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*Informationsportal zu Krebserkrankungen bei Kindern und Jugendlichen*

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## **Hodgkin Lymphoma – Brief Information**

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## Table of Content

1. General information .....	3
2. Incidence .....	3
3. Causes .....	4
4. Symptoms .....	4
4.1. General symptoms .....	5
4.2. Specific symptoms .....	5
5. Diagnosis .....	5
5.1. Obtaining a tumour sample (biopsy) .....	6
5.2. Tests to assess spread of the disease (Staging) .....	6
5.3. Tests before treatment begins .....	6
6. Treatment planning .....	7
6.1. Types of Hodgkin Lymphoma .....	8
6.2. Stages of Hodgkin Lymphoma .....	8
7. Treatment .....	9
7.1. Treatment methods .....	9
7.2. Treatment recommendation: classical Hodgkin lymphoma (cHL) .....	10
7.2.1. Chemotherapy .....	10
7.2.2. Radiotherapy .....	11
7.3. Treatment recommendation: Nodular Lymphocyte-Predominant Hodgkin Lymphoma (NLPHL) .....	12
8. Therapy optimising trials and registries .....	12
8.1. Registry GPOH-HD 2020 .....	13
9. Prognosis .....	13
Bibliography .....	15
Glossary .....	18

# Hodgkin Lymphoma – Brief Information

## 1. General information

Hodgkin lymphoma, also known as Morbus Hodgkin, Hodgkin's lymphoma or Hodgkin's Disease, is a cancer of the *lymphatic system*. Like the large entity of Non-Hodgkin-Lymphomas (NHL), it belongs to the group of malignant *lymphomas*. "Malignant lymphoma" literally means „malignant tumour of the lymph node“. Medically speaking, the term summarises all cancers that arise from *cells* of the lymphatic system (*lymphocytes*) and that can cause lymph node swelling.

Malignant lymphomas are classified into two major groups: Hodgkin's lymphoma, which is named after the physician and pathologist Dr. Thomas Hodgkin, and the *Non-Hodgkin lymphomas* (NHL). Differentiation between these two types of lymphomas is only possible by analysing the affected tissue under the microscope (histological examination).

Hodgkin lymphoma, which is named after the physician and pathologist Dr. Thomas Hodgkin, develops from transformed *B lymphocytes* (also known as B cells), a type of white blood cells (leukocytes) found in lymphatic tissue. Hodgkin lymphoma can arise from every organ comprised of lymphatic tissue. The most common location is the *lymph nodes*, however, liver, *bone marrow*, lungs or spleen can also be affected, especially in advanced stages of the disease. Without adequate treatment, Hodgkin lymphoma is a fatal disease in most patients.

**Good to know:** Until recently, the World Health Organization (*WHO*) classified malignant lymphomas into two major groups: Hodgkin lymphomas and the Non-Hodgkin lymphomas (NHL). Meanwhile, the WHO no longer uses the term NHL. Instead, lymphomas are classified according to their cell of origin into B-cell and T-cell lymphomas and, depending on the stage of maturity of the abnormal cells, into precursor-cell lymphomas and mature-cell lymphomas. Hodgkin lymphoma is a mature-cell B-cell lymphoma. Differentiation between Hodgkin lymphoma and other lymphomas is only possible by analysing the affected tissue under the microscope (histological examination).

## 2. Incidence

*Hodgkin lymphoma* is the most frequent lymphoma disease in childhood. According to the German Childhood Cancer Registry in Mainz, about 170 children and teenagers aged younger than 18 years are newly diagnosed with Hodgkin lymphoma in Germany per year. Thus, Hodgkin lymphoma accounts for approximately 7.4 % of all paediatric malignancies.

Hodgkin lymphoma is a rare diagnosis in children younger than 3 years. With increasing age, incidence gets more and more frequent, and in between ages 15 and 20 years, these tumours are most frequent. The incidence in children and adolescents (between 0 and 17 years) peaks at 15 years of age. Boys are slightly more affected than girls (gender ratio: 1.2:1).



### 3. Causes

The causes of Hodgkin lymphoma still have to be elucidated. It is known so far that the disease arises from the malignant transformation of cells in the *lymphatic system*, the B lymphocytes, and also, that this transformation is associated with *genetic* alterations of these cells. Why these genetic alterations exist and why they cause the disease in some children but not in others, is not known yet. Most certainly, Hodgkin lymphoma is caused by a specific combination of many factors.

A clustering of Hodgkin lymphoma patients within the white (Caucasian) population may suggest a genetic and also ethnological predisposition to the disease. Furthermore, children with certain congenital diseases of the *immune system* (such as *Wiskott-Aldrich syndrome*, *Louis-Bar syndrome*) or acquired immune deficiencies (for example due to HIV infection) are known to be at a higher risk to develop a Hodgkin lymphoma. Hereditary concomitant diseases associated with a predisposition to develop cancer are also called *cancer predisposition syndromes*.

In addition, the *Epstein-Barr virus* (EBV), which is well-known as the cause of *glandular fever* (infectious mononucleosis), seems to be associated with the development of Hodgkin lymphoma in some patients. Whether certain environmental factors (such as pesticides) promote Hodgkin lymphoma is currently being examined. However, for the majority of patients, no specific risk factor has been established yet.

