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Soft tissue tumours of the extremities in childhood - conservative of surgical procedures: Results

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All malignant tumours in children are treated in Germany in a standardized pattern. For each kind of a tumour there are study groups analogues to the organizations in whole world. Their findings will be continously transferred into latest therapeutical conceptions. The soft tissue sarcomas of the extremities are considered according to two studies, 81 and 86. The results had been worked out by Treuner at the university of Tübingen and are interpreted by Schweizer recently.

Insights into the biological behaviour of the soft tissue sarcomas, draw at the conclusion to consider the successes of therapy in dependance on sensibility for cytostatics as well as the primary operative, non-mutilating resectability. The variety of the soft tissue sarcomas are shown in slides I and II! Both-in the 81 study and in the study from Altmann and Schwartz from the USA the enormous significance of the rhabdomyosarcoma is conspicuous. From the comparison between the studies 81 and 86 follows that the number of the soft tissue sarcomas altogether has increased or that the recording has been better managed.

This preliminary report is based on a material of 460 children in 15 years in Germany collecting the cases of more than 40 clinics and departments.

In case of a very small sensibility against chemotherapeutics a primary part devolves upon the paediatric surgeon, independent of the local extension and the localisation of the growth.

Subsequently the surgeon has then to resect at several patients also once at the price of anatomic and functional mutilation.

In case of a high sensibility and a good response against cytostatics the operative part of the paediatric surgeon recedes into the background if the soft tissue sarcoma cannot be primarily resected under preservation or a good function. The primary surgical procedure must be in accordance with an oncologic-radical removal of the growth in free, not involved area without an endangering of vital essential structures and without any mutilations.

A primary amputation of extremities-in the past unfortunately much too often performed-disappeared totally from the conception of treatment.

1. A tumour involved muscle should always be resected in toto from origin to insertion. A group of muscles may require a removal of more than one muscle, but the loss of functions should be limited.

2. Of course, primary surgical therapy runs parallel to the sensitivity of tumour tissue to cytostatic drugs and/or to radiation. That does not mean to have to decide in every case to these characteristics. The staging and localisation at the time of establishing a diagnosis is of extreme importance.

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The following criterions are to be considered:

a) Influence of age to the prognosis; with increased age the prognosis becomes worse.
b) The localisation has decisive consequences in prognosis: a rhabdomyosarcoma of the extremity has an essential worse prognosis as for instance rhabdomyosarcomas of the urogenital tract.
c) Rhabdomyosarcomas of the alveolar type have, in a collection of all stages, a five-year-survival-rate of 45 %. Tumours of embryonal genesis possess a better prognosis-about 63 %. Otherwise rhabdomyosarcomas of extremities unfortunately very often belongs to the alveolar type. Therefore their prognosis is worse.
d) A tumour of small extension should and can always primarily be removed by operative way, also if it shows sensitivity against cytostatics. The more sensitive a tumour is against cytostatics, the less radically the surgical intervention must ensue. For that reason it is not so important in case of a good response to cytostatics, if microscopic visibly remnants remain in margin of resection or the removal of the growth is macroscopically not totally possible.

It is decisive to know the sensitivity to chemotherapy of each tumour! Under those with a high sensibility we find also the rhabdomyosarcoma, under those with a low sensitivity for instance the hemagio-sarcoma and leio-myosarcoma.

Almost to sensitivitiy is showing the fibrosarcoma and the neurofibrosarcoma.

But as nobody knows about the dignity of a tumour before the treatment, the paediatric surgeon has the following assignments in an oncological team:

He has to perform the biopsy to satisfy the diagnosis histologically, immunhistochemically, electronmicroscopically and in the future cytogenetically.

He has to determin the resectability and account for it. So, after all the paediatric surgeon must ask the following questions:

Firstly

Are the chances for a complete healing given away by a primary chemotherapy?

Secondly

Is it correct that by chemotherapy in stage III on a long-term basis equal or even better results can be obtained than by primary surgical procedure, even if this leadsto a mutilation?

Thirdly

Are mutilations avoidable after a primary chemotherapy ultimately and also in each case an anatomical and functonal mutilation, if a later operation however in the further course is getting necessary?

For the most common soft tissue tumour, the rhabdomyosarcoma the following results could be obtained:

Tumours stage I, primarily resectable, show a five-year-survival rate of 85 %. Tumours of stage III primarily non resectable and therefore cytostatically treated, have an even free survival of 59 %. Tumours of stage II reach an EFS of 63 %. Otherwise tumours of stage IV attend after five years only the small EFS of 11 %. Can tumours of stage III treated by an optimal cytostatic therapy be transformed in a stage I or at least in a stage II and might it be possible in this mode the five-year-survival-rate to approach to the 85 % line?

From our results of the 81 study it is evident that some patients with stage III initally and cytostatic treatment were transferred to stage I. In that way they achieved a five-years-survival-rate of 84 % and became parallel to the successes in the group of initial stage I-patients. In the meanwhile as a sensitive feature for this transformation the response grade after a seven weeks cytostatic therapy has been found out.

In case of weaker degree of response indeed the frequency of recurrence and the relapse of metastases increase obviously.

There are also children with a rhabdomyosarcoma of stage III who are under treatment with cytostatics reach a complete return of the tumour until the 7th week and thus a five-year-survival-rate of 95 %: At the moment of a second-look operation no remnance of growth are recocnizable. The duration of treatment also plays an important part. If the involution of the tumour is attained only after 16 weeks the five-year-survival-rate already declines to 70 %.

The 86 study shows that a tumour with partial return already after the 7th week, treated cytostatically has to be brought to a second-look-operation. Otherwise, if a tumour decreases a little, the radiation is required. In case of failing of a retrogression early operative resection is recommended, also at the price of mutilation.

One important question we are not able to reply with certainty:

Would the prognosis in children reduced by chemotherapy from stage III to stage I even be more agreeable of one had done an operative procedure before the chemotherapy starts thus primarily also in acceptance of a mutilation.

The peadiatric surgeon takes over three assignments within an oncological team:

Firstly

The biopsy they do not finding out the detailed diagnosis, the judgement of the resectability, the operative resection if a mutilation is avoidable.

Secondly

The decision in primarily non resectable tumours, if they do not reach after seven weeks a complete retrogression, if then is an operation justified, also at the price of partial mutilation.

Thirdly

The biopsy must be repeated 16 weeks after beginning of the therapy in children, who underwent a chemotherapeutic and radiological treatment.

Fourthly and last

In some cases a complication occurs through chemotherapy or radiotherapy which has to be removed in operative way.