Neuroblastoma

Information and support for parents

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About this booklet

This booklet is for parents and carers of a child who has been diagnosed with neuroblastoma. We hope it answers your questions and helps you to cope with some of the feelings you may have.

There is information about neuroblastoma, the treatments that are used and their possible side effects. It also discusses how a cancer diagnosis can affect you, your child and the rest of the family. You may also find the CCLG booklet ‘Children and Young People with Cancer: A Parent’s Guide’ helpful.

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 professionals that are looking after your child. Alternatively, if your child’s treatment has been completed, some of what is covered in this booklet may be different from your own experience.

This booklet is not intended to provide an alternative to discussion with members of the multidisciplinary team caring for your child. It is designed to provide a reference to the information you have been given already, or will be given in the future, and will act as a reminder of discussions you have had with your child’s treatment team.

This booklet provides general information about neuroblastoma. Every child is an individual and your child’s case must always be discussed with the treatment team caring for your child.

Throughout this booklet, we refer to children by which we also mean teenagers and young adults.
Neuroblastoma and its treatment

Neuroblastoma is a cancer almost exclusively of childhood. Childhood cancers are usually different from cancers affecting adults. They tend to occur in different parts of the body to adult cancers and look different under the microscope. They also respond differently to treatment.

Neuroblastoma is the most common solid tumour in children other than brain tumours. Fewer than 100 children in the UK are diagnosed each year with neuroblastoma, making up about 6% of the total number of childhood cancer diagnoses.
Neuroblastoma primarily affects younger children and is the most frequently occurring solid tumour in infants under the age of one year accounting for around a fifth (22%) of all cancers diagnosed at this age. The incidence of neuroblastoma is rare after the age of five. Only 2% of neuroblastoma is diagnosed in children over the age of 10 years and 0.5% in those over the age of 15 years.[1]

Neuroblastoma is very rare in teenagers, young adults and adults and current treatment is broadly the same as in younger children, but may change in the future. In these patient groups the disease is usually more slow growing. In general, the outlook is not as good as in younger patients, but the disease can often be controlled for many years due to its slow growth.

Neuroblastoma is an ‘embryonal tumour’, a type of cancer that develops from the cells left behind from a baby’s development in the womb. The cells that it develops from are called neuroblasts, giving rise to the name neuroblastoma:
• ‘neuro’ means nerve
• ‘blast’ means cells in an early stage of development
• ‘oma’ means a group of cells, or a tumour.

Neuroblastoma can occur anywhere in the body. The site of origin is either in one of the two adrenal glands situated in the abdomen (tummy) or in nerve tissue that runs alongside the spinal cord in the neck, chest, abdomen or pelvis.

The most common site for the tumour to grow is in the abdomen (tummy). 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. The adrenal glands are specialised glands found above the kidneys. They normally release hormones to maintain blood pressure, and enable us to respond to stress. In some cases, neuroblastoma can spread to tissues beyond the original site, such as the bone marrow, bone, lymph nodes, liver and skin.

When told that your child has been diagnosed with neuroblastoma, you may feel numb or as if you have been physically hit. Some of the feelings you have may include fear, denial, sadness, guilt and anger. These are normal reactions. It’s important to remember that it’s not your fault your child has cancer and it’s not because of anything you’ve done or anything you’ve not done.

Since your child’s diagnosis, you may have met many new faces, heard a lot of unfamiliar medical terms and your child may have undergone a series of investigations. The hospital staff will understand that you may need some time to absorb what has happened, and what you are being told about your child’s treatment. They will know that you may need to ask some questions several times.

Many parents cannot think of any questions to ask when a doctor is there, but think of all sorts of things as soon as they have gone. It is a good idea to write down things that you particularly want to ask, so that you can discuss your questions at the next opportunity.

If a particular moment is inconvenient, it may be suggested that the doctor and other members of the team could sit down with you later on and go over your concerns. This is not to ignore an urgent worry, but to provide a setting and a time when unhurried discussion can take place. The doctor may have to wait for results of certain tests before any realistic discussion can take place, but it is understood that such a time of waiting is a very difficult and anxious one for you and your family.

When a child is diagnosed with neuroblastoma, it has a big impact on the whole family. Your child’s routine is likely to change, they may have to stay in hospital and they’re likely to have regular hospital appointments. This may feel overwhelming for you, your child, and the rest of your family. There are many healthcare professionals and support organisations who can help you through this time.

Causes

As with most childhood cancers, the cause of neuroblastoma is unknown. It is not infectious and cannot be passed on to other people.

Hereditary disease

Around 1% of neuroblastoma is hereditary, most commonly due to genetic mutations present in a gene called anaplastic lymphoma kinase (ALK) or a gene called Phox2B.

These mutations occur in all cells as well as cancer cells and can then be passed on through families. Hereditary neuroblastoma often presents in patients who are younger than 18 months, 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.
Signs and symptoms

The symptoms of neuroblastoma may vary depending on where your child’s tumour is.

- If the tumour is in the abdomen, your child’s tummy may be swollen and they may complain of constipation or have difficulty passing urine.
- If the tumour affects the chest area, your child may be breathless and have difficulty swallowing.
- If the tumour occurs in the neck, it is often visible as a lump and occasionally affects breathing and swallowing. A tumour in the neck may cause different pupil size, reduced sweating, redness or a ‘droopy’ eyelid on one side of the face (Horner’s syndrome).
- Occasionally, there are deposits of neuroblastoma in the skin that appear as small, blue-coloured lumps.
- If the tumour is pressing on the spinal cord, children may have weakness in the legs and walk unsteadily. If your child is not yet walking, you may notice reduced leg movements. They may also have constipation or difficulty passing urine. There may also be associated back pain.
- Your child may be found to have high blood pressure.
- Very rarely, children may have jerky eye and muscle movements (opsoclonus-myoclonus-ataxia syndrome, or ‘dancing eyes syndrome’), and general unsteadiness associated with the neuroblastoma.

There are often vague and non-specific associated symptoms of tiredness, pale complexion, loss of appetite, weight loss, bone pain and generalised discomfort which can make a child rather irritable and unhappy.

Many children with neuroblastoma have little in the way of symptoms. Perhaps they have seemed ‘off colour’ for a little while or have had a loss of appetite, vague aches and pains or 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 presented with 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 a rare disease, the diagnosis is rarely suspected if only fairly vague symptoms are present. Neuroblastoma, particularly ‘high risk’ neuroblastoma, often presents at a late stage.

The NICE Referral Guidelines for Suspected Cancer recommend very urgent referral (for an appointment within 48 hours) for specialist assessment for neuroblastoma in children with a palpable abdominal mass (a lump in their tummy) or an unexplained enlarged abdominal organ[1].

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Diagnosis

In the UK and Ireland, there is a network of specialist hospitals with expertise in diagnosing and treating childhood cancer, known as Principal Treatment Centres. Once neuroblastoma is suspected, your child will be referred to a Principal Treatment Centre for diagnostic tests and a treatment plan. Although neuroblastoma is rare, these centres will see the majority of children who are diagnosed with neuroblastoma, and therefore have built up expertise in the disease and its management.

