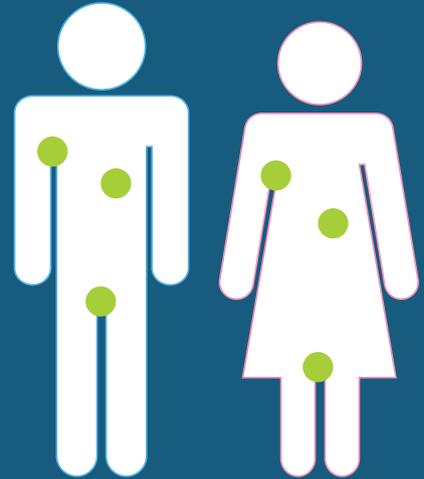


HS: DON'T DO IT YOURSELF

Understand more, and improve your life with HS

News from Hidradenitis Suppurativa research
for patients and their relatives

WHAT IS HS?



Hidradenitis Suppurativa (HS) is a chronic skin disease which occurs in repeated outbreaks of sore pimples or abscesses, located on the body where skin meets skin. The pimples and abscesses are, however, not ordinary pimples and abscesses - and the disease is not contagious.

HS is most often located in the armpits and in the groin, but may also occur under the breasts, in skin creases on the stomach, on the buttocks, around the genitals and around the anus. HS typically breaks out after puberty and three out of four patients are women. Approximately 1-2 % of the adult population suffers from HS in a varying degree.

HS: DON'T DO IT YOURSELF

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COVER LETTER

The European Patients' Associations with HS in Europe has tried to identify, together with clinicians, the most appropriate treatments to guarantee the best possible care.

Patients' expectations and the instruments available to clinicians have changed over time; a process has been triggered with the aim of creating a multidisciplinary alliance between all the stakeholders, coordinated by dermatologists and shared by patients.

Clinicians, patients and other health professionals are challenged every day to help relieve from- and, if possible, resolve difficult pathologies such as HS.

The long-standing experience of patients, reflected in their communication within the associations, has forced us to reconsider medical treatment with a different perspective on quality of care.

Patients have the task of requesting their involvement in all choices that concern their physical and mental wellbeing, that should ultimately result in a partnership between professionals and patients.

Each person affected by HS is invited to acquire all necessary knowledge to participate in an informed way in all decisions regarding his physical and mental wellbeing.

Patients' value, experience, knowledge and needs are an essential part of healthcare and, together with medical skills, allow them to be the protagonists in the history of their disease.

Health intervention can trigger difficult processes. Still, it is less difficult to manage these, than to live with the solitude that this disease brings with it.

In this perspective, the patient no longer has only rights, but also duties, as he/she must participate in the process. We are called to an historical transition: the bargaining between the clinical indication and the patient's preferences. In order to make sure that this transition takes place in the best way possible, patients' awareness and active participation in the governance of chronicity are essential.

Managing HS requires emancipated patients and doctors, free from the approaches of the past, when physical and psychological pain were not adequately taken into account.

Nowadays it is essential to take advantage of the most modern drainage and medical tools, as well as of diagnostic tools able to effectively control further symptoms associated with hidradenitis suppurativa. It is also important to implement new therapies, to take account of the urgent need to invest in research, to involve the institutional socio-health leaders and to disseminate correct information.

What is a good medicine for the HS patient?

Based on the experience with affected patients, and taking into account their preferences and values, it is important to develop and carry out health interventions that bring real benefits to patients, without neglecting safety. Health care must therefore reconcile different choices without giving up on scientific requirements.

Dermatologists and patients need to become leaders, capable of dragging other members and collaborators into the care team.

The ambitious challenge that EFPO poses on HS requires to end the self-management of the disease by both doctors and patients and focus on constant comparison between each other instead. The experience of many patients shows us that when this is achieved, it is possible to achieve the desired changes as well.

Giusi Pintori

President of Hidradenitis Europe
European Federation of HS
Patients' Organisations (EFPO)



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Chapter 1 | GENETICS

WHY DO I GET HS?

This is a question that patients are especially interested in finding out. Researchers are working hard for answers, and one thing they do know is that genetics is one of several factors that play an important role.

KNOWLEDGE EMPOWERS YOU

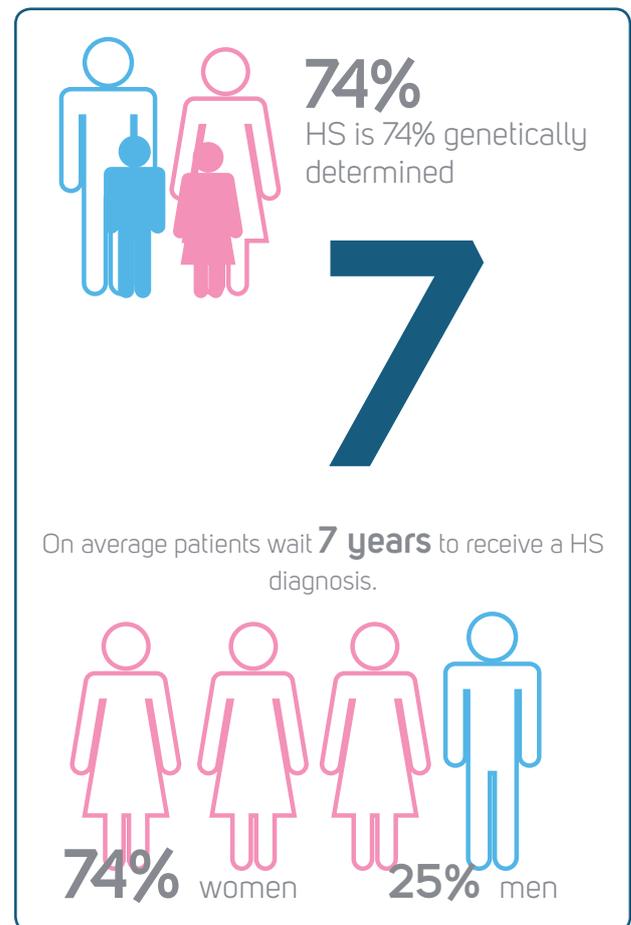
As a patient, you may get the feeling that it is your own fault that you have contracted HS. It is not unusual that patients experience that their own physician tells them to lose weight or to stop smoking. In such cases, knowledge about HS causes and the significance of genetics is important. Genetics play a part and it can be very liberating to have this knowledge. You have contracted a disease and you are not to blame. Knowing this can remove the feeling of guilt, which is paralyzing and that many HS patients unfortunately carry around with them.

Having knowledge about your disease empowers you

WHAT CAUSES HS?

Fortunately, there is lots of research being done on HS today and several different articles and scientific presentations addressed the issue of the cause of HS. In a Dutch twin-study is report that HS is 74% genetically determined; on the other hand, many studies only mentioned the connection with smoking and overweight. In other words, there seems to be a genetic component as well as other factors. Many smokers and people struggling with overweight do not have HS and vice versa, many HS patients do not have family members with the disease. Obesity and smoking habits play a role, but also slim people who never smoked have the disease. There are many patients who live with the disease without knowing that their condition has a name. They

haven't been diagnosed yet, because they haven't met a physician who could identify their condition. Many patients either do not push hard enough to get a diagnosis or have given up on getting help. On average patients wait 7 years to receive a HS diagnosis. Many patients have not been diagnosed and if they have, they might only be advised to lose weight or quit smoking



THINGS YOU CAN DO TO IMPROVE YOUR SITUATION!

BE AWARE OF THE SYMPTOMS

IF YOU OR OTHERS IN YOUR CLOSEST FAMILY HAVE HS, YOU NEED TO PAY SPECIAL ATTENTION TO THE DEVELOPMENT OF HS SYMPTOMS. WE KNOW THAT HS CAN BE HEREDITARY AND IF THE DISEASE RUNS IN THE FAMILY, IT IS IMPORTANT THAT YOU ARE AWARE OF HS SYMPTOMS AT AN EARLY STAGE, SO YOU OR YOUR FAMILY MEMBER IN DUE TIME CAN GET A REFERRAL TO A DERMATOLOGIST OR AN HS-EXPERT, WHO UNDERSTANDS THE COMPLEXITY OF THE DISEASE AND CAN HELP.

IF YOUR CHILD HAS HS

MAKE SURE TO TAKE IMMEDIATE ACTION IF YOU DISCOVER THAT YOUR CHILD HAS HS SYMPTOMS. GET A REFERRAL TO A DERMATOLOGIST OR AN HS-EXPERT TO ENSURE THAT YOU GET THE BEST POSSIBLE HELP.

IN ADDITION, YOU MAY HAVE A PATIENT ASSOCIATION IN YOUR COUNTRY THAT WILL BE ABLE TO HELP WITH RELEVANT INFORMATION.

THE INFLUENCE OF GENES ON HS

HS is a complex disease with a variation of elements that we know of, and some unknown elements. Therefore, research is both important and necessary. One of the key areas of interest for researchers is genetics.

“At the moment we know that HS is not an infection. We know that HS is more common in some families than in others. It occurs more frequently among women and in some segments of the population. HS is, for example, more common among African Americans than among Caucasian Americans. We also know that HS is connected to a number of related diseases, but we don't know which comes first - HS or the related diseases. This is not yet possible to determine with the data we have at hand.

The easy answer is that presently we do not know why some people get HS.

The more elaborate answer is that genetics play a part. We have found certain gene mutations that affect the immune system and the maturing and development of the cells.

We do not yet have the full understanding; however, we are certain that genes and biological inheritance are important for the development of the disease” says Professor dr. med Gregor Jemec, Zealand University Hospital, Denmark.

He explains that the hereditary component of the disease is considerable, but that it is only one of several possible causes that have yet to be determined.

“It would be nice if there was only one single cause to the disease. It is easy to understand that if e.g. an area is infected with bacteria, this causes an infection, and if you remove the bacteria, then the infection goes away. That would be comprehensible and logical, but unfortunately this is not the case with HS.”

“HS has several causes. You may inherit it through your genes, it is seen more frequently in people who smoke or are overweight, and you may lack a certain type of bacteria. These are all different ways to the same end.”

Gregor Jemec, Professor dr. med.

NEW RESEARCH IS ON THE WAY

According to Gregor Jemec, the research into HS is characterized by the fact that the disease, up until a few years ago, was an area of low interest. As a consequence, the scientific community is lacking basic scientific research and data that supports the existing knowledge. "Specific research on the link between the immune system and related diseases is being conducted. If it is possible to prove that the related diseases and HS have the same cause, then we have come far.

The related diseases are interesting for two reasons. They are relevant for the individual patient, which of course is the most important reason, and they are also of theoretical relevance. If we can determine that the cause of HS and the related diseases are identical, then we have located the mechanism. And if we succeed in finding a gene linked to the two diseases, then it will be possible to target the treatment towards what has gone wrong with the specific gene," says Gregor Jemec.

NEW RESEARCH FROM EHSF THE WORLD'S LARGEST CONFERENCE ON HS

HEADLINES OF SOME OF THE LATEST PUBLICATIONS RELATED TO HS AND GENETICS

A TWIN STUDY ON THE GENETIC BASIS OF HIDRADENITIS SUPPURATIVA

K.R. VAN STRAALEN ET AL.

A DUTCH STUDY SHOWS, IN POPULAR TERMS, THAT FOR 74 % OF ALL HS PATIENTS THE DISEASE IS GENETICALLY BASED.

