



# PEDIATRIC & ADOLESCENT OSTEOSARCOMA ... PROGRESS FROM THE PAST, PROSPECTS FOR THE FUTURE

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## Symposium Syllabus

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## QUALITY OF LIFE IN LONG TERM OSTEOSARCOMA SURVIVORS: AMPUTATION VERSUS LOWER LIMB SALVAGE

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Treatment protocols for osteosarcoma adopted over the past three and a half decades comprise preoperative (neoadjuvant) chemotherapy, surgical resection of the tumor and postoperative treatment. This strategy has improved the ability to perform safe limb-sparing procedures and has yielded a significant increase in overall survival. With modern therapy almost 70% of the osteosarcoma patients survive. As the number of survivors of childhood cancer increases, the impact of quality of life and function needs to be addressed. Data in regard to these aspects in osteosarcoma is limited. Limb salvage is the preferred treatment when patients have treatment options, yet the questionable long-term durability and complications of prostheses, combined with ambiguous function, leave doubt regarding the best clinical and surgical options. Comparisons between limb salvage patients, amputees and controls also require further investigation.

Indications for primary amputation are based on the location and local extent of tumor and the expected function of the extremity after tumor resection. Amputation would leave the patients with a life-long requirement for an external prosthetic leg associated with an overall limited walking distance. While artificial limbs are much more sophisticated than those used in the past, phantom limb sensations remain a substantial and unpredictable problem in the amputee. Complications such as stump overgrowth, bleeding, infection, although possibly rare, also require further elucidation.

Limb salvage surgery using endoprosthesis, allografts or reconstruction is performed in approximately 85% of patients affected by osteosarcoma located in the middle and/or distal femur. One drawback in limb-salvage surgery in the long term survivor is that endoprostheses have a limited life span with long-term prosthetic failure. The inherent high rate of reoperation remains a serious problem. Replacing a damaged, infected or severely worn out arthroplastic joint or its intramedullary stem is difficult, especially in the long-stem cemented endoprostheses used in the 80's. Limb lengthening procedures in patients who have not reached maturity must also be addressed.

Infection represents a major complication of prosthetic joint surgery, despite advances in design, surgical technique and antibiotic prophylaxis. Periprosthetic infections, compared to other indications for joint reconstruction, were found to be more frequent in patients treated for neoplastic conditions and their outcome can be devastating, resulting in total loss of joint function, amputation, and systemic complications. Numerous treatments have been proposed including irrigation, debridement and prosthesis retention, revision or excision arthroplasty and knee arthrodesis.

Quality of life in terms of function, psychological outcome and endpoint achievements such as marriage and employment apparently do not differ significantly between amputee and nonamputee osteosarcoma survivors. Among the instruments available to evaluate patients are: Quality of Life – Cancer Specific, a standardized self-report measure of spiritual, physical, social, and psychological well-being; Physical function by TESS –Toronto Extremity Salvage Score, a self-report of difficulty with daily activities and psychological outcomes of: 1) body image – assessed with the Amputee Body Image Scale and modified to include limb salvage participants ; 2) self esteem -- assessed by the Index of Self-Esteem and 3) Social support -- assessed by the Sarason Social Support Scale. Independent of the type of surgical procedure, lower function and quality of life scores appear to be associated with lower educational attainment and increased self-reported disability. Both amputation and limb-sparing techniques in osteosarcoma survivors of the lower extremity have benefits and drawbacks. Although these two categories are



functionally and cosmetically different, there do not appear to be any clear-cut advantages to either.

It has been reported that the overall risk of a subsequent amputation after limb salvage surgery with endoprosthesis replacement is 8.9% and is due to the following: 63% local recurrence, 34% infection, 2% mechanical failure of the prosthesis, 1% persistent pain.

There is a paucity of information in regard to upper extremity osteosarcoma survivors. However in the few patients seen at the Children's Cancer Hospital at the University of Texas MD Anderson Cancer Center, most agree that it is preferable to have an upper salvaged appendage of limited function rather than an amputation. Amputee patients nonetheless appear to have made satisfactory adjustments to their deficits with or without a (functional) external prosthesis.

A preliminary investigation of 100 pediatric and adult osteosarcoma survivors treated during childhood and adolescence at the University of Texas MD Anderson Cancer Center demonstrated that they adapted well to their motor and functional disabilities. It also appeared that amputee patients had a similar psychological and quality of life outcome as limb salvage patients. Assessments gleaned in amputees at the annual MD Anderson Cancer Center Ski Rehabilitation Program over the past three decades revealed that adult survivors had equanimity and economic independence. There was no evidence of excessive anxiety or depression or deficits in self-esteem compared with the normal population or matched controls. A number of amputee long term survivors also achieved high ranking in the professional and commercial work force. These positive aspects should be recognized and emphasized to patients and their parents when discussing outcome.

While information in regarding the quality of life in long term amputee and limb salvage osteosarcoma patients is limited, it must be recognized that publications on long term childhood cancer survivors have increased exponentially during the past decade. These publications address quality of life and complications which have occurred as a consequence of therapy administered principally to patients with neurological, hematological and solid tumor malignancies. Data are still being assembled on outcomes in osteosarcoma patients. It would not be unexpected to find similar chemotherapy induced side effects. The prominent complications include auditory dysfunction due to cisplatin, anthracycline induced cardiac dysfunction, sterility due to alkylating agents and second malignant neoplasms.