55.

WILMS' TUMOUR (WT) TRIALS AND STUDIES FROM THE INTERNATIONAL SOCIETY OF PAEDIATRIC ONCOLOGY (SIOP) 1971-1982. For the Trial Committee: P.A. Voûte, M.F. Tournade, H.J.M. Perry, J. Lemerle, J. de Kraker, Amsterdam, Villejuif/Paris.

Four studies were conducted. The first one concerning pre-operative radiotherapy (RT) to direct surgery followed by post-op. RT. A second randomisation was done in single versus multiple courses of actinomycin D (AD). From this study a preference was found for pre-op. treatment. No difference in single to multiple courses AD was found. Recurrence free survival (r.f.s.) being ± 50%, actuarial survival (a.s.) ± 70%. The second study was non-randomised. It concerned 6 months to 15 months of maintenance chemotherapy (CT) vincristine (VCR) and AD. No difference could be found with a r.f.s. of 79% and a.s. of 84%. 6 months CT is enough irrespective of stage. The 3rd WT study SIOP no. 5 compared pre-op. RT to pre-op. CT. 161 patients were included. For the CT arm and the RT arm r.f.s. and a.s. were: 76%, 66%; 90%, 85% respectively. Pre-op. CT is just as good as pre-op. RT. 50% of the patients with pre-op. therapy had a stage I tumour compared to 22% with immediate surgery. This is a great advantage, 50% of the patients do not need RT at all. Pre-op. therapy gives no problem in the diagnosis of so-called unfavourable histology. In the 4th WT study SIOP no. 6 all patients receive pre-op. CT with AD and VCR. Stage I patients are randomised in respectively 3 months and 6 months maintenance CT. Stage II with negative lymphnodes are randomised concerning post-op. RT yes or no. Stage II with positive lymphnodes and stage III patients are randomised in respectively intensified VCR and adding adriamycin in maintenance CT. Maintenance CT in the last 2 groups is 6 months. April 1982 207 patients were registered, 113 were suitable for trial. It is too early to draw conclusions. The study goes on. The conclusion from all the studies is: centralized treatment of this rare disease in children is necessary. Pre-op. CT should be given to all patients with a WT.

56.

NEPHROBLASTOMA STAGE IV. For the Trial Committee: J.de Kraker, P.Bey, D.Bürger, M.Carli, C.Nihoul-Fékété, P.A.Voûte, J.Lemerle. Amsterdam, Villejuif/Paris.

Under supervision of the organising committee of the nephroblastoma trials and studies of SIOP, a multi-center pilot study was designed. In this study the definition of Wilms' tumour stage IV patients will be restricted to those with haematogenous lung metastases at presentation. Rationals for the study were: a The still unfavourable prognosis of patients with stage IV disease. b The need for a more uniform treatment protocol, c To study the possibility of cutting on lung irradiation in some of these patients. To reach this, it was decided to initially intensify the pre-operative chemotherapy (CT). This will consist of Actinomycin D (AD) 15 µgr/kg/i.v. day 1, 2, 3 - 15, 16, 17 - 29, 30, 31. Vincristin (VCR) 1.5 mg/m^2/i.v. once a week for 6 weeks, and Adriamycin (ADR) 25 mg/m^2 day 8, 22, 36. After nephrectomy, metastatectomy will be considered. Postoperative treatment will depend on the fact if metastases are still demonstrable and the local stage of the tumour. Lung irradiation is necessary in case of incomplete resected or inoperable metastases. In case of a local stage II tumour with positive lymphnodes or stage III, radiotherapy is given to the abdomen according to the protocol SIOP VI. Postoperative CT will consist of AD 15 µgr/kg for three consecutive days and VCR 1.5 mg/m^2 once a week for at least 4 weeks. Maintenance therapy will be the same for all the patients. To start with VCR 1.5 mg/m^2/i.v. and ADR 50 mg/m^2/i.v. four times with a three week interval followed by 5 courses AD 15 µgr/kg/i.v. 5 days and VCR 1.5 mg/m^2 i.v. day 1 and day 5 every 3 weeks. Besides actuarial and disease free survival, cardiotoxicity and other treatment related sequelae will be of special concern.