Your child will undergo a clinical examination (the doctor’s physical examination), blood and urine tests, special scans, x-rays and bone marrow tests, as well as a ‘biopsy’ of the main tumour mass. All of these may seem frightening to both you and your child, but the reason for having each test will be discussed in detail.

Tests and scans

Tumour biopsy
A small piece of tumour is often taken for examination. This test is known as a biopsy. It involves an operation where your child has a general anaesthetic and a piece of the tumour is taken out through a small cut (incision) in the skin. Sometimes a piece of tumour may be drawn up through a needle; this procedure is known as a ‘needle biopsy’.

A series of tests may be carried out on the cells in this biopsy to find out more about the biology of the tumour. Knowing about tumour biology provides information that is used in deciding the best treatment for your child. These tests include looking at the chromosomes and other biological markers. One important biological marker is called MYCN, as when increased amounts of this marker are present, more intensive treatment may be needed.

Blood tests
Blood for testing may be taken from a vein in your child’s arm or by a finger prick. You are probably familiar with both of these procedures.

Urine tests
A simple special test in the diagnosis of neuroblastoma measures vanillyl mandelic acid (VMA) in the 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 the diagnosis to the paediatric oncologist. Sometimes a similar marker called homovanillic acid (HVA) is also measured. It is raised in 95% of neuroblastoma cases.

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

X-rays – x-rays may be used to see if the neuroblastoma has spread to certain bones.

CT scans – The CT scanner takes multiple x-ray images and these are converted by a computer to form a 3D view of either the whole body or of the part of the body under examination.

Ultrasound scans – The ultrasound scan will be a familiar procedure to all mothers who had this performed during their pregnancy. The sound waves produced by the scanner bounce from solid organs inside the body and are recorded on a screen. The doctors can see the outlines or shadows of normal organs and of any tumour inside the body.

mIBG scans – mIBG stands for ‘meta-iodobenzylguanidine’. This substance is naturally taken up by
neuroblastoma cells. mIBG contains radioactive material (at a level which is not harmful) and is given by injection into the blood stream. When mIBG accumulates in the neuroblastoma cells, the radioactive material can be detected by a machine called a gamma camera. This type of scan is a useful diagnostic tool as it gives a complete picture of the location of any tumour cells in the body.

**Bone scans** - A bone scan involves injecting a small amount of radioactive liquid (at a level which is not harmful) into a vein, usually in the hand or forearm. The radioactive material gathers in bones where the tumour has spread and these can be seen when pictures are taken on a gamma camera. Some centres use a mIBG scan as a way of obtaining the same information as that obtained by a bone scan.

**MRI scans** – an MRI (magnetic resonance imaging) scan relies on magnetism and is a very safe procedure as no radiation is used. It takes longer than a CT scan and is quite noisy. There are no known side effects to this type of scan.

Apart from the need for an injection of ‘contrast’ during some CT scans and the injection of radioactive liquid for a bone scan or mIBG scan, none of these investigations are painful to your child, but 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 sedation may be given. An anaesthetic may be needed for some children to have some of the tests.

**FDG-PET scans** - FDG stands for fluorodeoxyglucose and PET stands for Positron Emission Tomography. This is another type of radionuclide scan like a mIBG scan which can be useful to detect metastatic sites of neuroblastoma, particularly in cases where the mIBG scan is negative (around 10% of cases). In mIBG negative neuroblastoma the FDG-PET scan can be used to determine response to treatment.

**Bone and bone marrow**
The most common sites to which neuroblastoma cells spread are the bones and the bone marrow. To detect tumour cells in the bone, your child will probably undergo either a bone scan or a mIBG scan (see above), or both.

To examine the bone marrow for tumour cells, a needle is inserted into one of the larger bones like the hip bone and a small quantity of bone marrow, found at the centre of the bone, 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 site 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 site; 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.

It may take several days for any tests to be completed and the results analysed. Undergoing these tests and waiting for results can be a stressful time and lead to anxiety. However, exact assessment of the extent of your child’s disease before beginning treatment is very important. The results obtained will inform the type and length of treatment that will be given to your child.

**Genetic tests**
Doctors and scientists are continuing to learn more about neuroblastoma. Recently they have discovered that special biological tests can indicate how aggressive a particular neuroblastoma is likely to be. These include a genetic test called “MYCN amplification”.

MYCN is a gene which is found more often (amplified) in around 25% of neuroblastoma cases. MYCN amplification is more common in younger children and is very rare in older children or teenagers. The results of this test will help to determine the type of treatment your child has.

The MYCN test can be done very rapidly but some of the other genetic tests take longer to do and the results may take a few weeks to come back to your doctor. Most of these genetic tests are carried out in a very experienced special laboratory in Newcastle called the Newcastle National Reference Centre for Neuroblastoma Genetics.
Tumour ‘staging’

Doctors recognise several special categories of neuroblastoma that are grouped into different ‘stages’. Staging gives an indication of how far the tumour has spread in the body. The different stages of neuroblastoma have different treatments, as they carry with them different risks. Other factors that may affect your child’s prognosis are their age and the results of several laboratory tests that will be carried out during the initial investigation period.

For many years the International Neuroblastoma Staging System (INSS) has been used to assess individual children and these descriptions are likely to continue for some time alongside a more recently introduced International Neuroblastoma Risk Group Staging System (INRSS). The new system is aimed at getting a better match between the risks and side-effects of therapy (treatment) with the risk of disease. As these new staging definitions are adopted worldwide it will be possible to get a more accurate description of the effectiveness of different treatment strategies. The approximate INRSS stage is shown in brackets against the INSS stage below.

**Stage 1 neuroblastoma (INRSS stage L1)**
This means that the tumour is fairly small and confined to one site. It has not spread anywhere else in the body and is able to be removed completely by an operation. This type of tumour is usually curable by surgery alone.

**Stage 2 neuroblastoma (INRSS stage L1)**
As with stage 1, the tumour is confined to one site and has not spread to distant parts of the body. However it may be larger than a stage 1 neuroblastoma and, at operation, it may be more difficult to remove completely. Sometimes a lymph node or some glands near to the stage 2 neuroblastoma may also have been affected by the tumour. Stage 2 is usually treated by surgery alone but, depending on the site and results of certain tests, additional treatment such as chemotherapy (see page 13) may be needed.

**Stage 3 neuroblastoma (INRSS stage L2)**
This stage of tumour is also confined to the primary site in that it has not spread to other parts of the body. The tumour may be very large and said to have crossed the ‘mid-line’ of the body. This means that the tumour has grown right across the child’s abdomen or chest from the original side where it began. This type of tumour would usually be difficult to safely remove surgically. Chemotherapy will be initially recommended to try to shrink the tumour for subsequent safe surgical removal. After the operation, radiotherapy may be given.

