made so many new friends in those six months:

- I have many plans for the future of the organisation, some very long term but others that will be set in place early in 2016. So life promises to continue to be exciting! One of the things I am really looking forward to in 2016 is the Global Conference in Madrid, when I hope to meet and put faces to many of the people that at the moment are names. My first six months with HAE UK would not have been possible without the tremendous help and encouragement I have received from many people. Notable Ann and John Price, who gave me a very thorough grounding in how to run the charity. The other Trustees Ann Harding and Ed Price; our Medical Advisory Panel of Dr Hilary Longhurst, Dr Mark Gompels, Dr Tim Crouch, the Specialist nurses Christine Symons,

John Dempster and Fran Ashworth. Great thanks to Furkhanda Haxton who runs our Facebook page and was wonderfully busy and helpful at our Patient Day; and Rachel Annals, who as well as being our Executive Officer and keeping all the things like the website up and running, is a tremendous source of knowledge and expertise for me to call on. Only remains for me to wish everyone the 'Compliments of the Season' and a very Happy New Year. Forward to 2016!

### Women with HAE

Are you a woman who has HAE? Are you over 18? Then HAE UK and Teesside University invites you to take part in a unique and first of its kind study into HAE. The study aims to gather your experience of attacks and how they have affected your life, your family, your professional life and social life etc., and your experience of living with the condition and has how this has affected your family/ family life, your professional life, relationships etc. Furthermore, it is the aim to gather information on your experience of the healthcare system and medication in relation to HAE. Read more about the study at [www.haeuk.org/news-events](http://www.haeuk.org/news-events).



Peru [www.facebook.com/AngiodemaHereditarioPeru](http://www.facebook.com/AngiodemaHereditarioPeru)

Presently there are nine people diagnosed with HAE patients in Peru. There are two care center, both in Lima. Also, there are a few physicians treating HAE patients. Available HAE medication in Peru is Danazol and tranexamic acid, where there is no access to Berinert, Firazyr, or Ruconest at the moment.

## Appendix 2 – Email advertising study

----- Forwarded Message -----

**From:** "rachel.annals@haeuk.org" <rachel.annals@haeuk.org>**To:****Sent:** Monday, 11 January 2016, 22:02**Subject:** Calling all HAE UK female patients

Dear HAE UK member,

Please read about the study Pippa Adams is doing as her dissertation for Teesside Uni and contact her about taking part in it. It will only take you half an hour and this will be such valuable research into the experience of living with HAE.

The Lived Experience of Women who have Hereditary Angioedema

HAE UK and Teesside University has invited you to take part in a unique and first of its kind study into HAE. Are you a woman who has HAE? Are you over 18? People with types of HAE are welcome to take part in the study. The study will be conducted from November 2015 – January 2016.

The study aims to gather:

Your experience of attacks and how they have affected your life, your family, your professional life and social life etc.

Your experience of living with the condition and how this has affected your family/family life, your professional life, relationships etc.

Your experience of the healthcare system and medication in relation to HAE

There has been no previous study completed on women's living experience of the condition in which you would be the first to share your experience. You will be asked to complete a written account of your experience with HAE. The account should take approximately one hour to complete, however, you will be given two weeks to complete and return the account to the researcher.

The hope for the future of this research is to inform others and develop research into the lived experience of the condition.

If you wish to take part in this study please contact Pippa on [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk) for further details. This study has been approved by SSSBL ethics committee, Teesside University. The study is also being supervised by Stephanie Kilinc and you may contact her [S.Kilinc@tees.ac.uk](mailto:S.Kilinc@tees.ac.uk)

Thanks in advance for your help.

Best wishes,

Rachel

Rachel Annals

Tel: 07975 611789



[www.haeuk.org](http://www.haeuk.org)

[twitter.com/hae\\_uk](https://twitter.com/hae_uk)

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## Appendix 3- Advert on HAE UK website

### **The Lived Experience of Women who have Hereditary Angioedema**

HAE UK and Teesside University has invited you to take part in a unique and first of its kind study into HAE. Are you a **woman** who has HAE? Are you **over 18**? People with types of HAE are welcome to take part in the study. The study will be conducted from November 2015 – January 2016.

The study aims to gather:

- Your experience of attacks and how they have affected your life, your family, your professional life and social life etc.
- Your experience of living with the condition and how this has affected your family/family life, your professional life, relationships etc.
- Your experience of the healthcare system and medication in relation to HAE

There has been no previous study completed on women's living experience of the condition in which you would be the first to share your experience. You will be asked to complete a written account of your experience with HAE. The account should take approximately one hour to complete, however, you will be given two weeks to complete and return the account to the researcher.

The hope for the future of this research is to inform others and develop research into the lived experience of the condition.

If you wish to take part in this study please contact me on [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk) for further details. This study has been approved by SSSBL ethics committee, Teesside University. The study is also being supervised by Stephanie Kilinc and you may contact her [S.Kilinc@tees.ac.uk](mailto:S.Kilinc@tees.ac.uk)

Thank you for taking the time to read this and I hope you are able to help the development and furthering of the knowledge of HAE.

Sincerely,  
Pippa Adams  
Teesside University

## Appendix 4 – Materials used to collect the data

### **Title of project: The Lived Experience of Women who have Hereditary Angioedema**

Participant number:

Thank you for taking part in this research.

