Choroid plexus tumours - Brief information

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Kinderkrebsinfo is sponsored by Deutsche Kinderkrebsstiftung

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1. General information on the disease

Tumours of the choroid plexus, also known as choroid plexus tumours (CPT), are very rare tumours of the *central nervous system* (CNS). They arise from a brain tissue, the choroid plexus, which coats the fluid-filled cavities of the brain (called *cerebral ventricles*). Choroid plexus tumours most frequently occur within the lateral ventricles in the *cerebrum*, but the third ventricle in the *midbrain* as well as the fourth ventricle in front of the *brainstem* and the *cerebellopontine angle* can be origins of tumour growth.

Both benign and malignant choroid plexus tumours exist. Depending on the degree of malignancy as defined by the World Health Organization (*WHO classification*), the following grades of choroid plexus tumours are differentiated:

- (benign) choroid plexus papilloma (CPP, CNS WHO grade 1)
- (intermediate grade) atypical choroid plexus papilloma (APP, CNS WHO grade 2)
- (higher grade) choroid plexus carcinoma (CPC, CNS WHO grade 3)

All three types are comparably frequent. While choroid plexus papilloma type 1 and 2 mostly grow within the lateral ventricles, choroid plexus carcinoma tends to also invade adjacent brain tissue. However, all plexus tumours, including choroid plexus papilloma type 1 and 2, can spread (metastasise) within the brain and the *spinal cord* via the *cerebrospinal fluid* (CSF) that fills the brain cavities and the spinal canal.

A typical first sign of a choroid plexus tumour is (due to its origin) associated hydrocephalus (*see chapter „Symptoms“*).

2. Incidence

Choroid plexus tumours are very rare; they account for just about 2 % of all central nervous system (CNS) tumours and 0.4 % of all malignant diseases in children and adolescents. Toddlers are most frequently affected, especially one year-olds, while the disease is rather rare in teenagers and adults. The average age of patients (between 0 and 17 years) at diagnosis is is between 2 and 3.

According to the German Childhood Cancer Registry (Mainz), about 10 children and adolescents under 18 years of age are newly diagnosed with a choroid plexus tumour in Germany per year. This corresponds to an incidence of 1:1.000.000 children and adolescents (including all paediatric age groups). Since the incidence peaks in very young children, the proportion of plexus tumours

among all CNS tumours is much higher in this age group, amounting, for example, to up to 13 % for children in their first year of life.

3. Causes

Choroid plexus tumours arise from altered cells of the brain tissue that coats the inner walls of the brain's fluid-filled cavities (plexus choroideus). What causes the (malignant) transformation of these cells still needs to be elucidated. It is known so far, however, that children and adolescents with certain hereditary diseases, such as *Li-Fraumeni syndrome*, have an elevated risk to develop a choroid plexus tumour, in most cases a choroid plexus carcinoma. Since such genetic conditions are associated with a predisposition for tumours, they are also called *cancer predisposition syndrome*s. But the disease often appears in patients without any hereditary disease, too.

Aside from hereditary factors, choroid plexus tumour cells (especially plexus carcinoma cells) often show modifications on certain *gene*s or *chromosomes*. These modifications may lead to impaired cell development and dysfunctional communication between cells, thereby possibly contributing to the transformation of a healthy into a malignant cell. Such genetic alterations are usually not hereditary, they mostly happen early in development.

4. Symptoms

The major role of the plexus choroideus in the brain's ventricles is to produce the *cerebrospinal fluid* (CSF), which protects brain and spine from trauma as well as provides it with nutrients. As plexus tumours derive from this tissue, they can produce this fluid as well – according to their volume to such an extent that it accumulates in the ventricles, thereby leading to a *hydrocephalus*. Hydrocephalus can also develop when the tumour blocks the circulation and/or drainage of the cerebrospinal fluid due to its location in the brain ventricles.

Depending on the patient's age, the following general symptoms caused by increased production of cerebrospinal fluid may be present:

- Babies and toddlers with their soft spots (*fontanelle*s) still open may show an abnormal head circumference (*macrocephalus*). Also, personality changes, moodiness, failure to thrive and *neurological* impairments (for example squinting, torticollis), being hyper-agitated and screaming without an obvious reason may be presenting symptoms of a brain tumour in young children.
- In children whose soft spot is already closed, the space-occupying tumour and/or the excess cerebrospinal fluid may cause increased intracranial pressure, thereby leading to headaches, backpain, drowsiness, loss of appetite, nausea and vomiting (particularly in the morning after getting up), weight loss, fatigue, problems concentrating as well as personality and mood changes.