**Stage 4 neuroblastoma (INRSS Stage M)**
This means that the primary tumour may be of any size, but some neuroblastoma cells have broken away and spread to other organs of the body, most commonly bones, bone marrow or liver. Chemotherapy will be recommended for a stage 4 neuroblastoma to kill the tumour cells that have spread to different body parts and to shrink the primary tumour for later removal by an operation.

After surgery, further chemotherapy is given, using very large doses (see page 14). After this ‘high dose chemotherapy’, the next part of the treatment may involve radiotherapy (see page 13). If your child is enrolled on a clinical trial, this may be followed by immunotherapy (see page 15) which aims to boost the child’s own immune system to help kill any remaining tumour cells.
Ganglioneuroblastoma

This is a special type of neuroblastoma found in very young babies, under one year old. The tumour cells may have spread beyond the ‘primary’ site to other parts of the body, but the cells usually behave in a less aggressive fashion than in an older child. The sites of the body affected by stage 4s neuroblastoma in addition to a small primary tumour which is often in one of the adrenal glands are most typically the liver, skin and sometimes the bone marrow. When this pattern of disease is noted the oncologist may feel fairly confident that the child will get better with no, or very little, treatment as the the tumours can shrink and disappear spontaneously without any treatment. Sometimes if the tumour is causing clinical problems or if there are certain genetic changes in the tumour cells low doses of chemotherapy will be given to encourage the tumour to start shrinking.

In some of these 4s cases, months after the diagnosis, surgical removal of the primary tumour will be recommended.

Very occasionally, a baby who originally presented with a stage 4s neuroblastoma may develop signs and symptoms of the more ‘aggressive’ stage 4 neuroblastoma later on. The neuroblastoma cells typically reappear in the lymph glands or bones. When this happens the doctors are likely to decide to give chemotherapy.

Risk groups

Nowadays doctors categorise neuroblastoma according to the International Neuroblastoma Risk Group (INRG) Classification as low, intermediate (medium) or high risk.

Low and intermediate risk neuroblastoma account for around 50% of all neuroblastomas and include localised L1 and L2 neuroblastoma, and MS and M neuroblastoma in infants who are less than 12 months of age who do not have a genetic abnormality called ‘MYCN amplification’.

High risk neuroblastoma accounts for over 50% of neuroblastoma cases and includes patients with metastatic neuroblastoma (M) over the age of 12 months with or without MYCN amplification and infants less than 12 months with stage M neuroblastoma with MYCN amplification.

Ganglioneuroblastoma

Ganglioneuroblastoma is another type of tumour which is a ‘close relative’ of neuroblastoma. It can present in any age group, but the cells of this tumour are more ‘mature’ (i.e. less likely to spread) than those of the ordinary neuroblastoma. Like neuroblastoma, ganglioneuroblastoma stages 1, 2 and 4s, may be cured by surgery alone. Other stages will need chemotherapy in addition to surgery.

Immunotherapy is given alongside a treatment which makes neuroblastoma cells mature into normal nerve cells called 13 cis retinoic acid.

Most infants who present with stage 4 neuroblastoma who are diagnosed when they are under one year of age usually have less ‘aggressive’ neuroblastoma than that occurring in older children. Therefore, they may receive a less intensive course of chemotherapy than the treatment outlined above.

Stage 4s neuroblastoma (INRSC stage MS)

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Treatment

There are four main types of treatment that are used in the treatment of neuroblastoma: surgery, radiotherapy, chemotherapy and immunotherapy.

Treatment of neuroblastoma is given according to risk group:

- If your child has low risk neuroblastoma it is possible for your child to be treated with either surgery alone, chemotherapy and surgery, or in rare cases, not to be given any treatment at all. Your child will still be monitored closely for many months.

- If your child has intermediate (medium) risk neuroblastoma they will need chemotherapy, often surgery and occasionally radiotherapy.

- It is most common, however, for neuroblastoma to present as high risk neuroblastoma in the toddler or older age group, and this almost always requires strong drug treatment from the outset. The majority of patients with high risk disease are likely to require a combination of different treatments, including surgery, radiotherapy, chemotherapy and immunotherapy.

Surgery
This is an operation to remove the tumour if it is possible. Surgery may be involved at a later stage of treatment, often after several courses of chemotherapy have been given to shrink the tumour so that it can be more easily and therefore more safely removed by the surgeon.

Radiotherapy
This is treatment with high energy x-rays to try and kill the tumour cells. A treatment machine called a linear accelerator targets the x-rays at the tumour from outside the body.
**Chemotherapy**

This is the main treatment for medium and high risk neuroblastoma. Chemotherapy is the use of drugs, usually given through a drip into a vein (intravenously), to destroy cancer cells. Once the diagnosis of neuroblastoma is confirmed, the site, stage and sometimes the results from special genetic tests on the tumour will help the oncologist in charge of your child’s treatment to decide on the best treatment regimen, which is often called the protocol. The suggested treatment will be discussed fully with you. Further on in this booklet you can find a description of how the drugs chosen for your child will be delivered and an explanation of the potential side effects associated with chemotherapy.

For medium and high risk neuroblastoma, the general pattern of treatment in most European centres is to give chemotherapy to destroy neuroblastoma cells throughout the body. After a period of chemotherapy, the position and site of the tumour and any metastases will be reassessed carefully by bone marrow examinations, a CT or MRI scan, mIBG scan and bone scan.

Surgical removal of the primary tumour will then be attempted. Very occasionally, even if the main tumour has shrunk down very well, it may be near the main blood vessels or a delicate body organ that would make surgery complicated and risky. In this case, it may be suggested that the surgeon does not attempt to remove the tumour.

In older children with one type of medium risk neuroblastoma, radiotherapy to the site of the primary tumour may be given after surgery, together with more chemotherapy. This will be fully explained to you by your child’s paediatric oncologist and clinical oncologist (radiotherapy doctor).

**High dose chemotherapy**

After surgery, children with high risk neuroblastoma undergo further treatment with ‘high dose chemotherapy’. Very occasionally, high dose chemotherapy may be given before surgery. The drug or drugs used in this type of therapy may differ from patient to patient but usually comprises high dose busulfan and melphalan. Because the treatment is extremely strong, a child may need to be in hospital for a period of about four to six weeks and be nursed and cared for in an isolation cubicle. The treatment will lower the blood count for a long time during which time the child will be prone to life threatening infections and bleeding. To shorten the period when the blood count is low, children undergo a procedure known as autologous stem cell rescue.

This means that before the high dose chemotherapy is given, ‘stem cells’ are collected, or ‘harvested’, from your child. Stem cells are special cells found in the bone marrow that develop into the different types of blood cells. They can therefore help a child grow new bone marrow when their bone marrow is destroyed by the high dose chemotherapy. The stem cells collected from your child are given back to them after the high dose chemotherapy to allow the bone marrow to recover.

The stem cells come from your child’s bone marrow but are actually collected from the blood using a machine. A special tube (catheter) called a Vascath is put into a vein in your child 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 into a central venous catheter (see page 18). The collected stem cells are then frozen and stored and given back to your child after the high dose chemotherapy. The
Neuroblastoma

harvesting procedure takes about 3-4 hours once a day. Usually two to three days of harvesting are required. This procedure is painless and has very 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 special 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 for harvesting. G-CSF is started a few days after chemotherapy and is given on a daily basis until harvesting is finished. The whole of the harvesting procedure will be discussed in detail with you by your treatment team.