### 4. Symptoms

Hodgkin lymphoma begins subtly. First sign (*symptom*) of the disease is usually a very slowly increasing and painless swelling of one or more *lymph nodes*, for example in the regions of the throat or neck (most frequent sites), the armpits, clavicles (in the clavicular groove above the palpable clavicle), groins, or at multiple sites at the same time.

However, the disease can also arise from lymph nodes that cannot be seen or palpated at all, such as those behind the breastbone, in the chest, abdomen or along the spine. Since the cancer is continuously growing, the affected lymph nodes will soon become space occupying, thereby impairing inner organs and their functions. Therefore, enlarged lymph nodes in certain parts of the chest (*mediastinum*) may cause a dry cough or breathing difficulties, while others, in the abdomen for example, can result in diffuse abdominal pain and indigestion.

Enlargement of the spleen and liver (splenomegaly, hepatomegaly) due to lymphoma cell invasion is less frequent. If the lymphocytes in the *bone marrow* are involved, they occupy space within the hollow interior of bones. As a result, Hodgkin lymphoma can cause reduced production of red and white blood cells and, thus, *anaemia* and a predisposition for *infections*. However, these cases are rare.

Nonspecific (general) symptoms may include fever, weight loss, drenching night sweats, fatigue, and itchy skin. The first three symptoms are frequent in patients with Hodgkin lymphoma. They are called *B-symptoms*.

The following overview summarizes the most frequent symptoms caused by Hodgkin lymphoma.



## 4.1. General symptoms

- fever of unknown origin (over 38°C for three consecutive days) [B-symptom]
- night sweats [B-symptom]
- unexplained weight loss (more than 10 % in six consecutive months prior to admission) [B-symptom]
- fatigue, loss of appetite, malaise
- itchy skin

## 4.2. Specific symptoms

- in more than 90 % of patients: painless, palpable, superficial lymph node swellings, for example in the area of the neck (most frequent location), in the armpit, above the clavicle, in the groins or simultaneously at multiple sites
- chronic cough, shortness of breath (if thoracic *lymph nodes*, lungs or pleura are involved)
- abdominal pain, back pain, diarrhea (if abdominal lymph nodes or organs, such as liver or spleen, are involved)
- pallor due to lack of red blood cells (*anaemia*; if the *bone marrow* is involved)
- bone or joint pain (if bones are involved)

Symptoms and complaints usually develop slowly in patients with Hodgkin lymphoma (over weeks or months). They can vary individually as to which symptoms prevail and how pronounced they are.

**Good to know:** The occurrence of one or more of the above-mentioned symptoms does not necessarily mean that they are caused by a Hodgkin lymphoma. Several of these symptoms, such as lymph node swelling and fever, are exactly those often seen with common childhood diseases like common colds and other viral infections. Nevertheless, it is strongly recommended to have the child or teenager see a paediatrician, in particular, if symptoms persist or progress.

## 5. Diagnosis

If the paediatrician thinks that the young patient's history (*anamnesis*), *physical examination* and possibly even results from blood tests and/or imaging, such as *ultrasound* or *X-ray examination*, are suspicious of Hodgkin lymphoma, the child should be referred to a hospital with a childhood cancer program (paediatric oncology program), where further diagnostic tests can be initiated and performed by childhood cancer specialists. These tests serve to confirm or rule out the suspected diagnosis and to assess a possible spread of the disease ("staging").

## 5.1. Obtaining a tumour sample (biopsy)

For diagnosis, surgical removal and investigation of a lymph node or another tissue affected by the disease is required. Apart from *histological* investigation based on how the cells look like under the *microscope*, *immunohistochemical* investigations of these samples allow both the confirmation of the diagnosis and the determination of the type of Hodgkin lymphoma. Knowing the type of Hodgkin lymphoma is both essential for treatment planning as well as for the patient's *prognosis* (see *chapter "Treatment planning"*).

Molecular genetic testing of tissue and, more recently, of blood ('liquid biopsy') may in future become an important (non-invasive) supplementary test for identifying *prognostic factors*, as well as playing a key role in follow-up care. It makes it possible to predict disease relapses at an earlier stage without the need for complex *imaging* procedures.

## 5.2. Tests to assess spread of the disease (Staging)

Once the diagnosis of Hodgkin lymphoma has been confirmed, further tests are required to find out if and to which extent the cancer has spread and which organs are involved. These tests include *imaging*, such as *ultrasound* of the belly (abdomen) and lymph nodes, chest radiographs (*X-ray examination*), *magnetic resonance imaging* (MRI) of the abdomen and pelvis, *computed tomography* (CT), and *positron emission tomography* (PET). Total body-PET is usually combined with a CT scan (PET-CT) and/or an MRI (PET-MRI). Overall, MRI is the preferred imaging procedure, since it is not associated with radiation. However, to check the lungs and/or quickly assess the stage of the disease, CT is required.

Ultrasound scanning of the liver and spleen is also a standard examination and, when used in combination with PET and other imaging techniques (CT and MRI), can be very helpful for staging (assessing the extent of the disease). Since potential bone marrow involvement can now be assessed by PET, *bone marrow punch biopsy* and subsequent screening for lymphoma cells in the obtained bone marrow sample (which until recently was required in advanced stages of the disease) is no longer required.

