Overview

Issues of Survivorship and Rehabilitation in Soft Tissue Sarcoma

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Abstract

As the number of survivors of extremity soft tissue sarcoma increases, so does the need to understand the experience of survivors and develop measures, systems and services that support rehabilitation into normal life roles. Survivorship includes considerations of the physical, psychological and social domains, of which the physical sequelae of treatment are the best characterised in the literature. The survivorship experience may include disability, pain, lymphoedema, psychological problems, as well as difficulty with employment, relationships and lower quality of life. Rehabilitation strategies for extremity sarcoma patients must be personalised, holistic and begin early in the pathway, ideally before the first treatment intervention. The International Classification of Functioning, Disability and Health model is a useful framework for combining assessments, including objective outcome measures, which can be combined into a rehabilitation prescription. Research is needed to develop an evidence base for rehabilitation interventions to support patients with extremity soft tissue sarcoma.

Key words: Disability; impairment; rehabilitation; sarcoma; survivorship

Statement of Search Strategies Used and Sources of Information

During the research for this article the authors used the following search terms: survivorship, rehabilitation, sarcoma, impairment and disability.

Introduction

Sarcomas originate in connective tissues and other supportive structures such as bone, muscle and blood vessels. It is no surprise, therefore, that sarcoma treatment involving the removal of muscle and bone can have a dramatic impact on the locomotor system, routinely causing impairments, disability and restricting participation in normal life. Although the incidence of soft tissue sarcoma increases with age, many patients are young, increasing the burden over time on patients, their families and society. Incremental improvements in survival mean that there are an estimated 280 000 sarcoma survivors in Europe [1,2]. Patients have a wide range of survivorship experiences and require tailored specialist treatment, information and support.

This overview explores the survivorship experiences of patients treated for an extremity soft tissue sarcoma, the provision of rehabilitation services and discusses future directions. Some aspects may be relevant to patients with soft tissue sarcomas in other locations and some relevant studies from patients with bone sarcoma are included, but their detailed description is beyond the scope of this review.

What is Survivorship?

Survivorship is the lived experience of individuals after treatment for cancer. The term encompasses the physical, psychosocial and economic sequelae of cancer diagnosis and treatment in paediatric and adult survivors of cancer [3]. The recognised domains of survivorship are: pain; sexual function; cognitive functioning; employment, finance and return to work; emotional distress; depression; anxiety; and social needs. Here these are considered in three sections: physical (fatigue, physical functioning and pain); psychological (emotional distress, cognitive functioning, depression and...
anxiety); and social (sexual function, employment and social needs) [4].

**Physical**

Treatment for extremity sarcoma often leads to disability and participation restrictions, particularly after lower extremity tumours [4–8]. Physical functioning may worsen during treatment and then improve for a year or more, before plateauing [9, 10]. In the longer term, patients may remain less active than controls [11]. High-grade, large size tumours, resection of motor nerves and flap reconstruction are all associated with poorer outcomes, the latter possibly because tumours are often larger and need radiotherapy [12, 13].

Treatment leads to structural impairments (e.g. reduced joint movement), activity (walking, dressing) and participation restrictions (e.g. sports or employment). These are interdependent, for example higher physical impairment is associated with lower health related quality of life (QoL) [14–17]. Evaluating and understanding these different aspects requires standardised outcome measures and cohorts of patients [18].

Maintaining physical activity levels after cancer treatment is important, not only to improve general health but reduced physical activity may be linked with lower survival rates (e.g. after breast, colon or colorectal cancer) [19]. Therefore, patients treated for extremity soft tissue sarcoma may be at further increased risk of poor cancer outcomes. Furthermore, regular exercise may help to improve the lower exercise capacity (including VO$_{2}$max) seen in some survivors of paediatric cancer [20].

**Amputation or Limb Salvage?**

Most patients with extremity soft tissue sarcoma have limb-sparing surgery, but some undergo amputation. Survivorship experiences may differ, although QoL is similar after both [21]. Although amputation is associated with more disability, some patients have poor physical functioning after limb-sparing surgery and others function very well after amputation (particularly at the trans-tibial level) [17], but the latter depends on good limb fitting services, which vary widely in the National Health Service [22].

Both groups may experience pain and rehabilitation can be prolonged. Falls are also common after amputation (62% report falls) and limb-sparing surgery, for example after femoral nerve resection [17, 22, 23]. Falls cause fractures and other injuries, hence strategies to prevent and manage falls are important [22].

Decisions about limb-sparing surgery or amputation should consider the trade-offs between the two approaches. Publicity around paralympians in particular may be changing attitudes to disability and may lead to a greater acceptance of amputation [24].

**Fatigue**

Fatigue includes problems with concentration, motivation and physical activity, and affects people with several cancer types, including sarcoma, particularly during treatment [10]. Twenty-eight per cent of patients with benign or malignant bone and soft tissue tumours report severe fatigue, associated with treatment complications, less optimism and somatisation [25]. Fatigue is also associated with pain, anxiety and low self-efficacy [26]. Amputees may report less fatigue; it has been suggested that this is because the physical changes after treatment have to be accepted [27].

