

= THE POWER OF PARTNERSHIP: SHARED DECISION MAKING IN HAE



SHARING LIFE'S JOURNEY

This project was initiated and funded by BioCryst UK in partnership with HAE UK. This whitepaper has been collaboratively developed with input from multiple stakeholders. The research that underpins this report was led and conducted by ZPB Associates and insight was gathered through patient and HCP surveys, interviews and workshops.



PEOPLE HAVE NO CLUE WHAT IT FEELS LIKE TO HAVE THIS CONDITION. YOU DON'T KNOW DAY TO DAY IF YOU'RE GOING TO HAVE AN ATTACK OR HOW HAE IS GOING TO IMPACT YOUR LIFE.

Female, 46-55, Wales

HAE HAD AN IMPACT ON MY MENTAL HEALTH AS MY DAD PASSED AWAY BECAUSE OF IT. IT IS HARD LIVING WITH IT, AND I AM ANXIOUS ALL THE TIME. I TRY TO BE CALM AND NOT TO STRESS. THIS IS WHY I WORK PART TIME NOW, BUT I KNOW HAE WON'T GO.

Female, 26-35, London



MY CONSULTANT DUG INTO HOW ATTACKS ARE AFFECTING ME MENTALLY AND ASKED IF IT WAS AFFECTING MY RELATIONSHIPS. THEY WENT THROUGH EVERYTHING, AND THEY LISTENED.

Male, 46-55, Scotland



I WANTED THE CLINICIANS TO LOOK AT ME AS A PERSON RATHER THAN A FOLDER OF NOTES.

Male, aged 56+



FOREWORD

The view of some clinicians and patients living with hereditary angioedema (HAE) for many years is: 'If it ain't broke, don't fix it.' However, over the past decade, we have seen many new treatments become available, helping some HAE patients live their lives more fully, minimising the stress of having painful and debilitating attacks that can prove fatal if not managed promptly with medication.

Our in-depth research with people living with HAE has found that 25% of patients felt that coping with everyday life was a challenge. Additionally, less than 33% of patients discussed their individualised treatment plan at their last appointment, which suggests there is room for improvement in many of the conversations that patients are having with their healthcare team.

This report is the result of an ongoing and committed collaboration between patients (the majority of whom are HAE UK members), specialist immunologists, nurses and BioCryst. We aimed to understand and develop a framework for shared decision making in HAE, looking at how the conversations between patients and clinicians can be improved to develop better long-term management and improved quality of life for the 1,265 currently diagnosed patients in the UK who live with HAE.

The motivation for this report was set out by the 2021 update of the international World Allergy Organisation (WAO)/European Academy of Allergy and Clinical Immunology (EAACI) guidelines for the management of hereditary angioedema. It was the first time we saw reference to shared decision making in HAE guidelines due to increased HAE treatment options providing patients with the opportunity to have a greater role in decision making. The guidelines recommend that all patients have an action plan developed in partnership between them and their clinician.

Supporting patients

At HAE UK we work with patients to help them better understand their condition and to disseminate the latest information about treatment options. We want to see patients feel confident in having open and honest conversations with HCPs about treatment options and care, deciding together on a management plan.

Shared decision making

Shared decision making is a process that can help create stronger relationships between the HCP and patient. Better and more open discussions around their management plan and lifestyle will capture the points that are important to patients like pregnancy, menopause, the sometimes awkward or embarrassing side-effects of medication, and acknowledging the toll the disease has on mental health. It is important that everyone with HAE has the opportunity for regular check-ups with their HCPs, and shared decision-making tools can help to improve the quality of those interactions.

I extend my sincere thanks to those people with HAE and the healthcare professionals caring for them who have given up their time, played a role in developing this report, and for dedicated efforts to bring about positive change in the lives of those affected by HAE.

Angela Metcalfe
Chief Executive of HAE UK



PATIENT BURDEN OF HAE

Hereditary angioedema (HAE) is a rare genetic disease affecting around 1 in 50,000 of the UK population. People living with HAE experience unpredictable attacks that result in significant tissue swellings, and attacks that involve the throat are particularly dangerous as they can block the airway. While the cause of HAE attacks is not always known, stress, trauma, infection, and hormonal changes are recognised triggers.