Please write an account of how living with the condition has affected your life (e.g. your family, your social life, your professional life), experience of an attack, how medication and the healthcare system has affected your life and any other details you would like to include.

The account is divided into two sections. You can use the questions as prompts but you can also share information which isn't included in the questions. Please share as much or little as you would like and do not need to stay within the space provide. The account should take approximately an hour to complete. I would be grateful if you could complete your account and return it to me within two weeks at [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk)

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#### **Section 1**

Please write an account of how living with the condition has affected your life

#### **Section 2**

Please write an account detailing your experience of a particular attack you have had, including how you felt about it and your experiences just after the attack.

Appendix 5 – Participant sheet which provided participants information about the study

## PARTICIPANT INFORMATION SHEET

**Study Title:** The Lived Experience of Women who have Hereditary Angioedema

I would like to invite you to take part in a research study. Before you decide I would like you to understand why the research is being done and what it would involve for you. This should take about a hour. Talk to others about the study if you wish. Please ask if there is anything that is not clear.

Hereditary Angioedema (HAE) is a genetic disease in which women suffer more attacks than men .Research has been conducted into triggers and treatment but lacks in women’s experience of actually living with HAE.

### **What is the purpose of the study?**

The purpose of this study is to investigate the living experience women have who live in HAE and gain an understanding of what those living with HAE experience and how this affects their lives.

### **Why have I been invited?**

You have been invited to take part in the study as you have a diagnosis of HAE, you are over the age of 18 and you are a woman living with HAE.

### **Do I have to take part?**

If you agree to take part, I will then ask you to sign a consent form. You are free to withdraw from the study at any time and can ask for your data to be destroyed up 2 weeks after sending me your account, without giving a reason.

### **What will happen to me if I take part?**

You will be provided with a document to note down your experiences of living with HAE. There are two sections to this study: the first part is an account of your experience how HAE and the second an experience of an attack. Any information you would like to disclose about your experience of HAE would be greatly appreciated. This written account should take approximately one hour to complete, however, you will be given two weeks to complete and return the account to the researcher.

### **What are the potential disadvantages and risks of taking part in this study?**

There is a slight risk that by taking part within this study you may experience some upset when writing your personal account. If you become distressed during the course of the study please contact HAE UK [support@haeuk.org](mailto:support@haeuk.org) for further support. Also, if when writing the

account you feel unable to continue taking part in the study, you are free to withdraw by contacting me at [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk) and there will be no expectations for you to continue.

**What are the possible benefits of taking part?**

By taking part within the study you will help to provide a breath of knowledge about the reality of living with HAE and you could also help provide other women who suffer from HAE with what can be expected in certain episodes of their life.

**What if there is a problem?**

Any complaint about the way you have been dealt with during the study or any possible harm you might suffer will be addressed. If you have a concern about any aspect of this study, you should ask to speak to the Stephanie Kilinc who will do their best to answer your questions [s.kilinc@tees.ac.uk](mailto:s.kilinc@tees.ac.uk)

**Will my taking part in the study be kept confidential?**

Yes. I will follow ethical and legal practice and all information about you will be handled in confidence. All information which is collected about you during the course of the research will be kept strictly confidential, and any information about you will have your name and contact details removed so that you cannot be recognised.

**What will happen to the results of the research study?**

The results of the study will be wrote up into a report to report the findings of this study.

**Contact for Further Information**

You can contact me at [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk)

Thank you for reading this information sheet.

Appendix 6– Consent form so participants can provide consent to take part within the research

Date:

**CONSENT FORM**

Title of project: The Lived Experience of Women who have Hereditary Angioedema

Researcher: Pippa Adams

Thank you for agreeing to take part in this research. In agreeing to participate you have the following rights and protections as laid down in the British Psychological Society’s ethical guidelines.

- Your participation is entirely voluntary
- Under no circumstances will your real names or identifying information be included in the reporting of this research.
- You may withdraw your data from two weeks after you have submitted an account.
- Nobody, except myself and my research supervisor will have access to this anonymised material in its entirety.

**In agreeing to the terms of this consent form, participants should be aware that any anonymised material is solely for use in the current research project.**

Please initial

I confirm that I have read and understood the participant information sheet dated for ..... the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily.

I understand that my participation is voluntary and that I am free to withdraw at any time up until two weeks after submitting my account without giving any reason. To withdraw your data please contact n3060059@live.tees.ac.uk

I agree to take part in the above study.

-----  
Name of Participant                      Date                      Signature

-----  
Researcher                      Date                      Signature

1 for participant; 1 for researcher

## Appendix 7 – Debrief sheet provided to participants

### DEBRIEF

Title: The Lived Experience of Women who have Hereditary Angioedema

Researcher: Pippa Adams

Aim of research: The aim of the research is to gain an understanding of what life is like for living with HAE and to possibly provide information for other women with the HAE as what life is like living with HAE.