GENETICS IS A VERY STRONG FACTOR, BUT THERE ARE OTHER FACTORS PLAYING A ROLE AS WELL.

HIDRADENITIS SUPPURATIVA PROFOUNDLY IMPACTS THE QUALITY OF LIFE OF FAMILY RELATIVES

P. GUILLEM ET AL.

A FRENCH STUDY SHOWS THAT HS PATIENTS' FAMILY MEMBERS ARE NEGATIVELY AFFECTED BY THE DISEASE, INCLUDING PARENTS, CHILDREN, SIBLINGS AND SPOUSES.

5 QUESTIONS FOR THE PROFESSOR

1. IS IT POSSIBLE TO PREVENT HS?

No, that would require that we have a solid understanding of the cause of the disease, and we do not have that at the moment. We know that genes play a decisive role, so it is not possible to avoid HS.

2. WHAT IS THE SIGNIFICANCE OF OVERWEIGHT AND SMOKING

Overweight is related to the severity of the disease. But whether it also plays a role in the presence of the disease, we do not know. We also know that smokers have more severe HS. A recommendation would be to lose weight and smoke less – this should improve the severity of the disease. However, this does not mean that slim people and non-smokers do not get HS. The answers are never absolutely clear.

3. WHEN DO YOU GET HS?

HS usually occurs when people are in their 20's and not so often when people are in their 50's. This is one of the reasons why HS is so intrusive in a person's life. HS occurs in the most active part of your life e.g. when you are getting established with a partner, have children, get a home and start a career. A lot of things are going on that are affected by the disease. This is one of the reasons why HS, apart from the physical symptoms, may also cause mental and social problems.

4. IS THERE A CURE FOR HS?

There is no cure for HS, but being an inflammatory disease it tends to improve with age, as the immune system gets less active. However, we do not have good data that explains why HS becomes rarer with age. If you asks patients who were diagnosed 20-25 years ago how their HS is today, approximately one third will tell you that the disease has disappeared, one third will say it has become better, while the last third will say it is the same or worse. And this is hard to count on and not least to measure. You know if your symptoms are gone or not. But if you still have symptoms, then there is a tendency towards getting used to the level of the disease, and it is then difficult for us to say whether it is better or worse than previously.

5. WHICH OTHER SPECIALITIES ARE RELEVANT?

There are several relevant specialties, because HS is a disease that affects patients to a massive extent. But it is most likely that you may benefit from seeing a rheumatologist, a plastic surgeon, a gastroenterologist, a specialist of metabolic disorders, a gynecologist and a psychologist or psychiatrist over the course of your disease. (Read more about related diseases on page xx)

Chapter 2 | BACTERIA

NEW RESEARCH: HS PATIENTS MAY HAVE LESS BACTERIA THAN PEOPLE WITHOUT HS

It has been some time since it was discovered that HS is not just an ordinary bacterial infection, which can be cured with water, soap and antibiotics. New research now surprisingly indicates that HS patients have fewer bacteria than people who do not have HS.

If you are knowledgeable about HS, you also know that the disease has nothing to do with poor hygiene. Still, many HS patients are confronted with this prejudice, and it is often the physician who believes that the disease is caused by bacteria.

You can request a test of the skin either at your physician, specialist and hospital. A test may determine whether you have an infection that can be treated with antibiotics.

NEW KNOWLEDGE ABOUT BACTERIA

Our understanding of bacteria has undergone major development within recent years. Within the scientific field known as “microbiome research”, scientists are, trying to understand the interaction between so-called “good” bacteria and other microorganisms, and how they may affect your health. Recently, several Danish studies have found some interesting results concerning bacteria and HS.

“Both to the naked eye, and when looking closely at tissue samples of boils, you can see that there is a formation of pus and this might indicate that bacteria play

some kind of role in the development of HS,” says MD, Ph.D. Hans Christian Ring, Zealand University Hospital, Denmark.

Hans Christian Ring finished his Ph.D., on the significance of bacteria for HS, in the spring of 2017. His work has led to several new findings.

“Back in the day it was presumed that HS was caused by bacteria. This theory has been dismissed. Today we know that HS is a chronic inflammatory skin disease, which is not directly linked to well-known bacteria that form boils. And now the latest research indicates that HS patients lack bacteria of the sort “propioni bacteria,” which may be significant for the development of the disease. We have just examined the micro biological conditions before and after the boil has formed,” says Hans Christian Ring.

STUDY 1

THE ROLE OF BACTERIA PRIOR TO THE OUTBURST OF ABSCESSSES

“One of our research studies indicates that HS patients may lack certain bacteria before the draining abscesses appear. We took samples from the skin and compared the skin from the armpits of people with HS and without HS. We discovered that quantitatively more bacteria were present among people without the HS diagnosis. Healthy people had more of the so-called biofilm aggregates, which are bacteria sticking together and typically covered with a thin layer of mucus,” says Hans Christian Ring. He further explains that the discovery may be significant for the disease.

“Microbiome research has shown us that we all have bacteria and that they are important in relation to having healthy skin. If you lack bacteria it will result in an imbalance of the skin microbiome, and this might happen even before the abscesses appear. If there is an imbalance it might potentially lead to two things. Firstly, normal and harmless bacteria may change into harmful bacteria. Secondly, the immune system may react to the imbalance giving rise to a local infectious reaction,” says Hans Christian Ring.

STUDY 2 A DIFFERENT BACTERIAL COMBINATIONS

Another research study from the same project shows that an imbalance may also occur as a result of the combination of different types bacteria present. “In the second study we looked at the combination of the bacteria. We did that by looking at the genetic material (DNA) of the bacteria, and it is the first time this kind of study has been carried out within the area of HS. We examined tissue samples from very new lesions in HS skin, little red bumps at the pre-abscess stage, and we found that the combination of bacteria in early lesions was different from the combination in healthy skin. One bacteria in particular – propioni acne – is missing in HS patients. The bacteria is found at the sebaceous glands and we know from earlier that HS patients are missing sebaceous glands,” says Hans Christian Ring.

STUDY 3 BACTERIA ONCE THE ABSCESS HAS FORMED

A third research study has uncovered something interesting, once the abscess has formed. “We have also examined the chronic abscesses, which have a lot of pus. Under the microscope we can see that the big lumps of bacteria are surrounded by biofilm which prevents both the immune system and antibiotics from getting to the bacteria. The biofilm is typically the cause of inflammation, reddening, swelling, heat and pain. And when there is an abscess, and therefore ruptured skin, more bacteria accumulate, which maintains the abscesses. It turns into a classical chronic wound which is difficult to treat because of the biofilm,” says Hans Christian Ring.

According to Hans Christian Ring, this new knowledge may mean that the treatment of chronic abscesses to a greater extent must be targeted at the removal of the biofilm. It is still too early to determine how this can be done, but one way might be a combination of early surgery, when the classical lesions with tracts (sinus tracts) are identified, with antibiotics to target the biofilm.

IMPORTANT KNOWLEDGE ABOUT ACUTE PAIN

HS PATIENTS WHO GO TO THE EMERGENCY WARD FOR URGENT HELP WITH ACUTE PAIN FROM ABSCESES OFTEN EXPERIENCE THAT THE PHYSICIAN ON DUTY DECIDES TO CUT THE ABSCESS OPEN IN ORDER TO RELIEVE THE PRESSURE. IF A BOIL IS BIG AND ON THE POINT OF BURSTING WITH A LOT OF PUS THE BEST SOLUTION MAY BE TO DRAIN IT IN ORDER TO ALLEVIATE THE PAIN. THIS MAY HELP RELIEVE THE PAIN MOMENTARILY, BUT THE BOIL WILL ALWAYS RETURN. THERE IS ALSO ANOTHER SOLUTION THAT DOES NOT ENTAIL CUTTING OPEN THE BOIL. IT IS POSSIBLE TO INJECT A STEROID DIRECTLY INTO THE BOIL. THIS PROCEDURE RELIEVES THE INFLAMMATION, THE SWELLING AND NOT LEAST THE PAIN.

HS IS A CHRONIC DISEASE AND, IN ADDITION TO ACUTE TREATMENT, YOU SHOULD BE REFERRED TO AN HS SPECIALIST WHO HAS THE REQUIRED KNOWLEDGE OF THE COMPLEXITY OF THE DISEASE AND CAN HELP YOU FIND A LONG TERM TREATMENT, WHICH SPECIFICALLY WORKS FOR YOU, BY KEEPING YOUR SYMPTOMS AT BAY. THE EMERGENCY TREATMENT CAN NEVER STAND ALONE.

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

FOLLOW UP ON YOUR TREATMENT AND MAKE SURE TO DEFINE CLEAR TREATMENT GOALS WITH YOUR PHYSICIAN.

IF YOU EXPERIENCE THAT YOUR HS TREATMENT IS NOT WORKING SATISFACTORILY, THEN CONTACT YOUR PHYSICIAN AND DEMAND A CLEAR UNDERSTANDING OF WHAT THE TARGET FOR YOUR TREATMENT IS. FOR EXAMPLE, IF YOU RECEIVE ANTIBIOTICS AND ARE IN DOUBT ABOUT HOW MUCH TIME YOU NEED TO CONTINUE TO OBTAIN THE DESIRED TREATMENT EFFECT, THEN ASK FOR THIS INFORMATION.

IT IS ALSO IMPORTANT THAT YOU SEEK HELP FROM A DERMATOLOGIST IF YOU EXPERIENCE THAT YOUR PHYSICIAN DOES NOT HAVE THE REQUIRED KNOWLEDGE ABOUT HS, OR IF YOUR CURRENT PHYSICIAN IS NOT PREPARED TO HAVE A CONSTRUCTIVE DIALOGUE WITH YOU ABOUT TREATMENT OPTIONS, TREATMENT GOALS AND OF COURSE YOUR TREATMENT. IF YOUR PHYSICIAN IS NOT WILLING TO REFER YOU TO A DERMATOLOGIST OR A HS SPECIALIST, YOU MAY NEED TO CHANGE PHYSICIAN.

REQUEST A TEST IF YOU SUSPECT AN INFECTION

YOU CAN ALWAYS ASK YOUR PHYSICIAN OR OTHER HOSPITAL HEALTHCARE PRACTITIONERS FOR A TEST IF YOUR SKIN REDDENS, IS SWOLLEN OR SORE, AND YOU SUSPECT THAT THERE MIGHT BE A DEEP INFECTION, SUCH AS A STREPTOCOCCAL INFECTION. IF YOU HAVE AN INFECTION, IT CAN BE TREATED WITH ANTIBIOTICS.

SOURCE: PATIENTS' ASSOCIATION HS DENMARK

Chapter 3 | CLASSIFICATIONS

THIS IS HOW PHYSICIANS FOLLOW YOUR HS DEVELOPMENT

HS specialists around the globe use different methods to assess HS in relation to treatment and research. Therefore, there is a need for an international standard and a group of researchers are working on developing exactly that. The aim is to apply the standard within the next few years.

In the summer of 2016, 24 HS patients volunteered for an experiment at Zealand University Hospital, Denmark. The patients were examined by 12 HS specialists from 12 different countries. The specialists used different scoring methods – a total of eight different – to assess and classify HS. The specialists had to use the same methods to describe the same 24 patients. They reached very different results and were in total disagreement about the number of abscesses, sinus tracts (which are small tube-like corridors in the skin), scar tissue formation and finally the total score of the individual patient.