HAE is an inherited condition in which parents have a 50% chance of passing it on, but it can also occur spontaneously in people with no family history of the disease. It can take many years for HAE patients to receive a correct diagnosis due to varying levels of awareness of HAE. Often, they are referred to allergy services as abdominal attacks can mimic symptoms of gastrointestinal conditions.

The patient burden of HAE is substantial, affecting an individual's life in many ways, including physical health, emotional wellbeing, and overall quality of life. The unpredictable nature of attacks can make normal activities, such as work, study, and travel more difficult for people with HAE. The swelling experienced by those with HAE can be extremely painful and may interfere with daily activities and self care.

There is currently no cure for HAE, so treatments focus on the management of symptoms to improve quality of life for patients. This can involve treatment during attacks, avoiding factors or situations that can trigger attacks, and long-term preventative treatments aimed at reducing the frequency and severity of attacks.

YOU DON'T KNOW IF
TODAY'S THE DAY YOU
HAVE A BAD ATTACK.

Male, 46-55, Wales



Patient research

In a survey of HAE UK members in November 2022:



32% OF PATIENTS LISTED
UNPREDICTABILITY OF ATTACKS
AS THE BIGGEST CHALLENGE
OF LIVING WITH HAE¹



26% OF RESPONDENTS
FELT COPING WITH EVERYDAY
LIFE WAS A CHALLENGE¹



25% OF RESPONDENTS
FELT MANAGING THEIR
CONDITION WAS A CHALLENGE¹

Travel and mental health were the most commonly listed priorities when it came to living with HAE.

TREATING HAE

Treatments for HAE aim to either reduce the frequency and severity of attacks (prophylactic therapy) or manage attacks when they happen (on-demand therapy). The 2021 update of the international WAO/EAAACI guidelines for the management of hereditary angioedema recommend that all attacks are considered for on-demand treatment and are treated as soon as possible, with mandatory treatment of an attack that affects, or could affect, the upper airway.ⁱⁱ

The 2021 EAAACI/WAO guidelines state that the treatment goal for HAE is to effectively manage the disease to allow patients to live a normal life, which can currently only be achieved by long-term prophylaxis.ⁱⁱ Therefore, guidelines recommend patients are evaluated for long-term prophylaxis at every visit.ⁱⁱ

NHS England has developed a treatment algorithm based on clinical and cost effectiveness to aid decision making for HAE specialists and patients. The algorithm recommends licensed therapy for both on demand and prophylactic treatment. Patient choice and the ability of the patient or carer to use the required administration technique are both part of the decision-making criteria.

- The algorithm recommends on-demand treatment in patients who have less than two attacks per month. For patients over 12 years old experiencing two or more attacks per month, the algorithm recommends licensed oral prophylaxis.
- For people over 12 years old experiencing eight or more attacks per month despite licensed oral prophylaxis, injectable prophylaxis is recommended.

Importance of an individual treatment plan

HAE symptoms can manifest very differently for individual patients. Patients and their consultant should work together to create an individual treatment plan tailored to what is going on in a patient's life at that time. Our research indicates that these conversations may not be happening as often as needed during clinic consultations; less than 33% of patients discussed their individualised treatment plan at their last appointment and only 17% discussed deciding together with their consultant to try a different treatment.ⁱⁱ

Guidelines recommend individualised treatment plans are carefully developed by shared decision making and should address preventive measures, home care and self-administration, and include an effective emergency treatment plan to be used during attacks.ⁱⁱ

The choice of treatment and management plan should be determined based on the individual's current situation, the frequency and severity of attacks, the impact on quality of life, and individual preference. Regular patient education, monitoring of symptoms, and psychological support are integral components of the overall management plan.



WHAT ARE PATIENTS HOPING FOR WHEN MEETING WITH THEIR HCPs?

Our research indicated patients across treatment types may have a desire to change treatment but are limited by factors such as availability, changes to their healthcare team, the willingness of their HCPs or clinic to try new things, attack frequency, and severity of condition and availability of information from the healthcare team. Some patients have expressed feelings of guilt around the cost of treatment or using NHS resources, a self-imposed barrier that is sometimes reinforced by clinicians. Patients also experience anxiety around the thought of changing treatments if they have been on a certain therapy for a long time or had reassurance from previous consultants about the treatment they are already on.