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 on page 9. The bone marrow is then stored like stem cells and given back to your child after the high dose chemotherapy.

After high dose chemotherapy, children over the age of one year with stage 4 neuroblastoma will then receive radiotherapy to the site of the primary tumour, usually the abdomen (tummy). This will be fully explained to you by your child’s clinical oncologist (radiotherapy doctor). Radiotherapy is used to kill any tumour cells that could not be removed by surgery.

For children with high risk disease, additional treatment is given after recovery from the high dose treatment and radiotherapy. A drug that aims to ‘differentiate’ (to turn cells from a cancerous to non-cancerous state) any remaining tumour cells that may be present called 13-cis-retinoic acid is administered. Many children also receive immunotherapy with a monoclonal antibody ‘anti GD2 antibody’ (see below). These treatments are given for 6 months and your doctor will explain this treatment fully to you.

**Immunotherapy**

Immunotherapy is a new form of treatment for neuroblastoma which relies on cells of the body’s own immune system to kill cancer cells. There are different ways to do this but currently an antibody therapy is most commonly used. The antibody acts against a sugar-fat 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. This different way is called antibody dependent or complement dependent cytotoxicity.

There is evidence that other cells in the immune system, such as natural killer cells and macrophages, may promote neuroblastoma cell death. Drugs can be given to increase the number of these immune system cells, but these increased numbers may be responsible for serious side effects associated with the antibody treatment.

In the UK and Europe, the current high risk neuroblastoma clinical trial will soon be testing whether giving anti-GD2 antibody on its own is as effective in causing neuroblastoma cell death (and therefore improving the cure rate) as giving antibody together with interleukin-2 (IL-2) at a lower dose than was used in the trial a few years ago. A lower dose of IL-2 is being tested as the previous higher dose was associated with many side effects which in some cases resulted in the immunotherapy having to be stopped. IL-2 is a substance which increases natural killer cell activity and therefore may improve the effectiveness of the antiGD2 antibody. All patients also receive 13-cis-retinoic acid. You will find a general explanation of clinical trials on page 19.
**Beginning treatment**
When your child starts treatment, you may have to stay at the Principal Treatment Centre during the early stages (one or two cycles of chemotherapy). This will depend on the treatment protocol used.

Often, some of the treatment can be given at your local hospital, under the guidance of the nearest specialist centre. This is called ‘shared care’, and the local hospitals are known as Paediatric Oncology Shared Care Units (POSCU). If your child has any shared care, the decisions about their treatment will still be made by your child’s consultant at the Principal Treatment Centre.

**Other treatments**
If you have any queries about ‘new’ treatments which you may have heard about in the media or from friends, do discuss these queries with your child’s treatment team.

You may feel, very naturally, that you want to explore every avenue for your child and do not want to settle for just one doctor’s advice about treatment.

The oncologist will be able to reassure you that because paediatric oncology, and especially the treatment of neuroblastoma, is such a small and specialised area, all the doctors involved in it tend to know one another and meet frequently. They are naturally aware of the latest research in the field. Therefore if a new treatment were developed anywhere in the world, it is highly probable that your child’s doctor would know of it and be able to discuss it with you.
Possible side effects of treatment

**Nausea and vomiting**
These may occur when drugs are given or after a delay of a day or so. The vomiting caused by some drugs may last for a number of days. Most children are affected to some extent. It seems that babies and very young children have less nausea and vomiting than older children. Over the last few years, several new anti-sickness drugs have made this side effect less severe than in the past.

**Temporary hair loss**
This is the most visible side effect of chemotherapy. It will begin after starting treatment, perhaps in two weeks, and will usually continue until the whole treatment plan is completed, when the hair grows back quite quickly and normally. The idea of hair loss is usually very upsetting to parents and, of course, 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. Some older children may like to have a wig for special occasions. However, most children do not want to bother with this and wear a scarf, hat or nothing at all.

**Depression of the blood and bone marrow**
Normal parts of the blood, the neutrophils which fight bacterial infections, and the lymphocytes which combat viral infections, are damaged by chemotherapy and are, therefore, low in number. This means that your child is at risk of getting infections during the course of treatment, a number of which are caused by the ‘germs’ that normally live harmlessly in a child.

The blood cells are not killed permanently. However after each course of drug treatment their numbers become very low for some days, but then new ones begin to grow again.

The other parts of the blood, the platelets and the red cells, are also reduced in number by chemotherapy. If the platelets become low, then your child is at risk of bleeding more easily.

If, for example, your child has a troublesome and prolonged nosebleed whilst their platelets are low, a platelet transfusion will be given. Also your child might become anaemic because of low numbers of red cells. If this happens, they will receive a blood transfusion.

The amount of haemoglobin (measured by the number of red cells), the number of platelets and the number of neutrophils in the blood is referred to as the ‘blood count’. It is usually necessary to have a certain number of neutrophils and platelets before beginning each course of treatment with drugs. Therefore, the blood count is checked before starting chemotherapy. It is the recovery time of the blood cells that determines the minimum length of time between courses of chemotherapy.

If your child is receiving an intensive drug regimen, the drugs will usually be given according to the protocol, whatever the blood count. This may mean that your child's blood count is low for much of the treatment and consequently will need to be under close observation at all times for signs of infection, which would then be treated swiftly with intravenous antibiotics.

The reason that the hair cells and the blood cells are particularly affected by chemotherapy is that they are cells that multiply quickly. Neuroblastoma cells, in common, with other cancer cells, divide very rapidly and chemotherapy is designed to kill rapidly dividing cells, so the drugs kill off many of the blood and hair cells just as they do the cancer cells.

**Weight loss**
Both neuroblastoma and chemotherapy may cause weight loss. If this occurs, your child's doctor will consider different methods of providing adequate nutrition. This may be by dietary supplements (such as milkshakes and calorie powders), feeding through a tube passed via the nose into the stomach, or by feeding into the vein using the long line – this is called total parenteral nutrition (TPN).
**Damage to the kidneys and hearing**

Some drugs, such as cisplatin, can potentially damage the kidneys and also affect your child’s hearing. 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, an injection of radioactive substance (not harmful to your child) is given into a vein in the hand or arm. Blood samples are then taken.

The radioactive substance should be steadily flushed out of the blood through the kidneys and the level of the substance remaining in your child’s blood stream is a good indication of how well the kidneys are doing their job. If the kidneys were not functioning well as a side effect of chemotherapy, this would be noted and the chemotherapy adjusted if necessary.

**Constipation or diarrhoea**

Some drugs can change the way the bowel works, so your child may have diarrhoea or constipation. Let the hospital staff know if constipation becomes a problem, as laxatives can be given to relieve it.