I would like to emphasise that all information provided by yourself will be treated with strict confidentiality and under no circumstances will your name or any identifying characteristics of yourself will used be included in any reports or possible publications. If you have any further questions about this study I would be more than happy to assist and I can be contacted via e-mail on [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk). Alternatively, my supervisor can be contacted [s.kilinc@tees.ac.uk](mailto:s.kilinc@tees.ac.uk)

You can withdraw your data without giving a reason up to two weeks after sending your account, contacting the researcher at [n3060059@live.tees.ac.uk](mailto:n3060059@live.tees.ac.uk)

To receive support you can contact:

HAE UK: [support@haeuk.org](mailto:support@haeuk.org)

Thank you for your participation

## Appendix 8 – Table of themes from the women’s accounts of their experience of HAE

Double Edged Sword			
Theme	Quote	Participant	Lines
Meds	<i>“I was now given different medication, a self administrative syringe that i carry with me at all times. To be able to do this i had to attend 2 training days at the hospital”</i>	A1	60-61
	<i>“I was prescribed Danazol. I had no further abdominal attacks, and only had swellings (mostly my feet but occasionally my hands) when I forgot to take my medication.”</i>	C1	37-39
	<i>“I spent all of my 20’s and 30’s mostly unaffected by HAE, I went back to college, moved away to study dance, graduated, found a job which involved traveling over the country, had nights out, partied, went on holidays, etc. etc. All the typical things you do. I rarely gave HAE a second thought. I took a few danazol every other day, increased the dose slightly if I felt rundown or had a cold. Very occasionally I would get swollen feet, if I forgot to take them, but by working from home it meant that I rarely had to mention it at work.”</i>	C1	40-45
	<i>“my meds were changed to tranexamic acid, I was still mainly symptom free”</i>	C1	54
	<i>“I had my first swelling in years, around 6 months pregnant, in my arm. By 7 months I was in hospital with an abdominal swelling, it was 24 hours before C 1 was administered as I’d never had it before. I remember being blown away with how quick the C1 worked. I went from pain off the scale to sitting up drinking coffee in less than half an hour. “</i>	C1	55-59
	<i>“My consultant taught my husband to administer C1 intravenously to me in 2007 as I was pregnant with my second child and ill with abdominal attacks every 3 days... although I wasn’t experiencing anything like a full blown attack as the C1 would stop it, like magic.”</i>	C1	62-67
	<i>“In 2014 I was taught to treat myself intravenously, this was freedom. I as no longer reliant on my husband for my treatment, I could manage it myself, although we still haven’t spent time apart, we know we can, safe in the knowledge that my HAE can be controlled.”</i>	C1	84-86
	<i>“We were prescribed Epsikapron, an aminocaproic acid in powder form which was wonderful in that it controlled the HAE attacks and changed our lives completely.”</i>	E1	120-122
	<i>“I mentioned this to the doctor about 5 years later and he said it was now in tablet form and prescribed Cyklokapron, a tranexamic acid which did make life easier but unlike Epsikapron it didn't cause us to have diarrhoea... So I was finally able to get a job after Jonathon started school, something I had found impossible to consider with such frequent attacks.”</i>	E1	126-131
	<i>“[Females son] he was then put on Danazol. It was another 2 years after that that my medication was changed to Danazol when I started to get a swollen throat. Danazol does give 100%”</i>	E1	135-136
	<i>“But, having said that, Danazol has really changed our lives and we</i>	E1	157-

	<i>have no regrets about taking it."</i>		158
	<i>"Over time I was asked if I would consider home therapy and be trained for my husband and I infuse with C1 at home. There was no hesitation as I thought this was a god send."</i>	J2	35-36
	<i>"To date we still do this and my life has changed dramatically. I now feel in control of my condition and don't let it control me or my life. If I feel an attack coming on I will infuse as soon as I can and carry on, knowing it will settle down fairly quickly. There are still times however when I do feel really down and think why me and to be honest feel a little bit sorry for myself."</i>	J2	37-40
	<i>"Going on holiday and taking my meds with me gives me piece of mind and again control."</i>	J2	41
	<i>"I can't stress how much my life has changed since home therapy and how I can get on with living my life"</i>	J2	42-43
	<i>"I was 30 when a consultant finally recognised the symptoms and prescribed Danazol tablets, one per day"</i>	L1	11-12
	<i>"The only time I have attacks now are when I forget to take my pills, which is very rare so I am very lucky compared to other sufferers. I also have emergency medication – C1 inhibitor and Icatibant injections"</i>	L1	25-27
	<i>"I was put on danazol, on a very low dose of twice per week. This completely changed my life, I got to spend a lot more time at college, which allowed me to keep up with the workload and make more friends. I also managed to get my health back on track by regularly exercising and eating the right things."</i>	N1	20-23
	<i>"From going on danazol it allowed me to feel confident enough to go to university in Nottingham and study."</i>	N1	25-26
	<i>"The medication I began taking was Danazol, and for the next 17 years, combined with Tranexamic Acid, I remained fairly healthy with just a few attacks. This came at a great time, helping me through my GCSE exams and college years, and enabling me to secure a full time job."</i>	R1	25-28
	<i>"Many months later, and on my second appeal, I was finally granted the funding and was taught to self infuse. This was such a life changing time; I finally had the freedom and confidence to travel away from home and could plan a holiday with my family. Although HAE does have a big impact on my day to day activities, I am determined to not let it stop me doing the things I enjoy and leading a full and active life."</i>	R1	62-66
	<i>"whilst I prepared my medication and administered it at her kitchen table (she has seen me do this many times so was not shocked at the IV!) I then sat and rested on her settee for an hour until I felt almost back to normal, apart from the hand swelling which took a little longer to return to normal. I got over the abdominal swelling within an hour or so of having C1, the hand swell had returned to normal by the following morning."</i>	R1	80-84
	<i>"It was 2008 and I got married and my Husband started giving me my medication. He learnt how to infuse, he was shown by the nurses at Royal London Hospital. He was giving C1 – esterase inhibitor 2 vials twice a week and has been since. My life is a bit easier not having to travel to A&amp;E every 5 minutes but my attacks are frequent"</i>	R2	38-41