MY OLD CONSULTANT SAID, 'DO YOU KNOW HOW MUCH THESE COST, ARE YOU REALLY ILL?' WHEREAS MY NEW CONSULTANT TOLD ME NOT TO WORRY ABOUT DRUG COSTS AS A TRIP TO A&E WOULD COST A LOT MORE. SO, I FELT BETTER ABOUT TAKING THE DRUGS WHEN I NEEDED THEM.

Female, aged 50-55

- Hesitancy to try new treatments: Sometimes treatments for HAE can cause side effects that can impair quality of life for patients. A fear of potential side effects or a lack of perceived efficacy can result in patient hesitancy to try new treatments – even if their current treatment is not optimal.
- Treatment cost: Emphasising cost when deciding on a treatment can influence patient preference and negatively impact shared decision making. Some patients don't use injections for all of their attacks as they think they are expensive or that they should just 'ride the attack out' – which can put them at increased risk of a serious or fatal attack and lower their overall quality of life.

DON'T ROCK THE BOAT. I MEAN, THERE'S A LITTLE BIT OF CONCERN I THINK UNDERNEATH FOR THE LONG-TERM AFFECTS.

Male, 56+, Wales

- Lack of engagement with care: Patients can lack motivation to engage in care when they have not had attacks recently or know they can access emergency care when they need it. Increased attack frequency would need to coincide with their upcoming appointment, otherwise it wouldn't feel important to discuss.
- Feeling like their condition is not understood: Our research indicated that some HAE patients feel like their healthcare teams do not fully understand their condition, particularly calling out a lack of GP knowledge around HAE. There is a clear need to support communications with non-specialist HCPs.
- HCP treatment preferences: Patients feel that some HCPs will try to steer the patient towards drugs they don't like or away from drugs they do, which can impact shared decision making.

"We know some patient enquiries about change of treatment receive a response focussed on the costs rather than on explaining the clinical criteria that denote availability of the specific treatment."

Angela Metcalfe,
Chief Executive, HAE UK



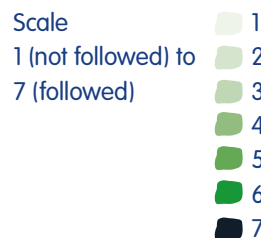
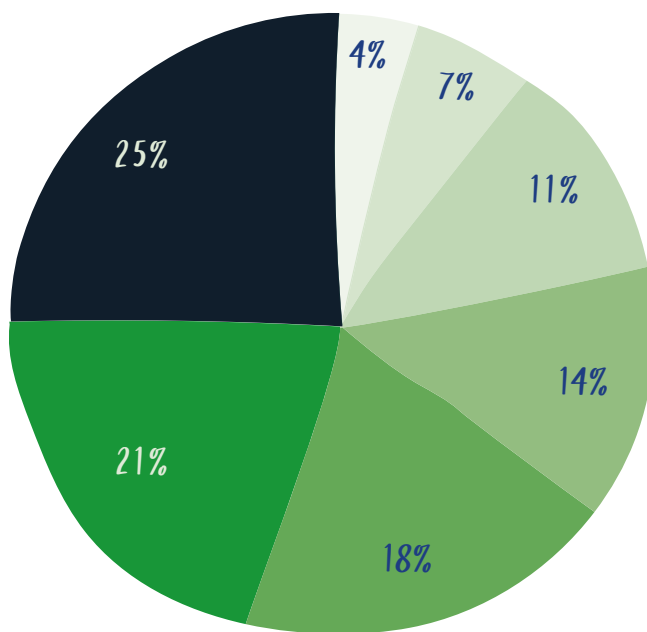


WHAT PATIENTS SAY ABOUT SHARED DECISION MAKING

What is shared decision making?

Shared decision making is a process in which healthcare professionals work with their patient to reach a decision about care. This process involves healthcare professionals sharing information on different options and ensures the patient's preferences, lifestyle, and priorities are considered when making a plan of action. When done well, shared decision making enables patients to take a more active role in decisions regarding their treatment and care.

