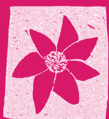
A parent's guide to

Neuroblastoma

Information and support for when your child is diagnosed with a type of cancer called neuroblastoma.



Neuroblastoma
AUSTRALIA

This Parent Leaflet has been based on the Parent Handbook created by Neuroblastoma UK and the Children's Cancer and Leukaemia Group in 2020 (visit www.neuroblastoma.org.uk) which kindly provided permission to Neuroblastoma Australia to use. Information has been adjusted for Australia and this has been reviewed by Dr. Toby Trahair (Paediatric Oncologist at Sydney Children's Hospital).

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About this guide

This guide has been written with the help of experts and parents whose children have had neuroblastoma. It aims to help parents and carers following their child's diagnosis and gives specific information about neuroblastoma, its treatment and possible side effects.

If your child has only just been diagnosed, there may be information in this booklet that has not yet been discussed with you by the medical team looking after your child. This guide provides a reference to the information you have already been given, or will be given in the future, and will act as a helpful reminder for any discussions you have with your child's treatment team.

This guide is a general overview of neuroblastoma. It is important to remember that every child is an individual and your child's specific diagnosis must always be discussed with the treatment team caring for them.

We hope it helps answer your questions so that you understand more about neuroblastoma and its treatment.

We strongly recommend this guide is read in conjunction with information provided to you by your treating oncologist and hospital.

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Beginning the journey

When you are told that your child has neuroblastoma, it can feel as if your life has turned upside down overnight. You may be feeling numb, scared, not believing that any of this is really happening, angry as to why this is happening to your child and maybe feeling guilty that your child's cancer is because of something you have or haven't done even though this isn't true. All of these feelings are completely normal and many parents say that they felt the same.

Since your child's diagnosis, you may have met many new people, heard a lot of unfamiliar medical terms and your child may have undergone a series of tests. This can feel very overwhelming and daunting as a parent. Don't worry, hospital staff fully understand that it takes time for you to digest what is happening and what the next steps might be. They are there to help you through this difficult time with information and reassurance.

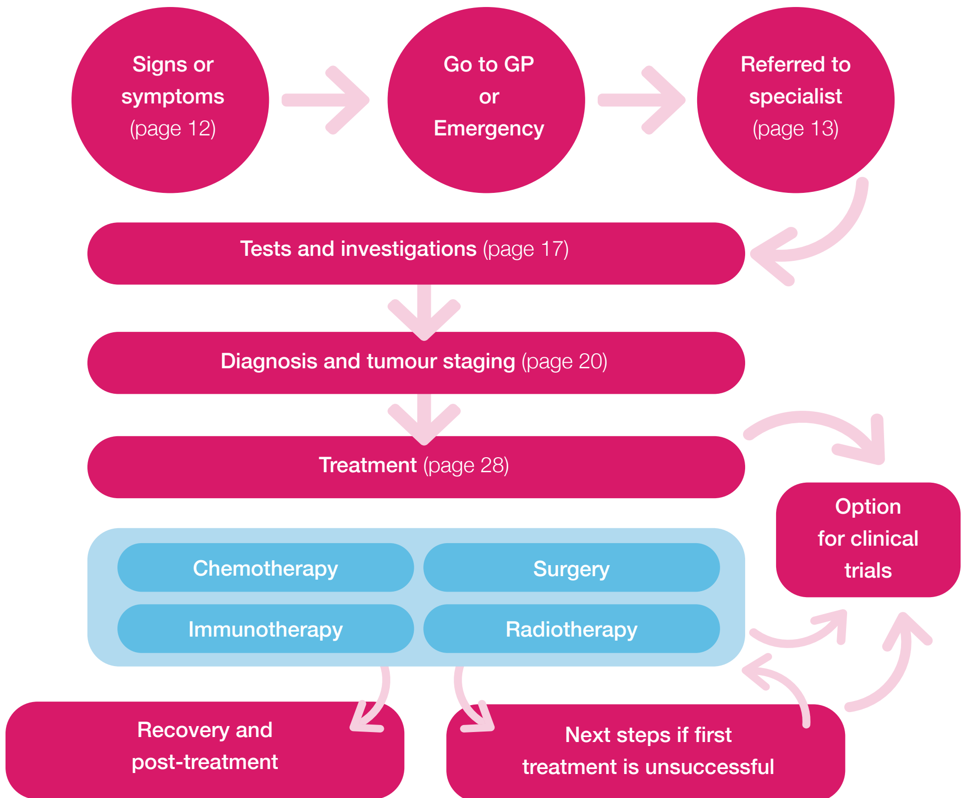
Many parents cannot think of any questions to ask during a hospital consultation but think of all sorts of things as soon as they get home. It is a good idea to write down questions as soon as you think of them so that you can discuss them at the next opportunity.

When a child is diagnosed with cancer, it has a huge impact on your whole family. Your child's routine is likely to change with hospital stays and regular appointments and this can feel overwhelming for you, your child and the rest of your family. There are many support organisations who can help you through this time and some of these organisations are listed at the end of this guide, but you should also discuss your feelings with the team looking after your child.

TOP TIP

Write down your questions at the back of this guide and fill in the answers during your clinic appointments.

Neuroblastoma – your child's healthcare journey



Neuroblastoma

Neuroblastoma is a cancer that is almost always found in children. Childhood cancers are usually different from cancers affecting adults. They tend to happen in different parts of the body to adult cancers and behave differently. Cancers in children also respond differently to treatment.

What is neuroblastoma?

It is the second most common solid tumour in children after brain tumours.

Around 50 children in Australia are diagnosed each year with neuroblastoma, making up about 6% of the total number of childhood cancer diagnoses.

It primarily affects younger children and is the most common solid tumour in infants under the age of one, making up around a fifth (22%) of all cancers diagnosed at this age. It is rare for children to be diagnosed with neuroblastoma over the age of five. Only 2% of neuroblastomas are diagnosed in children over the age of 10 years and 0.5% in those over 15 years old.[1]

Neuroblastoma is classified as an **embryonal tumour**, a type of cancer that develops from the cells left behind from a baby's development in the womb. The cells from which it develops are specific cells in the nervous system called neuroblasts, giving neuroblastoma its name:

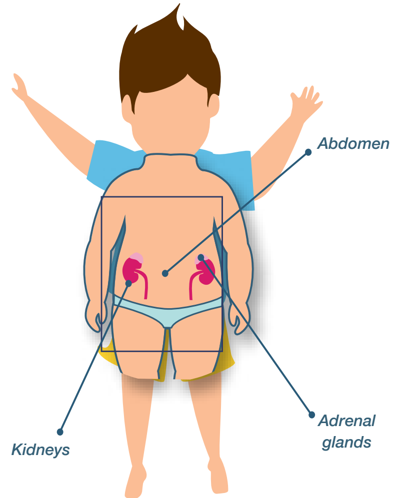
- 'neuro' means nerve
- 'blast' means cells in an early stage of development
- 'oma' means a group of cells, or a tumour

1 Cancer Research UK. (2014). Children's cancer incidence statistics. [ONLINE] Available at: www.cancerresearchuk.org/health-professional/cancer-statistics/childrens-cancers/incidence#heading-Ten. Last accessed 01/04/2019.

Neuroblastoma can happen anywhere in the body. The most common place for the tumour to grow is in the abdomen.

Some tumours grow at the back of the chest and occasionally even higher up towards the neck. About 50% of tumours start in the adrenal glands, which are found above the kidneys. These glands release hormones such as adrenaline, to maintain blood pressure and enable us to respond to stress.

In some cases, neuroblastoma can spread to tissues beyond the place it started, such as the bone marrow, bone, lymph nodes, liver and skin.



What causes neuroblastoma?

As with most childhood cancers, the cause of neuroblastoma is unknown. It is not infectious and cannot be passed on to other people. Around 1% of neuroblastoma cases are hereditary, most commonly due to genetic changes in DNA present in a gene called anaplastic lymphomakinase (ALK) or a gene called Phox2B. These genetic changes happen in all cells as well as cancer cells and can be passed on through families.