“It is very difficult to count the individual lesions, especially if there are many and they merge. It is also impossible to see what is going on under the skin, and the result of this experiment showed that the existing methods are inadequate,” says Linnea Thorlacius, PhD. Student at the Dermatological Department at Zealand University Hospital, Denmark, and part of the research team.

HOW IMPORTANT IS CLASSIFICATION FOR PATIENTS?

HS severity cannot be rated without asking the patient. The classification is important in determining what kind of treatments can be offered to the patient.

“An abscess may hurt just as much in Hurley I as in Hurley III (see page xxx) and the classification does not say anything about the activity level of the disease. It is the inflammation that causes pain, and even though you only have one single boil and you may only be rated a Hurley I that one abscess might easily ruin everything

for you, if it is located at the edge of your knickers and is constantly flaring up.”

Bente Villumsen, chairperson of the Patient Association HS Denmark.

ALWAYS EVALUATE THE EFFECT OF YOUR THERAPY WITH YOUR PHYSICIAN

But how do the different classifications affect the treatment you are offered?

This is explained by Ditte Marie Saunte, clinical research lecturer, Institute for Clinical Medicine at Copenhagen University and Chief Physician at Zealand University Hospital, Denmark.

“HS is a disease that can progress, meaning that you as a HS patient may move up in the Hurley levels. It is impossible to predict whether the disease will progress and at what pace it will progress for the individual patient. It is recommended that you use the treatment, which has been prescribed to you, and that you evaluate the effect with your physician. Sometimes a treatment works well for a period, but if the medication ceases to have the desired effect or if the disease progresses, it must be considered whether your treatment is still the right one. This is where Hurley comes into the picture as a help to define the treatment options. A low score will consequently lead to a mild treatment. As an example, a patient at Hurley level I is often capable of getting by with local treatment on the skin (topical therapy) while a patient at Hurley level III mainly gets systemic therapy i.e. pills or injections, possibly in combination with surgery,” says Ditte Marie Saunte.

WHAT DOES CLASSIFICATION MEAN FOR YOUR TREATMENT?

"In order to choose the appropriate treatment, you need to be able to describe the status and development of the disease, but with regard to the individual patient the specific method that is applied is not important," says Ditte Marie Saunte.

"In connection with clinical studies, classification and scoring systems are important, but for the individual patient the only thing that matters is how they feel and whether the treatment is working." Ditte Marie Saunte,

Chief Physician and Clinical Lecturer.

"Does the treatment have the intended effect on the disease? Is there less activity? Do new abscesses with pus and soreness appear on new areas of the body? Or is there a stationary activity in the same area?

If it for instance is the same sinus tract under the armpit that swells and oozes pus, the patient will benefit from having it surgically removed. If the activity has moved to new randomly scattered areas, then we have to look into the possibilities of medical treatment, which can reduce the disease activity. Is the patient in pain, or is there no pain,

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

PLAY AN ACTIVE ROLE

AS A PATIENT YOU HAVE A JOINT RESPONSIBILITY, ALONG WITH YOUR PHYSICIAN, FOR YOUR TREATMENT, NOT LEAST REGARDING THE DIAGNOSIS AND CLASSIFICATION. IT IS IMPORTANT THAT YOU TAKE AN ACTIVE PART IN YOUR TREATMENT PROCESS AND TELL YOUR PHYSICIANS AND NURSES HOW YOU FEEL, ALSO MENTALLY. YOUR PHYSICIAN CANNOT LOOK AT YOU AND DETERMINE WHETHER YOU HAVE SLEPT BADLY, ARE IN PAIN, OR FELL DEPRESSED. THE PHYSICIANS AND NURSES REALLY WANT TO HELP – BUT TO DO THIS, THEY NEED YOU TO GIVE THEM THE FULL PICTURE OF YOUR SITUATION.

MAKE THE MOST OF YOUR TIME WITH YOUR PHYSICIAN

TIME IS OFTEN LIMITED AT A CONSULTATION. AS A PATIENT YOU TYPICALLY HAVE 10-20 MINUTES WITH YOUR PHYSICIAN, SO USE THE TIME WISELY. BE SURE TO ASK THE QUESTIONS THAT ARE IMPORTANT FOR YOU, AND MAKE SURE THAT THE PHYSICIAN IS AWARE OF YOUR CURRENT SITUATION AND HOW YOUR SYMPTOMS ARE INFLUENCING YOUR LIFE.

WRITE DOWN WHAT YOU NEED TO SAY BEFORE THE CONSULTATION SO THAT YOU DO NOT FORGET SOMETHING IMPORTANT AND TAKE NOTES DURING THE CONSULTATION. BRING A FRIEND OR A RELATIVE TO SUPPORT YOU. FINALLY, NEVER LEAVE THE PHYSICIAN'S OFFICE WITHOUT A NEW APPOINTMENT OR A CLEAR AGREEMENT ON HOW YOU SHOULD DEAL WITH YOUR DISEASE AND YOUR TREATMENT UNTIL YOUR NEXT CONSULTATION.

how is the disease developing and how is the treatment working? In brief, we examine the patient from one case to the next and here the scoring method is not so important, as long as we use the same methods every time and are skilled at using them,” says Ditte Marie Saunte.

RESEARCH NEEDS CLASSIFICATION AND COMMON STANDARDS

When it comes to research, Ditte Marie Saunte stresses that the classification has a different and more important function. “It is important to emphasize that there is a big difference between sitting with our patients, and talking about clinical studies. At present, we do not have a common international standard for monitoring the disease when assessing patients in a clinical trial setting. Therefore, it is difficult to compare results across different studies. At the moment, research into HS is developing rapidly, and it is very important that we are able to compare research results of treatment with different medicinal products or surgical methods. This way it will be easier to choose the treatments that are best for each patient. But that requires common standards,” says Ditte Marie Saunte.

According to Ditte Marie Saunte, HS specialists use a number of different methods to create a coherent understanding of how the patient is feeling and how the disease is developing. Here she gives an overview of the most commonly used methods for classification of HS.

CRITERIA FOR THE DIAGNOSIS OF HS

- THERE MUST BE TYPICAL LESIONS: PAINFUL, DEEP-LYING BOILS OR LUMPS (PIMPLE-LIKE) PERHAPS CONNECTED UNDER THE SKIN (SINUS TRACTS) AND POSSIBLE SCAR TISSUE.
- THE LESIONS TYPICALLY OCCUR WHERE SKIN MEETS SKIN: UNDER THE BREASTS, UNDER THE ARMPITS, THE GROIN, SKIN CREASES ON THE STOMACH, ANUS, AROUND THE GENITALS OR ON THE BUTTOCKS. THERE MAY BE ONE OR MORE LESIONS, AND A SINGLE OR SEVERAL SKIN AREAS INVOLVED.
- IT IS A CHRONIC AND RECURRING DISEASE, AND THE ABSCESSSES MUST OCCUR AT LEAST TWICE WITHIN A 6-MONTH PERIOD BEFORE A HS DIAGNOSIS IS POSSIBLE.

It may support the diagnosis process if other family members have HS. When you have been diagnosed, it is possible to determine the Hurley-stage.

HURLEY STAGES

According to Ditte Marie Saunte, Hurley Staging gives a snapshot of HS. It does not say anything about HS activity of the disease and development over time, but it may be good for an overall grading.

- HURLEY STAGE I IS A MILD DEGREE OF HS, CHARACTERISED ONE OR SEVERAL PIMPLE-LIKE BOILS. THERE ARE NO SINUS TRACTS, NO SCAR TISSUE AND THE BOILS MIGHT SOMETIMES BE PERCEIVED AS ORDINARY BOILS. MOST HS PATIENTS HAVE HURLEY STAGE I
- HURLEY STAGE II IS A MODERATE DEGREE OF HS, CHARACTERISED BY ONE OR MORE RECURRING BOILS AND BOIL-LIKE AREAS AND THERE MAY BE SINUS TRACTS AND SCAR TISSUE.
- HURLEY STAGE III IS A SEVERE DEGREE OF HS. THE PATIENT HAS AN ENTIRE BODY AREA AFFECTED WITH CONNECTED SINUS TRACTS AND ABSCESSSES. ONLY FEW PATIENTS ARE CATEGORIZED AS HURLEY STAGE III.

SARTORIUS SCORE

This method is a scoring system, which describes HS severity by assigning it a number. The physician counts the number of boils and sinus tracts and indicates the size of the areas involved on the body. You give points, add them up and reach a result which constitutes the score. Next time you count, you can see how the disease has progressed.

DERMATOLOGY LIFE QUALITY INDEX (DLQI)

DLQI is a questionnaire that has been used internationally since 1994 to measure quality of life in different skin diseases. It is one of the most commonly used methods for measuring quality of life within dermatology. The method shows how severe the patient rates their condition and how much it affects everyday life. It consists of 10 questions, for example, how itchy, painful and sore was your skin during the last week? There are five options: Very much, very, some, not at all, or not relevant.

ULTRASOUND

When looking at the skin with the naked eye, you can see if it is red and if there is pus, but you cannot see how deep the HS lesions are under the skin or whether there is fluid or tracts under the skin. According to Ditte Marie

Saunte, ultrasound is a good tool in this situation, e.g. to define the area that you need to operate. Ultrasound is very precise and gives a picture of how HS looks under the skin. Sometimes, you see that the lesions lie deeper and cover a larger area than what the physician was able to see with the naked eye.

Furthermore, the method also works in connection with clinical studies where you want to measure how any given treatment works. Ultrasound is one of the new initiatives within HS. It provides you with the big picture and thus a better possibility for classifying HS correctly.

MAGNETIC RESONANCE (MR) SCANNING

With an MR scan it is possible to look further into the body than with ultrasound. MR scanning is especially effective when examining how widespread the HS sinus tract system is. It is particularly suited for patients who have

HS around the anus, since the scan clarifies whether there is a connection between the sinus tract system and the anus. It is important to have this clarified prior to surgery, since such a connection might indicate a bowel disease - and in such a case it might be necessary to carry out an examination of the gut prior to a given surgery.

Did you know:
People living
with HS
have a worse quality
of life than
cancer patients

NEW RESEARCH FROM EHSF THE WORLD'S LARGEST CONFERENCE ON HS

HEADLINES OF SOME OF THE LATEST PUBLICATIONS RELATED TO HS AND CLASSIFICATIONS

ULTRASOUND USE MODIFIES THE STAGING OF PATIENTS WITH HIDRADENITIS SUPPURATIVA

A. MARTORELL ET AL.

A SPANISH/CHILEAN RESEARCH TEAM HAS STUDIED HOW ULTRASOUND MAY SUPPORT HS CLINICAL EXAMINATION. FAR FROM EVERYTHING IS VISIBLE ON THE SURFACE OF THE SKIN AND CONSEQUENTLY ULTRASOUND IS A GOOD SUPPLEMENTARY EXAMINATION METHOD THAT ALLOWS YOU TO SEE THE EXACT SPREAD OF THE DISEASE SUBCUTANEOUSLY. THE RESEARCH SHOWED THAT THE SPREAD OF THE DISEASE IS OFTEN UNDERESTIMATED, WHEN ULTRASOUND IS NOT USED.

HIGH-FREQUENCY ULTRASOUND IN HIDROSADENITIS SUPPURATIVA

V. DINI ET AL.

AN ITALIAN STUDY HIGHLIGHTED THE POSSIBILITIES OF USING ULTRASOUND WHEN ASSESSING HS DEVELOPMENT AND SPREAD.