We asked HAE patients if they feel that they share decisions about their care with their healthcare professional.



SHARED DECISION MAKING NEEDS TO BE A CONTRACT BETWEEN THE PATIENT AND CLINICIAN. THERE NEEDS TO BE MUTUAL RESPECT FOR YOU AND YOUR EXPERTISE AS A CONSULTANT AND ALSO FOR YOUR PATIENT'S AUTONOMY, HUMANITY, AND THEIR EXPERIENCE OF DISEASE. SO, I FEEL SHARED DECISION MAKING IS A CONTRACT AND AGREEMENT BETWEEN ME AND MY PATIENT.

Dr Sorena Kiani,
Consultant Immunologist



Patient survey results

Our research indicated a high awareness of shared decision making amongst HCPs and patients; however, implementation is inconsistent: 26% of patients are unfamiliar with the concept of shared decision making. Patients were also asked to answer on a scale of 1-7 if they felt like the process of shared decision making is followed by them and their main healthcare professional when discussing their treatment. 36% of patients felt as though this was average or below average.¹ Satisfaction with care teams ranged hugely for patients, with most praise focused on honest discussions and exploring options.

I ALWAYS FEEL THAT MY CONSULTANT IS COMPLETELY OPEN TO MY IDEAS AND HOW I WANT TO BEST MANAGE MY CONDITION. WITHIN REASON, OBVIOUSLY, BUT IT'S VERY MUCH A COLLABORATIVE DECISION RATHER THAN ONE THAT'S MADE BY JUST ONE SIDE.

Female, 36-45, West Midlands



MY DOSAGE ISN'T REALLY CHANGING; MY MEDICATION ISN'T CHANGING. WE'LL QUICKLY LOOK AT IF I'M HAVING ANY ATTACKS, THEN HE'LL JUST SAY THERE'S OTHER OPTIONS, BUT WE ALWAYS REACH THE SAME CONCLUSION.

Male, 26-35, West Midlands



Lack of detailed guidance for shared decision making in HAE

There is variation in how shared decision making is carried out, both at organisational and individual HCP level. HCPs agreed that for HAE, formal guidance or protocols could be of benefit to specialists in training or used as a basis for audit to reduce variability between centres. Most support and training in shared decision making takes place within specialist HAE centres, and there appears to be limited sharing of best practice among HCPs outside these centres, leading to a variability in patient experience.

Communication with non-specialists

Both patients and HCPs feel patients are involved in care; however, there is a need to support communication with non-specialists, with patients often feeling their condition is not taken seriously by non-specialist HCPs. This can lead to patients feeling as though there is not a holistic approach to their HAE care.

I WANTED THE CLINICIANS TO LOOK AT ME AS A PERSON RATHER THAN A FOLDER OF NOTES.

Male, aged 56, London



I THINK THE NURSES SEEM TO UNDERSTAND THE CONDITION A LITTLE BIT MORE, WHEREAS I THINK WITH THE CONSULTANT, IT'S MORE OF A BLANKET TREATMENT.

Female, 46-55, North West England



WE HAVE HAE SPECIFIC CLINICS EVERY TWO MONTHS. THE CLINICS ARE SET UP SO PATIENTS MOVE THROUGH A PATHWAY, SEEING DIFFERENT MEMBERS OF THE MULTI-DISCIPLINARY TEAM THROUGHOUT THEIR APPOINTMENT. BY ALLOWING PATIENTS TO GO THROUGH A JOURNEY, THEY GET ALL THE INFORMATION THEY NEED AND CAN ASK QUESTIONS THROUGHOUT THE DAY ONCE THEY HAVE DIGESTED THAT INFORMATION.

Consultant Immunologist, East of England



The patient-clinician relationship

HCPs noted that shared decision making is a process that cannot be achieved straight away. It takes time to develop trust between patients and their clinician and for clinicians to be able to satisfy patients. Therefore, if a patient doesn't see the same clinician consistently, a strong patient-clinician relationship cannot form, which is crucial to shared decision making. One patient, female, aged 20, said, "They don't even know my name. I have to start again with who I am and all my extended family medical history each time."

