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## Treatment of non metastatic peripheral Primitive Neuroectodermal Tumour (PNET)/Extra osseous Ewing's sarcoma (EOES): experience in the SIOP MMT 89 study

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**Objectives:** To report the outcome of treatment for non metastatic PNET/EOES treated in the SIOP MMT 89 study.

**Patients & treatment:** PNET/EOES = 42/199 (22%) of all patients with non Rhabdomyosarcoma MMT registered and histologically reviewed in MMT 89.

32 (76%) were non metastatic. M:F ratio 1.9:1. Median age 9 yrs (3 months - 16.8 yrs). Median follow up 7 years. Localised, complete primary excision (Stage I pT1, pT2) received chemotherapy (CT) with VA x 2; Localised, incomplete initial excision (Stage I, II pT3a (microscopic residual), pT3bc (macroscopic residual) received IVA x 6; Stage III (node positive) received intensified "6 drug" CT. Radiotherapy (RT) was given only for residual disease after initial CT ± second surgery. Sites of disease were limbs (13), trunk/walls (12), head & neck (7).

**Results:** Five year overall (OS) and event free (EFS) survival were 59% (42-76) and 44% (27-61). Initial causes of failure were: no CR (5 = 16%), local relapse (5 = 16%) or metastatic relapse (9 = 28%). Seventeen patients were alive in 1st or 2nd+ CR of whom 9 had been treated with CT alone (1) or CT + conservative surgery (8), 7 had received RT and 1 radical surgery (amputation). Size of primary had a significant effect on 5 year EFS (86% if T < 5cm (n = 7) vs. 30% if T > 5 cm (n = 23) p = 0.05).

**Conclusions:** Outcome for non metastatic patients with PNET/EOES is less favourable than for RMS (5 year OS 71%). The most common cause of failure was metastatic relapse (first event in 28% vs. 6% in RMS). Tumour size seems more important to prognosis than in RMS.

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## Synovial sarcoma in children and adolescents: experience of the International Society of Paediatric Oncology (SIOP)

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**Objective:** To review outcome for non metastatic patients with a diagnosis of Synovial Sarcoma (SS) confirmed by central pathology review, treated in SIOP MMT 84 & 89 studies (1989-1995).

**Patients & Methods:** Forty patients with median age 12 yr (3m - 16 yr). M:F 1.9:1. Primary site was Limbs (30 = 75%) (lower limb 22/30); Trunk/Walls (7); Head & Neck (3). TNM Clinical Stage I (23), II (17), III (0). Post surgical stage pT1, pT2 (9); pT3a (12), pT3b(18). All patients received chemotherapy - Stage pT1, pT2 (complete primary resection) received IVA (Ifosfamide, Vincristine, Actinomycin D)x 3 in MMT 84 and VA x 2 in MMT 89; patients with incomplete primary surgical resection (pT3ab) received IVA x 6-10 in MMT 84 & 89 (to a maximum cumulative Ifosfamide dose = 60 g/m2). Local therapy (second surgery +/- RT) was given to patients not achieving complete remission with primary surgery +/- chemotherapy.

**Results:** 5 year overall and event free survival was 87% (73-94) and 69% (58-81) at a median follow up of 98 (3-149) months. Multivariate analysis revealed Clinical Stage (p=0.01) as the only significant prognostic factor. Twenty five patients (62%) are alive in 1st CR. Progressive disease occurred in two patients, both died. Thirteen patients relapsed (local 9, local and metastatic 1, metastatic 3). Overall 11/33 long term survivors (defined as follow up > 3 yr from last event) received RT and 2 required amputation.

**Conclusion:** Non metastatic SS has an excellent prognosis. Most patients can be cured with chemotherapy and conservative local therapy. Relapse was predominantly local and chemotherapy appears to offer a role in improving local control and reducing the risk of distant metastases.

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## Hemithorax irradiation in Ewing tumors of the chest wall

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**Purpose:** In the CESS 86 and the EICESS 92 trials, hemithorax irradiation was performed in patients with Ewing tumors of the chest wall involving the pleura or contaminating the pleural cavity. The results of these patients were evaluated and compared to patients with chest wall tumors who did not receive hemithorax irradiation.

**Methods:** Between 1985 and 1996, 138 patients presented with a non metastatic Ewing tumor of the chest wall. They were treated in a multimodal treatment regimen including polychemotherapy, surgery and/or radiotherapy depending on the tumor characteristics. Hemithorax irradiation was performed with 15 Gy for patients < 14 years and with 20 Gy for patients > 14 years. 42 patients received hemithorax irradiation (group 1), 86 patients did not (group 2). In 10 patient, no sufficient data was available.

**Results:** Comparing both groups, initial pleural effusion, pleural infiltration and intraoperative contamination of the pleural space were significantly more frequent in group 1. Event free survival after 7 years was 63% for patients in group 1 and 46% for patients in group 2 (n.s.). 7 year local relapse rates including combined relapses were 12% in group 1 and 10% in group 2. The corresponding systemic relapse rates were 22% vs. 39%.

**Conclusion:** In the unfavorable subgroup of patients who received hemithorax irradiation, there is a non significant improvement in EFS due to reduced systemic relapses. Local control is equivalent.

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## Prognostic factors in patients with localised tumor of the Ewing family (ET). Update of the EW88 study of the French Society of Pediatric Oncology (SFOP)

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**Purpose:** 1) To improve survival in patients with ET using semi-continuous chemotherapy (CT) and performing resection of the primary, as often as possible, 2) To identify prognostic factors.

**Patients and methods:** 141 patients with localised tumour entered the trial between 01.88 and 12.91. Induction CT consisted of 5 courses of Cyclophosphamide, 150 mg/m2 x 7 days, followed by Doxorubicin, 35 mg/m2 IV on day 8. Surgery was recommended. The delivery and the doses of radiation therapy (RT) was based on the quality of resection and the histological response to CT. Maintenance CT consisted of vincristine + actinomycin and cyclophosphamide + doxorubicin.

**Results:** After a median FU of 8.5 years, the OS at 5 years was 66% and DFS was 58%. In patients treated by surgery, the only prognostic factor was histological response to CT: DFS was 75% for good responders (< 5% of residual cells), 48% for intermediate responders and only 20% for poor responders (> 30% of cells) p < 0.0001.The tumour volume by itself had no influence on DFS in these patients. In contrast, it had a strong impact on DFS in patients treated by RT alone. Age had no impact on outcome.

**Conclusion:** Trials for localised ET should be based on the histological response to chemotherapy or on the tumour volume according to the modality of local therapy.

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## Management of ewing's family of tumors of the chest wall (EFTCW) in childhood: a twenty-year single-institutional experience with 41 consecutive patients

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**Introduction:** In the study period 1975-1995 we treated 41 consecutive children with EFTCW at onset. Despite an improvement of the diagnostic