

KNOWLEDGE ON HS

- UNDERSTAND MORE, AND IMPROVE YOUR LIFE LIVING WITH HS

News from Hidradenitis Suppurativa
research for patients and their relatives



HS develops where
SKIN MEETS SKIN.

The condition is most often
located in the armpits, under
the breasts and in the groin.

WHAT IS HS?

Hidradenitis Suppurativa (HS) is a chronic skin disease which occurs as repeated outbreaks of sore pimples or abscesses, located on parts of the body where skin meets skin. The pimples and abscesses are, however, not ordinary pimples and abscesses, and HS 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 starts after puberty and three out of four patients are female. Studies have shown that 1%-2% of the adult population suffers from HS to varying degrees.

CONTENT

	FOREWORD	04
Chapter 1	GENETICS	
	Why do you get HS?	06
Chapter 2	BACTERIA	
	What role do bacteria have in HS?.....	09
Chapter 3	CLASSIFICATIONS	
	How do physicians monitor the development of your HS?	11
Chapter 4	IMMUNOLOGY	
	What is the connection between the immune system and HS?	15
Chapter 5	COMORBIDITY	
	What other diseases should you be aware of?....	17
Chapter 6	TREATMENT	
	How is HS treated?	22
	GLOSSARY	26
	QUESTIONS	
	Relevant questions for your next physician’s appointment	27

HS RESEARCH IS DEVELOPING RAPIDLY

Hidradenitis suppurativa (HS, also known as acne inversa or Verneuil's disease) is a chronic skin disease, which occurs as repeated outbreaks of sore skin reactions that resemble abscesses or lesions.

Although HS is considered to be relatively common in the population, it is not well-known among the general population and many physicians. Many patients suffer in silence and are not aware of their diagnosis and treatment options.

We still see patients who drift around randomly in the health system without a diagnosis. These patients experience the consequences of having a disease that only few people are aware of and where treatment offers may, in many cases, be inadequate. HS as a condition has often been tabooed and overlooked. However, change is coming and the progress that has been made in understanding HS is now finally starting to materialise – not least within the field of HS research, which has never been more intensive than right now.

RESEARCH RESULTS IN PROGRESS that benefits you and me

What treatment is the best for you? How do you deal with the pain associated with HS? And what do you do about your sleeplessness? Although there are many questions and few answers, there is now a lot of help, good advice and tips on living

with HS. But it is important that you as a patient can distinguish between the knowledge derived from research and the advice that is based on individual experiences. Special diet plans or a cream that may prevent new abscesses may be nice to try, as long as it does not harm you. The advice may work for some, while others do not experience the desired effect.

However, scientific research is different. Research examines theses and theories within a controlled framework. Here, there are comprehensive requirements for impartiality and documentation, and it is, for example, extremely demanding to get approval for a new drug. There are many requirements on how many patients should be included in studies and that results should be made publicly available for everyone to see. Impartiality entails that results will only be regarded as valid if different researchers conclude the same results.

Research takes time and is difficult, but it is the only sure way to acquire safe knowledge and better treatments.

Knowledge gives HOPE

Today, HS research is developing rapidly. New knowledge, therapies and collaborations among researchers, therapists and patient associations give hope for you, who live with HS. A good indicator of this rapid development is the number of HS research studies being published. Worldwide, the number of HS-related articles has risen dramatically over time. By 1971, just 62 HS scientific articles had ever been published. By 2000 there were 353 articles, and by 2017 the total number of published articles rose to more than 1700. Despite this massive increase in HS research, there remain many unanswered questions and therefore, there are many good reasons to continue the work.

The world's biggest HS conference

This positive development was clear to all who attended the 6th Conference of the EHSF 2017 – the world's largest HS conference – which took place in Copenhagen in February 2017. At the annual EHSF conferences, dermatologists, surgeons, gastroenterologists, rheumatologists, nurses, patients and other experts from around the world engage in HS discussions. These experts interact across their fields of specialty, united in their shared interest in HS research. The intensity in the activities, cooperation and efforts has never been higher. While previously HS conferences were only ever filled small venues, participants have now moved into the biggest venues. The conference gave hope to you and the others that comprise the 1%-2% of the world's population living with HS.

