Norwegian Childhood Cancer Society

Solid tumours in children and adolescents

Symptoms, treatment and follow-up

Our values

We give *hope* to each other and the world.

We have the *courage* to speak
out on behalf of our families.

We have the *strength* to support
children and families who are affected.

We create *joy* in daily life.

Solid tumours in children and adolescents

When children and adolescents have cancer, it has a major impact on the daily lives of both the child and the family. Treatment of cancer is demanding and lengthy. This brochure is written to provide insight into the challenges that lie ahead when a child or adolescent has cancer. Such a brochure can never accommodate all the questions and thoughts you might have, but our desire is to provide facts about both the types of cancer and the treatment.

We have emphasised information that we know parents and networks around the child wonder about. It is also important to remember that no families are the same, and that every family will have its own experiences and feelings when one of the children in the family is diagnosed with cancer.

The information in this brochure is intended always to be valid. Therefore, we have omitted some details. We would also encourage you to look at the websites of the Norwegian Childhood Cancer Society and the Childhood Cancer Portal, where information is updated at all times.

This brochure will discuss the most common solid tumours. These are neuroblastoma, Wilms tumour, rhabdomyosarcoma, osteosarcoma, Ewing sarcoma, and retinoblastomas.

→ **Read further to learn more about cancerous tumours.**

What is meant by solid tumours?

Children and adolescents get different types of cancer than adults.

The two largest groups are acute leukaemia (blood cancer) and tumours of the central nervous system (brain and spinal cord), which account for about a third each. The final third are solid tumours outside the central nervous system. These are the ones we will look at in more detail in this brochure.

Such tumours, or "lumps", are composed of malignant cells, and they usually grow gradually, some slowly, some quite rapidly. They can be found in many different places in the body, they can be large or small, and they can be "kind" or more aggressive. The treatment depends on what type of tumour it is.

What types of solid tumours are found in children and adolescents?

The tumours often originate in different tissue in the body, both connective tissue and bone tissue, and because these are located in many parts of the body, tumours can grow from a number of sites.

This is different from cancer in adults, where the cancerous tumour often appears in specific organs such as the prostate, bowel and lung, to name a few. In children, the location in the body will at best give a clue as to what type of tumour it may be, but it will never be sufficient to achieve a correct diagnosis. We therefore always need a tissue sample of the tumour, also called a biopsy, to make a definite diagnosis.

Solid tumours can appear practically anywhere in the body, and symptoms will therefore vary greatly. Most symptoms can also occur in other contexts—and most often they are not a sign of cancer.

The main types of solid tumours in children and adolescents

Neuroblastoma

Neuroblastoma is a rare tumour, with fewer than 10 cases in Norway per year.

The tumour originates in the voluntary nerve system, and is found primarily in the adrenal glands, but also in the ganglia along the spinal column. Neuroblastoma is most common in the first two years of life and is rare after childhood. Over two thirds of patients experience spreading of the disease ("metastases").

Neuroblastoma is an enigmatic disease — it comes in many forms, which can be more or less aggressive. In infants, who often have a "kind" form of the tumour, we sometimes see the tumour disappear spontaneously, while older children with apparently the same extent of the disease, require intensive treatment to have any chance of recovery. Research has shown that there are different markers that tell us how aggressive the tumour is. The most important factor is the n-myc gene. If the n-myc is increased, the prognosis is poorer. Infants often, but not always, lack these unfavourable factors.

Typical symptoms are anaemia, lethargy, weight loss, fever or abnormal amount of sweating, in rarer cases also high blood pressure. Often the first symptom is a lump in the abdomen that can be seen or felt. If the disease has spread, there will be skeletal pain, bleeding around the eyes and bone fractures without any proper trauma. Paralysis may indicate pressure on nerves or on the spinal cord.

The diagnosis is made using imaging such as an MRI and CT, a biopsy and findings of typical tumour markers. Most neuroblastomas secrete adrenaline substances (catecholamines) that can be detected in the urine. In addition, there are typical markers in the blood. A bone marrow examination is also important, as cancer cells can spread to the bone marrow.

