From 1960 to 1996 we followed up 64 children aged from 1 to 18 years, which is about 18.1% of all DT patients (352). There were 26 boys and 38 girls. Eleven children had abdominal desmoids (AD), 51 had extraabdominal desmoids (ED), and 2 children had a combination of AD and ED. Congenital DTs were registered in 12 children, mothers of three of them having DT in combination with diffuse polyposis of the colon. Primary DTs were in 37 children, 27 presented with recurrences, 20 of them with multiple ones. DT sites were femur (26), gluteal area (18), abdominal wall (11), shin (10), foot (8), thoracic wall (7), hand (6), neck (5), shoulder (4), lumbar area (4), perineum (2), abdominal wall and femur (2), cheek and lower jaw (2). The total number of desmoid lesions was 105. The size of DT ranged from 5 to 35 cm, its maximum weight was 3.8 kg.

105 DTs in 61 children were subjected to surgical treatment, 3 patients were administered no surgery. After surgical treatment relapses occurred in 60% of the cases. After combined modality treatment they occurred two times rarer and after a longer period of time. Remote results are known in all the patients: 14 persons are alive for 3–5 years, 6 – for 6–10 years, 18 – for 11–15 years, 13 – for 16–20 years after the first operation. Of all the children, only one girl has an inoperable tumor. The rest are healthy. In none of the cases malignization of DT, conversion into fibrosarcoma or metastasizing were observed. In one girl a spontaneous regression of DT recurrence was registered at the beginning of menses and subsequent two childbirths. The mother and children are healthy.

563 ORAL

Nasopharyngeal carcinomas of childhood

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Aim: This study reviews the authors' experience, from 1980 through 1995, in management and outcome of 56 children with nasopharyngeal carcinomas.

Methods: There were 23 females and 33 males, their ages ranging from 7 to 19 years (median, 16 years). Twelve children had WHO type 1, 4 had type 2 and 40 had type 3 carcinomas. Six children were at Stage II, 9 at Stage III and 41 at Stage IV (TNM-AJC). Sixteen children had T4 tumors and 18 had bilateral nodal involvement. External beam radiation therapy consisted of 50 to 70 Gy (median, 64 Gy) to primary tumor and 50 to 74 Gy (median, 66 Gy) to involved nodes, delivered in 1.8 to 2 Gy daily fractions. Fourteen children with T1–3 tumors received 1 to 3 (median, 2) fractions, each of 5 Gy, of HDR intracavitary brachytherapy boost. Thirteen children received neoadjuvant, 4 neoadjuvant and adjuvant and 7 concurrent chemotherapy.

Results: Follow-up ranged from 0.3 to 16.8 years (mean, 9.6 years). Complete primary tumoral response was achieved in 49 out of 55 and complete nodal tumoral response in 39 out of 40 evaluable children. Overall survival (OS) and disease-free survival (DFS) were 52.6% and 48.3% respectively, at 5 years and 52.6% and 48.3%, respectively, at 10 years. There were 6 primary, 3 nodal and 2 primary and nodal tumoral failures among children exhibiting complete response. There were 11 systemic failures among all children. In univariate analysis: T-stage and bilateral nodal involvement were significant prognostic factors for OS and T-stage. N-stage and bilateral nodal involvement for DFS. In multivariate analysis; age. T-stage. N-stage and addition of chemotherapy were significant prognostic factors for OS and T-stage, N-stage, and addition of chemotherapy for DFS. Two children died of adjuvant chemotherapy-related toxicity. Of 23 children with longer than 5 years of follow-up, 12 had mild neck atrophy, 3 shortening of clavicles, 2 trismus, 1 hypothyroidism and almost all xerostomia and dental caries.

Conclusion: Although radiation therapy alone is effective in achieving primary and nodal tumoral control, it should be accompanied with chemotherapy in attempt to improve OS and DFS.

564 POSTER

Specificities and optimization of peripheral blood stem cell collection in children: Treatment for malignancies can be mobilization

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Autologous peripheral blood stem cells (PBSC) is currently used to support high dose chemotherapy for children with solid tumors. However the volume of extracorporel blood loss required by leukapheresis and the veinous access could limit the procedure in small children.

Patients: Between 06/88 and 08/96, 136 children (median age: 96 months, median weight: 16 kg) with solid (n = 126) or hematologic malignancy (n = 10) underwent 141 stem cell mobilization episodes with hematopoietic growth factor (HGF) alone (86 first pts) or in combination with cytotoxic chemotherapy (CT) (different regimen prescribed for the malignancy, 50 pts). To optimize the PBSC collection, we have monitored the number of CD34+ cells in blood.

