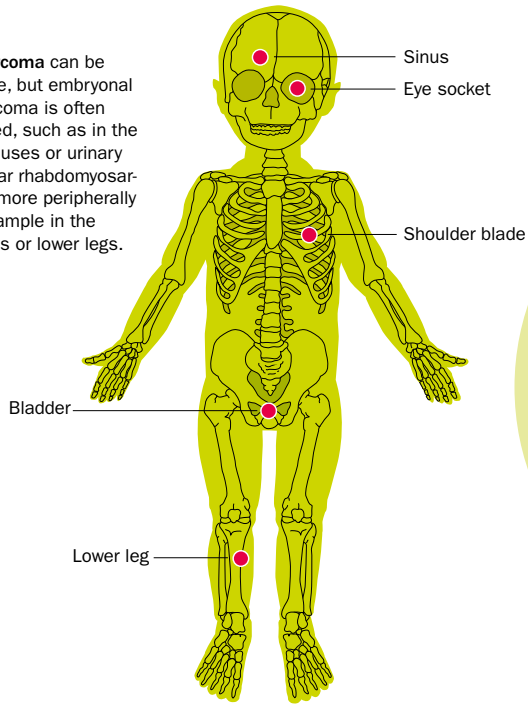
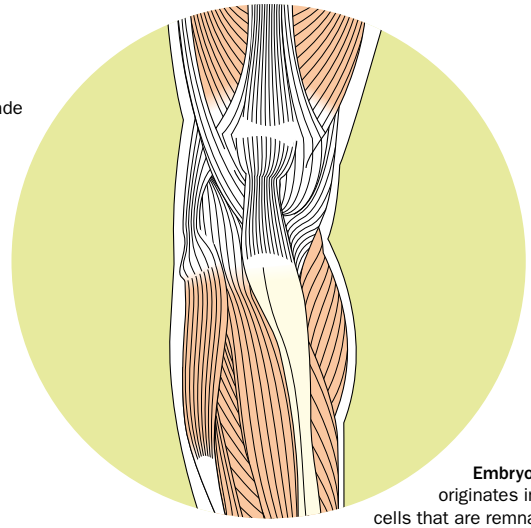
RHABDOMYOSARCOMA

Rhabdomyosarcoma originates in muscles and is the most common type of sarcoma in children. About six to ten children in Sweden are diagnosed with it every year.

Rhabdomyosarcoma can be found anywhere, but embryonal rhabdomyosarcoma is often centrally located, such as in the eye socket, sinuses or urinary bladder. Alveolar rhabdomyosarcoma is often more peripherally located, for example in the shoulder blades or lower legs.





Embryonal rhabdomyosarcoma originates in very immature muscle cells that are remnants of the foetal stage. Alveolar rhabdomyosarcoma originates from slightly more mature muscle cells.

RHABDOMYOSARCOMA

In principle rhabdomyosarcoma may occur anywhere in the body where muscles are being formed – especially skeletal muscles. About 60% of all rhabdomyosarcoma cases originate in the head and neck region. The most common location, found in about 10% of cases, is the eye socket. Another common location is around the urinary bladder.

SYMPTOMS

Symptoms of rhabdomyosarcoma can be highly varied. A nodule that grows or fails to disappear can affect nerves or various organs. For example, it can be difficult to urinate.

The tumour itself does not hurt, but the surrounding tissues may be painful. A tumour in the eye socket may cause the eye to bulge, while a tumour in the ear may become visible in the auditory canal.

Bleeding from any body orifice may be the first sign of rhabdomyosarcoma.

DIAGNOSIS

To make a diagnosis, doctors take a cell sample or tissue sample from the tumour. The area surrounding the suspected tumour is examined by x-ray.

The tumour most commonly spreads to the lungs, which are also x-rayed. Doctors also take x-rays of the bones and a bone marrow sample to see whether cancer cells have spread to the skeleton.

If the tumour is located near the brain or spinal cord, a spinal fluid sample is also taken. Many sarcomas can be precisely identified through analysis of their genetic deviations.

MAIN TYPES

Rhabdomyosarcoma is divided into two main types, embryonal and alveolar. The type plays a major role in treatment and prognosis.

Embryonal rhabdomyosarcoma originates in very immature muscle cells that are remnants of the foetal stage. They are often centrally located in the body, for example in the eye socket, sinuses or urinary bladder.

This type of rhabdomyosarcoma usually affects young children.

Alveolar rhabdomyosarcoma originates from slightly more mature muscle cells and develops more peripherally in the body, such as the shoulder blades or lower legs.

Alveolar sarcoma, which is more

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common among older children and teenagers, is more aggressive and resistant to treatment.

TREATMENT

Children diagnosed with rhabdomyosarcoma almost always undergo a combination of chemotherapy, surgery and/or radiotherapy.

Most children with rhabdomyosarcoma are first given chemotherapy to shrink the tumour.

Alveolar rhabdomyosarcoma usually requires radiotherapy, regardless of how successful surgery has been. Afterwards, the child usually again undergoes several courses of chemotherapy. The treatment period is usually six months, but in certain cases may be extended for an additional six months of maintenance therapy.

PROGNOSIS

Survival, regardless of the type of rhabdomyosarcoma, has radically improved in recent years. However, the type of cell from which the tumour originates is of great importance, as is the location of the tumour. Currently, between 70 and 90% of children with rhabdomyosarcoma recover from the disease. However, between 20 and 30% suffer a recurrence.

Alveolar rhabdomyosarcoma is generally more difficult to treat than the embryonal type, as are tumours in children over the age of ten. The same applies to tumours that are larger than five centimetres in diameter and to tumours that have already spread at the time of diagnosis, which is the case in one of five children.

The location of the tumour is also highly important when doctors determine how much local treatment can be given. This applies to surgery where damage to healthy tissues and organs must be avoided, and to radiotherapy where extensive late complications need to be considered.

For example, treating tumours near the urinary bladder or genitals poses a risk of affecting bladder function, the child's future sex life and the ability to have children.

Fact-checked by Gustaf Ljungman, Associate Professor and Senior Lecturer at the University Children's Hospital in Uppsala, September 2017.