

## Lectures (L)

### L1 Unscrambling the genomic chaos in osteosarcomas

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### L2 Genomic analysis of soft tissue tumors

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Human neoplasms display a myriad of acquired genetic alterations. The types and patterns of mutation are highly variable and heterogeneous among different tumor entities, ranging from ploidy shifts to single-base substitutions and epigenetic alterations. Soft tissue tumors are no exception – genetic analyses have shown that the clinical and biological variation among these neoplasms is reflected in their genotypes. The malignant tumors, the sarcomas, may be broadly dichotomized into one group characterized by specific gene fusions and one group typically showing massive rearrangements leading to recurrent, but non-specific, gains and losses; several target genes within these regions have been identified. It has been shown convincingly that the benign soft tissue tumors are as genetically heterogeneous as the sarcomas, but they usually have near-diploid chromosome counts with only a few structural and/or numerical aberrations, distinguishing them from many of their malignant counterparts. Also for a growing number of the benign lesions, the molecular consequences of the chromosomal rearrangements have been clarified.

Genetic studies of benign and malignant soft tissue tumors not only have provided cell biologists with decisive information on genes that are essential for tumor development, but they have also supplied clinicians involved in the management of these patients with valuable biomarkers. The current presentation will provide some examples of how the information on genomic changes in soft tissue tumors can be used for clinical purposes.

### **L3 CircSarc-Circulating Tumor DNA in sarcoma**

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### **L4 Searching for fusion genes – A 20-year Odyssey**

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### **L5 CINSARC and more.....**

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The metastatic spreading of cancers, and sarcomas in particular, remains the major pejorative factor for patient management. Until now, estimation of metastatic risk in sarcoma has been based essentially on histological and clinical criteria. At the clinical level, factors deteriorating prognosis are essentially penetration and invasion of neighboring tissues. Subsequently, tumor prognosis is clarified during the histological study of tumors with the FNCLCC histological grading (Trojani et al, 1984). Since the beginning of the 2000s, with the evolution of high throughput technologies for genome and transcriptome characterization (Array-CGH and expression-array), molecular signatures have successively been identified in various tumor types, starting with breast cancer (Van de Veer et al, 2002; Van de Vijver et al, 2002). In our published study (Chibon et al, Nat. Med 2010), using an original approach based on biological mechanisms of the tumors, we were able to identify a prognostic signature. The Gene Ontology analysis of the 67 genes showed that all the annotated genes were involved in the same biologic processes: the control of mitosis and chromosome integrity. This signature was then named CINSARC for Complexity INDEX in SARComas and successfully applied to GISTs (Lagarde et al, Clin Can Res 2012; Lartigue et al, Eur J Can 2015) and synovial sarcomas (Lagarde et al, JCO 2013; Chakiba et al, Ann Oncol 2014).

CINSARC and GI signatures are now validated enough to go further to clinical application.

To this goal we performed RNAseq analysis of 150 sarcomas to validate the CINSARC prognostic value by this technology (with FFPE samples).

Our goals are now to identify the genetic determinism of CINSARC expression and to understand the mechanism linking CINSARC expression and metastatic spreading. New findings in these topics will be presented.

## **L6 Recent advances in Forme Fruste sarcomas: synovial sarcoma, myxoid liposarcoma, epitheloid sarcoma and clear cell sarcoma**

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## **L7 NoSarcC**

*O. Myklebost*

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## **L8 FET oncogene carrying tumors, molecular biology, diagnostics and clinical trials**

*P. Åman*

Gothenburg University Hospital , Sweden

## **L9 Lytic peptides in human sarcoma treatment**

*Ø. Rekdahl*

University of Tromsø, Norway

## **L10 The microenvironment of sarcoma as sources of therapeutic targets**

*D. Heymann*

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Except for Ewing's sarcoma, which involves a fusion protein playing an oncogenic role, or specific inherited syndromes (*e.g.*, *p53*, *Rb*), the causes of most bone sarcomas are not well known. Most people with osteosarcoma do not have any specific risk factors and the etiology of osteosarcoma is now based mainly on the 'seed and soil' theory proposed by Stephen Paget at the end of the 19<sup>th</sup> century. This theory has led to the concept of a « niche » which is a specialized microenvironment promoting the emergence of tumor stem cells and providing all the required factors for their development. A vicious cycle is established between the bone microenvironment and cancer cells during the progression of bone metastases and primary bone tumors as well. This concept was initially described for normal hemopoiesis that is sustained by the local osteoblastic niche. Although the bone niche plays a key function in tumor growth, the bone microenvironment contributes to the enhanced resistance of tumors to therapy and can sustain cancer cells in a quiescent state. The bone tumor microenvironment is then described as a sanctuary that contributes to the drug resistance patterns and may control at least in part the tumour growth. Based on these observations, tumour microenvironment constitutes a key source of potential new therapeutic targets (bone cells, immune cells, blood vessels, etc).

Ref: INSERM, UMR 957, "Pathophysiology of Bone Resorption and Therapy of Primary Bone Tumours", Equipe Ligue Contre le Cancer 2012, University of Nantes, France

## **L11 Sirtuins as therapeutic targets for sarcoma**

*B. Brodin*

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## **L12 Optimal treatment of desmoid tumors**

*L. Jeys*

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## **L13 Giant cell tumors: imaging features in the era of denosumab**

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**Background:** The imaging features of giant cell tumor of bone (GCTB) are well known. The initial diagnosis of GCTB in the extremities is often made on radiographs, showing a geographic osteolytic destruction without sclerotic borders, often adjacent to the articular surface of the knee or in the proximal humerus. Computed tomography (CT) and magnetic resonance imaging (MRI) mostly serves as staging modalities, to evaluate possible cortical destruction and possible soft tissue extension. MRI often shows a typical signal pattern characterized by low to intermediate signal intensity at T1- and T2-weighted sequences in the majority of cases. Denosumab has been used as neoadjuvant treatment for tumors where joint-sparing surgery is considered hazardous, and for patients with inoperable or disseminated GCTB. After denosumab treatment there usually is a dramatic change in appearance of the tumor.

**Methods:** 7 patients with GCTB treated with neoadjuvant denosumab have been evaluated by clinical findings, radiology, surgery and histopathology.

**Results:** Tumor pain disappeared rapidly. There was marked reduction of the osteolytic components in all patients, with new bone formation, osteosclerosis and sclerotic rim formation observed with radiography and CT. Devitalization of large portions of the tumors was seen with MRI with tumor tissue being replaced by bone.

**Interpretation:** With denosumab there is a dramatic change in tumor appearance with all imaging modalities, with clearer delineation of tumor boundaries on radiography and CT, but not always with distinct boundaries between new bone formation and remaining viable tumor.

## **L14 Chordoma versus benign notochordal cell tumor – radiological aspects**

*I. Taksdal*

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Benign Notochordal Cell Tumour, BNCT, was introduced as a new diagnosis in the 2013 edition of the WHO Classification of Tumours of Soft Tissue and Bone. It is defined as a benign tumor showing notochordal differentiation. The incidence is uncertain, its origin is controversial, and the relation of BNCT to its malignant counterpart, chordoma, is not yet clear.

From the radiologist's point of view, chordoma usually presents in a symptomatic, middle-aged or elderly patient as a midline lesion, most commonly in the spheno-occipital or sacro-coccygeal region. It shows bone destruction on CT, a soft-tissue component on MR, low signal on T1, very high signal on water sensitive sequences and some contrast enhancement. BNCT is easily detected on MR with signal and location like chordoma, only it does not enhance. It is usually an incidental MR finding in an asymptomatic person. BNCT does not destruct bone and may be invisible or slightly sclerotic on CT.

In an otherwise healthy individual, a central bone marrow lesion in the sacrum is often interpreted as an atypical hemangioma, without visible fat. Hemangiomas are frequent incidentalomas and probably many BNCTs are not even noticed or described. The threshold for biopsy is high in an asymptomatic person.

In a patient with known malignancy, e.g. prostatic cancer, BNCT may be interpreted as metastasis. In a cancer patient, a midline single bone lesion which fulfils the radiological criteria for BNCT may warrant a biopsy to avoid overstaging.

Knowledge of this new entity is important both for radiologists and clinicians. Examples of notochordal cell tumors and their differential diagnoses will be presented.

## **L15 Differentiating lipomatous tumors with MRI-technique**

*M. Skorpil*

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## **L16 Current concepts and future perspectives for surgical treatment of visceral and retroperitoneal sarcomas**

*A. Gronchi*

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## **L17 Current concepts and future perspectives for surgical treatment of GIST**

*J. Åhlén*

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## **L18 Radiotherapy for retroperitoneal sarcomas: how and when and for whom?**

*R. Haas*

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In this presentation 3 issues will be addressed as to the role of radiotherapy (RT) in retroperitoneal sarcomas (RPS); the “how”, the “when” and the “for whom” questions.