Results: 380 leukapheresis were performed with a median number of 3 per pt before 1996, and 2 in 1996 with a significant ($p = 0.05$) higher median of total CD34+ cells harvested ($8.99.10^6$ vs $6.74.10^6$ CD34+/kg). 80% of children had central lines for the PBSC harvest. We observed 45 clinical problems (hypocalcemia, hypovolemia, shock, and minor events: tiredness, isolated or in combination). 68% of pts were transfused at least once time with RBC and 8% required platelet support before the procedure. There is no difference between HGF alone and CT + HGF in term of mobilization failure. A significant correlation between the number of blood CD34+ cells (the day before and day of collection) and the leukapheresis product was found ($r = 0.59$ and 0.8 respectively). All the pts with $>1.10^6$ CD34+/kg in leukapheresis product had more than 11200 CD34+ cells/ml in blood. **In conclusion:** PBSC harvest required strategic care, especially for children who weight less than 10 kg. We have observed that most CT regimen used in pediatric malignancies can mobilize PBSC. The monitoring of daily blood CD34+ level at the end of aplasia is useful to select the best day to begin leukapheresis in order to optimize the number of collections.

565 POSTER

Thyroid cancer in childhood – Value of total thyroidektomy

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The prognosis of children with thyroid cancer is good despite frequent local invasion or metastases. The value of thyroidectomy is controverse.

Methods: Between 1970–1996, 20 children aged 4–16 years underwent surgery for thyroid cancer (papillary: n = 15; follicular: n = 3; medullary: n = 2). Clinical and pathologic findings, therapy and outcome were analyzed. Median follow-up was 7.5 years (1–25 y).

Results: 4 children had extrathyroidal tumor invasion (20%), 5 other had multifocal intrathyroidal tumor spread (25%). Cervical lymph node metastases were found in 9 (45%), distant metastases in 4 children (20%). All underwent total thyroidectomy, 4 as completion procedure. Radioiodine therapy was performed in 16 children (80%). Surgical complications included 1 vocal cord paralysis and 2 temporary hypocalcemias.

Cervical lymph node recurrences developed in 4 children. All were cured by surgical and radioiodine therapy. One girl died after 4 years from multiple metastases of a medullary thyroid carcinoma. All other are alive and free of disease.

Conclusions: Our surgical approach for thyroid cancer in children is total thyroidectomy with selective lymph node dissection. Indications are (1) a high frequency of multifocal and/or metastatic disease and (2) a smaller dosis of postoperative radioiodine.

566 POSTER

Evaluation of minimal and residual disease (MRD) in Ewing's tumors (ET) at diagnosis and during treatment

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Purpose: As Ewing's cells express specific chimaeric transcripts as a result of the t(11;22)(q24;q12) chromosome translocation or of its variant t(21;22), the reverse transcriptase-polymerase chain reaction (RT PCR) technique can detect MRD.

Material and Methods: Using a one tube nested PCR amplification procedure, blood and/or bone marrow (BM) from 113 patients with ET were evaluated for the presence of tumor cells at various times before and during treatment.

Results: At diagnosis, 16/62 patients had circulating tumor cells. The spontaneous occurrence of these circulating cells before biopsy could be established in some cases, but in other the detection might be related to the mobilisation of cells linked to the tumor sampling. The presence of circulating tumor cells was not correlated with the size of the primary tumors nor with the presence of metastasis. Ewing cells infiltrating BM were detected in 13/41 patients. The presence of RT-PCR positive BM was most frequently observed in patients with metastasis (8/17 vs 5/24), but, half of the patients (9/17) having lung or bone metastasis did not had RT-PCR positive BM.

Tumor cells could still be found in the BM after initial chemotherapy in two patients: both demonstrated rapid clinical progression.

Conclusions: 1) RT PCR positive circulating blood cells have no prognostic value in localised ET. 2) Successive analysis of BM by RT-PCR might be useful for the monitoring of response to treatment and ultimate prognosis.

567 POSTER

Port-Systems in children with malignancies; Our experience with 111 children under cancer management

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Background: Children with malignancies, especially those with high malignant grades, need intensive chemotherapy as well as supportive therapy. In order to accomplish this, one needs a very reliable venous access. From the whole range of the to date available indwelling devices e.g. Port-Systems, Hickman-catheters, central venous catheters, etc., one has got to take into consideration the advantages and disadvantages of the devices so as to fulfil the respective requirements. For our requirements (a sparsely populated federal state with a very large collecting area, we decided to use the Port-Systems.

Methods: 123 Port-System implantations were performed in 113 pediatric patients (average age = 7.1 yr) with malignancies. Sixty-seven (67) patients (60%) had solid tumors and 44 suffered from systemic malignancies. In most of the cases, the devices were implanted after confirmation of the diagnosis. The following company Port-Systems (age-related), in order of frequency, were used: Braun/Dexon (n = 106), Vygon and Fresenius. Sixty-eight (68%) percent of the patients received the Port-Systems in the cephalical vein, 28% in the internal jugular vein; in rare cases insertions were performed in the external jugular and/or the subclavian vein. Most of the systems were implanted in the right veins. The average operation time, in general anesthesia, was 45 minutes. A special leaflet was developed in order to ensure certainty and uniformity in the maintenance of the devices through the well-trained staff.

