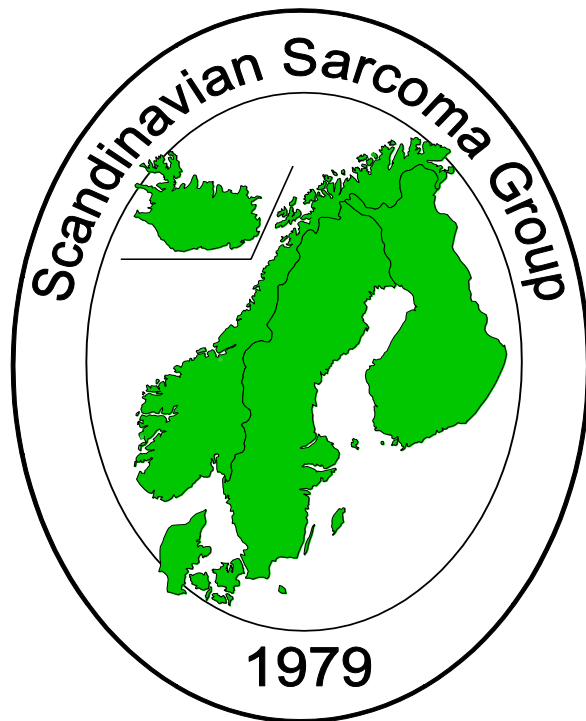


**The Scandinavian Sarcoma Group
annual report
on extremity and trunk wall
soft tissue and bone sarcomas
2012-2016**



The SSG annual report on extremity and trunk wall soft tissue and bone sarcomas.

The Scandinavian Sarcoma Group has a long-standing tradition of monitoring sarcoma diagnostics and treatment in population based prospective registers. The Scandinavian register was established in 1986 and has been collecting data continuously. The past couple of years, the participating countries have developed independent national quality registers. Throughout the process, a close collaboration has maintained conformity in the registered parameters. The data in this report are pooled from Denmark's, Iceland's, Norway's and Sweden's national quality registers on sarcoma diagnostic and treatment. The report focuses on diagnostics, referral patterns and tumor characteristics. Some treatment data is included but no data on outcome.

Each national register has extracted its own data and thereafter the data has been merged. For some parameters the registers have slight variations in how data is recorded as explained below.

The national registers have a high coverage of all sarcoma cases within the entire populations. In Sweden for instance, the completeness of the national quality register for extremity and trunk wall sarcomas was 92% in 2015 and 88% for 2016. Over time, we have learnt that there is a delay in registration why the coverage will probably rise to above 90% also for 2016.

In this report, only data on extremity and trunk wall sarcomas are included. From next year, also data on retractorperitoneal and visceral sarcomas will be included.

Due to the rarity of sarcomas, some data is presented per center and year while other parameters are pooled into center data for the entire time period.

The SSG Central Register Committee

Emelie Styring (Chairman)

Peter Holmberg Jørgensen

Halldór Jónsson jr

Olga Zaikova

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Soft tissue sarcomas

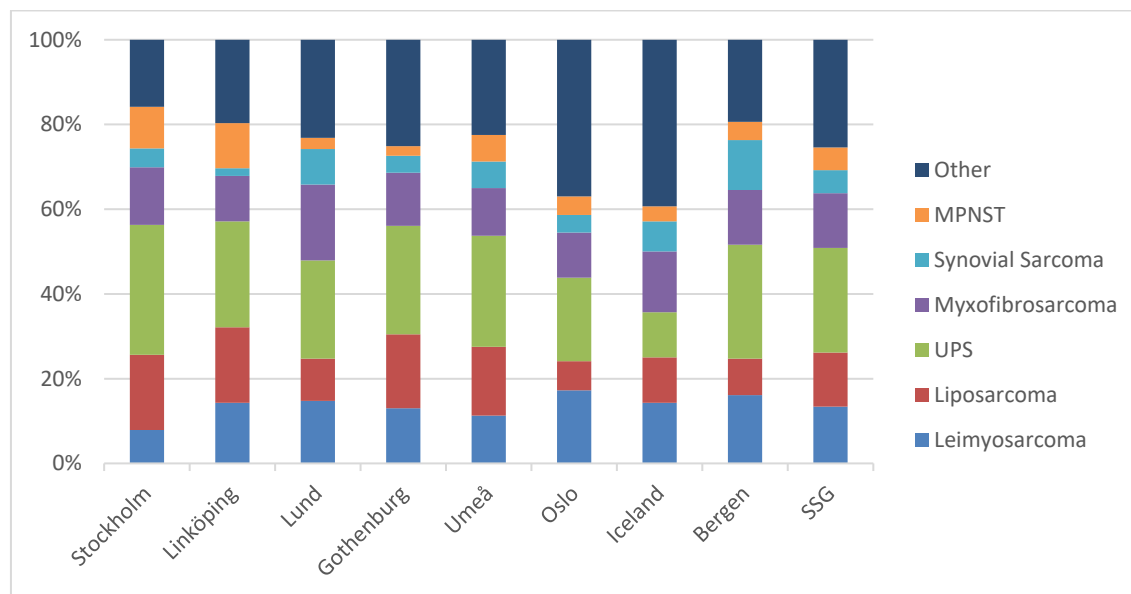
The initial part of the annual report describes key terms for the entire time period, 2012-2016, per center. The data is not presented for each center and year since each group would be too small for analysis.

