Bone and soft tissue sarcomas of the trunk wall and extremities amount to approximately 1% of all malignancies (Gustafson 1994). Due to the low frequency and complex management, it is generally recommended that such patients be referred to tumor centers for treatment by a multidisciplinary team mainly including orthopedic surgeons, oncologists, radiologists, cytologists and pathologists. This concept has been generally adopted in Scandinavian countries (Rydholm 1983; Wiklund et al. 1996).

In 1986, the Scandinavian Sarcoma Group (SSG) started a register of patients with sarcoma who had been referred to tumor centers in Finland, Norway, and Sweden. The aim was to create a large database of patients treated during a limited period, according to uniform accepted guidelines, for comparison with non-SSG centers, to assess how SSG treatment guidelines were implemented and to permit multicenter studies of patients with rare tumors.

This report is based on patients with bone (BS) or soft tissue sarcoma (STS) of the extremities or trunk wall diagnosed between March 1, 1986 and December 31, 1997 and reported to the SSG Register. Data of patients entered in SSG treatment trials of osteosarcoma (SSG II, SSG VIII, ISG/SSG I) or Ewing’s sarcoma (SSG IV, SSG IX) are also included in the SSG Register. Patients with STS located in the head and neck, viscera or retroperitoneal space were not included nor were patients with Kaposi’s sarcoma or dermatofibrosarcoma protuberans. Data were reported from all sarcoma centers in Norway (3) and Sweden (5). Patients from Helsinki University Hospital have been reported 1988–93 to the Register and from Turku and Tampere University Hospitals regularly since 1994 (Table 1).

Patient accrual

3152 patients were registered. There were 1031 patients with bone sarcomas and 2121 with soft tissue sarcomas.

Site and location

Among BS, the commonest sites were the femur, tibia and humerus and among STS, the thigh, trunk wall and lower leg (Table 2). Only 20% of BS were intracompartmental as compared to 63% of STS. Among intracompartmental STS, approximately half were subcutaneous and half intramuscular.

Metastatic status

At diagnosis metastases were found in 13% of BS patients and 9% of STS. Among BS, metastases were especially common in malignant fibrous histiocytoma (24%) and osteosarcoma (17%) and among STS in rhabdomyosarcoma (30%), angiosarcoma (21%) and unclassified sarcoma (21%).

**Table 1. Participating centers and number of patients reported**

<table>
<thead>
<tr>
<th>Center</th>
<th>BS</th>
<th>STS</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Finland</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Helsingfors University Hospital</td>
<td>152</td>
<td>184</td>
<td>336</td>
</tr>
<tr>
<td>Tammerfors University Hospital</td>
<td>7</td>
<td>29</td>
<td>36</td>
</tr>
<tr>
<td>Turku University Hospital</td>
<td>7</td>
<td>25</td>
<td>32</td>
</tr>
<tr>
<td>Norway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Radiumhospital, Oslo a</td>
<td>234</td>
<td>438</td>
<td>672</td>
</tr>
<tr>
<td>Haukeland Univ. Hospital, Bergen</td>
<td>72</td>
<td>126</td>
<td>198</td>
</tr>
<tr>
<td>Regional Hospital, Trondheim</td>
<td>30</td>
<td>84</td>
<td>114</td>
</tr>
<tr>
<td>Sweden</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Karolinska Hospital, Stockholm</td>
<td>216</td>
<td>464</td>
<td>680</td>
</tr>
<tr>
<td>Sahlgrenska Hospital, Gothenburg</td>
<td>179</td>
<td>344</td>
<td>523</td>
</tr>
<tr>
<td>University Hospital, Lund</td>
<td>70</td>
<td>273</td>
<td>343</td>
</tr>
<tr>
<td>University Hospital, Umeå</td>
<td>35</td>
<td>77</td>
<td>112</td>
</tr>
<tr>
<td>University Hospital, Linköping</td>
<td>24</td>
<td>77</td>
<td>101</td>
</tr>
<tr>
<td>Total</td>
<td>1031</td>
<td>2121</td>
<td>3152</td>
</tr>
</tbody>
</table>

a includes patients operated at Ullevål Hospital, Oslo
Table 2. Site of BS and STS

<table>
<thead>
<tr>
<th>Site BS</th>
<th>n</th>
<th>%</th>
<th>Site STS</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
</table>
| Vertebra | 44| 4% | Trunk wall | 306| 15%
| Sacrum  | 68| 7% | Gluteal  | 138| 6%
| Pelvis   | 156| 15% | Groin  | 89| 4%
| Rib  | 64| 6% |
| Clavicle | 6| 1% | Shoulder | 119| 6%
| Scapula | 31| 3% | Upper arm | 141| 7%
| Humerus | 94| 9% | Elbow  | 42| 2%
| Radius | 10| 1% | Lower arm | 96| 5%
| Ulna  | 8| 1% | Hand  | 35| 2%
| Hand  | 10| 1% |
| Femur | 355| 34% | Thigh | 710| 33%
| Tibia | 136| 13% | Knee  | 106| 5%
| Fibula | 35| 3% | Lower leg | 251| 12%
| Foot  | 15| 2% | Foot  | 82| 4%