A common EFFORT

There is more good news. Today, we witness strong collaborations between researchers across national borders. For many years, HS was a low-interest area, and we are therefore excited to experience the great support expressed at the EHSF conferences and in HS research in general. HS patients and patient associations are joining forces across borders, which is unique. They are being heard and are actively contributing to global research projects. It is thus, patients, physicians and nurses that pull together.

It's about QUALITY OF LIFE

Whether we are talking about drugs or surgery, quality of life is important. A treatment that does not improve your quality of life is of limited use. Living with a chronic disease can be tough, and even small things can make a big difference. The more you know about your condition, and the better you can explain how the

disease and the treatments affect you, the better you and your physician can work together in bringing the disease to rest.

RESEARCH RESULTS that are relevant to you

In the following pages we have collected some HS research results relevant to you and everyone living with HS. These pages give you an insight into the fundamental immunological aspects of HS, the importance of genetics, HS classifications, important complications that you should be aware of, and an overview of the various HS treatment options.

The ambition of this publication is to gather and communicate insights from patients and specialists, since both are indispensable in HS treatment. We hope that this publication can contribute to a more nuanced understanding of HS and thereby strengthen the cooperation between patients and physicians.

February 2018



Bente Villumsen
Chairperson of the Patient Association
HS Denmark



Christos C. Zouboulis
Professor, Dr. med.
President of the European Hidradenitis
Suppurativa Foundation (EHSF)



Gregor Jemec
Professor, Dr. med.
President of the EHSF-conference 2017
in Copenhagen

GENETICS MATTER

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.

Why me? This is a question that occupies the minds of many HS patients including Bente Villumsen, chairperson of the Patient Association HS Denmark.

"My father had HS, and when I was 17 years old I didn't want to have children myself because I wouldn't risk passing on the disease to them. My mother said that by then a cure would probably have been found. This, however, requires research and at that time no research was being carried out. Back in the days, we didn't know anything, not even that the disease had a name. And if we find it difficult to talk about HS today, it was completely impossible back then," says Bente Villumsen.

Knowledge empowers you

Bente Villumsen says that 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 which many HS patients unfortunately carry around with them. Having knowledge about your disease empowers you," says Bente Villumsen.

"In regards to my own children, I have been very aware of the fact that they might contract HS. As a consequence, my daughter was diagnosed and received the adequate treatment at a very early stage."

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

Be aware of the symptoms

If you or others in your 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 your 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.

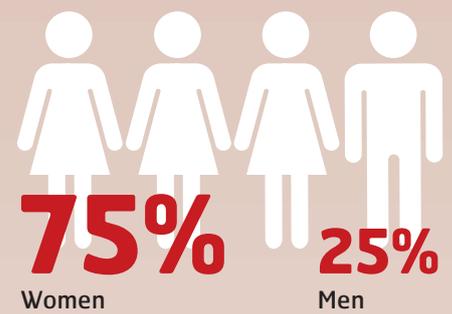
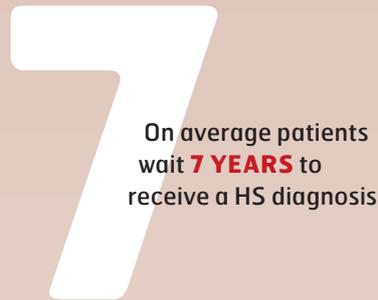
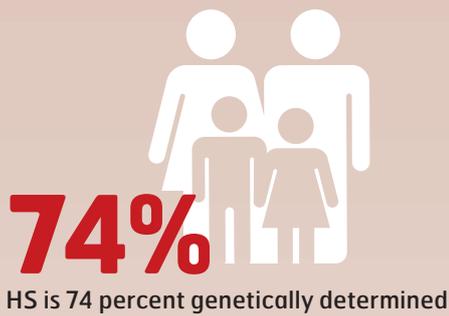
If your child has HS

Make sure to act immediately 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 early. In addition, you may have a patient association in your country that will be able to offer additional help.

What causes HS?

Fortunately, there is lots of current HS research and Bente Villumsen points to the latest EHSF conference, where many different articles and scientific presentations addressed the issue of the causes of HS.