Diarrhoea usually gets better without medication. It’s important for your child to drink plenty of fluids if they have diarrhoea. 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**

Some drugs can cause a sore mouth, which may lead to mouth ulcers. Mouth care is very important – 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.

**Veno-occlusive disease (VOD)**

VOD is one of the less common but still serious complications which can occur during high dose chemotherapy and stem cell transplant (SCT). Veno-occlusive disease is not another illness, but a complication that affects the liver.

Veno-occlusive disease may also be referred to as sinusoidal obstruction syndrome (SOS). VOD can range from mild to severe, and occurs in approximately 10 - 30% of children/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.

In VOD the chemotherapy causes damage to cells in the liver, so the veins within the liver become increasingly occluded (obstructed) by cellular debris (dead or damaged cells), causing the blood flow from the liver to back up. The protein-rich fluid content of the blood leaks out into the peritoneal cavity (a potential space between the two membranes that separate the organs in the abdominal cavity from the abdominal wall). There will be an increase in weight as the fluid collects in this space. As the tummy gets bigger it will become uncomfortable and may be quite painful requiring pain relief medicine.

**Fertility**

Parents are also usually concerned about the effects of chemotherapy on their child’s potential fertility, or ability to have children. A very long term outcome of present treatments for neuroblastoma is difficult to predict, but, for example, it is known that the drugs cyclophosphamide, busulfan and melphalan particularly when given in high doses, are usually damaging to fertility. However, this side effect has to be weighed against the potential benefit of using these drugs. Busulfan and melphalan were found to be superior to another high dose chemotherapy regimen in the current high risk neuroblastoma clinical trial in treating neuroblastoma.
Central lines and ports

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

**Central lines**
A central line (central venous catheter) is a thin, flexible plastic tube that is inserted into a vein, usually near the collarbone. A central line is often called a Hickman line, after the person that developed it. You may also hear it referred to as a ‘wiggly’, a more child-friendly name used in many centres.

A central line is inserted under a general anaesthetic. The surgeon makes a small cut into a vein near the collarbone and feeds the tube down until the tip is in one of the large veins near the heart.

The other end of the tube is then tunneled under the skin and comes out on the front of the chest. A removable bung is attached to the end of the line, which allows samples of blood to be taken or medicines to be injected. It can also be used to give blood transfusions.

A central line needs to be kept dry when showering or bathing – a plastic dressing can be used for this. The central lines have clamps on them which should be closed when the line is not being used.

**Implantable ports**
Some children may have an alternative type of central line, called a port (sometimes called a ‘Port-a-cath’). Again, this is used to give drugs and take blood. A port is a completely implantable central venous catheter system. It is different from other lines in that it has no part exposed outside the skin. The tubes don’t come out through the skin. Instead, they end in a reservoir or port that is under the skin below the collarbone, so they aren’t clearly visible.

Implantable ports are inserted under general anaesthetic. A small needle is pushed through the skin into the port to give chemotherapy or take blood. The skin over the port can be numbed beforehand with an anaesthetic cream or spray. The needle can stay in place for up to seven days and is secured in place with a dressing.
Clinical trials

Most children being treated for neuroblastoma at one of the Principal Treatment Centres in the United Kingdom are offered cancer treatment that is part of a clinical trial. Trials are done for a number of reasons. Remember, any treatment suggested for your child will be the best treatment for them. If a trial is suggested, you’ll be given information about it and what is involved. You will have to consent to the trial as well as to the treatment itself.

Improvements in treatment of the disease have come through clinical trials. These ensure that the effects and outcomes of particular aspects of treatment are measured consistently and reliably over time, to see if they improve survival. Clinical trials may also involve the comparison of different treatments where it is not known whether one drug or procedure is more effective than another.

Informed consent
Before your child is asked to take part in a clinical trial, the treatment team will explain what the trial is aiming to achieve, and the risks and benefits of taking part in it. It will only be after you have had the trial fully explained and had time to consider the matter that you will be asked whether you give your consent for your child to take part. If you do agree you will be asked to sign a form giving your informed consent. The treatment team will not put pressure on you to agree, and your child will receive the best treatment available whatever you decide.

Deciding on whether to take part in a trial can add to the stress of coping with a sick child and the bewildering range of processes associated with treatment. It may seem as though the treatment team is passing over to you responsibility for deciding what treatment your child should be given. In practice, however, the treatment team will only invite you to join a trial when it is considered in the best interests of your child, and where both you and your child and the advancement of neuroblastoma treatment are likely to benefit.

Randomisation
If you agree to take part in a trial the decision about which of the treatments being tested will be given to your child may be decided by random allocation, or randomisation. This takes the decision out of the hands of the treatment team, ensures that there is no bias in who receives which treatment, and ensures that the results of the trial are valid and meaningful.

Because this is such a rapidly changing field, it is not possible to give details about any particular procedures now being tested, but you may be reassured that your child will never be subjected to any test, procedure or ‘new’ drug without your full knowledge and permission.

Information about current clinical trials for neuroblastoma can be found on Neuroblastoma UK’s website, www.neuroblastoma.org.uk

The CCLG factsheet ‘A guide to clinical trials’ is designed to help parents and young people understand more about clinical trials and answer some of the many questions that people have about entry onto clinical trials.
Treatment abroad

There has been some media coverage of families who have decided to take their child with neuroblastoma abroad for treatment. The decision to take a child abroad for treatment for neuroblastoma is always one best taken by the child’s family and the consultant paediatric oncologist caring for the child. A lot of factors are taken into account in reaching that decision. Some children might not be eligible for a particular treatment in the UK, for example immunotherapy. For other children, going abroad will provide the opportunity to receive unique experimental therapies that are not available in the UK but are available in single centres in other countries.

In the UK, all patients in first remission registered on the current European High Risk Neuroblastoma Trial are eligible for immunotherapy. For UK patients who relapse there are a number of treatment options available in the UK including further chemotherapy, early phase clinical trials, targeted radiotherapy and, increasingly, access to immunotherapy treatment for patients who did not receive it in first remission.

Unfortunately, with very few exceptions, children with relapsed high risk neuroblastoma will not survive their disease. However, prolonged periods of disease control can sometimes be achieved with treatments available both here in the UK and elsewhere.

Relapsed or refractory high risk neuroblastoma

If high risk neuroblastoma comes back during or after the initial treatment (relapsed disease), it is often possible to control the disease for years, although it is usually very difficult to achieve a complete cure.

Sometime high risk neuroblastoma does not respond to the initial treatment (refractory disease). If your child has relapsed or refractory high risk neuroblastoma, choosing which treatment is right for your child will depend on a number of things, including where their disease is and what treatment they have previously been given. Your child’s own doctor will be able to help you decide which the most appropriate treatment for your child is at this time.
Coping with neuroblastoma
Talking to your child

One of the hardest parts of caring for a child with cancer is knowing what to say and how much information to give them.

Answering questions honestly is best. 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 doctors or nurses for guidance on how to talk to your child. There are also booklets available from CCLG, CLIC Sargent and Macmillan Cancer Support 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. Older children may be more frightened of pain. It can help to explain that there are good painkillers available to help control any pain they have. Doctors and nurses will be happy to explain more about this and can help you reassure your child.