Due to different ways of recording data this part of the report includes the numbers from Iceland, Norway and Sweden but not Denmark.

Totals per center 2012-2016

Histotypes

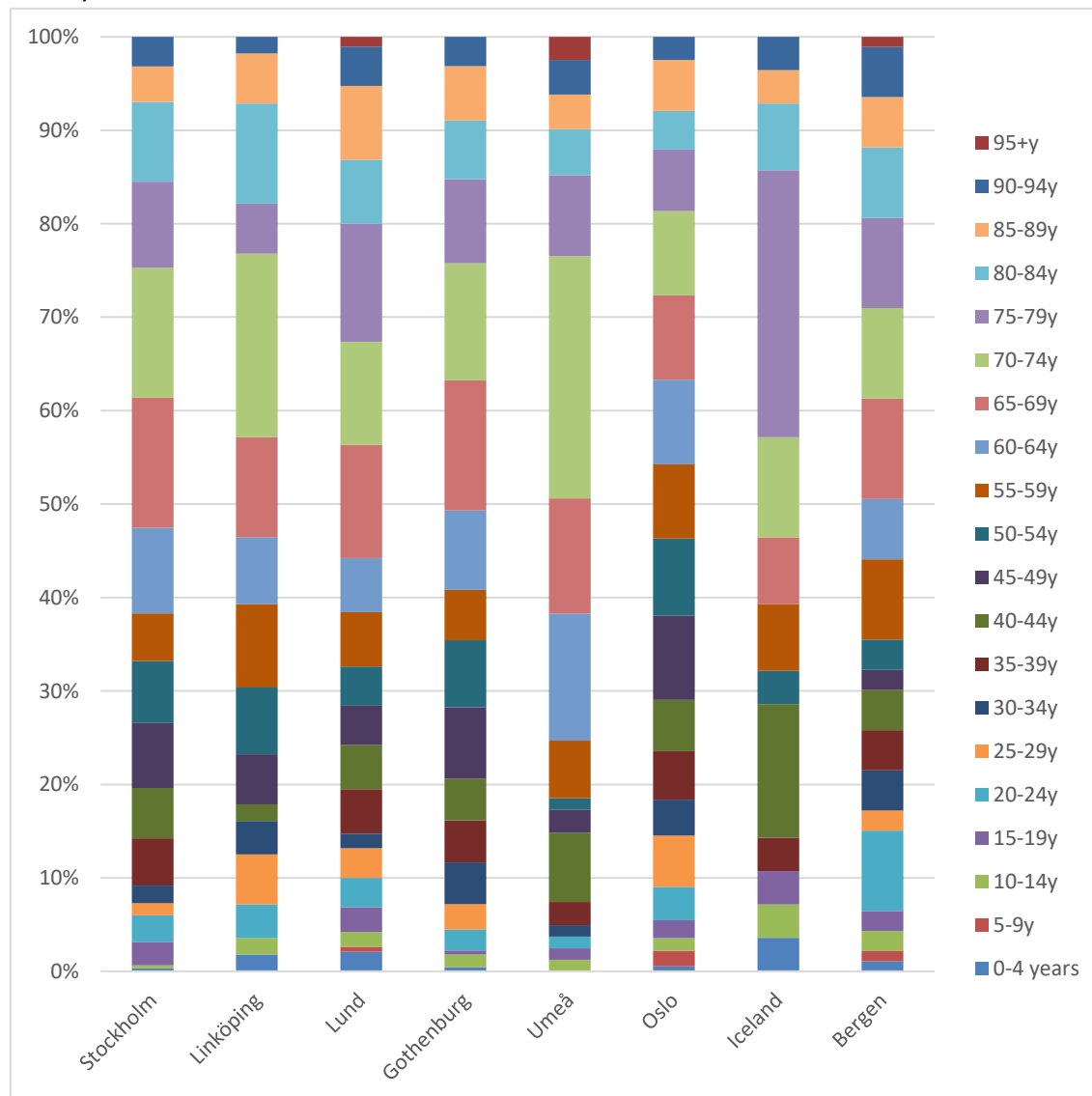
Soft tissue sarcomas (STS) are classified according to the WHO Classification of Tumours of Soft Tissue and Bone. World Health Organization classification of tumours, 4th edition¹. Due to the rarity of certain tumor types only the most common types are presented per center. Other tumor types, which may occur only in one case per year or less in all of Scandinavia, are presented together as "Other".



Liposarcoma includes myxoid liposarcoma, dedifferentiated liposarcoma and pleomorphic liposarcoma but not atypical lipomatous tumor.