INTERNATIONAL INITIATIVE NEW STANDARDS FOR HS CLASSIFICATION

HISTORIC is the name of a large international initiative, which is aimed at defining an international standard for HS assessment. The first methods may be ready to use within a couple of years. Patients, physicians and nurses from 19 different countries on four continents have been asked: What are the relevant 'factors' to measure when you need to assess HS development? Almost 100 specialists have participated in an online poll regarding the factors they perceive as the most important to measure. Some of the specialists have also met face-to-face several times to try to agree. "It is a major problem within the scientific field of HS that the things which are measured, to see if treatments works, are very diverse. There is no agreement, and it makes it impossible to compare treatments. What is the best way to measure? This is the question we are attempting to answer with HISTORIC, where we as something quite new, have also been asking the patients," says Linnea Thorlacius who is part of the international research team.

PATIENTS AS RESEARCHERS

Bente Villumsen, chairperson of the Patient Association on HS Denmark, is one of the patients participating in the project and she is a member of the project's steering committee. She is pleased that the researchers listen to the patients when working on finding and selecting methods for measuring HS. "In this research project we are several patients who participate as researchers. We are not medical experts,

but we have experience in living with HS and can communicate what is important to us and how HS affects our lives on a practical level, e.g. which clothes to wear, challenges when going to the public swimming pool, and why patients sometimes stay away from social settings. When physicians find it difficult to measure how active the disease is, it is important that they ask us – the patients – how we feel," she says.

A LONG WISH LIST

At this point of the process, the task is to identify the many inputs and make a shorter and more manageable list. "We started with a long list of 56 different measurement criteria. They were identified by looking at literature and at what had been used until now. We also carried out many patient interviews, 20 interviews in Denmark and 20 in USA, and asked both physicians and nurses in a survey. That provided us with data sets that were made into a long list, which the participants have since been voting on twice. The purpose is to group the many questions into broad categories covering pain, quality of life, physical signs and symptoms," says Linnea Thorlacius. At present, the list has been comprised to the following themes: Pain, clinical signs, disease development, patient and physician angle, HS-specific quality of life and symptoms.

Chapter 4 | IMMUNOLOGY

THE IMMUNE SYSTEM PLAYS AN IMPORTANT PART IN GAINING FURTHER HS UNDERSTANDING

With HS, the immune system is overactive and, in rough terms, attacks the body by developing an inflammation – even if there are not many bacteria present. In the following, you can read about the link between the immune system and HS.

When people without HS develop a boil, it is usually caused by a bacterial attack from outside the body. The boil develops when the immune system attacks the bacteria. But with HS, the boil develops from within, because the immune system attacks the tissue around hair follicles. The presence of bacteria is not required. Knowledge about immune system is fundamental because that can improve the quality of life of HS patients: it is extremely complex to understand and use terms such as cytokine, lymphocytes and interleukins, but it is important that patients take care about themselves and these are three tips you could consider trying out:

THREE TIPS

REST WHEN YOU ARE TIRED;

IF THERE IS A THING YOU KNOW YOU SHOULD NOT DO, THEN SAY "NO" WHEN ASKED;

AND FINALLY, ASK FOR HELP WHEN YOU NEED IT.

A classic example is HS typically flaring up when you are going on vacation. You are stressed because of the packing for yourself and maybe for your family, and often we travel to places where it is hot and moist, which can provoke an outbreak.

THE IMMUNE SYSTEM IS ONE OF OUR MOST POWERFUL BIOLOGICAL MECHANISMS

Professor dr. med Gregor Jemec, Zealand University Hospital, Denmark describes the immune system as one of the most powerful biological mechanisms in the body. "It can both save our life and destroy it. It is constructed to detect things which are out of the ordinary, such as lumps, infections, new tissue growth or sick cells. Things the immune system must rid us of. This requires two things: Firstly, the immune system must be able to distinguish between what is sick and what is not. Rheumatism is a very good example of this as the immune system in this case detects its own joints and attacks them.

"When the immune system recognizes healthy tissue and attacks it you have an autoimmune disease." Gregor Jemec, professor, dr.med

"The other thing that needs to be present in a well-functioning immune system is a switch that can turn the system on and off. The immune system is as mentioned a very powerful mechanism which in principle, is able to kill people if it is too active. If it is turned on too quickly and turned off too late it will lead to disease. In this case you cannot turn off the inflammation that is being activated in turn it will be active for too long and with too much force. This is what is called an auto-inflammatory disease," says Gregor Jemec.

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

TAKE CARE OF YOURSELF

WHEN YOUR IMMUNE SYSTEM ATTACKS SOMETHING WHICH IT ISN'T SUPPOSED TO ATTACK, YOUR HS MIGHT FLARE UP AND YOU MIGHT EXPERIENCE A PHYSICAL REACTION FROM YOUR BODY. MANY PEOPLE WITH HS STILL DON'T KNOW THAT THERE IS A CONNECTION BETWEEN THEIR IMMUNE SYSTEM AND THEIR DISEASE, SO THIS IS IMPORTANT TO ACKNOWLEDGE. SOME HS-PATIENTS REACT BY GETTING A FLU-LIKE OUTBREAK WITH FATIGUE AND SORE MUSCLES. SO WHAT CAN YOU DO? SOME HAVE THEIR D-VITAMIN LEVEL MEASURED AND TAKE A SUPPLEMENT TO OBTAIN A BETTER IMMUNE SYSTEM. OTHERS TRY TO EAT AN ANTI-INFLAMMATORY DIET E.G. AVOIDING SUGAR AND WHITE BREAD, AND IN RETURN TAKE SUPPLEMENTS OF GINGER AND FISH OIL. IT IS IMPORTANT TO STRESS THAT SCIENTIFIC RESEARCH DOES NOT COME UP WITH SPECIFIC RECOMMENDATIONS, BUT IN GENERAL IT HELPS IF YOU TAKE GOOD CARE OF YOURSELF. GET ENOUGH SLEEP, AVOID STRESS, EAT A HEALTHY AND VARIED DIET AND EXERCISE – EVEN A SHORT WALK COUNTS. IF YOU WAKE UP IN THE MORNING AND FEEL POORLY, IT ACTUALLY HELPS TO GET OUT OF BED AND GO OUTSIDE. WHEN YOU ARE ACTIVE, THE SORENESS AND INDISPOSITION IS THROWN INTO THE BACKGROUND AND IT BECOMES EASIER TO FOCUS ON SOMETHING ELSE.

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION!

PAY ATTENTION TO THE SYMPTOMS (P 16)

HS IS A DISEASE THAT COMES AND GOES AND IN THIS SAME WAY YOUR SYMPTOMS CHANGE. A VARIETY OF THINGS AFFECT YOUR IMMUNE SYSTEM IN DIFFERENT WAYS. THEREFORE YOU NEED TO PAY SPECIAL ATTENTION TO HOW YOUR BODY REACTS. WHEN YOU HAVE HS, YOU HAVE AN INCREASED RISK OF A NUMBER OF RELATED DISEASES WHICH YOU CAN READ MORE ABOUT IN CHAPTER 5.

WHAT HAPPENS TO THE CELLS?

Professor dr. med Gregor Jemec explains that research shows that in HS, the cells do not communicate properly. They produce too many transmitter substances, which promote inflammation and produce too few that inhibit it. This is an important insight in the field of research in order to understand the disease and develop appropriate treatment options. Researchers are now working on finding the specific transmitter substances that communicate with each other. If there is, for e.g., too much of transmitter substance 'a' in connection with HS and it is possible to find a product inhibiting 'a', one could presume that this inhibitor could have an effect on the disease.

The biological treatments of today inhibit overactive transmitter substances, and more results in this field are expected,' says Gregor Jemec. He stresses that research down this path has resulted in the development of biological treatments. However, we still have not found the answer to the cause of the disease.

NEXT STEP IN THE RESEARCH

It is natural to ask where we lack knowledge about the significance of the immune systems in HS? The short answer is, everywhere! "We need research on the

relationship between bacteria on surface of the skin and the immune system. We have indications from some early studies, but it is extremely complex. You need to find viable bacteria, you also need to cultivate and monitor them without them dying in the process. It might prove that some bacteria are good to have and that they cause the disease to break out due to their absence rather than by their presence"" says Gregor Jemec.

Gregor Jemec also mentions related diseases as a field of immunological research.

"When the immune system is overly activated it becomes inaccurate, and this might explain a number of the related diseases. Let's say the immune system has to hit the area which is black, but it also hits some which is dark, grey or speckled grey. That may lead to a number of the related diseases e.g. rheumatism and inflammatory bowel disease." Gregor Jemec, professor, dr.med

"Finally, there is a strong indication that the problems with the immune system are affected by the genes. A few studies indicate this, but they haven't been confirmed by new studies. The area is very complex and is reflected in the fact that no one has been interested in this disease for years. Therefore, there is a need for researchers to find answers, and we are well under way," says Gregor Jemec.

NEW RESEARCH FROM EHSF THE WORLD'S LARGEST CONFERENCE ON HS

HEADLINES FROM SOME OF THE LATEST PUBLICATIONS RELATED TO IMMUNOLOGY

EFFECT OF CIGARETTE SMOKE EXTRACT AND ELECTRONIC CIGARETTE LIQUID ON CYTOKINE PRODUCTION BY IMMUNE CELLS FROM HIDRADENITIS SUPPURATIVA PATIENTS

A. MALARA ET AL

AN IRISH STUDY SHOWS THAT SUBSTANCES IN CIGARETTE SMOKE AND E-CIGARETTE FUMES AFFECT THE IMMUNE SYSTEM, BUT IT IS NOT QUITE CLEAR HOW. IN SOME WAYS IT ACTIVATES THE IMMUNE SYSTEM, IN OTHER WAYS IT DAMPENS IT. THE CONNECTION BETWEEN HS AND SMOKING IS THEREFORE STILL A RIDDLE.

Chapter 5 | COMORBIDITY

WHAT OTHER DISEASES SHOULD YOU BE AWARE OF?

SOME GOOD ADVICE:

Know the relevant symptoms, talk to your physician if you get the symptoms – and do not go around worrying unnecessarily.

You easily lose courage when you look at the list of disease related to HS: cardiovascular diseases, diabetes, inflammatory bowel disease, depression, joint pains, rheumatism and sleep disturbances. It is a concern which many people with HS live with.

For HS patients it is important to know that there might be a connection between HS and specific related diseases. It can be scary to examine the list of the related diseases – as if it wasn't more than enough just to have HS. However, it is important to remember that it is far from all HS patients who contract the related diseases.

DISEASES MOST COMMONLY RELATED TO HS

When you have HS, research shows that there are different diseases that you have an increased risk of contracting. On the following pages you can read about the most common diseases related to HS. We have divided the diseases into three groups.

- [DISEASES THAT ARE CONNECTED TO OVERWEIGHT \(THE METABOLIC SYNDROME\)](#)
- [DISEASES THAT LIKE HS ARE CAUSED BY AN IMBALANCE IN THE IMMUNE SYSTEM \(RHEUMATISM AND INFLAMMATORY BOWEL DISEASE\)](#)
- [PSYCHOLOGICAL REACTIONS AND DISEASES](#)

We start with the diseases related to the metabolic syndrome.