Coping with tests

Whatever your child’s age, a truthful approach to what is happening is also needed when helping to prepare them for tests and investigations. It is very tempting to try and calm a frightened toddler by the reassurance that “….it won’t hurt…..”. If this is not true, your child may lose trust in you and feel very insecure. If you can, it is better to find something positive in a nasty situation. You may say, for example, “…Yes, this finger prick will hurt a bit just for a moment, but Mummy/Daddy will stay with you all the time and then we’ll go to the playroom/make a drink/read a story….etc.”.

Asking your child to be a “big girl” or “brave boy” can be unhelpful when the child is frightened. Approaches such as “We will all shout ‘ouch’ together when the prick happens” can help the child to know that the adults appreciate that it hurts a bit and it isn’t shameful or babyish to cry.

Specialist members of the treatment team, like the play specialist and the school teacher, will have books and games and specially adapted toys through which to familiarise your child with the procedures and to help them express feelings through play. A favourite with the young child is often the ‘hospital corner’ of the playroom or, for a child in bed, a bandaged teddy who receives more than his fair share of ‘injections’. This often incidentally helps your child to make sense of, and have some control over, what is happening to them.

Older children and teenagers need other types of ‘distractions’. Often, they need to feel that they have some control over their treatment, for example, if possible, choosing whether they have a scan this afternoon or tomorrow.
You and your family

Having a child diagnosed with cancer will affect you as parents or carers, your other children, and the people close to you. Understandably, you’ll feel worried and stressed as you come to terms with the diagnosis and what it means for your child to go through treatment. You’ll worry that your child will suffer and that your family life is going to be completely disrupted. At first, you may worry that your child is going to die.

When you’re first told the diagnosis, you may feel numb, confused, or unable to hear or remember information about your child’s diagnosis or treatment. You may also feel overwhelmed by painful and powerful emotions. These reactions are normal – remember the doctors and nurses are there to help you at this time. There are also other people and organisations that can offer support (see pages 34-35).

Seeking information
You may want to find out as much as possible about the cancer and its treatment. There’s a lot of information on the internet but not all of it is reliable, so talk to the doctors about where to look. The best advice is to use a website by a recognised organisation such as CCLG, Macmillan or Cancer Research UK.

The CCLG can send you a leaflet entitled ‘How can the internet help us?’ It gives tips on finding reliable and user-friendly information about cancer care and treatment on the internet.

Everyone is different – you may read about a child in a similar situation to yours, but they may have a very different experience. Remember, too, that statistics are only helpful to a degree. They’re based on large numbers of people and can’t say what will happen with your child in particular.

You can ask the doctors and nurses questions again if you’ve been unable to take it all in. It may help to have someone else with you, and to write down your questions beforehand.

Feelings and emotions
Some of the feelings and emotions you may have are described briefly below. Your feelings are likely to change over time and you may not experience all the emotions described here.

Shock
This is one of the most common feelings experienced when first faced with a cancer diagnosis. This is a completely normal reaction which can in the short term cause a range of physical and emotional symptoms such as lack of sleep, lack of appetite, nausea and anxiety.

Fear and denial
You may feel scared, anxious, and panicky about what’s going to happen and what the future holds. Understandably, parents often want to deny that such a terrible thing could happen to their child. Some people may feel tempted to take their child from one doctor to another. At times, you may feel the fear is almost too much to bear, such as when your child is going into the operating theatre and you wave goodbye.

Sadness
It’s natural to feel sad or depressed at times. Every parent wants their child to be healthy, happy and carefree. Cancer and its treatment can have a big impact on you and your child’s life. At times, you may have feelings of hopelessness. You may find it difficult to eat or sleep, or feel as though you have no energy for the things you need to do each day.
Parents often say that they feel overwhelmed by the enormity of the situation. These painful and unpleasant feelings can’t be avoided, and you’re likely to have them at various times during your child’s illness. It’s important to have support to help you through these times.

**Guilt**
It’s very common for a parent to feel guilty if their child has cancer. Some people wonder if it was something they did or didn’t do that caused the cancer, or they feel that it’s a punishment for something they did in the past. Sometimes, parents blame themselves for not noticing their child’s symptoms quickly enough.

Many parents will have strong feelings of guilt, but it’s important to remember that you’re not responsible for causing your child’s cancer.

**Anger**
It’s normal to feel angry at times. You may feel angry with the hospital staff for putting your child through tests and treatment. You may feel angry that you have to cope with such uncertainty, and the unfamiliar world of hospitals, doctors and nurses. Some people even find that they’re angry with their child, as it’s their illness that is causing so many problems for the family. This can be distressing but it’s also normal.

Parents can feel angry at each other, especially if they have different ways of coping with their child’s illness. For example, one parent might want to talk about it a lot and the other might just want to get on with normal life as much as possible.

You may also feel angry with family or friends who make thoughtless remarks or are too busy to give you support. Or you might feel frustrated with people who avoid you because they don’t know what to say.

**Looking after yourself**
It’s important to take care of your own needs and to not feel guilty about doing so. For example, eating and sleeping well, exercising if possible, dealing with any health problems and taking regular breaks will help you cope and care for your child.

Parents often find it hard to express their grief and fears to each other, with the result that they bottle up their feelings, become tense and quarrel more than usual.

Different people have different ways of coping with stress and what helps one can irritate another. If you can, try to talk to each other as openly as possible, and be as supportive and patient as you can.

You may find the following CCLG booklets helpful:
- Children and young people with cancer: A parent’s guide
- Family life and cancer
- Grandparents
- Helping brothers and sisters
Brothers and sisters

The brothers and sisters of a child with cancer may have many or all of 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 need to be cared for by family members or friends. They may have a lot of time away from you and find their daily routine keeps changing. As well as worrying about their brother or sister’s health, they may also feel resentful of all the attention they’re getting. This can make them feel very left out and angry. They may worry that they’re also going to get ill.

It can help to plan some time to let your other children be the centre of attention for a while, perhaps by going out for a meal or to see a film. Even 10 minutes at the end of a day can make a big difference. If your child with cancer is having some special attention – for example, having sweet treats or their favourite things to eat – you can do the same for other children in the family so they don’t feel left out.

The needs of brothers and sisters can sometimes be overlooked, particularly in the early months when you may spend most of your time caring for your ill child in hospital or at home. Many brothers and sisters keep their feelings bottled up inside to avoid worrying their parents.

Often, the place where siblings may show how they feel is at school. They may:

- withdraw and become very quiet
- become disruptive in the classroom
- cry easily
- become frustrated and have outbursts of anger
- fall behind in class work
- get lower marks than usual
- start missing school
- become rebellious towards teachers
- have arguments and fights with friends and other children in their class

It will help to let your children’s head teacher(s) know that their brother or sister has been diagnosed with cancer. You can ask for the school’s help and support for your children. The teachers will understand that feelings may be expressed through behaviour at school, once they’re aware of the stresses facing the family.