Age at diagnosis

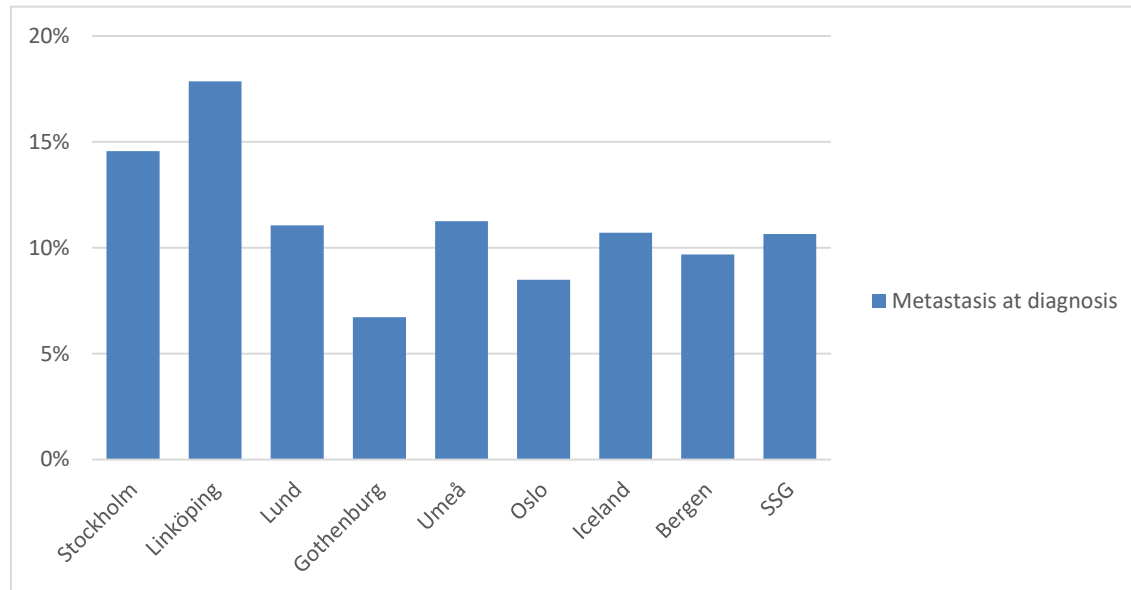
STs are most common in patients 50 years old or older as shown below. The patients' ages at diagnosis is fairly similar in distribution between the different centers.



Metastasis at diagnosis

On average, 18% of all STS patients presented with metastatic disease of those diagnosed between 2012-2016.

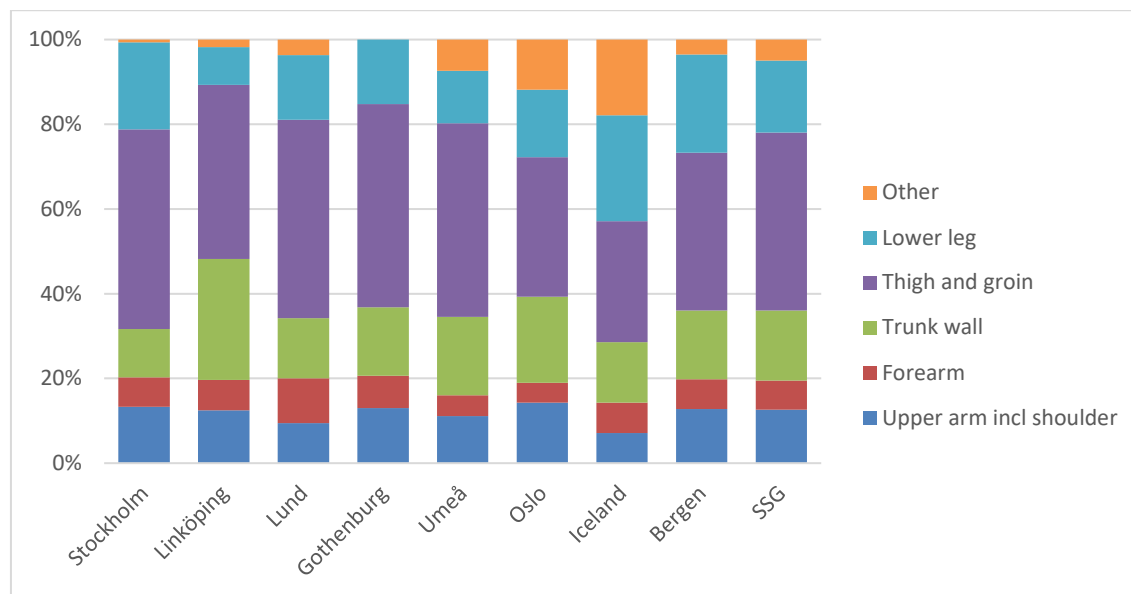
Please note that the y-axis ends at 20%.