THE METABOLIC SYNDROME

The metabolic syndrome is a collection of different factors that increase the risk of developing heart or vascular

conditions. The syndrome upsets the metabolism of fat and sugar and with this follows overweight, diabetes, elevated cholesterol, arteriosclerosis, and similar conditions. The syndrome is often connected to lifestyle factors such as diet, smoking, alcohol and exercise, which may be hereditary and is more often seen with people who have HS. The following is related to the metabolic syndrome:

OVERWEIGHT

“Many HS patients struggle with overweight, and the severity of HS is statistically connected to the Body Mass Index (BMI) number, the more overweight, the more severe the disease.” Ditte Marie Saunte, Senior Consultant and Clinical Lecturer.

Currently, there are no studies showing exactly what effect weight loss has on HS activity. A Danish study (in connection with gastric bypass surgery, where part of the stomach was removed) shows that if weight was reduced by approximately 15%, the HS improved for a good number of the patients, while a few experienced a worsening.

Another Danish study shows that HS settles down more easily in people of normal weight. “We know that overweight influences the level of disease activity. We also know that fat cells may contribute to the inflammation process in the immune system, where the skin reacts spontaneously against itself and creates an inflammation. Furthermore, the mechanical friction when skin rubs against skin may trigger or maintain the disease, which is why losing weight also might help. However, we do not know the exact reduction of the disease activity in connection with weight loss.

An important question is also what comes first, HS or overweight? We do not have this answer either,” says Ditte Marie Saunte, Associate Professor, MD, Ph.D. at Zealand University Hospital, Denmark.

LOSING WEIGHT REQUIRES HELP, SUPPORT AND HS TREATMENT

Many HS patients are advised to lose weight, and this suggestion must never stand alone.

Overweight is important when dealing with HS, however, patients too often experience that the message from the physician is to lose weight. But losing many kilos is not that easy. Patients need help, support and motivation to take one step at a time. And it is important to stress that HS patients need to be treated for their HS at the same time.

DIABETES (TYPE 2)

Diabetes is also a part of the metabolic syndrome and is also more common among people with HS. It is unknown whether diabetes develops due to HS or if it is the overweight which triggers diabetes. Symptoms related to diabetes are feeling more thirsty than normal or if you urinate more than usual. If you have any of these symptoms, we advise you to go and see your general practitioner (GP).

IMPACT ON LIPIDS (FAT) IN THE BLOOD

Dyslipidaemia is also a part of the metabolic syndrome. Here, the balance between the different types of cholesterol in the blood is affected. There is an increase in triglycerides – the “bad” type of cholesterol, and

a decrease in the high density lipoprotein (HDL), the “good” type. This shift increases the risk of cardiovascular diseases.

HIGH BLOOD PRESSURE

There is no direct link between HS and high blood pressure, but as blood pressure is a part of the metabolic syndrome, which is seen more frequently amongst HS patients.

IMMUNOLOGICAL DISEASES CONNECTED TO HS

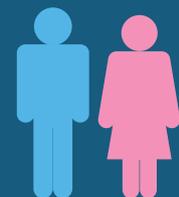
HS is an immunological disease where the immune system is overly active. When you have HS, you have an increased risk of having other immunological diseases as well. Not all physicians are aware of this connection, and we therefore recommend that you are aware of the symptoms so you can get adequate help and treatment.

JOINT PAIN AND SWOLLEN JOINTS MAY BE A SIGN OF RHEUMATISM

HS patients have an increased occurrence of joint pain and rheumatoid symptoms compared to people who do not have HS. Many experience that it worsens when the HS is more active. Just like HS, rheumatism is an immunological disease.

“If you experience joint pain, it is important that you react.

IF YOU.....



ARE OVERWEIGHT AND HAVE HS IT IS A GOOD IDEA TO SEE YOUR PHYSICIAN REGULARLY TO HAVE YOUR BLOOD PRESSURE MEASURED TOGETHER WITH YOUR BLOOD SUGAR LEVEL, CHOLESTEROL LEVEL, KIDNEY FUNCTION ETC. A TALK ABOUT HEALTH PROMOTING HABITS SUCH AS HELP TO LOSE WEIGHT, DIETARY ADVICE AND EXERCISE, MIGHT ALSO HELP.

Talk to your physician or a dermatologist so that you can be referred to a rheumatologist.” Bente Villumsen, chairperson of the Patient Association HS Denmark
It is very important that you do not wait, but make sure to get treatment in due time. Inadequate treatment can result in irreparable joint damage which may be disabling. Keep an eye on joint pain, lower back back, morning stiffness and swollen joints.

INFLAMMATORY BOWEL DISEASES

Other examples of inflammatory diseases are the intestinal diseases colitis ulcerosa and Crohn’s disease that are both more frequent among people with HS. Pay special attention to stomach aches, chronic diarrhoea, blood in your stool, weight loss and fatigue. If you experience these symptoms we recommend that you go and see your general practitioner (GP) and ask to be referred to a gastroenterologist.

PSYCHOLOGICAL REACTIONS AND DISEASES

QUALITY OF LIFE

HS is a disease affecting many aspects of the patients’ lives and challenges the quality of life for many. When HS is active it naturally has a physical strain, but as HS is a highly tabooed disease which is difficult to talk about, many patients experience that they are alone with their

worries, thoughts and unanswered questions.

This obviously affects your mood, your energy, and your sleep.

Studies have shown that HS patients have a poorer quality of life than skin cancer patients, measured on The Dermatology Life Quality Index (DLQI)

On the next page you will find a list of the most common psychological reactions and diseases related to HS.

THE MOST COMMON PSYCHOLOGICAL REACTIONS AND HS-RELATED DISEASES

DEPRESSION

Many HS patients feel depressed and lack energy, and among HS patients there is a lower self-reported quality of life than among the general population. Pain and low self-esteem may affect personal relations, sexual activity and work life. It can be difficult to have a sexual relationship with somebody and HS patients have an increased number of sick days from work. A Danish study shows, for example, that there is a larger unemployment rate among people with HS than the general population. Furthermore, surveys show that HS patients have a higher percentage of suicidal thoughts than the general population.

40% experience that they are anxious or depressed

have had suicidal thoughts **16%**

20% have considered not having children because of the disease

SLEEP DISTURBANCE

It is clear that you sleep poorly when you are in pain. Some patients experience a lot of itching, which, when combined with pain, can affect your sleep.

A good night's sleep is of great significance for general health and the quality of life. New research shows that disturbed sleep is common among HS patients.

BEING TIRED

Many HS patients complain about being tired. According

to Ditte Marie Saunte, it is difficult to measure fatigue levels, because what is fatigue? Being tired is an individual experience and you can get used to being tired and interpret it in many ways. It is also an area that lacks research. Until now, research has shown that HS patients show signs of fatigue – a kind of tiredness where you feel weak and exhausted, lack energy and constantly are tired.

Tiredness that patients feel is very noticeable and affects daily life even after a good night's sleep.



SMOKING

SMOKING IS NOT RELATED TO THE METABOLIC SYNDROME, AND IS NOT A COMORBIDITY, BUT THE INCIDENCE OF SMOKERS IS FAR HIGHER AMONG HS PATIENTS THAN IN THE GENERAL POPULATION. ACCORDING TO DITTE MARIE SAUNTE, THE ASSUMPTION IS THAT SMOKING WORSENS HS, AND SOME STUDIES SHOW THAT THE MORE YOU SMOKE, THE WORSE YOUR HS IS. HOWEVER, NO STUDIES HAVE YET SHOWN AN EFFECT OF QUITTING SMOKING.

“Studies show that the more packets of cigarettes, the worse the condition. We have studies showing one thing and a few others showing something else. So smoking is another area without total agreement.”

“We do know that smoking may also affect the microflora in the skin hereby activating the cytokines, which can increase the level of inflammation. In other words there is a connection.”

Ditte Marie Saunte, Associate Professor, MD, Ph.D.

NEW RESEARCH FROM EHSF THE WORLD'S LARGEST CONFERENCE ON HS

HEADLINES OF SOME OF THE LATEST PUBLICATIONS, COMORBIDITYV

QUALITY OF LIFE AND PSYCHOSOCIAL IMPLICATIONS IN PATIENTS WITH HIDRADENITIS SUPPURATIVA

C. DESSINIOTI ET AL

A GREEK STUDY SHOWS DETERIORATION OF THE QUALITY OF LIFE FOR HS PATIENTS. THE RESEARCHERS RECOMMEND PSYCHOSOCIAL SUPPORT TO WITHSTAND SOCIAL ISOLATION.

CLINICAL CHARACTERISTICS OF PRURITUS AND PAIN IN HIDRADENITIS SUPPURATIVA PATIENTS

L. MATUSIAK ET AL

A POLISH STUDY TESTS THE ITCHING AND PAIN THAT IS WELL-KNOWN SYMPTOMS FOR HS PATIENTS, BUT WHICH MANY HS PATIENTS THINK THEY ARE THE ONLY ONES TO SUFFER FROM. THIS STUDY DOCUMENTS THAT ITCHING IS A MAJOR PROBLEM. IN THE STUDY, IT APPEARS THAT ALMOST ALL PATIENTS FEEL PAIN AND THAT MORE THAN 40 PER CENT OF HS PATIENT EXPERIENCE ITCHING. THE ITCHING SEVERITY IS ALMOST AT THE SAME LEVEL AS THE PAIN.

ITCHING IS A WELL-KNOWN SYMPTOM FOR HIDRADENITIS SUPPURATIVA: A CROSS SECTORAL STUDY

A.R.VOSSEN ET AL

A DUTCH STUDY ALSO DOCUMENTS THAT ITCHING IS A COMMON PROBLEM FOR HS PATIENTS. IN THE STUDY, IT IS CONCLUDED THAT APPROXIMATELY 60 PER CENT OF THE PATIENTS WERE BOTHERED BY ITCHING. THE RESEARCHERS CONCLUDE THAT ITCHING IS OFTEN OVERLOOKED IN HS.

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

YOU ARE CONTROL OF YOUR LIFE

HS may affect many aspects of your life. It may affect your sleep, how you dress, your ability to work and your general mood. Some swallow the pain and pretend everything is okay. This need not be the right solution. There is much you can do to improve your situation and determine how you wish to live your life.

SPEAK TO YOUR PHYSICIAN ABOUT YOUR SYMPTOMS

Your optimal care is not the sole responsibility of your physician. As a HS patient, you have a different and larger responsibility for your body than those without HS. The physician, for example, cannot see how your mood is, or if you have diarrhoea, urinate more than usual and so on. If you experience these symptoms it is your responsibility to tell your physician.

SLEEP

Active HS may affect you sleep. Many experiences disrupted sleep and only sleep a few hours at a time. A bad night's sleep influences the morning mood which can start with pain and a buzzing feeling in the body, a little like having the flu. All in all, it can be difficult to get the day started. The bad night's sleep is not only caused by the pain - poor sleep can also be a result of the unrest, which the body experiences when it is fighting the inflammation.

Some people with HS experience that it helps to sleep in a cool room. Sweat, heat and moisture may trigger active HS lesion. The solution might be to turn off or turn down the heat or purchase a special pillow that feels cool to lie on. You can also buy duvets or mattress toppers that regulate the temperature so that it does not get too hot.

REST

Take a nap or sit down with your feet up. Fatigue accompanies the abscesses and the inflammation so it is quite natural that you need to take extra breaks. Take the breaks with a clean conscience. Also, meditation may help you to find peace and to relax. Many patients experience that a 'body scan exercise' may help to obtain peace of mind, to focus and to ignore the pain. On YouTube you can find many free body scan exercises.

