

overall survival rate at five years from diagnosis was 15%. Three children were found to have a family history of polyposis coli affecting the mother and maternal relatives. Interestingly two of the three children with the family history of polyposis coli are alive and disease free more than seven years after total resection of their tumours. The third child died from intestinal obstruction secondary to adhesions three years after complete removal of the hepatoblastoma but with no evidence of recurrent disease. The histology of these cases has been reviewed and in all cases the hepatoblastoma was of a well differentiated type. Detailed chromosome banding studies with particular reference to chromosome 2 have been carried out on the mothers of the three children in our initial series, the two surviving children and their siblings. Polyposis coli is also rare with an estimated incidence of approximately 1 in 8,000 births (0.13%). In this series of 113 children with hepatoblastoma three of the mothers (2.7%) have polyposis coli thus the rate among these mothers is about 200 times that found in the general population. The outcome for these children suggests that cases of hepatoblastoma associated with polyposis coli may have better prognosis than usual.

66.

PRELIMINARY RESULTS OF A GERMAN COOPERATIVE EWING'S SARCOMA STUDY WITH INITIAL CHEMOTHERAPY AND DELAYED LOCAL THERAPY
H. Jürgens, Pediatric hospital, University of Duesseldorf, Moorenstr. 5, D-4000 Duesseldorf, West Germany
(for the German Pediatric Oncology Society trial committee)
In 1980 a cooperative German Ewing's sarcoma study was initiated with primary chemotherapy and delayed local therapy. Chemotherapy consisted of four 9 week courses of combination chemotherapy with VACA (vincristine, actinomycin D, cyclophosphamide and adriamycin) and patients were randomized for additional bleomycin. Patients with evaluable tumor regression under primary chemotherapy received local therapy having completed 2 courses of chemotherapy. Local therapy was individualized and consisted of either radical surgery with complete resection of the bone involved, or incomplete resection of the involved bone followed by radiation with 3600 rad, or radiotherapy only. Patients with radiation only for local therapy were randomized for 4600 rad versus 6000 rad tumor dose.
In April 1982 35 patients were entered into the trial. 27/35 patients are considered protocol patients. Reasons for excluding patients from the protocol were: metastatic disease at diagnosis or primary local therapy. 23/27 patients are free of disease on April 15, 1982 with a median period of observation of 8 months. All 4 patients with radical surgery for local therapy remained free of disease.
1/11 patients with radiotherapy alone developed pulmonary metastases 8 months from diagnosis having received 6000 rad tumor dose.
3/7 patients with surgery followed by radiation developed metastases 12, 10 or 4 months from diagnosis. All 9 patients with distal extremity tumors are disease free, 1/7 patients with proximal extremity lesions and 3/10 patients with central lesions developed metastases.

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A. Feldges, P. Imbach, A. Lüthy, H.J. Plüss, E. Signer, H.P. Wagner and M. Wyss.
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STRATEGY IN THE TREATMENT OF CHILDHOOD HISTIOCYTOSIS X
From 1976 to 1981 30 children with Histiocytosis X were entered in a prospective national study. The goal of the study was to diagnose all children with Histiocytosis X in Switzerland and to give a uniform treatment to all disseminated forms. In 14/25 evaluable cases with histologically confirmed diagnosis the disease was limited to bone. In 6 children with solitary eosinophilic granuloma no progression was observed for at least 1 year after surgery and/or radiotherapy. 6 of 8 children with multiple bone lesions achieved complete remission on vinblastine (VLB) and prednisone (P) and were maintained with 6-mercaptopurine (6-MP). One relapsed and 5 live with no evidence of disease, 4 off therapy for 1 - 1½ years. None of the 8 patients with multiple bone lesions died.

11 children with soft tissue - and/or organ involvement were treated more aggressively with VLB/P pulses and 6-MP and methotrexate for maintenance. One child with pulmonary disease did not respond and died. The remaining 10 patients have a stable disease and in 5/10 therapy could be stopped after 2 years.

68.

CENTRAL VENOUS CATHETERIZATION IN PEDIATRIC ONCOLOGY.
M. Meignier, X. Rialland, A. David, J. Crambert, J.L. Harousseau, Unité d'Hématologie et Oncologie Pédiatriques, 44035 NANTES CEDEX, France.

Advantages of long-term central venous catheterization (CVC) are well recognized for children who require prolonged intravenous therapy. We investigated this technique in Pediatric Oncology for administration of chemotherapy. In 14 children aged 2 to 14 years (mean 8,5 y.), a percutaneous central venous catheter was performed in 26 instances (subclavian vein 12, internal jugular vein 6, brachial vein 6, femoral vein 2). The catheter was left in place for a duration of 2 to 45 days (mean 25,8 days). In 16 cases (65 %) catheter was removed after completion of chemotherapy without complication. A complication inherent to catheter occurred in 10 cases (dislodgment 2, phlebitis 3, bacteremia 2, catheter occlusion 3). No death was imputable to these complications. C.V.C. permitted a better supportive care for these children in spite of a potential high risk inherent to thrombopenia and neutropenia. More, it greatly improved the comfort of these patients who required long-term intravenous infusions.

69.

AGGRESSIVE TREATMENT OF B-CELL NON HODGKIN LYMPHOMA (NHL) - A PROTOCOL OF THE FRENCH PEDIATRIC ONCOLOGY SOCIETY - PRELIMINARY RESULTS ON 60 PATIENTS. C. Patte, E. Benz-Lemoine, T. Philip, F. Demeocq, A. Bernard, C. Rodary, J. Lemerle, for the organizing committee.

Based on results of a previous study and on our experience in treating recurrences, a new protocol was designed for stages III and IV and some stage II B-cell NHL. The original protocol was as follows: A. Induction: 1) Two successive courses: CPM 1 g/m²/dx3; VCR 2 mg/m²x3; PRED 2 mg/kg/dx10; ADR 60 mg/m²; HDMTX 3 g/m²; IT MTX. 2) ARA-C 100 mg/m²/dx5 in continuous infusion (CI); ASP 1000 U/kg/dx5; HDMTX 3g/m²; ADR 45 mg/m²/dx2; IT MTX; IT ARA-C. 3) BCNU 60 mg/m²; ARA-C 100 mg/m²/dx5 CI; CPM 0,5 g/m²/dx3; 6-TG 150 mg/m²/dx5. B. No radiotherapy at all. C. Maintenance: two monthly alternative courses with 1) HDMTX, CPM, ADR, VCR, PRED and IT MTX. 2) BCNU, ARA-C IT ARA-C, ASP, 6-TG. The treatment was to be completed within one year. Due to toxicity, the protocol was changed: CPM dose was cut in the 1st induction course and ADR removed from the 3rd.

In June 1982, 11 centers in France had included patients (pts) in this non-randomised study. 32 pts were treated according to the original protocol. 30/32 went into complete remission (CR). 3/30 recurred at 5%, 8, 8 months while on maintenance: 1 in bone marrow, 2 in CNS. 5 died from toxicity during induction. 22 are in first remission, follow-up being 8 to 16 months. Actuarial recurrence-free survival is 69 %. 28 pts are included in the modified protocol: 11 have completed induction phase. 10 went into CR. 2 died from toxicity. 15 are in induction phase.

From these preliminary data, survival and recurrence free survival, despite toxicity in the first protocol appear to be superior to those previously obtained.

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