If a sibling is obviously having difficulty dealing with the situation, talk about it with staff at the hospital such as the specialist nurses or the social worker. They can arrange for counselling, help and support for you and your other children. Some hospitals have support groups for siblings.
Going home

As soon as the diagnostic tests and first treatment cycles are over, you may be able to go home. Despite your obvious relief at being told you can leave hospital, and your pleasure at being reunited with the rest of your family, getting back to normal routine may take a little time. Probably you have pinned a lot of hope on this as the time when your child will pick up, eat more normally again and temporarily forget about hospital.

All these things will hopefully be the case, but there may be some snags at first. There may be some sense of anti-climax as you and your child may have become ‘hospitalised’ to some extent.

You may find it difficult to sleep and eat normally at first. You may even, perhaps to your surprise, miss the company of other parents, the sharing of hospital routine and the security offered in the ward surroundings. Your child may be extra clingy and demanding just when you want to try and give more attention to other family members who have missed you.

Many parents who have previously been quite confident in their manner of caring for and bringing up their child find themselves worried and doubtful about applying even the most common sense rules.

The team at your child’s Principal Treatment Centre are still there to offer you advice and support. You may also find your GP and the community team an important source of support.

Behaviour
The question of discipline is a difficult one. Maintaining normal discipline is reassuring for your child and can help them feel secure.

Sleeping
If your child has had you by their side constantly in hospital, they may be more clingy at home or reluctant to sleep at first in their own bed again. However, it is usually best to remind them, as gently and firmly as possible, that home habits and hospital ones are separate and that at home they have their own cot or bed. At first your child may need to check your presence constantly by calling or coming into your room. Hopefully they will settle into the home routine again when they know that will not ‘give in’ and allow behaviour that was not allowed before their illness.

Eating
In the case of neuroblastoma there are no particular restrictions on diet and no set medicine to take between hospital treatments. Some parents have found that as treatment progresses the child does regain energy and appetite between or after each course of chemotherapy.

Eating is often an issue that causes concern to many parents. Your child may have lost weight, firstly because of the neuroblastoma itself and then because of the drugs that may have caused sickness and loss of appetite. Some children do not regain their former appetite or weight until the whole course of drug treatments has finished.

Unless your child’s doctor is worried about excessive weight loss it will be considered fairly normal for them to remain rather thin throughout treatment. You may find it helpful to talk to the dietician at the hospital, who may be able to suggest way of presenting nourishing and weight building food. Sometimes supplementary feeding through a tube from the nose to the stomach, or even TPN (Total Parenteral Nutrition) through the central line is needed.
Reducing the risk of infection

It is very tempting to become overprotective of your ill child and perhaps of your healthy ones too in view of what has happened. You have probably been told by the treatment team to allow your child to do whatever they feel able, and it is very good to encourage them to regain their old skills and confidence as they feel better.

Much of your desire to protect your child may be the result of medical advice to keep them away from sources of infection. Many parents agonise about the extent to which their child should mix with other children. You should follow the advice of your child’s doctor, but on the whole they should enjoy the benefits of mixing with others, with sensible precautions.

You will have been told that measles and chicken pox are diseases your child should avoid if possible. Whilst this is not always easy, it helps to enlist your neighbours, friends, child’s teacher, possibly health visitor and your local doctor, to let you know if any child in your community has either got, or been in contact with, these infections. This also applies to the teachers of any of your other children. You cannot disrupt their schooling, but you should be extra cautious with your sick child if a brother or sister has measles or chicken pox in their class.

Even if your sick child does come into contact with measles or chickenpox there is no need to panic, but you must let a doctor know immediately. It may be possible to give your child a special injection in case of chickenpox, (called ‘ZIG’ which stands for Zoster Immune Globulin) either to reduce the attack or to prevent it completely. Some centres use an alternative drug called acyclovir instead of ZIG for this purpose. Zoster is the name of the chickenpox virus. It is identical to the virus causing shingles, which should also be avoided. If you have concern about possible contact, telephone the hospital or your doctor for advice.
Children with cancer often have gaps in their education. This can be due to going into hospital, the side effects of treatment, or generally not feeling well enough to fully take part in daily school life. Most children’s cancer hospitals have education departments that can support your child while they’re in hospital. The teaching staff at the hospital will contact your child’s teachers to make sure they can continue their schooling whenever they feel well enough. It’s even possible for children to take exams in hospital if necessary.

As your child’s health improves and if treatment allows, going back to school may be a relief or a challenge. For many children, school is a refuge from the world of hospitals and procedures – a place for fun, friends and learning. Going back to school can be a sign that life is returning to normal.

However, some children, especially teenagers, may dread going back to school. This may be because of temporary or permanent changes in their appearance. Or they may worry that they’ll have missed a lot of work, or that being away will have affected their relationships with their friends.

Bullying may be an issue for some children at school, especially more vulnerable children. You can get advice and support from the school and other organisations (see pages 34-35) if you think your child is being bullied.

If treatment has affected your child’s ability to learn, this can be a major frustration for them and may affect their confidence and self-esteem. The school can give extra help for children with learning difficulties. Talk to the teachers at school if you think your child may have problems.

Teachers have an important role to play in helping your child settle back into school, and helping other pupils understand what has happened to your child.

Keeping teachers informed
It’s important to let the school know how your child is doing. As soon as your child is diagnosed, contact the head teacher to tell them what’s happening. It can help to let the school know about the plans for treatment. The school teachers can then work with the hospital education department to make sure they cover the same work as the rest of the class. It can help for the teachers at the hospital to let the school know if your child is emotionally or physically fragile. The school teachers can then take this into account.

At any stage of treatment, your child should be involved in letting the teacher know what information they would like to be shared with their classmates.

Risk of infection at school
For most children on cancer treatment returning to school is recommended, even when their immune system is low. It is important that they carry on with as normal a routine as possible. Most infections that children on cancer treatment pick up are not from other people or children. Chickenpox, measles or shingles can be dangerous to children who have a low immunity due to cancer treatment.

The school can develop a system to let other parents know that they should notify their child’s teacher if their child develops chickenpox, measles or shingles, so that appropriate action can be taken.

If your child has been exposed to chickenpox and has not had it before, contact the hospital straight away. It may be necessary to give your child some medicine to prevent chickenpox developing.

It can be difficult to get the balance right between letting your child mix with their friends and worrying that they might pick up an infection. You can discuss this with both the hospital staff and the teachers at school to make sure you’re happy with what your child does.
Keeping up with schoolwork

It's important for your child to try to keep up with schoolwork whenever they can. Learning can continue outside school. By speaking regularly to the teacher, you’ll know which subjects are being covered. Often, the teacher will send assignments and materials home with siblings or arrangements can be made to collect them.

Your child should have an individual education plan in place. This plan can be shared between your child’s school and hospital school to ensure there is consistency in what is being taught.

To help your child keep up in school, you may need to ask for a special education statement. This qualifies your child for extra help.