The treatment depends on whether the tumour is "kind or aggressive", on the extent and on prognostic factors such as n-myc. Surgery is the primary treatment and for localised tumours without risk factors, surgery is sufficient. Others need surgery and some chemotherapy. In cases with extensive disease, intensive chemotherapy and surgery must be combined with high-dose treatment with autologous stem cell support, using the child's own stem cells (HMAS), in addition to radiation and immunological maintenance therapy.

Neuroblastoma is considered a particularly difficult form of childhood cancer, but survival has increased significantly. The prognosis for localised tumours is good, while treatment of extensive disease remains a challenge.

Wilms tumour

Wilms tumour is a solid tumour of the kidneys.

It was named after the German physician Max Wilms, who first described the tumour. Wilms tumour is also called "nephroblastoma" (for nephros [Greek] = kidney). Wilms tumour is rare. In Norway only five to eight children are diagnosed annually, and the children are usually under five years old when diagnosed. The tumour can spread to lymph nodes and to the lungs, among other places.

Children with certain congenital conditions, for example hemihypertrophy or Beckwith Wiedemann syndrome, have an increased incidence of Wilms tumour. Children with these conditions are included in a screening programme, where blood samples and ultrasounds of the kidneys are taken at regular intervals throughout infancy.

The tumour is often discovered because the child's abdomen bulges or a mass can be felt. Often this can be detected by chance in an otherwise healthy child. Other symptoms are abdominal pain, blood in the urine and, in rare cases, high blood pressure.

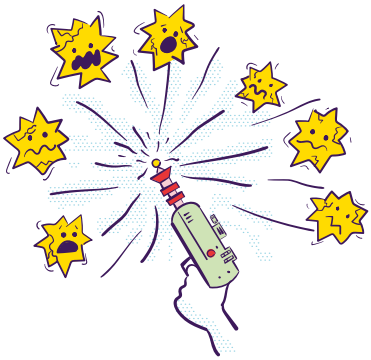
General symptoms such as reduced appetite or fever are rare.

The investigation typically includes an ultrasound and MRI of the abdomen, as well as a CT of the lungs to rule out metastases. In typical cases of Wilms tumour, the biopsy (tissue sample) is often skipped at first, but the diagnosis is confirmed by examination of the tumour tissue after surgery.

The treatment for Wilms tumour is a combination of chemotherapy and surgery. Usually, chemotherapy is given for 4 – 6 weeks, and then the tumour and the remainder of the kidney are removed. After surgery, depending on the stage and aggressiveness of the tumour, a new round of chemotherapy can last from a few weeks to six months. In some cases it may be necessary with additional radiation therapy to the tumour area and/or to the lungs.

Survival of Wilms tumours is good, with nine out of ten children surviving. A few cases need aggressive treatment to have a chance of cure. Relapses of Wilms tumour can often be completely cured, but the child must undergo a new round of treatment, which often will include radiation therapy.

Rhabdomyosarcoma



Rhabdomyosarcoma is the most common form of cancer of the soft tissue, such as muscle and connective tissue, but it is rare in children.

In Norway, about five children a year are affected by this form of tumour, and the age at diagnosis is often under five years. The tumour can occur in many different parts of the body, but the most common are the head and neck, genitals and urinary tract, as well as in the arms or legs.

There are two variants of the disease:

- Embryonal rhabdomyosarcoma is the most common form and often has a favourable prognosis.

- Alveolar rhabdomyosarcoma is rarer. This form is more often localised in the arms or legs, and has a tendency to spread and relapse.

Around 20 per cent of patients have spreading to other areas of the body, most often to the lungs, but also to lymph nodes, bone marrow or the skeleton.

Symptoms depend on the location of the tumour. There may be swelling around the eye, lumps in the ear and nose, blood from the urinary tract or vagina, difficulty emptying the bladder when urinating, or visible tumours in other parts of the body. Tumours in the head and neck region may cause neurological signs such as facial paralysis or paralysis of the eye muscles due to pressure on, or infiltration of, cranial nerves.

The diagnosis is made using imaging techniques such as ultrasound and MRI of the tumour area, in addition to X-rays and CT of the lungs. The blood tests are usually normal and there are no specific tumour markers. Tissue examination (needle biopsy under anaesthesia) is always necessary. The biopsy is examined using various special methods.