PAIN

It is hard to live with pain, and it affects your quality of life, your sleep and all other aspects of life. Apart from pain killers, a locally active cream containing lidocaine may soothe when it hurts, which you can buy at a pharmacy. Apply a thin layer on the area where it hurts. Remember to talk to your physician or nurse about the pain.

EXERCISE

Some find it hard to exercise because sweating triggers active lesions which can hurt and it make it difficult keep bandages in place. Even though abscesses, bandages and pain may get in the way, there is no doubt that many experience that exercise helps. Exercise does not only give more energy, but also can improve mood. Many patients experience that exercise has a positive effect on the disease itself. If breakouts get in the way of a jog or a trip to the indoor swimming pool, consider taking a walk or riding your bicycle or any form of exercise that you can tolerate. That also counts as exercise. Some experience that yoga is a good alternative. You do not perspire the same way and you do something good for your body when you can control what exercises you feel comfortable doing.

Chapter 6 | TREATMENT

THE FOLLOWING CHAPTER OUTLINES THE TREATMENT OPTIONS FOR HS PATIENTS BASED ON THE NEWLY PUBLISHED EUROPEAN GUIDELINE FROM THE LEADING CLINICAL EXPERTS ON HS IN EUROPE

The leading clinical experts on HS published a common European guideline for the treatment of HS in 2015. The guideline is the experts' recommendations for treating patients with HS. In the meantime, emerging knowledge on the treatment response of HS has led to an update of the European guideline in 2016. Notably, the guideline has not yet been implemented in all European countries, and thus there may be some differences between clinics and countries on how HS is treated. The European guideline suggests that the treatment of

HS should be decided based on the individual impact of the disease and its severity (Fig. 2). In the 2015 guideline, the severity of HS is classified in three stages (Hurley I, II, III). In the 2016 update, HS is classified in inflammatory and non-inflammatory types of the disease. Skin that is inflamed appears as red, soar and warm. The severity classification and treatment are adjusted to the disease type. Inflammatory disease is classified to mild, moderate and severe according to the International Hidradenitis Suppurativa Severity Score System.

INTERNATIONAL HIDRADENITIS SUPPURATIVA SEVERITY SYSTEM SCORE

(IHS4) IS USED FOR ASSESSMENT OF A PATIENTS DISEASE SEVERITY

IT IS CALCULATED AS FOLLOWS:

INFLAMMATORY NODULES	NUMBER X 1 =	<input type="text"/>
+ ABSCESSSES	NUMBER X 2 =	<input type="text"/>
+ DRAINING TUNNELS (FISTULAE AND SINUSES)	NUMBER X 4 =	<input type="text"/>
= SEVERITY SCORE		<input type="text"/>

MILD HS: < 3 POINTS
MODERATE HS: 4-10 POINTS
SEVERE HS: > 11 POINTS

Source: Zouboulis CC, Desai N, Ermetstam L, et al. Partial actualization of the European S1 guideline for the treatment of hidradenitis suppurativa/acne inversa. Exp Dermatol. 2016; 25(suppl 2):13. (modified).

This classification helps determine which treatment will be appropriate for the individual HS patient. The more disease activity and scar tissue, the higher the score and classification.

The appropriate treatment for the individual patient must also be based on a subjective assessment of the patient's previous treatments and the effects of these. Overall, it is important to give the patient treatment that is as gentle as possible and have the least adverse effects.

MEDICAL TREATMENT

HS can be treated locally (topical treatment) by applying prescribed products onto the skin. As shown in the figure resorcinol and clindamycin can be prescribed. Injections with corticosteroids into active elements can also be used. Besides the topical treatments, systemic treatments can also be relevant. Systemic treatment is most commonly oral (tablets or capsules). Such treatment is often an antibiotic, which normally requires around ten – twelve weeks of treatment before the effects should be assessed. Antibiotic treatment kills bacteria and inhibits the immune system's overreaction. In severe cases combination therapy, with two types of antibiotics, is an option. In some cases when the above-mentioned treatments are not sufficient, biological treatment can be relevant. Such treatment works by inhibiting the immune system and its overreaction. This kind of treatment has been applied to patients with arthritis, psoriasis or inflammatory bowel disease over the last 15 years. Medical treatments can be combined with LASERs and surgery.

SURGICAL TREATMENT

Surgical treatment should be considered, especially in smaller, limited areas. There are different kinds of surgery. LASERs are used for removing affected tissue in limited areas. In some cases a method called "deroofing" can be used. In this case, fistulas are split open, the "roof", is so to speak, removed. The bottom of the tubes are covered with immature skin, and when this is exposed, it heals up fast. In more severe cases, plastic surgery, where larger areas of skin are removed, can be performed.

OTHER TREATMENTS

Besides the medical and surgical treatments, it is also important to try to minimize other factors that can contribute to aggravating HS, in particular being overweight and smoking. We know from research that many patients with HS experience an improvement in their condition if they lose weight or stop smoking.

Christos C. Zouboulis
professor, Dr. med.

RECONSTRUCTIVE SURGICAL TREATMENT OF HIDRADENITIS SUPPURATIVA

Surgical treatment of Hidradenitis suppurativa is referred to patients who cannot be assisted by medical, local e nutritional therapy.

Plastic and reconstructive surgical procedures aim to limit, in an aesthetic/reconstructive point of view, scars and their visibility even in case of minimum demolition, according to the mean young age of the patients.

Surgical treatment should be considered in case of:

- nodular and/or abscessual singular recurrent form,
- nodular confluent abscessual form,
- sub-total or total aesthetic units extended form,
- extended genital form.

Treatment of a singular lesion consists of a demolitive excision extended to the inflammatory region next to the primary site of the pathology. Fistulography using methylene blue sometimes could be performed to obtain a complete removing of the fistula. Scars usually should follow minor tension lines of the body or could be hidden into cutaneous natural sulcus or plica to optimize aesthetic results.

Multiple confluent nodular lesions must be removed by extended demolitive surgery followed by reconstructive strategy using local or loco-regional cutaneous flaps. Loco-regional cutaneous flaps sometimes are preferred by the reconstructive surgeon, even if they offer poorer aesthetic results, because they are rely on skin adjacent to the affected area less prone to recurrency of the disease. Nodular confluent abscessual forms, which involves sub-total or total aesthetic units, are treated by removing all the interested area (axilla and groin); it is necessary to consider a reconstructive strategy in this case harvesting loco-regional cutaneous flaps which allows a huge transposition of healthy tissue.

Extended nodular/abscessual genital form usually shows local infections which causes emotional and sexual discomfort; immediate reconstruction is preferred by surgical techniques based on pedicled or perforator island flaps harvested from surrounding healthy areas.

It is possible to obtain excellent aesthetic and functional results using, for selected patients, common aesthetic

procedure as abdominoplasty, thigh lift or mammoplasty. These procedures allow elevation and transposition of wide healthy skin flap with final scars concealed as in ordinary aesthetic surgery procedures.

In case of wide anatomic region involved by hidradenitis suppurativa, for example the gluteal region, it is possible to follow a flowchart:

- total excision of the lesion or the involved region,
- therapy ex- vacuum (VAC Therapy),
- healing by secondary intention and/or by using skin grafts in a selected cases similar to the treatment of II and III degree burned patients.

Considering age, sex, functional and relational discomfort and aesthetic outcomes according to the stage of disease, it is possible to get the best surgical strategy in each patient.

Alessio Caggiati MD PhD

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EMERGENCY DEPARTMENTS AND HS

CARE OF HS IN EMERGENCY DEPARTMENTS

Emergency care is a key element of any healthcare system, and there is a growing interest in increasing the satisfaction of patients and their families or carers with this healthcare setting.

Healthcare quality comprises both the scientific and technical quality of the care received, and the quality perceived by the patient during the healthcare process.

The user's point of view represents one more parameter for assessing any healthcare intervention.

User satisfaction should be understood in terms of satisfied expectations and perceived quality.

At this point, it is appropriate to note a piece of information deduced from the HS Barometer: more than 60% of the patients consulted stated that they were unsatisfied or not at all satisfied with the care that they and their family members received from the healthcare system.

When they were specifically asked about their satisfaction with the healthcare that they received in relation to HS,

more than 72% stated that they were unsatisfied or not at all satisfied.

These figures clearly contrast with the satisfaction surveys that the different regional health services regularly carry out, which, in general, reflect very high levels of satisfaction.

In Spain, although the public health system has other resources for emergency care, hospital emergency departments represent the most common care level, receiving more than 26 million visits per year, according to the data from the Spanish Ministry of Health.

Consistent with the data published by the Spanish Ministry of Health in the healthcare Barometer, hospital emergency departments are regularly used by only 16 percent of the population.

This assumes that some layers of the population use these departments several times per year, many of them within potential vulnerable groups such as paediatric, multi-disease, elderly, chronic, oncology, palliative-care, rare-disease and psychiatry patients.

Emergency departments constitute not only one of the two gateways to the healthcare system, together with primary care, but also a sort of safety net for the system itself.

As indicated in the introduction to this chapter, HS patients, whether or not they have been diagnosed with HS and regardless of how long they have had the disease, represent one group that has to make recurring use of emergency care.

DIAGNOSIS OF HS AT IN-HOSPITAL AND OUT-OF-HOSPITAL EMERGENCY SERVICES

As explained in the previous chapters, the diagnosis of HS is eminently clinical. As indicated in the diagnostic algorithm "Diagnosis of HS", visual examination of the lesions and their location and recurrence, together with other minor criteria, are the elements needed to identify HS.

An intervention on other levels that directly addresses the difficulty of diagnosing these patients and their dispersion within health systems is recommended.

The establishment of emergency reference or quick consultation offices may be very useful, as they allow the patient to be quickly referred to dermatology and seen by his or her specialist within a few days.

This type of visit would solve the problem detected by

professionals of the difficulty of follow-up of patients who, driven by the need to make a diagnosis or by the search for a solution to painful, relapsing lesions, wander through the healthcare system from one specialist to another without the professional managing to trace an accurate history of the disease which aids in diagnosis and management.

Emergency reference offices would achieve suitable healthcare continuity, once the intervention in the emergency department generally intended to relieve pain has been performed, with a procedure if minor surgery is required. This type of quick consultation could also be extrapolated to other specialities to improve the management of certain diseases in which recurrence is essential for establishing an accurate diagnosis.

MANAGEMENT OF HS IN EMERGENCY DEPARTMENTS

In relation to that explained above, the recommended management by an emergency medicine professional when faced with a patient with HS or with lesions suspected to correspond to HS appears in graphic form below. For the implementation of that management, it must be stressed that the professional should be familiar with the lesions characteristic of the disease: Hurley stage I, largely follicular nodular lesions or abscesses; Hurley stage II, presence of relapsing abscesses or of fistulas in the characteristic areas; Hurley stage III, multiple abscesses and fistulas.