Some patients reported finding it easier to speak to their nurses about their condition as a result of the different kind of relationship that nurses are able to form with patients.

Support for HCPs and patients on shared decision making can be found:
<https://www.england.nhs.uk/personalisedcare/shared-decision-making/>
<https://www.nice.org.uk/guidance/ng197>

No decision about me without me: making shared decision making a reality by Angela Coulter, 2009:

<https://www.kingsfund.org.uk/publications/making-shared-decision-making-reality>



PRINCIPLES FOR BETTER COMMUNICATION BETWEEN CLINICIANS AND PATIENTS

Shared decision making is a two-way process, and both patients and HCPs need to engage with the process. The following steps can be taken by HCPs and patients to enhance shared decision making in practice.

Patients

- Patients should be supported to advocate for themselves and to share with clinicians the impact of their condition and their aspirations objectively. This would provide their clinician all the information to inform shared decision making conversations and make treatment decisions together.
- Keeping a log or diary of when attacks occur can help add detail and paint an accurate picture of what has happened since the last appointment, ensuring nothing is missed.
- Some patients may feel more comfortable speaking about what is on their mind by preparing what they want to talk about before their appointment or by bringing a loved one to support them.
- If patients do not feel comfortable or feel that they don't have the right kind of relationship with their consultant for any reason, they should be able to request to see another member of the team without this being seen as a criticism.

HCPs

- Clinicians should take thorough notes in every consultation so that other clinicians or junior staff can pick up a patient's file and know what is going on.
- Clinicians should consider asking more in depth, open-ended questions, such as 'what are you worried about?' 'how is it impacting you?' or specifics about their life in order to build a fuller picture of their condition and better relationships based on deeper understanding. Specific questioning around recognised treatment side effects (e.g, anxiety or facial hirsutism) can help uncover side effects patients may not have thought to mention.
- HCPs should encourage patients to record all evidence of disease activity to provide the full clinical picture, which may indicate the need for a change in treatment.
- HCPs should keep patients informed about new treatments available in languages that they will understand. Everyone is different, and some patients like detailed information at appointments, while others find it too much to take in at one time. The HCP should learn the patient's preference and tailor their approach accordingly.
- There is an opportunity to expand the role of nurses in the pathway. Going through the shared decision-making process with nurses separately from the physician consultation can enhance the patient experience. Nurses can relay important information to the consultant if they speak to the patient prior to the consultation or can talk with patients after their consultation once they have had time to digest information.



MY CONSULTANT DUG INTO HOW ATTACKS ARE AFFECTING ME MENTALLY AND ASKED IF IT WAS AFFECTING MY RELATIONSHIPS. THEY WENT THROUGH EVERYTHING, AND THEY LISTENED.

Female, 46-55, Scotland



The psychological strain of living with HAE

There is a significant emotional and psychological burden for people living with HAE. Those who have had family members die of HAE or who have experienced a throat attack – a terrifying and life-threatening event – often have high levels of anxiety. A study found HAE patients have a 39% chance of being diagnosed with depression. Patients can experience anxiety around attacks or aspects of their treatment, such as switching medications, self-injecting, or self-care. It is a vicious cycle, as poor treatment experiences and attacks lead to more stress, and in turn, increased anxiety can lead to more attacks.

All HAE patients should have access to mental health services to support their wellbeing. This is likely to have additional benefits, such as improving their resilience and willingness to explore new treatment options.

HAE HAD AN IMPACT ON MY MENTAL HEALTH AS MY DAD PASSED AWAY BECAUSE OF IT. IT IS HARD LIVING WITH IT.

Female, 26-35, London



The right to see a specialist

Everyone with HAE should be treated by a specialist with specific expertise in HAE, as recommended in the international WAO/EAACI guideline for the management of HAE.ⁱⁱ Access to a specialist HAE consultant and specialist nurse as part of the patient pathway has been found to have been beneficial to HAE patients and clinicians. Acknowledging that there is only a limited number of HAE specialists and treatment centres, this may not be possible for all, but patients should always be able to access at least one of these specialist roles.