Detection of specific genetic alterations in the cancer cells may be indicative of particular subgroups of rhabdomyosarcoma.

Treatment consists of a combination of chemotherapy, and usually either surgery or radiation therapy. Chemotherapy is always given, but in rhabdomyosarcoma, local tumour control by surgery or radiation is particularly important, as the greatest risk is local tumour regrowth. In cases where surgery of the tumour is difficult or impossible (especially in cerebral = parameningeal sites), radiation is given as local therapy, even if it involves a risk of late effects.

Examples of late effects are asymmetries of the face due to one-sided irradiation of growing bone tissue, or hormonal problems due to irradiation of the pituitary gland.

Survival for the entire group in the Nordic countries is around 80 per cent, but varies widely from localised, radically operated tumours that have the best prognosis, to metastazised tumours that remain difficult to treat.

In children, the location in the body will at best give a clue as to what type of tumour it may be.

Bone tumours

The two most common, but still rare, forms of skeletal tumours are osteosarcoma and Ewing's sarcoma.

Both are most common in the age group 10 – 20 years, but Ewing's sarcoma in particular occurs also in younger children.

Osteosarcoma

In osteosarcoma, the tumour is most often located near the large joints of the femur or the tibia, but other sites in the body are also common. The main symptoms are pain where the tumour is located (often also at night), limping and sometimes a visible swelling. Ordinary skeletal X-rays may in typical cases provide a strong indication of osteosarcoma. In addition, MRI and possibly CT examinations of the area in question are performed. After the images have been taken, a biopsy (tissue sample) of the tumour is also performed. This is done either by needle biopsy, in which a thin, hollow needle is inserted and a sample taken, or an open biopsy. The blood tests are usually normal and there are no known tumour markers in the blood.

Ewing's sarcoma

In Ewing's sarcoma, tumours may be located in the leg, but also in the pelvis, spinal column, humerus or thoracic wall (ribs). Symptoms are usually a painful swelling, but in contrast to what is found in osteosarcoma, general symptoms such as fever, lethargy, anaemia and high blood sedimentation are common. Blood tests may show increased white blood cells and increased LD (lactate dehydrogenase). A biopsy (tissue sample) is needed for definite diagnosis. Detection of a specific genetic alteration in the tumour cells is indicative of Ewing's sarcoma.

The treatment for skeletal tumours is the same as for adults, with slight modifications. Before surgery, the child receives a period of chemotherapy (preoperative or "neo-adjuvant" chemotherapy). This is done to shrink the tumour before surgery. During the surgery, as much of the tumour as possible is removed, before more chemotherapy is given. In some cases, radiation therapy is also needed, and in selected cases, high-dose chemotherapy with autologous stem cell support, using the child's own stem cells, is the best option.

The prognosis for survival of Ewing's sarcoma is around 80 per cent, strongly dependent on whether or not there is spreading. The same applies to osteosarcoma, but overall survival is lower than for Ewing's sarcoma.

Retinoblastoma

This is a typical solid tumour affecting young children originating in the retina of the eye.

On average, four children a year in Norway develop this tumour. The diagnosis is often made in infancy. Some cases are hereditary, and tumours often occur in both eyes. Herditary cases also have an increased risk of developing other types of cancer later in life.

Retinoblastomas are treated with local measures, often using a catheter to inject a chemotherapy drug into the blood vessels supplying the eye. Sometimes systemic chemotherapy treatment is needed. In rare cases, the eye must be removed. Radiation therapy is currently avoided, as this would increase the risk of new cancer developing in the irradiated area.

Survival of retinoblastoma is close to 100 per cent, but many patients have late effects such as impaired vision, and in some cases blindness.

Other solid tumours

There are a number of rare forms of cancer in children and adolescents, such as liver tumours (hepatoblastoma and hepatocellular carcinoma), germ cell tumours and others. Investigation and treatment follow the same principles as for the other cancers described.

TIPS

When cancer is suspected in a child or adolescent, the investigation will take place at a department with expertise in childhood cancer.

What do we know about causes of childhood cancer?

