RENAL TUMOURS

ABSTRACT NO.: P-093

Results of treatment of children with bilateral nephroblastoma

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Key words: treatment, results, nephroblastoma, children
Introduction. Wilms tumor is the most common renal cancer in children. Approximately 5 % of children with Wilms tumor present with disease in both kidneys. We retrospectively reviewed our institutional experience of treatment patients with bilateral Wilms tumor (BWT) from 1980 till 2013.
Aim. To improve the results of treatment of children with bilateral nephroblastoma
Materials and methods. During the period from 1980 to 2013 at the Institute of Oncology and Hematology were examined and treated 82 children with bilateral nephroblastoma. In all patients, the diagnosis was confirmed morphologically. The main peak of incidence of bilateral nephroblastoma accounts for the period from age 3 to 5 years – 47 (57.3 %). All children received neoadjuvant chemotherapy vincristine, dactinomycin, doxorubicin. Surgical treatment was 78 of 82 children. The other four children have not received surgical treatment due to progression of the disease on the background of the treatment. Prior to surgery all children performed radioisotope study of kidneys and urine test for endogenous creatinine clearance. 59 children (75.6 %), surgical treatment was carried out in two stages, first at the least affected kidney tumor, then – on the contralateral organ. 19 patients (23.9 %), surgery was performed in one step. Postoperative chemotherapy and radiotherapy (if necessary) is carried out, depending on the stage of disease and histological variants f the tumor, according to the accepted protocol in the clinic. Since 2010, children with bilateral nephroblastoma, receive treatment protocol International Society of Pediatric Oncology (SIOP) 2001.
Results. 7 patients were re-operated because of local recurrence. Four of them were anti-relapse chemotherapy after repeated operation and are in dynamic observation over 2 years, 3 patients died of disease progression. 2-year OS rate – 84.7 %, 5-year OS rate – 77 %, 2-year EFS – 79 %.
Conclusion. The correct diagnostic and modern strategy therapy can improve overall survival in children with bilateral nephroblastoma.

ABSTRACT NO.: OP-097

Renal cell carcinoma in children: report of two cases

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Key words: renal tumors, renal cell carcinoma, children
Introduction. Renal cell carcinoma is a rare tumor in children. This disease constitutes 0.1–0.3 % of all tumors and 1.8–6.3 % of renal neoplasm in childhood and adolescent younger than 15 years.
Aim. To show two cases of the successful treatment of such a rare tumor in pediatric population as a renal cell carcinoma.
Materials and methods. From January 2012 to January 2016 77 patients with renal neoplasm were operated on. On histology the majority of the patients had nephroblastoma –

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57 (74.0 %); also we observed mesoblastic nephroma – 6 (7.8 %), rhabdoid tumor – 5 (6.5 %), clear-cell sarcoma – 4 (5.2 %), carcinoma – 2 (2.6 %), nephroblastomatosis – 1 (1.3 %), cystic nephroma – 1 (1.3 %) and neuroepithelial tumor – 1 (1.3 %).

Case 1. A 6 y.o. girl. On admission complaints of an abdominal pain and a painful urination. Abdominal US and CT scan showed a tumor of the lower pole of the right kidney measuring 41 × 37 × 49 mm (the size of the right kidney was 84 × 30 mm). Chest CT scan showed no metastasis. No pre-existing condition and no inherited syndrome were found. A 4 weeks of neoadjuvant chemotherapy according to SIOP 2001 protocol for localized stage I of a disease (ACT-D, VCR) were administered. CT scan after chemo showed the same size and structure of a tumor. The patient was operated on. The surgical procedure included laparotomy, modified radical nephrectomy with preservation of the adrenal gland, biopsy of lymph nodes of the ipsilateral and contralateral renal hilum and paracaval lymph nodes. Histology was associated with translocation Xp11.2/transformation of gene *TFE3* (TFE3 antibody positive reaction on immunohistochemistry, no cytogenetic examination was performed), grade 3, stage I. No postoperative treatment was performed. At the present time the patient is under observation.