"I especially noticed a Dutch twin-study that concluded that HS is **74%** genetically determined. This corresponds to my own observations. My father had HS and we believe my grandfather had it as well – and my daughter also has the disease, so I have always known that it was hereditary. At the same time you might be confused, as many other studies at the same conference 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. In my experience hormones and stress also have a negative impact on the



disease. Personally, my HS flares when I am too busy and my stress level increases. As HS patients, we have very good reasons to be confused, and I have a feeling that there are still a million of questions we cannot answer yet”, says Bente Villumsen.

Smoking and obesity are not the answers to everything

Bente points out that there are still 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 haven't been diagnosed and if they have, they might only be

“As patients we constantly experience running into blind alleys in the healthcare system, often because the physicians do not know of the disease.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

advised to lose weight or quit smoking. As an HS patient you almost get the feeling that you have yourself to blame for the disease and that you are not doing the right things, and consequently that the disease is self-inflicted. I am aware that obesity and smoking habits play a role, but it is also a fact that slim people never smoked also have the disease,” says Bente Villumsen.

The influence of genes on HS

HS is a complex disease with various 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, HS research is characterised by the fact that the disease, up until a few years ago, was a low-interest among researchers. Consequently, the scientific community is lacking basic scientific research and data that supports the existing HS 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.



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 psychological 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 ask 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 gynaecologist and a psychologist or psychiatrist over the course of your disease. Read more about related diseases on page 17.

*Source: Gregor Jemec.
Professor, Dr. med.*

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

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.

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 indicates that HS patients have fewer bacteria than people who do not have HS.

If you are knowledgeable about HS, you also know that having HS 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.

“The important message is that HS is not caused by poor hygiene. It is not an infection and the abscesses are not contagious, even though they look like ordinary boils.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

“This is why we sometimes refer to it as skin reactions that look like boils. The body reacts as if it has a boil, but it isn't a boil. This also makes you wonder why you are recommended to wash with e.g. disinfectant soap, which is used in hospitals, but I have used it myself during periods. Not that it helps against HS, but when the skin is damaged it may be an entry point for bacteria. It is a very good reason to disinfect and cover the wound as well,” says Bente Villumsen

chairperson of the Patient Association HS Denmark.

She recommends that you react if the skin gets extra red, swollen or sore. You can request a test of the skin either at your physician, specialist or at the 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.”

*Hans Christian Ring,
MD, Ph.D.*

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 microbiological conditions before and after the boil has formed,” Dr. Ring explains.



Study 1 The role of bacteria prior to the outburst of abscesses

"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 HS.

"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 because 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 an interesting finding concerning 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 must be targeted at biofilm removal. 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 HS lesions with tracts (sinus tracts) are identified, with antibiotics to target the biofilm.

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: Patient Association HS Denmark.

IMPORTANT KNOWLEDGE ABOUT ACUTE PAIN

HS patients who go to the hospital emergency ward for urgent help with acute pain from abscesses often experience that the physician on duty decides to cut the abscess open to relieve the pressure. If a boil is big and on the point of bursting releasing a lot of pus, the best solution may be to drain it to alleviate the pain. This may help relieve the pain momentarily, but the abscess will return. There is also another solution that does not involve 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 a 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.

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 – 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, Ph.D. student at the Dermatological Department at Zealand University Hospital, and part of the research team.

How important is classification for patients?

According to Bente Villumsen, chairperson of the Patient Association HS Denmark, HS severity cannot be rated without asking the patient. “The classifications itself is of no direct importance to us as patients, but the classification is important in determining what kind of treatments we are being offered. That being said, your own experiences determine how severe your HS is.”

“An abscess may hurt just as much in Hurley I as in Hurley III (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.”

16%

have been suicidal*

20%

consider **NOT** having children because of the disease*

**of Danish HS patients.
Source: KORA 2016*

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*



"In this light, it makes good sense to use different classification methods and then combine them," says Bente Villumsen.

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, Associate Professor, MD, Ph.D at Copenhagen University and Zealand University Hospital.

"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,
Associate Professor, MD, Ph.D*

"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, 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 HS 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 few 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.

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 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 feel 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 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.

Criteria for HS diagnosis

- There must be typical lesions: painful, deep-lying boils or lumps (pimple-like) perhaps connected under the skin (sinus tracts) and possibly 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 abscesses 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 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 abscesses. Only few patients are categorised 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

” As a patient, you can help by using the prescribed treatment and evaluate the effect together with your physician. Sometimes a treatment works for a period of time. If the treatment does not work, or the disease progresses, you have to reconsider whether it is the correct treatment.”