Hereditary neuroblastoma often appears in patients who are younger than 18 months old, with more than one primary tumour. In the situation where two or more family members have neuroblastoma, screening of other family members with genetic testing for the above genes, urine testing and ultrasound examination is recommended and will be discussed with you, together with referral to a clinical geneticist.

What are the signs and symptoms?

Many children with neuroblastoma have little in the way of symptoms. Perhaps they have seemed unwell for a little while or have not been eating much, complaining of vague aches and pains or unexplained sweating. Unless a parent or doctor feels a lump while bathing, dressing or examining the child, a diagnosis of neuroblastoma may not be initially considered.

It is possible that many of the symptoms your child has are similar to those of more common, less serious childhood illnesses. Many parents wonder if they or a doctor should have noticed something sooner, but because this is such an uncommon disease, neuroblastoma is rarely suspected if only fairly vague symptoms are present.

Neuroblastoma, particularly 'high risk' neuroblastoma, often shows itself at a late stage when it has spread around the body.

Urgent referrals

Urgent referral for specialist assessment by a paediatric oncologist is recommended for any child with a palpable abdominal mass (a lump in their tummy) or an unexplained enlarged abdominal organ.

Diagnosis

If cancer is suspected, your child will be referred by your GP, paediatrician or Emergency Department staff to your nearest children’s hospital which has a specialist children’s cancer (called paediatric oncology) centre. These centres are located in a network of 9 hospitals in Australia with expertise in managing and treating all childhood cancers, including neuroblastoma.

Childhood Cancer Centres in Australia

1. Sydney Children’s Hospital
2. The Children’s Hospital at Westmead
3. Queensland Children’s Hospital, Brisbane
4. The Royal Children’s Hospital, Melbourne
5. Monash Children’s Hospital, Melbourne
6. Women and Children’s Hospital, Adelaide
7. John Hunter Children’s Hospital, Newcastle
8. Perth Children’s Hospital
9. Royal Hobart Hospital, Tasmania

Your child will undergo many different tests in order to confirm whether your child has cancer, what type and which treatment will be best.

Tests, assessments and scans

Your child's medical team will run a whole range of tests, assessments and scans to get an accurate diagnosis. All of these may seem frightening to both you and your child, but it is important for doctors to see where the cancer is in your child's body, whether it has spread, and to assess the general health of your child. This will then help your child's doctor to decide which treatment will be best. It may take a couple of weeks for all the test results to come back.

Tumour and bone marrow biopsy

A biopsy involves removing some of the cells from your child's tumour to look at them under a microscope. This is done during an operation where your child is given a general anaesthetic, and a piece of the tumour is taken out through a small cut (or incision) in the skin. Sometimes, a small piece of tumour may be drawn up through a needle; this procedure is known as a 'needle biopsy'.

The tumour sample is then sent to a laboratory to find out if the tumour is made up of neuroblastoma cells and to look at the biology of the neuroblastoma by looking at DNA changes and biological markers.

Knowing about your child's tumour biology gives important information that is used in deciding the best treatment for your child.

Blood tests

Blood for testing may be taken from a vein in your child's arm or by a finger prick. This gives important information about your child's current health, blood group, and any infections. It also acts as a good way of monitoring the side effects of treatment. Blood tests are common throughout cancer treatment and the clinical team can apply local anaesthetic cream to the skin to make the needle less painful.

Urine tests (Urine Catecholamines)

A simple specialised test in the diagnosis of neuroblastoma measures 'vanillylmandelic acid' (VMA) in your child's urine. You may hear this test referred to as either the 'VMA' or 'urine catecholamine' test. VMA is a chemical found in the urine in raised amounts when a child has neuroblastoma, and this is a good indicator of diagnosis. Sometimes a similar marker called 'homovanillic acid' (HVA) is also measured. These urinary markers are raised in nine out of 10 cases of children with neuroblastoma. As these VMA/HVA chemicals are produced by the tumour, as well as being useful to help with diagnosis, they can be used to measure tumour activity during treatment, so are sometimes referred to as tumour markers.

Scans and x-rays

Your child will have a number of x-rays and specialist scans to confirm the diagnosis of neuroblastoma. The scans will show where the main neuroblastoma ('primary tumour') is in their body, and if there are neuroblastoma cells which have spread to other parts of their body ('secondary tumours' or 'metastases').

Apart from the need for an injection of 'contrast' during some CT scans, or the injection of radioactive liquid for a bone scan or mIBG scan, none of these investigations are painful to your child. However, it is appreciated that they may feel unsettled or frightened. Some of the scans require that your child remains still for quite some time and to assist in this a general anaesthetic (where your child would be asleep) may be needed to have some of the tests. These will be discussed with you prior to any scans being done.

X-rays, Ultrasounds, CT and MRI scans

X-rays may be used to see if neuroblastoma has spread to certain bones. The ultrasound will be a familiar procedure to all mothers who had this performed during their pregnancy. The sound waves produced by the ultrasound can be recorded on a screen and outline normal organs and tumours inside the body. The CT or MRI scanner takes multiple x-ray images to build up a 3D picture inside the body. The MRI (magnetic resonance imaging) uses magnetic fields to build up a detailed picture of a part of the body, and no radiation is used. MRI takes longer than a CT scan and is quite noisy. Your child may be given a sedative or general anaesthetic to make sure that they lie still.

mIBG scans

This type of scan looks for abnormal cell growth within in the body. It involves injecting a type of isotope called meta-iodobenzylguanidine (mIBG) into your child's veins which then travels around your child's body and is naturally absorbed by neuroblastoma cells. mIBG contains a harmless level of radioactive material so when mIBG builds up in the neuroblastoma cells, a scanner called a gamma camera can detect them as 'hot spots'. mIBG scanning is useful at diagnosis as it gives a complete picture of where the tumour cells are in the body. mIBG shows up in neuroblastoma cells in around 90% of cases. mIGB scans can be used to monitor treatment response. Where neuroblastoma doesn't take up mIGB, a different scan, the FDG-PET CT scan, can be used to monitor neuroblastoma.

FDG-PET scans

This is another type of radionuclide scan, like an mIBG scan, which can be useful to see where neuroblastoma has spread to, particularly in cases where the mIBG scan is negative (around 10% of cases). In some children, whose mIBG scan does not detect neuroblastoma tumours, the FDG-PET scan can be used to assess how your child may be responding to treatment. FDG stands for 'fluorodeoxyglucose' and PET stands for 'positron emission tomography'.

Bone scans

Bone scans can show how much the cancer has spread to the bones. It involves injecting a small dose of radioactive liquid into a vein, usually in the hand or arm. The substance is absorbed by the bone but affected areas will absorb more so they will be highlighted on the scanner as 'hot spots'. Bone scans are only used if mIBG scans are negative. Bone scans are less commonly done than previously.

Bones and bone marrow

The most common places which neuroblastoma cells spread to, are the bones and the bone marrow. To detect tumour cells in the bone, your child will undergo either an mIBG, PET-CT or bone scan (see above).

To examine your child's bone marrow (the spongy material in the middle of a bone), a needle is inserted into one of the larger bones (like the hip bone) and a small quantity of bone marrow is drawn out. This is called an aspirate. A trephine (a core of the bone marrow) involves taking a very small piece of bone at the place where the marrow is drawn out.

To make sure that the test is as accurate as possible, aspirates and trephines may be taken from more than one location, usually from the hip bones on either side of the body. Your child will always be given a general anaesthetic before these tests are undertaken.

Genetic tests

One important biological marker to test for in neuroblastoma cases is the gene called MYCN (pronounced 'mikken'). When additional numbers of copies of this gene are present, this is called MYCN amplification and happens in around 25% of neuroblastoma cases. It is more common in younger children and it indicates that it is a more aggressive cancer that is likely to spread. If your child has MYCN amplification then we know that more intensive treatment is needed from the start.