### ***How?***

Most experience in irradiating RPS patients is derived from papers addressing external photon beam regimens from linear accelerators. Nevertheless, in this presentation boost techniques, tomotherapy, brachytherapy and proton beam RT will also be addressed.

### ***When?***

Although in sarcoma patients, in general, RT can be applied on indication both before and after surgery, in this presentation, from a toxicity point of view, arguments for preoperative RT are made.

### ***For whom?***

Up to now, the knowledge on RT in RPS patients is derived from retrospective series, be it that some of them are indeed very large. The ultimate proof of its value will only be produced as soon as the now successfully accruing EORTC 62092 – 22092 “STRASS” study is published. The trial prospectively randomizes the standard extensive surgery alone versus the same surgery preceded by neoadjuvant RT.

## **Denosumab in giant cell tumors L19-21**

### **L19 Pathology**

*P. Rissler*

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### **L20 Experiences of implementing denosumab in the treatment of Giant Cell Tumor of bone**

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**Background.** Giant cell tumor of bone (GCTB) is a rare, osteolytic tumor which causes severe bone destruction, sometimes with soft tissue extension and most often located in the distal or proximal parts of long bones. It is considered a benign tumor but still metastasizes in 1-5% of cases. Primary surgery with curettage is the mainstay of treatment, but is not always possible due to location and extension of the lesion. Local recurrence is dependent on type of surgery, location and maybe to the use of local adjuvant treatment. Denosumab, a human monoclonal antibody against the tumor-driving pathway of RANK-RANK ligand is a potent inhibitor of the osteolytic activity of the tumor. It has been used as neoadjuvant treatment for tumors where surgery with joint-sparing intention is considered hazardous, and for patients with inoperable or disseminated GCTB .

**Methods.** With a back-drop of current knowledge of denosumab use in GCTB 7 patients, who had neoadjuvant treatment, have been clinically, radiologically, surgically and histopathologically evaluated.

**Results.** All patients responded well with prompt regress of symptoms, reduction of lytic components and new bone formation observed with radiography and MRI. Surgery was performed without problems from soft-tissue components, with remarkably less bleeding but without clear demarcation of the tumor.



Histopathology revealed disappearance of giant cells but the spindle-/tumor cells remained.

**Interpretation.** Denosumab was very effective with respect to clinical symptoms and down-staging of soft tissue components. Surgery was performed without complications, but joint-sparing treatment was not always possible. However, although pathology confirms a less lytic tumor the spindle cells remain. The consequences of these findings will be discussed.

## **L21 Surgery**

*F. Vult von Steyern*

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## **L22 Challenges for new trials in osteosarcoma and Ewing sarcoma**

*J. Whelan*

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## **L23 Orthopaedic challenges for osteosarcomas and Ewing sarcomas**

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The Orthopedic challenges for Osteosarcoma (OS) and Ewings sarcoma (ES) are quite different. In OS the place for surgical excision in the oncological treatment is unchallenged whereas in ES the place for surgery is not as clear.

In conventional OS, i. e. around the knee, proximal femur and humerus local control can be achieved in approximately 90 % of patients with local excision. Reconstruction based on modular megaprosthesis give long term good functional results. The challenge is to develop even better prosthetic systems without jeopardizing the good results already achieved. Osteosarcoma in older patients, often arising in more uncommon locations, has gained more attention in later years. As we realize that these patients also benefit and can tolerate neo-adjuvant chemotherapy, indications for surgical treatment also increase. Here we need to gather results of surgical

treatment and reconstructions from a large number of institutions to get any meaningful information.

In ES the value of surgical excision in pelvic locations has been challenged. There are no randomized trials but there appears to be little evidence that surgery improves outcome as an adjunct to chemotherapy and radiotherapy. We must therefore question whether there is a place for surgery in other sites as well. Clearly, the functional results of radiotherapy as opposed to surgery is much better in the majority of locations. However, we know little about the risks of radiotherapy in terms of growth deformity, secondary neoplasms, stress fractures, and late local recurrences. On the other hand even when we think that a wide surgical margin can be attained, pathology is often disappointing, showing areas of viable tumor close to the margins, necessitating radiotherapy postoperatively anyway. Would such patients fare better with only radiotherapy? Here we need smart clinical trials from a large number of institutions to answer these questions.

## **L24 Computer navigation of pelvine tumors**

*L. Jeys*

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## **L25 Challenges in today's radiology in sarcoma diagnosis, staging and therapy response**

*D. Vanel*

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## **L26 Findings from the postmarketing adult osteosarcoma surveillance study in the Nordic countries**

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**Background and purpose:** A 10-year postmarketing drug surveillance study was initiated in 2004 to evaluate a potential association between teriparatide treatment and osteosarcoma in humans. The objectives were to

identify incident cases of adult osteosarcoma and determine if patients had a prior history of teriparatide treatment and to collect data on other potential risk factors for osteosarcoma. Patients were identified in Denmark, Finland, Iceland, Norway, and Sweden through collaboration with the Scandinavian Sarcoma Group (SSG) registry; data collection activities concluded 31Dec2013.

**Methods:** Incident cases of histologically confirmed primary osteosarcoma diagnosed from January 2004 through December 2013 in adults aged 40+ years were identified through the SSG registry and the Finnish and Swedish National Cancer Registries. Following patient consent, demographic information, treatment history, and lifestyle and occupational exposures were abstracted from the patient's medical record.

**Results:** 112 patient medical records were abstracted. None of these patients had a record of teriparatide use. Most of the patients were men (56%), and the mean age at diagnosis was 60 years. The most commonly coded histologies were osteosarcoma not otherwise specified (84%) and chondroblastic osteosarcoma (13%), and the most common tumor site was the leg bones (46%). Possible pre-existing risk factors for osteosarcoma were radiation (22%) and injury or infection at tumor site (11%). Site or region of radiation treatment and tumor matched for 84% of radiation cases.

**Interpretation:** Despite study size limitations, no association between teriparatide and osteosarcoma was detected. The study provides a population-based characterization of osteosarcoma in adults.

## **L27 Nordic studies on second malignant neoplasm (SMN) after childhood**

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**Background and purpose:** 5-year survival in childhood cancer has increased from 20% in the late 1960ies to 80% today. Development of a second malignant neoplasm (SMN) is one of the most serious of the several late treatment complications that may occur.

**Methods:** There are several Nordic studies on SMN. A first cohort study (Olsen et al. 1993) included 30 880 persons under the age of 20 with a first malignant neoplasm diagnosed during the period of 1943 – 1987. A second study (Olsen et al. 2009) expanded the cohort to 47 697 persons aged 0 – 19 years with a first malignant neoplasm diagnosed during 1943 – 2005. Another study (Sankila et al. 1996) assessed the risk of developing SMN among 1 641 persons, diagnosed with Mb. Hodgkin before 20 years of age. In 2 case-control studies (Garwicz et al. 2000 and Svahn-Tapper et al. 2006) the risk of developing an SMN was correlated to the treatment given for childhood cancer.

**Results:** The relative risk of developing an SMN was around 3 – 4. It was increased in all age groups even for cohort members approaching 70 years of age. The age-specific incidence rates were highest for cohort members treated in the era of intensive, multiple-agent chemotherapy (1975-2005). Patients with Hodgkin’s lymphoma had a much higher risk (8). The risk of developing an SMN was above all related to radiation therapy whereas chemotherapy greatly increased the risk when given together with radiation therapy. Hereditary factors were important for the occurrence of SMN, independent of different kinds of therapy.

**Interpretation:** Long-term survivors of childhood cancer have a persistent excess risk for a second primary cancer throughout their lives. Radiation therapy is the main causative factor of these second malignant neoplasms. Appropriate surveillance programs are needed.

**References:** Olsen et al. *BMJ*. 1993 Oct 23;307(6911):1030-6.  
Olsen et al. *J Natl Cancer Inst*. 2009 Jun 3;101(11):806-13.  
Sankila et al. *J Clin Oncol*. 1996 May;14(5):1442-6.  
Garwicz et al. *Int J Cancer*. 2000 Nov 15;88(4):672-8.  
Svahn-Tapper et al. *Acta Oncol*. 2006;45(4):438-48.