**Good to know:** PET/MRI and/or PET/CT are essential both for determining the stage of Hodgkin's lymphoma and, later on, for monitoring the disease's response to treatment. The advantage of PET over other imaging techniques is that it detects metabolic activity in the tissue, thereby making living tumour tissue visible throughout the body (*also see chapter "Treatment planning"*).

## 5.3. Tests before treatment begins

For treatment preparation, tests on the patient's cardiac function (*electrocardiography* and *echocardiography*) are performed. Furthermore, additional blood tests are needed to assess the patient's general health condition and to check whether the function of certain organs (such as liver and kidneys) is affected by the disease and whether there are any metabolic disorders to be considered prior or during therapy. Any changes occurring during the course of treatment can be assessed and managed better based on the results of those initial tests, which thus help to keep



the risk of certain treatment-related side effects as low as possible. Also, the patient's *blood group* needs to be determined for the (very) rare case a *blood transfusion* is required during treatment.

**Good to know:** The tests mentioned above are mandatory for every patient. In addition, further tests may be required. Your caregivers will inform you and your child, which diagnostic procedures are individually required in your case and why.

#### *Psychosocial Care*

A child's cancer is a stressful situation for the whole family. The psychosocial team of the clinic or later the aftercare facility provides advice and support to patients and their relatives from diagnosis to completion of treatment as well as during aftercare. Don't hesitate to take advantage of this offer. It is an integral part of the treatment concept of all paediatric oncology centres in many countries. Here you will find comprehensive information on this.

## 6. Treatment planning

After having established the diagnosis, the doctors will plan the treatment. In order to provide a therapy that is specifically designed for the patient's individual situation (risk-adapted therapy), the doctors will take into consideration certain factors that have been shown to have an impact on the *prognosis* (so-called risk factors or prognostic factors).

The following characteristics of Hodgkin lymphoma represent important *prognostic factors* and major criteria for treatment planning:

- **histological characteristics** (meaning the microscopically assessed type of Hodgkin lymphoma): The type of Hodgkin lymphoma decides according to which therapy protocol or therapy optimizing trial the patient will be treated.
- **stage of the disease:** The extent of the disease in- and outside the lymphatic tissue as well as the presence (or absence) of other stage-defining factors (such as *B-symptoms*, elevated *erythrocyte sedimentation rate*, high tumour load) are crucial for assigning a patient to the appropriate treatment level. Three treatment levels are currently being differentiated, considering patients with early, medium and advanced stages of the disease. Treatment intensities differ accordingly. This risk-adapted approach provides a strategy by which also patients with advanced stages of the disease have a chance of cure.
- **response of the disease to chemotherapy:** The speed and extent to which Hodgkin lymphoma responds to treatment are key factors in determining whether *radiotherapy* is required or not. An early, late or generally unsatisfactory response is assessed by means of positron emission tomography and a 5-point scale (Deauville score 1–5).

The following chapters provide information on the histological classification of Hodgkin lymphoma and on the different stages of the disease.

## 6.1. Types of Hodgkin Lymphoma

Hodgkin lymphoma can be divided into different types based on specific morphological and *immunohistochemical* characteristics. Experts refer to this division as a “classification”. The World Health Organization (*WHO*) recognises two forms of Hodgkin lymphoma in children and adolescents (*WHO classification 2022*):

1. Classical Hodgkin lymphoma (cHL)
2. Nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL)

The two types vary regarding incidences, courses of the disease as well as prognosis. The majority of Hodgkin lymphomas are of the classical type. Nodular lymphocyte-predominant Hodgkin lymphoma accounts for around 8–10% of cases. It is considered a separate disease and treated differently than classical Hodgkin lymphoma.

## 6.2. Stages of Hodgkin Lymphoma

Staging of Hodgkin lymphoma is crucial for both treatment planning and estimating prognosis. The stage of the disease is primarily assessed based on its spread at the time of initial diagnosis. It describes which lymph node regions of the body are involved and how many.

Staging also helps to assess whether the disease has spread to organs or tissues outside the lymphatic system (extranodal or extralymphatic disease). Such “extranodal/extralymphatic” involvement occurs in the lungs, liver and skeletal system and is referred to as stage “E” when the lymphoma has spread – from the affected lymph node region(s) – to a single, adjacent non-lymphatic organ or tissue (known as “extranodal spread”).

The stages of Hodgkin lymphoma are classified according to the updated *Ann Arbor staging system* using the terms I through IV as shown in the following table:

### Hodgkin lymphoma stage grouping including E-stages:

Stage of Disease	Definition
Stage I	Lymphoma is found in one single lymph node region (stage 1). It may as well extend to one single extralymphatic organ or site, such as chest wall, heart sac or lung (stage IE).
Stage II	Lymphoma is found in two or more lymph node regions on the same side of the <i>diaphragm</i> (stage II). It may as well extend to a single adjacent extralymphatic organ or site, such as chest wall, heart sac or lung (stage IIE).
Stage III	Lymphoma is found in lymph node regions on both sides of the diaphragm (stage III). It may as well extend to an extralymphatic organ/site (stage IIIE) and/or the spleen (stage IIIES or IIIS, respectively).



Stage of Disease	Definition
Stage IV	Non-contiguous involvement of one or more extralymphatic organs or tissues (such as lungs, liver, bone, <i>bone marrow</i> ) with or without involving (distant) <i>lymph nodes</i>

**Abbreviations:** **E** – refers to a lymphoma that has spread from the affected lymph node region(s) to a single, adjacent non-lymphatic organ or tissue; **S** – spleen, notes cancerous involvement of the spleen.