	<i>and controlled now.”</i>		
Side Effects	<i>“The move meant a new consultant, this time in immunology. She was amazing. At my first appointment she was horrified that my meds [danazol] hadn’t been reviewed since 1989, and incredibly concerned that wasn’t taking HAE seriously and seemed unaware of the seriousness of it. I was diagnosed with hepatic haemangiomas in the liver in 2004 and my meds were changed”</i>	C1	51-54
	<i>“Epsikapron it didn't cause us to have diarrhoea.”</i>	E1	129
	<i>“Danazol does give 100% control but does come with side effects. We have both suffered blood clots and are on Warfarin for life. I have also had 2 other episodes where Danazol could be to blame.”</i>	E1	136-138
	<i>“where Danazol could be to blame. The first one was a suspected TIA when I woke during the night unable to use my right hand and arm. When I tried to get out of bed my right leg collapsed under me and I immediately thought that I'd had a mini stroke as I struggled to walk to the bathroom. The effects wore off after a few minutes and I resisted [females husband] suggestion that he take me to the hospital. The next morning I was worried enough to see my GP who, having listened to the symptoms and taken my sky high blood pressure sent me to hospital. I underwent all the tests and I hadn't got blocked carotid arteries nor any bleeding from the brain so the doctors were unsure what had been the cause of the brief paralysis.”</i>	E1	138-146
	<i>“The following year it was found that I had blood in my urine but a cystoscopy showed there was nothing wrong and I was discharged. It was later when I looked up the Danazol side effects on the internet I found that it can give the appearance of having had a stroke and also can cause blood in the urine”</i>	E1	146-149
	<i>“Another aspect of the Danazol is the number of years we have now been taking it, 29 years for Jamie and 27 years for me, when it should only be used for 6 - 9 months when used for something like endometriosis as it can cause high blood pressure and high cholesterol levels.”</i>	E1	151 - 153
	<i>“The side effects of the high doses of anabolic steroids that I was given when I was first diagnosed 27 years ago, have left me with hair loss and my scalp shows through. I was given wigs but have not got the courage to wear them. Increased body and facial hair, polycystic ovaries that have robbed me of having children, weight gain, mood swings, high cholesterol and feeling like a freak. People look and point at me in shops like NEXT. Going into the changing rooms leaves me in tears as I see the body fat, the scalp showing my hair loss and body hair that is on most of my body. Children have been known to ask if I am a man. I have no self esteem.”</i>	F1	6-12
	<i>“I miss my fertility the most. I was diagnosed by a consultant who whacked me on a high dose of anabolic steroids and said that would be me for the rest of my life.”</i>	F1	17-19
	<i>“I immediately became pregnant but didn’t worry as I was very healthy and in a happy and stable marriage. When I told the consultant at my next visit he said that I couldn’t carry the baby to term because the Danazol would damage the fetus and so I had to have a termination at 18 weeks, via an a very painful and prolonged</i>	L1	13-23

	<i>induced live birth. I was not told this would happen and had no family there to support me, they thought I was having an operation and would come back later. Obviously I was extremely distressed and upset, particularly at the hospital where I was treated with disgust and I couldn't understand why. Both the nursing staff and other patients ignored me and I gave birth alone in a side ward, and was left for 30 minutes before a nurse came to take the baby away. This is something I will never forget and I will never forgive the doctor for not informing me of this risk. I had a sterilization at the same time so this horrible event wouldn't be repeated."</i>		
	<i>"Although I was told initially that Danazol couldn't cause high blood pressure, something I have had since my teens, more recent studies have shown that it can indeed cause high blood pressure and I do take tablets to lower my BP, which is under control at the moment. I have regular blood tests and liver function tests, liver scans and tests for H Pylori (?) to reduce the risk of stomach ulcers."</i>	L1	27-31
	<i>"I moved off danazol, as it was really affecting my hormones and my working life, I was taking a lot of time off work more than 2 days at a time usually."</i>	N1	30-32
	<i>"This didn't come without side effects though, and being a woman, I did find it difficult at times when I had more facial and body hair than all of my friends, my voice was deeper and I had very good muscle tone. My symptoms over the years have changed frequently."</i>	R1	28-31
	<i>Because of the prolonged use of Danazol, I had regular blood tests to monitor my C1 levels as well as ultrasound scans on my liver area. "Over time I developed a small adanoma on my liver which caused me a little discomfort, and was monitored closely with regular MRI and ultrasound scans."</i>	R1	41-43
	Restriction		
Theme	Quote	Participant	Lines
Daily life	<i>"As i attended a small primary school it was apparent that every student and teacher was now aware of my condition that was not a big deal to me. After that, every fall i had, every bruise and scratch was questioned by teaching staff as if it was a result of my condition. They were given an instruction pack containing what to look for when attacks occur and bruising, scratches and falls where not a part of this. Primary school i felt wrapped me in cotton wool."</i>	A1	14-19
	<i>"I was always a physically active child, participating in as many sports as i could. Once i was diagnosed i was unable to participate in any contact sport incase it sparked an attack."</i>	A1	19-21
	<i>"I would wake up, run to the loo to vomit, then exhaustion induced sleep, followed by off the scale uncontrollable pain, until I had sipped enough water to cause vomiting, which lead to the exhaustion induced sleep etc. This cycle would last days. I would be off school for a week at least."</i>	C1	9-11
	<i>"Going back to work was impossible I wasn't well enough. I was in a cycle of doing as much as I could with the children or home, before I spent another day(s) on the sofa"</i>	C1	64-66
	<i>"My husband regularly worked from home and looked after our children as I was either ill, in pain or exhausted, or getting over an abdominal attack. I'm not sure how we would have managed if his</i>	C1	68-70

