



kinderkrebsinfo

Informationsportal zu Krebserkrankungen bei Kindern und Jugendlichen

Tumours of the salivary glands

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Tumours of the salivary glands

Both benign and malignant tumours can develop in the salivary glands. This, however, happens rarely both in children as well as in adults. In Germany, approximately ten children or teenagers are diagnosed with such a tumour per year.

Hence, there are no therapy studies providing the optimal treatment strategy that has been proven for many patients, as there are for other tumours in childhood and adolescence. However, the experts of the Registry for Rare Diseases in Paediatrics (German abbreviation: STEP, for “Seltene Tumorerkrankung in der Pädiatrie”) have summarized their experiences as well as the experiences of international research groups on the following pages.

Since every child is different, it is important to individually adjust the treatment to every single patient. Therefore, “STEP” offers free advice via a tumourboard attended by multidisciplinary experts. Feel free to have your attending physician contact the experts at “STEP” at step@klinikumdo.de. Also, by registering your child with the „STEP“ registry, you get the option to help expanding the experience with these rare tumours, thereby helping other children who are diagnosed with such a disease in the future.

1. Introduction to the salivary glands

The salivary glands are located in the area of the lower jaw. They are responsible for producing saliva. Saliva moistures our intaken food while chewing, thereby making it smooth. Saliva contains certain enzymes (so-called amylases) which predigest – to a limited degree – carbohydrate-rich food. Saliva also contains an antibacterial substance that promotes the inactivation of pathogens.

There are three pairs of large salivary glands:

The largest are located in front of the parotid glands (*Glandulae parotidaeae*). The facial nerve (*Nervus facialis*), which innervates the facial muscles and controls facial expression, goes through them.

In addition, there are the two submandibular (*Glandulae submandibularis*) and the sublingual salivary glands (*Glandulae sublingualis*).

2. Disease: what is a tumour of the salivary glands?

Salivary gland tumours arise from salivary gland tissue. Most of the tumours develop in the parotid glands, however, the other salivary glands can also be affected.

There are a few different *tumours* in this area. Benign and malignant tumours are equally common.

The most common benign salivary gland tumour is pleomorphic adenoma. The most frequent malignant tumour in children and adolescents is the so-called *mucoepidermoid carcinoma*, followed



by *acinar cell carcinoma*. These tumours behave less aggressively than other *carcinomas* mostly occurring in adults. This means that they grow more slowly and spread less frequently into the *lymph nodes* or other organs (*metastasis*).

3. Cause: how does a tumour of the salivary glands develop?

The exact cause of developing a salivary gland tumour is unknown. Most tumours occur sporadically. That means that there are no familial *genetic* risks. Different from adults, there are also no known risk factors for most children.

It is known, though, that the risk for these tumours is increased after *radiotherapy* of another cancer in this area. Occasionally, salivary gland tumours have been observed after *chemotherapy* for *leukaemia* treatment.

4. Symptoms: what are the signs of the disease?

Patient with a salivary gland tumour may not have any complaints for a while. First indication of the disease is often a visible and palpable, usually indolent, swelling. Some children may have impaired facial expression, such as a wry smile or slurred speech, which indicate damage of the facial nerve.

The following signs of the disease (*symptoms*) may occur, particularly with advanced stage:

- swelling
- decreased salivation, dry mouth
- pain, facial sensory deficits
- unilateral paralysis of facial muscles

5. Diagnosis: what kinds of tests are required?

Usually, the paediatrician will first take a thorough medical history (*anamnesis*) and perform a *physical examination*. There are no laboratory results that are specifically indicative of a salivary gland tumour. When palpation and/or symptoms are suspicious of a tumour, *imaging* diagnostics will help find out whether this is the case.

Ultrasound (sonography), *magnetic resonance tomography* (MRT, layered scanning of the body using magnetic fields and radio waves) and/or *computed tomography* (CT, detailed x-ray slices of the body) help to obtain information on the tumour's location, its size, extent and growth pattern. When dealing with children and adolescents, not using techniques based on *X-rays* is the goal, therefore, *ultrasound* and magnetic resonance tomography are the standards.