## **L28 Hypoxia induced gene-profile in sarcoma patients: A new prognostic marker**

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**Background and purpose:** The prognosis of soft tissue sarcoma (STS) patients has not changed in decades and new treatment modalities are needed. It is known that tumor-hypoxia is important in tumor progression and resistance to treatment for several cancers. However, little attention has been paid to the role of hypoxia in sarcomas. We investigated the prognostic value of a hypoxia induced gene-profile in sarcoma patients.

**Patients and methods:** A validated hypoxia-induced gene-profile of head and neck cancer was explored by RT-qPCR in diagnostic biopsies or resected specimens from 74 patients diagnosed with localized high grade STS in the period 1990-2008. The patients were allocated to 2 groups: a more hypoxic and a less hypoxic group, according to the median rank of the gene-profile. The primary endpoint was disease specific mortality estimated using Kaplan-Meier and the proportional hazard model. Adjustments were made for comorbidity and stage.

**Results:** The 5-year disease specific mortality was 25% (95%CI: 13-43) for patients with less hypoxic tumours compared to 44% (29-62) for patients with more hypoxic tumours. The adjusted Hazard ratio (HR) was 2.3 (1.01-5.1). Radiation treated patients allocated to the group of more hypoxic tumours (n=18) had a significantly higher disease specific mortality compared to radiation treated patients allocated to the less hypoxic group (n=19). HR: 4.2 (1.3-15).

**Interpretation:** This hypoxia-induced gene-profile may have prognostic value in STS patients and it may facilitate improvements in the choice of treatment modality for patients with hypoxic tumors.

## **L29 Low-grade and dedifferentiated osteosarcoma in Norway 1975-2009**

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**Background:** Despite osteosarcoma (OS) being the most common primary malignant bone tumor, it is a rare disorder with a great span in tumor biology and prognosis as documented in 3 previous reports from Norway[1-3]. The present study describes clinical characteristics and treatment outcome of low-grade and dedifferentiated OS (DOS).

**Method:** A nationwide cohort comprising all histologically verified low-grade and DOS between 1975 and 2009, based on registry sources supplemented with clinical reports from all hospitals involved in sarcoma management[1].

**Results:** 54 patients were identified; 12 were DOS. The incidence for all patients was bimodally distributed by age, with a dominant peak in the 20ies and a smaller one in the 6ties. 3 patients had primary metastatic disease, whereas 7 developed metastases during follow up and 14 experienced local relapses, respectively. Patients with DOS dominated the group with metastatic relapse (6 cases). 5-year sarcoma specific survival was 91 %, with no improvement over time. Wide surgical margin after resection of primary tumor had positive impact on survival. As expected both local relapse and metastases during follow-up were associated with a poor outcome. Chemotherapy and radiotherapy predicted also poor outcome in the analyses, due to the selection bias of patients in need for such treatment. Neither higher age nor axial tumor localization were associated with a poor outcome.

**Interpretation:** Low-grade OS has excellent prognosis when surgically resected with a wide margin. However, it has the potential to dedifferentiate and metastasize with a dismal outcome.

1. Berner K, Johannesen TB, Berner A, Haugland HK, Bjerkeheggen B, Bohler PJ, Bruland OS. Time-trends on incidence and survival in a nationwide and unselected cohort of patients with skeletal osteosarcoma. *Acta Oncol.* 2015 Jun 24;54:25-33.

2. Berner K, Hall KS, Monge OR, Weedon-Fekjaer H, Zaikova O, Bruland OS. Prognostic factors and treatment results of high-grade osteosarcoma in Norway: a scope beyond the "classical" patient. *Sarcoma*. 2015;2015:516843.
3. Berner K, Bjerkehagen B, Bruland OS, Berner A. Extraskelletal osteosarcoma in Norway, between 1975 and 2009, and a brief review of the literature. *Anticancer Research*. 2015. In press.

### **L30 Risk stratification of patients with malignant peripheral nerve sheath tumors and identification of new treatment options**

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**Background and purpose:** The aggressive malignant peripheral nerve sheath tumor (MPNST) is a soft tissue cancer with high mortality and no standardized therapy beyond surgery. Almost half of the patients have a predisposing genetic syndrome, neurofibromatosis type 1 (NF1). The purposes of our studies were to identify molecular markers that distinguish high- from low-risk tumors, thus selecting patients who would qualify for more aggressive treatment, and to identify new strategies for treatment.

**Methods:** 30 fresh frozen MPNST patient samples and 5 MPNST cell lines were submitted to gene expression analyses. 63 formalin fixed MPNST samples were analyzed for protein expression by immunohistochemistry *in situ*, and the 5 cell lines were submitted to high-throughput cell viability and cytotoxicity screen of a panel of 310 different oncology drugs.

**Results:** Based on gene expression and protein expression data, we identified a 3-protein profile, BIRC5 (survivin), TK1 (thymidine kinase 1) and TOP2A (topoisomerase 2a), that clearly stratified the patients into poor and good outcome groups, independent of NF1 status and surgical remission status [1].

From the *in vitro* drug screen, we confirmed that the BIRC5 inhibitor YM155 was among the most potent cytotoxic drugs and that the TOP2A inhibitor doxorubicin had a cytostatic effect. In addition, several drugs that interfere with thymidine metabolism had a strong cytotoxic and/or cytostatic effect.

**Interpretation:** The 3 proteins BIRC5, TK1, and TOP2A, are all prognostic markers for MPNST patients and druggable targets. Further preclinical studies validating these targets for clinical use are warranted.

[1] Kolberg, M. *et al.* Protein expression of BIRC5, TK1, and TOP2A in malignant peripheral nerve sheath tumours – A prognostic test after surgical resection. *Mol. Oncol. in press* 2015

### **L31 Megaprosthesis reconstruction for chondrosarcoma of the limbs: Evaluation of the functional and oncological outcome**

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**Background:** Wide tumor resection is the treatment of choice in high- and intermediate-grade chondrosarcoma. However, high local recurrence rates and metastasis are common, despite aggressive surgery.

**Methods:** Between 1992 and 2006 we treated 78 patients with chondrosarcoma of the limb with megaprotheses. Mean age was 55 years. Most frequent tumor sites were distal- (n=24) and proximal femur (n=24). 17 patients had a grade I (GI), 42 a GII, 7 a GIII, and 12 a de-differentiated chondrosarcoma. Median follow-up was 62 months.

**Results:** Overall survival was significantly linked to the tumor grade. 21 patients died of disease. Despite 91% wide resection margins, local recurrence rate was 8% for all tumors, highest (42%) for high-grade chondrosarcoma. 26% developed metastases irrespective of the resection margins. Adjuvant therapy was given to 13 patients, without any effect on the overall prognosis.



Mean MSTS score was good. The main prosthesis related complications were infection (n=4) and aseptic loosening (n=3). 12 % required surgical revision due to implant related complications and 23% due to oncological complications. Secondary amputation was necessary in 9 patients (8 oncological complications, 1 infection). Prosthetic survival at the latest follow up was 77 % for the primary implant.

**Interpretation:** Endoprosthetic replacement of limb chondrosarcoma provides good functional outcome, low rates of implant related complications, and a long prosthetic survival.

However, in absence of effective adjuvant therapies, even wide tumor resection results in high rates of local recurrences and metastatic disease in high grade chondrosarcoma compared to other primary bone sarcoma.

### **L32 RESPECT – Rehabilitation including social, physical activity and education in children and teenagers with cancer**

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Background and purpose: Cancer treatment reduces children's quality of life and impairs their academic and physical performances. Accordingly, there is a need for interventions that ameliorate the children's inability to participate in their normal school, leisure activities, and interact with peers during treatment. We examined the feasibility of a multimodal intervention rehabilitation program including i) an age adjusted educational program on childhood cancer targeting the child with cancer, classmates, and schoolteachers; ii) selection of 2 classmates as ambassadors, who bi-weekly visit the child when hospitalized; and iii) participation in individual and joint (patients and ambassadors) hospital based physical activity programs. Patient and methods: The first 30 consecutive children aged 6-18 years newly diagnosed with cancer enrolled in the intervention group are included in this feasibility study.

Results: 30 of 31 eligible patients consented to participate. Median time from diagnosis to inclusion was 6 days (75% range:0-13). The educational program (30/30) was performed within a median of 14 days (75% range:6-39). Median time for ambassador assignments was 9 days thereafter (75% range:5-20). In average one third of all classmates (1-14 applications) applied to become an ambassador. Median travel distance was 49 km (75%

range:14-168, max:340 km). The median number of ambassador visits was 8. None of the patients or ambassadors discontinued study participation. Interpretation: The RESPECT study includes highly acceptable intervention components that are feasible for the children with cancer, their school classes and ambassadors.