**Each of the four stages are subgrouped either into the A- or B-category, depending on**

**A:** the absence of general symptoms

**B:** the presence of the following symptoms (*B-symptoms*):

- unexplained body weight loss of 10 % or more in the six months prior to admission and/or
- persistent or recurrent fever (above 38°C) of unknown cause and/or
- drenching night sweats (for example, wet hair, soaked pyjamas)

The presence or absence of B-symptoms is labelled for all stage groups by the suffix B or A, respectively (for example: stage IB or IA).

**Further criteria for stage assessment:** In addition to E-stages and B-symptoms, two further important factors are taken into account in order to determine the stage of the disease and, consequently, the treatment more accurately: elevated *erythrocyte sedimentation rate* ("sed rate"; ESR) and large tumour load ("bulky disease", "bulk").

## 7. Treatment

Treatment of children and adolescents with *Hodgkin lymphoma* should take place in a children's hospital with a paediatric oncology program. Only such a treatment centre provides highly experienced and qualified staff (doctors, nurses and many more) that is specialised and focussed on the diagnostics and treatment of children and teenagers with cancer according to the most advanced treatment concepts. The doctors in these centres collaborate closely with each other. Together, they treat their patients according to treatment plans (protocols) that are continuously optimised. The aim of treating patients with Hodgkin lymphoma is to maintain the well-known high cure rates whilst minimizing acute or long-term side effects as much as possible.

### 7.1. Treatment methods

Treatment options for Hodgkin lymphoma include **chemotherapy**, **radiotherapy** and **high-dose chemotherapy** followed by **stem cell transplantation**.

- Central backbone of current treatment concepts for Hodgkin lymphoma is *chemotherapy* (for special considerations see chapter on *nodular lymphocyte-predominant Hodgkin lymphoma*)



*below*). Chemotherapy uses drugs (so-called cytostatic agents) that can kill fast-dividing cells, such as cancer cells, or inhibit their growth, respectively. Since one cytostatic agent alone may not be capable of destroying all the lymphoma cells, a combination of *cytostatics* that function in different ways is given (polychemotherapy). The goal is to eliminate as many malignant cells as possible.

- For some patients, low-dose *radiotherapy* (radiation therapy) of the affected regions is additionally recommended. Both conventional radiotherapy (with *photons*) and *proton therapy* may be considered. However, in order to reduce radiation-induced late effects, radiotherapy has been used continuously less during the last years. Today, only certain patients receive radiotherapy, for example when their disease does not sufficiently respond to chemotherapy.
- In rare situations, when standard chemotherapy and radiotherapy do not prove sufficiently effective or in the event of recurrent disease (relapse), *high-dose chemotherapy* followed by *stem cell transplantation* (SCT) may be considered as an effective treatment option. The high doses of cytostatics given according to this treatment strategy are capable of eliminating the resistant lymphoma cells. Since high-dose chemotherapy also leads to the destruction of the blood-forming cells in the *bone marrow*, the patient will receive blood-forming stem cells in a second step. Usually, these stem cells are obtained from the patient's blood or bone marrow prior to high-dose chemotherapy and are given back right after this treatment (so-called *autologous stem cell transplantation*).

**Good to know:** The duration and intensity of chemotherapy, as well as the necessity of radiation or stem cell transplantation, and last but not least, the patient's individual *prognosis* is dependent on the extent (stage) of the disease at initial diagnosis and how it responds to treatment. The type of Hodgkin lymphoma (classical HL or NLPHL) plays an important role in treatment planning.

## 7.2. Treatment recommendation: classical Hodgkin lymphoma (cHL)

The following information describes methods and courses of treatment for patients with classical Hodgkin lymphoma. These are based on the current treatment recommendations of the GPOH-HD Study Commission, which in turn are guided by the results of the most recent therapy optimising trial (EuroNet-PHL-C2). Chemo- and radiotherapy are important treatment modalities used in these protocols. If *radiation therapy* is indicated, it is usually performed after cessation of *chemotherapy*. The decision whether radiotherapy is necessary or not is primarily based on the response of the disease to chemotherapy (*please see below*).

### 7.2.1. Chemotherapy

Chemotherapy for patients with classical Hodgkin lymphoma usually consists of multiple treatment cycles or blocks (blocks of *chemotherapy*). The quantity of blocks and, thus, treatment duration and intensity are based on the stage of the disease and the treatment level (TL) they have been assigned to. Usually, patients with

- early stages of the disease (TL 1) receive two or three cycles of chemotherapy,



- intermediate stages of the disease (TL 2) receive four cycles of chemotherapy,
- advanced stages of the disease (TL 3) receive six cycles of chemotherapy.

Every treatment block takes two to four weeks and the different cycles partially contain different combinations of cytostatic agents. For example, "OEPA", a combination of vincristine (oncovin; "O"), etoposide (VP-16; "E"), prednisone ("P") and adriamycin (doxorubicin; "A"), is the current standard for the first two blocks, the so-called "induction therapy".