Obtaining a tissue sample is not mandatory, since surgical tumour removal is scheduled at the beginning of treatment.



Psychosocial Care

A child's cancer is a stressful situation for the whole family. The psychosocial team of the clinic or later the aftercare facility provides advice and support to patients and their relatives from diagnosis to completion of treatment as well as during aftercare. Don't hesitate to take advantage of this offer. It is an integral part of the treatment concept of all paediatric oncology centres in many countries. Here you will find comprehensive information on this.

6. Therapy: how is treatment done?

Treatment of children and adolescents with a tumour of the salivary glands should take place in a children's hospital with a paediatric oncology program, in cooperation with an ear, nose and neck (ENT) clinic. Only such a childhood cancer centre provides highly experienced and qualified staff (doctors, nurses and many more), since they are specialised and focused on the *diagnostics* and treatment of children and teenagers with cancer according to the most advanced treatment concepts.

Therapy mainly consists of surgery aiming at tumour removal, sometimes followed by radiotherapy. Chemotherapy is applied very rarely.

6.1. Surgery

The major therapeutic step is gross total tumour removal. Since remaining tumour tissue is frequently followed by recurrent disease (relapse), tumours are being removed with a safety margin. That means that not only the tumour itself is being resected, but also an appropriate amount of adjacent healthy tissue as well. Taking out the entire salivary gland or its affected lobe in total is considered safest.

However, if the tumour is located in the parotid gland, the facial nerve needs to be spared so that *surgery* does not cause subsequent facial paralysis. This procedure may be complicated, particularly when dealing with larger tumours. Therefore, tumour resection should always be performed by an experienced ENT surgeon who also has experience with treatment of these tumours. For its protection, the facial nerve is permanently monitored during surgery (*neuromonitoring*).

In case of lymph node involvement, surgery needs to be extended to the affected *lymph nodes* and adjacent tissue. This procedure is known as neck dissection.

6.2. Additional treatment options

Benign tumours (adenomas) do not require additional treatment following surgery. For most malignant salivary gland tumours in childhood and adolescence, therapy is ceased after gross total resection, too. If complete removal is not feasible or in case of *metastasis*, additional therapy needs to be considered. In general, chemo- or radiotherapy are the options.

For localised tumours, radiotherapy is more efficient. However, the risk of *radiotherapy*-associated long-term sequelae require consideration. According to the currently available data, children and



adolescents more frequently develop complications after radiotherapy than adults (for example impaired facial growth, secondary malignancies). Therefore, radiotherapy should be only applied for selected patients and after careful consideration, for example when the tumour could not be completely removed or in case of an aggressive tumour type.

Only in extremely rare scenarios, *chemotherapy* is required in children and adolescents. It is usually combined with radiotherapy. Since this is required in rare scenarios only, advice should be given by the inter-disciplinary tumour board for rare diseases.

Note: please confer in advance, feel free to contact the STEP Registry step@klinikumdo.de.

7. Prognosis: what are the chances of cure?

Children and adolescents with a tumour of the salivary glands usually have a very good *prognosis*. Most patients in this age group become completely healthy. The outcome is mostly impacted by the tumour type and the success of surgery. In general, gross total tumour resection is associated with a more favourable prognosis than partial removal.

It is good to know that benign tumours can also relapse after incomplete tumour resection. In this case, a benign tumour may occur again at the same location. However, malignant tumours, thus salivary gland carcinomas, in childhood and adolescence are usually rather minimally malignant (low-grade). This means that they metastasize very rarely and don't grow aggressively locally.

The STEP Registry has analysed the data of all registered patients. About 10 % of children and adolescents developed a local relapse in the salivary gland or in the adjacent tissue, which was subsequently treated surgically again. Only extremely rare, very aggressive variants, such as the NUT midline carcinoma, take a malignant course. Hence, this tumour type requires the choice of a very intensive treatment from start.