	<i>employer had not so understood."</i>		
	<i>"HAE became something that we considered all the time, weather is was questions about where we would store my meds when we went on holiday (camping, B&amp;B's hotels etc) whether we could commit to certain events, either a toddler group or out with friends. Or if it was a good idea to go out, play in the park, meet friends. I found it didn't matter what I did or didn't do. What I ate or didn't eat I would have abdominal attacks, swelling in a hand or foot, but it would always end up as an abdominal attack, It was impossible for us to spend time apart. What would I do without my husband to treat me? If I was away with the children who would look after them if I was ill? We did feel very restricted, and frustrated. My husband was my carer! That felt ridiculous."</i>	C1	70-78
	<i>"As a child I suffered an HAE attack approximately every 4 weeks and these attacks were always the internal swellings of the intestines which caused severe stomach pains and acute sickness and lasted anything from 24 - 48 hours. Added to this my stomach was always sore for sometime afterwards due to the excessive vomiting. This meant I that I regularly had time off school which didn't go well with the headmistress of the convent school I attended."</i>	E1	1-5
	<i>"it was difficult to make any plans as there was no guarantee that Jamie or I would be attack free. I also started to have swellings on my arms legs and genitals but not my face. With Phil having changed jobs there were no more early afternoon finishes as with his other job and I had to cope with my attacks and looking after the 3 boys as best I could."</i>	E1	81-85
	<i>"It leaves me feeling as if I have been robbed of a normal and fulfilled life. It has lost me the chance to get a degree, divorced from a husband who eventually found someone else as we didn't do anything with our down time as I would be in pain or resting."</i>	F1	1-3
	<i>"It has lost me 4-5 jobs. Numerous absences that could happen so suddenly meant that I became unreliable and colleagues began to resent having to pick up my workload. It has meant I have spent too many years trying to live on benefits and scraping by."</i>	F1	3-6
	<i>"Trying to find a partner is a nightmare as who wants to live with a freak like me? Appearance is normally what attracts men and there is nothing physical about me that would appeal to them. Those that are interested are only looking for sex! 16 years of being on my own and I have given up. I'm destined to be alone forever"</i>	F1	12-15
	<i>"Trying to find a partner is a nightmare as who wants to live with a freak like me? Appearance is normally what attracts men and there is nothing physical about me that would appeal to them. Those that are interested are only looking for sex! 16 years of being on my own and I have given up. I'm destined to be alone forever"</i>	F1	15-17
	<i>"Trying to keep a full time job with HAE became impossible and then being diagnosed with chronic fatigue syndrome (CFS) was another blow. Suddenly I had to learn how to pace my life and work part time. I am limited in the type of jobs I can do. I have to declare that I have this very rare medical condition to potential employers and they have no idea of what it is and what they do read online is about how it can kill so it scares them."</i>	F1	23-28

	<i>"My life was not meant to be like this. I was supposed to get my degree, get a job, meet a guy and have a family and HAE has robbed me of all that."</i>	F1	33-34
	<i>"The caring NHS let me down greatly and eventually led to 2 years of being put through the sickness &amp; absence procedure until I told them to terminate me as the stress of it all wasn't helping the HAE. Stress is a major trigger for this condition. I loved working in the NHS and had worked as a nursing auxiliary and lab assistant which were both full time and demanding jobs. I moan having to leave the NHS as I saw it as a career."</i>	F1	75-79
	<i>"Stress impacts my experience of HAE. Stressful work in particular will lead to an increase in episodes. To demonstrate this, I moved from a job which was full time and demanding of my time and attention outside of working time, to a part time job with few demands outside of my working pattern. I went from an episode every two weeks to not having an episode for a year. Therefore, I know that I can best manage my health when I am working in a part time position in which I know my time off is my own."</i>	J1	4-9
	<i>"Going on holiday was always a risk and one year when I went on holiday with my then fiancée and both my hands and arms swelled up and I had to stay in the room for 3 days( putting my hands in bowls of cold water to try to take swelling down)."</i>	J2	19-22
	<i>"The impact on my life in the early days in relation to work was that I had to have quite a lot of time off, which I didn't like and only did this if I was having a really bad attack. Socially it would stop you in your tracks as it could take days to recover from an attack."</i>	J2	23-26
	<i>"This condition has affected every aspect of my life since childhood. At school I missed many lessons and exams mainly because of sickness, diarrhoea and abdominal cramps. As I got older the swellings became more common in my extremities."</i>	L1	1-3
	<i>"My working life was affected considerably but fortunately I had very understanding and sympathetic employers"</i>	L1	9-10
	<i>"From the age of three I was diagnosed with the condition and ever since I have had weekly/twice a week attacks. Throughout my educational life it meant a lot of time off school/college/uni, which has meant being severely behind on work and having to catch up as quickly as I possibly could."</i>	N1	1-4
	<i>"However, this really affected my social life as a child as I wasn't able to keep many friends, as by the time I was back they had all moved around friendship groups etc. I remember feeling very alone"</i>	N1	6-7
	<i>"My teenage years was probably the time when it most affected my life to the point where I was just constantly having attacks every 2-3 days and them being very severe. Due to this it ended up so bad, I ended up being very depressed, gaining a lot of weight and reaching breaking point. From all of this built up depression, lack of friends and tension I ended up in intensive care with a bad throat swelling."</i>	N1	10-15
	<i>"However, I noticed during my last year all the stress and tension from my final project meant I was paying regular visits to A&amp;E with throat swellings. I noticed that after the year was complete my swellings went down to a minimum again and seemed to be more manageable."</i>	N1	26-29

