



**kinderkrebsinfo**

Informationsportal zu Krebserkrankungen bei Kindern und Jugendlichen

## **Pineoblastoma – Brief information**

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# Pineoblastoma – Brief information

## 1. General information on the disease

A pineoblastoma is a malignant brain *tumour*. It arises from cells of the *pineal gland* (Glandula pinealis), a small endocrine organ in the *diencephalon*, which is located in the centre of the brain. Since pineoblastoma directly originate from the *central nervous system* (CNS), they are also called *primary CNS tumours*, thereby differentiating them from malignant tumours of other organs that have spread (metastasised) to the CNS.

Pineoblastoma originate from extremely immature (*undifferentiated, embryonal*) cells of the central nervous system, which divide at a high rate. Therefore, these tumours grow very fast. Due to their aggressive growth behaviour, they are defined as high-grade malignant tumours.

Pineoblastoma may spread from the area of the pineal gland into other regions of the brain and *spinal cord* as well. Metastasis outside the CNS, for instance to bones, *bone marrow*, lung, or *lymph nodes*, is rare.

In the past, many authors considered pineoblastomas and embryonal, non-rhabdoid CNS tumours – due to their rarity and similarity regarding the course of the disease – as one group. Today, however, it is well-known that a pineoblastoma differs substantially from other embryonal CNS tumour on the *molecular* level and, therefore, needs to be dealt with as an independent tumour type.

## 2. Incidence

Pineoblastomas are very rare: with about three or four newly diagnosed patients under 18 years of age per year, they account for less than 1 % of all CNS tumours in childhood and adolescence in Germany. Pineoblastoma is mostly diagnosed in children and young adults. The patients' average age at diagnosis is approximately 18 years.

## 3. Causes

A pineoblastoma is caused by a malignant transformation of *nerve tissue* cells. The reasons for tumour development have not been completely found out yet. It is well-known, though, that *radiotherapy* of the brain, for example as received by children with certain forms of *leukaemia* or with eye cancer (*retinoblastoma*), leads to an increased risk of developing a CNS tumour later in life.

In addition, it has been shown that pineoblastoma are sometimes associated with certain *genetic* and *chromosomal* abnormalities in the tumour cells. The resulting impairments of cell development and cell communication may be contributing factors promoting the transformation of a healthy into a cancer cell. However, since pineoblastomas are rare, only a few *molecular* abnormalities that might be responsible for causing the disease have been identified yet.



**Good to know:** rarely, pineoblastoma can be associated with hereditary retinoblastoma and thus genetic alterations of the *retinoblastoma gene* (so-called trilateral retinoblastoma). More information on trilateral retinoblastoma can be found in our [Retinoblastoma](#) chapter.

## 4. Symptoms

Due to the uncontrolled and aggressive growth pattern of pineoblastoma, *symptoms* typically develop and deteriorate fast. The presenting symptoms of this tumour (like other tumours of the *central nervous system*) primarily depend on the patient's age, the site and size of the tumour as well as its pattern of spread within the CNS. The following general (nonspecific) and local (specific) symptoms can occur:

### 4.1. General (nonspecific) symptoms

Unspecific general symptoms occur independently of the tumour's location. They may be similar to and therefore mimic other, non-CNS diseases. General symptoms of a child or adolescent with a CNS tumour may include headaches and/or back pain, dizziness, loss of appetite, nausea and vomiting (particularly after getting up in the morning), weight loss, increasing fatigue, inability to concentrate, school problems, mood swings, and character changes as well as developmental delay, to name a few.

Major reason for these symptoms is the slowly but continuously increasing intracranial pressure (ICP). An elevated intracranial pressure may be caused by the growing, thus more and more space-occupying tumour within the bony skull. It may as well be due to the tumour blocking the regular flow of the *cerebrospinal fluid*, thereby forming *hydrocephalus*. In babies or small children with soft spots (open *fontanelles*), elevated intracranial pressure and hydrocephalus typically present with a bulging fontanelle or a larger than expected head circumference (*macrocephalus*), respectively.

