

Neuroblastoma



Neuroblastoma occurs when malignant cancer cells form in the nerve cells of the sympathetic nervous system involved in the development of the nervous system and other tissues.

Neuroblastoma most commonly occurs in one of the adrenal glands situated in the tummy or in the chest, neck, abdomen, pelvis or the nerve tissue that runs alongside the spinal cord. The adrenal glands are specialised glands that release hormones that help the body respond to stress and maintain blood pressure.

Neuroblastoma may be present at birth, but generally presents in early childhood, before the age of 5 years. In most cases, by the time it is diagnosed the cancer has usually already spread to areas outside of the original site, often to the lymph nodes, bones, bone marrow, liver, and skin.

Approximately 25% of newly diagnosed neuroblastomas are in children under the age of 1 year, and for some reason this age group has the best cure rate, at 90%.





Signs and Symptoms

Symptoms of neuroblastoma only begin when the tumour starts pressing on nearby tissue as it gets bigger, or when the tumour spreads to bone. The symptoms mimic those of many other diseases, which makes diagnosing neuroblastoma very difficult.

Some of the typical signs and symptoms of Neuroblastoma are:

Symptoms vary depending on where the tumour is situated. Initial symptoms are very vague and could easily be missed. These include loss of appetite, tiredness, or pain in the bones.

More specific symptoms depend on where the neuroblastoma started:

-  **Abdominal:** If the tumour started in the abdomen, the child may have a swollen tummy and complain of difficulty in passing urine or a stool
-  **Neck:** A tumour in the neck can often be seen as a lump; it may affect breathing and swallowing
-  **Chest:** Tumours in the chest area may make it difficult for the child to breath or swallow; they may also have a persistent cough
-  **Spinal Cord:** If the tumour is near and pressing on the spinal cord, the child may experience a weakness in the legs and have difficulty walking. If the child is not yet walking,

neuroblastoma can present as reduced or weakened leg movements. Difficulty in passing urine and constipation may also occur

- 🦿 **Skin:** Deposits of neuroblastoma sometimes appear as small blue-coloured lumps
- 🦿 **Blood Pressure:** The child may have high blood pressure
- 🦿 **Eye & Muscle Movements:** Jerky eye or muscle movements and general unsteadiness may occur, but this is rare

Other symptoms can include easy bruising or bleeding; tiredness; swelling of the ankles, legs, feet, or scrotum; severe watery diarrhoea; fever; bone pain; dark circles around the eyes ("black eyes"); a droopy eyelid; backache; loss of appetite, nausea, or weight loss; irritability, or listlessness

If your child presents with any of the above symptoms, please contact a doctor as soon as possible.

Tests and Diagnosis

Neuroblastoma virtually always produces certain chemicals that are passed in the urine. These chemicals are called vanillylmandelic acid (VMA) or homovanillic acid (HVA) and are present in high levels in 99% of children with neuroblastoma. As these chemicals are produced by the tumour cells, they are known as tumour markers and can be used to monitor whether the tumour is growing or shrinking during treatment.

If neuroblastoma is suspected, a variety of other tests will be done to confirm the diagnosis and at what stage the tumour is.

Neuroblastoma can be diagnosed or staged using the following tests and procedures:

- 🦿 **VMA and HVA tests:** These are specific urine tests to check the VMA and HVA levels in the urine
- 🦿 **MIBG Scans:** MIBG (metaiodobenzyl guanidine) is a substance that is given by injection and is taken up by neuroblastoma cells. A small amount of radioactive iodine is attached to the MIBG, which enables the tumours to be seen by a radiation scanner
- 🦿 **A Biopsy:** An operation done under general anaesthetic; a small sample of cells is taken from the tumour and studied under a microscope by a pathologist. Various laboratory tests, collectively referred to as tumour biology, are done to study the chromosomes and 'biological markers' in the tumour cells. One of these 'markers' is called MYCN and a certain amount of these in the cells can suggest that the tumour is an aggressive one, which indicates a more aggressive treatment regimen.

Other tests that may be done include x-rays, blood and bone marrow tests, and CT or MRI scans,

Treatment Options

Treatment options depend on the age of the child, the position and size of the tumour, the tumour biology (MYCN status), and whether the tumour has spread.

Treatment options include:

- 🎗️ **Surgery:** The tumour can often be removed by surgery if it is localised and has not spread in to the lymph nodes or other parts of the body. If the tumour has spread or is classed as high risk, further treatment with chemotherapy and of radiotherapy may be required.
- 🎗️ **Chemotherapy:** The use of a combination of anti-cancer drugs to destroy or shrink cancer cells is the preferred treatment for children, and may be done prior to and after surgery
- 🎗️ **High-dose Chemotherapy with Stem Cell Support:** If the neuroblastoma is high-risk with MYCN amplification or has spread, high doses of chemotherapy may be used to wipe out any remaining neuroblastoma cells. Unfortunately this also wipes out the body's bone marrow. To combat this, stem cells are collected from your child via a drip before the chemotherapy is given, and frozen and stored. Once the chemo treatments are done, the stem cells are returned to your child's body via a drip; they make their way back to the bone marrow and grow and develop into mature blood cells within 14-21 days.
- 🎗️ **Radiotherapy:** High-energy rays that destroy the cancer cells but do minimal harm to normal cells.

Some of the treatment options may result in after-effects such as nausea, vomiting, irritation or soreness of the skin from radiation, hair loss, risk of infection, fatigue, bruising and bleeding or diarrhoea.

Awareness Ribbon Colour

The awareness ribbon colour for Neuroblastoma is Light Purple

Article researched and written on behalf of [Little Fighters Cancer Trust](#) by Billi du Preez of [Red Feather Scribes](#)

Academic Editing by Dr Marc Hendricks MBChB (UCT), DIP PEC (SA), DCH (SA), FCPaeds (SA), CMO Paeds (SA) Senior Specialist, Paediatric Oncologist Red Cross War Memorial Children's Hospital

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