Another genetic test involves looking at the number of each chromosome; a healthy cell should have two copies of 23 chromosomes. In neuroblastoma, there may be gains or losses of all or parts of chromosomes. This test is particularly important for deciding treatment in children with low- and medium-risk neuroblastoma. Children who have changes to whole chromosomes (gains or losses of whole chromosomes) are less likely to require treatment than those with changes to parts of chromosomes (called segmental chromosomal abnormalities).

Recently mutations and amplifications in the gene ALK (anaplastic lymphoma kinase) have also been shown to present in 10-15% of high-risk neuroblastoma tumours at diagnosis. Some clinical trials are looking to see if treatment with an ALK inhibiting drug can improve outcomes.

The results of these genetic tests will help to determine the type of treatment your child has. The MYCN test can be done very quickly but other genetic tests looking at all the chromosomes can take slightly longer to do, and the results may take a few weeks to come back to your child's doctor. Most genetic tests are carried out at specialist laboratories.

There is a small chance that some genetic tests will reveal that your child had an increased hereditary risk of getting neuroblastoma. This information will be shared with you when it is in the best interests of your child and will involve referral to a genetics specialist.

It will likely take more than a few days for all the tests to be completed and the results analysed. Undergoing these tests and waiting for results is an extremely anxious and stressful time for families. However, exact assessment of the extent of your child's tumour before beginning treatment is very important. The results found will allow your child's doctor to select the right type and length of treatment for your child.

Tumour 'staging'

When your child is diagnosed, you will also be told what stage the cancer is at. This considers the size of the tumour and whether it has spread beyond the part of the body where it started.

Doctors recognise several categories of neuroblastoma that are grouped into different 'stages' and 'risk groups'. The treatment your child has for their neuroblastoma depends on their tumour 'stage' and 'risk group'. Knowing the stage of your child's cancer helps doctors make sure your child is in the correct risk group so that they can give the right treatment and care.

Doctors use imaging tests such as CT or MRI scans to look for particular risk factors for surgery within a child's tumour. These factors can be a sign that the neuroblastoma is not suitable for surgery initially and may require chemotherapy to shrink it, or that it could simply be observed. These factors, known as image defined risk factors (IDRF), include whether the tumour is wrapped around an organ or blood vessels and other findings on a CT/MRI scan.

The International Neuroblastoma Risk Group (INRG) Staging System

The (INRG) Staging System is currently used to assess the tumours of individual children to match the right treatment with the biology of their tumour taking into account risk factors and possible side effects. The INRG staging system has replaced an older system called the International Neuroblastoma Staging System (INSS). The stages are explained opposite.

The INRG stages:

The stage is based on the results of imaging tests taken before surgery as well as the assessment of any Image Defined Risk Factors (IDRFs).

Stage L1: The tumor is located only in the area where it started; no IDRFs are found on imaging scans, such as a computed tomography (CT) or magnetic resonance imaging (MRI) scan.

Stage L2: The tumor has not spread beyond the area where it started and the nearby tissue; IDRFs are found on imaging scans, such as a CT or MRI scan.

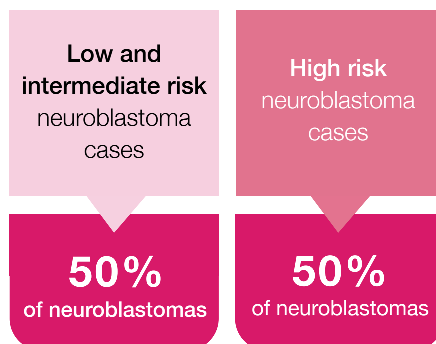
Stage M: The tumor has spread to other parts of the body (except stage MS; see below).

Stage MS: The tumor has spread to only the skin, liver, and/or bone marrow (with less than 10% bone marrow involvement) in a patient younger than 18 months.

The International Neuroblastoma Risk Group Risk Classification

Knowing your child's neuroblastoma risk group (INRG)

Treatment of neuroblastoma is tailored to each child and is given according to their risk group. Doctors categorise neuroblastoma according to the International Risk Group (INRG) Risk Classification as low- and intermediate- (medium), or high-risk. The risk classification takes into account factors such as the child's age at the time of diagnosis, stage of tumour (described above in the INRG Staging System), presence or absence of MYCN amplification, specific biological features of your child's tumour, specific genetic abnormalities, or if your child is showing severe clinical symptoms. The risk classifications will be explained on the following pages and you can also refer to a Risk Stratification Table on page 48.



Ganglioneuroblastoma

Ganglioneuroblastoma is another type of tumour which is closely related to neuroblastoma. It can be seen in any age group, but the cells of this tumour are more 'mature' than those of the ordinary neuroblastoma. When fully mature, they form a ganglioneuroma.

There are two types of ganglioneuroblastoma – intermixed and nodular. An intermixed ganglioneurobastoma is a benign (non-harmful) tumour which is either INRG L1 or L2 and, if it cannot safely be removed by an operation, it can often simply be observed.

A nodular ganglioneuroblastoma can be INRG L1, L2, or M. The nodules of neuroblastoma may break away from the main tumour and in these cases (M disease) will need chemotherapy and the other treatments given to children with high-risk neuroblastoma.

Low-risk neuroblastoma

This group includes children with neuroblastoma with favourable genetics (no MYCN amplification and no segmental chromosome changes) and favourable pathology:

- **Infants under 18 months at diagnosis with localised neuroblastoma (stage L1)**
- **Infants under 18 months at diagnosis with localised inoperable neuroblastoma (stage L2)**
- **Infants under 18 months old with metastatic neuroblastoma (Stage MS or M) disease**
- **Any age child with localised (stage L1 or L2) maturing ganglioneuroma, or ganglioneuroblastoma intermixed**

Around 15 children are diagnosed with low-risk neuroblastoma in Australia each year and more than 90% are cured of their disease. Children with low-risk localised neuroblastoma who have symptoms which could present a risk to their health or whose tumours contain certain genetic changes, may require treatment with chemotherapy and possibly subsequent surgery. In these cases, treatment is with chemotherapy followed by re-assessment of the position and extent of the tumour. This may include CT or MRI scan, mIBG scan, urinary catecholamine measurement or FDG-PET scan before consideration of removing the tumour by surgery later.

In some cases, children with low-risk localised neuroblastoma that is not causing any symptoms and whose tumour shows favourable biology (such as lack of MYCN amplification and no segmental chromosomal abnormalities) may not require chemotherapy or surgery but can be closely monitored as most of these tumours (>60-80%) naturally decrease in size over weeks to months. A small number of children may require surgery or other treatment if there is an increase in the size of the tumour.

Stage MS neuroblastoma

This is a type of low-risk neuroblastoma with a particular distribution of secondary tumours which can be to skin, liver, bone marrow, distant lymph nodes but not bone, lung or the brain. Most infants with MS neuroblastoma do well but some young infants (under 2 months) sometimes can get very sick from rapid growth of neuroblastoma in the liver. A small number of these young babies will need urgent treatment often with chemotherapy.

For most children diagnosed with MS neuroblastoma, your child's doctor may feel confident that your child will get better with no or very little treatment, as the tumours can shrink and disappear naturally without any treatment.

Sometimes, if the tumour is causing clinical problems or if there are certain genetic changes (presence of segmental chromosomal abnormalities) found in the tumour cells, then low doses of chemotherapy may be given to encourage the tumour to start shrinking. In some children with MS neuroblastoma, removing the primary tumour by surgery will be recommended some months later.