### 4.2. Local (specific) symptoms

Local symptoms may indicate the tumour location and, thus, which functional regions of the CNS might be affected. Pineoblastoma, for example, can cause trouble with eye movements, in particular limitations of upgaze. This vision impairment results from the tumour's specific location in the *diencephalon* and is also known as Parinaud syndrome. Furthermore, a tumour in the diencephalon or in the hemispheres of the *cerebrum* can be associated with *seizures* and/or motor deficits. Also, vision impairments, speech disorders, behavioural and sleep problems, as well as moodiness and altered appetite regulation may, although to a lesser extent, be indicative of tumour location.

**Good to know:** Not all patients presenting with one or more of the symptoms mentioned above do have a pineoblastoma or another type of brain tumour. Many of these symptoms may also occur with other, harmless diseases that are not associated with a brain tumour at all. However, if certain symptoms persist or get worse (for example repetitive headaches or rapid increase of head circumference in a young child), a doctor should be seen to find the underlying reason. In case it is a brain tumour, treatment should be started as soon as possible.

## 5. Diagnosis

If the paediatrician thinks that the young patient's history (*anamnesis*) and *physical examination* are suspicious of a tumour of the central nervous system (CNS), the child should immediately be referred to a hospital with a childhood cancer program (paediatric oncology unit), where further diagnostics can be initiated and performed by childhood cancer professionals. Very close collaboration between various specialists (such as *paediatric oncologists*, paediatric neurosurgeons, paediatric *radiologists*, to name a few) is required, both to find out, whether the patient really suffers from a malignant CNS tumour and, if so, to determine the tumour type and the extension of the disease. Knowing these details is absolutely essential for optimal treatment and *prognosis*.

### 5.1. Tests to secure diagnosis

The initial diagnostic procedures for a young patient presenting with a suspected CNS tumour at a childhood cancer centre include another assessment of the patient's history, a thorough physical/ *neurological* exam and *imaging* diagnostics, such as *magnetic resonance imaging* (MRI) or (less often) *computed tomography* (CT). These tests help find out exactly whether the patient has a tumour of the central nervous system. Also, localisation and extent of the tumor, its demarcation regarding adjacent tissue as well as a potential *hydrocephalus* can be diagnosed by these imaging techniques very well.

In order to validate the final diagnosis, *histological* and *molecular* analysis of surgically obtained tumour tissue (*biopsy*) is required. Usually, this is done using the tissue obtained during surgical tumour removal.

The extent of histological and, especially, *molecular genetic* workup has been substantially increased over the past years. Today's option of using modern lab techniques makes it possible to identify molecular tissue characteristics that do not only help finalize the diagnosis, but can also provide information on what to expect regarding the course of the disease (such as growth behaviour).

### 5.2. Tests to assess spread of disease

Once the diagnosis of a pineoblastoma has been confirmed, additional tests are required to assess the extent of the disease within the central nervous system (CNS). Apart from *MRI* scans of the complete CNS (brain and spine) for macroscopic metastases, these tests also include microscopic checking of the *cerebrospinal fluid* (CSF) for tumour cells in the *spinal cord* (which are not visible by MRI scan). Cerebrospinal fluid is mostly obtained from the spine in the lower back (*lumbar puncture*), since the risk of the puncture needle damaging the spinal cord is lowest at the lower back level.

### 5.3. Tests before treatment begins

In preparation for the intensive treatment of the brain tumour, further investigations are performed, such as *electrocardiography* (ECG) and *echocardiography* to check cardio function. 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.

The condition and function of the hormonal glands will be checked in order to detect and manage potential tumour- and/or treatment-associated endocrinological impairments as early as possible. For the same reason, neuropsychological testing [see *neuropsychology*] might be done prior to cancer treatment. 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 in case a *blood transfusion* is required during treatment. In sexually mature females (which means after they have experienced their first menstruation), a pregnancy test is recommended prior to treatment as well.

**Good to know:** Not every patient needs the complete check-up. On the other hand, tests might be added that haven't been mentioned here, depending on the individual situation of the patient. Your caregivers will inform you and your child, which diagnostic procedures are individually required in your child's situation 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

Once the diagnosis and extent of a CNS tumour has been confirmed, treatment planning starts. In order to provide the patient with the best possible individual and risk-adapted therapy, the treatment team considers specific factors that are known to have an impact on the *prognosis* (so-called prognostic factors).

Important *prognostic factors* in case of a pineoblastoma are the type, localization, extent and spread of the tumour. Also, the biological (molecular) features of a tumour increasingly impact the choice of optimal treatment. In addition, the patient's age and overall physical condition play an important role. Age at diagnosis, above all, determines whether the patient may receive radiotherapy or not. All these factors are included in treatment planning in order to achieve the best outcome possible for each patient.

