

БЪЛГАРСКО
НАУЧНО ДРУЖЕСТВО
ПО ДЕТСКА ХИРУРГИЯ



III НАЦИОНАЛЕН КОНГРЕС ПО ДЕТСКА ХИРУРГИЯ

С МЕЖДУНАРОДНО УЧАСТИЕ

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Our 20 years experience in the treatment of hepatoblastoma

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Introduction

Hepatoblastoma – rare tumor – below 1% of all malignancies in childhood.

More often <18 months of age.

Can be associated with Beckwith-Wiedeman syndrome and hemihypertrophy.

The right hepatic lobe is more commonly affected.

The most common metastases – in lungs, brain, regional lymph nodes etc.

Tumor markers – α FP, β HCG.

Material and methods

For the period of 20 years (1991-2011) - 14 children.

Sex: males : females -11 : 3.

Age: under the age of 2 years – 12 patients, older than 10 years – 2 males.

Localization: right hepatic lobe – 7 patients, left – in 3 patients, multifocal – in 4 patients.

Histology: fetal type – in 6 patients, embrional – in 5 and anaplastic – in 3 children.

Clinical stage: stage II – 7 patients, stage III – 7.

Presenting features

The initial demonstration of the disease in most cases – an abdominal mass, very often – asymptomatic.

10 children (71,4%) - in good healthy condition, in 4 boys – anorexia, weight loss and anemia, in 1 child – hepatomegaly.

In one boy 1,5 year after radical surgery isosexual precocity with penile and testicular enlargement with insufficient presentation of pubic hair predicted the lung and later the brain metastases.

Laboratory data

Anemia below 6g/L - in one child.

Thrombocytosis over 800 000 – in 7 children, up to 700 000 - in 6 children and below 450 000 – in one child.

Tumor markers

In all 14 children α FP was elevated at diagnosis.

In 3 children with local relapses and in 2 children with lung metastases during the course of the disease increased levels of β HCG was registered.

Treatment

Surgery: All 14 children were surgically treated and diagnosis was confirmed histomorphologically. Eleven children received neoadjuvant chemotherapy (3-7 cycles with Cis-platin, Farnorubicin and Carboplatin). Five children underwent second surgery for relapses and 2 boys underwent surgical treatment for lung metastases.

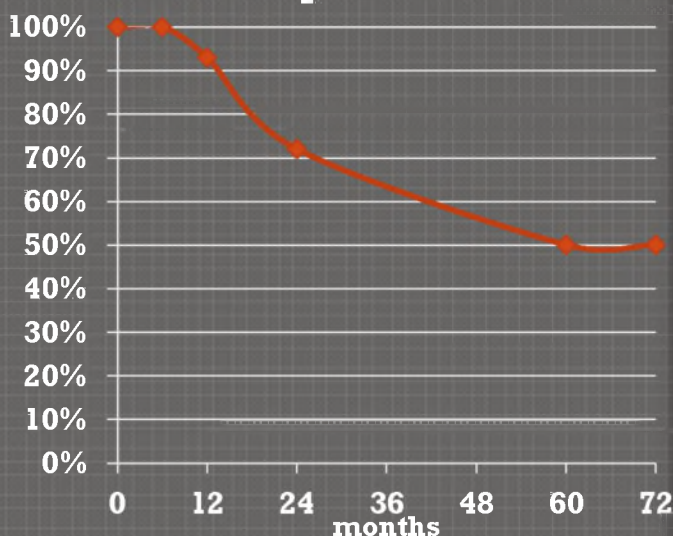
Chemotherapy: according the SIOPEL 2 protocol.

Conclusion

The prognosis of the disease depends on clinical stage, histological type, localization as well as possibilities for radical surgery and techniques for liver resection.

Our experience for 20 years shows 50% survival in 14 complex treated children with hepatoblastoma, considered in the past as with hopeless prognosis.

Survival of 14 patients with hepatoblastoma



Prognostic factors

| Clinical stage | Alive | % | Dead | % | Total |
|----------------|-------|----|------|----|-------|
| II | 6 | 86 | 1 | 14 | 7 |
| III | 1 | 14 | 6 | 86 | 7 |

| Histological type | Alive | % | Dead | % | Total |
|-------------------|-------|-----|------|-----|-------|
| Fetal | 6 | 100 | 0 | 0 | 6 |
| Embrional | 1 | 20 | 4 | 80 | 5 |
| Anaplastic | 0 | 0 | 3 | 100 | 3 |

| Localization | Alive | % | Dead | % | Total |
|--------------|-------|-----|------|-----|-------|
| Left lobe | 2 | 67 | 1 | 33 | 3 |
| Right lobe | 5 | 70 | 2 | 30 | 7 |
| Multifocal | 0 | 100 | 4 | 100 | 4 |

| Sex | Alive | % | Dead | % | Total |
|--------|-------|-----|------|----|-------|
| Female | 3 | 100 | 0 | 0 | 3 |
| Male | 4 | 36 | 7 | 64 | 11 |