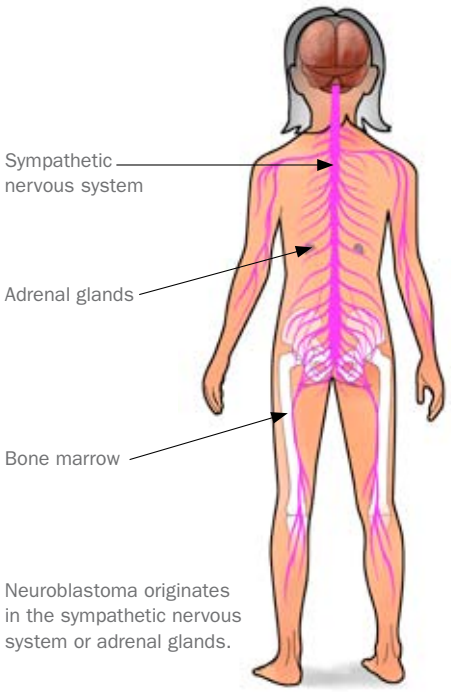
FAKTA OM

NEUROBLASTOMA



Neuroblastoma originates in the sympathetic nervous system or adrenal glands.

Neuroblastoma is mainly a disease of very young children and practically non-existent in adults. Most of those who develop the disease do so before the age of two. It is the most common form of cancer in children behind leukaemia and brain tumours. Around 20 children in Sweden are affected each year.

BACKGROUND AND CAUSE

Neuroblastoma is a type of tumour that affects very young children. The average age at diagnosis is about two years, but babies can be born with the disease. Neuroblastoma occurs in and during the development of the sympathetic nervous system, which is a branch of the autonomic nervous system – the parts of the nervous system that cannot be controlled by will, such as digestion and blood pressure. The sympathetic nervous system runs down the spinal column and branches out in the torso to the adrenal glands and elsewhere. As it is spread across large parts of the body, neuroblastoma can occur in any of these places, but it is most common in the adrenal glands.

It is believed that neuroblastoma originates in the stem cells that go on to form the sympathetic nervous system. The reason why children are affected by neuroblastoma is unknown. The risk of the disease is the same across much of the world, which suggests that the tumour cannot be linked to the parents' lifestyle (diet etc) or an underlying viral infection. One possibility is that the tumour results from entirely random changes in the stem cells' DNA.

SYMPTOMS AND DIAGNOSIS

Children with neuroblastoma often have no symptoms, and the disease is discovered when their parents or doctors feel a lump. The tumour can sometimes release hormones that can cause diarrhoea, sweats and other symptoms. When the tumour grows or spreads, it may put pressure on other organs, resulting in symptoms.

It is normally easy to make a diagnosis by taking a sample straight from the tumour. Additional tests may include urine samples, X-rays, ultrasound, MRI, bone marrow samples and scintigraphy.

TREATMENT

With the help of known risk factors, it is possible to classify children with neuroblastoma into those with benign, moderate or aggressive disease. This classification decides how the child will be treated. Those with a benign tumour will undergo surgery and then be kept under observation. One special form of neuroblastoma can disappear of its own accord without treatment, while children with more advanced disease need to be treated with chemotherapy, surgery, radiotherapy and special high-dose chemotherapy with stem cell support and vitamin A. All Swedish children with neuroblastoma are treated on the basis of a European protocol which is being improved all the time as our understanding of the disease grows.

RESEARCH

Extensive research has focused on identifying which genes change in neuroblastoma. Most important from a clinical point of view is the

almost 30-year-old discovery that the MYCN gene is often amplified (multiple copies). This is seen in around 20-30% of children with neuroblastoma and is very closely linked to an aggressive form of the disease with an increased risk of recurrence. Other factors used to classify the risk are the child's age, the stage and histopathological maturity of the tumour, and other genetic changes.

Intensive research into neuroblastoma is under way, both basic and clinical. At a basic level, scientists are trying to discover what happens when cells in the sympathetic nervous system cause tumours, and which genetic changes lead to this. This is going hand-in-hand with clinical research where new forms of treatment are being tested, often based on discoveries made in the basic research. The Swedish Child Cancer Foundation is supporting a number of researchers in Sweden working on increasing our understanding of the disease in order to find better treatment options, especially for children where the disease has already spread at the time of diagnosis.