	<i>"The first time I did it was me just sat at home with some instructions online (silly I know – but it worked). Now I regularly inject every 4-5 days with 3 x 500 iu C1 Inhibitor but I am still unable to keep a stable work and social life, as sometimes I wake up with bad swellings. Now my main swellings I have are stomach ones. Sometimes even c1 inhibitor can take a while to even counteract them, which can mean 2 days of pain and exhaustion."</i>	N1	33-37
	<i>"I would say living with the condition has affected many aspects of my life such as day to day tasks, walking and writing etc. my social life, being unable to meet friends due to illness or being absent from school/work thus missing out on big events, my education, having to constantly catch up on missed work and try keep on track as best I could and mainly my job, I work in marketing so any stress or tiredness really effects my condition, thus I have a lot of time off work with sickness."</i>	N1	38-42
	<i>"My health was at an all time low, I had lost all my friends and gained a large amount of weight from the pressures of school and trying to avoid my illness at all costs. I paid a large price!"</i>	N1	66-68
	<i>"Throughout my early school years I was regularly sick and suffered abdominal cramps, at least once or twice a fortnight, and on the odd occasion I had swelling in my hands or feet. This was quite embarrassing as I was growing up and I did feel like an outcast on those days."</i>	R1	4-7
	<i>"Growing up was particularly difficult because I was not diagnosed and had no idea what was wrong with me, which did make me feel like an outsider, I lost confidence and became quite shy."</i>	R1	66-68
	<i>"Growing up I was always ill especially having so much time off school, not going out as much like my friends did."</i>	R2	7-8
	<i>"I couldn't wear high heel shoes, tight clothing, walk a long distance, standing on my feet long periods of time, always prone to infection all brought on my attacks."</i>	R2	8-10
	<i>"I couldn't have a normal life like my friends it was so upsetting I didn't do very well at school. Having so much time off school and being anxious with exams resulted in more swelling....."</i>	R2	14-16
	<i>"I started working as a PA around 20 years old, I loved my job but my health was awful I had so much time off work. I couldn't really progress because my health stopped me from doing that"</i>	R2	31-32
	<i>"When i had been diagnosed with became clear that when i was 6 years old i had experience an episode of swelling in my abdomen, resulting in 3 days off school."</i>	A1	8-10
Attacks	<i>"When i was 14 years old i had to have a tooth removed due to over crowding in my mouth. This meant i had to have 2 drips over the course of two days with an overnight stay in the RVI hospital in Newcastle (where my consultant is now based). This was a traumatic and what i felt an over the top experience. Before i was diagnosed i had numerous fillings in my teeth and never had any problems with it. But dental trauma is an apparent trigger."</i>	A1	29-33
	<i>"But dental trauma is an apparent trigger. I have very crooked teeth but was unable to have a brace because of the constant dental trauma that i would undergo for around 1 year. I now have to live with teeth that are not straight and people making comments like</i>	A1	33-37