Very young children

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 at nursery 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 nursery you may feel there’s 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 nursery or pre-school
• your child’s social needs can be met by siblings and/or other children outside of the nursery
• your child is well enough to attend nursery or pre-school
• your child has already had chickenpox

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

The CCLG booklet ‘Welcome back!’ is a guide for teachers helping children and young people returning to school after a diagnosis of cancer.
After treatment, and follow-up

Understandably, children who have been in hospital can often be difficult and demanding when they go home. Younger children may behave in a more childish way, whereas older children may be aggressive or jealous of their healthy brothers and sisters. Maintaining discipline and having their friends over to visit can help children get back into everyday life.

While it’s often a relief to get to the end of treatment, you may find that you feel more anxious and worried now that you and your child are not attending the hospital so regularly. This is normal – all of a sudden you don’t have the immediate reassurance from the doctors and other staff.

You may also be worried that symptoms will return once treatment ends. Remember, the hospital staff are still there to help you and will understand your concerns. It’s important to contact them if you have any worries about your child’s health. Despite all the improvements in cancer treatment, sometimes it doesn’t work. In this case another treatment may be available, but this isn’t the case for all children.

There’s a lot of support for families when a cancer comes back. Further information is available from CCLG, Macmillan and CLIC Sargent.

If your child cannot be cured

Sadly, sometimes parents are faced with the devastating news that there are no more treatment options available for your child, which means that a cure is now no longer possible.

The goal shifts from being able to cure your child’s disease to providing the best quality of life for your child and managing their symptoms. You and your child will be looked after by a specialist team with expertise in managing symptoms and providing emotional and practical help and support to the whole family.

Palliative care
Palliative care is the phase of treatment where cure is no longer possible, and is concerned with managing symptoms and quality of life.

The aims of palliative care services are:
- To arrange services to meet the specific needs of your child and your whole family
- To manage symptoms promptly
- To provide care where you want it (as far as possible)
- To provide your whole family with as much support as you need

CCLG has a range of booklets offering support if your child cannot be cured.
Further information
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 neuroblastoma 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.

**Alopecia**
Loss of hair

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

**Biopsy**
Removal of a small piece of tissue for examination, 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. See p11 for further information.

**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 (found more often than normal) 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 user 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 scans
The sound waves produced by a scanner bounce from solid organs inside the body and are recorded on a screen. Allows doctors to see the outlines or shadows of normal organs and tumours.

Vanillyl mandelic acid (VMA)
A substance found in the urine in raised amounts when a child has neuroblastoma
Useful organisations

Neuroblastoma UK
www.neuroblastoma.org.uk
Tel: 020 8940 4353
Email: info@neuroblastoma.org.uk

Neuroblastoma UK is the leading UK charity devoted to funding research into the causes and treatment of this aggressive childhood cancer. Neuroblastoma affects about 100 children each year in the UK and, while outcomes have improved overall in the past 20-30 years, they remain poor for those with the higher-risk variants of the disease. Founded in 1982 as the Neuroblastoma Society, we have funded millions’ of pounds worth of research and at any one time have upwards of 10 research projects under way, some in conjunction with other charities. Our Board of Trustees comprises parents and others impacted by neuroblastoma who take responsibility for all aspects of our activity, plus specialist medical trustees who also advise on clinical and research matters. Some 95% of our revenue goes directly to support research.

In addition to funding research, we facilitate communications and collaboration between UK and international researchers, notably through our biannual research symposium which attracts up to 200 researchers, and support UK participation in Europe-wide clinical trials. We work closely with other organisations, such as CCLG, to raise awareness and campaign for better funding and treatment of childhood cancer. Finally, through successive editions of this booklet and other channels, we provide information and support to families affected by neuroblastoma.

Children’s Cancer and Leukaemia Group (CCLG)
www.cclg.org.uk
Tel: 0116 252 5858
Email: info@cclg.org.uk

CCLG is the professional association for those involved in the treatment and care of children with cancer in the UK and Ireland. Through a network of Principal Treatment Centres, CCLG members together with other professionals are responsible for organising cancer treatment for children in the UK and are dedicated to improving best practice and outcomes.

CCLG is also a registered charity, providing high quality, award-winning information about childhood cancer to patients and families. We also fund and support research into childhood cancer.

SIOPEN
www.siopen.org
SIOPEN brings together clinicians from across Europe and beyond to improve the outcomes for patients with neuroblastoma.

International collaboration is vital because neuroblastoma is uncommon. Collaboration ensures there are enough patients to do clinical studies. This allows us to learn more about the condition and improve treatments. SIOPEN runs a number of large clinical trials examining how best to treat neuroblastoma.

In addition, it leads research into the biology of neuroblastoma to provide a better understanding of how the disease develops and how these different types respond to treatment. Speciality Committees focus on specific areas such as biology, pathology and surgery.
Action for Sick Children
www.actionforsickchildren.org
Helpline: 0800 074 4519
Provides advice and information for parents with children going to hospital. Offers counselling for parents and has leaflets and videos for children.

Cancer Research UK
www.cancerresearchuk.org
Information on all cancer types and a key funder of research into cancer.

CLIC Sargent
www.clicsargent.org.uk
0300 330 0803
Provides practical support and advice for children and young people affected by childhood cancer and their families and offers accredited information for patients.

Macmillan Cancer Support
www.macmillan.org.uk
0808 808 0000
Provides practical medical and financial support to anyone who is affected by cancer.

Solving Kids Cancer Europe
(Formerly Neuroblastoma Children’s Cancer Alliance)
www.solvingkidscancer.org.uk
Solving Kids’ Cancer(Europe) helps families affected by high-risk childhood cancers by supporting access to treatment, research, parent education and raising awareness.

Teenage Cancer Trust
www.teenagecancertrust.org
Raises funds and supports teenagers fighting cancer.
Children’s Cancer and Leukaemia Group is a leading children’s cancer charity and the UK and Ireland’s professional association for those involved in the treatment and care of children with cancer. Each week in the UK and Ireland, more than 30 children are diagnosed with cancer. Two out of ten children will not survive their disease.

We bring together childhood cancer professionals to ensure all children receive the best possible treatment and care. We fund and support research into childhood cancers, and we help young patients and their families with our expert, high quality and award-winning information resources.

If you have any comments on this booklet please contact us at the address below

CCLG publications on a variety of topics related to children’s cancer are available to order or download free of charge from our website

Children’s Cancer and Leukaemia Group 0116 252 5858
University of Leicester info@cclg.org.uk
Clinical Sciences Building www.cclg.org.uk
Leicester Royal Infirmary
Leicester LE2 7LX

Registered Charity number 286669

Neuroblastoma UK is the leading UK charity devoted to funding neuroblastoma research. We work closely with other charities to achieve this, and also to provide information and support to families and to raise awareness of the disease among the public, government bodies and clinical professions.

Neuroblastoma UK 020 8940 4353
54 Forest Road info@neuroblastoma.org.uk
Richmond www.neuroblastoma.org.uk
TW9 3BZ

Registered Charity number 326385

NEUROBLASTOMA UK
FIGHTING CHILDHOOD CANCER

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