It may be beneficial for immunology services to organise dedicated HAE clinics as these can optimise focus on HAE, utilise specialist skills and resources efficiently, and allow opportunities for patients to speak to their peers if this is desired.

This and other models of care require further evaluation to ensure patients' concerns and preferences can be addressed.

International guidelines recommend that HAE-specific, comprehensive, integrated care is available for all patients.ⁱⁱ However, regional variation across treatment centres for angioedema means HAE patients are receiving different models of care across the UK.

This can lead to inequalities in patients' satisfaction with their care and their ability to access certain treatments. Variability across treatment centres could be due to variation in resources, the volume of HAE patients, or variation in numbers of HAE specialist staff or HAE-specific clinics. Variations in staff responsibilities across different centres can further exacerbate inconsistencies in care. Some consultants reported more effective shared decision making with involvement of specialist nurses in the patient pathway. Rotation of clinic physicians means that patients may experience many different consultants; however, there are examples of specialist nurses providing much-needed continuity in the clinical relationship with patients. These findings suggest that a broadening of the specialist nurse role across all HAE centres should be evaluated, subject to achievement of the nurse competencies recently developed and published by the British Society of Immunology (BSI).

I AM ANXIOUS ALL THE TIME. I TRY TO BE CALM AND NOT TO STRESS. THIS IS WHY I WORK PART TIME NOW, BUT I KNOW HAE WON'T GO.

Female, 26-35, London



“Some patients are very well read on the treatments that are currently available and those that are in trial and likely to become viable in the next few years. However, the majority will sit with their current treatments as the thought of change is frightening. When there is perhaps a dedicated nurse that the patient feels more comfortable asking questions about treatments with patients definitely feel more able to agree to transition to a new treatment. However, there are an identified number of vacancies in immunology services for both nurses and consultants at present, meaning that not all specialist centres are able to support patients with both at the same appointment. This needs to be addressed potentially even with telephone appointments from nurses to back up and support face-to-face clinician visits.”

Angela Metcalfe, Chief Executive, HAE UK

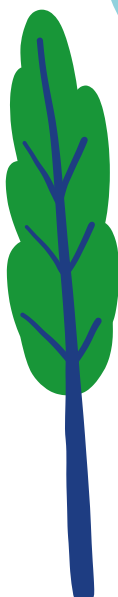
PHYSICIANS AT A&E NOT UNDERSTANDING MY CONDITION LEADS TO DELAY IN APPROPRIATE CARE OR INAPPROPRIATE TREATMENT. THIS CARE IS NOT EASY TO ACCESS, IT'S DIFFICULT TO GET TO ON PUBLIC TRANSPORT, AND IT'S NOT WORTH THE TROUBLE UNLESS LITERALLY DYING.

Female, aged 56, London



MAKE SURE YOU'RE IN THE RIGHT CLINIC WITH THE NEWEST TREATMENTS, AND IF YOU'RE NOT, THEN IT CAN TAKE A MUCH LONGER TIME TO GET ACCESS TO THINGS.

Male, aged 46, East of England



OUR RECOMMENDATIONS

We are calling on patients, HCPs, and the healthcare system to make a change. There is an opportunity for HAE patients and HCPs to make shared decision making the norm in HAE. Our research has shown both patients and clinicians recognise its value and the potential to improve the lives of people living with the disease.

NHS England guidance recognises that the implementation of shared decision making is a complex intervention. As our research with people with lived experience of HAE and with clinicians and stakeholders has found, HAE patient support and shared decision making should address the unpredictability of attacks and mitigate against the serious and negative impact of the disease on people's lives.

THE SYSTEM

Specialist HAE clinics

1. Care should be delivered by a specialist immunologist within a dedicated HAE clinic that includes a broader role for specialist nurses in line with the BSI-published HAE nurse competencies. We propose, where possible, to broaden the role of nurses in the clinic to be more comprehensively involved in the care of patients with HAE to offer a more holistic approach and strengthen SDM conversations. This model of care means services are designed to provide a dedicated clinic for HAE, which means resources can be used efficiently, and HCPs are able to focus and provide patients with the opportunity to get to know other patients.