	<i>"why did you never get a brace?!". I have never been happy with this decision. I fought for a brace, i don't want to be the only person my age with awful teeth while everyone else was offered a brace to help them."</i>		
	<i>"After this the lady asked me what my plans where for the future, in regards to family. I told her that it is all i have ever wanted. I want children as soon i was in a position to have a child and fully care for them. ...My family and friends always knew that i was in love with the idea of having children. Always have been and always will be. Until the geneticist told me that it might not be possible to conceive naturally. At that point my world crumbled around me... get some funding on the NHS because there is a medical reason behind the reasons for IVF. This is something i would consider. She then went on to tell me that the child is not the only one at risk. During pregnancy, just like puberty the risk of a HAE attack increases massively due to hormones. I had always been told that the sites of swelling are hands, feet, abdomen and airways. I was never told that swelling can occur in the uterous and the surrounding areas, especially throughout pregnancy and birth. This leaves a massive risk on the child and the carrier. The risks are massive i was told"</i>	A1	69-82
	<i>"I developed a problem where the ring and small finger on my right hand curled up after I had rung a cloth dry. The pain was terrible and it took 3 months for doctors to decide it was a trapped nerve in my elbow which could only be cured by an operation. I asked the consultant if my having HAE caused any problem and he snapped at me that no it wouldn't. I thought he felt I was trying to tell him how to do his job but it seemed a reasonable question to me. On the morning my operation was due I was sitting in the hospital bed when a nurse came and asked what I would like to eat for my dinner. Puzzled I told her I was having an operation at noon and wasn't allowed to eat anything. She I wasn't having an operation that day because the anaesthetist had read a paper on HAE a fortnight earlier and was refusing to give me anaesthetic until I'd had the necessary preparation!"</i>	E1	198-206
	<i>"I have mentioned that stress is a trigger for HAE but unbelievably, good stress is also a trigger. We can't get excited about things like family events as that can trigger a swell! The number of times I have made plans to visit family or friends and had to cancel at the last moment as a swell has been triggered. We have to learn to stay calm."</i>	F1	47-50
	<i>"During my teens it was unusual for me to go more than two weeks without having 2 or 3 days of intensive vomiting and diarrhoea. I was always very thin - I weighed 6½ stone when I got married at 19 and only weighed 8 stone when 9 months pregnant! I was often hospitalised for dehydration"</i>	L1	6-9
	<i>"My first attack was when I was 8 years old. I visiting the dentist for a routine checkup not realising a day later. I woke up looking like the elephant man. My parents and I always wondered why and how what happened that day. After a week later my face seemed to go down without any medication was in bed hiding could hardly eat or drink. I was so upset and so worried what happened"</i>	R2	3-7

	Rarity of the Condition		
Theme	Quote	Participant	Lines
Legitimacy	<i>"I remember mentioning that I had HAE swellings at work and on one occasion I took a day off as my feet were swollen so I couldn't get in. I wasn't believed; everyone thought I'd taken time off with a hangover as there was no "evidence" of swelling the next day."</i>	C1	29-31
	<i>"She was fierce and terrifying to a small child and in front of the entire class would sarcastically note that I had been spoiled yet again by my grandparents who allowed me have time off school whenever I felt like it."</i>	E1	5-7
	<i>"[Females husband] I has always been very supportive and helpful about my HAE attacks and it was he would insist I go to see my GP about the frequent attacks when I was always reluctant to go (I'm still the same, I will do anything to get out of seeing a doctor if possible) but the psychosomatic diagnosis shook him and he admits he began to doubt me. I couldn't believe that anyone could think I would put myself through all the misery of such frequent attacks and hours of pain and being physically sick, let alone someone who had seen me in the throes of such attacks so many times. He says now he didn't really doubt me but it did make him wonder."</i>	E1	178-184
	<i>"My colleagues, I felt, didn't really understand how bad this pain was and I always got the impression that they thought I was making it out to be something more than it was. I always worked when I had attacks and some shifts, I would be on very strong opiate painkillers and still trying my best to do the job."</i>	F1	70-73
	<i>"Because the condition is not common, people struggle to really understand it. This includes employers, colleagues, friends etc."</i>	J1	14-15
	<i>"feeling as though a lot of people were judging me, teachers included for "skiving", as they didn't understand nor wanted to understand my illness."</i>	N1	8-9
	<i>"It was difficult though, as friends and teachers didn't understand, and I had no reason to explain why I had severe cramps and felt sick, tired, faint and unable to concentrate at times."</i>	R1	12-15
	<i>"This went on for years and years I use to think people thought I was making it up and especially my family. The swelling was visible"</i>	R2	11-13
Professional Awareness	Therefore from the age of 13 i was put on the combined pill to help them [period] be regular and i was told that once that was sorted the rest would sort it out itself. I was on this pill until [her consultant] informed me that I should have never been put on that pill. The combined pill in fact triggers attacks in HAE patients. From there on i was on the mini pill, therefore now i do not have a period and i started to feel normal....The problem with going to my GP is that they did not have a clue what HAE actually was with it being such a rare disease, therefore they were unable to prescribe the correct medication leading to unnecessary problems for me.	A1	52-59
	<i>"My parents never took me to the GP with these attacks as my mother had the same symptoms and had be told that there was nothing wrong with her other than "her body couldn't cope with her emotions" and she should "pull herself together"."</i>	C1	11-14