### 6.1. Classification of tumours of the pinealis region

Pineoblastoma belongs to the group of so-called pineal tumours. Various types of tumour can arise in the *pineal gland*, which differ from each other both in terms of their appearance under the *microscope* (i.e. *histologically*) and in terms of their *molecular* tissue properties. Sometimes they



are also associated with different *prognosis* for patients. Pineoblastoma, the most frequent tumour of the pineal region, is defined as a high-grade malignant tumour by the World Health Organization (WHO).

According to the current *WHO classification* for tumours of the central nervous system (WHO classification 2021), pineal tumours are differentiated as follows:

- Pineoblastoma (CNS WHO grade 4)
- Papillary tumour of the pineal region (PTPR) (CNS WHO grade 2 or 3)
- Pineal parenchymal tumour of intermediate differentiation (CNS WHO grade 2 or 3)

Pineal tumours other than pineoblastoma are less malignant (CNS WHO grade 2 or 3), but very rare in childhood and adolescence.

## 7. Treatment

Treatment of children and adolescents with pineoblastoma should take place in a children's hospital with a paediatric oncology program. Only in such a childhood cancer centre, highly experienced and qualified staff (doctors, nurses and many more) is guaranteed, since they are specialized and focus 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 goal of the treatment is to achieve higher cure rates while avoiding side effects as much as possible.

Current treatment concepts involve neurosurgical **tumour removal**, **chemotherapy** and, depending on the patient's age, **radiotherapy**. In selected patients, **high-dose chemotherapy** followed by **autologous stem cell transplantation** may also be an option.

Pineoblastomas are very rare diseases and their therapies are constantly being further developed. The treatment options presented below are based on recommendations from the study/registry centre, however, these must be discussed individually in each case. They do not claim to be exhaustive at this point. The exact course of treatment for individual patients will be decided by the attending physician in consultation with the patient or their relatives.

### 7.1. Surgery

The first step in treating a pineoblastoma is *surgery*. Goal of surgery is gross (microsurgical) total tumour removal. This means that at the end of the surgical procedure, no tumour tissue can be identified through the surgical microscope. Frequently, however, total tumour resection cannot be achieved due to the localization of these tumours.

In most patients, neurosurgical intervention results in normalising the flow of *cerebrospinal fluid* (CSF). Patients initially presenting with *hydrocephalus* may need a transient hydrocephalus **drainage prior to** tumour removal or, sometimes, even a permanent drainage system later.

## 7.2. Additional, non-surgical treatment

Since pineoblastomas tend to infiltrate adjacent tissue and, furthermore, often spread into other parts of the central nervous system via the cerebrovascular fluid (CSF), treating the tumour locally only is not sufficient. Therefore, surgery is followed by additional non-surgical treatment, comprising *radiotherapy* and/or *chemotherapy*.

Chemotherapy uses drugs (so-called cytostatic agents or *cytostatics*) that can kill fast-dividing cells, such as cancer cells, or inhibit their growth, respectively. In order to eliminate as many of the cancer cells as possible, a combination of several cytostatics is usually applied. Frequently-used agents are, for example, carboplatin, etoposide, methotrexate, vincristine, cyclophosphamide, lomustine, cisplatin and/or temozolomide.

Radiotherapy is done using energy-rich, *electromagnetic* radiation, given through the skin to the tumour region. Radiation causes *DNA* damage in tumour cells, thereby leading to cell death. Modern techniques, such as *intensity-modulated radiotherapy* (IMRT), help minimise radiation damages in healthy tissue. Instead of conventional radiotherapy with *photons*, particle-radiation with *protons* (*proton therapy*) can be an option in suitable cases as well. This type of radiotherapy allows to reduce the effects of radiation in healthy tissue even better and is, therefore, gaining an increasing importance in the treatment of children and teenagers with *solid tumours*.

Decision upon which therapy is to be applied (treatment modalities, intensity of chemo-/radiotherapy) is based on the patient's age, the histological and molecular subtype of the tumour, certain biological risk factors, as well as on the extent of both metastases and surgical tumour removal (see chapter "Therapy planning").

Possible treatment strategies are as described below.

