Neuroblastoma

Neuroblastoma is a cancer affecting developing nerve cells. Neuroblastoma is a rare cancer and most often occurs in children under 5 years old. Neuroblastoma often starts in the tummy (abdomen) but can spread to other parts of the body, especially bones, liver and skin.

The treatments for neuroblastoma include surgery, chemotherapy and radiotherapy. The outcome (prognosis) depends on the stage of the neuroblastoma when it is first diagnosed.

What is neuroblastoma?

Neuroblastoma develops from particular types of nerve cells called neuroblasts. 'Neuro' means nerve cell and 'blast' means in an early stage of development. Neuroblastoma therefore means a tumour of developing nerve cells.

Neuroblastoma often starts in the tummy (abdomen) and often affects the adrenal glands or the nerves at the back of the tummy. Neuroblastomas can spread to other parts of the body, especially bones, liver and skin.

Neuroblastomas often produce chemicals called catecholamines. Catecholamines are normal chemicals in the body. However, some neuroblastomas produce large amounts of catecholamines and the effects include increased heart rate, high blood pressure and diarrhoea.

See separate leaflet called Cancer - A General Overview for more general information about cancer

How common is neuroblastoma?

Neuroblastoma is a rare cancer that affects children, mostly under the age of 5 years. About 100 children are diagnosed with neuroblastoma each year in the UK. Very rarely neuroblastoma can also occur in older children, teenagers and adults.

What are the causes of neuroblastoma?

The cause of neuroblastoma is not known. There may be a history of other family members also having a neuroblastoma but this is rare.

Neuroblastoma symptoms

Neuroblastoma usually develops in the tummy (abdomen) and causes a lump (tumour) in the tummy. This may also cause swelling of the tummy with discomfort or pain.

Less often the neuroblastoma affects the spinal cord, causing numbness and weakness in the legs. Neuroblastoma may also first be seen as a lump in the neck.

Neuroblastomas spread to other parts of the body by the time of diagnosis in about half of all affected children. The symptoms will often include tiredness, high temperature (fever), weight loss and loss of appetite.

Other symptoms again depend on which part of the body is affected:

- Bones are the most common place to which a neuroblastoma may spread. Symptoms include pain and swelling over the bone. If the spine is affected then this may also cause numbness and weakness in the legs.
- Bone marrow. This may cause tiredness and looking very pale due to a low red blood cell count (anaemia), bruising or bleeding (low platelet count) and frequent or prolonged infections (low white cell count).
- Skin. This may cause the affected skin to become blue-black in colour, similar to a bruise.

- Liver. This may cause pain and swelling in the tummy. The liver may not work properly and this may lead to skin or the whites of the eyes becoming a yellow colour (jaundice).

if the neuroblastoma makes high levels of the chemicals called catecholamines then this may cause weight loss, increased sweating, redness of the skin (flushing), a fast heart rate and watery diarrhoea.

What tests are used to diagnose neuroblastoma?

Any child or adult who has any symptoms suggesting the possibility of a cancer such as neuroblastoma should be seen urgently (within a maximum of two weeks) by a specialist.

The tests to make a diagnosis and to see whether the neuroblastoma has spread to other parts of the body will include: blood tests, chest X-ray, CT scan, MRI scan, and a bone scan. Urine tests may be used to measure the amount of catecholamines in the body.

A nuclear medicine scan called a MIBG scan can be used to diagnose neuroblastoma. Taking a sample (biopsy) of the lump (swelling), and a having bone marrow biopsy will also help to confirm the diagnosis and help to decide the best way to treat the neuroblastoma.

What are the stages of neuroblastoma?

The stage of a cancer such as neuroblastoma is used by doctors to decide on the best treatment for the neuroblastoma. Systems used to find the stage of the neuroblastoma often include CT scans. The stages for neuroblastoma are:

- **Stage 1 and 2 (also called stage L1)**. The neuroblastoma is in one area of the body and has not spread to any other part of the body.
- **Stage 3 (also called stage L2)**. The neuroblastoma has spread but only into nearby structures.
- **Stage 4 (also called stage M)**. The neuroblastoma has spread to distant parts of the body.

- **Stage 4S (also called stage MS)**. This stage has a better outcome (prognosis) than other stage 4 neuroblastomas. Stage 4S means the child is younger than 18 months at diagnosis, the neuroblastoma has not spread to the bones, and less than 1 in 10 of the cells in the bone marrow are neuroblastoma cells.

As well as the different stages, risk groups are used to assess the risk of the neuroblastoma coming back after treatment. The three risk groups are low-risk, intermediate-risk and high-risk. The risk group is calculated from:

- Age. Children younger than 18 months are at lower risk.
- The appearance of the tumour under the microscope (histology). Tumours with more normal-looking cells are lower-risk.
- Genetic changes. Some neuroblastoma tumours have too many of a gene called MYCN, which controls the growth of the cell. These cells grow quickly and are less likely to develop properly. Tumours with too many MYCN genes are higher-risk. Other gene changes in the neuroblastoma cells can also help to determine risk. See separate leaflet called Genetic Testing for more details.

What are the treatments for neuroblastoma?

The treatment depends on the stage and the risk group. Very young children with low-risk neuroblastoma (stage 4S or MS) may not need any treatment. This type of neuroblastoma can sometimes disappear on its own. Therefore, these children are closely monitored by specialist doctors but may not need any treatment.

The treatments for other children with neuroblastoma may include surgery, chemotherapy and radiotherapy. The treatment will depend on the risk group. Children with low-risk neuroblastoma may just need surgery, with or without chemotherapy. Children with intermediate-risk or high-risk neuroblastoma usually need a combination of surgery, chemotherapy and radiotherapy.

High-risk neuroblastoma (stage M)

Treatment for high-risk neuroblastoma usually starts with chemotherapy to reduce the spread of the neuroblastoma (this is called induction treatment). This is then followed by surgery and then radiotherapy.

Further treatment for high-risk neuroblastoma involves high-dose chemotherapy with a stem cell transplant. This is followed by maintenance treatment to reduce the chance of the neuroblastoma coming back after treatment. Maintenance treatment may include a combination of a medicine called isotretinoin and monoclonal antibody treatment.

Neuroblastoma coming back after treatment (recurrent disease)
The treatments will again include a combination of surgery, chemotherapy and radiotherapy. Disease which returns may need more intensive treatment than the initial treatment that was needed for the neuroblastoma.

What are the complications of neuroblastoma?

The complications of neuroblastoma depend on which parts of the body are affected. Although modern treatments are very effective and life-saving, the treatments may also cause long-term complications.

The risk of complications caused by treatments is very small compared with the benefits of treatments, which are often life-saving and successfully cure the neuroblastoma. The complications of treatments are also becoming less common as treatments improve. However, they may include infertility in later life, hearing problems and an increased risk of developing another cancer.

What is the outcome (prognosis)?

The outcome depends on the stage and risk group. The outcome can be excellent if the neuroblastoma has not spread. However, the outcome for children with high-risk neuroblastoma may be poor, with a high risk of disease returning.

Further reading

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