57.

THE FIBROADENOMATOUS TYPE OF NEPHROBLASTOMA. J.F.M.Delemarre, B.Sandstedt, M.F.Tournade, for the S.I.O.P. Nephroblastoma Trial and Study, secretary: Emma Kinderziekenhuis, 1018 HJ Amsterdam, The Netherlands.

After revision of the microscopical sections of 889 patients from the files of the SIOP Nephroblastoma Study and Trial 25 cases of a special subtype were recognised. Because of the likeness of the histological structures with those seen in fibroadenoma of the ovary this type is called: fibroadenomatous type of nephroblastoma. After studying clinical and histological factors in correlation with actuarial survival and recurrence free survival in comparison with the whole series of SIOP Nephroblastomas, it was possible to recognise this subtype as a clinicopathological entity with favorable prognosis.

58.

CISPLATIN IN THE TREATMENT OF RECURRENT WILMS' TUMOR. A.T. Meadows, J.Wilimas, J.Champion, J.Belasco, Children's Hospital Cancer Research Center, Philadelphia, Pennsylvania, and the St. Jude Children's Research Hospital, Memphis, Tennessee, U.S.A.

Thirteen patients received cisplatin for recurrent or unresponsive Wilms' tumor and 3 of the 12 who were evaluable had transient and partial responses. The inevaluable patient was a 6 year old female with recurrent pulmonary metastases who died hours after the infusion. All had received Vincristine, Actinomycin D and Adriamycin prior to the administration of cisplatin and most had received pulmonary and abdominal irradiation. Two patients demonstrated partial clearing of pulmonary metastases which lasted for 2 and 3 months, while one patient had a decrease in the size of his abdominal recurrence. This lasted 3 months but grew back just prior to a scheduled laparotomy for removal of the residual tumor. The dose of cisplatin used was fairly uniform: 90 mg/m^2 or 3 mg/kg.

The characteristics of these patients differed somewhat from the usual population of Wilms' tumor patients; 8 were male and 4 were female. The majority (9) were Stages III or IV at diagnosis. It is noteworthy that while the median age of the patients was 4 years, the 3 patients with early stage disease in this group were 7, 9 and 14 years of age. In addition, of the 3 patients with unfavorable histologic types, two were the only black patients and one was a 13 month old white male.

We conclude that cisplatin may have some efficacy in the therapy of Wilms' tumor and studies of its use earlier in the progression of disease should be considered.

Supported in part by a grant from the National Institutes of Health (CA 14489).

59.

IS TOTAL NEPHRECTOMY ALWAYS ADVISABLE IN THE TREATMENT OF NEPHROBLASTOMA ? . A.Gentil Martins, M.Sousinha, J.Valença de Sousa, M.A.Picanço, J.O. Queiroz. - Clínica Oncológica IV (Pediatria), Instituto Português de Oncologia de Francisco Gentil, Lisboa - Portugal.

It has been standard policy almost all over the world to perform total nephroureterectomy for malignant tumors of the kidney, mainly for nephroblastoma.
Due to advances in chemotherapy and the improved results obtained we felt it justified, whenever technically possible (section on normal tissues) to preserve the non-involved protion of the kidney.
Of a total of 80 patients 10 had heminephrectomy performed (10%). These 10 patients had 12 heminephrectomies (in one case for bilateral tumor and another for recurrence in a horseshoe kidney). Follow up is respectively 8y, 6y, 4y, 3y, 2y(x3), 1y(x2), 1w. Only in the case of a horseshoe kidney there was local recurrence. This patient was lost to follow-up 1 year latter (died?) and only the patient with bilateral tumor has died from pulmonary metastases. All others are so far alive and well.
We fell that there is a case to be conservative in the surgical treatment of Nephroblastoma, namelly in polar tumors, with no increased risk of local recurrence if surgery respects the basic principles of tumor ressection.