Involve patients

2. Support the call for greater patient involvement and representation. HAE patients desire greater involvement and representation in commissioning committees. Healthcare leaders must support this call and champion the patient voice.

Make available training and resources on shared decision making

3. Optimise multidisciplinary care by offering training on shared decision making for consultants, immunology nurses, and specialist pharmacists. Generalists like GPs who have patients with HAE and A&E teams in areas where there are known family clusters of patients with HAE should have knowledge of HAE attacks what to look for, and when to involve an immunologist.



PATIENTS

What matters most to you

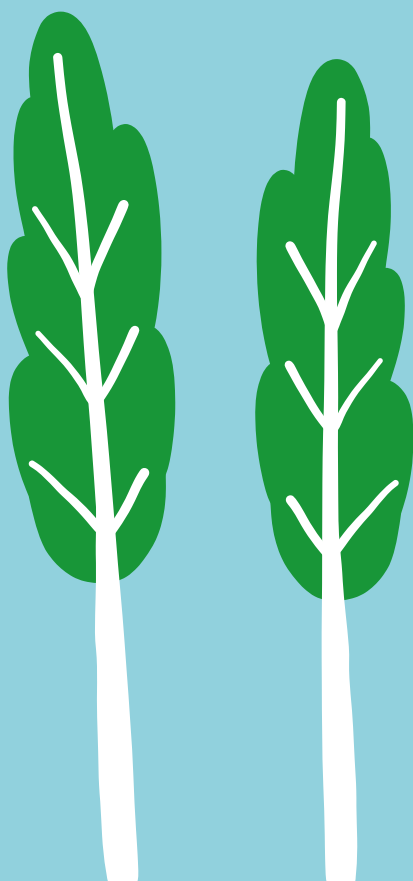
1. Patients should work with clinicians to develop a management plan together. The process should involve shared decision making with clinicians that includes professional guidance and, importantly, the patient's unique needs, taking into account their preferences, beliefs, and values.

Don't let HAE get in the way of living your life

2. Patients should take a proactive approach to their care and should be supported by the team around them. Patients should take it upon themselves to understand treatment options and should be able to ask for help if they have concerns about treatment side effects or issues such as their mental health.

A management plan that works for you

3. For the very best support for their condition, patients should be able to attend a HAE specialist clinic. Appreciating differences in communication styles, if a patient does not feel that they are having the right conversations, they should feel comfortable to ask to be managed by a different specialist, ideally a clinician specialising in HAE.



HCPs

Putting the patient first

1. HCPs should use shared decision making to develop trust and an effective and strong relationship with patients to come to agree a suitable care plan. This may mean the HCP accepting that the wishes and priorities of the patient differ from theirs. HCPs should also be flexible and embrace the use of different tools or methods that work for the patient, e.g, paper for some, apps for others, phone vs face-to-face interactions.

See your patient as a person

2. HCPs should take a holistic view of a patient, getting to know them and looking past the disease. Often, the disease symptoms are just one factor to consider, with the impact of treatment options on a patient's opportunities at different stages of their life being equally important to take into account.

Use tools to enable better shared decisions

3. Use shared decision making tools to help relay up to date evidence-based information in a way that patients can understand. Use literature and global assessment measures such as the Angioedema Control Test (AECT), to your advantage. Seek out training if required.



METHODOLOGY

A mixed methodology research approach was used to explore how SDM is used across the UK in the management of HAE, which formed the basis of this report. BioCryst commissioned ZPB Associates, a UK healthcare communications consultancy, to conduct research on their behalf.

The research objectives were:

- Understand HCP and patients' knowledge and current experience of Shared Decision Making (SDM)
- Identify areas of differing viewpoint between HCPs and patients on SDM
- Understand patients' motivations and the desire to be involved in decision making, e.g., where prophylaxis may give patients agency
- Identify current barriers and drivers for effective SDM
- Identify opportunities to address the barriers and drivers for both patients and HCPs and enhance SDM

Desk research

To understand the current HAE landscape, ZPB Associates conducted desk research into current guidelines, examples of shared decision making in practice, barriers and drivers, key stakeholders, and the management process.