Results: The average dwelling time of the Port-Systems was 529 days (max. 2308 days = 77 months); the cumulative duration for all the systems totalled 65099 days (= 178.4 yr). The cumulative dwelling time of the devices increased during the examination period (1989 to 1995) from 8000 to 12506 days; significant average increase in dwelling time from 277 to 299 days. We could not establish a significant difference between patients with solid tumours and those with systemic malignancies. The incidence of infection was 0.67, that of irreversible occlusions 0.31 and of port-defects 0.26/1000 catheter days. The bacterial port-colonisation and infection was mainly caused by Staphylococcus epidermidis. Occlusions were removed with the help of Urokinase and the defective systems were totally removed.

Summary: The treatment and management of pediatric malignancies have been made easier with the introduction of the Port-Systems; these devices have been accepted both by the patients and their parents. However, in order to avoid complications and thus guarantee long life, special catheter care should a prerequisite. The co-operation and education of the parents play a pivotal role in regards the handling of the systems. Our results show that intravascular devices can be used throughout the whole therapy period, even days/weeks/months after therapy, without complications.

568 POSTER

External beam radiation (EBRT) for retinoblastoma (RB): The Hospital Vall d'Hebron experience and review of literature

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Purpose: To analyze treatment results and patterns of failure following EBRT for RB in our institution and to compare them with previously published data.

Methods: 29 patients with RB received EBRT between January 1985 and December 1995. The mean age was 14 m. (6 m.–4 y.). 36 eyes were treated, 32 with a preservation finality. Reese-Elsworth (R-E) staging of these 32 eyes was as follows: group I, n = 5; group II, n = 9; group III, n = 11; group IV, n = 2; group V, n = 5. EBRT doses ranged from 30–50 Gy (mean 39.8 Gy) in 200 cGy fractions, 5 day/week. 10 patients were treated with an anterior electron beam technique, 16 with 2 lateral opposed photon beam technique, 6 with an oblique photon beam field and 4 with a single lateral photon field.

Results: 29/36 (80%) patients had a response after EBRT. Eye preservation rates in the R-E groups I to V were: 5/5, 8/9, 9/11, 2/3 and 5/8 respectively. 13/36 (36%) developed a local failure, 8 were local relapses and 5 were new tumors in previously uninvolved retina. Cataract formation rate was 100% with the anterior techniques but 55% with the lens sparing lateral field techniques.

Conclusion: EBRT provides adequate tumor control in retinoblastoma eyes with an acceptable toxicity. Our results are compared with previously reported data.

569 POSTER

Postoperative treatment of medulloblastoma in childhood 1986–1996

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Purpose: Evaluation of medulloblastoma patients who received radiotherapy and adjuvant chemotherapy after surgical resection.

Materials and Methods: Postoperative treatment was performed in 52 children with posterior fossa tumour whose median age was 7.5 years (range 2–17) based on administration of Vincristine, DBD, Natulan, MTX, VP-16 and Cysplatin immediately after surgery.

The radiotherapy started 6 weeks after the tumour resection: 30 Gy craniospinal irradiation in low risk cases and 35 Gy in high risk cases. The boost dose was 20 Gy to the posterior fossa. The treatment was performed with linear accelerator using 6 or 9 MV X-ray beams.

Results: The median survival rate was 35 months, ranged 3–117 months. The recidiva free median survival rate was 25 months. 19 patients died (36.5%). The cause of death was 8 local recurrence, by 8 metastases within the central nervous system and by 3 intercurrent disease.

Local recurrence was observed in 12 patients: 8 died, 4 are living with complains.

33 patients are living: 15 (45%) with normal activity, 10 (31%) with mild complain and 8 (24%) with severe complains.

Conclusion: The correctly planed and achieved radiotherapy with adequate doses can avert the local recurrence and the metastases within the central nervous system.

570 PUBLICATION

Childhood cancer mortality in Austria, 1980–1992

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Purpose: To describe trend analyses focusing on cancer mortality among children aged 0–14 in Austria covering the period 1980–1992.

Methods: The data were abstraced from the official Austrian mortality statistics of the years 1980–1992. Age-specific and age-standardized rates were calculated. Time trends were tested for significance with Spearman's rank correlation test. Sex specific rates were calculated but showed no significant differences.

Results: In the period 1980–1992 cancer was the cause of death of 718 children. Leukaemia was responsible for 34.5% of all childhood cancer deaths and 1.5% of all childhood deaths. The mortality of all malignant neoplasms in children decreased significantly ($p = 0.0004$) from 54.8 per million in 1980 to 33.9 per million in 1992. This trend is mainly due to the reduction in mortlaity of leukaemia (from 22.3 per million in 1980 to 13.1 per million in 1992).

Conclusions: The decreasing trends are primarily caused by better diagnostic and therapeutic procedures and most probably not by decreasing incidence. New concepts of treating childhood leukaemia have resulted in decreasing mortality rates. For the small number of cases observed among the other causes of cancer deaths it is not possible to prove a similar influence of these modern measures, but it can not be excluded.