Intermediate-risk neuroblastoma

This group includes children with neuroblastoma without MYCN amplification but who have unfavourable histology, and/or diploid tumours and/or segmental chromosome changes:

- **Infants under 18 months with metastatic neuroblastoma (Stage M) and diploid tumours**
- **Infants under 18 months with localised inoperable neuroblastoma or ganglioneuroblastoma nodular (Stage L2) with unfavourable histology**
- **Infants under 18 months with localised inoperable neuroblastoma (Stage L2) with segmental chromosomal changes**

Around 10 children are diagnosed with intermediate-risk neuroblastoma each year in Australia and over 80% are cured of their disease. Children with intermediate-risk neuroblastoma are initially treated with a period of chemotherapy. This is followed by careful reassessment of the position and extent of the tumour, including bone marrow examination, CT or MRI, mIBG, CT-PET or bone scan. In some children, it may then be possible to proceed with surgical removal of the tumour, or some children may require further chemotherapy.

Children over the age of 18 months with localised, unremovable (L2) neuroblastoma, whose tumour biology shows a higher level of cancer activity, will also receive radiotherapy and differentiation therapy with an oral medication called 13-cis-retinoic acid after chemotherapy and surgery.

Very rarely, a child has stage L1, MYCN amplified, intermediate-risk neuroblastoma. In these cases, the child will also receive radiotherapy to the site of the primary tumour after surgery, as well as 13-cis-retinoic acid. This will be fully explained to you by your child's doctor. Very occasionally, some children with intermediate-risk neuroblastoma will be discussed at a monthly meeting with national experts on the national tumor board panel, organised by the Australia and New Zealand Children's Haematology/Oncology Group (ANZCHOG) and high-risk type treatment may be recommended.

High-risk neuroblastoma

This group includes:

- **any child with MYCN amplification (other than stage L1)**
- **any child over 12 months old with neuroblastoma that has spread to other areas of the body (stage M). It should be noted that infants in this group are considered high risk by SIOPEN (clinical trial group) but are not eligible for some of the high risk treatments.**

Around 50% of children with newly-diagnosed neuroblastoma are diagnosed as INRG high-risk category. In Australia, this is around 25 children per year, approximately 50% of whom will be cured of their disease.

Children with high-risk neuroblastoma require intensive treatment of different types from the start because their neuroblastoma cells are highly aggressive. The majority of children will need a combination of different treatments including intensive induction chemotherapy, surgery to remove the primary tumour, radiotherapy, high-dose consolidation therapy (with autologous stem cell transplant), radiation therapy and post-consolidation therapy with differentiation therapy and immunotherapy (an anti-neuroblastoma antibody).

Children with high-risk neuroblastoma are initially treated with a period of induction chemotherapy, followed immediately by careful re-assessment of position and extent of tumour locations. This includes bone marrow examination, CT or MRI scans, urinary catecholamine testing, and mIBG (or FDG-PET CT) scan. The aim of induction therapy is to shrink the primary tumour and clear sites of metatastic disease. A small number of children may need further chemotherapy if the response to induction therapy has been suboptimal. After induction chemotherapy it is possible to remove the tumour by surgery in most children. Children with high-risk neuroblastoma are then considered for consolidation therapy, treatment with very high-dose chemotherapy and autologous stem cells, also referred to as 'myeloablative therapy'. Following consolidation therapy, most children have treatment with local radiotherapy to the area where the tumour was removed at surgery, followed by differentiation therapy with the oral medication called 13-cis-retinoic acid, and immunotherapy.

Starting treatment

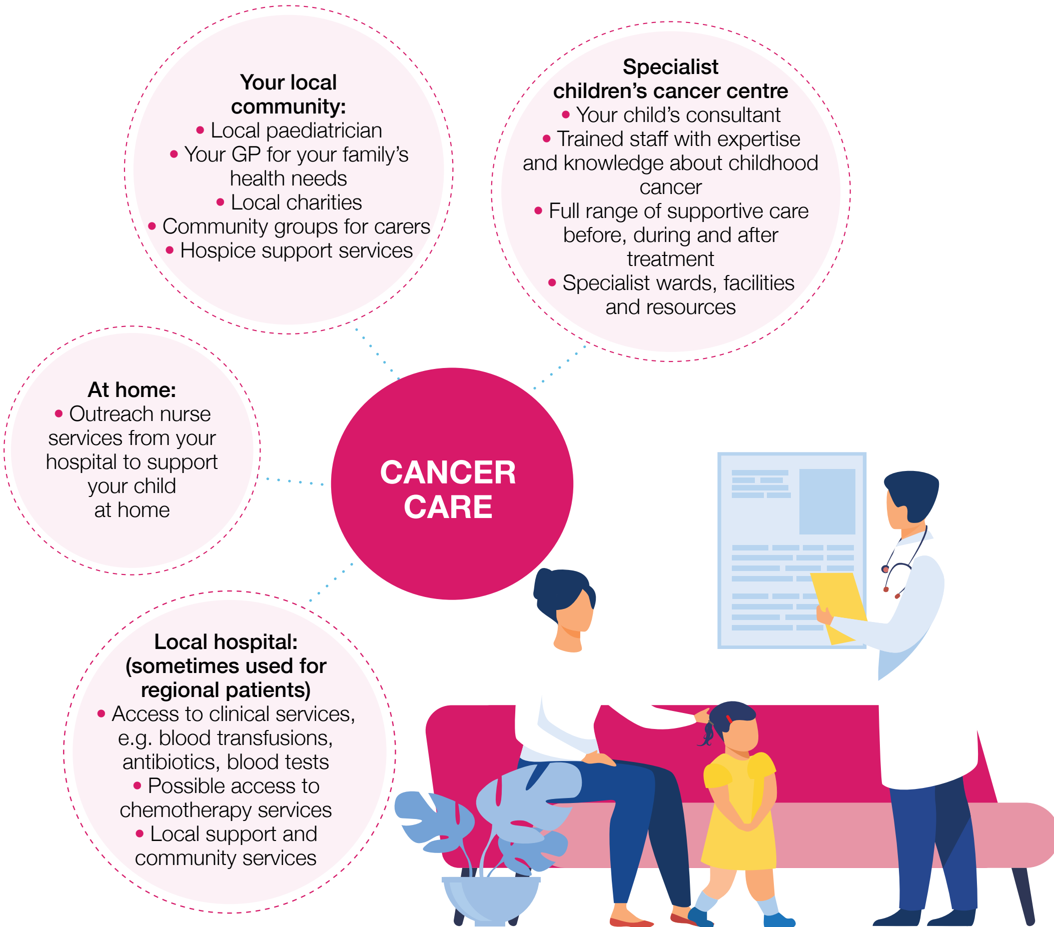
Once your child has a confirmed diagnosis of neuroblastoma, the medical team will be keen to get your child started on treatment as soon as possible at your nearest children's cancer centre. This means that your child will get highly specialised care and may need to stay in hospital for a while so that they can be monitored when starting treatment.

Your child's treatment and care will be managed by:

- Paediatric oncologist – a doctor who specialises in treating all children with cancer
- Paediatric surgeon – a doctor who specialises in the surgical management of cancer in children
- Paediatric radiation oncologist – a doctor who specialises in using radiotherapy to treat children's cancer
- Clinical nurse specialist – a nurse who specialises in caring and supporting children with cancer

A lot of different professional staff will be involved in looking after your child, both at home and while they are in hospital. A team of specialists called the 'multi-disciplinary team' (MDT) will meet regularly to discuss your child's cancer, treatment options, clinical trials, and any areas in which your child may need support.

What your child's cancer care might look like:



Types of treatment

Surgery

Your child's tumour will be removed during an operation if it is possible and safe to do so. In some children, surgery may be involved at a later stage of treatment. This is often after several cycles of chemotherapy have been given to shrink the tumour so that it can be more easily (and therefore more safely) removed by the surgeon. The extent of surgery differs for each child and will depend on factors such as the location of the original tumour and involvement of surrounding organs or structures. If your child has received chemotherapy before surgery, then it is important to make sure that your child has made a full recovery before surgery takes place. This will be discussed with you.