All other blocks ("consolidation therapy") include either "COPDAC", a combination of cyclophosphamide ("C"), vincristine ("O"), prednisone ("P") and dacarbazine ("DAC"), or "DECOPDAC", a slightly more intensive treatment involving six cytostatics (doxorubicin, etoposide, cyclophosphamide, vincristine, prednisone und dacarbazine). According to recent study findings, the latter can help reduce the need for radiotherapy, particularly in advanced stages of the disease. COPDAC is administered in a 28-day cycle, whilst DECOPDAC is administered in a 21-day cycle.

There are treatment breaks of about two weeks between single blocks. Duration of all chemotherapy is between two to six months if no relapse develops during or after treatment.

## 7.2.2. Radiotherapy

According to current treatment recommendations, less than a quarter of all patients receive *radiation therapy* following chemotherapy. The prime decision-making factor regarding radiotherapy is not the stage of the disease (as it was a while ago), but the response of the disease to chemotherapy. A distinction is made between an early response (following induction therapy) and a late response (following consolidation therapy).

### **Current standard treatment recommendations are:**

- Patients whose disease shows good (adequate) early response after two blocks of chemotherapy (assessed by *positron emission tomography*, PET) do not receive radiotherapy, regardless of the patient's treatment group or stage of the disease.
- Patients whose disease does not sufficiently (not adequately) respond to both the first two blocks of chemotherapy and to subsequent cycles (consolidation therapy) will undergo radiotherapy once chemotherapy has been completed.

**Note:** "Good response" means, that the tumour as found at initial diagnosis now does not contain any live tumour cells any more, thus is PET-negative and also decreased in size for about 50 % of its initial volume (Deauville-Score (DS) 1-3, see *chapter "Treatment planning"*). .

Radiotherapy usually starts about two weeks after cessation of chemotherapy, which is – depending on the patient's treatment level (TL) – after a total of two (TL1), four (TL2) or six (TL3) cycles. Radiotherapy may be directed at the lymph nodes or body regions affected at the time of diagnosis and/or at regions that remain metabolically active (PET-positive) following completion of chemotherapy; the decision in each individual case depends on the stage of the disease and the type of chemotherapy previously administered.

The standard total radiation dose is 20–30 *gray* (Gy), vulnerable organs are treated with lower doses. In order to spare the healthy tissue that is surrounding the cancer, the total radiation dose is not given all at once. Instead, patients receive smaller portions of a maximum of 1.8 Gy per treatment. Duration of radiotherapy comes to two to three weeks total. Radiotherapy is usually not performed over the weekends.

### 7.3. Treatment recommendation: Nodular Lymphocyte-Predominant Hodgkin Lymphoma (NLPHL)

For children and adolescents with nodular lymphocyte-predominant Hodgkin lymphoma, there are certain differences in treatment:

In contrast to patients with classical Hodgkin lymphoma, patients with early stage NLPHL (stage IA) do usually not receive any *chemotherapy* (and *radiotherapy*), as long as only one *lymph node* is affected and can be easily removed completely and without putting the patient in danger or at risk of mutilation (this is very important!). Experience has shown that about two thirds of these patients will defeat their disease without chemo- and radiotherapy. However, regular follow-up examinations are necessary to closely observe the course of the disease (observatory approach). In case of recurrent disease, intensive treatment is recommended.

Patients in stage IA with a residual tumour that is not completely resectable and patients in stage IIA initially receive – also in contrast to classic Hodgkin lymphoma – a mild chemotherapy consisting of three cycles of cyclophosphamide, vinblastine and prednisolone (CVP for short). The necessity of additional treatment measures depends on how well the disease responds to this chemotherapy. In case of unfavourable treatment response, two further courses of chemotherapy (an OEPA regimen comprising vincristine, etoposide, prednisone and adriamycin (=doxorubicin)) are given. If the response remains inadequate, additional radiotherapy is administered to the areas of the body affected at the time of diagnosis (19.8 Gy).

Treatment as for classical Hodgkin lymphoma is recommended only for those with advanced disease, meaning higher stages (III-IV) of NLPHL: Following induction therapy (with two cycles of OEPA), two or four cycles of COPDAC-28 are administered; in advanced cases, the DECOPDAC-21 combination may be used, too (*see chapter "Treatment recommendations: Classical Hodgkin lymphoma"*). In individual cases, the monoclonal antibody rituximab may be used as an adjunct to standard therapy [see *monoclonal antibodies*]. If the response to chemotherapy is poor (early and, in particular, late), radiotherapy is recommended for consolidation.

**Good to know:** The majority (over 80–85%) of patients with lymphocyte-predominant Hodgkin lymphoma are diagnosed at stage IA or IIA.

## 8. Therapy optimising trials and registries

In Germany, treatment of almost all children and adolescents with Hodgkin lymphoma is performed according to the treatment plans (protocols) of *therapy optimising trials* or registries. The term “therapy optimising trial” refers to a form of controlled clinical trial that aims at improving current

treatment concepts for sick patients based on the current scientific knowledge. With many treatment centres being involved in this kind of standardised treatment, such studies are also called “multicentred” and “cooperative”, and most often many countries participate.

Patients who cannot participate in any study, for example because none is available or open for them at that time, or since they do not meet the required inclusion criteria, respectively, are often included in a so-called **registry**. The patients are generally treated according to the recommendations of the trial centre, thus receiving the current best therapy available.