Tumor site

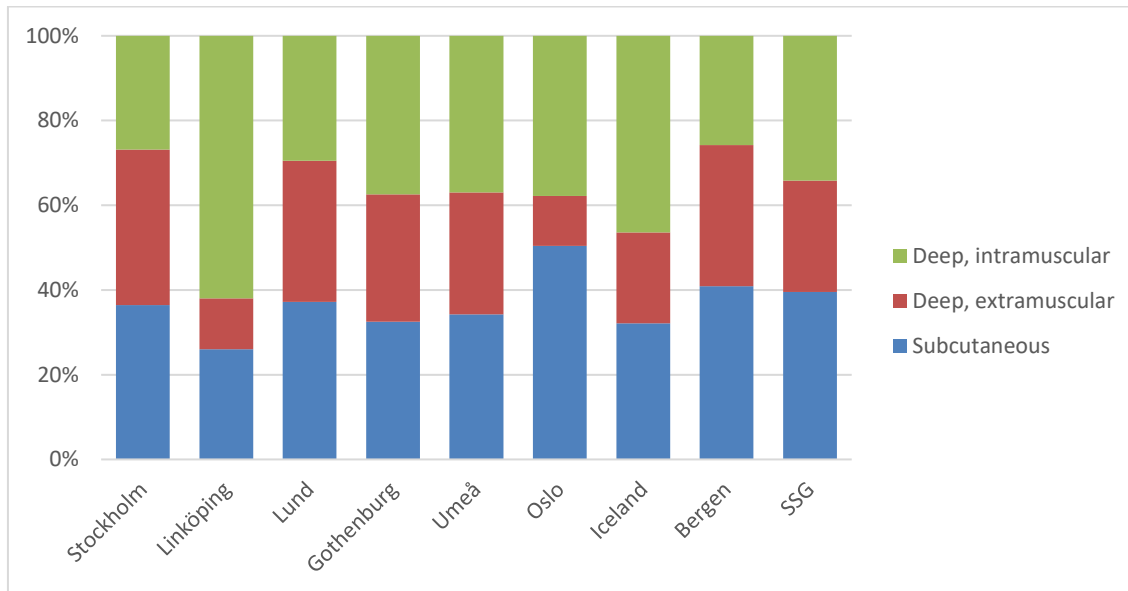
The relative distribution of tumor sites varies between the centers mainly because the data from Oslo and Iceland includes head and neck STS also in this parameter.

Among extremity and trunk wall STS, the most common tumor site is the thigh, hip and groin area as expected.



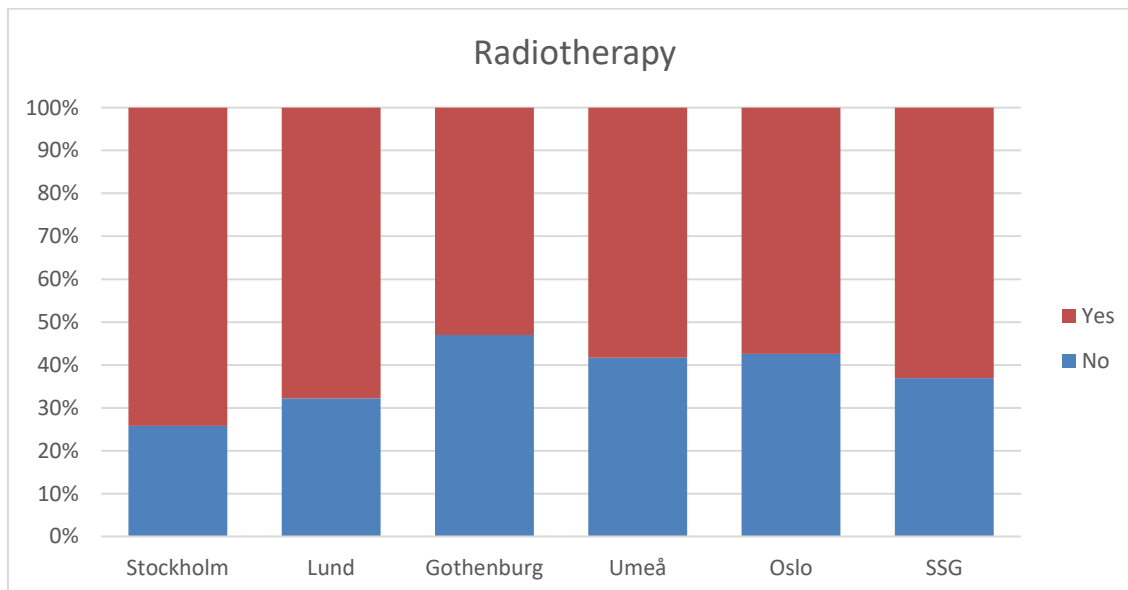
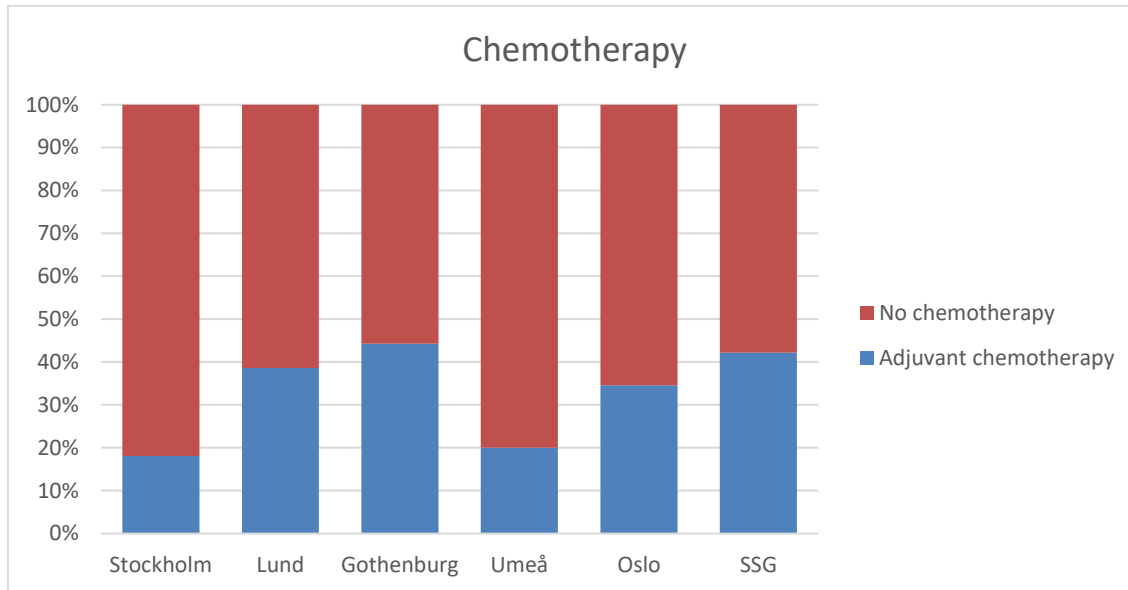
Tumor depth