Chemotherapy

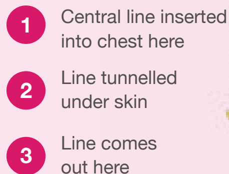
This is the main treatment for intermediate- and high-risk neuroblastoma. Chemotherapy is the use of drugs to destroy cancer cells. They can be given in different ways but the most common way is intravenously – into a vein – whether through injections, cannula, or a line such as a central line, implantable port or peripherally inserted central line (PICC). Chemotherapy is usually given as a combination of multiple different drugs. Once the diagnosis of neuroblastoma is confirmed, the results from the tests will help your child's doctor to decide on the best treatment regimen often called 'protocol'. The suggested treatment will be discussed fully with you.

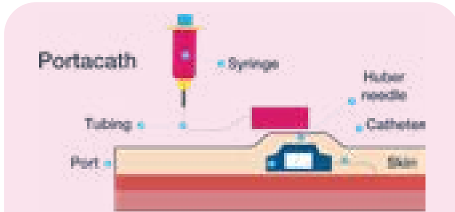
Giving chemotherapy to a child

Central lines and ports are similar devices which make it easier to deliver chemotherapeutic drugs to your child and to take blood samples.

A central line (sometimes referred to as a wiggly) is a thin, flexible plastic tube that is inserted into a large vein near the collarbone under anaesthetic. This allows drugs to be injected and blood samples taken without having to use a needle.

The line can be seen taped to the child's chest and is normally covered by clothing. It is securely attached, should not fall out and should be kept dry.





A portacath or port is an alternative device which does the same thing and is implanted under the skin usually in the chest. A small needle is pushed through the skin into the port to give drugs or take blood and has no part of it exposed outside of the skin. This has the advantage that swimming and sports are less of a problem.

High-dose consolidation chemotherapy

After surgery, children with high-risk neuroblastoma may undergo further treatment with consolidation therapy with autologous stem cells, sometimes call myeloblative therapy. The aim of consolidation therapy is to kill off residual neuroblastoma cells. Rarely, high-dose chemotherapy may be given before surgery. In some high-risk neuroblastoma treatment programs, two courses of consolidation therapy are used, also called 'tandem transplantation'. As consolidation treatment is intensive, your child will be in hospital for a period of about four to six weeks and may be cared for in an isolation cubicle. The treatment will lower your child's count for a prolonged period during which time they will be prone to infections and bleeding. To shorten this period, children receive autologous stem cells (aka peripheral blood stem cells) after consolidation therapy.

Autologous stem cell transplant

Before high-dose chemotherapy is given, stem cells are collected, or 'harvested', from your child and frozen safely until they can be given back to your child after high-dose chemotherapy so that the bone marrow can recover.

Autologous stem cell transplant

Stem cells are found in the bone marrow which makes many different types of blood cells for the body. The bone marrow can become damaged from high-dose chemotherapy so putting healthy stem cells back into the body afterwards can re-populate the child's bone marrow with new blood cells.

What happens?

Stem cells are collected from your child's blood using an apheresis machine which can be connected to the Hickman central line or a special collection catheter called a vascath. In most cases, stem cells can be collected through a Hickman central line. In cases where that doesn't work, a vascath is put into your child's vein under general anaesthetic. This catheter allows blood to be sucked into the machine through the vascath. The machine then collects the stem cells and returns the rest of the blood back to your child through a catheter. The collected stem cells are then frozen, stored and given back to your child after the high-dose chemotherapy.

The harvesting procedure may take most of the day and may need to be repeated on a second day to collect enough stem cells. This procedure is painless and has few side effects. Harvesting is performed 10 to 14 days after a course of chemotherapy. To increase the number of stem cells in the blood before harvesting, a drug called G-CSF is given to your child. This drug helps the stem cells to move out of the bone marrow and into the blood ready for harvesting. The whole of the harvesting procedure will be discussed in detail with you by your child's treatment team.

Very rarely, as an alternative to stem cell harvest, some children may have stem cells collected straight from the bone marrow in a procedure called a bone marrow harvest. This process is performed under general anaesthetic and is very similar to the bone marrow aspirate described previously but takes longer to collect enough bone marrow cells. The bone marrow is then stored like stem cells and given back to your child after high-dose chemotherapy.

Radiotherapy

Radiotherapy treats cancer by using high-energy rays to destroy cancer cells in a specific part of the body. Children with high-risk neuroblastoma will have radiotherapy after their tumour has been removed by surgery and after having high-dose chemotherapy. Some children with intermediate-risk neuroblastoma will also have radiotherapy after surgery but this decision is taken on an individual basis. Radiotherapy is focused on the area where the primary tumour was removed at the time of surgery. It is used to try and kill tumour cells that can sometimes remain after surgery.

Radiotherapy is painless and the machine does not touch your child – it is like having an ordinary x-ray. The total radiation dose is spread out over time and often involves treatment every day for 3-5 weeks, usually excluding weekends. In some situations, the radiotherapy course can be shorter or longer and your child's doctor will discuss this with you.

Radiotherapy requires careful preparation and planning to decide where in your child's body to treat and the best way to give the radiotherapy and can involve several steps. Some young children may struggle to lie still for radiotherapy so sometimes need general anaesthetic each day for radiotherapy planning and treatment. This will be discussed with you by your child's clinical team.

Differentiation therapy/immunotherapy

For children with high-risk neuroblastoma whose tumours have responded to induction chemotherapy, surgery, high-dose chemotherapy and radiotherapy, additional treatment is recommended. This involves a drug called 13-cis-retinoic acid which 'differentiates' (turns cells from being cancerous to non-cancerous) any remaining tumour cells that may be present. Alongside this, children will receive immunotherapy with a monoclonal antibody called anti-GD2 antibody.

Immunotherapy treatment for neuroblastoma relies on your child's own immune cells to kill cancer cells. There are different ways to do this and currently antibody therapy is the most commonly used.

The antibody targets a molecule present on nearly all neuroblastoma cells called GD2 (disialoganglioside). When the antibody binds to GD2 on the neuroblastoma cells, the cells die in a different way than after chemotherapy or radiotherapy.

There is evidence that other cells in the immune system may also promote and contribute to destroying neuroblastoma cells. Drugs can be given to increase the number of these immune system cells during treatment with the anti-GD2 immunotherapy. In North American treatment programs, a cytokine called GM-CSF is most commonly used with the anti-GD2 antibody. In European treatment programs, the anti-GD2 antibody is commonly given alone (ie without GM-CSF). In Australia, both North American and European immunotherapy programs have been used to treat children with high-risk neuroblastoma. The anti-GD2 antibody is used along with 13-cis-retinoic acid for children with high-risk neuroblastoma whose tumours have responded to initial therapy.

If you have any queries about new treatments that you may have heard about in the media or from others, we encourage you to discuss these queries with your child's treatment team. You may naturally feel that you want to explore every avenue for your child and do not want to settle for just one doctor's advice about treatment.

It is reassuring to know that your child's treatment will be discussed during the MDT meeting with other oncologists based at the same hospital. Children's cancer, especially in the field of neuroblastoma, is a small and specialised area of medicine and all doctors will know each other and meet regularly. They will be aware of the latest research in the field through being a member of the solid tumour group of the Australian and New Zealand Children's Haematology & Oncology Group (ANZCHOG), the Children's Oncology Group (COG) and/or SIOPEN, the European association involved in the research and care of children with neuroblastoma. Paediatric oncologists in Australia have a broad network of national and international collaborations. Therefore, if a new treatment has been developed anywhere in the world, it is likely that your child's doctor will know of it and be able to discuss it with you.

In some situations, it may be appropriate (with your consent) that your child's case will be discussed with national experts on the national tumour board panel which is held monthly.

Possible side effects of treatment

Unfortunately, treatment can cause some side effects. Thankfully, these are mostly temporary and there are often ways of controlling or reducing them through supportive care. Your child may be well enough to play, have fun and have a good quality of life during treatment. Your child's cancer team will be able to tell you more about what side effects your child is most likely to have, and how they plan to manage them.