HCP interviews

In June 2022, ZPB Associates carried out 5x30-minute interviews with HCPs working in HAE on the topic of shared decision making. The interviewees included Consultants and Registrars in Immunology and Allergy, Specialist Nurses, and Consultant Practitioners.

The interview objectives were to:

- Find out more about how SDM is currently carried out 'on the ground' in the NHS with HAE patients
- Define the benefits of SDM
- Explore barriers to effective SDM
- Gain an understanding of what tools and resources are currently being used and what further support -would be welcomed by HCPs

Patient survey

To understand the patients' perspectives, the current experience and feeling of SDM, and where there is divergence with HCPs, ZPB Associates conducted a survey with patients and carers in November 2022 through HAE UK's network. 131 respondents were surveyed (alongside HAE patients, this included one carer, two partners, and four parents). In April 2022, a follow-up survey of 29 respondents (17 female, 12 male) from the first survey was then conducted to quantify the quality of SDM that patients experienced. The aim was to dive deeper into understanding what goes on during appointments with HCPs, what patients perceive as 'good' or 'bad' care and communication, and find out what would optimise disease management.



Patient interviews

In May 2023, ZPB Associates conducted interviews with nine HAE patients (from 29 screened respondents) about their experiences of shared decision making. Three patients from each of the following treatment groups were interviewed: licensed long-term prophylaxis, on-demand treatment, and androgens or tranexamic acid. Across each group, a range of geographies, genders, ages, time since diagnosis, and attack rates were captured.

Co-creation workshops

Using the findings from the survey and the interviews, ZPB Associates ran two co-creation sessions with HCPs and patients to validate findings, identify pain points around shared decision making, and ideate solutions to these problems.

HCP workshop

The HCP workshop, held in October 2023, lasted for two hours and 15 minutes and was attended by seven HCPs: five Consultant Immunologists, one Immunology Specialist Nurse, and one Immunology Nurse Consultant. The session was conducted as a peer-to-peer learning event, an opportunity for clinicians to discuss the challenges they face when managing HAE patients, share examples of how shared decision making is undertaken, and how it could be improved going forward. This workshop was organised by BioCryst Ltd in partnership with HAE UK and ZPB Associates Ltd. Materials were planned and developed collaboratively and paid for by BioCryst Ltd.

Patient workshop

The BioCryst UK Ltd co-creation workshop, August 2023, held in partnership with HAE UK, was attended by six participants with HAE. The group included a mix of age, gender, background, and treatment to as far as possible represent the wide range of people living with HAE. The session was conducted as a peer-to-peer learning event, an opportunity for patients to meet and work with others with HAE, have their say on how HAE is managed, and share examples of how shared decision making is undertaken and how it could be improved.

Limitations

It is to be noted that most patients who were involved in this research were a part of the HAE UK community and, therefore, are likely to be more engaged with their care. It is acknowledged that there are a group of patients who aren't engaging with care, and whilst we discussed solutions that may support them, they weren't involved in this research. The survey sample of 131 patients can be taken as reasonably representative of the UK HAE community; however, the subsequent survey was in a smaller subgroup of 29 patients consenting to more in-depth questioning, which may not therefore, be representative of the main population of patients with HAE in UK.

¹ Quantitative survey conducted with HAE UK membership n=131 November 2022 and Quantitative survey conducted with HAE UK membership n=29 April 2023 / ² The international WAO/EAACI guideline for the management of hereditary angioedema- The 2021 revision and update - PubMed (nih.gov) / ³ A National Survey of Hereditary Angioedema and Acquired C1 Inhibitor Deficiency in the United Kingdom - The Journal of Allergy and Clinical Immunology: In Practice (jaci-inpractice.org) / ⁴ El-Shanawany T. Hereditary angioedema. Available at <https://www.immunology.org/public-information/bitesized-immunology/immune-dysfunction/hereditary-angioedema>. Access date: June 2023 / ⁵ BioCryst UK Ltd co-creation workshop conducted with HAE UK membership n=6 August 2023 / ⁶ www.nice.org.uk/about/what-we-do/our-programmes/nice-guidance/nice-guidelines/shared-decision-making / ⁷ www.ncbi.nlm.nih.gov/pmc/articles/PMC4211935/