Depending on the tumour's location in the central nervous system (CNS), thus on which brain regions are being impaired by the tumour, so-called "region-specific" symptoms may also be observed. For example, a tumour growing within the brain hemispheres (*cerebrum*) or in the

midbrain can be associated with palsies and/or seizures, whereas typical symptoms of a tumour in the *cerebellum* or the *brainstem* are impaired balance and/or cranial nerve palsies. Symptoms like these can help the doctor to get an idea of the tumour site.

Good to know: Not all patients presenting with one or more of the symptoms mentioned above do have a choroid plexus or another type of brain tumour. Many of these symptoms can also occur with other, harmless diseases that are not associated with a brain tumour at all. However, if certain symptoms persist or worsen (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 doctor thinks that the young patient's history (*anamnesis*) and *physical examination* are suspicious of a tumour of the *central nervous system* (CNS), the patient should immediately be referred to a childhood cancer centre 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 planning and *prognosis*.

5.1. Tests to confirm 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 diagnostic *imaging*, such as *magnetic resonance imaging* (MRI). These diagnostic tools help to confirm or rule out the presence of a CNS tumour and to determine a possible spread of the disease in other parts of the central nervous system, including the spinal canal. Also, tumour size and site, its extent with regard to the adjacent tissue, and a hydrocephalus can be assessed by these imaging techniques.

If in a baby or toddler magnetic resonance imaging (involving the use of a *contrast agent*) reveals a contrast-enhancing tumour in the region of the *cerebral ventricles*, a choroid plexus tumour can be suspected. Final confirmation of this diagnosis requires *histological* examination of a tumour sample, which is obtained during neurosurgical tumour removal (*see chapter "Treatment*").

5.2. Tests to assess spread of the disease

Once the diagnosis of a choroid plexus tumour has been confirmed by histology, 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), these tests also include checking of the *cerebrospinal fluid* (CSF) for tumour cells. Cerebrospinal fluid is mostly obtained from the spine in the lower back (*lumbar puncture*).

5.3. Diagnostics by the Reference Centres

In order to secure the validity of the results, having the MRI results as well as the cerebrospinal fluid reviewed by a second expert (so-called referee expert) is recommended. This strategy helps preventing inaccurate assessment of metastasis and its subsequent impact on treatment planning.

5.4. Tests before treatment

Further tests prior to treatment include examination of the heart function by *electrocardiography* (ECG) and/or *echocardiography* (echo). Also, various blood tests serve to find out about the patient's general health status and whether the functions of certain organs (such as the liver, kidneys and endocrine system) are intact, since if not, this needs to be considered before and during 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.

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 diagnosis has been confirmed, therapy is planned. In order to design a highly individual treatment regimen for the patient, adapted to his clinical situation and risk of relapse, certain individual factors influencing the patient's *prognosis* (called risk factors or *prognostic factors*) are being considered during treatment planning (risk-adapted treatment strategy).

The most relevant prognostic factor for patients with a choroid plexus tumour is the (histological) type of the disease. It defines the tumour's growth behaviour and, thus, its grade of malignancy (CNS WHO grade, see chapter "General information on the disease"). Therefore, the CNS WHO grade is the major factor to be considered when determining the optimal treatment plan for a patient with choroid plexus tumour. Also, checking for *Li-Fraumeni syndrome* is important, since this disease is usually associated with a less favourable prognosis for the patient and can affect family members. Furthermore, it requires special considerations for long-term follow-up of these patients.

Additional prognostic factors are the anatomical site, size and spread of the tumour as well as the extent of surgical removal and the response of the disease to chemo- and/or radiotherapy. Also, the patient's age and clinical condition play an important role. Patient's age at diagnosis particularly determines whether radiotherapy is an option or not. All these factors are considered in order to achieve the best possible outcome for every patient with choroid plexus tumour.

7. Treatment

Treatment of children and adolescents with a choroid plexus tumour (CPT) should take place in a children's hospital with a paediatric oncology program. Only such a childhood cancer centre provides highly experienced and qualified staff (doctors, nurses and many more), since they are specialised and focused 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 high cure rates while avoiding side and late effects as much as possible.

