

# Hodgkin's disease

You have just been diagnosed with Hodgkin's disease.

Hodgkin is the name of the physician who first identified this disease in 1862. Hodgkin's disease is a form of lymphoma also known as Hodgkin's lymphoma. A lymphoma is a disease due to the proliferation of immune cells located in the lymph nodes in the lymph, the spleen and the bone marrow.

Pathogens are normally neutralised by immune cells in the lymph nodes.

The occurrence of Hodgkin's lymphoma is due to the uncontrolled proliferation of abnormal B-lymphocytes. These abnormal lymphocytes are called 'Reed-Sternberg cells' and have specific features. Lymphocytes are white blood cells involved in antibody production. They circulate in the blood and lymph vessels. As soon as one of them becomes abnormal, it proliferates as it multiplies faster and lives longer than normal cells. These malignant cells eventually build up into one or more malignant tumours which usually develop within the lymph nodes, most often in the neck, but also in the spleen, the bone marrow or other organs.

The causes of Hodgkin's lymphoma remain unknown. However several risk factors have been identified, such as Epstein Barr viral infections (responsible for mononucleosis or glandular fever), but genetic factors may also account for it. Just like any other cancer Hodgkin's lymphoma is not contagious.

Hodgkin's lymphoma is a rather rare form of cancer. In France about 1,300 new cases are diagnosed each year, that is to say about 0.5% of all new cancer cases. This disease is not age-related, but is more common among young adults and slightly more common among men (54%) than among women. There are two frequency spikes around 30 and after 60 years of age respectively so that the median age at diagnosis is 35 years.

## Medecin's notes :

### First symptoms of the disease

The most common early sign of the disease is the swelling of one or more lymph nodes. They are rarely painful, are located in the neck or the armpits, but may also be felt in other body parts such as the chest and the abdomen in particular. When they are markedly swollen they may cause other troubles, mainly coughing and problems breathing in case of chest lymph node involvement.

Other more general and not so specific symptoms may be experienced, such as unexplained weight loss, profuse night sweats, itching and fatigue.

### Diagnosis

The diagnosis of Hodgkin's lymphoma is formally made from a biopsy which consists in removing a sample of tissue from the enlarged lymph nodes. The cells sampled are then examined under a microscope by a pathologist, that is to say a physician specialised in the study of tissues. The presence of Reed-Sternberg cells allows to confirm the diagnosis of the disease and identify its subtype according to the kind of other cells found around the Reed-Sternberg cells in the diseased lymph node.

Other investigations are usually performed to assess the extent of the disease and its stage. Several imaging techniques are then used to screen for deeper lesions, that is to say lesions that cannot be felt on a clinical examination. As a rule your physician will prescribe a chest Xray and a CT scan of your neck, thorax, abdomen and pelvis. Positron-Emission Tomography (PET scan) is also used to detect any active foci of the disease. Blood tests are also requested to measure markers of disease activity. Depending on each patient's health other investigations may be necessary to make a thorough assessment.

As some drugs may make patients, especially males, sterile, it is possible to preserve some sperm after the disease is diagnosed.

# Hodgkin's disease (suite)

## *Disease stages*

Ann Arbor's classification, set up in 1971 by a group of international experts, allows to distinguish four stages in Hodgkin's disease:

- Stage I : the disease is limited to one lymph node region or to one organ.
- Stage II : the disease is present in several groups of lymph nodes on the same side of the diaphragm (either in the upper or in the lower part of the body).
- Stage III : the disease has spread to several groups of lymph nodes on both sides of the diaphragm (both in the upper and in the lower part of the body)
- Stage IV : the disease has spread to one or more internal organs (the lungs, the liver or bones for example).

Each stage is further divided into two main subtypes, A and B. A refers to symptom-free disease, whereas B means significant symptoms (fever, night sweats, weight loss for instance) are present. In this classification the disease may be said to be IA or IIIB.

**Your treatment may induce adverse effects and carry risks. Your doctor will keep you informed and tell you what symptoms to watch out for before you start on the suggested treatment.**

## *Participating in a clinical trial*

*The best way to contribute to the improvement of disease management is to treat patients in the context of clinical trial. If your doctor suggests this could apply to you, he will explain its purpose, protocol, expected benefits, potential risks and will give you an information leaflet.*

*Participating in a trial of course means you will first have to give your written informed consent.*

## *Useful contacts:*

- **Secretarial / appointment:**
- **Nursing consultation:**
- **Consulting psychologist:**
- **Social worker:**
- **In an emergency:**

## *Management*

In most cases the treatment of Hodgkin's lymphoma rests on multiple drug chemotherapy, sometimes followed with radiotherapy. The type of chemo- and radiotherapy as well as their duration is tailored to the stage of the disease and its risk of progress. Most treatments are administered on an outpatient basis, i.e. without your staying in hospital overnight. As of today, with the treatments available, lasting full remission is possible followed by recovery in over 80% of Hodgkin's disease patients.

In case of a relapse a new treatment plan must be set up. It usually consists of chemotherapy so as to achieve a new remission. In order to improve treatment success rates in case of a relapse intense chemotherapy may be used followed with stem cell transplantation. High dose chemotherapy aims to destroy most if not all cancerous cells. The drawback of this treatment is the sharp drop in normal blood cells (so called aplasia). The body then lacks immune defences against infections. To shorten aplasia time and help normal cells to be quickly restored an autograft is performed; before high dose chemotherapy stem cells likely to produce all blood cells are removed from the patient. These stem cells are retrieved during cytopheresis, a process in which a device is passed through the blood to collect only stem cells which are then frozen. Once intensive treatment is over, the stem cells are thawed and re-injected into the patient so as to replenish his blood cells. This technique calls for several weeks' hospitalisation.

## *Follow-up*

Once remission is achieved after treatment regular follow-up with a specialist is essential. This usually consists of blood tests, series of pictures and a thorough clinical examination in order to monitor remission and, in case signs occur, to make an early detect a relapse as early as possible. Moreover this also enables the physician to suggest some form of management for the likely long-term adverse effects of the treatment. The follow-up is then tailored to each patient.