The proportion of deep tumors is similar at all larger sarcoma centers. That 1/3 of the tumors are subcutaneous implies that the registers are truly population based with a high level of completeness.



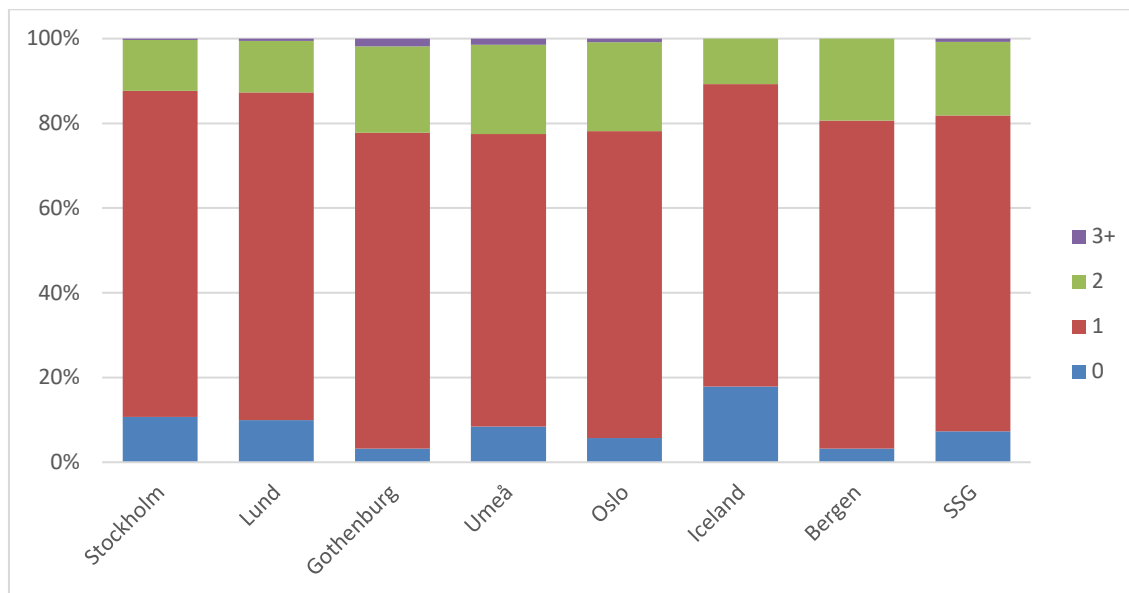
Adjuvant chemo- or radiotherapy

The use of neoadjuvant and adjuvant chemo- or radiotherapy for localized, high grade STS (grade 2 and 3) varies between the different centers. Currently, there is no ongoing SSG study on adjuvant chemotherapy for STS of the extremity and trunk wall which may, in part, explain the differences demonstrated. Data on radiotherapy refers to deep-seated tumors.