### 7.2.1. Treatment options for pineoblastoma patients

After maximal possible tumour removal, patients with non-metastasised pineoblastoma, who are older than four years of age, will receive *radiation* of the complete central nervous system (craniospinal radiotherapy), followed by an additional boost to the tumour site. Radiotherapy is followed by a so-called maintenance *chemotherapy*, which includes multiple cytostatic agents. In cases of *metastasis*, treatment will be intensified, for example by giving higher doses of radiotherapy combined with a preceding chemotherapy (induction chemotherapy).

In children under four years of age, radiotherapy should be avoided or delayed in order to minimize the risk of serious late effects. Instead of radiotherapy after surgery, patients will receive chemotherapy with multiple agents. In some patients, radiotherapy may be an option later on. Some patients are also eligible for *high-dose chemotherapy* followed by *autologous stem cell transplantation* to increase the chances of survival.

There are no standard therapies for patients who develop a papillary tumour of the pineal region or a pineal parenchymal tumour with intermediate differentiation in childhood and adolescence (see chapter "Treatment planning – Classification"). These tumour types are extremely rare diseases. Therapeutic strategies must be confirmed on an individual basis.





**Important to know:** The details of how an individual patient is treated will be discussed between the responsible physician, the patient and the family.

## 8. Therapy optimising trials and registries

In Germany, children and adolescents with pineoblastoma and relapses (*recurrence*) of this tumour are generally treated individually and monitored in the framework of registry studies. Where available, participation in *therapy optimising trials* or early phase studies (phase I/II studies, in cooperation with the pharmaceutical industry) is considered together with the patients and/or their relatives. There are various early phase networks in Germany for finding suitable early phase studies.

Therapy optimising trials are standardised and controlled clinical trials that aim at steadily developing and improving treatment concepts for sick patients based on the current scientific knowledge.

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**. One goal of a registry is to collect treatment data for research questions. Furthermore, the registry centre supports the doctors at site with (non-committal) treatment recommendations based on the most recent data on best treatment options, in order to provide the patient with optimal therapy even without the framework of a clinical study.

The following registries are available at the moment:

- **I-HIT-MED Registry:** Patients with pineoblastoma can be enrolled the International HIT-MED Registry, regardless of the treatment given. These patients will receive treatment as per individually designed treatment plans. The goal of the registry is not to assess the feasibility of an ongoing trial, safety or efficacy of a certain treatment. It rather aims at collecting individual patient data for future analysis. The headquarters of the registry are located in the Children's Cancer Centre at the University of Hamburg, Germany. The head of the registry is Prof. Dr. med. Stefan Rutkowski.

## 9. Prognosis

The cure rates for children and adolescents with pineoblastoma are about 60–70 % (5-year-survival rate). However, in individual patients, *prognosis* is dependent on various factors. In particular, the stage of the disease and the patient's age play a significant role. Hence, children and adolescents with metastasized disease have generally a more unfavourable prognosis than patients with localized disease. In young patients who cannot receive *radiotherapy* as part of their treatment, the prognosis is significantly worse, while cure rates of over 80 % have been described in older children with non-metastasised disease.



**Note:** The survival rates mentioned in the text above are statistical values. Therefore, they only provide information on the total cohort of patients with childhood Medulloblastoma. They do not predict individual outcomes.

In the context of cancer, the term "cure" should rather be referred to as „free of cancer“, for even if current treatment regimens may help remove the tumour, the the tumour's growth may have caused irreparable damage to the brain or the treatment may be associated with late effects. Early detection and appropriate management of these long-term secondary effects typically require intensive *rehabilitation* and thorough long-term follow-up care.

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# Glossary

|                                      |  |
|--------------------------------------|--|
| 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.   |
| 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. |
| biopsy                               | removal of a tissue sample for subsequent (mainly microscopic) examination; this can be done, for example, by puncture with a hollow needle, with the use of special instruments (such as forceps, punching instruments, probes) or surgically with a scalpel.   |
| 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).                                     |
| central nervous system               | comprises the brain and spinal cord and is separated from the so-called peripheral nervous system; as a central organ of integration, coordination and regulation, it serves to process external sensory impressions as well as stimuli produced by the organism itself.   |
| cerebrospinal fluid                  | fluid produced by cells of the cerebral ventricles; it floats around the brain and spinal cord to protect them from injury and provide them with nutrients.  |
| cerebrum                             | largest and most highly developed section of the brain; it consists of two hemispheres connected by a thick bundle of nerves (corpus callosum). Each hemisphere of the brain is specialized on specific tasks. The outermost layer of the cerebrum, the cerebral   |