Case 2. A 10 y.o. boy. On admission complaints of a high blood pressure controlled by p.o. intake of enalapril. Abdominal US and CT scan showed a tumor of the upper pole of the right kidney measuring 38 × 35 × 37 mm (the size of the right kidney was 89 × 36 mm). Chest CT scan showed not numerous subpleural 2–3 mm foci (metastasis?). Bone scintigraphy showed no metastasis. No pre-existing condition and no inherited syndrome were found. A 6 weeks of neoadjuvant chemotherapy according to SIOP 2001 protocol for unilateral lesion and distant metastasis (ACT-D, VCR, DOX) were administered. CT scan after chemo showed the same size and structure of a tumor and of lung foci. The patient was operated on. The surgical procedure included laparoscopy, modified radical nephrectomy with preservation of the adrenal gland, biopsy of lymph nodes of the ipsilateral renal hilum and aortocaval lymph nodes. On histology it was clear-cell renal cell carcinoma, grade 1, stage I. No postoperative treatment was performed. The lung foci were considered as not metastatic. At the present time the patient is under observation.

Results. There was no blood loss during both surgery procedures. The duration of the first operation was 90 minutes and of the second one 220 minutes. No intra- or postoperative complications were observed. In both cases the tumor was less than 5 cm. Both patients were classified as stage I. The post-surgery follow-up is 2 months in case 1 and 1.5 months in case 2. There is no evidence of relapse according to the 1st US investigation data.

Conclusion. Patients with localized renal cell carcinoma could be treated with surgery alone. But they should receive careful post-operative monitoring of lungs and liver to prevent a spread of a disease.

ABSTRACT NO.: PP-120

Diagnosis of renal tumours in neonates

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Key words: renal tumours, neonates, ultrasound diagnosis

Introduction. Renal tumors are quite common in childhood, ranking fifth among all pediatric tumors. Wilms' tumor is the most frequent renal tumor in childhood (represents 95 % of kidney tumors). Wilms' tumor is the more frequent in infants till 1 year and are its ratio with mesoblastic nephroma is about 2:1. But if we focus on the age group of infants under 6 months this ratio already can be is 1:2.

Aim. The aim of this study to learn the frequency and sonographic characteristics of main types of human embryonic tumors of kidney that manifest themselves in the perinatal and neonatal periods.

Materials and methods. A retrospective analysis data of first month of life infants with a confirmed diagnosis of a solid tumor of kidney was made. Depth of retrospective was 15 years.

Results. Proportion of patients with major forms of kidney tumors in the neonatal period amounted to 3.4 % from all children till 14 years with this disorder. These were 7 cases with a confirmed diagnosis of a solid tumor of kidney in neonates. In 6 of 7 of cases in neonates with renal tumors had mesoblastic nephroma. All 6 cases clinically have been classified as "definitely congenital" tumors, 3 of which were diagnosed prenatally at 33–35 weeks of pregnancy. On postnatal sonograms were determined the rounded forms solid tumors of the kidney 9–40 cm^3. Acoustic tumor density was middle or low, degree of vascularization was average or high. Indices of resistance for arteries in different sections of tumors were ranged from 0.50 to 0.79. If tumor was localized in the upper segment of the kidney we need to differentiated it and the neuroblastoma adrenal. We observed only one case of neonatal nephroblastoma. It was coming from lower pole of the left kidney, a solid, with a fuzzy on the border with parenchyma of kidney contour of the tumor. Tumor volume was 15 cm^3. Were determined of average tumor vascularization, deformation and expansion of renal pelvis, lymph nodes along renal pedicle. Was marked by rapid growth of neonatal renal tumor (tumor volume doubling time was 8–13 days). At this point it should be noted that in infants till 1 year in the ¾ cases tumors of kidney they have been diagnosed in volume exceeding 250 cm^3.

Conclusion. Ultrasound as the primary method of postnatal (neonatal) diagnosis of kidney tumors in all cases was. Prenatal detection of the visually similar (on malignant) benign forms of the renal neoplasms is indicate on a certain efficiency of the prenatal diagnosis of the renal tumor as such. 50 % effectiveness of prenatal ultrasound diagnosis for neonates with mesoblastic nephroma were noted. There is a low frequency of occurrence of nephroblastoma in the newborn (0.48 % of all cases of this tumor). Mesoblastic nephroma predominates in a ratio of 6:1. Significant dimensions of tumors in infants till 1 year with one hand and a low incidence of neonatal nephroblastoma with other hand are confirming the high growth rate of the nephroblastoma in age group till 1 year.