	<i>"It wasn't until 2003 when I was moving away, buying a house and talking about having a family that I thought I should see my consultant again (I hadn't seen him or any other consultant since 89), in the hope that he could answer questions about the new treatments available for HAE and suggest where I could be referred when I moved. I never saw him personally, just a junior doctor in his team and a SHO who knew very little about HAE, and were no help"</i>	C1	46-50
	<i>"The family referred to my frequent sickness as bilious attacks and these began when I was 3 and I remember as a small child being taken to see various doctors by my granny though none of them could find what was causing the attacks. Some of the advice given was a bit bizarre such as saying I mustn't sit on anything cold when playing out and that I should have my long hair cut as it was draining my strength"</i>	E1	10-14
	<i>"When [females son] had his first facial swelling he had eaten a prawn the day before so it was decided that he was either allergic to prawns or penicillin. The result of this was that whenever he was taken to hospital we were asked if he had been eating prawns which felt like an accusation that we were the sort of parents who were uncaring enough to let that happen"</i>	E1	88-91
	<i>"On one occasion when my right leg was swollen I went to see the doctor and he said he would refer me to the local hospital. Of course, by the time I went to the hospital my leg was back to normal and the consultant decided I wanted an operation to remove fat from thighs. With my constant bouts of sickness I weighed about 6 stone so I hadn't really any fat to remove and I felt he was thinking I was just wasting his time"</i>	E1	94-98
	<i>"When I went back to see the GP to find out the result of my hospital visit I was given some more pills. I took them but they were no more help than any other pills and then to my surprise I discovered that they were tranquillisers! I returned to my GP to ask him why I'd been prescribed tranquillisers. He replied that it was because my attacks were psychosomatic. In shock I just said "What"? He simply said "It means you're doing it to yourself. It's all in your head". I truly couldn't believe that anyone could possibly think I would inflict those horrendous sickness attacks on myself but I could tell from the doctor's tone and delivery that he totally did believe this."</i>	E1	98-104
	<i>"[Females consultant] has said not to blame the doctors who failed to understand the problems as HAE is very rare."</i>	E1	186-188
	<i>"When I saw my GP after [sons birth] birth because my attacks had increased he put it down to tiredness as I had a toddler and a baby. It was frustrating as I knew I was tired but my problem was the frequent sickness bouts."</i>	E1	189-191
	<i>"When I was expecting [my son] I asked my GP why I was free of attacks during my pregnancy and he said "Because you're content". I had not expected this answer at all. I had asked a serious question because I felt there must be some medical reason the attacks stopped when I was pregnant and found his comment infuriating and patronising"</i>	E1	192-195
	<i>"Years later Jamie was in hospital he was left in a cubicle with his</i>	E1	224-

	<i>medical notes and, bored with the long wait, had a look inside. He found the report of that conference and was shocked to read the comment that "The mother was lying about her swellings because she didn't mention them on the first day!" He found it incredible that, despite being diagnosed I wasn't believed. It still really annoys me to this day to think about that comment."</i>		228
	<i>"Despite telling the staff about HAE they didn't believe that was the problem and insisted I must have swallowed a fish bone. I told them I hadn't eaten any fish but had had chicken the previous evening so they decided I had a chicken bone stuck in my throat which I knew I hadn't but they persisted with this theory for some time"</i>	E1	236-240
	<i>"Even the ESA assessment that I had to undergo to get onto benefits was a struggle as the health professional doing the assessment said it was classed as cardiac because of the Angio part of the name! We argued for 10 minutes over this!"</i>	F1	28-30
	<i>"When I have to present at A&amp;E I am aware that there is always the possibility I will have a bit of a battle on my hands. Some A&amp;E doctors want to be the expert in the situation, even though they may have never even seen someone with HAE before. I can overhear the nurses tell them what it is and what they need to do about it, yet they try to pretend that they are knowledgeable about it."</i>	J1	33-37
	<i>"My mum would always ask the doctors if my symptoms were something to do with me taking the pill (as she had problems with this many years ago and as I had the same blood group she just wondered). They all said no and that it would benefit me to keep taking it as it would regulate my periods. IF ONLY THEY HAD LISTENED"</i>	J2	7-4
	<i>"I spent a lot of time in and out of hospital seeing different doctors and consultants only to be given different diagnosis's each time such as: Gastro-enteritis Irritable bowel syndrome And even told it was psychological and I was making myself ill. Along with this I underwent various operations listed below before I had a confirmed diagnosis. At the age of 19 I had my appendix removed At the age of 21 I had an ectopic kidney re-routed (situated in my pelvic area)At the age of 23 I had to have my kidney removed as was banging against my pelvic bone as they had moved it during the previous operation"</i>	J2	8-17
	<i>"By this time I couldn't talk and was that upset I didn't know what to do. I took my emergency letter in and flashed my bracelet only for them to tell me to sit down and wait to be seen. Once my sister came in and played holy war with them and fully explained they rushed me through to resus. I was given oxygen and put on heart monitor. By this time my husband arrived and was able to tell them everything they needed to know as I couldn't. It took them quite a while to get hold of some C1 and during this time I had the camera up my nose and down my throat a few times"</i>	J2	52-59
	<i>"Throughout childhood I was told by doctors that I had to pull myself together because the illness was caused by nerves, or the vomiting and cramps were caused by acidosis or allergies, although sensitivity tests did not reveal anything. I was also told I would grow out of it as I got older, when in fact the conditions worsened!"</i>	L1	3-6

	<i>“Not many people have heard of it and very few health professionals have actually witnessed the condition. This means that when I take the C1 inhibitor to A&amp;E for treatment, quite often they refuse to administer it for fear of reprisals - even after reading the letter from my consultant. Only after speaking to my consultant over the phone and discussing it with their colleagues and superiors, do they agree to administer the C1 inhibitor – sometimes too late to be effective! This does worry me enormously, especially when travelling abroad”</i>	L1	42-47
	<i>“The whole of my face swelled up, my eyes were completely shut and the doctor who visited me at home called an ambulance, he thought my husband had beaten me up! The ambulance crew also thought that I had been beaten and didn’t believe my story. No one had heard of HAE.”</i>	L1	54-57
	<i>“When I was about 18 months old my parents began to notice that I was being sick on a regular basis, but the doctors were not convinced there was anything abnormally wrong with me.”</i>	R1	2-4
	<i>“Over the years I was tested for various food allergies and spent months on special food diets, but none of these seemed to make a difference. My problems were classed by doctors as migrane tummy, for which I received no treatment.”</i>	R1	8-10