For patients with a choroid plexus tumour the tmajor treatment columns are surgery, chemotherapy and – depending on the patient's age – radiation.

Important note: The treatment options described in the following paragraphs are recommendations by the CPT trial/registry centre. They are based on the experiences and results obtained from the treatment study CPT-SIOP 2000 (closed in 2020), which have been analysed by the Brain Tumour Group of the European Society for Paediatric Oncology (SIOP-E BTG) (*see also chapter "Trials and registries"*.) The treating physician will decide, together with the patient and/or his relatives, how the individual treatment of the particular patient is going to look like.

7.1. Surgery

First and major step in the treatment of a patient with choroid plexus tumour is *surgery*. While considering the associated risk of surgery, the goal is to remove as much of the tumour as possible, since the extent of neurosurgical resection (neurosurgery) can impact the subsequent course of the disease. Therefore, if, after an incomplete tumour removal, *histological* analysis of the tissue confirms a choroid plexus tumour, the doctors will discuss the options of a second surgery in order to remove the remaining tumour tissue as well.

7.2. Watch-and-wait approach or non-surgical treatment

Following surgery, either close monitoring of the course of the disease (watch-and-wait approach) or subsequent (adjuvant) cancer treatment are the options, depending on the patient's individual situation, which includes the histological type of tumour, the stage of the disease at diagnosis as well as the extent of surgical removal.

7.2.1. Watch-and-wait (monitoring)

Patients with a non-metastasised classic plexus papilloma (CPP, CNS WHO-grade 1) and patients after complete resection of an atypic plexus papilloma (APP, CNS WHO-grade 2) usually do not receive further treatment after *neurosurgery*. However, the doctors closely monitor the course of the

disease (by regular *magnetic resonance imaging*, MRI). Further treatment is only recommended if recurrent tumour growth occurs. But many patients remain tumour-free after only one surgery.

7.2.2. Non-surgical treatment after surgery (adjuvant therapy)

For patients with metastasised plexus papilloma (CPP, CNS WHO grade 1), with incompletely removed atypical plexus papilloma (APP, CNS WHO grade 2), or with plexus carcinoma (CPC, CNS WHO grade 3), surgery alone is not enough. Since the risk of progressive disease or relapse, respectively, is very high in these patients, the doctors will recommend additional non-surgical treatment, such as a *chemotherapy* and, if necessary, also *radiation therapy* (radiotherapy).

With chemotherapy, the patient receives a combination of substances that inhibit cell growth, thereby stopping the tumour cells from growing or even killing them (cytostatic therapy). Radiotherapy uses highly energetic, *electromagnetic* radiation, which is usually given from a machine through the skin onto the tumour site, thereby causing *DNA* damage and subsequently death of the tumour cells.

Modern techniques, such as *intensity-modulated radiotherapy* (IMRT), help minimise radiation damages in healthy tissue. Instead of conventional radiotherapy with *photon*s, particle radiation with *proton*s (*proton therapy*) can be an option for some patients 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.

Final decision with regard to the best treatment strategy for each individual patient especially takes into consideration the patient's age at diagnosis as well as the histological tumour type. In general, all patients requiring adjuvant treatment (according to the criteria mentioned above) should receive chemotherapy. Whether this is followed by radiotherapy is decided individually.

7.2.2.1. Chemotherapy

Chemotherapy usually consists of a combination of different *cytostatics* (polychemotherapy) given over several treatment cycles. In Germany, the standard chemotherapy regimen for patients with choroid plexus tumour includes a combination of the agents carboplatin, etoposide and vincristine, which are given every four weeks – for a total of six times – as an intravenous *infusion* (also known as *systemic* treatment). Depending on the individual disease situation, more treatment cycles as well as additional agents are an option. Other combinations of cytostatic agents, for example like those applied in North America, are possible, too.

In rare situations and in case of recurrent disease, certain chemotherapeutic agents may be given directly into the *cerebral ventricles* (*intraventricular* therapy). Prior to this type of treatment, a brief neurosurgical procedure is required in order to temporarily implant a so-called *Ommaya reservoir* (a little cushion-like device of the size of a kidney bean) under the scalp. Via this device, the doctors can

both give chemotherapy and take cerebrovascular fluid (CSF) for checking on / ruling out remaining tumour cells floating in the CSF.