Feeling and being sick

This may happen when chemotherapy drugs are given or after a day or so. The sickness caused by some drugs may last for several days and most children are affected to some extent but anti-sickness drugs have made this common side effect much less severe. Babies and very young children seem to have less sickness than older children.

Losing hair

This is the most visible side effect of chemotherapy and it affects all hair – not only on the head but also eyebrows, eyelashes and other body hair. Losing hair begins after starting treatment, most often within two weeks. Hair usually grows back within a few months of stopping treatment.

The idea of hair loss is usually very upsetting to parents and quite a shock to your child. Most children quickly get used to their appearance and parents have often remarked that once their child's hair is lost it does not seem to bother the child at all. Children might like to wear bandanas, hats, scarves or a wig. Your child's cancer team can give you help and advice with this.

Effects on the blood and bone marrow

Low resistance to infection (called neutropenia)

Many chemotherapy drugs reduce the production of white blood cells which lowers your child's immunity and makes them prone to infection (neutropenic) during treatment. This effect usually begins about seven days after treatment is given. After a few days, the number of blood cells will then increase steadily to return to a safe level before the next chemotherapy drugs are due.

Bruising or bleeding

Other parts of the blood such as platelets and red cells, are also reduced in number by chemotherapy. If the platelets become low, then your child is at risk of bleeding more easily, such as nosebleeds and bleeding gums. For example, if your child has a troublesome and long nosebleed whilst their platelets are low, a platelet transfusion will be given. Your child might also become anaemic because of low numbers of red cells. If this happens, they will receive a blood transfusion.

Before each round of chemotherapy, your child will have a blood test called a 'full blood count' or FBC to make sure that all blood cells such as levels of haemoglobin, number of platelets and the number of neutrophils in the blood have recovered. If levels are still low, then the next course of treatment may be delayed until your child's blood reaches a safe level again.

If your child is receiving an intensive chemotherapy, the treatment will be given according to the protocol. For some treatments like rapid COJEC the chemotherapy may be given regardless of the blood count.

In other chemotherapy programs there may need to be adequate recovery of blood counts before the next cycle is started.

For most children with high-risk neuroblastoma, blood counts are likely to be low for much of the treatment. Consequently, your child will need to stay under close observation at all times for signs of infection so they can be quickly treated with antibiotics if needed.

Losing weight

Weight loss may be caused by both neuroblastoma and chemotherapy, and is very common. Your child's doctor will consider different ways of making sure your child still receives enough nutrition if this happens. This may be through dietary supplements such as high-calorie milkshakes and powders or by feeding through a nasogastric tube passed via the nose into the stomach. Your child's medical team, which includes a fully qualified dietitian, will talk this through with you if required.

Constipation or diarrhoea

Some drugs can change the way the bowel works so your child may have diarrhoea or constipation. We encourage you to let hospital staff know if constipation becomes a problem as laxatives can be given to relieve it. Diarrhoea usually gets better without medication and it's important for your child to drink plenty of fluids. Occasionally, anti-diarrhoea medicines may be needed. If your child has persistent diarrhoea, please let the hospital know as it is possible that they may become dehydrated.

Sore mouth (called mucositis)

Some drugs can cause a sore mouth which may lead to mouth ulcers. Mouth care is very important, and the nurses will show you the best way to care for your child's mouth. The doctor may prescribe mouthwashes or other medicines to help.

Effects on the liver

This is called veno-occlusive disease (VOD) and is one of the less common, but still serious complications which can occur during high-dose chemotherapy and stem cell transplant. VOD is not another illness but a complication that affects the liver. It can range from mild to severe and occurs in approximately 10-30% of children or young people who have received busulfan, or high-dose melphalan as part of their conditioning treatment before a stem cell transplant. Although it is serious, it is usually a temporary problem, but it can be more complicated or even cause long-term problems. Your child's consultant will discuss this in more detail with you.

Effects on hearing and the kidneys

Some drugs, such as cisplatin, can affect your child's hearing and potentially damage their kidneys. Therefore, your child will undergo regular hearing tests (audiograms) as well as tests to measure how well the kidneys are working (the 'glomerular filtration rate' (GFR) test). In this test your child receives an injection of radioactive substance (not harmful to your child) into a vein in the hand or arm. Blood samples are then taken.

Effects on the heart

Some drugs such as doxorubicin can potentially affect the way your child's heart works by causing damage to the heart muscle. Your child will undergo regular tests, such as a heart ultrasound scan (called an echocardiogram) to assess how well their heart is working both during and after completion of their treatment.

Effects on the nerves

Some chemotherapy treatments such as vincristine and cisplatin can potentially cause damage to your child's nerves. This can cause your child to have numbness, tingling or painful hands and feet. Some children find it can affect their walking or fine hand movements like picking up small objects.

Future fertility

Parents can also be concerned about the effects of chemotherapy on their child's ability to have children in the future. A long-term outcome of present treatments for neuroblastoma is difficult to predict but it is known that some of the chemotherapies used to treat high-risk neuroblastoma can be harmful to fertility. There are some treatments aiming to maintain fertility for children undergoing treatment for neuroblastoma and your child's medical team may discuss this with you. For example, it is known that most drugs used in high-dose consolidation therapy such as cyclophosphamide, busulfan and melphalan can be damaging to fertility when given in high doses. However, this side effect has to be weighed against the potential benefit of using these drugs.

Reaction to immunology/antibody treatment

Many children with high-risk neuroblastoma will receive treatment with immunotherapy, using an anti-neuroblastoma antibody which targets GD2. The most common anti-GD2 drugs used in Australia and New Zealand are unituxin (dinutuximab) and qarziba (dinutuximab beta). This treatment can commonly cause a reaction whilst your child is receiving the infusion. Some children can notice abdominal (tummy) pain, numbness or tingling, a rash, generalised swelling, dizziness or a sensation of the heart racing. Your child will be very closely monitored throughout antibody treatment and you should alert a member of staff if you have any concerns.

Taking part in clinical trials

Sometimes in Australia your child will be offered cancer treatment within a clinical trial. These are research studies carried out to try and find new and better treatments for cancer such as a new drug or combination of treatments. By doing this through clinical trials, we can make sure that comparisons are measured consistently and reliably over time to see if one treatment is better than another. This is why improvements are made every year in treating and curing cancer in children.

Your child will receive the best possible treatment regardless of whether they are on a trial or not. If a trial is suggested, you will be given information about it and what is involved. You will need to consent to the trial as well as to the treatment itself. Your child's consultant will be able to discuss potential clinical trials with you.

Giving your consent

Before your child is asked to take part in a clinical trial, your child's medical team will explain what the trial is aiming to achieve and the risks and benefits of taking part. Once you have considered everything you need to know, you will be asked to give your consent for your child to take part and sign a form. There is no pressure to take part and your child will receive the best treatment available whatever you decide. When deciding whether to take part in a trial, it can add to the stress of coping with your sick child and the bewildering range of processes associated with treatment. It may seem as though the treatment team is passing over responsibility for deciding what treatment your child should be given. However, clinical trials are designed to see if it is possible to continue to improve on currently accepted treatments. Most clinical trials use what is regarded as the current best available therapy and are asking questions on how to make treatments more effective.

Randomisation

For some trials, the researchers carry out a process called randomisation, particularly when the trial is comparing two treatments to find out if one treatment is better than another. In this situation the researchers don't know if one treatment is clearly better than the other. This means a computer will randomly allocate your child to have a particular treatment in the trial. This is done so that each treatment group has a similar mix of children of different ages, sex and general health. It makes sure that researchers and doctors can't decide who should get which treatment to avoid bias that could skew the findings.