Two major international trials were completed some time ago: the EuroNet-PHL-C2 trial for patients with newly diagnosed classical Hodgkin lymphoma and the EuroNet-PHL-LP1 trial for patients with early-stage lymphocyte-predominant Hodgkin lymphoma. There are currently no ongoing therapy optimising trials in Germany. However, all patients can be registered in the GPOH-HD 2020 registry.

## 8.1. Registry GPOH-HD 2020

The international registry GPOH-HD 2020 of the Hodgkin Study Group was opened October 1, 2020 with the aim to ensure optimal treatment of children and adolescents with Hodgkin lymphoma also during times without active clinical studies. Eligibility includes patients with first diagnosis or relapse of a classic Hodgkin lymphoma (cHL) or nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL). Patients from previous studies are eligible to register with the registry as well. The study centre provides therapy recommendations that are based on the results of the most recent active trials, EuroNet-PHL-C2 and EuroNet-PHL-LP1, respectively (see *chapter "Treatment"*).

The international and German study centre for the above-mentioned registry is located at the “Zentrum für Kinderheilkunde und Jugendmedizin der Universitätsklinik Gießen” (Department of Paediatrics, University of Gießen, Germany). The Principal Investigator is Prof. Dr. med. Christine Mauz-Körholz.

## 9. Prognosis

### Prognosis for newly diagnosed patients

Nowadays, long-term survival rates of children and teenagers after treatment of Hodgkin lymphoma are high: More than 9 out of 10 (99 %) patients can be cured today, thanks to current modern diagnostics and standardised treatment concepts – and regardless of how advanced the disease had been at diagnosis.

This is only possible due to the current treatment strategies that adjust the intensity of therapy (number of *chemotherapy* cycles, *radiotherapy*) to the patient’s individual situation by considering different treatment groups. Patients with more advanced stages of the disease (treatment levels II and III) need a more intensive therapy than patients presenting with earlier stages (treatment level I) in order to provide a comparably favourable *prognosis*.

### Prognosis for patients with recurrent disease



In about 5–10 % of patients aged younger than 18 years the disease does either not respond to current treatment strategies (progressive disease) or the patients later develop recurrent disease (*recurrence*, relapse). In general, favourable long-term outcomes can be achieved for patients with relapsed Hodgkin lymphoma, too. Individual prognosis, however, depends primarily upon the timepoint of relapse and how intense primary treatment has been.

Patients with late relapse (more than one year after completion of therapy), who receive a second chemo- and radiotherapy for relapse treatment, have high survival rates (10-year survival of more than 90%). This applies in particular to patients with early stage of the disease at primary diagnosis (treatment group 1) and/or those who have not received any radiotherapy as part of initial treatment.

Until now, patients with early relapse (between three and twelve months after cessation of primary treatment) and patients with non-response or progressive disease have had a less favourable outcome with conventional chemo- and radiotherapy for second therapy. Previous trials reported 10-year survival rates of approximately 75 % and 50 % respectively.

Nowadays, even in these patients (in standard relapse situations or even with tumour progression), very high response rates can be achieved by means of intensive chemotherapy and/or new treatment options (such as *immunotherapy* and targeted *antibody* therapies), so that subsequent *high-dose chemotherapy* and *autologous stem cell transplantation* offer good prospects of cure (in the range of 80–90%). In patients with so-called low-risk relapses, equally high cure rates can be achieved by immunotherapy followed by consolidation radiotherapy (provided that *remission* has been achieved through immunotherapy).

**Note:** The survival rates mentioned in the text above are statistical values. Therefore, they only provide information on the total cohort of patients with Hodgkin lymphoma. They do not predict individual outcomes. However, statistics help to estimate probabilities of survival.

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# Glossary

anaemia	„lack of blood“; reduction of the red blood pigment (haemoglobin) and/or the proportion of red blood cells (haematocrit) in the blood below the normal value typical for a given age. Signs of anaemia include palor, headache, dizziness, and fatigue.
anamnesis	medical interview, a patient's history, development of signs of illness; the type, onset and course of the (current) symptoms as well as any risk factors (e.g. hereditary diseases) are evaluated during a medical interview.
Ann Arbor staging system	system for staging malignant lymphomas, especially Hodgkin lymphomas and certain forms of Non-Hodgkin lymphomas
antibody	Antibodies are proteins from the group of globulins, which the bodys immune system forms as a defensive reaction to invading foreign substances or foreign structures (antigens). The antibodies bind specifically to these antigens and lead (in various ways) to the elimination of the pathogen. Antibodies are produced by a group of white blood cells, so-called B lymphocytes, which are known as "plasma cells" at the stage of antibody production.
autologous stem cell transplantation	(re)transfer of blood stem cells, e.g. after a chemotherapy or radiotherapy; the patient receives his own cells that were previously taken from their own bone marrow or blood. Autologous stem cell transplantation may be an option, for example, for certain patients with lymphoma, neuroblastoma, soft tissue sarcoma, or a brain tumour.
B lymphocytes	subtype of lymphocytes; they develop in the bone marrow and are responsible for the recognition of pathogens and the formation of antibodies.
B-symptoms	certain non-specific symptoms that often occur simultaneously in cancer patients: recurrent fever (above 38°C) for no apparent reason, night sweats and unintentional weight loss (more than 10% of body weight in six months). The three symptoms are grouped together under the term B symptomatology. In this combination, they occur primarily in patients with Hodgkin lymphoma and Non-Hodgkin lymphoma and are associated with an unfavourable prognosis.
blood group	hereditary, usually stable, structural characteristics (blood group antigens) of blood components (e.g. AB0 blood groups) located on the cell walls of blood and other tissue cells;