### **L33 Teenagers losing a parent to cancer. Experience, modifiable risk factors and long-term outcome**

*T. Bylund Grenklo*  
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### **L34 Benefits and rationale for exercise during cancer therapy**

*M. Quist*  
Rigshospitalet, University of Copenhagen, Copenhagen. Denmark

In comparison with other areas of clinical medicine, exercise therapy has received comparably less attention in persons following a diagnosis of cancer. The precise reasons for this are unknown but likely stem from the prevailing dogma that a cancer diagnosis is associated with poor survival, a compromised immune system that may be more compromised by exercise training, and other debilitating side-effects that preclude participation in and benefit from exercise training. Nevertheless, the past two decades has witnessed a dramatic change in attitude with significant increased research and clinical interest in the role of exercise therapy following a cancer diagnosis.

The assumption is that a patient has already experienced the detrimental impact of the cancer treatment management plan and now is undergoing an exercise training program to restore / recover pre-treatment cardiorespiratory fitness levels. However, arguably a potentially more effective approach is to establish a 'prevention paradigm', in which patients undergo exercise training prior to or during therapy to mitigate and/or prevent therapy-induced toxicities.

This presentation will focus on the rationale, benefits, safety and efficacy of exercise training during cancer therapy in persons diagnosed with early and late stage cancer. Rationale and benefits will be described from both a physical and mental view.

## **L35 Sarcoma patient's advocacy groups in Europe**

*E. Lecoïnte*

Sarcoma Patients Euro Net (SPAEN)

## **L36 How and when to treat advanced GIST – with examples from real life in Eastern Germany**

*P. Reichardt*

HELIOS Klinikum Berlin-Buch, Department of Interdisciplinary Oncology, Sarcoma Center Berlin-Brandenburg, Berlin, Germany

Imatinib is considered the treatment of choice in all patients with locally advanced or metastatic GIST. Treatment should start immediately after the diagnosis. This also applies to patients who have undergone complete surgical removal of metastatic disease. A dose of 400 mg/day is considered standard for all patients except those with known exon 9 mutation, where 800 mg/day is recommended. Treatment with imatinib should not be terminated once started in metastatic disease, even in complete remission or after resection of residual disease. Surgical resection of remnant responding disease is an option that can be considered on an individualized basis. Survival estimates for patients with advanced GIST do not exceed 20% after 10 years.

After failure of or intolerance to imatinib, sunitinib, a multifunctional tyrosine kinase inhibitor is considered standard of care based on superiority in progression-free survival in a randomized phase III-trial compared to placebo. Flexible individualized treatment schedules as well as treatment beyond progression in case of continued clinical benefit have been shown to be beneficial with respect to progression-free and overall survival. The oral multikinase inhibitor regorafenib demonstrated significant superiority in progression-free survival after failure of both imatinib and sunitinib in the GRID phase III trial and is the treatment of choice in third line. Finally, in patients having failed all available treatment options, imatinib rechallenge proved significantly superior to best supportive care alone.

## **L37 How and when and for how long to use chemotherapy – with examples from real life in Texas**

*B. Benjamin*

MD Anderson Hospital, Houston, Texas, USA

Chemotherapy for soft-tissue sarcomas has limited efficacy, and we continue to hope for breakthroughs. Nonetheless, optimization of existing regimens can result in cures of patients with metastatic disease, and it is important to prepare for success. When surgery can convert a responding patient to CR status with acceptable morbidity, we incorporate it into the treatment regimen. Even when cure is clearly not possible, disease control can be prolonged by maximizing the efficacy of each regimen, and that frequently means more than 6 cycles of chemotherapy; but it is also essential to know which patients need treatment and which patients should not get treated. Adriamycin, the most effective single agent overall, is limited by cumulative cardiac toxicity, but by administering the drug by prolonged (at least 48-hour) infusion, or by giving it in conjunction with dexrazoxane, cumulative doses of 600-2000 mg/m<sup>2</sup> can be administered with no more cardiac risk than with 400 mg/m<sup>2</sup> by short infusion. For most sarcomas, the combination of adriamycin and ifosfamide (AI) is clearly more effective than single-agent therapy and is our usual standard (and we add vincristine for small-cell histologies), but there are clearly exceptions. Patients older than 65, those with renal insufficiency, and those with a single kidney do not tolerate AI. Patients with myxoid liposarcoma and those with vascular leiomyosarcomas can be treated as effectively with adriamycin and dacarbazine (ADIC), which we also use for many patients older than 65. We frequently use AI as initial chemotherapy for uterine leiomyosarcoma before gemcitabine-docetaxel (gem-tax). Since gem-tax is also effective in many sarcomas, we use it after initial AI in the majority of patients (but not in synovial sarcoma or myxoid liposarcoma), usually before relapse. Gem-tax is our preferred front-line regimen for epithelioid sarcoma, where we have seen prolonged remissions. It can also be used as initial chemotherapy for elderly patients who might not tolerate an adriamycin combination. When neuropathy or edema limits continued docetaxel, we continue with single-agent gemcitabine or add pazopanib. We have not had access to trabectedin for the majority of patients, so its addition to the armamentarium has been limited by protocol requirements. Examples of patients treated with some of these strategies will be presented.

## **L38 How and when and for whom to use targeted therapy – with examples from real life in Lombardia**

*S. Stacchiotti*

National Cancer Institute, Milan, Italy

## **L39 Scandinavian Sarcoma Group: Bone sarcoma trials**

*K. Sundby Hall<sup>1</sup>, S. Smeland<sup>2</sup>*

<sup>1,2</sup> Dept of Oncology, The Norwegian Radium hospital, Oslo University Hospital, Oslo, Norway. <sup>1</sup>Institute for Clinical Medicine, University of Oslo, Oslo, Norway

EURAMOS-1 was closed June 2011 and the results from the good-responder arm were published in 2015. With the current follow-up, EFS for MAPIfn was not superior to MAP alone. However, one quarter of the patients did not start Interferon, half of those who started terminated therapy early and follow-up continues. The data from the poor-responder arm were released June 2014 and showed that adding ifosfamide and etoposide to MAP is associated with additional morbidity and has no effect on survival. EURAMOS-1 does not support adaption of postoperative chemotherapy based on histological response.

The good-responders arm (MAP without Interferon) is the current standard treatment for resectable osteosarcoma utilized by SSG and the other participating groups in the EURAMOS collaboration.

EUROBOSS (bone sarcomas 40-65 y) was closed Dec. 2014. About 400 European patients including 64 SSG patients are recruited. Before response and toxicity data are available the treatment arm for good histological responders should be applied also for poor responders.

SSG participates (currently Norway and Denmark) in EURELOS, a register study with the aim of collecting detailed treatment and outcome data for recurring osteosarcoma. Regulatory applications for Sweden will be submitted soon.

Skåne University Hospital, Lund, is participating in a phase I/II trial testing Modufolin (an active metabolite to leucovorine), as rescue after high dose Methotrexate treatment for osteosarcoma. Other SSG centers may later join this study.

SSG is a partner of the Euro-Ewing Consortium and will participate in rEECur, a randomized study on second line chemotherapy in recurrent and primary refractory Ewing sarcoma for patients <50y. Some funding has been received from the European Union. Applications to regulatory authorities will soon be completed for Sweden and Denmark, whilst Norway and Finland are ready to start recruitment.

## **L40 Scandinavian Sarcoma Group: Soft tissue sarcoma trials**

*M. Eriksson*

Skane University Hospital, Dept of Oncology, Lund, Sweden

During 2014 SSG completed inclusion in our trial of adjuvant chemoradiotherapy in soft tissue sarcoma, SSG XX. This trial has been recruiting patients with soft tissue sarcomas in extremities or trunk wall and a high risk of recurrence defined by biological features. In total, 160 patients with primary surgery were included in Group A. Furthermore, 20 patients were enrolled in Group B; patients with risk of intralesional margins at primary surgery were given pre-operative chemoradiotherapy. The selection of patients and the use of interfoliated shortened radiotherapy between chemotherapy cycles are the two characteristics of the trial. The main analyses are expected next year.

GIST is the most common type of sarcoma, and after the SSG XVIII/AIO adjuvant trial, showing that three years of adjuvant imatinib in high risk patients was superior to one year, we have recently launched a new adjuvant trial, SSG XXII, which will compare three years with five years of therapy by randomizing patients after the standard treatment of three years to two additional years or nothing more.

SSG XXI, "PAGIST", is a trial of pazopanib in third line treatment for advanced GIST who has failed both imatinib and sunitinib. In October 2014 all planned 72 patients were enrolled, and the results are expected during the year. Finally, SSG will now together with AGITG and EORTC initiate a new trial, ALT-GIST, in first line advanced GIST comparing standard imatinib with alternating imatinib and regorafenib.