**Pain**

Pain is common in survivors, regardless of local treatment [21, 28, 29]. Pain is a regular feature in over 90% of patients after amputation (severe in 9.5%) [17] and has a significant and negative impact on physical functioning, QoL and employment. Although pain may improve from the diagnostic to treatment phase, it can last for many years [10, 17, 21, 29, 30].

**Lymphoedema**

The interruption of lymphatic drainage by surgery or radiotherapy makes lymphoedema an expected complication, affecting about a third of patients after extremity soft tissue sarcoma, some severely [31]. Factors associated with lymphoedema include larger (>5 cm), deep tumours and vein resection [32]. The large volume limbs that result are associated with poorer outcomes [33].

**Psychological**

Psychological distress is reported by about a third of patients. At diagnosis, anxiety predominates, and a year after treatment, depression is predominant [34]. There is a high rate of anxiety and depression from diagnosis to follow-up [35], which may persist. In one study of paediatric sarcoma survivors, 77% reported long-term psychological distress [36]. Psychological functioning is closely linked to physical functioning and social domains, and access to psychological therapy, support and counselling is important. Body image may change, particularly after amputation, and this has been linked to QoL outcomes [21].

**Social**

A sarcoma diagnosis may accentuate social isolation and affect relationships and sexual functioning [37]. Single patients have a higher risk of sarcoma-specific death, possibly because spouses facilitate treatment either financially or through logistical support [38].

Despite poor fitness, adult survivors of childhood extremity sarcoma are as likely to be married, live independently, be employed and attend college as controls [16]. Lack of participation in life roles, marital status, age and professional status predict QoL [10, 39]. Psychological distress and pain may be barriers to employment [40, 41] and most patients would benefit from vocational counselling [33].

Sarcoma treatment may affect participation in sports, with potential consequences for long-term health and
fitness. Problem-focussed approaches that substitute other sports, such as golf or snooker, in place of contact sports may be helpful [42], as may other exercise interventions that have positive effects in terms of QoL, fatigue, sleep and cognitive function [43]. Patients with limb sparing or amputation seem to participate in sports at about the same rates [44]. Participation may reflect pre-morbid activity, but targeting motivation, fear and affect may help return to sports [45].

Rehabilitation

Rehabilitation enables patients to reach and maintain optimal physical, sensory, intellectual and social functional levels [46]. Good rehabilitation emphasises a return to normal psychosocial functioning [47]. Rehabilitation should begin early in treatment, ideally at diagnosis. Early and honest discussion about outcomes, home and work environments and roles can manage patient and family expectations and support informed decision making. This is particularly important if treatment aims to palliate rather than cure. Patients need a personalised multidisciplinary plan but delivery can be challenging as services vary and patients are widely distributed geographically and require remote monitoring [22]. Furthermore, there are few evidence-based disease-specific rehabilitation models and treatment strategies incorporating concepts of disability and QoL [48,49].

Intervention between diagnosis and first treatment (‘prehabilitation’) can reduce surgical complications and length of stay and lower health costs [50]. Prehabilitation typically includes assessments of baseline physical function, comorbidities and cardiorespiratory function. However, early assessment of home environments, employment and other roles is also beneficial [47]. Early interventions to improve cardiopulmonary fitness and total body strength may improve physical functioning after treatment, and these should be developed for sarcoma patients [50].

Good communication between the surgeon and the rehabilitation team is important to agree an appropriate regimen, considering limb weakness, swelling, neurological injury and weight bearing. Early pain control, wound management and pressure care are important to avoid wound or other complications that have a major effect on rehabilitation. Rehabilitation can also be delayed by chemotherapy and radiotherapy [51]. Daily stretching during radiotherapy may reduce fibrosis and joint contracture. Orthoses may be helpful if there is neurapraxia and distal limb weakness [47]. After amputation, support is required to maintain movement in unaffected limbs, stump bandaging and early mobilisation and prosthesis use.

After the acute phase, an emphasis on role development and return to work is helpful. In the longer term, good general health, diet and exercise should be encouraged. Exercise interventions should be personalised and consider the recognised barriers of pain, fear and anxiety [45]. New symptoms or a change in the limb should alert the team to the risk of local recurrence.

The International Classification of Functioning, Disability and Health (ICF) model (Figure 1) provides a useful conceptual framework for understanding the multidimensional needs of patients. The ICF supports data compilation, analysis, policy monitoring, service provision and communication between health professionals [53,54] and has been used to develop rehabilitation models [54,55]. A structured approach allows interventions to be targeted at specific domains [56], allows the interaction of domains to be explored and supports the development of evidence for interventions [53]. Physical rehabilitation and self-management programmes using the ICF framework can improve QoL in cancer survivors [48,57].