Source:

*strategic health initiative to determine the standard of care
FOR PATIENTS WITH HIDRADENITIS SUPPURATIVA*

booklet made by HERCULES:

<https://bit.ly/2nWnGHQ>

PSYCHOLOGICAL ASPECTS

Hidradenitis suppurativa is known to profoundly and negatively impact on patients' quality of life, interpersonal relationships, self-esteem, workability, and perception of self-image and public image. Indeed, it may be associated with a wide spectrum of psychiatric disorders including depression, anxiety, embarrassment, stigmatization, and sexual health impairment. The psychological burden might also cause inactivity, working disability, unhealthy behaviors (i.e., high-calories food intake or

limited physical and leisure activities), and social life avoidance. Depression seems to be the most common psychiatric disorder with a prevalence ranging from 5.9% to 42.9%, while the estimated prevalence for anxiety is 3.9%. Recently, a multicentric Italian study, performed in Dermatology Units belonging to 3 University Institutions: University of Pisa, University of Rome Tor Vergata, and Catholic University of the Sacred Heart in Rome, described a peculiar psychological disturbance, named alexithymia, associated with HS. This disorder constitutes a personality trait defined as difficulty in recognizing, distinguishing, describing feelings, constricted imaginative processes, paucity of fantasy, and it is associated with unhealthy behaviors such as poor nutrition and a sedentary lifestyle, poor social support networks, low self-confidence and low confidence in others. Furthermore it impairs intimate relationships, both personal and sexual, which greatly influence physical and mental well-being. This study showed that 37.2% of HS patients were affected by alexithymia and 24.4% by borderline alexithymia. This is the first study reporting the association between HS and alexithymia, adding another psychological disorder to HS burden which might be clinically relevant as it may constitute the "warning light" of a broader and more burdensome psychological impairment and distress occurring in HS patients. The presence of alexithymia should increase physician attention on the psychological impact related to HS, referring this peculiar HS patient subgroup to psychologist for a deeper assessment of the overall psychological condition.

Key messages

- HS is a psychologically burdensome condition
- It is associated with various psychiatric disorders
- About 37% of patients are alexithymic
- Alexithymia a "warning light" of a broader and more burdensome psychological impairment and distress occurring in HS patients

Andrea Chiricozzi

*Researcher at the Department of Translation and Clinical Medicine,
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HEALTHY LIFESTYLE

Promoting healthy lifestyles can have a major impact on the progress of HS and can significantly improve the overall health status of patients.

HS is related to overweight, obesity and metabolic syndrome, attention must be paid to the quality of food and this also affects normal-weight patients. The correlation between food and inflammatory processes is well known; in fact, there are foods that can promote inflammation in the body and others that can counteract it. Studies conducted on patients with HS have analyzed and confirmed the incidence of certain foods on inflammatory processes and on skin lesions such as yeast and gluten, lactose, casein, and simple and refined carbohydrates.

The Mediterranean diet is known for its anti-inflammatory properties. Unfortunately, it has been changing over time leading to the emergence of all those diseases of the XXI that have a minimum common denominator: subclinical inflammation.

You should go back to the original Mediterranean diet. Therefore, increase the consumption of extra virgin olive oil, fish, legumes, unrefined cereals, fruits and vegetables rich in fibers, polyphenols, vitamins such as D, A, C and E and fatty acids -3. At the same time, it is essential to reduce the consumption of saturated fats, refined foods, industrial products, simple sugars and to rebalance the physiological relationship -6 / 3 which, in a ratio of 5:1. Alongside nutrition, exercise exerts a beneficial effect, increasing oxidative metabolism and reducing inflammation.

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ABOUT STEROID DRUGS

The use of steroid drugs is not an elective therapy of HS, but it can be used in emergency conditions, particularly in the phases of intense exacerbation, also possible during systemic therapies, both antibiotic and biological. The administration can take place both in systemic and intralesional form. In our experience, we have favoured systemic use for very short periods, such as pulsed intravenous therapy, for 5 days; it was a successfully therapy, in particularly in acute and painful cases; after that, we used basic therapy. In regards of local administration, international guidelines consider intralesional infiltration of steroids as 2nd choice therapy and with low evidence, (category of evidence IV° and strength of recommendation D, therefore at lower limits). The intralesional use is unfortunately burdened by pain, in places already inflamed and painful. Perhaps, it may help to cool the area and add anaesthetic solution before administration.

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OTHER THERAPIES-HYPERBARIC OXYGEN THERAPY

Hyperbaric oxygen therapy (HBOT) is a hospital medical treatment technique consisting of inhaling pure oxygen or an oxygenated mixture at a pressure above ambient atmospheric pressure. The pressure levels and the duration of a session of HBOT depend on the pathology to be treated.

At the level of the body, the HBOT will thus induce an increase in the partial pressure of the oxygen in the blood but also at the level of the interstitial tissue spaces. This result in HBO-specific effects on hypoxic tissues or organs, effects that could not be reproduced by the simple breathing of oxygen to the mask in a hospital room at ambient atmospheric pressure: oxygen supply, vasomotion phenomena in tissue microcirculation, anti-infectious effects. Finally, a metabolic action in healing phenomena by stimulation of some cutaneous cells of the body (fibroblasts) and by activation mediators involved in the genesis of small blood vessels.

HBOT thus facilitates healing on difficult terrain such as

the advanced stages of Verneuil's disease, particularly those requiring surgical procedures with wide or deep ablations of the affected tissues. It also reduces the risk of post-operative complications such as infections.

In practice and in hyperbaric center of Lyon, the session of HBO lasts 105 minutes during which the patient breathes pure oxygen at 2.5 times the atmospheric pressure. He can sit or lie down. In general, only one session per day is performed and it takes about 30 sessions to reach the scar goal (sometimes much more when the patient is smokers). Medical consultations are also planned throughout the hyperbaric treatment to follow the patient and his skin evolution

Doctor Thierry Joffre

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Chapter 7 | PATIENTS' EXPERIENCE

EXPERIENCE IS PERSONAL AND HOW A PATIENT, THEIR FAMILY OR CARER FEELS ABOUT THEIR EXPERIENCE OF A SERVICE IS VITAL.

IMPROVING PATIENTS' EXPERIENCE IS A KEY AIM FOR EFPO.

BY ASKING, MONITORING AND ACTING UPON PATIENT FEEDBACK IT ENABLES ALL OF US TO MAKE IMPROVEMENTS IN THE HS.

GIANFRANCO'S EXPERIENCE

Sometimes I remember the late summer day of 2013 when, after a terrible week with a fever over 40 degrees and a brutal incision in my right armpit, a dermatologist gave a name to my illness. "Hidradenitis Suppurativa" she said.

I remember the following days them as the darkest of my life. "There is no cure, there are only therapies but no certainty". Response: "Antibiotics and retinoids".

But meanwhile, not even a month later I was sick again, the other armpit was inflamed.

My almost twenty years seemed suddenly heavier. The fear of not being able to predict and manage the following acute episode paralyzed me.

I did not want to talk about my fear to reassure my relatives.

Looking on the internet I discovered the Inversa Onlus association and in bad luck I discovered that I was also "lucky".

Someone could give me a hand, advices, support and the information that could help me.

A few months later I went to see the doctor who was recommended to me. "Antibiotics and retinoids", again! but with the promise to meet again soon if there were no improvements.

But the situation continued worse with increasingly frequent suppuration and increasingly painful abscesses.

I went back to the doctor: "Let's try an anti-inflammatory diet". Another year slipped away, I have lost almost 10 kg, but the situation did not improve.

"Let's try with botulinum toxin for at least 8-10 months". Nothing, no improvement.

I did not lose hope, but this condition became increasingly difficult to manage alone and without my family by my side due to the distance.

However, comparing myself with members of the patient association, learning more about HS and how to medicate, helped me to develop a positively change in the approach of managing the disease.

I did not want to give up, I went back to the doctor and this time after a series of ultrasounds the response changed: "The situation is serious, your abscesses are very extensive and deep, we have to try with a drug for autoimmune diseases, cyclosporin A".

Within six months, I started feeling better, but I still had acute episodes, but at least after couple of months and not after few weeks.

A year later the size of my abscesses had been reduced a lot!

"Ok, let's continue with the therapy and add the ND-Yag laser".

The situation has continued to improve until May 2017 when I underwent to a deroofting surgery in the hands of the same doctor who had followed me since 2013.

Today I cannot say I'm healed, but I can say I feel better,

without important relapses and I finally started to rejoice in life.

What did I learn? Do not be discouraged, you need to ask for help and you have to change your lifestyle.

You need to know your illness to know how to fight and you must trust your doctor.

But above all, we must thank Inversa Onlus, without which today I would probably still be in my bed crying without answers.

*Gianfranco Lombardo
Italy*

MONICA'S EXPERIENCE

I did not know what the HS was, not even at my first surgery which, according to the surgeon who operated on me, was simply ingrown hairs that then during the same, he realized that I was full of small cysts and he took them off with the laser.

I heard about HS for the first time 3 years ago in my second speech.

Unfortunately, during all my illness I have taken a lot of antibiotics that have not always helped me.

Before getting to know the Inversa Onlus Association I have been engraved many times without anesthesia and in the face of my complaints and screams during this practice I have been treated very hard. This happened several times and with different doctors.

In the meantime, since I never gave up, I did research on my own and finally I found My guardian angels ... Giusi Pintori, the Inversa Onlus Association and patient support. They directed me to health experts and I went through a new clinical and surgical path. I found a little more sensitivity and understanding.

The support that carries out the association has been my salvation several times in fact the "hs monster" is devastating for the body but also for the mind and being helped by those who know what it entails in every aspect is essential.

The HS has conditioned my life a lot, but I have to recognize that now thanks to the incessant support of the Association I can fight it with more energy and hope.

*Monica Benedetti
Italy*

MARIE-FRANCE'S EXPERIENCE

WHEN FROM HELP IS REBORN HOPE

The disease appeared late in my case. Married, I already had my three children. I worked as a Clinical Research Associate and life was sweet and beautiful. Until 1991, when the first lesions appeared. Despite a rapid diagnosis of Hidradenitis Suppurativa (HS), and a first intervention in the wake, things have not really improved. A year later I found myself at the most severe stage of the disease and interventions began to follow one another, each time a little wider, a little more mutilating. Some were so extensive and deep that several transplants were needed. The work stoppages followed one another until an occupational doctor finally declared me unfit for work and put me on disability. That day, it is not only a job that I lost, it's a whole part of my life, my future. Revolted, angry at this injustice that I could not understand, I wanted fights me. So I looked for help, that I found it in Canada in the English-speaking area. That's when I decided to create an association in France. After a year and a half of work, the first AFRH website was put online in November 1999 and the AFRH officially founded in February 2000. Investing me in the association really helped me overcome my own suffering, the many treatments that often made me sick, and all the operations I had to undergo. I learned to know about my illness, learned not to be ashamed of it. The word frees more than one can believe. You have to dare to speak, and do not hesitate to ask for help! Today, after more than 18 years fighting for and alongside patients, I am convinced that it is together that we can and will advance knowledge about HS in Europe. Our federation is a chance that we are giving to all European patients to get the opportunity to come out of the shadows and hope to see soon emerge a cure.

*Marie-France Bru-Daprés
President and Founder of AFRH (Association Française pour la
Recherche sur l'Hidrosadénite)- www.afrh.org*

CHRISTEL'S EXPERIENCE

My journey, my life with Verneuil's disease also called 'Hidradenitis suppurativa'.

Let me introduce myself. My name is Christel, my daughter and I suffer from the disease. I've been suffering since the age of 17, I am actually 46. Just like many people in the same situation, I've had to wait many agonising years before being able to put a name on what was wrong. I was finally diagnosed at the age of 31.