Patients with parotid gland tumours can suffer injury of the facial nerve (*Nervus facialis*) and subsequent facial paralysis as a treatment complication after surgery. This happens in about 8 % of cases; the risk can be reduced by means of intraoperative neuromonitoring (IOM).

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Glossary

acinar cell carcinoma	malignant tumour resulting from certain glandular cells (acinar cells) of the salivary glands; acinar cells are found in the glandular ducts of various organs, especially in the salivary glands of the lower jaw, but also in the pancreas, liver and lungs.
anamnesis	medical interview, a patient's history, development of signs of illness; the type, onset and course of the (current) symptoms as well as any risk factors (e.g. hereditary diseases) are evaluated during a medical interview.
carcinoma	malignant tumour resulting from degenerated epithelial tissue (e.g. skin, mucous membranes, glandular tissue);
chemotherapy	here: use of drugs (chemotherapeutic agents, cytostatics) for the specific inhibition of tumor cells in the organism
computed tomography	imaging, X-ray diagnostic procedure; it produces an image by computer-controlled evaluation of a large number of X-rays from different directions. This makes it possible to produce sliced images of body parts (tomograms, transverse or longitudinal sections of the human body)
diagnostics	methods / measures for the detection of a disease
genetic	concerning the (level of) inheritance or genes; inherited
imaging	diagnostic procedures generating images of the inside of the body, such as ultrasound and X-ray examination, computed tomography, magnetic resonance imaging, and scintigraphy
leukaemia	malignant disease of the blood forming (haematopoietic) system and the most common cancer in children and adolescents (about 33%); depending on the origin of the malignant cells, a distinction is made between lymphoblastic and myeloid leukaemias. Depending on the course of the disease (fast or slow), a distinction is made between acute and chronic leukaemias.
lymph nodes	small lenticular to bean-shaped organs that are part of the body's immune system and are located in many parts of the body; they serve as filter stations for the tissue water (lymph) of a region of the body and contain cells of the immune system.
metastasis	1. tumour spread from the primary site of tumour to other parts of the body; characteristic feature of malignant tumours (cancer). 2. collective term for a disease process characterized by malignant



	cells spreading from their primary site to other areas of the body via the bloodstream and/or the lymphatic system.
mucoepidermoid carcinoma	rare malignant tumour of the salivary glands
physical examination	an important part of diagnostic examinations; includes palpation and listening to certain body organs as well as testing reflexes to obtain indications of the nature or course of a disease.
prognosis	prediction of the course and outcome of a disease / prospect of recovery
radiotherapy	controlled use of ionizing (high-energy) radiation for the treatment of malignant diseases
surgery	surgical intervention on or in the body of a patient for the purpose of treatment, less often also in the context of diagnostics; the surgical intervention is carried out with the help of special instruments, generally with the patient under anesthesia.
symptom	sign of illness
tumour	groups of abnormal cells forming a growing lump, both benign and malignant
ultrasound	an imaging technique used to examine organs, in which ultrasound waves are sent through the skin into the body; at tissue and organ boundaries, the sound waves are reflected back, picked up by a receiver (transducer) and converted into corresponding images with the help of a computer.
X-rays	high-energy electromagnetic radiation, discovered by W. C. Röntgen in 1895; X-rays can partially penetrate matter, so that insights into the interior of the human body are possible, among other things. Since X-rays have an ionising effect (ionising rays), they can also change matter, e.g. damage cells and possibly cause cancer. In diagnostics, X-rays are used to examine certain parts of the body. Depending on the type of irradiated tissue, the radiation is intercepted (absorbed) to varying degrees and displayed on a film plate as a two-dimensional image. Since every X-ray examination is associated with a certain amount of radiation, particularly sensitive parts of the body (such as gonads) must be protected. In the context of X-ray therapy (e.g. radiotherapy), very high-energy X-rays are used to kill tumour cells.