



About Neuroblastoma

Overview

If your child has been diagnosed with neuroblastoma or you are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- [What Is Neuroblastoma?](#)

Research and Statistics

See the latest estimates for new cases of neuroblastoma in the US and what research is currently being done.

- [What Are the Key Statistics About Neuroblastoma?](#)
- [What's New in Neuroblastoma Research and Treatment?](#)

What Are the Differences Between Cancers in Adults and Children?

The types of cancers that develop in children are often different from the types that develop in adults. Childhood cancers are often the result of DNA changes in cells that take place very early in life, sometimes even before birth. Unlike many cancers in adults, childhood cancers are not strongly linked to lifestyle or environmental risk factors.

There are exceptions, but childhood cancers tend to respond better to treatments such as chemotherapy. Children's bodies also tend to tolerate chemotherapy better than adults' bodies do. But cancer treatments such as chemotherapy and radiation therapy can have some long-term side effects, so children who survive their cancer need careful attention for the rest of their lives.

Since the 1960s, most children and teens with cancer have been treated at specialized centers designed for them. Being treated in these centers offers the advantage of a team of specialists who know the differences between adult and childhood cancers, as well as the unique needs of children with cancer and their families. This team usually includes pediatric oncologists, surgeons, radiation oncologists, pathologists, pediatric oncology nurses, and nurse practitioners.

These centers also have psychologists, social workers, child life specialists, nutritionists, rehabilitation and physical therapists, and educators who can support and educate the entire family.

Most children with cancer in the United States are treated at a center that is a member of the Children's Oncology Group (COG). All of these centers are associated with a university or children's hospital. As we have learned more about treating childhood cancer, it has become even more important that treatment be given by experts in this area.

Any time a child is diagnosed with cancer, it affects every family member and nearly every aspect of the family's life. You can read more about coping with all these changes in our documents about children with cancer.

- [References](#)

[See all references for Neuroblastoma](#)

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What Is Neuroblastoma?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see [What Is Cancer?](#)

Neuroblastoma is a type of cancer that starts in certain very early forms of nerve cells found in an embryo or fetus. (The term *neuro* refers to nerves, while *blastoma* refers to a cancer that affects immature or developing cells). This type of cancer occurs most

often in infants and young children. It is rarely found in children older than 10 years.

To understand neuroblastoma, it helps to know about the sympathetic nervous system, which is where these tumors start.

About the sympathetic nervous system

The nervous system consists of the brain, spinal cord, and the nerves that reach out from them to all areas of the body. The nervous system is essential for thinking, sensation, and movement, among other things.

Part of the nervous system also controls body functions we are rarely aware of, such as heart rate, breathing, blood pressure, digestion, and other functions. This part of the nervous system is known as the *autonomic nervous system*.

The *sympathetic nervous system* is part of the autonomic nervous system. It includes:

- Nerve fibers that run along either side the spinal cord.
- Clusters of nerve cells called *ganglia* (plural of ganglion) at certain points along the path of the nerve fibers.
- Nerve-like cells found in the medulla (center) of the adrenal glands. The adrenals are small glands that sit on top of each kidney. These glands make hormones (such as adrenaline [epinephrine]) that help control heart rate, blood pressure, blood sugar, and how the body reacts to stress.

The main cells that make up the nervous system are called *nerve cells* or *neurons*. These cells interact with other types of cells in the body by releasing tiny amounts of chemicals (hormones). This is important, because neuroblastoma cells often release certain hormones that can cause symptoms (see the section, [Signs and Symptoms of Neuroblastoma](#)).

Neuroblastomas

Neuroblastomas are cancers that start in early nerve cells (called *neuroblasts*) of the sympathetic nervous system, so they can be found anywhere along this system.

A little more than 1 out of 3 neuroblastomas start in the adrenal glands. About 1 out of 4 begin in sympathetic nerve ganglia in the abdomen. Most of the rest start in sympathetic ganglia near the spine in the chest or neck, or in the pelvis.

Rarely, a neuroblastoma has spread so widely by the time it is found that doctors can't tell exactly where it started.

There is a wide range in how neuroblastomas behave. Some grow and spread quickly, while others grow slowly. Sometimes, in very young children, the cancer cells die for no reason and the tumor goes away on its own. In other cases, the cells sometimes mature on their own into normal ganglion cells and stop dividing. This makes the tumor a ganglioneuroma (see below).

Other autonomic nervous system tumors in children

Not all childhood autonomic nervous system tumors are malignant (cancerous).

Ganglioneuroma is a benign (non-cancerous) tumor made up of mature ganglion and nerve sheath cells.

Ganglioneuroblastoma is a tumor that has both malignant and benign parts. It contains neuroblasts (immature nerve cells) that can grow and spread abnormally, similar to neuroblastoma, as well as areas of more mature tissue that are similar to ganglioneuroma.

Ganglioneuromas are usually removed by surgery and looked at carefully under a microscope to be sure they don't have areas of malignant cells (which would make the tumor a ganglioneuroblastoma). If the final diagnosis is ganglioneuroma, no other treatment is needed. If it's found to be a ganglioneuroblastoma, it's treated the same as a neuroblastoma.