Number of surgeries

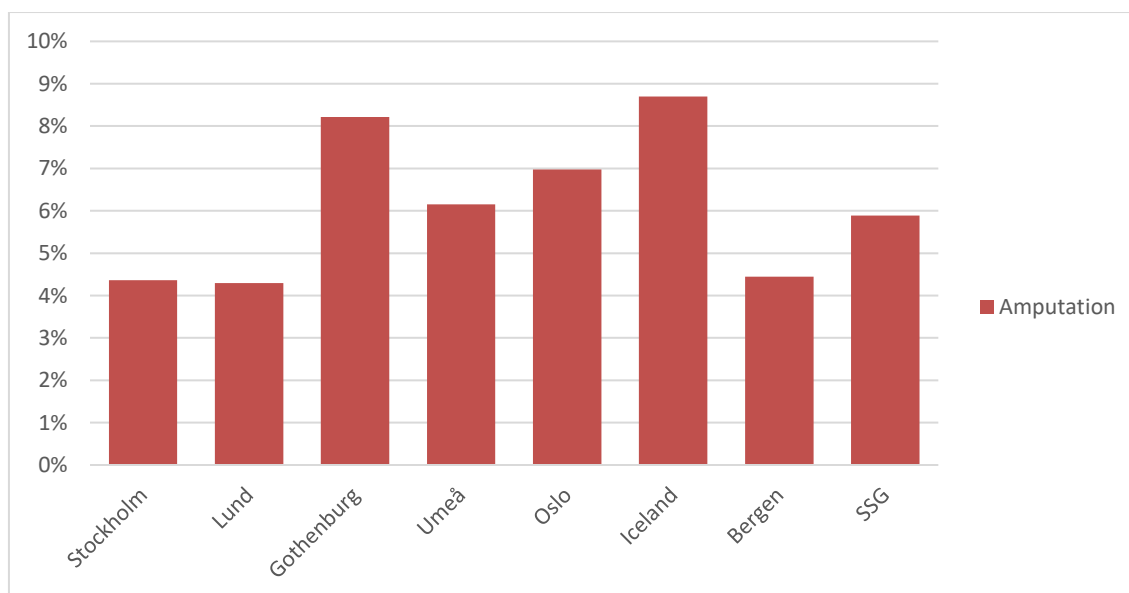
At all centers, most patients are treated with 1 surgery for their STS.



Only centers with reasonable reporting rates for this parameter are included in the diagram.

Type of operation (final surgery)

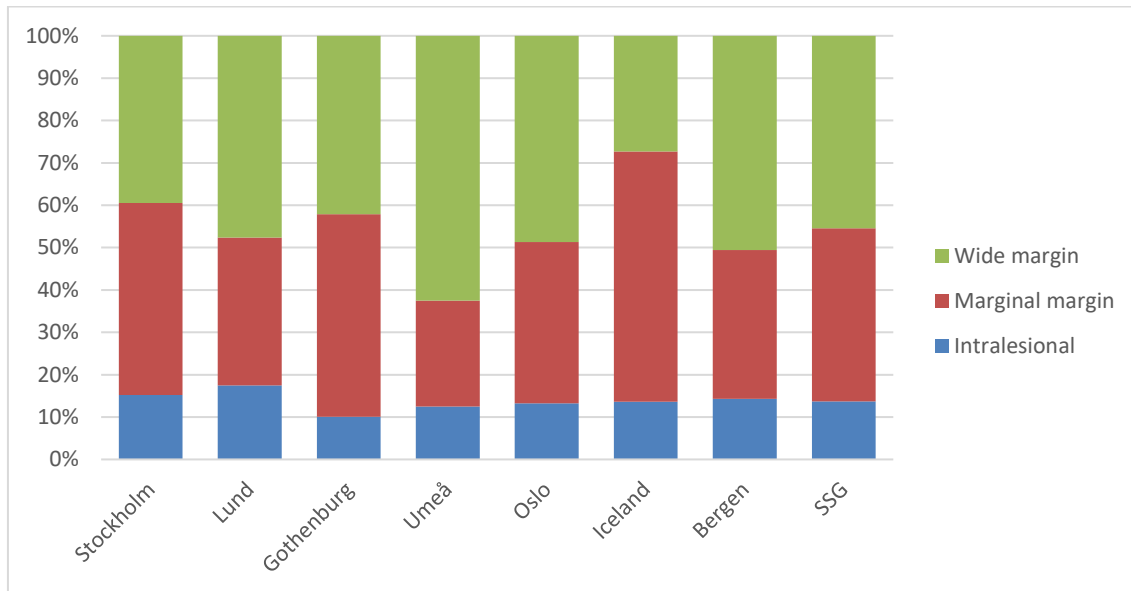
The amputation rate for STS is below 6 % in all of Scandinavia. It varies between 4 and 8 % from center to center but the actual number of patients undergoing amputation is low why a few extra cases one year may lead to an increased percentage. The differences between centers need to be monitored over time before any conclusions may be drawn. Please note that the y-axis ends at 10%.



Only centers with reasonable reporting rates for this parameter are included in the diagram.

Surgical margin

The final surgical margin, after 1 or more surgeries, is wide in more than one third of all cases. The rate of intralesional surgical margin is very similar between the different centers.



Only centers with reasonable reporting rates for this parameter are included in the diagram.