In most cases, we don't know what causes solid tumours in children. It seems that environmental factors, electromagnetic radiation, socioeconomic status of the parents or infections have no significant impact. The development of cancer in children is also unrelated to pregnancy, and it is definitely not because their parents have done something wrong.

However, we are aware of a few risk factors. Previous radiation therapy, usually for a different cancer, increases the risk of cancer developing in the irradiated area.

Also, previous chemotherapy can increase the risk of cancer. Certain congenital conditions increase the risk of developing cancer in childhood. An example is neurofibromatosis type 1 and 2.

There is a lot of research on hereditary predisposition to cancer, but for the time being we find only a few cases where cancer is linked to hereditary factors.

Side effects of the treatment

The treatment is demanding in many ways, and unfortunately, it has some side effects. Thanks to research, the treatment is evolving, and in the future, it will be increasingly personalised.

Anaemia

Low red blood cell counts cause symptoms such as lethargy, pallor and shortness of breath. Red blood cells can be replenished by a blood transfusion, which is quite common to have several times during the treatment period.

Increased bleeding tendency

Low platelet counts lead to an increased risk of bleeding. The child will bruise easily, and even small wounds or scratches can bleed for a long time before the bleeding stops. For example, you should not take rectal temperature readings, as the mucous membrane can easily bleed. Blood platelets can be replenished by a transfusion.

Sore mucous membrane

Some chemotherapeutic drugs may cause mucosal damage, which can be very painful. In the case of sore mucous membranes, good pain relief and good oral hygiene are important to avoid infections. More frequent dental check-ups are recommended for everyone throughout the treatment period.

Hair loss

Some drugs can cause hair loss, but the hair will grow back after the end of treatment, sometimes already during treatment. For the youngest children this is usually not a big problem, but older children and adolescents may find this difficult. However, all children are entitled to wigs and special headwear during the treatment period, and the hospital will help to arrange this.

Impaired immune system

Low white blood cell counts lead to increased vulnerability to infections. It is therefore very common that the child will regularly undergo intravenous antibiotic treatment during the treatment period.

The white blood cells cannot be replenished by a transfusion. When the white blood cell count is low, we have to wait for

the bone marrow to start producing white blood cells again. In some cases, the production of white blood cells can be accelerated by giving drugs containing growth factors. This is determined by the treating physician.

Anti-infection measures

Because the child is more vulnerable to infections throughout the treatment period, all hospitals have some advice and guidelines for patients and families to reduce the risk of infections. In general, the family is advised to be extra careful with regard to infection, i.e., to prevent the child coming into direct contact with people who have infectious conditions such as colds, stomach flus, coughs, or the like. Each family will need to speak to their doctor, as recommendations vary somewhat from hospital to hospital.