The described projects will be discussed

## **L41 Scandinavian Sarcoma Group: Recommendations for radiotherapy**

*N. L. Jebsen<sup>1</sup>, J. Engellau<sup>2</sup>, A. Safwat<sup>3</sup>, P. Nilsson<sup>4</sup>, J. Karlen<sup>5</sup>, K. Nilsson<sup>6</sup>, B. Söderer<sup>7</sup>, T. Hølmekbakk<sup>8</sup>, C. Trovik<sup>9</sup>*

<sup>1,9</sup>Haukeland University Hospital, Bergen, Norway; <sup>2,4</sup>Skåne University Hospital, Lund, Sweden; <sup>3</sup>Aarhus University Hospital, Aarhus, Denmark; <sup>5,7</sup>Karolinska University Hospital, Stockholm, Sweden; <sup>6</sup>University of Uppsala, Uppsala, Sweden; <sup>8</sup>Oslo University Hospital, Oslo, Norway.

**Background:** Radiotherapy (RT) is frequently indicated in treatment of bone and soft tissue sarcoma and may be administered pre- or postoperatively, as definitive treatment in radiosensitive sarcoma types, in cases of inoperable primary tumors or metastases causing local symptoms. Given the diversity of tumor localization and histological subtypes resulting in the heterogeneous biological features of sarcoma, different RT techniques, fractionation schedules and total doses will apply for various clinical scenarios.

In adjuvant treatment of extremity and trunk wall soft tissue sarcoma (ETSTS) in adults, the Scandinavian practice has been to give RT to doses typically lower than in other reported series. Nevertheless, local control rates in SSG are comparable to reported results from sarcoma centres outside Scandinavia. SSG guidelines for adjuvant RT in ETSTS in adults were initially outlined in the SSG XIII protocol of 1998. The succeeding SSG XX only holds recommendations for high-risk scenarios involving accelerated RT interposed with adjuvant doxorubicin and ifosfamide, and no comprehensive RT guideline for clinical settings outside the SSG XX have been formalized. In treatment of bone sarcoma and pediatric sarcoma, specific international or Italian/Scandinavian treatment protocols should be followed.

**Recommendations:** Based on Scandinavian practice and scientific reports, the aim of the project group was to present current recommendations for adjuvant RT in adult ETSTS of various risk categories (Table). RT guidelines for ongoing study protocols in pediatric STS or bone sarcoma used in Scandinavia are referred to. Additionally, general treatment prescriptions are presented.

Grade FNCLCC G1-3	Margin	Depth	Radiotherapy
G1	Wide	sc/deep	No
G1	Marginal	sc	No
G1	Marginal	deep	Consider RT
G2-3	Wide	sc	Consider RT
G2-3	Wide	deep	RT 50 Gy/25 fractions
G2-3	Marginal	sc/deep	RT 50 Gy/25 fractions
G1-3	Intralesional: micro/macro positive	sc/deep	RT 60 -70 Gy (2 Gy fractions)
G1-G3	Inoperable	sc/deep	68-74 Gy (2 Gy fractions)

Table. Recommendation for radiotherapy in patients with extremity and trunk wall soft tissue sarcoma.

## L42 Gastrointestinal stromal tumor (GIST) and surgery

*T. Hølmebakk*

Department of Abdominal and Paediatric Surgery, Oslo University Hospital, Oslo, Norway

The following issues will be presented and discussed: classification of GIST, diagnostic work-up, preoperative surgical considerations, intra-operative surgical considerations, surgery for metastatic disease.



## **L43 Medical treatment and research in GIST**

*M. Eriksson*

Skane University Hospital, Dept of Oncology, Lund, Sweden

Gastrointestinal stromal tumor (GIST) is the single most common sarcoma subtype, with most cases arising in the stomach or the small bowel. Since the introduction of tyrosin kinase inhibitors the prognosis for advanced GIST has improved dramatically. Imatinib is approved as the first line drug in advanced disease, benefitting about 85% of all patients. Sunitinib is approved in second line, as recently also regorafenib. The primary treatment for non-metastatic disease is surgery. Certain risk factors for recurrence after surgery have been identified: size, number of mitoses, location of the tumor (stomach with best prognosis), and tumor rupture during surgery.

To reduce the risk of recurrence, 3 large trials of adjuvant imatinib have been undertaken. SSG XVIII/AIO has been the most influential, showing that 3 years of adjuvant imatinib for high risk patients is better than 1 year, both in terms of progression-free and total survival. Based on these results, SSG now launches a new adjuvant trial to compare 3 with 5 years of treatment, SSG XXII.

SSG is also involved in trials in advanced GIST. The PAGIST study has enrolled 72 patients who had progressed on imatinib and sunitinib, and were given pazopanib in third line. Results are expected soon. Now, SSG in cooperation with AGITG and EORTC, will also launch a new trial for first line treatment of advanced non-operable GIST comparing the standard treatment of imatinib with an alternating regimen with imatinib and regorafenib.

All the trials will be briefly discussed.

#### **L44 Health-related Quality of life, anxiety and depression symptoms in patients who have had surgery for abdominal sarcoma**

*S. Grindberg, L. Smedberg*

Karolinska University Hospital, Stockholm, Sweden

This is a prospective longitudinal study with a quantitative approach carried out at a surgical unit in a hospital in Stockholm. We describe 30 patients with gastrointestinal soft tissue sarcoma (GIST, leiomyosarcoma, liposarcoma, malignant fibrous histiocytoma and solitary fibrous tumor) who had health-related quality of life, depression and anxiety measured before and after surgical treatment.

Health-related quality of life, depression and anxiety were measured with questionnaires before surgery (baseline), when discharged from nursing ward and at repeat visit after surgery. The instruments used were the EORTC QLQ-C30, developed by the European Organization for Research and Treatment of Cancer and the Hospital Anxiety and Depression Scale.

We found that patients experienced a loss of physical function at discharge from the ward compared with before surgery and that anxiety symptoms peaked before surgery.

#### **L45 I will return 50....belive it or not! My 19, soon 20 years as a GIST patient**

*S. Thorbjørnsen*

Oslo, Norway

## **L46 Social life for children and teenagers during chemotherapy**

*M. af Sandeberg*

Department of Pediatric Hematology and Oncology, Karolinska University Hospital, Stockholm, Sweden.

Missing friends and school is considered a major concern for children undergoing treatment for cancer. Self-reported 'health related quality of life' is diminished during cancer treatment and positively related to school attendance. Trying to live a normal life, including friends and school is, according to children, during and after cancer treatment the best way to cope with the cancer experience. Social isolation during different developmental stages in childhood may be detrimental and 'the Swedish national guidelines for social life when a child is treated for cancer' encourages social interaction and school attendance. Thus, the aim of the Swedish national network of consultant nurses is to prevent children with cancer from being separated from a normal social life. Nevertheless, contacts with friends, preschool and school are limited due to treatments, adverse events and examinations. In the meantime it is, however not always due to recommendations based on evidence. The children themselves may avoid social interaction due to changes in appearance and therefore support from peers is particularly important. Furthermore, in a developmental stage primarily characterized by activity and action the children describes that they are, despite known positive consequences from physical activity constantly being told that they should rest. Endless nagging about food, regardless of risk of malnutrition is also affecting social life. Fear of increased risk of infection is the most common reason for not recommending social interaction and school absence. Studies have, however shown that school attendance is not a risk factor for treated infections during cancer treatment.

## **L47 Rehabilitation conversations with cancer patients in hospitals**

*L. Bjerrum-Thisted*

Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark.

**Background and purpose:** Systematic and coherent efforts in identifying cancer patient needs for rehabilitation and palliation initially, during, and after treatment, based on the patient's perspective and need of support, has been mandatory in Denmark since 2012 according to the National Cancer Disease Management Program. I present the experiences with development of a dialogue tool used in conversations with patients about needs for support during and after cancer treatment.

**Methods:** The composition of the tool was based on literature, clinical experiences from experts in palliative and rehabilitation care, the conceptual framework and WHO model for rehabilitation, ICF, and with inspiration from Distress Thermometer and Guided Self Determination. The tool was tested and evaluated (2013-14) by using questionnaires, focus group interviews, user board meetings, observations and individual interviews with staff and patients.

**Results:** A new tool was developed where patients and relatives are invited to be involved and prepare themselves for conversations about rehabilitation by answering different questions at home. The following dialogues often led the patient or the healthcare professional to take action. The tool is in spring 2015 implemented across all hospitals (n=6) and municipalities (n=29), in the Capital Region of Copenhagen and will be evaluated after a year with qualitative methods and journal audit.