|                     |  |
|---------------------|--|
|                     | <p>cortex, houses the ability to learn, speak and think, as well as consciousness and memory, amongst other things. This is also where the processing centres for information from the sensory organs (e.g. eyes, ears) are located.</p>   |
| chemotherapy        | <p>here: use of drugs (chemotherapeutic agents, cytostatics) for the specific inhibition of tumor cells in the organism</p>  |
| chromosomal         | <p>referring to the chromosomes, carriers of the genetic material (see chromosomes)</p>  |
| CNS tumour          | <p>tumour of the central nervous system; a primary CNS tumour is a solid tumour that originates from brain or spinal cord tissue. Secondary CNS tumours are metastases of tumours located in other organs or tissues.</p>  |
| computed tomography | <p>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)</p>   |
| cytostatics         | <p>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.</p>   |
| diencephalon        | <p>vital part of the brain with function for numerous life processes; it connects to the brainstem towards the cerebrum and consists of functionally different sections. The "thalamus", for example, decides which sensory impressions should penetrate into consciousness and are to be sent to the appropriate processing centers. The "hypothalamus" serves as a mediator between the hormonal and nervous systems and controls, among other things, important metabolic processes (e.g. heat and water balance, carbohydrate, fat, protein metabolism, blood pressure). Together with the pituitary gland, it regulates the activity of subordinate glands. Other parts of the diencephalon are responsible for muscle activities and for controlling the day-night rhythm.</p> |
| DNA                 | <p>abbreviation for deoxyribonucleic acid; it carries the genetic information and is found in all living beings. DNA contains the genes that provide the information for the production of ribonucleic acids (RNA) or proteins. It is a large molecule consisting of two nucleic acid chains twisted into a double helix. The individual chains consist of a sequence of four different building blocks</p>  |



|                        |   |
|------------------------|---|
|                        | (bases), the order (sequence) of which determines the genetic code.   |
| drainage               | here: drainage of pathological or increased natural body fluids to the outside, for example drainage of cerebrospinal fluid from the cerebral ventricles or of pathological fluid accumulation from the pleura (pleural drainage);  |
| 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  |
| electromagnetic        | electromagnetic rays (also known as electromagnetic waves) consist of coupled electric and magnetic fields; examples of electromagnetic radiation are X-rays and gamma rays as well as radio waves, thermal radiation and light.  |
| embryonal              | here: in an early stage of development, immature;   |
| fontanelle             | soft spot on an infant's head, due to the bony plates not having connected yet; the final closure usually occurs before the age of two.   |
| genetic                | concerning the (level of) inheritance or genes; inherited   |
| 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).  |
| hydrocephalus          | medical term for abnormal buildup of cerebrospinal fluid in the cavities (ventricles) in the brain; it is caused by a dilation of the brain's ventricles due to various causes.   |
| 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  |



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| intensity-modulated radiotherapy | modern radiation technology, which provides maximum protection for the surrounding healthy tissue from radiation exposure by means of a highly precise distribution of the radiation dose at the tumour site; the intensity of the radiation dose can be precisely adjusted to the irradiation field only; this may also allow the use of a higher radiation dose.   |
| leukaemia                        | malignant disease of the blood forming (haematopoietic) system and the most common cancer in children and adolescents (about 33%); depending on the origin of the malignant cells, a distinction is made between lymphoblastic and myeloid leukaemias. Depending on the course of the disease (fast or slow), a distinction is made between acute and chronic leukaemias.  |
| lumbar puncture                  | puncture of the spinal canal in the lumbar spine, e.g. to remove cerebrospinal fluid (CSF) or for the purpose of administering medication (so-called intrathecal treatment); in the case of cancer, a sample and examination of cerebrospinal fluid can be used to detect malignant cells; in the case of increased intracranial pressure due to a CNS tumour, cerebrospinal fluid removal (CSF) is also used to relieve pressure. |
| lymph nodes                      | small lenticular to bean-shaped organs that are part of the body's 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.   |
| macrocephalus                    | large head, which can be caused by a hydrocephalus (hydrocephalus) in a child with unclosed fontanelles, but also by a large tumour or both  |
| 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.   |
| metastasis                       | 1. tumour spread from the primary site of tumour to other parts of the body; characteristic feature of malignant tumours (cancer). 2. collective term for a disease process characterized by malignant cells spreading from their primary site to other areas of the body via the bloodstream and/or the lymphatic system.   |
| microscope                       | an instrument that allows you to magnify objects or certain structures of objects that are not visible to the human eye  |