7.2.2.2. Radiotherapy

Whether radiotherapy is indicated after chemotherapy and if so, how intensive it should be, requires individual assessment for every single patient. The decision in favour or against radiotherapy particularly depends on whether the tumour responded well to chemotherapy and whether there is remaining tumour after chemotherapy. Additional factors include the presence of metastases at diagnosis [see *metastasis*] and the patient's age. Patients with *Li-Fraumeni syndrome* usually don't benefit from radiotherapy.

For patients with Li-Fraumeni syndrome, (conventional) radiotherapy is generally not recommended, but focal proton treatment (*see above*) may be an option.

Diagnostic tests during treatment: Diagnostic imaging (such as *ultrasound* and *magnetic resonance imaging*) as well as testing the cerebrospinal fluid are recommended at defined timepoints of treatment in order to assess the disease's response to therapy. This helps to find out if treatment may require adjustment or can be continued as before.

8. Trials and registries

IIn Germany, children and adolescents with a tumour of the central nervous system are usually treated according to the treatment plans (protocols) of clinically controlled trials or registries. The clinical trials aim at improving current treatment concepts for 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 who do not meet the required inclusion criteria, respectively, may be included in a so-called **registry**. Such a registry mainly serves to accompany patients scientifically during treatment and follow-up. It pools patient data for further scientific analysis in order to acquire evidence-based information, based on which both current and future treatment strategies can be optimised and valid recommendations given. To ensure optimal treatment for patients not registered in a study, experts from assigned trial panels usually provide recommendations and advice to the local caregiver team.

A long-term international trial for the treatment of children and adolescents with choroid plexus tumours (CPT) was closed in 2010: trial CPT-SIOP 2000. There is currently no open trial for CPT patients. A new study is in development in collaboration with North America due to the rarity of the disease. The international **CPT-SIOP Registry** has been open to newly diagnosed patients since March 2010. Since the CPT study group joined the HIT-MED study group in 2024, registration of CPT patients in the **I-HIT-MED Registry** has been possible and preferred within Germany.

The current treatment recommendations given by the principal investigators at the registry's study centre are based on the interim results obtained from trial CPT-SIOP 2000 and those from international trials as well as on the ongoing data analyses within the registry. If needed, treating physicians can contact the registry centre. The development of standardised European treatment

guidelines is currently carried out in the context of the ERN PaedCan project. However, after all, individual treatment planning is left to your child's oncologist's discretion.

The national CPT registry and trial headquarters are located at the University Children's Hospital in Hamburg, Germany, with Dr. med Denise Obrecht as the head (see *www.uke.de/cpt*).

9. Prognosis

The outcome (prognosis) of a patient with a choroid plexus tumour (CPT) mainly depends on the type of CPT. In particular for patients with malignant types of choroid plexus tumour, the biological behaviour and the spread of the tumour, that is, the presence or absence of *metastasis* (spread into the cerebrovascular fluid), play a prognostic role.

Patients with classic plexus papilloma (CPP, CNS WHO grade 1) usually have a very favourable prognosis with a five-year survival rate of up to 100 %. Also, patients with atypical plexus papilloma (APP, CNS WHO grade 2) show good prognosis: their average five-year survival rate is – according to information provided by the CPP-SIOP Registry – about 95 %, with the outcome for less than 2-year-olds being a bit more favourable than for older children (100 % or 85 %, respectively).

Patients with plexus carcinoma (CPC, CNS WHO grade 3) have an average five-year survival rate of about 60 % after combination therapy, which, however, individually strongly depends on the success of surgery and the options of additional cancer therapy as well as on the presence (or absence) of a certain *genetic* modification in tumour cells (so-called *TP53 mutation*). However, the risk of recurrent disease (relapse) is high for patients with plexus carcinoma. Nevertheless, there are currently a good number of long-term survivors of plexus carcinoma in Germany.

Note: The above-mentioned survival rates are statistical values. Therefore, they only provide information on the total cohort of patients with this kind of childhood brain tumour. They do not predict individual outcomes.

In the context of cancer, the term "cure“ should rather be referred to as „free of cancer“, because current treatment regimens may help getting rid of the tumour, but they are also frequently associated with numerous late-effects. Early detection and appropriate management of these long-term secondary effects typically require intensive *rehabilitation* and thorough long-term follow-up care, although a patient may have been "cured“ from the cancer.

