with MTX than MTX alone. However, all children reach an normal range IQ in validated psychometric tests. MRI lesions cannot predict neurocognitive function.

1242 POSTER

Cancer in relatives of children with haematological malignancies

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Familial aggregation has been reported for virtually every form of human cancer. Accordingly, many multiple-case families with haematological malignancies have been described and this led to the suggestion of genetic susceptibility to these diseases. We undertook a family study of the frequency and type of cancer in relatives of 76 children affected by acute leukaemia and malignant lymphoma treated at the Division of Haematology and Ontology, University Paediatric Clinic Rijeka, in the period from 1980 to 1999. The information was obtained from interviews with the parents of affected children regarding the occurrence and type of cancer in relatives. Whenever possible, medical records and death certificates were sought for reported tumours. In the absence of medical confirmation, a questionnaire covering the same information was sent to the interviewed parents to cross-check the information. If there was any discrepancy, a second interview was scheduled to reconcile the differences. The control group consisted of 76 healthy children of the same age and sex. Our results show that there is a significant excess of cancer in the families of children affected by leukaemia and lymphoma. The higher frequency was obtained for the first- and second-degree relatives, while there was no difference in the incidence of cancer among more distant cases' and controls' relatives. Regarding the type the cancer, leukaemia, gastric cancer, and cervical cancer were significantly more frequent. Our results suggest that genetic factors play an important role in the aetiology of haematological malignancies in children. Continued large epidemiological and molecular genetic studies are needed to estimate precisely the inherited fraction of childhood leukaemia and lymphoma and to identify individuals at risk.

1243 POSTER

High dose chemotherapy in children with malignant lymphoma

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Purpose: This retrospective analysis was undertaken to determine overall survival and prognostic factors of children and adolescents with primary refractory or recurrent malignant lymphoma, treated with high-dose chemotherapy with autografting.

Methods: Forty-five children with poor-prognosis malignant lymphoma, of whom 27 were boys, underwent megatherapy between January 1992 and April 2000. The reasons for high-dose chemotherapy were: poor initial response to first-line chemotherapy (14x) or relapse (31x). There were 28 patients with Hodgkin's disease and 17 with non-Hodgkin's lymphoma. The median age was 15,7 years. The conditioning for Hodgkin's disease patients contained cyclophosphamide, etoposide and busulfan or carmustine. Patients with non-Hodgkin's lymphomas received cyclophosphamide, etoposide and busulfan or total body irradiation. Bone marrow was used as the source of hematopoietic stem cells in 10 patients, peripheral blood in twenty-eight, and both in seven.

Results: After a median follow-up of 47 months the overall survival is 61%. Eleven patients died of disease progression, 4 secondary to infectious complications, one of car accident. Median time to relapse after transplantation was 7,5 months. In the univariate analysis, minimal residual disease before transplantation was significantly associated with improved survival.

Conclusion: Further improvement of these results will require earlier transplantation, improved preparative regimens or early posttransplant immunotherapy.

Paediatric solid tumours

1244 POSTER

The prognostic value of the timepoint of relapse in paediatric rhabdomyosarcoma-like tumors

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Introduction: Rhabdomyosarcoma (RMS)-like tumors are a tumor entity consisting of embryonal (RME), alveolar (RMA), RMS not other specified (RMS), extraosseous ewing sarcomas and primitve neuroectodermal tumors (PNET). Prognosis (EFS and SUR) correlates with subtypes (favourable: RMS, RME, unfavourable: RMA, EES, PNET).

We evaluated EFS (time from first to second event) and SUR after first relapse of RMS-like tumors depending on the time of relapse. Early, intermediate and late relapsing patients were evaluated concerning histology and type of relapse.

Patients: Of 1461 patients with RMS-like tumors 1323 met evaluation criteria (no pre-treatment for malignancy, no second malignancy, no relapse at registration, age <21 years). Of these 294 relapsed after the end of therapy. 184 patients had a local, 110 a metastatic relapse. 166 patients had favourable, 81 unfavourable histology.

Results: Of 294 patients 34%, 32% and 34% relapsed within 6 (early), 6 to 12 (intermediate) and more than 12 (late) months respectively after the end of therapy. 5 year EFS to second event was 10%, 18% and 29% for early, intermediate and late relapsing patients respectively ($p<.001$). 5 year survival was 12%, 20% and 32% for early intermediate and late relapsing patiens respectively ($p<.001$). 5 year EFS and SUR after local relapse was 18% and 19%, 32% and 35%, 38% and 41% for early, intermediate and late relapsing patients ($p<.001$ for SUR and EFS). 5 year EFS and SUR after metastatic relapse was 0% and 0%, 0% and 5%, 8% and 12% for early, intermediate and late relapse ($p<.001$ for EFS and SUR). Favourable histology showed 5 year EFS and SUR of 13% and 13%, 27% and 30%, 38% and 45% for early, intermediate and late relapse ($p<.01$). For unfavourable histology 5 year EFS and SUR was 8% and 8%, 8% and 8%, 8% and 10% for early, intermediate and late relapses ($p<.01$).

Discussion: Relapsing patients with RMS-like tumors have an unfavourable prognosis both concerning EFS and SUR. Whether relapse occurs within 6, 6 to 12 months or after 12 months has significant influence on prognosis. EFS and SUR after metastatic relapse and/or unfavourable histology is almost null, after local relapse and/or favourable histology and late relapse SUR remains at 45%.

1245 POSTER

Response rates and prognosis depending on response in non-rhabdomyosarcomas. A report from the CWS study

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Objective: To asses response rates in non-rhabdomyosarcoma (NRMS). To evaluate prognosis depending on response rates in NRMS.

Patients: 101 patients with localized NRMS (4 largest groups: synovial sarcomas (SS): n=20, neurofibrosarcomas (NFS): n=18, undifferentiated sarcomas(UDS): n=13, malignant rhabdoid tumor (MRT): n=10) registered with the CWS study between 1981 and 1997 met evaluation criteria (age <22 yrs, no pretreatment, surgery within 8 weeks of begin of chemotherapy, no second malignancy, no relapsed patient at registration). Response was measured by CT or MRI. Depending on tumor-regression response was assessed after 9 to 16 weeks of chemotherapy: complete regression: complete response (CR), regression >2/3: good response (GR), regression <2/3 but >1/3: partial response (PR), regression <1/3: non response (NR) evidence of tumor growth: progressive disease (PD).

Results: Response rates were: CR 11%, GR 24%, PR 14%, NR 45% and PD 6%. Response rates for different histologies were: UDS: 85% SS: 45%, MRT: 40%, NFS 39%. EFS for non-responders vs. responders was 69% vs. 29% ($p<.0001$). Prognosis correlated well with response (EFS: CR 82%, GR 74%, PR 50%, NR 30%; PD 17% $p<.007$). In NR irradiation went along with unsignificantly better prognosis (EFS with irradiation 37%, without 22%). Secondary resection (sR) influenced prognosis significantly (EFS: sR0 67%, sR1 60%, sR2 0%; $p<.01$).

Conclusion: Response is an important predictor for prognosis in NRMS. There is a linear relation between prognosis and response. Irradiation may improve outcome but numbers are non-significant. Treatment of choice in non-responding NRMS remains secondary - if possible complete -resection.