**Interpretation:** The tool provides efficient use of patients and nurses time and the opportunity for a more targeted and qualified conversation. The dialogues became more focused on the patient's actual concerns and more delicate matters like sexuality and anxiety were discussed

## **L48 “No one mentioned the word sex”. Women’s and men’s experiences of information about sexual health in relation to cancer treatment**

*E-M. Rasmusson*

Department of Oncology and Radiotherapy, Skane University Hospital, Lund, Sweden

**Background and purpose:** Research shows that the sexuality of both men and women is affected by cancer and the treatment. Healthcare staff are obliged to inform about treatments and their side effects. Studies show that the information about sexual health is deficient. 2 studies investigate to what extent patients with cancer needed such information, and what information they had been given.

**Methods:** The qualitative study (study I) is based on interviews with 11 women with gynecological cancer. In the quantitative study (study II) 106 men and women with different kinds of cancer answered a questionnaire. Both studies are predominantly descriptive. The results were analyzed and discussed using the theory of sexual scripts.

**Results:** In study I, women with gynecological cancer wanted health care professionals to initiate conversations about sexuality and partner relationships. It is important for health care professionals who provide care for these women to be open and sensitive toward their questions. Further, health care professionals need to be sensitive to when it is the best time for the individual woman to talk about her situation and to address her questions.

Study II shows that 48 % of the respondents had not had any information regarding the areas of inquiry. There was a discrepancy between the information they had wished for and the information they had received. A considerably larger number of men than women had been given information about fertility and sexual desire.

## **L49 Nausea and vomiting: treatment by evidence and history**

*D.M, Sørensen*

Department of Oncology, Vejle hospital – part of Lillebaelt Hospital, Denmark.

**Background and purpose:** This is a presentation regarding work by a Special Interest Group, organized through the Danish Nurses' Organization and Danish Cancer Nurse society. Previous studies showed differences between professionals a patients experience and expectations of nausea and vomiting in relation to chemotherapy. The aim of the group is to implement evidence-based antiemetic guidelines for best supporting antiemetic treatment.

**Methods:** Group-meetings 5-6 days a year, presentations in our own departments, participation in 1 vomiting and nausea local groups, presentations at national and international conferences. This presentation will address the methods of gathering information regarding the patients experience of nausea and vomiting, which international antiemetic guidelines that are being used in Denmark and how we, as a Special Interest Group try to ensure both evidence-based and best practice in the treatment of nausea and vomiting.

**Results and conclusion:** The work of the Special Interest Group shows a difference between the international guidelines and the antiemetic treatment given. We find it important to ensure the knowledge regarding evidence-based antiemetic guidelines and the methods and effect of gathering information regarding the patients experience of nausea and vomiting.

## **L50 Oral complications of cancer therapies**

*S. Bier Jensen*

Department of Odontology (School of Dentistry), University of Copenhagen, Copenhagen, Denmark

This lecture will discuss oral complications of chemotherapy and evidence-based clinical management guidelines. The main focus will be on oral mucositis, salivary gland hypofunction and xerostomia, and oral fungal infections. Oral mucosal lesions from targeted therapies and management strategies will also be addressed

## **L51 Important issues in care for TYA (teenagers and young adults) treated for cancer**

*M. Olsson, M. Jarfelt, P. Pergert, K. Enskär,*

Department of Pediatrics, Institute of Clinical Sciences, University of Gothenburg, Sahlgrenska Academy, Sweden

**Purpose:** This study explores the needs of teenagers and young adults (TYA) in care situations. The overall aim of this project is to highlight issues that are important for the TYA patient to develop a questionnaire based on these issues.

**Method:** The chosen method is focus group interviews and personal interviews with former cancer patients. The inclusion criteria were patients who had finished treatment not longer than 3 years ago and were between 15-29 years when treated. Participants were recruited from university hospitals in Sweden. In every focus group there were a number of 2-6 participants. The interviews were recorded, transcribed into text and analyzed through content analysis.

**Results:** Results can be summarized into 4 categories: personal professional interaction, knowledge and participation, age-appropriate environment, and support. Important TYA care needs vary over time due to individual situations. The time line of the cancer experience can be described as a continuum; at diagnosis, during treatment, and in life- after cancer treatment.

**Interpretation:** TYAs treated in Sweden have special needs that are not being satisfied, whether at pediatric or adult units. Areas that need closer attention are: close relatives' participation in the care, information on sex and fertility, age-appropriate social physical environments during treatment, and psychosocial support after treatment. The result will be a base for the development of a questionnaire for TYAs cancer care.

## **L52 Municipal rehabilitation in Copenhagen Centre for Cancer and Health**

*K. Birtø*

Center for Kræft & Sundhet, Copenhagen, Denmark

## L53

*W. P. Bekkering*

Department of Orthopedics, Rehabilitation and Physical Therapy of the Leiden University Medical Center / Willem-Alexander Children's Hospital, Leiden, the Netherlands.

Departments of Orthopedics and Pediatric oncology of the Academic Medical Center / Emma Children's Hospital, Amsterdam, the Netherlands.

**Background:** Survival rates of patients with malignant bone tumors improved over the last decades. Similarly, various extremity salvage procedures became available leading to a decline of amputation rates. Quality of Life (QoL) is an important outcome measure in children and young adults after surgery for a malignant tumor of the lower extremity.

**Methods:** QoL scores in comparison with healthy peers, other pediatric cancer patients and differences between different surgical options will be presented based on current literature.

**Results:** Patients after lower extremity bone tumor surgery report significantly lower functional QoL scores in comparison with healthy peers and most other pediatric cancer patients. However, mental QoL scores appear to be better than scores in healthy peers. In the available literature, no consistent differences were reported between limb-salvage and ablative surgery. Prospectively, after two years since surgery no further improvements were achieved at functional QoL scores and mental QoL scores remain favorable in comparison with healthy peers.

**Discussion:** The implication of the data published so far for children and young adults is hampered by the selection of predominantly elderly patients and the lack of objective and child adequate measures. Furthermore, research is limited to generic QoL measures and disease specific measures with interest into sportive and cosmetic aspects that characterize bone tumor surgery are lacking. Different initiatives to improve QoL like earlier and extensive information provision, shared decision making and support groups will be presented.



## **L54 Adolescent and your adult patients with cancer, a clinical practical guideline**

*K. Stokke, K. Tveten, M. Gudim, KM. Antonsen, HM. Riddervold, S. Næss, AK. Bergan, J. Lebesby, SN. Jensen*  
Oslo University Hospital, Norway

**Background and purpose:** More young people are cured of cancer, but long-term side effects and follows can strike hard at a time when the young develops from childhood to adulthood. Adolescence is characterized by great expectations and changes. Young people separate from parents, take education and establish themselves. When an adolescent gets cancer, it causes that every day is marked by illness and treatment. Treatment takes time and effort that should be spent on so much else. Youth is in growth and development both physically, mentally and socially, and undergoes the cancer treatment in a particularly vulnerable period. Being seriously ill can make it difficult to follow the studies, maintaining friendships, establish dating relationship and maintain social networks.

Adolescents who receive treatment in hospitals are a special group in the hospital environment. They are treated both at the pediatric ward and the adult department. None of the departments are organized for this patient group. It is nevertheless important to emphasize that there is much good work being done today. At the same time that random experience and inadequate knowledge about youth can make communication between youth and health workers not as good as both parties want. Oslo University Hospital has therefore the need for a professional guideline for psychosocial follow-up of adolescents with cancer.

**Methods:** The guideline is developed by evidence-based method that means that the recommendations are based on systematic obtained research combined with the clinician's experience and expertise and patient values and preferences. In order to the users' perspective there were two representatives from the Youth group in Cancer Society and a youth representative from the User council at the hospital represented.

**Results:** The guideline addresses the examination phase, the diagnosis and treatment phase, life after treatment, progression during treatment, relapse

of the disease, palliative care and follow-up of survivors to reduce psychosocial consequences for patients and relatives.

**Interpretation:** The work is however not finished when the guideline is finalized. Then the main job begins; to implement it in the hospital. The procedures and guidelines must live the life out there in the hospital where patients and health workers is housed so that youth and their families will be attended in the best possible way.