Data per center and year

Number of cases

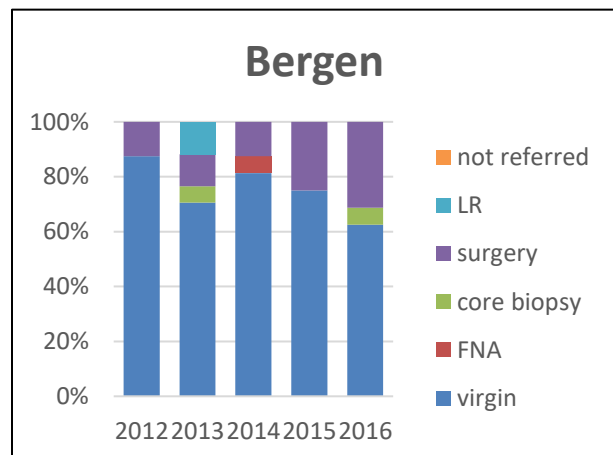
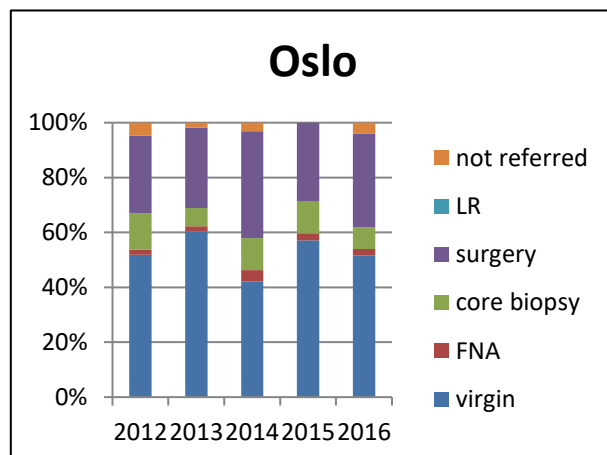
| | 2012 | 2013 | 2014 | 2015 | 2016 | Total |
|-------------------|------|------|------|------|------|-------|
| <i>Oslo</i> | 72 | 66 | 69 | 58 | 100 | 365 |
| <i>Bergen</i> | 24 | 17 | 16 | 20 | 16 | 93 |
| <i>Stockholm</i> | 77 | 58 | 55 | 82 | 44 | 316 |
| <i>Linköping</i> | | | 15 | 20 | 21 | 56 |
| <i>Lund</i> | 41 | 32 | 40 | 37 | 40 | 190 |
| <i>Gothenburg</i> | 38 | 40 | 50 | 53 | 42 | 223 |
| <i>Umeå</i> | 18 | 13 | 21 | 14 | 15 | 81 |
| <i>Iceland</i> | 6 | 2 | 6 | 9 | 5 | 28 |
| <i>Total</i> | 276 | 228 | 272 | 293 | 283 | 1352 |

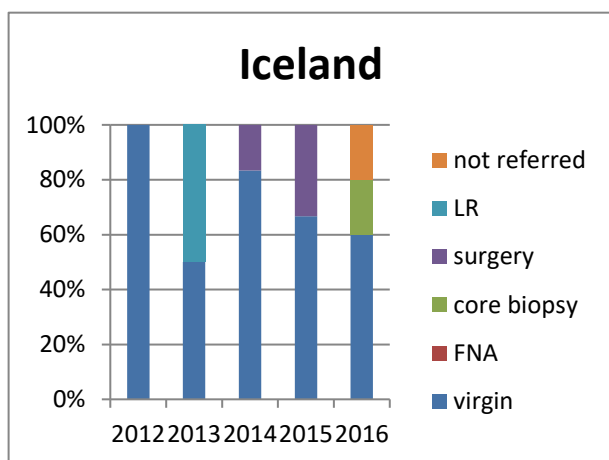
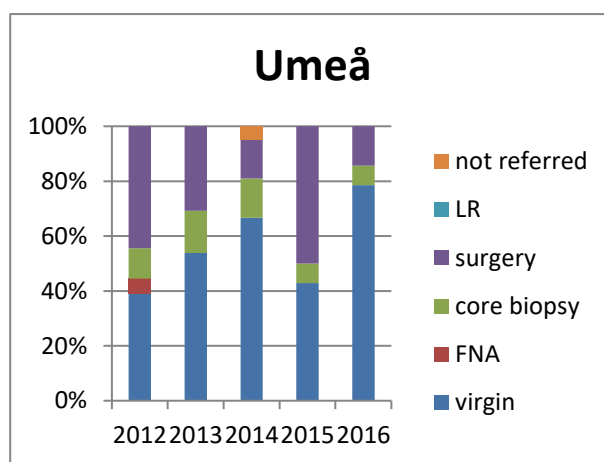
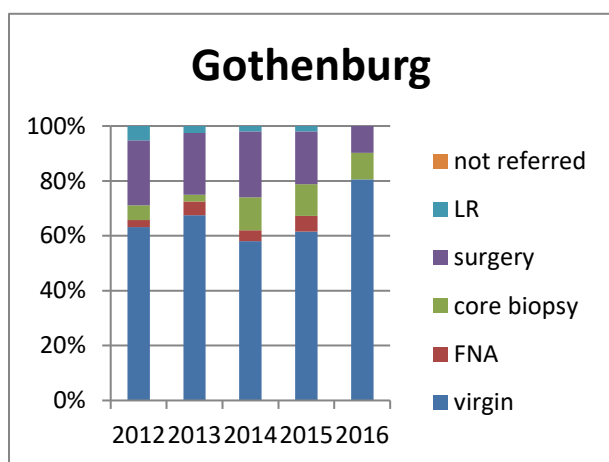
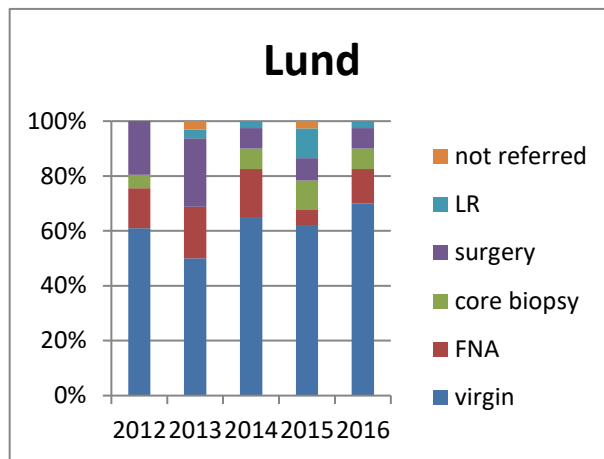
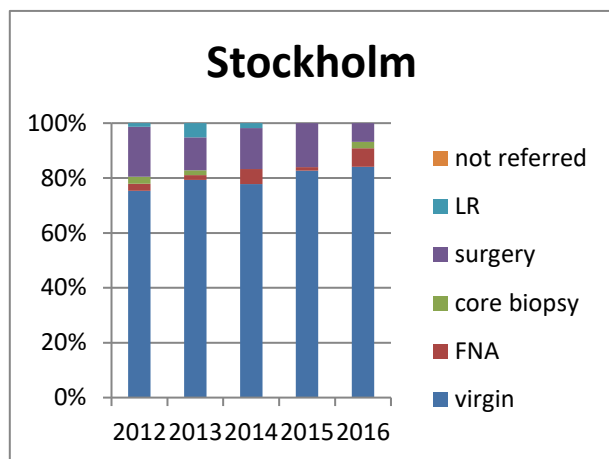
Referral pattern