blood transfusion	transfer of blood (whole blood) or blood components (e.g. red blood cells or platelets) from a donor to a recipient
bone marrow	site of blood formation; spongy tissue with a strong blood supply that fills the cavities inside many bones (e.g., vertebrae, pelvic and thigh bones, ribs, sternum, shoulder blade, and collarbone); in the bone marrow, all forms of blood cells develop from blood progenitor cells (blood stem cells).
bone marrow punch biopsy	removal of bone marrow tissue for the purpose of examining the cells; with the help of a special hollow needle, a tissue cylinder about 2 cm long is punched out of the bone. The examination is always carried out under anesthesia. A bone marrow punch biopsy may be necessary in addition to or instead of a bone marrow puncture if the latter does not provide sufficient tissue for a reliable examination. Like the bone marrow puncture, it is usually performed from the posterior iliac crest bone. There, the bone marrow is only separated from the skin by a relatively thin layer of bone, so that the removal can take place without significant risk.
cancer predisposition syndrome	genetic disorders that can include malformations and intellectual disability in addition to an increased risk of tumors; according to current knowledge, about 10% of childhood and adolescent cancers develop due to a known hereditary change or cancer predisposition syndrome. Cancer predisposition syndromes include Louis Bar syndrome (= ataxia telangiectatica), Beckwith-Wiedemann syndrome, Down syndrome, Hippel-Lindau syndrome, Li-Fraumeni syndrome, MEN syndrome, neurofibromatosis and WAGR syndrome. The familial form of retinoblastoma is also part of it.
cell	the smallest functional unit in organisms with the ability to perform metabolism, involuntary muscle movement and reproduction and to respond to stimuli; each cell contains a nucleus and a cell body (cytoplasm) and is externally bounded by the cell membrane.
chemotherapy	here: use of drugs (chemotherapeutic agents, cytostatics) for the specific inhibition of tumor cells in the organism
computed tomography	imaging, X-ray diagnostic procedure; it produces an image by computer-controlled evaluation of a large number of X-rays from different directions. This makes it possible to produce sliced images of body parts (tomograms, transverse or longitudinal sections of the human body)
cytostatics	drugs that inhibit cell growth; cytostatics can affect the metabolism of different types of cells, thereby destroying them and/or



	preventing them from multiplying. Cells that divide frequently are particularly affected.
diaphragm	a thin muscle below the lungs; separates the chest and abdominal cavities and supports breathing
echocardiography	ultrasound examination of the heart to check its performance (cardiac function); the position or structure of the heart valves and walls, the wall thickness of the heart muscle, the size of the heart and the ejected blood volume (pumping function of the heart) are examined and assessed, among other things.
electrocardiography	method of measuring the electrical activity of the heart
Epstein-Barr virus	causative agent of glandular fever;
erythrocyte sedimentation rate	determines how quickly the red blood cells (erythrocytes) sink within one hour (sometimes additionally within two hours) in a special measuring tube; for example, the test can indicate inflammation in the body. In certain diseases, erythrocytes settle in the blood either more slowly or faster than usual.
genetic	concerning the (level of) inheritance or genes; inherited
glandular fever	common, often harmless viral disease that occurs mainly in children and young adults; is caused by the Epstein-Barr virus (EBV) and affects the lymphatic tissue (e.g. lymph nodes, spleen). Pfeiffers glandular fever is associated with characteristic changes in blood counts (conspicuous increase in white blood cells; leukocytosis).
gray	unit of measurement for the dose of energy caused by ionising radiation (e.g. in the context of radiotherapy) and absorbed by a given mass (kilogram of body weight)
high-dose chemotherapy	the use of a particularly high dose of cell growth-inhibiting drugs (cytostatics); in the case of cancer, it aims to destroy all malignant cells. Since the haematopoietic system in the bone marrow is also destroyed, the patients own or foreign blood stem cells must then be transferred (autologous or allogeneic stem cell transplantation).
histological	concerning the tissues of the body; in a histological (fine tissue) examination, tissue samples are examined under the microscope after special preparation (preparation of tissue sections and use of certain staining techniques).



