



PATIENT INFORMATION LEAFLET

SWEET SYNDROME

WHAT ARE THE AIMS OF THIS LEAFLET?

This leaflet has been written to help you understand more about Sweet syndrome. It tells you what it is, what causes it, possible treatments and where you can find out more about this condition.

WHAT IS SWEET SYNDROME?

Sweet syndrome, also called acute febrile neutrophilic dermatosis, is a rare skin disorder. It is characterised by fever and the appearance of tender red or purple lumps or patches on the skin that may turn into ulcers. It is not contagious, not hereditary, and not a form of skin cancer. The condition is classified as a neutrophilic dermatosis - this refers to a group of skin disorders where a type of white blood cell called neutrophils gather in the skin. Neutrophils are responsible for fighting off infections. They work by surrounding and destroying bacteria that enter the body. In Sweet syndrome, neutrophils accumulate in the thick layer of tissue (dermis) that sits just below the outer layer of the skin (epidermis).

WHAT CAUSES SWEET SYNDROME?

Sweet syndrome can affect people of any age, sex, ethnicity and skin type. In about half of cases, the cause is impossible to identify. In the other half of cases, Sweet syndrome most commonly results from upper respiratory tract or gastrointestinal infections, or reactions to medications, such as non-steroidal anti-inflammatory drugs (NSAIDs).

In some people, Sweet syndrome can result from another condition. This can include inflammatory bowel disease, lupus erythematosus or rheumatoid arthritis. In a few cases, it is a result of hormonal changes such as in pregnancy or thyroid gland disorders. More rarely, it may be a sign of cancer such as blood, breast or bowel cancer.

Classical Sweet syndrome in adults affects women more often than men. This female

preponderance has not been seen in cancer-associated or drug-induced Sweet syndrome.

Most likely, the disorder results from multiple, complex factors including immunological, and environmental factors. There is also speculation that Sweet syndrome occurs as an allergic reaction (reactive process) to an unknown agent. In such instances, there is a hypersensitivity reaction by the immune system to a specific agent such as an infection, cancer, or a certain drug.

WHAT DOES SWEET SYNDROME OF THE SKIN LOOK AND FEEL LIKE?

There could be a single or multiple red, pink or purplish and tender skin lumps of different sizes, occurring at any site on the body. The colour may vary across different skin tones; in brown or black skin tones, the patches may have a subtle redness and on close look may appear dark brown, purple, or pinkish in colour. The arms, face and neck are affected most often. Sometimes, blisters or pus-filled blebs (pustules) can be seen, and the rash may appear at the site of an injury to the skin.

In some individuals, neutrophils may build up in the fatty layer of tissue just below the skin (subcutaneous fat) rather than in the dermis. Affected individuals often develop **redness** and small bumps (nodules) on the skin. The arms and legs are most often affected.

Sweet syndrome may cause some or all of the following symptoms arising together over a period of hours or a few days:

- a rash (described above)
- tiredness, lack of energy and feeling unwell
- high fever (temperature)
- aching joints and muscles
- mouth ulcers
- sore red eyes



HOW WILL SWEET SYNDROME BE DIAGNOSED?

Tests that are useful in Sweet syndrome include:

1. **Skin biopsy.** This is an important test. A sample of the abnormal skin may be taken by your dermatologist. The skin sample is then examined under a microscope in the laboratory. It takes time for the sample to be processed and analysed by a pathologist (specialist doctor) so you will not get the result on the day.
2. **Blood tests** such as high levels of a type of white blood cells called neutrophils, can help to make a diagnosis of Sweet syndrome.
3. Your doctor may recommend other blood tests or imaging studies, depending on your symptoms. These may include X-ray or CT scans that look for an underlying cause of the Sweet syndrome, if suspected.
4. If a medication is thought to be responsible, your doctor may recommend stopping it for a while to see if this improves your symptoms.

CAN SWEET SYNDROME BE TREATED?

In some cases, Sweet syndrome may resolve on its own without treatment. This can take weeks to months. Due to the rarity of the condition, there are no treatment trials that have been tested on patients. Various treatments have been reported in the medical literature as part of single case reports or small series of patients. However, the condition can recur (comes back) despite therapy.

Treatments can be divided into topical and oral medications

1. Applications to the skin (topical treatments)
 - **Steroid creams or ointments** may be prescribed, these come in different potencies and strengths. Your doctor will advise you on what is best for your situation. Occasionally, **steroid injections** into the lesions can be used for very swollen or painful lumps.

2. Oral treatments (tablets)
 - **Steroid tablets** (e.g. prednisolone) can be a very effective treatment for Sweet syndrome. They are usually given at a high dose to start with and then reduced slowly over a number of weeks to prevent the rash coming back.
 - Other medications such as **dapsone, colchicine**, potassium iodide or indomethacin can also be very effective and may be preferred by your doctor if you cannot take steroid tablets because of other medical problems such as diabetes or high blood pressure.
 - Immunosuppressive agents such as **ciclosporin** can reduce the action of the body's own defence system (the immune system) and can also be very useful for treating Sweet syndrome.

Some of the above medications can have certain side effects and you may require regular blood tests for monitoring while prescribed these treatments.

If an underlying medical problem is found, treating this itself may lead to an improvement in the Sweet syndrome. Recurrences are possible and, therefore, treatment may need to be reintroduced until the skin lesions settle down. In rare cases, the condition can persist, and treatment would then be given as a maintenance therapy.

DO THE SKIN LESIONS LEAVE ANY SCARS?

The skin lesions associated with Sweet syndrome usually heal without scars, unless open sores (ulcers) were present. The affected area may remain discolored for months after the lesion has been resolved.

WHERE CAN I GET MORE INFORMATION ABOUT SWEET SYNDROME?

Patient support groups providing information:

Sweet Syndrome UK

Web:

<https://helpforsweetssyndromeuk.wordpress.com/>



Weblinks to other relevant sources:

<https://www.dermnetnz.org/topics/acute-febrile-neutrophilic-dermatosis/>

<http://www.healthline.com/health/sweets-syndrome#Overview1>

<http://www.pcds.org.uk/clinical-guidance/sweets-syndrome-syn.-acute-febrile-neutrophilic-dermatosis-gomm-button-dise>

<http://www.patient.co.uk/doctor/febrile-neutrophilic-dermatosis>

Jargon Buster

<https://www.skinhealthinfo.org.uk/support-resources/jargon-buster/>

Please note that the British Association of Dermatologists (BAD) provides web links to additional resources to help people access a range of information about their treatment or skin condition. The views expressed in these external resources may not be shared by the BAD or its members. The BAD has no control of and does not endorse the content of external links.

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

**BRITISH ASSOCIATION OF
DERMATOLOGISTS**
PATIENT INFORMATION LEAFLET

PRODUCED | MAY 2012
UPDATED | APRIL 2015, JULY 2018,
APRIL 2024
NEXT REVIEW DATE | APRIL 2027