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

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 a gut examination prior to a given surgery.

HS AFFECTS

1-2%

of the population

DID YOU KNOW

People living with HS have a worse quality of life than cancer patients.



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 method 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 work, 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, 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 comprises the following themes: pain, clinical signs, disease development, patient and physician angle, HS-specific quality of life and symptoms.

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

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 under the skin. 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.

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.

“When I talk to other HS patients about our immune system being overactive, there are always many questions: is that why I get ill so often? Or does my overactive immune system mean that I am less ill than others? As a patient, I would like to have the theoretical understanding of my disease and understand what is going on in the immune system, but it is extremely complex to understand and use terms such as cytokine, lymphocytes and interleukins.”

“I simply acknowledge that the immune system is a key area in understanding HS, and an area which is being massively researched at present.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

Bente Villumsen points out that understanding the importance of the immune system is knowledge that can be transferred into actions that can improve the quality of life for HS patients: “We know that sugar and stress affect the immune system, so this is an area where we can improve the situation. If you drink two litres of soft drinks a day, it might be a good idea to reduce that amount. In general, it is important that you take care of yourself, and here I usually use the following three tips which 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

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

Take care of yourself

When your immune system attacks something which it is not supposed to attack, your HS might flare up and you might experience a physical reaction from your body. Many people with HS do not 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 vitamin D level measured and take a supplement to boost their immune system. Others try to eat an anti-inflammatory diet, for example avoiding sugar and white bread, and in return take ginger and fish oil supplements. 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 helps. If you wake up in the morning and feel poorly, it helps to get out of bed and go outside. When you are active, the soreness and condition 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

HS is a condition that comes and goes and in this same way your symptoms change. Various 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 some related diseases (see Chapter 5).

outbreak. "Here it would be obvious to decline some tasks or ask for help," says Bente Villumsen.

The immune system is one of our most powerful biological mechanisms

Professor dr. med Gregor Jemec, Zealand University Hospital, 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 autoimmune disease," says Gregor Jemec.

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 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 such research has resulted in the development of new 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 system 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 HS CONFERENCE

Headlines of 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.

WHAT OTHER DISEASES SHOULD YOU BE AWARE OF?

If you have HS you have an increased risk of getting other related diseases.

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 diseases 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.

“What is merely suspicion, and what is well-founded knowledge that we have to take seriously? 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. And as a patients' association we must always stay balanced when communicating about diseases related to HS,” says Bente Villumsen, chairperson of the Patient Association HS Denmark.

“ In order to help HS patients, we want to inform and increase the level of knowledge about diseases related to HS. On the other hand, we do not want to create unnecessary fear.”

*Gregor Jemec,
professor, Dr. med.*

Diseases most commonly related to HS

When you have HS, research shows that there are different diseases that you have an increased risk of developing. 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,
Associate Professor, MD, Ph.D*

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 body 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.

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 and related tests. A talk about health-promoting habits such as help to lose weight, dietary advice and exercise, might also help.

Losing weight requires help, support and HS treatment

Many HS patients are advised to lose weight, and according to Bente Villumsen this suggestion must never stand alone. “I am well aware that overweight is important when dealing with HS, however, we too often experience that the message from the physician is that you need to lose weight. But losing many kilos is not that easy. You 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.”

“ When you live with boils and daily pain it can be very difficult to be organized enough to eat healthy and almost impossible to exercise. A good and effective treatment is therefore an important precondition for weight loss.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

Diabetes (Type 2)

Diabetes is also a part of the metabolic syndrome and is also more common in 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 blood pressure is a part of the metabolic syndrome, which is more frequent 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 these symptoms worsen 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. 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 pain, 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 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 with 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.

40% experience that they are **anxious or depressed**

16% have had **suicidal thoughts**

20% have considered **not having children because of the disease**



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. With HS, 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 incidence of suicidal thoughts than the general population.

Sleep disturbance

It is clear that you can 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 exactly 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.

Bente Villumsen stresses that the tiredness that you feel as a HS patient is very noticeable and affects your daily life even after a good night's sleep.