After many operations, many local as well as oral antibiotic treatments, my personal and professional life changed drastically. The disease prevented me from doing many things such as playing with my kids, taking them swimming... The pain was often so unbearable that I couldn't even look after them. I often left them with their grand-mother 's. I felt so guilty and considered myself like a bad mother. But what else could I do? The pain was so intense that I couldn't carry them nor take them to school.

Professionally speaking, the pain was the reason why I've had to resign from a job I loved so much. Travelling became unbearable (120kms a day) the more tired I was the more eruptions appeared. Even though my post was re attributed to me under the 'disabled worker' status, my doctor and I came to the same conclusion. I wouldn't be able to carry on working. I was then labelled too 'disabled ' to work, but not enough to benefit from the 'disability allowance'. The illness took my life and my social status. So I've decided to get involved in this association to help and support other people suffering from Verneuil's disease. Getting involved allowed me to build a social network and helped me get through rough times.

*Christel Vincent
France*

HÉLÈNE'S EXPERIENCE

It's been Twenty years now that I have HS. I was 37 years Old when the disease appeared. I was already married, I had two children, a good job when an abscess appeared in the hollow of my armpit. I did not pay attention to this the first time, thinking that it was going to disappear as quickly as he had come, but day after day, he became bulky and more painful. I couldn't do anything normally, doing simple gestures becomes hard. My doctor was not able to tell me right away how badly I was suffering and it took me two years to put a name on this disease. I realized that most patients had a lot of trouble finding competent doctors and that the time to diagnose was abnormally long. I quickly met other patients and we decided to regroup and create an association to make known the disease and reduce this delay. "Solidarité Verneuil" was created in 2009 in Lyon (France) and since then we have been present throughout France thanks to the network ho help us to make of medical contacts. The Prevention and the personalized accompaniment of patients is undoubtedly the key to a return to a life closest to normal for all patients with Hidradenitis Suppurativa (HS). With the absence of a clearly defined health policy on the treatment of this pathology, the mobilization of patients in associations at national and European level is essential for an awareness of public authorities and the medical profession throughout the world. 'Europe. EFPO is the spokesperson for thousands of patients and hopes to finally find the treatments they absolutely need for a return to a better quality of life.

*Hélène RAYNAL,
Présidente fondatrice de Solidarité Verneuil/HS France
<http://www.solidarite-verneuil.org/>*

CHRISTELLE'S EXPERIENCE

My name is Christelle, I am 47 years old and I live in the south-west of France. HS arrived in my life when I was 17. The first sign was an abscess in the left armpit. My family told me that it was a boil, until I was 20 years old, I didn't know what to do so I tried to put an antiseptic on it. With the time, it was more regular, so I went to the emergency department to meet a surgeon. It was done regularly, then it was every two weeks, he was sure there was a cause, so he sent me to several hospitals and dermatologists, I came home knowing nothing more, nobody could tell me what was happening to me. They told me that the reason was the stress and they told me that my Hygiene was not good, and it was maybe another reason. For 16 years, I wandered doctors, dermatologists, and surgery was the only way for me to feel better.

At 36 years old, a physiotherapist gave me the name of a dermatologist In Bordeaux, he told me that I had Verneuil's disease (HS) and that there was no treatment except surgery. to teach alone to understand what this disease was. I have learned from those 13 years learning about it on the internet and Facebook groups, I know the disease well and a lot of doctors and surgeons. I talk about it often and make my best to help people to get solutions and help them to find answers more quickly. I wish there was more communication on this sickness, I will fight for it because after what happened to me any many others it can't continue like that. I will never give up. HS won on my body, but she will never have my heart and my motivation.

*Christelle Cologni
France*

SILVIA'S EXPERIENCE

Saying that a disease doesn't define you may sound literary, but the truth is that it's not that way. When your condition is a chronic one, it shapes you to its liking, it makes you in its image and likeness and it changes your behavior to the way it wants, even though you don't want to. You learn to live under its guideline, under what and when it allows you to do. Sometimes you think you are winning the battle but it's just an illusion.

This is the reality every patient suffering from a chronic and weakening disease has to struggle with.

Pain, frustration, loneliness, constantly going to the doctors', hospitals, surgeries, medical cares, more and more invasive medications, abandonment, sadness, not being able to live a normal life, to go out and have a coffee, to make that trip you've always wished, to have a family of your own, to work out, to wear the clothes you like, to go to a live concert, to study at the university, to go on Erasmus, to swim or to go to a SPA, to wear that bikini you love, or that lingerie, or those jeans...

When you are not able to say a word, when everything is decided by your condition, when it invades everything, and the rest of the world doesn't understand it, and even more, they don't care is when you realize how big its tentacles are, how much it's taking from you.

At this very moment you realize how strong you are, how your mind has been able to adapt to every single obstacle this condition has put in your way. Between sobs and disappointments, you find out you can still achieve something positive from these terrible things. That strength keeps you going and fighting and it makes you assume as your own other patient's tears.

And that's the reason why I passed from staying in a bed for one year without being able to move to handle the Spanish HS Association. I turned my pain into strength, my anger into energy, my desperation into hope, my embarrassment into joy and this way I became the image and the voice of the native Spanish speakers from the world. Nowadays Asendhi is a reference on an international level. We are a big family.

That is my strength, knowing that everyday when I wake up I'm going to make other's lives better.

*Silvia Lobo Benito
President of Asociación de Enfermos de Hidrosadenitis- ASENDHI
www.asendhi.org*

GLORIA'S EXPERIENCE

I'm forty-one. I got married fourteen years ago and I have two daughters. One is nine and the other one is ten. I'm a pensioner for permanent disability due to my condition, Hidradenitis Suppurativa, Hurley stage III.

It all began as a fairy tale and it became a horror story. Two days after my little one was born, I felt some small bumps in my breast and in my right armpit. At the beginning doctors thought it was mastitis but three years later they found out it was HS. They were three years having temperature, pain, suppuration, fear and no understanding at all. Three years with no help, no medical, work or social support.

But, despite this, I feel lucky because most of the cases are diagnosed after nine years of struggling with this condition.

It was very hard being ill all the time and learning this is a condition nobody knows about. I used to feel so lost... The worst things were rejection at work, no empathy from co-workers and bosses, humiliation because I felt that I was no longer who I used to be. I had work so hard to get some labor recognition, a status and it was worthless. I was ill, and it didn't matter how hard I tried, I could never achieve my goals. Stress at work made my condition worse, because this is one of the reasons why more boils appear. I needed to hide my dressings and my scars because I felt that if I they found out at work, they were not going to count me in anymore.

I started having cortisone, immunosuppressant and surgeries came. And I didn't care about my job anymore and sometimes even about life either. By that time I felt smaller and smaller everyday.

Nobody helped me. I fought to have a disability recognized and after several years, humiliations, deplorable medical inspections and trials, I finally got it and with it some peace of mind, at least about money.

My best support and medicine are my closest relatives, most of all my daughters, who have been my engine. Without them I wouldn't have had the strength to keep on going. My husband is my nurse. He knows how I feel just looking at me. He is my biggest supporter. And, of course, my mother who keeps me going. The truth is that I can only count on my closest relatives and a few friends.

I want to keep on fighting through our association to get new better medicines and research breakthroughs

because I have two daughters who haven't been able to see her mother being healthy as any other kid their ages and I don't want them to suffer the same way I have. And if in the future they suffer from this terrible condition I don't want them to have the same problems I've faced and I want medicines or treatments to help them having a normal life and to be happy.

*Gloria María García Cervi
Spain*

GLOSSARY

ABSCESS

A painful collection of pus that has built up within the tissue of the body

AUTOIMMUNE DISEASE

A disease where the immune system produces antibodies against a normal tissue. The antibodies then attack the body's own tissue and causes inflammation (see "inflammation" below)

AUTO-INFLAMMATORY DISEASE

If the immune system turns on too quickly and turns off too late, the disease is called auto-inflammatory. This leads to inflammation which burns too long and with too much force

BIOFILM

A slimy layer of microorganisms, such as bacteria, that stick together

COMORBIDITY

The existence of more than one disorder or disease in an individual at the same time

DERMATOLOGIST

A physician who is specialized in diagnosing and treating skin disorders

DYSLIPIDAEMIA

"Dys" means "wrong", and "lipid" is fat in the blood. Blood fat is composed by cholesterol and triglycerides. At dyslipidaemia, there is too much of the fats which increase the risk of heart disease, and too little of fats which prevent it

ENDOCRINOLOGIST

A physician who is specialized in diagnosing and treating hormone-related conditions, such as diabetes

FISTULA

A tube-like connection between the skin surface and a mucous surface, e.g. the bowels

HURLEY STAGES

A staging system used in HS to describe how severe the disease is, from stage I to III

INFECTION

Inflammatory state due to an invasion of microorganisms, such as bacteria

INFLAMMATION

The immune system's response to stimulus, not necessarily infection

METABOLIC SYNDROME

Disturbance of the fat and sugar metabolism which induces overweight, diabetes, high cholesterol, arteriosclerosis etc. Increases the risk of heart and circulatory diseases

SINUS TRACT

A tunnel under the skin, from skin surface to skin surface

STEROIDS

Synthetic hormones which dampens inflammation

SYSTEMIC TREATMENT

Treatment with tablets or injection

TOPICAL TREATMENT

Local treatment on the skin

WE HAVE LISTED SOME QUESTIONS THAT CAN BE RELEVANT WHEN YOU TALK TO YOUR PHYSICIAN

Always try to be as precise and concrete as possible when you speak with your physician. Remember that the physician can only help you if you give him or her the full picture of your situation.

It is important that you and your physician put together a holistic treatment plan which deals with your specific situation and needs.

SINCE YOUR LAST CONSULTATION

- How have you been feeling since your last consultation?
- How many boils and abscesses have you had lately and where are they located?
- How does the disease affect you mood and can you cope? Or does it make you feel depressed?
- Does the condition affect your social life, your relationship with your family and your romantic relationships?
- How does HS influence your ability to manage your work or your educational situation?
- Do you get enough sleep and rest?

TREATMENT

- Is your disease at rest?
- Does your treatment work as expected?
- Do you need other or a supplementary treatment?
- Do you experience side effects from the medicine?

SUPPORTIVE TREATMENT

- Do you get sufficient help to relieve the pain?
- Would you like help to lose weight or exercise?
- Do you need support to quit smoking?

RELATED DISEASES

- Do you have other skin problems?
- Do you have stomach problems, e.g. pain, bad stomach, diarrhoea or blood in the stool?
- Do you have swollen or sore joints?
- Have you had heart palpitations, dizziness or general malaise?

Source: Patient Association HS Denmark

HS research is in rapid development. New knowledge, therapies and stronger cooperation between researchers, therapists and patient associations give hope to you and all those with HS.

The ambition of this publication is to gather and communicate the insights from both patients and specialists, since both perspectives are of major importance for HS understanding and treatment.

We hope that this publication can contribute to a more nuanced understanding of HS among patients and relatives, and in this way, strengthen the cooperation and partnership between patients and physicians.



WHAT IS EFPO

The Hidradenitis Europe – European Federation of HS Patients’ Organisations (EFPO) is an international non-profit organisation for patients’ organisations of people with Hidradenitis suppurativa (HS).

EFPO was founded in 2016 by the patients’ organisations on HS from France, Spain, The Netherlands, Italy, Belgium and Denmark. The main objective of the organisation is to increase the awareness on the disease and support you – HS patients across Europe – in the fight against HS.