Table 3. Histotypes

<table>
<thead>
<tr>
<th>Sarcoma</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone sarcoma</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| Osteosarcoma                 | 321| 31%
| Chondrosarcoma               | 320| 31%
| Ewing sarcoma/PNET           | 145| 14%
| Malignant fibrous histiocytoma | 65| 7%
| Chordoma                     | 52| 5%
| Malignant giant cell tumor   | 34| 3%
| Parosteal osteosarcoma       | 21| 2%
| Osteosarcoma, other subtypes | 18| 2%
| Fibrosarcoma                 | 9| 1%
| Leiomyosarcoma               | 8| 1%
| Other types                  | 43| 4%
| Soft tissue sarcoma          |   |   |
| Malignant fibrous histiocytoma | 845| 40%
| Liposarcoma                  | 279| 13%
| Leiomyosarcoma               | 173| 8%
| Synovial sarcoma             | 166| 8%
| Malignant schwannoma         | 104| 5%
| Fibrosarcoma                 | 74| 4%
| PNET/Ewing                   | 41| 2%
| Myxoid chondrosarcoma        | 36| 2%
| Rhabdomyosarcoma             | 33| 2%
| Malignant pericytoma         | 27| 1%
| Osteosarcoma                 | 26| 1%
| Epithelioid cell sarcoma     | 24| 1%
| Clear cell sarcoma           | 21| 1%
| Hemangiopericytoma           | 15| 1%
| Alveolar cell sarcoma        | 10| 1%
| Others and unclassified      | 153| 7%

Table 4. Referral status in percentages

<table>
<thead>
<tr>
<th>Referred after</th>
<th>BS</th>
<th>STS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Untouched lesions</td>
<td>75</td>
<td>41</td>
</tr>
<tr>
<td>Fine needle biopsy</td>
<td>5</td>
<td>13</td>
</tr>
<tr>
<td>Coarse needle/cut biopsy</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Incisional biopsy</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>Excision</td>
<td>6</td>
<td>27</td>
</tr>
<tr>
<td>Local recurrence</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

Histological type and grade

The histological types and malignancy grades presented are those reported by the different contributing centers. The commonest histotypes among BS were osteosarcoma (31%), chondrosarcoma (31%) and Ewing’s sarcoma (14%) (Table 3). Among STS, malignant fibrous histiocytoma (45%) and liposarcoma (14%) were the only histotypes accounting for more than 10% of the lesions.

Histological grading of STS on a 4-grade scale (Broders et al. 1939, Angervall et al. 1986), based on biopsy or surgical specimens, showed that 78% of STS were of high grade (III–IV). Among chondrosarcoma, 26% were grade I, 42% grade II and 32% grades III–IV.

Referral

The SSG recommends referral to tumor centers of all patients suspected of having a sarcoma without prior open biopsy or excision. Specifically, patients having solitary bone lesions, without other malignancy or deep–seeded soft tissue tumors or those with subcutaneous lesions larger than 5 cm should be referred.

84% of BS patients, but only 58% of STS patients were referred to a SSG center before open biopsy or surgical treatment (Table 4). 10% and 17%, respectively, of patients referred before surgery had a fine– or coarse–needle biopsy before referral. Interestingly, older BS patients tended more often to have been operated on before referral, whereas among STS patients, it was the younger patients who were less often referred primarily. Only 1/3 of patients with subcutaneous STS were referred before surgery as compared to 2/3 of those with deep lesions.

Biopsy

Surgical treatment without prior biopsy was performed in 60% of STS patients treated before referral and in 17% of SSG center treated patients.

Open biopsy was used to make a preoperative diagnosis in 50% of BS patients, followed by fine–needle aspiration cytology in 24% and by coarse–needle or true–cut biopsy in 15%. Cytology was the preferred method among STS patients.

Treatment

Treatment of patients with osteosarcoma and Ewing’s sarcoma patients according to SSG protocols, is discussed in separate chapters. Among 2121 STS patients, 1953 were treated surgically. 64% were operat-
ed on primarily at a SSG center. 23% were operated on outside but were referred and had a second operation for a primary tumor at a sarcoma center. Hence, only 11% of patients had all surgical treatment for a primary tumor outside a sarcoma center. Limb-sparing surgery for extremity STS was performed in 90% of the patients.

**Surgical margins**

In STS, a wide or compartmental (radical according to Enneking’s (1980) terminology) margin was achieved in only 15% of patients operated on outside a sarcoma center, as compared to 64% of those operated on at a center. The reported margins among center-operated patients were 7% intralesional, 29% marginal, 59% wide, and 5% compartmental.

**Adjuvant treatment**

Postoperative radiotherapy in STS has been recommended after a marginal margin. Radiotherapy was also indicated after an intralesional margin if the margin could not be improved by further surgery. Adjuvant radiotherapy was not generally considered indicated after wide or compartmental margins (Alho et al. 1989). Overall, 25% of operated STS patients had adjuvant radiotherapy. Among patients with an intralesional or marginal surgical margin, 54% had postoperative radiotherapy. The SSG had no recommendations or trials during the study period for adjuvant chemotherapy of primary STS. Adjuvant chemotherapy was reported in only 5% of operated STS patients.