Hodgkin lymphoma	malignant disease of the lymphatic system; belongs to the malignant lymphomas and accounts for about 5% of malignant diseases in childhood and adolescence.
imaging	diagnostic procedures generating images of the inside of the body, such as ultrasound and X-ray examination, computed tomography, magnetic resonance imaging, and scintigraphy
immune system	the bodys own system for maintaining a healthy organism by defending against foreign substances and destroying abnormal body cells (e.g. cancer cells); the immune system has the ability to distinguish between self and foreign or dangerous and harmless; mainly the organs of the lymphatic system as well as cells distributed throughout the body (e.g. leukocytes) and molecules (e.g. immunoglobulins) are involved.
immunohistochemical	in an immunohistochemical or immunohistological examination, proteins or other cell or tissue structures are visualized with the help of labeled antibodies (e.g. bound to dyes).
immunotherapy	a form of treatment that affects the immune system with the aim of fighting off or fighting tumours or other diseases
infection	penetration of the smallest organisms (e.g. bacteria, viruses, fungi) into the body and subsequent multiplication within it. Depending on the characteristics of the microorganisms and the immune system of the infected person, various infectious diseases can occur after infections.
Louis-Bar syndrome	hereditary disease; it is mainly characterized by degeneration of the central nervous system (CNS), an impairment of the immune system (immunodeficiency), dilated blood vessels of the eyes and skin (so-called telangiectasias) and an increased risk of cancer (so-called cancer predisposition syndrome). Degeneration of the CNS is associated with various neurological disorders, such as movement disorders (ataxia) and abnormal eye movements. The immunodeficiency often causes recurrent infections.
lymph nodes	small lenticular to bean-shaped organs that are part of the bodys immune system and are located in many parts of the body; they serve as filter stations for the tissue water (lymph) of a region of the body and contain cells of the immune system.
lymphatic system	collective term for lymphatic vessels, lymphatic vessel trunks, lymph nodes, lymphatic tissues (lymphocytes in connective tissue, mucous membranes, glands) and lymphatic organs (spleen, pharyngeal tonsils, bone marrow, thymus gland).



lymphocytes	subgroup of white blood cells that are responsible for the body's own defenses, especially the defense against viruses; there are B and T lymphocytes. They are formed in the bone marrow, but partly only mature to full functionality in the lymphatic tissue (e.g. lymph nodes, spleen, thymus gland). They eventually enter the blood via the lymphatic vessels, where they take over their respective tasks.
lymphoma	collective term for lymph node enlargement of various causes
magnetic resonance imaging	diagnostic imaging method; very precise, radiation-free examination method for the visualization of structures inside the body; with the help of magnetic fields, cross-sectional images of the body are generated, which usually allow a very good assessment of the organs and many organ changes.
mediastinum	middle section of the thoracic cavity located between the two lungs
microscope	an instrument that allows you to magnify objects or certain structures of objects that are not visible to the human eye
monoclonal antibodies	antibodies produced by the derivatives of a single B lymphocyte (cell clone) that are completely identical; they can be genetically engineered for diagnostic and therapeutic purposes and target a small molecular segment (epitope) of a specific antigen.
Non-Hodgkin lymphoma	a large group of malignant diseases of the lymphatic system, which can provoke lymph node swelling as a main feature; like Hodgkin lymphoma, NHL is a malignant lymphoma. It accounts for about 7 % of malignant diseases in childhood and adolescence.
photon	from ancient Greek light; smallest unit of electromagnetic radiation; each photon transports energy.
physical examination	an important part of diagnostic examinations; includes palpation and listening to certain body organs as well as testing reflexes to obtain indications of the nature or course of a disease.
positron emission tomography	an imaging, nuclear medicine procedure based on the principle of scintigraphy, which can be used in cancer medicine to visualize tumours or metastases. To detect tumour tissue, a radioactively labeled sugar compound is administered. Since tumours have a higher metabolism than healthy tissue, the radioactive substance is increasingly taken up and stored by the tumour cells. The tumour cells enriched with this substance emit signals that are



	captured by a special camera (PET scanner) and converted into an image (tomogram).
prognosis	prediction of the course and outcome of a disease / prospect of recovery
prognostic factors	factors that allow an approximate assessment of the further course of the disease (i.e. the prognosis);
proton therapy	modern form of radiotherapy using protons for the treatment of malignant tumours; compared to conventional radiotherapy with photons, this type of radiation can specifically target the tumour area, thereby sparing adjacent, healthy tissue from the effects of radiation.
radiotherapy	controlled use of ionizing (high-energy) radiation for the treatment of malignant diseases
recurrence	relapse, recurrence of a disease after recovery
remission	temporary or permanent decrease or disappearance of the signs of cancer.
stem cell transplantation	transfer of haematopoietic stem cells after conditioning chemotherapy, radiation or immunosuppression of the recipient; the stem cells can be obtained either from the bone marrow or from the bloodstream. In the first case, the procedure of their transfer is called bone marrow transplantation, in the second case peripheral stem cell transplantation. Depending on the type of donor, a distinction is made between two forms of SCT: allogeneic SCT (stem cells from a foreign donor) and autologous SCT (own stem cells).
symptom	sign of illness
therapy optimising trial	a controlled clinical trial (study) that aims to provide the best possible treatment for patients and at the same time to improve and develop treatment options; therapy optimisation is aimed not only at improving the chances of recovery, but also at limiting treatment-related side effects and long-term effects.
ultrasound	an imaging technique used to examine organs, in which ultrasound waves are sent through the skin into the body; at tissue and organ boundaries, the sound waves are reflected back, picked up by a receiver (transducer) and converted into corresponding images with the help of a computer.



WHO	abbreviation for World Health Organization; international federation for cooperation in the field of public health
WHO classification	international guideline developed by the World Health Organization (WHO) for classification, diagnosis and differentiation/grading of (malignant) diseases
Wiskott-Aldrich syndrome	congenital disease with coagulation disorder and immunodeficiency; typical features include skin bleeding, increased susceptibility to infections, eczema-like skin lesions and a tendency to allergic reactions as well as an increased risk of cancer. Wiskott-Aldrich syndrome is, therefore, also one of the cancer predisposition syndromes (CPS).
X-ray examination	imaging procedure that uses X-rays to visualize organs or parts of organs