ABSTRACT NO.: P-255

The first experience of nephron-sparing surgery in children with Wilms tumor using a single laparoscopic access

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Key words: Wilms tumor, partial nephrectomy, nephron-sparing surgery, children

Introduction. In recent years, the question of preserving treatment in children with Wilms tumor is being discussed in the international literature. Unfortunately, preserving surgery can be performed not in all patients. But the surgical techniques improve at an incredible rate. Of course, the problem of nephron-sparing surgery in children with Wilms tumor is not limited by the improvement of surgical techniques. Oncohematologists, chemo- and radiotherapists, anesthetist, pathologists and other specialists are working on the problem. Only the joint efforts of various experts can cure the patients with such disease.

Aim. To identify the use of nephron-sparing single laparoscopic access surgery of children with Wilms tumor.

Materials and methods. Two children (3 and 4 years-old) with Wilms tumor were treated in the Children's Republican Clinical Hospital of Tatarstan Republic in 2015. Pre- and postoperative therapy was carried out in accordance with the Protocol SIOP 2001 / GPOH. The decision about the preserving surgery in the first case was made due to the bilateral process and in the second case to the volume of tumor lesions less than 30 % of the kidney. In both cases, as a surgical technique were performed partial nephrectomy by a single laparoscopic access. In both cases, the tumor was located in the left kidney. That is why the access to the abdominal cavity was performed through a 2.5 cm length incision on the left semicircle of umbilicus, using SILS-port (Covidien, USA), and CO2 insufflation into the peritoneal cavity up to 10 mm Hg, installation of the 5 mm laparoscope 30°. After the abdomen revision, the left retroperitoneal space was opened from splenic angle on the lateral border of the colon using dissector-roticulator, clinch-roticulator and ultrasonic scissors Harmonic (Johnson & Johnson, USA). Doing so, the upper pole of the left kidney was visualized. Initially, the ultrasonic scissors and dissector denuded the renal pedicle. The rubber band was applied on the renal vessels with the exposition of no more than 40 minutes. All the tumor arteries and veins extending from the renal pedicle were clipped and intersected. The tumor was dissected entirely after the renal resection at the level of the healthy parenchyma (5 mm from the tumor). The tumor was removed using a laparoscopic evacuation bag (in one case skin incision was increased to 4 cm). A retroperitoneal sanation was carried out. The renal wound was closed with hemostatic mesh and was sprinkled with hemostatic powder. The top of the wound were applied with kidney fat. Then the rubber band was removed from the renal pedicle. Intraoperative blood loss in both cases were about 25 ml.

Results. The removed renal segment with a tumor in the first case had a size of approximately 45 × 30 mm (weight about 50 g), in the second case about 95 × 80 mm (weight about 220 g). Six months later the surgery both children were tested on the computerized tomography, which showed no signs of metastasis or recurrence of the tumor. Parents of the patients were satisfied by the cosmetic results of the surgical treatment.

Conclusion. Thus, the partial nephrectomy in children with Wilms tumor using of a single laparoscopic approach may be the choice with good functional and cosmetic results.

ABSTRACT NO.: PP-260

Renal tumors in infants less than 6 months of age

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Key words: nephroblastoma, mesoblastic nephroma, infants

Introduction. Renal tumors are rare in infants less than 6 months of age. Nephroblastoma and mesoblastic nephroma (MN) are the most common tumors.

Aim. The aim of this study is to determine their frequency in this age group, and to define their clinical, histopathological characteristics, treatment of these tumors and outcomes of the patients.

Materials and methods. In this retrospective study, performed in pediatric surgery department of university hospital Mustapha Pacha of Algiers, we reviewed our 20 years experience, from january 1996 to december 2015.

Results. Among 23 cases of renal tumors in this category of patients, the nephroblastoma leads in the first position (15 cases), and 8 patients suffered from mesoblastic nephroma. the mean age was estimated at 3.24 months. The female prevalence was noted with a sex ratio of 0.7 (13 females against 10 males). In all these cases, the main symptom consisted of abdominal mass. All the 23 patients underwent a widened nephro ureterectomy; 14 of them had first surgery (8 MN and 6 nephroblastoma). The 9 other cases underwent preoperative chemotherapy. The most common of histologic type found was intermediate (6 cases). However 5 patients presented nephroblastoma with unfavorable histology. All patients with mesoblastic nephroma are alive, and 3 with nephroblastoma died; one of them following a head trauma.