**Follow-up**

Follow-up has been reported in 85% of BS and 92% of STS patients. Among both BS and STS patients, 42% had died, 5% were alive with disease and 53% showed no evidence of disease at the last follow-up. The median follow-up of patients still alive was 5 years.

**Comparison among centers**

Comparison of data reported from participating centers revealed large differences in demography and treatment (Table 5). All patients treated for STS in the Southern Health Region of Sweden, whether treated at the center in Lund or at other hospitals, were reported from Lund, and their data can be used as a basis for comparison, since it is population-based. The patients from Lund were older than at other centers. Patients from Helsinki were younger and also had smaller lesions. Centers in Sweden received two thirds of their patients without prior surgery and in Bergen, the referral practice was almost as good. In Oslo and Helsinki, however, only one third of the patients were referred before surgery. The amputation rate in Gothenburg was twice as high as in other centers and, conversely, Gothenburg reported better surgical margins.

**Discussion**

The SSG Register aimed to assess treatment and results in all sarcoma patients treated at SSG centers, not only those participating in clinical trials. We also wished to assess how SSG guidelines were followed and whether compliance had led to better results and, finally, to have a large data-bank to analyze specific questions.

Patients were reported to the Register from SSG centers in Finland, Norway and Sweden, but participation was not compulsory. Since the SSG Register also includes the population-based series from Lund we can detect differences in referral patterns in various regions of Scandinavian (Rydholm 1983, Röös 1987, Gustafson 1994). In Sweden and Norway, all inhabitants were encompassed by SSG centers, whereas in Finland, reporting has been inconsistent. Patients are now also being reported from Herlev, Denmark. One major concern regarding tumor registers is the quality of the data. This has been ensured by consensus on terminology in SSG and retrospec-

### Table 5. Key data from centers reporting more than 100 STS patients in percentages

<table>
<thead>
<tr>
<th>Center</th>
<th>Age &gt;65 years</th>
<th>Referred before surgery</th>
<th>Subcutaneous</th>
<th>Size &gt;7cm</th>
<th>Amputation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stockholm</td>
<td>45</td>
<td>68</td>
<td>34</td>
<td>41</td>
<td>7</td>
</tr>
<tr>
<td>Oslo</td>
<td>43</td>
<td>33</td>
<td>38</td>
<td>51</td>
<td>8</td>
</tr>
<tr>
<td>Gothenburg</td>
<td>46</td>
<td>70</td>
<td>29</td>
<td>47</td>
<td>14</td>
</tr>
<tr>
<td>Lund</td>
<td>55</td>
<td>73</td>
<td>32</td>
<td>43</td>
<td>5</td>
</tr>
<tr>
<td>Helsinki</td>
<td>27</td>
<td>27</td>
<td>36</td>
<td>39</td>
<td>8</td>
</tr>
<tr>
<td>Bergen</td>
<td>44</td>
<td>56</td>
<td>41</td>
<td>44</td>
<td>9</td>
</tr>
</tbody>
</table>
tive comparison of recorded data with patient charts. The principle of referring BS patients to SSG centers before surgery or biopsy is closely adhered to in Scandinavia. In some centers older patients are operated on before referral because the clinician presupposes a metastasis and does not suspect a primary malignant lesion. The referral practice in STS is less well established, especially in Finland and parts of Norway. Sweden has borne the fruits of more than 20 year’s experience in southern Sweden to centralize STS treatment in Lund (Rydholm 1983). The referral practice in the Scandinavian countries compares favorably to that in the U. S. A. (Mankin et al. 1996).

Since peer review has not been concluded, the histological diagnoses were those made at the different centers. The results of retrospective analysis by the SSG Pathology Board are presented separately, but show that the validity of the histopathological diagnostics is high. Patients with osteosarcoma and Ewing’s sarcoma, entered in SSG trials, are continuously being reviewed as regards diagnosis, surgical margins chemotherapy.

Fine-needle biopsy for cytological diagnosis of soft tissue lesions was developed and popularized in Sweden and has also been generally used in Norway (Åkerman et al. 1985). An increasing use of such biopsies at local hospitals may be one reason for the high referral rate before surgery to SSG centers in Sweden; Swedish surgeons refrain from open biopsies or excisions without a cytological diagnosis.

Accrual of new patients, validation of reported data and continuous follow-up should be ensured, as the SSG Register is used for clinical studies of musculoskeletal neoplasia. Projects based on the SSG Register, such as on local recurrence of STS and synovial sarcoma, provide a quality controlled basis for treatment protocols regarding diagnostics guidelines, surgical treatment and adjuvant chemo- and radiotherapy.

Acknowledgments
SSG secretaries, Evy Nilsson and Ingrid Dahlberg, were responsible for data management. The SSG Register was supported by the Nordic Cancer Union, the Swedish Cancer Foundation and the Swedish National Board of Health and Welfare.