Safety

Ensuring safety of children in clinical trials is the top priority. All trials are approved by an ethics committee (Human Research Ethics Committees in Australia) and regulatory committees, and they are reviewed on an ongoing basis. Most trials have independent monitoring committees to make sure that the trial is progressing well and that there is no undue concerns about toxicities and side effects for participants.

After treatment

When treatment finishes, your child will be given a summary of the treatment they have received and an aftercare plan involving regular follow-up checks in hospital over the next few years. This is to make sure there are no signs of the cancer coming back (called 'relapse') and also to look for and treat any long-term effects that might happen as a result of the cancer and its treatment (called 'late effects'). These possible late effects can affect their heart and lungs, growth hormones, and they may be at a higher risk of developing cancer again in the future, so it is important that your child attends their follow-up clinic appointments.

If neuroblastoma comes back during or after treatment

Occasionally children with low-risk or intermediate risk neuroblastoma will experience a relapse or recurrence of their neuroblastoma. In most cases these children can be effectively treated and cured with appropriate therapy.

When high-risk neuroblastoma comes back during or after initial treatment, it is called **relapsed disease** and it may be possible to control the disease for years. However, it is usually very difficult to achieve a complete cure. Sometimes high-risk neuroblastoma does not respond well to the initial treatment – this is called **refractory disease**.

If your child has relapsed or refractory high-risk neuroblastoma, choosing which treatment is right for your child will depend on many things. This includes where their disease is and what treatment they have previously received. Your child's consultant will be able to help you decide which is the most appropriate treatment for your child at this time.

Going for treatment overseas

There has been some media coverage of families who have decided to take their child with neuroblastoma for treatment abroad. The decision to take a child abroad for treatment is always best taken by you and your family and the consultant paediatric oncologist caring for your child. A lot of factors will need to be considered to reach that decision. Most internationally approved therapies are either approved in Australia or can be accessed. There are an increasing number of early phase trials for promising new cancer treatment for children in Australia. In some cases going abroad may provide the opportunity to receive unique experimental therapies that are not available in Australia but are available in single centres in other countries. For patients in Australia who have relapsed neuroblastoma, there are a number of treatment options available. These include further second and third line chemotherapy, early phase clinical trials, and targeted radiotherapy.

If treatment doesn't work

For some children, parents are told the devastating news that their child's cancer is not curable. Your child's care will change to focus on giving the best quality of life for your child and managing their symptoms. There is a huge amount of practical and emotional support for families at this time, such as from your child's hospital, charities, and organisations.

Your treating hospital has a range of resources to help parents to prepare and plan for the next steps if treatment doesn't work.

Caring for yourself and other family members

Your child's diagnosis will have an emotional effect on everyone in your family, as well as many friends and even acquaintances in the wider community. This section addresses your needs because, as a parent, your wellbeing is crucial to your child and the rest of your family.

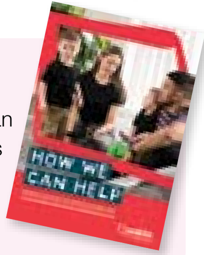
Looking after yourself

Having a child diagnosed with neuroblastoma has a huge impact on a parent or carer. You will likely have different feelings at different times, with periods of frustration, anger, fear, anguish, panic and grief. At some other times, you may feel quite calm, as you and your child settle into the routine of treatment.

You will probably find that your emotions go up and down a lot during the days and weeks following diagnosis, and that your feelings change over time.

When you notice a difficult or uncomfortable emotion, try to calm yourself and notice what you are feeling, rather than pushing it away. Sometimes, your feelings might spur you to take action or make a change. Other times, talking things through with someone you can trust is all that is needed.

One of the best things you can do for your family is to take care of your own needs. Eating and sleeping well, getting fresh air outside, addressing any health problems and taking regular breaks. By meeting your own needs, you can be there to help everyone else.



Looking after yourself

Useful tips on self care can be found under resources on these websites: **pics.org.au** and **redkite.org.au**

Supporting your child

The effect of a cancer diagnosis and treatment will depend on the age of your child. As well as feeling unwell and coping with side effects, they may be missing home, family and friends, and may struggle with the change in routine. If they are not able to say how they are feeling, they may express this through behaviour such as clingi-ness, tantrums or tearfulness.

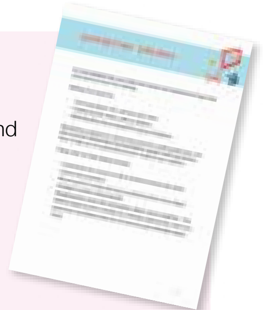
One of the hardest parts of caring for a child with cancer is knowing what to say and how much information to give them. Many families feel that answering questions honestly is best, giving a little information at a time. Some children may not ask questions, but this doesn't mean they don't want to know what's happening. They may be frightened and uncertain of many things. Some children may even wonder if they have done something wrong and that's why they have cancer.

You can ask your child's medical team for guidance on how to talk to your child. There are also booklets for young children available from Redkite that can help you explain the illness and treatment.

Younger children may be frightened about being separated from their parents. It's important to reassure them that any separation is only temporary. Doctors and nurses will be happy to explain more about this and can help you reassure your child.

Supporting siblings

Brothers and sisters of a child with cancer may have many or all the same feelings and emotions that you have. If you need to spend a lot of time in the hospital with your child, your other children may have to be cared for by family members or friends. They may have to spend a lot of time away from you and find their daily routine keeps changing. As well as worrying about their sibling's health, they may also feel resentful of all the attention they're getting. This can make them feel left out and angry. Being there for them or showing that you still love them can help to reassure and comfort them.



Supporting Siblings

Useful tips can be found on websites including Cancer Council and Redkite.

For a full list visit the **resources section** on Neuroblastoma Australia.

Supporting your child at school

As your child's health improves, they may be able to go back to school. This is important for their educational, psychological, and social development, but it is also important for the whole family as school routines can help everyone return to a sense of 'normal life'. As soon as your child is diagnosed, contact your childcare or school's principal to tell them what's happening and keep open the lines of communication. It can help to let them know about the plans for treatment.

Many children diagnosed with neuroblastoma are very young and have not yet started school. As a parent, you may have to choose between having your child in childcare throughout treatment or keeping your child at home. Keeping your child at home may mean they have less chance for social growth and development, but if they stay at childcare you may feel there is a risk of infection.

There is no right or wrong decision – it's a personal choice for you to make. You may want to think about whether your child:

- is already settled at daycare or pre-school
- has their social needs met by siblings and/or other children outside of childcare
- is well enough to attend daycare or pre-school
- has already had chickenpox which can be harmful for children on treatment

It may be useful for you to talk to your specialist nurse or social worker about childcare or school attendance and the support they can offer to help with this.

Supporting your child at school

Find out more information at the Ronald McDonald House Charities website under Learning Program.

The Australian Government Cancer Website (**childrenscancer.canceraustralia.gov.au**) also has information.

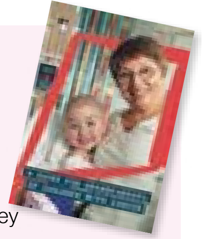
Supporting grandparents

Being told that their grandchild has cancer will be a huge shock for your parents. They will worry not only about their grandchild but also how you will deal and cope with this news. Most are also concerned about the effects it will have on any other children within your family and, of course, how they will cope themselves.

As parents, you will usually have access to doctors and others who can answer your questions. It is not so easy for your parents to get information first hand and this can lead to feelings of stress and isolation. Keeping them involved and allowing them to help you and your family if they are able to can help them play a valuable role in supporting their family.

Seeking information

You may want to find out as much as possible about the cancer and its treatment. There is a lot of online information but not all of it is reliable, so talk to the doctors about where to look.



Supporting grandparents

This booklet from Redkite answers some of the many questions which grandparents of children with cancer told Redkite they would have liked answered when their grandchild was diagnosed and during their grandchild's treatment phase.

Resources

Resources

The following is a list of Australian consumer organisations and community-funded organisations providing support and high-quality information for families affected by neuroblastoma.