Conclusion. In our study, the renal tumors observed in infants less than 6 months of age represente 6.33 % of all tumors of kidney in the child. We have noted a high frequency of nephroblastoma (65.2 %) compared to the mesoblastic nephroma which takes the second rank in this type of tumors. This study has been carried in order to improve the medical care of the patients. In this age group, we have to think first of malignant tumors, and to better analyze the imaging supports and the extension of the tumors.

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ABSTRACT NO.: OP-266

Neoadjuvant transcatheter arterial chemoembolization combined with systemic chemotherapy for treatment of clear cell sarcoma of the kidney (CCSK)

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Key words: transcatheter arterial chemoembolization, clear cell sarcoma of the kidney

Introduction. Clear cell sarcoma of the kidney (CCSK) is a rare type of pediatric malignant renal tumor. It is known as an aggressive tumor with poor prognosis.

Aim. The aim of the study was to evaluate the efficacy of neoadjuvant transcatheter arterial chemoembolization (TACE) combined with systematic chemotherapy for the treatment of CCSK in children.

Materials and methods. From January 2010 to December 2015, 5 patients (2 boys and 3 girls; median – 2.9 years, range 0.9–7.1 years) with unilateral CCSK were treated with preoperative TACE combined with systemic chemotherapy. All of these patients were diagnosed histologically by percutaneous core-needle biopsy before treatment. At diagnosis, the mean maximal tumor diameter was 11.7 cm (range: 6.7–14 cm). Two patients (2/5) presented with lung metastasis, 1 (1/5) with bone metastasis, and 1 (1/5) with inferior vena cava (IVC) thrombus.

Patients subjected to TACE by Seldinger's method. A catheter was placed into the involved renal artery. Chemoembolization emulsion consisted of cisplatin (80–90 mg/m^2), pirarubicin (40 mg/m^2), vindesine (3 mg/m^2) , normal saline (120–180 ml) and iodized oil (5–10 ml) were infused into the renal artery. Preoperative systemic chemotherapy with vindesine (3mg/m^2/day, on day 1), ifosfamide (1200mg/m^2/day, on days 2–4), and etoposide (100mg/m^2/day, on days 5–7) was administered 3 weeks after TACE. Surgical resection carried out 3 weeks after neoadjuvant intravenous chemotherapy. Postoperative received radiotherapy and chemotherapy with a combination of carboplatin, ifosfamide, etoposide, pirarubicin, vindesine for 24 weeks.

Results. No cardiotoxicity, renal insufficiency, or hepatic dysfunction were found in all patients. Grade III marrow suppression developed in 2 patients. In terms of RECIST criteria, 3 patients had a partial response (PR), 1 had stable disease (SD) and 1 showed progressive disease (PD) after neoadjuvant therapy. Four patients underwent radical nephrectomy after neoadjuvant therapy. Complete surgical removal of the tumor achieved in 3 patients and 1 had intraoperative tumor rupture. Surgical stages of this 4 patients were stage II in 1, stage III in 2, and stage IV in 1 patients. Pathologic examination of surgical specimens found tumor necrosis > 90 % in all 4 patients. This 4 patients were recurrence free survival up to now with a median follow-up of 44 months (range, 25–62 months). Another patient with tumor progress after neoadjuvant therapy failed to surgery, died of lung metastasis and bloody ascites.

Conclusion. Neoadjuvant transcatheter arterial chemoembolization combined with systematic chemotherapy for the treatment of clear cell sarcoma of the kidney in children is safe and effective.

ABSTRACT NO.: OP-268

Inhibition of autophagy in nephroblastoma and the potential therapeutic significance

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Key words: nephroblastoma, pediatric tumor, autophagy, chloroquine

Introduction. Nephroblastoma (NB) is the most common abdominal malignant neoplasms and contributes to 6 % of malignant pediatric tumors. Currently, it is treated by surgery in combination with chemo- and radio-therapy. NB has overall good prognosis, however, patients still suffer from the side effects of traditional therapies. Autophagy is a self-digestion physiological event to maintain cellular homeostasis. Deregulation of autophagy has been proved to participate in the onset and progression of various malignant tumors. Investigations of the autophagic activity in NB have not been reported in the literature. However, abnormal expressions of Bcl-2, β-catenin and p53, which directly regulate the autophagy activity, are manifested in NB. Hence we hypothesize a potential link between autophagy deregulation and the onset of NB. Furthermore, regulation of autophagy can be a potential strategy as adjuvant therapy to increase the efficacy and to decrease the side effects of traditional therapeutics.