Nausea

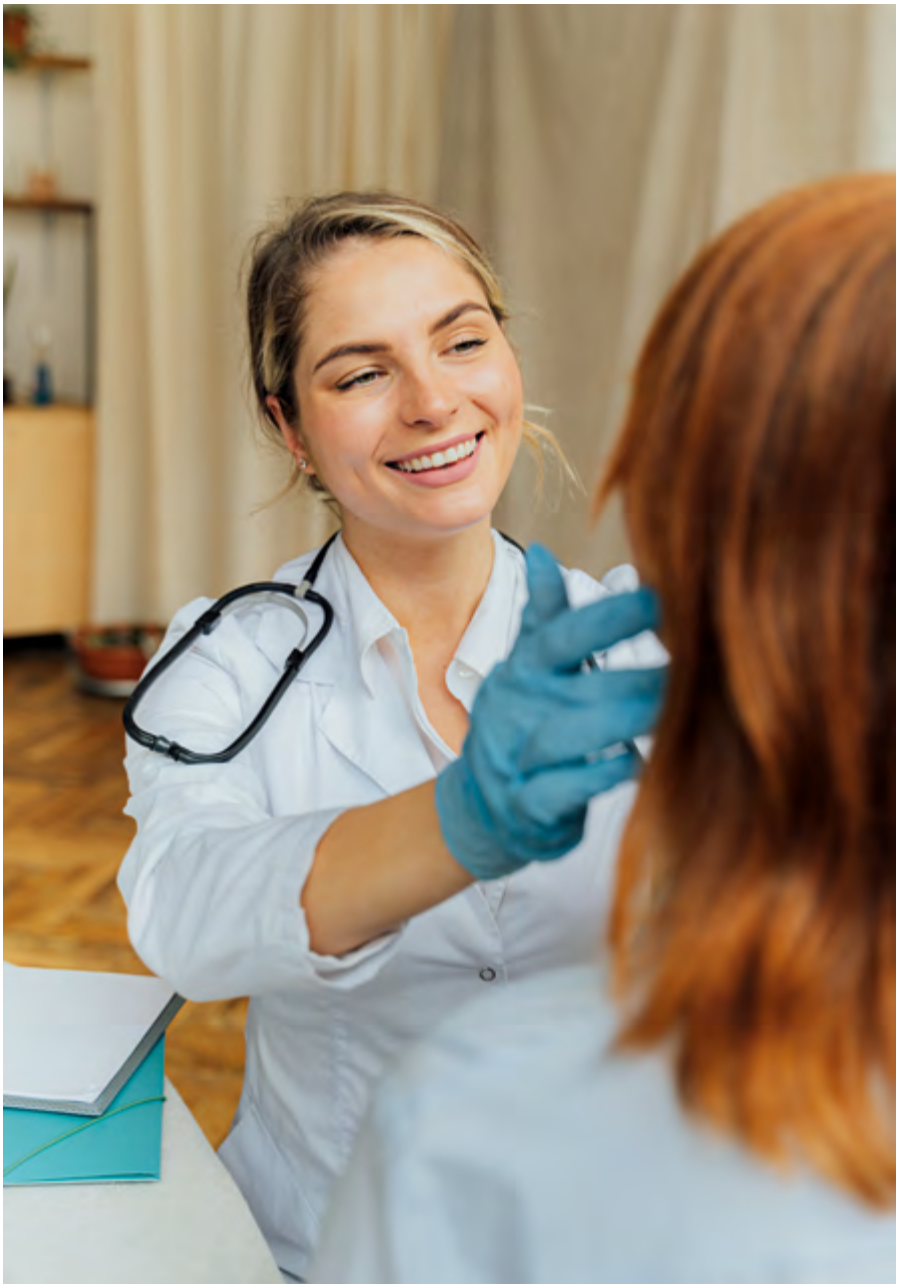
Nausea is common during chemotherapy. It depends on the type of chemotherapy the child receives, and is also very individual. It is important to take the child's nausea seriously and try different medicines if satisfactory relief is not obtained at first. Today there are very effective anti-nausea medications, so most people should be able to go through the treatment without too many problems with nausea.

Reduced appetite

Reduced appetite is a common problem and is caused by nausea, altered sense of taste, mucosal sores and psychosocial factors. Children need proper nutrition to ensure growth and development. Early consideration should be given to nutritional drinks, tube feeding and possibly the insertion of a "button" (percutangastrostomy PEG) in the stomach where the child can be fed. The clinical nutritionist at the hospital can be of great help with this. In a few cases, intravenous nutrition may be necessary.

→ **See our nutrition brochure for more advice.**

The development of cancer in children is not related to pregnancy, and it is definitely not because their parents have done something wrong.



Follow-up after completion of treatment

There is close and careful follow-up by the hospital throughout the treatment period. A regional hospital has the main responsibility for investigation and treatment, while much of the treatment itself can be done at the local hospital.

After completion of treatment, the child will be followed up according to specific guidelines. The follow-up consists of a clinical examination, blood tests and often some imaging examinations, such as ultrasound, X-ray or MRI.

Check-up frequency varies according to the type of tumour, and they are more frequent in the first few years, often every 1–3 months in the first and second year. The purpose of the check-ups is to look for signs of relapse, and for signs of late effects. They ensure that the patient and family can cope with the disease, side effects and late effects in the best possible way.

Late effects

The aim of cancer treatment is for the child to be cured from cancer. Unfortunately, often treatment has to be so intense that there is a risk of late effects. This also applies to solid tumours.

