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.

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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.

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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.