“The fatigue we experience is not the same as if you have been at work a whole day or have had too little sleep. Our fatigue is a flu-like tiredness in the body which is still there even when you wake up after 12 hours sleep.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

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*



THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

You are in 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 inform your physician.

Sleep

Active HS may affect your sleep. Many experience 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 due to the physical unrest, which the body experiences when fighting inflammation.

Some people with HS report 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 a break 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 many 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.

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

Headlines of some of the latest publications, comorbidity.

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, Ł. Matusiak et al.

A Polish study tests the itching and pain that is a common symptom for many HS patients, but which many think they are the only ones to suffer from. This study documents that itching is a major problem. In the study, it almost all patients report pain and more than 40% of HS patients 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 approximate 60% of the patients were bothered by itching. The researchers conclude that itching is often overlooked in HS.

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 is adjusted to the disease type. Inflammatory disease is classified to mild, moderate and severe according to the International Hidradenitis Suppurativa Severity Score System (Fig. 1).

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 style="width: 50px; height: 20px; border: 1px solid white;" type="text"/>
+ Abscesses	number x 2 =	<input style="width: 50px; height: 20px; border: 1px solid white;" type="text"/>
+ Draining tunnels (fistulae and sinuses)	number x 4 =	<input style="width: 50px; height: 20px; border: 1px solid white;" type="text"/>
= Severity score		<input style="width: 50px; height: 20px; border: 1px solid white;" type="text"/>

Mild HS: ≤ 3 points

Moderate HS: 4-10 points

Severe HS: ≥ 11 points

Fig. 1.

Source: Zouboulis CC, Desai N, Emtestam 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).

” We clearly recommend our members to get a new physician if their GP isn’t willing to refer them to a dermatologist.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

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.

1 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.

Treatments are improving

Bente Villumsen Chairperson of the Patient Association HS Denmark has, as a patient and active in the patient association, been following the development of new treatments close. “Some of our members have been drifting around in the health system for decades and have been treated differently depending on where they have ended up, and without getting the diagnosis HS.

” If the treatment doesn’t make you feel better, the treatment isn’t good enough.”

*Bente Villumsen,
chairperson of the Patient
Association HS Denmark*

2 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 “deroofting” 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.

3 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.

*Source: Christos C. Zouboulis
professor, Dr. med.*

Because of this, we are very excited about the development that is underway; Research is accelerating and the treatment options are improving. At the same time the awareness of HS is growing day by day and even though the disease is still tabooed, hard to talk about and hard to live with, things are finally moving in the right direction”, says Bente Villumsen.