It is difficult to say at the start of treatment what late effects your child might experience. This depends on age, type and site of the tumour, type of chemotherapy, use of radiation therapy, and host factors and more. Many patients experience no late effects, but more than half will get at least one significant late effect.

The main late effects after treatment for solid tumours are the following:

- In several types of tumours, the chemotherapy group anthracyclines (such as doxorubicin) are given. These drugs can affect the heart muscle, both during treatment and for many years afterwards, causing heart failure. This is carefully monitored throughout the treatment period and must also be checked at regular intervals during follow-up. The cardiac follow-up will depend on the total dose of anthracyclines the child received during the treatment.

- Some chemotherapy drugs (especially cisplatin) can cause hearing damage. During the treatment, hearing tests are done and if there are signs of incipient damage, the treatment is adjusted to prevent further damage.

- Radiation therapy can lead to poor skeletal growth of the irradiated area. It can also cause hormonal changes due to irradiation of the pituitary gland or other hormonal glands. Radiation therapy also carries an increased risk of new cancers in the irradiated area.

- Fertility may be affected by chemotherapy and radiation therapy. In certain cases where it is possible, the possibility of freezing sperm for boys will be assessed. The possibility of sperm banking depends on the age and sexual maturity of the male. For girls, in some cancer diagnoses, ovarian tissue freezing is an option. The vast majority, both girls and boys, retain their fertility if they have not undergone a stem cell transplant.

→ **See our late effects brochure for more information.**

It is difficult to say at the start of treatment what late effects your child might experience.

The Nordic co-operation (Det nordiske samarbeidet)

There is close co-operation between the Nordic countries in childhood cancer treatment, and all children and adolescents up to the age of 18 are treated according to the same treatment protocols in the Nordic countries.

A treatment protocol is the “recipe” for how the treatment should be given to the child. The protocol starts on day 1 of the treatment, and states what is included in the treatment every day and week throughout the treatment. The protocol tells which chemotherapy drugs to give when. The doctors responsible for the treatment will go through this list with the family at regular intervals.

About the Norwegian Childhood Cancer Society

The Norwegian Childhood Cancer Society is a voluntary and nationwide organisation.

Our office is in Oslo, and we have county associations run by families who have or have had children with cancer. The associations work for the families on a voluntary basis. Our goal is that no child should die of cancer.

The Norwegian Childhood Cancer Society exists to help children and adolescents with cancer and their families. We are there for the whole family, meaning that the sick child, siblings and parents are all included. Some of the sick children have recovered, some are living with symptoms, some are under treatment, while others we have unfortunately lost.

At the hospitals, our peer contacts organise parents' meetings with the opportunity for new families to talk to someone. When your child is diagnosed with something as serious as cancer, it can be good to have someone to talk to who has experienced what you have. We also provide positive experiences for children who have to stay in the hospital for long periods.

The Norwegian Childhood Cancer Society wants to be the largest driving force in Norway to focus on childhood cancer in the media and society.

We also contribute to research and education to combat childhood cancer.

Become a member

→ Register on barnekreftforeningen.no

What does membership of the Norwegian Childhood Cancer Society mean?

As a member you have a unique ability to have an impact on conditions for children and adolescents with cancer and their families.

Membership in the Norwegian Childhood Cancer Society means access to a community of families who are, or have been, in the same situation. The association provides information, advice and support at all stages of a child who has, or has had, cancer.

TIPS

Contact your county association or the staff of the Childhood Cancer Society and tell us what is important to you.

Contact

If you have any questions about membership or the Norwegian Childhood Cancer Society, please contact us.

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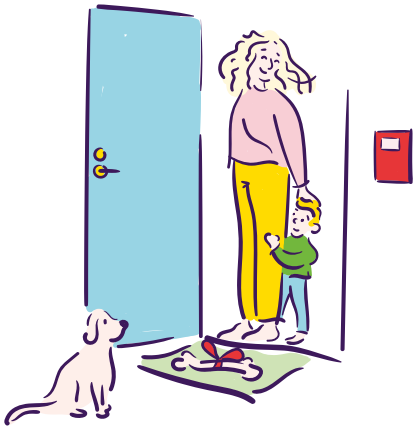

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