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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.
brainstem	the section of the brain that forms the transition between the brain and the spinal cord; it controls vital functions, such as breathing, heart rate and blood pressure, and is responsible for important reflexes such as the blinking, swallowing or coughing reflex, lacrimation and saliva production. This is also where the roots of the cranial nerves are located.
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.
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.
cerebellopontine angle	niche in the posterior region of the brain and part of the cerebellum; this is where the roots (nuclei) of ten of the twelve cranial nerves are located in a very small space.
cerebellum	part of the brain that is located in the posterior fossa of the skull, between the cerebrum and the brainstem; it is mostly responsible for the coordination of all body movements and also for maintaining balance.
cerebral ventricles	cerebral ventricles filled with cerebrospinal fluid; the four cerebral ventricles represent the continuation of the spinal canal merging into these four chambers in the brain.

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 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.
chemotherapy	here: use of drugs (chemotherapeutic agents, cytostatics) for the specific inhibition of tumor cells in the organism
CNS tumour	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.
contrast agent	substance that can be used to improve the visualization of structures and functions of the body in imaging techniques; contrast agents are mainly used in X-ray diagnostics (X-ray examination, computed tomography), magnetic resonance imaging (MRI) and ultrasound examinations.
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.
DNA	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 (bases), the order (sequence) of which determines the genetic code.
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.
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.
gene	unit of genetic information in the genome of living organisms; a gene contains the genetic information – the blueprint – for a specific gene product (protein or RNA). In most organisms, the entirety of all genes, the genome, is present as a deoxyribonucleic acid chain (DNA), which forms the chromosomes in the cell nucleus. The information of a gene is mediated by a certain sequence of the nucleic acid building blocks adenine, guanine, cytosine and thymine.
genetic	concerning the (level of) inheritance or genes; inherited
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
infusion	introduction of fluids into the body, usually over a long period of time and via a central venous catheter; an infusion is given, for example, to supply water, electrolytes, proteins and/or medication as part of intensive treatment.
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.

intraventricular	into the ventricular system / in the ventricular system, i.e. into / in the cerebrospinal fluid
Li-Fraumeni syndrome	cancer predisposition syndrome characterized by the increased occurrence of various solid tumours within a family; in childhood and adolescence, tumours of the adrenal glands, as well as soft tissue sarcomas, leukaemias and CNS tumours are most commonly observed, in adulthood mainly bone tumours (osteosarcomas), breast cancer and lung tumours. In most cases, there is a change (mutation) of the so-called tumour suppressor gene TP-53 (protein p53).
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.
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.
midbrain	part of the brainstem; adjacent to the diencephalon at the top and the bridge at the bottom, the latter of which is also part of the brainstem. In the midbrain, there are important pathways that ascend and descend between the brain and the spinal cord. The midbrain is also home to the eye muscle nerves (cranial nerves III and IV) and to the nerve cell nuclei for certain muscle activities (e.g. of the face and neck).
mutation	alteration of genetic material; it can arise without any identifiable external cause (so-called spontaneous mutation) or be caused by external influences (induced mutation). External influences

	include, for example, ionizing radiation or certain chemical substances (mutagens). If somatic cells are affected, it is referred to as a somatic mutation, and if germ cells are affected, it is referred to as a generative mutation. Somatic mutations are not heritable, while germ cell mutations can lead to hereditary damage. Depending on the extent of the change (single or multiple genes, larger chromosome segments or complete chromosomes), a distinction is made between point and block mutations as well as numerical and structural chromosomal aberrations.
neurological	referring to the function of the nervous system / nerve tissue
neurosurgery	a branch of surgery that includes parts of the diagnosis and surgical treatment of diseases of the nervous system
Ommaya reservoir	a small plastic reservoir that can be implanted under the scalp and is then connected to one of the brain's ventricles; the shape of the Ommaya reservoir is reminiscent of a small pillow. At its bottom, it is connected by a tube (ventricular catheter) to one of the cerebral chambers (usually the right lateral ventricle) or another cavity in the brain filled with cerebrospinal fluid (CSF) (e.g. arachnoid cyst). The Ommaya reservoir (or Rickhams reservoir, another model with a similar mechanism) is implanted as part of a short, neurosurgical procedure. Such a reservoir can be connected to a shunt system for the long-term treatment of hydrocephalus or to a ventricular catheter.
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.
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.
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, protheses and other measures
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 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.
systemic	covering/includiing the entire body
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 classification	international guideline developed by the World Health Organization (WHO) for classification, diagnosis and differentiation/grading of (malignant) diseases