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| molecular             | at the level of molecules   |
| molecular genetic     | referring to structure, formation, development, function and interactions of cells and cell building blocks (e.g. nucleic acids, proteins) at the molecular level; the focus is on the analysis of the genetic information stored in the nucleic acids (DNA and RNA) and its processing in the context of protein synthesis as well as gene regulation. |
| MRI                   | abbreviation for magnetic resonance imaging, a very precise, radiation-free examination method for imaging structures inside the body   |
| nerve tissue          | tissue of the nervous system; it consists of nerve cells (neurons) and its own special connective tissue, the glial cells.  |
| neurological          | referring to the function of the nervous system / nerve tissue  |
| paediatric oncologist | paediatrician who is specialized on the management of children and adolescents with cancer  |
| 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.   |
| pineal gland          | hormone gland attached to the diencephalon between the two cerebral hemispheres; its function is the production of melatonin, a hormone that makes the body respond to changes in light conditions.   |
| 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                | a positively charged particle within an atom; together with the electrically neutral neutrons, it forms the atomic nucleus. Protons form the counterpart to the negatively charged electrons of the atomic shell.   |
| 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.   |



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| radiation           | controlled use of ionizing (high-energy) radiation for the treatment of malignant diseases   |
| radiologist         | a physician specialized in diagnostic imaging and radiotherapy   |
| radiotherapy        | controlled use of ionizing (high-energy) radiation for the treatment of malignant diseases   |
| recurrence          | relapse, recurrence of a disease after recovery  |
| rehabilitation      | medical, social, psychosocial and occupational measures after an illness for reintegration into society, work and private life, which may include, among other things, the restoration of abilities through exercise treatment, prostheses and other measures  |
| retinoblastoma      | a rare malignant tumour of the retina that occurs almost exclusively in children; there are hereditary and non-hereditary forms of the disease. Either one or both eyes can be affected (unilateral or bilateral retinoblastoma). In very rare cases, hereditary retinoblastoma can also occur together with a brain tumour (e.g., pineoblastoma); in this case, it is called trilateral retinoblastoma.   |
| retinoblastoma gene | tumour suppressor gene called RB1, which causes the development of a malignant retinal tumour (retinoblastoma) when genetically altered (mutated); it is located on the long arm of chromosome 13.   |
| seizures            | uncontrolled electrical activity between nerve cells in the brain; a distinction is made between focal and generalized seizures. Focal seizures are limited to a specific area of the brain; depending on the area of the brain, the symptoms vary: e.g. twitching of one side of the body, an arm or a leg. Generalized seizures spread over large areas of the brain and lead, for example, to twitching of the limbs, sudden absence and loss of consciousness. |
| solid tumour        | solid, localized increase of the body's own tissue; solid tumours can originate from various tissues and can be benign or malignant; only the malignant ones are considered as cancers. Solid tumours include all cancers that do not affect the haematopoietic or lymphatic system. The latter are systemic malignancies. The most common solid tumours in childhood and adolescence are brain tumours, followed by neuroblastoma and soft tissue sarcomas.       |
| spinal cord         | part of the central nervous system; its main function is to transmit messages between the brain and other organs of the body. The  |



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|                          | spinal cord is protectively enveloped by the three spinal cord membranes and the bony spinal canal.  |
| surgery                  | surgical intervention on or in the body of a patient for the purpose of treatment, less often also in the context of diagnostics; the surgical intervention is carried out with the help of special instruments, generally with the patient under anesthesia.  |
| 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. |
| tumour                   | groups of abnormal cells forming a growing lump, both benign and malignant   |
| undifferentiated         | here: immature, not yet functional and usually capable of unlimited division (e.g. stem cells); the development from undifferentiated to differentiated cells and tissues (differentiation) takes place in stages. Accordingly, there are many different degrees of differentiation.                                     |
| 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   |