Neuroblastoma is a form of cancer that is made up of cells that are found in nerve tissues of the body. These cells are called neuroblasts, and they are early nerve cells found most commonly in the adrenal glands (near the kidneys), and along the tissues around the spinal cord in the neck, chest, abdomen and pelvis. Many neuroblastomas start in the adrenal glands.

Neuroblastoma occurs most commonly in infants and children under 5 years of age, and rarely in children over 10 years of age. Very rarely, neuroblastoma occurs in adults.

Risk factors

A risk factor is anything that increases a person's chance of developing a certain condition or disease, such as cancer. In adults, lifestyle and environmental factors (such as smoking or exposure to certain chemicals) can be significant risk factors for developing certain types of cancer. In children, very few risk factors have been identified that increase the chance of developing cancer. For most children with cancer, the underlying cause is unknown.

Even if your child has a risk factor, it does not mean they will develop cancer. Many children with a risk factor will never develop cancer, while others with cancer may have had no known risk factors. Even if a child with a risk factor develops cancer, it is usually hard to know how much that risk factor contributed to the development of their disease.

The causes of neuroblastoma are not well understood, but factors associated with a higher chance of developing neuroblastoma include the following:

Family history

In rare cases (around 1% to 2% of children with neuroblastoma), neuroblastoma runs in families. When this happens, the cancer usually develops at a younger age than in children with no family history of the disease, and often in multiple parts of the body.

Knowledge of your family's history relating to cancer may change over time.

Genetic conditions

A small number of children have genetic or inherited risk factors that increase their chance of developing cancer during childhood. These genetic conditions can run in families, or can start in the child from a change in their DNA. Neuroblastoma that runs in families tends to be associated with faulty *ALK* or *PHOX2B* genes.

Certain genetic disorders can increase a child's risk of developing neuroblastoma. These include:

- Hirschsprung disease

- congenital central hypoventilation syndrome
- neurofibromatosis type 1.

If your child is diagnosed with one of these genetic conditions, they will need specific follow-up. The health care team will advise which ongoing tests your child will need.

Cancers in children that are linked to genetic conditions may also affect the risk for other family members. Speak to your child's treatment team to see whether genetic counselling is recommended for you or your family.

For more information about genetic conditions, see the children's cancer glossary or the Centre for Genetics Education.

Symptoms

Most symptoms of neuroblastoma are caused by growth of the cancer into nearby tissues or organs, or its spread to distant parts of the body such as bones or bone marrow. Symptoms may include:

- lump or swelling in the abdomen (belly), neck or chest, or under the skin
- stomach pain, feeling full or not wanting to eat, usually with weight loss
- swelling in the arms or legs (if a tumour is pressing on blood vessels or lymph vessels)
- problems going to the toilet (if the cancer grows into the bladder or bowel)
- bulging eyes, or dark circles around the eyes
- jerky, uncontrolled eye movements
- bone pain
- other unexplained pain
- weakness or paralysis (if the cancer is pressing on the nerves or spinal cord)
- trouble breathing or swallowing.

In some cases, neuroblastoma produces certain hormones that can cause diarrhoea, high blood pressure, rapid heartbeat, sweating and flushing of the skin.

Note about symptoms

Many conditions – including common childhood infections – can cause these symptoms, not just neuroblastoma. If your child has any of these symptoms and you are concerned, talk to your child's doctor.

Diagnosis

Your child will have a number of tests to investigate their symptoms and confirm a diagnosis of neuroblastoma, including:

- medical history and physical examination
- blood tests
- urine tests
- medical imaging, which may include:
    - X-ray
    - ultrasound
    - computed tomography (CT) scan
    - magnetic resonance imaging (MRI)
    - bone scan
    - metaidobenzylguanidine (MIBG) scan
    - positron emission tomography (PET) scan
- biopsy – where a small sample of the cancer is removed to be examined under a microscope. The sample can also be tested for genetic changes that can help determine the best type of treatment for your child
- bone marrow aspiration and biopsy – where a sample of bone marrow is taken with a small piece of bone to be examined under a microscope. This is done from 2 separate places in the body at the same time (called a bilateral bone marrow aspiration). Your child will probably have more than one of these tests.

These tests are explained in more detail in How is cancer diagnosed?.

The tests are designed to diagnose neuroblastoma, as well as determine the size of the tumour, its exact location in the body and whether it has spread to other parts of the body. The most common places it spreads to are the lymph nodes, liver, bones and bone marrow, and sometimes the skin.

Staging

If your child is diagnosed with neuroblastoma, some of the diagnostic tests will also help to stage the tumour. Staging determines where the tumour is, how big it is, which nearby organs are involved and whether the cancer has spread to other parts of the body. This is important to determine the outlook (prognosis) for your child, and to decide on the best options for treatment.

There are different ways of staging neuroblastoma – the following is the most commonly used:

Stage 1

The tumour is only in 1 part of the body, and all of the tumour that can be seen has been removed by surgery.

Stage 2

Stage 2 includes stages 2A and 2B:

- Stage 2A – the tumour is only in 1 part of the body, but not all of the tumour that can be seen has been removed by surgery
- Stage 2B – the tumour is only in 1 part of the body, and all of the tumour that can be seen has been removed by surgery. However, there are neuroblastoma cells in nearby lymph nodes.

Stage 3

Stage 3 involves 1 of the following:

- The tumour has not been completely removed by surgery and has spread to the other side of the body or to nearby lymph nodes
- The tumour is on 1 side of the body but has spread to lymph nodes on the other side of the body
- The tumour is in the middle of the body and cannot be completely removed by surgery. It has spread to tissues or lymph nodes on both sides of the body.