THE HS TREATMENT CHOICES

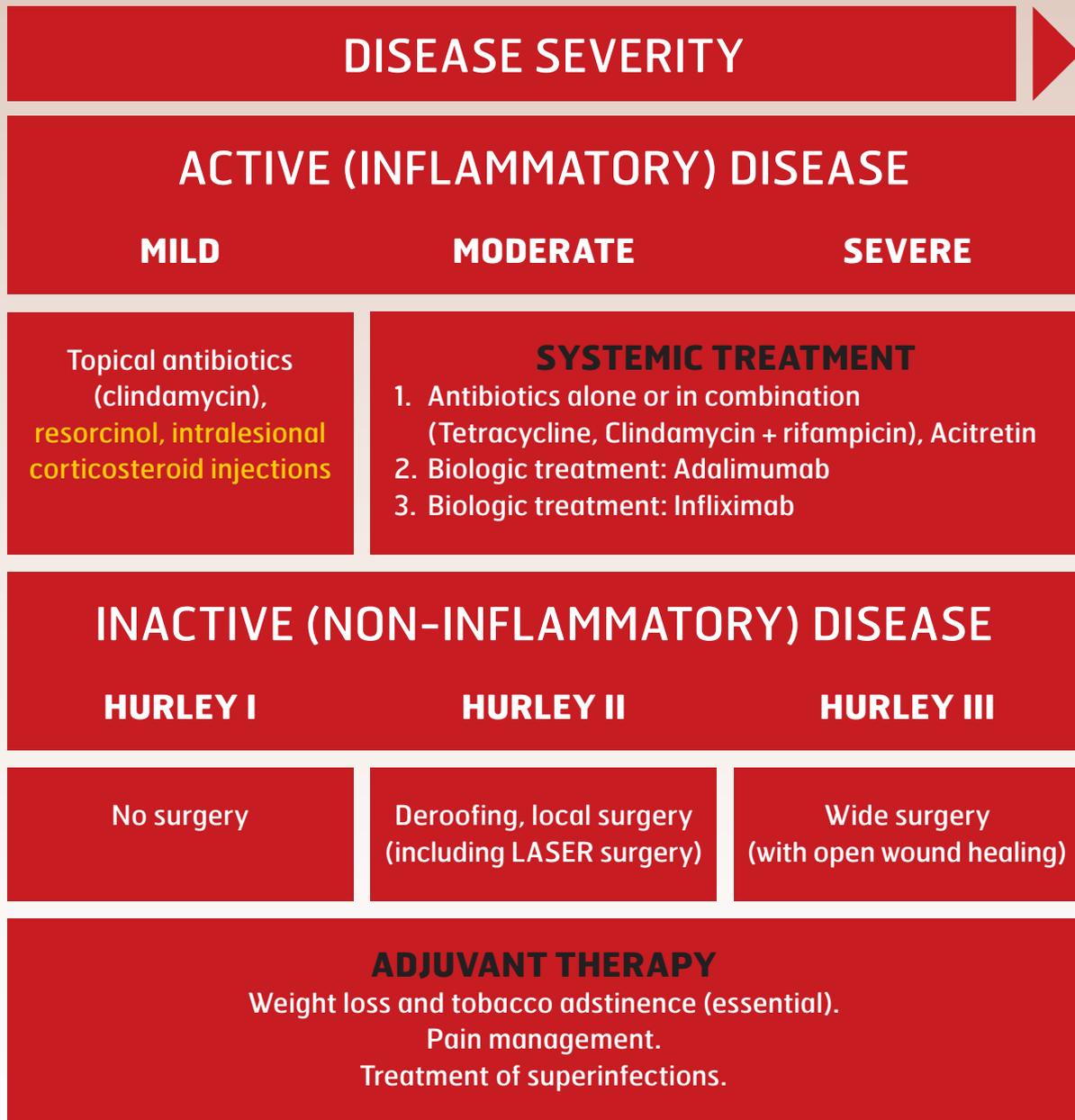


Fig. 2. The HS treatment choices (modified European guideline 2016 – yellow: minor evidence).

Source: Zouboulis CC, Tzellos T, Kyrgidis A, et al. Development and validation of IHS4, a novel dynamic scoring system to assess hidradenitis suppurativa/acne inversa severity. Br J Dermatol 2017;177:1401-1409. (modified).

THINGS YOU CAN DO TO IMPROVE YOUR SITUATION

Find your HS expert

1%-2% of the European population is suffering from HS, but many patients are lost or have given up and turned their back on the health care system: A lot of patients hide their condition and do not go to the doctor – or simply never make it to a dermatologist or a hospital ward, because their family doctor does not recognize or understand the chronic aspects of the disease.

Hygiene has nothing to do with HS

It is important to stress that bad hygiene has nothing to do with HS, even though many have believed so over the years and some still believe so today. HS is first and foremost an immunological disease and is not contagious. It is a myth that has only contributed to the taboo of HS and this cannot be repeated often enough.

Talk to a specialist

It is important to talk to a dermatologist who is specialised in HS. This is the first and most important step in getting the right treatment, which is targeted at the chronic aspects of the disease. During the last few years, new and effective treatments have been developed and approved by the European and US health authorities. As a consequence, many HS patients are now being treated for the chronic aspects of the disease, and are thus more in control of their disease than vice versa.

As a HS patient, it is important that you are referred to a HS expert who understands the complexity of the disease. It is important that you are in close dialogue about different treatment options and how the treatment works for you. If the treatment you are being offered is not helping you, it is not good enough – and you and your dermatologist have to discuss new possibilities.

You are the expert on how HS affects your body and your ability to live your life. The dermatologist is the expert regarding treatment options. Ideally you and your dermatologist have to work together as a team and determine the right treatment, which can enable you to live the life you want to live.

Source: Patient Association HS Denmark / European Reference Network for Rare and Complex Diseases of the Skin.



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 them 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 your 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?

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.