Rehabilitation prescription forms based on the ICF support collection of information and outcomes and can guide care delivery (Figure 2) [56,58]. The model can include performance scales, such as the Eastern Cooperative Oncology

![Fig 1. International Classification of Functioning, Disability and Health (ICF) [15,52].](http://dx.doi.org/10.1016/j.clon.2017.04.001)
Group score, the Musculoskeletal Tumour Society Score (a measure of impairment) and the Toronto Extremity Salvage Score (a measure of activity limitation). In future, objective measures of physical functioning (such as those derived from small body worn monitors containing accelerometers) may increasingly be included in routine assessment [59]. QoL measures include aspects of the physical, mental and social domains of life. There is a relationship between physical activity, participation restrictions and QoL: inability to participate in life roles has the largest impact on QoL [60]. After amputation, disability is strongly related to QoL [17]. Programmes to improve QoL therefore have to

![Fig 2. Examples of rehabilitation interventions.](http://dx.doi.org/10.1016/j.clon.2017.04.001)
consider improving physical performance, psychological distress and employment [61,62].

**Delivering Rehabilitation and Survivorship Care**

The four essential components of survivorship care are:

(i) Prevention of recurrent and new cancers, and other late effects;
(ii) Surveillance for cancer spread, recurrence or second cancers; assessment of medical and psychosocial late effects;
(iii) Intervention for consequences of cancer and its treatment, e.g. medical problems, symptoms, psychological distress and concerns related to employment, insurance and disability;
(iv) Rehabilitation: patients should have access to physiotherapy, occupational therapy, psychological support, prosthetic and pain services, and other services, including financial support [63].

Delivery clearly requires a multidisciplinary team, including rehabilitation, psychological support and sexual health services, expert pain management and return to work [4]. Coordination between specialist and non-specialist providers is essential to deliver appropriate care closer to home, although local services may be highly variable [22].

Rehabilitation services are required in the acute setting, before or after surgical care, in outpatients or following readmission in the specialist centre or closer to home. Care has to be meaningful to patients faced with many appointments, with different teams, over a prolonged period.

In the acute setting, enhanced recovery programmes (involving measures such as locoregional anaesthesia and compressive bandaging of the limb) may reduce hospital stay [64]. A system for central coordination of care for bone sarcoma patients delivered close to home has been proposed and may be helpful for soft tissue sarcoma survivors [65]. In it, rehabilitation is supervised by a specialist care coordinator with telephone consultations and remote supervision of care delivered by non-specialists. Better online information can help spread good practice [66].

Readmission for inpatient rehabilitation may be of some benefit. A 4 week programme for children and adolescents showed improvements in gait cadence variables in patients with sarcoma and an immediate and sustained impact on QoL [67].

After treatment, wellness days in which services including physiotherapy, occupational therapy, pain management, benefits advice and psychotherapy are available and patients can receive peer support are helpful.

**Discussion**

The survivorship and rehabilitation needs of patients treated for extremity soft tissue sarcoma are diverse but important parts of service delivery. Patient-centred and holistic rehabilitation programmes embedded throughout care can have a positive effect on survivorship. However, delivery within financially challenged health care systems may be difficult.

Understanding of the survivorship experience has improved over the last two decades and the care of survivors has gained prominence. The challenge to translate this into improved services and treatments remains. The ICF model is a useful tool for capturing and interpreting the dimensions of survivorship and will be enhanced by the evidence-based use of objective measures of physical functioning, as technologies such as body worn monitors are integrated into clinical practice. This model supports the targeting of impairments that underpin poor global outcomes (such as balance or gait training).

A further challenge is to communicate differences in the survivorship experience to patients early in treatment in a way that supports shared decision making. Formal elicitation of priorities about future outcomes and matching these to available treatments in a systematic and evidence-based way has not yet become normal practice in sarcoma treatment.

The diversity of clinical presentations, variation in treatments, demographic and other factors makes generalisation about survivorship difficult. Most studies rely on small sample sizes and differing designs that may introduce bias and many studies rely on historical cohorts that do not reflect contemporary clinical practice. This variation demands a systematic approach to assessment leading to individualised rehabilitation strategies.

**Future Directions**

The next few years may see incremental reductions in the morbidity of treatment that improve outcomes for survivors. Surgical resections guided by imaging and visualisation of margins may reduce the volume of normal tissue resected. Newer radiotherapy techniques may lead to less limb toxicity and fewer long-term effects. For example, the reduced toxicity of proton therapy could lead to less lymphoedema – postulated in the treatment of axillary lymph nodes in breast cancer [68].

In parallel, evidence-based developments in rehabilitation are needed, including prehabilitation, falls prevention and management and remote monitoring. These are suitable for multicentre clinical trials informing service provision and management.

**Conclusion**

The treatment of extremity soft tissue sarcoma has a lasting impact as patients attempt to return to normal living. Physical impairments, disabilities and pain dominate the experience of many. Services need to be personalised and comprehensive, including aspects that may be difficult to provide in some health systems. The refinement of new treatments should not only consider the impact on the disease, but also on survivorship. At the time of diagnosis and treatment planning, patients should be closely involved...
in shared decisions about treatment considering their effectiveness and the impact on the survivorship experience. Further research is needed to develop the evidence base for rehabilitation interventions.

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References