- **Camp Quality** delivers family support, hospital, education and recreation programs.
- **Cancer Advisor** is a one-stop website for the families of children, teenagers and young adults with cancer, and young people with a diagnosis.
- **Cancer Australia** is a website with information on children's cancers as well as guidance and advice for children, parents, grandparents, relatives, friends and the community on living with children's cancer.
- The **Cancer Council** in your state or territory provides general information about cancer information on resources and support groups in your local area. The **Cancer Connect** service provides online peer-based support and information for people living with, and affected by, cancer. You can call the Cancer Council Helpline from anywhere in Australia for the cost of a local call on 13 11 20.
- **Cancer Hub** is a website where charities (CanTeen, Camp Quality and Redkite) have come together to offer a wide range of practical and emotional cancer support services for families facing cancer (with children aged 0-25), providing a one-stop shop.
- **CanTeen** helps young people (aged 12-24) cope with their own cancer, or cancer in their family. Also see Canteen's freely available online books and resources for young people who have cancer (suitable for ages 12-15 years). For young people with cancer, the Youth Cancer Service provides specialist, age-specific treatment and support.
- **CanTeen Connect for Parents** is a community designed to connect parents affected by cancer and provide support and resources to help with your family's cancer journey.
- **Cancer Voices Australia** supports and advocates for Australians affected by cancer.
- **Children's Cancer Foundation** aims to help children with cancer to access world-leading treatment and support. It also supports families throughout the treatment process.
- **Kids Helpline** is a free, private and confidential 24/7 phone and online counselling service for young people aged 5 to 25 in Australia. Visit Kids Helpline or call 1800 551 800.
- **Kids with Cancer Foundation Australia** helps children with cancer and their families with finances.
- **Make-A-Wish Foundation** grants wishes to children in Australia with life-threatening medical conditions.
- **MissingSchool.org.au** is an online parent-run organisation that provides advice and parent support on education issues for your child.
- Neuroblastoma Australia supports and helps families of children with neuroblastoma and organises fundraising activities for neuroblastoma research. It provides an online support group for carers.

- **Redkite** provides financial assistance, emotional support and educational assistance to children with cancer and their families. It also has scholarships available for specific educational goals.
- **Ronald McDonald House Charities** provides programs including educational support and accommodation near treatment centres.
- **Rare Cancers Australia** supports patients with rare and less common cancers, including several children's cancers.
- **Starlight Children's Foundation** provides a range of programs to ensure the wellbeing of seriously ill children. Livewire is a free online community powered by Starlight, which connects teens living with illness or disability & their siblings, in Australia or New Zealand.
- **The Bereavement Care Centre** has counselling and support services for terminally ill children and their families and recently bereaved people.

Your treating hospital may offer different supports to help you and your family through the cancer journey. The treatment and support team at the hospital is there to help you, your child and your family. Ask your child's treatment team about the hospital services available for your family, and how to access them, such as:

- your child's educational needs,
- accommodation if you travel a long way for appointments,
- psychosocial support including access to a psychologist or social worker,
- culturally or language specific information, or
- other supportive care including palliative care.

At the end of treatment, there are services available in the hospital including outreach clinics and cancer survivorship clinics to help you manage the long-term effects of treatment for childhood cancer.

Appendix

Below is a table which shows how a risk group is arrived at by your child's oncologist.

INRG Risk Stratification Table

INRG Stage	Age (months)	Histologic Category	Grade of Tumour Differentiation	MYCN	11q Aberration	Ploidy	Pretreatment Risk Group
L1/L2		GN maturing; GNB intermixed					A Very low
L1		Any, except GN maturing or GNB intermixed		NA			B Very low
				Amp			K High
L2	< 18	Any, except GN maturing or GNB intermixed		NA	No		D Low
					Yes		G Intermediate
	≥ 18	GNB nodular; neuroblastoma	Differentiating	NA	No		E Low
					Yes		
			Poorly differentiated or undifferentiated	NA			H Intermediate
				Amp			N High
M	< 18			NA		Hyperdiploid	F Low
	< 12			NA		Diploid	I Intermediate
	12 to < 18			NA		Diploid	J Intermediate
	< 18			Amp			O High
	≥ 18						P High
MS	< 18			NA	No		C Very low
					Yes		Q High
				Amp			R High

Cohen et al. The International Neuroblastoma Risk Group (INRG) Classification System. An INRG task force report. J Clin Oncol. 2009: 27 (2): 289-297.

Glossary

Adrenal glands
Specialised glands above the kidneys that release hormones to maintain blood pressure and enable us to respond to stress. About 50% of neuroblastomas start in the adrenal glands.

Anaplastic lymphoma kinase (ALK)
This is a protein involved in the normal development of the nervous system. The gene that controls the production of this protein is mutated in around 8-10% of cases of neuroblastoma of all risk groups.

Anaesthetic
Drug which stops feeling, especially pain. A general anaesthetic makes you unconscious. A local anaesthetic stops feeling in one part of the body but you are still awake.

Biopsy
Removal of a small piece of tumour for testing to establish a diagnosis.

Bone marrow
The substance at the centre of long bones that makes blood cells.

Catheter
Tube that is passed into the body to drain fluid.

Central line (Hickman line, Port-a-cath)
Long plastic tube that is inserted into a large vein near the heart under anaesthetic. Central lines are used to take blood samples and give drugs.

Chemotherapy
Treatment using one or more anti-cancer drugs.

CT scans
Multiple x-rays are taken by a CT scanner and converted by a computer to form a 3D view of the part of the body under examination.

Ganglioneuroblastoma
A type of tumour that is a 'close relative' of neuroblastoma.

Genetic
A condition caused by abnormal genes (may be inherited).

Immune system
The body's defence against infection, disease and foreign substances.

Immunology
The study of the body's immune system, which fights infection.

Immunotherapy
A form of treatment which relies on cells of the body's own immune system to kill cancer cells.

Immunosuppressive
Lowering the body's ability to fight infection.

Intravenous (IV)
Into a vein, for example, when drugs are given directly through a drip.

Malignant
Cancerous.

Metastases
Cancer that has spread from the place where it started (also known as secondary cancer).

mIBG (meta-iodobenzylguanidine)
A radioactive substance taken up by neuroblastoma cells used in a type of scan that helps to locate neuroblastoma cells in the body.

MYCN
A gene which is amplified (present in many more copies than the normal number of two in a cell) in around 25% of children with neuroblastoma. A test for MYCN amplification may be used to determine how aggressive a particular neuroblastoma may be.

Nausea
Feeling sick.

Neutrophils
A type of white blood cell which fights infection.

Oncologist
A doctor who specialises in the treatment of cancer.

Oncology
The study and treatment of cancer.

Paediatric
To do with children.

Palliative
Relief of a symptom (for example, pain) rather than cure of the disease.

Prognosis
The outlook or expected outcome of a disease and its treatment.

Radiotherapy
The use of radiation to treat cancer.

Refractory
Resistant to treatment.

Relapse
The return of symptoms of a disease after a period of good health; re-occurrence of a tumour after treatment.

Remission
A period of good health where there is no longer any visible cancer.

Surgery
An operation.

Stem cell
Early (immature) blood cell from which other blood cells are made.

Therapy
Treatment.

Tumour
An abnormal lump of tissue formed by a collection of cells. It may be benign (non-cancerous) or malignant (cancerous).

Ultrasound scan
The sound waves produced by a scanner bounce from solid organs inside the body and are recorded on screen. Allows doctors to see the outlines or shadows of normal organs and tumours.

Vanillylmandelic acid (VMA)
A substance found in the urine in raised amounts when a child has neuroblastoma.

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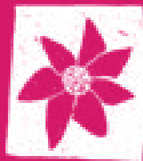

Neuroblastoma
AUSTRALIA

Neuroblastoma Australia is a registered charity supporting families affected by neuroblastoma.

For more information visit www.neuroblastoma.org.au