Aim. Our study attempts to explore the relationship between autophagy and NB with the purpose to find optimized protocol by autophagy regulation.

Materials and methods. The following experiments were performed.1. Expression of several autophagy-related genes (ATGs) were analyzed in the mRNA and the protein levels in NB tissues. 2. In NB cell lines, novel therapeutic strategies were tested by combinational use of autophagy-targeting drugs and conventional chemotherapeutics to increase the efficacy and to decrease the side effects. 3. In nude mice NB tumor model, the synergistic antitumor effect of chloroquine, a typical autophagy-suppressant, with the antimetabolite vindesine was tested.

Results. 1. There was suppression of autophagy in NB tissues prior to chemotherapy compared with the surrounding normal kidney tissues; autophagy was activated after chemotherapy and might participate in chemoresistance development. 2. The microtubule-targeting antimetabolites vincrestine and vindesine had synergistic antitumor effect with both chloroquine (autophagy-suppressing drug) and rapamycin (autophagy-promoting drug). The anti-proliferative effect of rapamycin on NB was at least partly through activation of autophagy. 3. The synergistic antitumor effect of chloroquine with vindesine was confirmed by in vivo experiments in nude mice.

Conclusion. Through this project, a standardized tumor tissue bank has been built in our hospital and will serve as the tool and the resource for future research. We have examined the background autophagy levels in the NB tissues, and found autophagy was suppressed and could be re-activated by chemotherapy. In both in vitro and in vivo experiments, inhibition of autophagy (by chloroquine) was shown to decrease tumor growth synergistically with microtubule-targeting chemotherapeutics. From this project, new insights were gained in the tumorigenesis and chemoresistance mechanisms of NB, and novel therapeutic strategies could be further investigated based on the current data.

ABSTRACT NO.: OP-295

Rare pediatric renal tumors: always keep in mind. An experience of one laboratory

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Key words: renal tumors, rare tumors, pediatric renal tumors

Introduction. The most well known and diagnosed pediatric renal tumor is nephroblastoma. But wide age limits in childhood (0–18 years) should always be taken into account to not miss a rare tumor.

Aim. To show wide spectrum of pediatric renal tumors of all ages in group, including newborns and older children.

Materials and methods. 231 renal tumors were studied in our department since January 2012. There were 187 pediatric cases including 77 children operated on in our clinic. Patients's age varied from 1 month to 17 years. Only 100 entire kidneys were delivered to the laboratory and could be examined fully and carefully. The rest of tumors were biopsied or cut not in the right way so they couldn't be graded. Histologically there were nephroblastoma (119 – 63.6 %), rhabdoid tumor (14 – 7.5 %), nephrogenic rests (14 – 7.5 %), congenital mesoblastic nephroma (11 – 5.9 %), clear cell sarcoma of kidney (10 – 5.3 %), translocational carcinoma (5 – 2.7 %), metanephric adenoma (5 – 2.7 %), clear cell carcinoma (2 – 1.06 %), cystic nephroma (2 – 1.06 %), high grade sarcoma NOS (2 – 1.06 %), medullary carcinoma (1 – 0.5 %), juxtaglomerular cell tumor (1 – 0.5 %), metanephric stromal tumor (1 – 0.5 %). Almost in all consultative cases there was no clinical information except age and location of the tumor. All the tumors were diagnosed histologically and in some cases by using immunohistochemistry as well.

Results. The diagnosis was confirmed in 107 cases (57.2 %), 52 tumors were diagnosed primary (27.8 %), in 28 cases the incoming diagnosis was changed cardinally (14.9 %). None of benign tumors were diagnosed correctly by the local pathologist or suspected by clinician. None of consultative rhabdoid tumors were diagnosed in local departments of pathology and only half of it were suspected by clinician.

Conclusion. Not only nephroblastoma should the first diagnosis in pediatric renal tumors. The patient should be examined fully and, moreover, complete clinical information must be provided to the pathologist to avoid rough diagnostic mistakes and misdiagnosing the tumor.