- [References](#)

[See all references for Neuroblastoma](#)

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What Are the Key Statistics About Neuroblastoma?

Neuroblastoma is by far the most common cancer in infants (less than 1 year old). It

accounts for about 6% of all cancers in children. There are about 700 new cases of neuroblastoma each year in the United States. This number has remained about the same for many years.

The average age of children when they are diagnosed is about 1 to 2 years. In rare cases, neuroblastoma is detected by ultrasound even before birth. Nearly 90% of cases are diagnosed by age 5. Neuroblastoma is very rare in people over the age of 10 years.

In about 2 of 3 cases, the disease has already spread to the lymph nodes or to other parts of the body when it is diagnosed.

Statistics related to survival are discussed in the section [Survival Rates for Neuroblastoma Based on Risk Groups](#).

Visit the [American Cancer Society's Cancer Statistics Center](#) for more key statistics.

- [References](#)

[See all references for Neuroblastoma](#)

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What's New in Neuroblastoma Research and Treatment?

Important research into neuroblastoma is under way right now in many university hospitals, medical centers, and other institutions around the world. Each year, scientists find out more about what causes the disease and how to improve treatment.

Genetics of neuroblastomas

Researchers now have better lab tests to look for changes in the genes of neuroblastoma cells. They have made a great deal of progress in recent years in figuring out which neuroblastomas are likely to be cured with standard treatment, and

which will need more aggressive treatment.

For example, using newer lab tests, researchers have found that certain DNA changes on the short arm of chromosome 6 (6p22) are more likely to be seen in neuroblastomas that grow more aggressively. More recently, researchers have found that neuroblastoma cells in older children are more likely to have changes in the *ATRX* tumor suppressor gene. Tumors with this gene change tend to grow more slowly, but they are also harder to cure. This may help explain why younger children with neuroblastoma tend to do better in the long term than children who are older when they are diagnosed.

Doctors are now looking to use these and other findings to help choose the best treatments. Newer [staging systems](#) and [risk group classifications](#), which take advantage of some of these findings, should be in use within the next few years.

Treatment

Survival rates for neuroblastoma have gotten better as doctors have found ways to improve on current treatments.

Chemotherapy

Doctors continue to search for the best combinations of chemotherapy drugs to treat neuroblastoma.

Several chemotherapy drugs that are already used to treat other cancers, such as topotecan, irinotecan, and temozolomide, are now being studied for use against neuroblastoma.

Other studies are looking to see if children with low or intermediate risk neuroblastoma can be treated with less (or even no) chemotherapy. The goal is to still have the same good results, but with fewer side effects from treatment.

Stem cell transplants

Doctors are also trying to improve the success rate with high-dose chemotherapy and stem cell transplants, using different combinations of chemotherapy, radiation therapy, retinoids, and other treatments. Some clinical trials are studying the use of more than one stem cell transplant in the same patient (known as a *tandem transplant*). Others are looking to see if using stem cells donated from another person (an *allogeneic stem cell*

transplant) might help some children with hard-to-treat tumors.

Retinoids

Retinoids such as 13-cis-retinoic acid (isotretinoin) have reduced the risk of recurrence after treatment in children with high-risk neuroblastoma. Newer, potentially more effective retinoids, such as fenretinide, are now being studied in clinical trials.

Targeted drugs

Knowledge about what makes neuroblastoma cells different from normal cells may lead to new approaches to treating this disease. Newer drugs that target neuroblastoma cells more specifically than standard chemo drugs are now being studied in clinical trials. For example, doctors are now studying medicines that target the pathways inside neuroblastoma cells that help them grow, such as crizotinib (Xalkori) for the ALK pathway and alisertib (MLN8237) for the aurora A pathway.

Crizotinib is a drug that targets cells with changes in the *ALK* gene. Up to 15% of neuroblastomas have changes in this gene. In an early study, crizotinib was found to cause some neuroblastomas to shrink, although it's not clear how long this might last.

Some other drugs that work differently from standard chemo drugs are being studied against neuroblastoma as well. Examples include bortezomib, vorinostat, lenalidomide, temsirolimus, sorafenib, nifurtimox, and lestaurtinib.

Immunotherapy

Immunotherapy is the use of medicines to help a patient's own immune system fight cancer.

The [monoclonal antibody](#) dinutuximab (Unituxin), which targets GD2 on neuroblastoma cells, is now used routinely for children with high-risk neuroblastoma, to help immune system cells find and destroy the cancer cells. Clinical trials are now testing the effectiveness of several other antibodies that target GD2. One example is hu14.18-IL2, an antibody that is linked to interleukin-2 (an immune-boosting cytokine). Early results have found that this antibody/cytokine combination may help some children in whom other treatments are no longer working.

Several cancer vaccines are also being studied for use against neuroblastoma. For these vaccines, modified neuroblastoma cells or other substances are injected into the

body to try to get the child's own immune system to attack cancer cells. These treatments are still in the early stages of clinical trials.

- [References](#)

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