Stage 4

Stage 4 includes stages 4 and 4S:

- Stage 4 – the tumour has spread to other parts of the body such as distant lymph nodes, liver, bones or bone marrow, or the skin
- Stage 4S (also called special neuroblastoma) – the child is less than 1 year old. The tumour is only in 1 part of the body, and all the tumour that can be seen may be removed by surgery. However, the cancer has spread to the liver, skin or bone marrow, and maybe the lymph nodes near the tumour.

Treatment

Treatment and care of children with cancer is usually provided by a team of health professionals called a multidisciplinary team. Members of this team are specialists in children's cancers – they understand the differences between children's cancer and adult cancer, and each team member brings different skills in managing care to meet the needs of both you and your child.

The team will be led by a childhood cancer specialist (paediatric oncologist). Other members of the team depend on the age of your child and their type of disease, and may change over time as your

child's needs change. A list of team members who might make up the multidisciplinary team can be found in The treatment team.

Treatment for neuroblastoma depends on the age of your child, the stage of the disease, the biological features of the cancer and other factors identified during diagnosis. Treatment will be tailored to your child's particular situation, and may involve one or more of the following (see How is cancer treated for more detail).

Surgery

Your child is likely to have surgery to remove all or part of the tumour. If the entire tumour can be removed and it has not spread to other parts of the body, your child may not need any other treatment.

Chemotherapy

Chemotherapy uses anti-cancer medicines to destroy cancer cells. It is often given as a combination of medicines to try to prevent the cancer cells from becoming resistant to just one or two medicines.

Chemotherapy medicines are given together in courses, often over a few days. Once the body has recovered from the side effects, the next course is given. Most children receive multiple courses of chemotherapy.

Chemotherapy may be used:

- before surgery (to shrink the tumour and make it easier to remove)
- after surgery (to destroy any remaining cancer cells)
- as the main treatment (if the cancer has spread to other parts of the body, or if surgery is not a good option)
- to relieve symptoms of neuroblastoma.

Radiation therapy

Radiation therapy (also called radiotherapy) uses high-energy X-rays or other types of radiation to destroy cancer cells or stop them from growing. Radiation therapy is commonly used to treat children with high-risk neuroblastoma. If radiation therapy is included in your child's treatment, special care will be taken to reduce the risks.

Radiation therapy can be used to relieve symptoms of neuroblastoma, usually after other treatments have been tried.

Stem cell transplant

Some children with high-risk neuroblastoma may be treated using a stem cell transplant (also known as

a bone marrow transplant), in combination with high-dose chemotherapy or radiation therapy.

After the transplant, your child will be given additional treatment known as maintenance therapy to destroy any remaining cancer cells. Maintenance therapy is given for 6 months and may include:

- a vitamin-like medicine that slows the cancer's ability to make more cancer cells
- antibodies and other compounds to stimulate the body's immune system to destroy any remaining cancer cells.

Careful observation

For a few carefully selected patients, specific treatment may not be needed. If your child has a tumour that is not growing or spreading, they might be monitored closely but not given any treatment until they develop symptoms, or until their symptoms change. In some children (especially very young children), a neuroblastoma will disappear by itself, and no treatment is needed.

Support

Diagnosis of cancer in a child is a very difficult time for the child, their family and their friends. You might feel overwhelmed, scared, anxious or angry. These are all normal feelings. It is very important to seek support from family, friends, health professionals or other services to help you, your child and your family cope with cancer.

Talk to your child's treatment team if you are having difficulties coping.

Living with children's cancer has information about physical, emotional and practical issues during and after diagnosis and treatment. There is also a page with helpful links on where to find support.

The Cancer Council in your state or territory can give you general information about cancer, as well as information on resources and support groups in your local area. Call the Cancer Council Helpline from anywhere in Australia for the cost of a local call on **13 11 20** .

For additional specific information about childhood cancer, contact any of the major children's hospitals and networks in your state or territory.

Chance of cure

Many children with cancer are cured of the disease. Children's bodies have great capacity for healing. Also, huge improvements have been made in the treatment of childhood cancer in the past few decades. In the 1980s, around 65% of children diagnosed with cancer were alive more than 5 years after their diagnosis. Today, around 83% of children are successfully treated and become long-term survivors.

Long-term survival (also called the outlook or prognosis) and treatment options depend on a range of factors, including:

- age of your child at diagnosis
- extent or stage of the cancer
- appearance of the cancer cells under the microscope (the shape, function and structure of the cells)
- how the cancer responds to treatment
- cancer or tumour biology, which includes
    - the patterns of the cancer cells
    - how different the cancer cells are from normal cells
    - how fast the cancer cells are growing.

Talk to your child's doctor about your child's individual disease, treatment options and outlook.

Clinical trials

Researchers are trialling new ways to diagnose and treat different types of cancer. Your child may be invited to be part of a clinical trial to test new ways of treating neuroblastoma.

New treatments have to go through very strict regulation and approval processes before they can be used in a clinical trial. Your child's doctor will explain everything about the trial and give you detailed written information. You will need to give special permission for your child to be part of the trial.

Participating in a clinical trial may or may not directly benefit your child, but the results of clinical trials today will help children with cancer in the future.

See Clinical trials and research for more information, including whether there are any clinical trials your child can join.

More information

For more information about neuroblastoma, see:

- Neuroblastoma, from the American Cancer Society
- Neuroblastoma treatment (PDQ®), from the National Cancer Institute (United States).