ABSTRACT NO.: OP-330

Five year experience of Wilms tumor at a tertiary care centre, where we stand? A developing country perspective

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Key words: WT

Introduction. We present a retrospective study, looking at demographics and outcome of children with WT presented to the hematology and oncology department of the Children's Hospital Lahore between January 2009 and December 2013.

Aim. The main objective of the study is to discuss presentation and outcome of children with Wilms tumor (WT).

Materials and methods. All children diagnosed as WT on radiological findings and proven on histopathology after needle biopsy were included. Data regarding age, gender, initial staging and outcome were recorded and analyzed. CT scan abdomen, chest and needle biopsy were done in almost all patients. According to SIOP WT 2001 (UKCCSG) Protocol all patients were treated with Pre-operative chemotherapy, surgery followed by post-operative chemotherapy and radiotherapy in some cases.

Results. A total of 175 patients were included. Males and females were equal in number 51.4 % and 48.6 % respectively. Majority 88 (50 %) were between 2–5 years of age, 44 (25 %) patients were above 5 years and 43 (24.6 %) below 2 years. Sixty two (36 %) presented in stage III, 37 (21 %) stage IV metastatic disease, 26 (15 %) stage II and only 5 % in stage I and V each. Seventy four (42.2 %) patients had completed treatment advice, 20 (11.4 %) died and 10 (5 %) had relapse while 65 (22.2 %) were lost to follow up with missing record.

Conclusion. Most of patients presented at less than five year of age with advance disease stage III and IV. Treatment outcome is fair 42.2 % with abandonment and 67 % without abandonment with overall mortality 11.4 %. Abandonment due to multiple social reasons is major factor affecting the overall survival rate that need to be addressed to improve ultimate outcome.

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ABSTRACT NO.: OP-355

The treatment of Wilms' tumour, a single institution experience

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Key words: paediatric renal tumours, Wilms' tumour, nephroblastoma

Introduction. There is no national protocol for treatment of Wilms' tumour (WT) in Russian Federation yet. The Department of Paediatric Haematology and Oncology of Clinical City Hospital 31 is not a participant of SIOP 93-01/GPOH and SIOP WT 2001 but we were allowed to follow diagnostic and treatment recommendations of these protocols.

Aim. The aim of our study was to evaluate the results of a treatment in patients with WT who were treated according to the protocols in City Hospital 31 in St.Petersburg.

Materials and methods. We analyzed a retrospective data review of 51 patients (pts) with WT over a period of 16 years (1999–2015). There were 28 girls and 23 boys in our group. Forty seven pts developed an unilateral WT and in 4 pts both kidneys were involved. The median age was 2 years (1 mo – 8 yrs). A pre-treatment aspiration biopsy was provided in 23 children (45 % cases).

Results. The median follow up time was 42 months (1–178 months). Five years event free survival (EFS) was 84.9 ± 5.2 % and the overall survival was 86.9 ± 5.7 %. The EFS of children with stage I (5 pts) was 100 % and it was 95.2 ± 4.6 % in stage II tumor group (24 pts). EFS at 5 years for stage III (8 pts) and IV (12 pts) were 87.5 ± 11.7 % and 58.3 ± 14.2 % respectively. Two patients with bilateral WT without lung metastases are in the complete remission now and have a normal renal function. There were only 2pts in the low risk group and both are alive. EFS at 5 years for 42 children in the intermediate risk group (IRG) was 91.8 ± 4.5 % and 50.0 ± 20.4 % for 6pts in the high risk group (HRG). Six pts relapsed. The median time of recurrence was 7 months: one child achieved a second remission (in IRG) but was lost to follow up, 4 pts died of the recurrence (in HRG), and another patient from IRG with a local relapse of stage II WT continues his treatment now. There were 2 treatment-associated deaths: 1st- during initial preoperative chemotherapy from disseminated intravascular coagulation, 2nd- in complete remission of bilateral WT stage IV (IRG) due to an acute intestinal obstruction at the end of the postoperative chemotherapy.

Conclusion. The present study demonstrates the favorable outcome in the local stage cases with an initial preoperative chemotherapy followed by a surgery and a postoperative chemotherapy. However, in the stage IV cases and in the high risk group the survival rate is not satisfactory.