## **L55 Young and cancer**

*T. Lindahl Greve*

Ung Cancer, Sweden

## **L56 Physiotherapy following surgery**

*T. Thorkildsen*

Oslo University Hospital, Radiumhospitalet, Oslo, Norway

## **L57 Functional ability and physical activity in children and adolescents after lower extremity bone tumor**

*P.W. Bekkering*

Department of Orthopedics, Rehabilitation and Physical Therapy of the Leiden University Medical Center / Willem-Alexander Children's Hospital, Leiden, the Netherlands. Departments of Orthopedics and Pediatric oncology of the Academic Medical Center / Emma Children's Hospital, Amsterdam, the Netherlands.

**Background:** Survival rates of patients with malignant bone tumors have improved over the last decades. Similarly, various extremity salvage procedures became available leading to a decline of amputation rates. Functional ability and the level of physical activity are important outcome measures in children and young adults after surgery for a malignant tumor of the lower extremity. However, extensive research report disappointing outcome scores in comparison with healthy peers and other pediatric cancer survivors and has not been able to determine consistent advantages for either limb-salvage or ablative procedures.

**Methods:** Functional ability and physical activity scores in comparison with healthy peers, other pediatric cancer patients and differences between different surgical options will be highlighted based on current literature.

**Results:** Patients after lower extremity bone tumor surgery report significantly lower functional ability and physical activity scores in comparison with healthy peers and most other pediatric cancer patients. Furthermore, patients report no consistent differences at physical ability and functional activity levels between limb-salvage or ablative surgery. However, different sportive choices were made based on the fragility of the reconstructed extremity. No further improvements were achieved after 2 years since surgery.

**Discussion:** The implication of the data published so far for children and young adults is hampered by the selection of predominantly elderly patients and the lack of objective and child adequate measures. Furthermore, research is limited to daily functioning and activities with little interest into sportive and intensive activities that characterize childhood and adolescence. Different initiatives to improve functional ability and physical activity levels like oncological training and sportive rehabilitation will be presented.

## **L58 Physiotherapy and surgery, clinical exercise**

*M. Lia Johansen*

Oslo University Hospital, Radiumhospitalet, Oslo, Norway

## **L59 Pain and sarcoma, reflections from a physiotherapist**

*H. Hallström*

Karolinska Universitetssjukhuset, Stockholm, Sweden.

## **L60 Social life for children and teenagers during chemotherapy**

*M. Quist*

Rigshospitalet, University of Copenhagen, Copenhagen, Denmark

In comparison with other areas of clinical medicine, exercise therapy has received comparably less attention in persons following a diagnosis of cancer. The precise reasons for this are unknown but likely stem from the prevailing dogma that a cancer diagnosis is associated with poor survival, a compromised immune system that may be more compromised by exercise training, and other debilitating side-effects that preclude participation in and benefit from exercise training. Nevertheless, the past two decades has witnessed a dramatic change in attitude with significant increased research and clinical interest in the role of exercise therapy following a cancer diagnosis. The increased interest in exercise has occurred in conjunction with the emergence of cancer survivorship and the growing importance of managing the late-effects of cancer therapy in persons who are now living much longer following a cancer diagnosis.

The evidence of exercise offer to patients with cancer undergoing chemotherapy is growing and the effect has been established in several meta-analyses. This presentation will focus on the approach towards patients with cancer undergoing chemotherapy with early and late stage cancer and how this approach can be beneficial from both a physical and mental view.

## Posters (P)

### **P1 Denosumab in patients with giant-cell tumor of bone in Norway; results from a nationwide cohort**

*K. Boye<sup>1</sup>, N. L. Jebsen<sup>2</sup>, O. Zaikova<sup>3</sup>, H. Knobel<sup>4</sup>, A. M. Løndalen<sup>5</sup>, C. S. Trovik<sup>2</sup>, O. R. Monge<sup>2</sup>, K. Sundby Hall<sup>1</sup>*

<sup>1</sup>Department of Oncology, Oslo University Hospital, Oslo, Norway. <sup>2</sup>Department of Oncology, Haukeland University Hospital, Bergen, Norway. <sup>3</sup>Department of Orthopaedic Surgery, Oslo University Hospital, Oslo, Norway. <sup>4</sup>Department of Oncology, St. Olavs University Hospital, Trondheim, Norway. <sup>5</sup>Department of Nuclear Medicine, Oslo University Hospital, Oslo, Norway

**Background and purpose:** Denosumab is a relatively new treatment option for patients with giant-cell tumor of bone (GCTB). We report the results for patients treated in Norway.

**Methods:** Patients treated with denosumab for GCTB were identified from the clinical databases at the Norwegian sarcoma reference centers. Data was collected by retrospective review of patient records.

**Results:** 18 patients were identified. Denosumab was given for recurrent disease in 7 patients and as first line treatment in 11 patients, of which 6 received therapy as part of a neoadjuvant/adjuvant strategy and 5 for surgically unsalvageable primary tumor. 10 of 12 patients with recurrent or unresectable disease are still on denosumab without progression with median treatment duration of 25 (6-40) months. 2 patients discontinued treatment due to osteonecrosis of the jaw and reduced compliance, respectively. In the adjuvant group, 3 patients had disease recurrence after stopping denosumab, 2 patients are disease-free after 11 and 7 months, and 1 patient currently receive adjuvant treatment. In 3 of 6 patients, the extent of surgery was reduced due to the neoadjuvant therapy. 17 of 18 patients underwent 18F-FDG PET/CT response evaluation at median 4.7 weeks from starting denosumab. Median baseline SUVmax was 11 and median SUVmax at evaluation was 4.9 (p<0.001).

**Interpretation:** In a nationwide GCTB patient cohort, denosumab was an effective agent and durable responses were observed. Adjuvant therapy seems questionable. 18F-FDG PET/CT could be a valuable tool for early response evaluation.

## P2 Computer-guided pelvic tumor resection and reconstruction with a custom made prosthesis produced in a 3-D printer. A case report

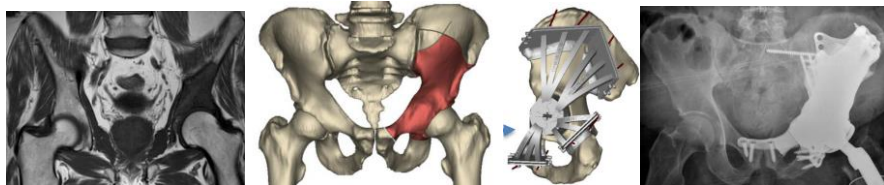
*R. Löfvenberg, V. Otten, M. Farhang, S-E. Persson, K. Strigård.*

Department of Surgery and Perioperative Sciences, Umeå University, Umeå, Sweden

**Background and purpose:** The complex geometry of the pelvis and large tumors with limited visualization, whether with eyesight or x-ray, make an accurate resection difficult. We report our experience with computer-guided surgery using patient-specific instruments and a 3D printed reconstruction prosthesis

**Patient and method:** A 56-year-old-man had a chondrosarcoma (grade II) in his left pelvis. No metastasis. The tumor invaded os ischium, os pubis and os ileum. In collaboration with Mobelife ([www.mobelife](http://www.mobelife)) a patient specific instrument (saw gig, bore gig) was constructed as well as a custom made prosthesis including a manual. The operative team included 3 orthopedic surgeons, 1 vascular surgeon and 1 surgeon specializing in abdominal wall surgery. The operative time was 13 hours.

**Result:** The operative procedure was satisfactory with wide margins. The patient is satisfied but is still using crutches 1 month after operation.



Planned resection and demonstration of saw gig (lateral view)

**Reference:** Gouin et al. Computer-Assisted Planning and Patient-Specific Instruments for Bone Tumor Resection within the Pelvis: A Series of 11 Patients. *Sarcoma*. 2014; 2014: 842709

Bellanova et al. Surgical guides (patient-specific instruments) for pediatric tibial bone sarcoma resection and allograft reconstruction. *Sarcoma*. 2013;2013:787653. doi: 10.1155/2013/787653. Epub 2013 Mar 4.

### **P3 Serum beta-HCG as a tumor marker in a patient with osteosarcoma: case report**

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Beta-HCG serum levels can be helpful in patients with various malignancies, but is not routinely used in osteosarcoma to monitor treatment response and disease progression. I present a woman with an osteosarcoma of the 7<sup>th</sup> rib and increased pre-treatment beta-HCG level. A 22-year-old woman was admitted to our hospital in January 2011 with a mass in the right hemithorax. Pre-operative routine laboratory investigations revealed a high level of serum beta-HCG 240 IU/L (normal range 0.0-7.0 IU/L). Pregnancy was excluded. After radical operation with a pathohistological confirmation of centrally osteosarcoma, repeated measurements of beta-HCG levels showed that they normalized 17 days after the operation. The patient received adjuvant chemotherapy. During adjuvant treatment levels of serum beta-HCG remained normal till May 2011 when suddenly raised to 169 IU/L. The patient now had severe pain in the back. Bone and also lung metastases were radiologically confirmed. The patient was treated with an operative stabilization of Th6, L1 and L3 and postoperative radiotherapy. The salvage chemotherapy was not delivered to the patient due to poor performance status. Levels of serum beta-HCG were continuously rising as the disease progressed. An extremely high maximum value of beta-HCG 58123 IU/L was reached in August 2011 a few days before the patient died.

According to some previous reports, we confirmed that serum beta-HCG may be used as an additional diagnostic, prognostic and follow-up marker in some osteosarcoma patients, but needs further investigation with a larger sample size.

## **P4 The microenvironment of soft tissue sarcomas. Predominance of tumor associated macrophages and expression of immunomodulating molecules.**

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**Background and purpose** Interaction of malignant cells with non-malignant connective tissue cells that comprise the stroma of the tumor forms a microenvironment which is important for tumor growth. We describe in detail the molecular microenvironment of high-grade soft tissue sarcomas. A special interest lies in correlating the profile of the tumor with patient prognosis.

**Methods** Paraffin-embedded sections are stained by immunohistochemistry in order to detect the presence of tumor-associated macrophages, T regulatory lymphocytes and growth factors. RNA is isolated and quantitative analysis of gene expression is performed. Data are correlated to histological parameters that are known to affect prognosis.

**Results** Tumor associated macrophages were abundant in the majority of specimens tested, and their presence correlated with the expression of the immunosuppressive cytokine IL-10 and the pro-inflammatory mediator cox-2. Large tumors showed higher expression of the pro-inflammatory cytokine IL-1b. T regulatory lymphocytes were present in a minority of the specimens tested.

**Interpretation:** Tumor associated macrophages appear to have a key role in the biology of soft tissue sarcomas, and they may be a target for novel therapies.



## **P5 High tenascin C expression is associated with shorter time to recurrence in desmoid tumors**

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**Background and purpose:** Desmoid tumours are benign lesions that have a high rate of recurrence. Although the tumours lack metastasizing ability, they can cause severe morbidity and even death. Predicting the clinical course of desmoid tumours is challenging and there is controversy concerning the treatment protocol. The extracellular matrix glycoprotein tenascin C is highly expressed during embryonic development, tissue repair, and in pathological disorders such as chronic inflammation and cancer but expression is low in normal tissue. Drugs targeting the expression or function of tenascin C are currently being developed. Our aim was to evaluate the role of tenascin C in desmoid tumours.

**Methods:** We used tissue microarray (TMA) technique and immunohistochemistry to study the expression of tenascin C in 91 patients with surgically resected desmoid tumours and compared its immunoexpression with clinical data. Tissue microarrays were constructed, and the specimens were immunohistochemically stained for tenascin C expression. Each tissue specimen was manually assessed on a staining scale ranging from 0 (no staining) to 3 (strong staining) and correlated with various clinicopathological parameters.

**Results:** Immunohistochemistry revealed tenascin C expression in 48 % ( $n = 42$ ) of the tumours. High Tenascin C protein expression was significantly associated with a shorter time to progression ( $p = 0.04$ ).

**Interpretation:** We conclude that tenascin C expression may have prognostic significance in desmoid tumours.

## **P6 Sirtuin expression, activity and inhibition in pediatric soft tissue sarcomas**

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**Background and purpose:** Sirtuins are NAD<sup>+</sup> dependent deacetylases and ADP-rybosyl transferases active on histone and non-histone substrates. The first sirtuin was discovered as a transcriptional repressor of the mating-type-loci (*Silent Information Regulator sir2*) in the budding yeast, where it extended life span. 7 mammalian sirtuins (SIRT1-7) have been now identified. These proteins regulate cellular processes such as metabolism, cell survival, differentiation, DNA repair and are implicated in the pathogenesis of solid tumors and leukemias. We investigated the effects of sirtuin expression, activity and inhibition on the survival of pediatric sarcoma cell lines.

**Material and methods:** We analyzed the expression of SIRT1 and SIRT2 in pediatric sarcoma tumors cell lines and normal cells and evaluated the activity of the sirtuin inhibitor and p53 activator tenovin-6 (Tv6) in synovial sarcomas and rhabdomyosarcomas. We found that SIRT1 is overexpressed in synovial sarcomas biopsies and cell lines in comparison to normal mesenchymal cells. Pharmacological inhibition of Tv6 inhibited tumor cell proliferation in sarcoma cell lines and impaired sirtuin enzymatic activity. Interestingly, tumor cell death following Tv6 treatment was associated with an impairment of autophagy as shown by an increment of autophagy associated proteins lipidated LC3 (LC3II) and p62 following exposure to Tv6. Using siRNA to knock down SIRT1 and SIRT2, we found that the expression of both proteins is crucial for the survival of rhabdomyosarcoma cells and that the loss of SIRT1 expression results in a decreased LC3II expression. Tv6 administration in SCID mice carrying human rhabdomyosarcoma xenografts significantly reduced tumor growth and induced nuclear translocation of p53, SIRT1 and SIRT2.

**Interpretation:** SIRT1 and SIRT2 expression is crucial for the survival of synovial sarcomas and rhabdomyosarcomas. Our findings show, for the first time, a role for sirtuin deacetylating activity on the survival of synovial sarcomas and rhabdomyosarcomas and associate sirtuin activity with autophagy.

## **P7 The significance of major and minor tumor rupture of small bowel gastrointestinal stromal tumors (GISTs)**

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**Background and purpose:** Tumor rupture is an established risk factor for recurrence of GIST, but the term has not been defined. We propose definitions of major and minor rupture and have investigated their impact on frequency and pattern of recurrence.

**Method:** Patients treated surgically for small intestinal GIST without metastases from 2000 through 2012 were retrieved from our prospective sarcoma database. Information on tumor rupture was supplemented by retrospectively reviewing surgical reports, and pathological slides were re-examined for possible tumor involvement of the peritoneal surface. Major rupture included surgical biopsy, blood-tinged fluid at laparotomy, bowel perforation at tumor site, tumor fracture/piecemeal resection, and microscopic tumor infiltration into adjacent organ. Minor rupture included percutaneous needle biopsy, naked tumor at surgery, iatrogenic serosal rupture, and microscopically involved margins/surfaces without macroscopic evidence of rupture.

**Results:** 72 patients were identified. Radical surgery (R0/R1) was performed in all but 1 case. No rupture was recorded in 32, minor rupture in 21 and major rupture in 19 patients. 7 patients completed adjuvant imatinib treatment; 1 patient received neoadjuvant treatment. Of 27 recurrences, 19 were peritoneal. Estimated 5-year peritoneal recurrence rates were 48 %, 25 % and 23 % after major, minor and no rupture, respectively ( $P=0.006$ ). Correspondingly, estimated overall 5-year recurrence rates were 61 %, 28 %, and 34 % after major, minor and no rupture ( $P=0.003$ ).

**Interpretation:** Peritoneal recurrences are increased only after major tumor rupture. Minor rupture should not be used as a sole criterion for adjuvant treatment.

## **P8 Surgery alone is sufficient in patients with liposarcoma of the spermatic cord: our experience of 5 patients**

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**Background and purpose:** Liposarcoma of the spermatic cord is rare annually affecting approximately 0.25 persons per million. No consensus exists regarding best treatment. We describe our experience of 5 patients.

**Methods:** 5 cases of liposarcoma of the cord from 2011 to 2014 was included in our retrospective series. Data collated included the patient's age, tumor size, presentation, site of tumor, World Health Organisation (WHO) histotype, tumor grade, adequacy of tumor resection, and the use of adjuvant chemotherapy or radiotherapy. Following surgery, decision for adjuvant therapy was decided by a Multidisciplinary Tumour Board. Patients were routinely followed up at least every 6 months. Our study end points included duration of disease-free survival (DFS) as well as overall survival (OS).

**Results:** The median age was 67 (33 – 72) years. 3 patients presented with primary disease. 3 patients had well-differentiated and 2 patients had dedifferentiated liposarcoma. 2 patients both had primary disease first identified post-operatively by histology, required re-excision to obtain R0 resection margins. None of our patients underwent adjuvant chemotherapy or radiotherapy following achievement of R0 resection margins. None of our patients have developed loco-regional recurrence or distant metastases. Median disease-free survival was 30 (10 – 45) months

**Interpretation:** After surgery with R0 resection margins alone, adjuvant chemo- or radiotherapy may not be required in the management of spermatic cord liposarcoma.