The rate of patients referred as virgins (i.e. with untouched tumors) varies.

The SSG's referral guidelines recommend referral of all deep-seated lesions in the extremities and trunk wall, irrespective of size, and of superficial lesions larger than 5 cm. Thus small, superficial tumors may have been excised before referral and still have been referred according to guidelines.

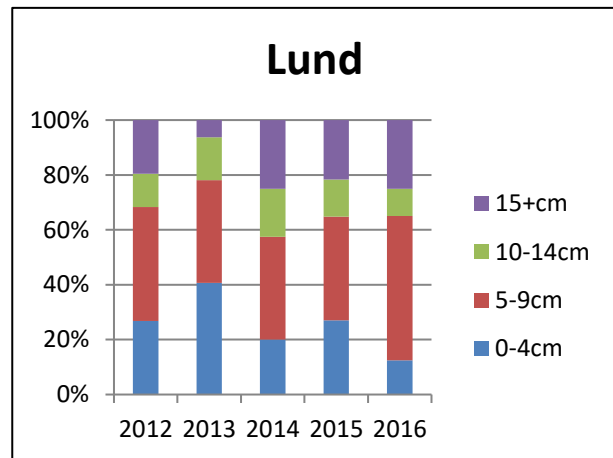
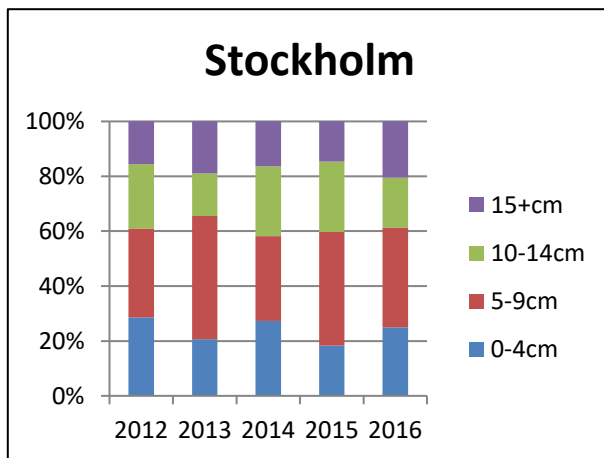
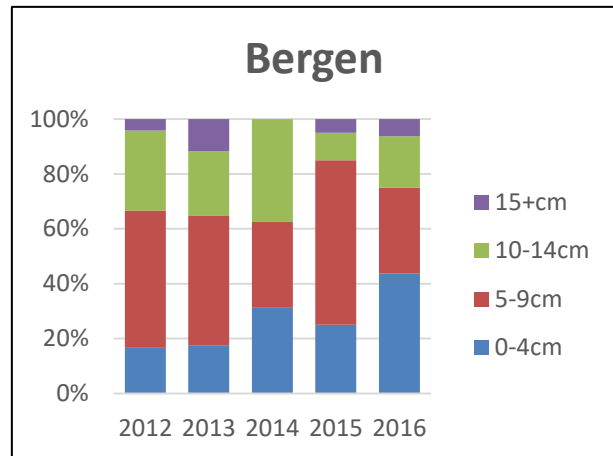
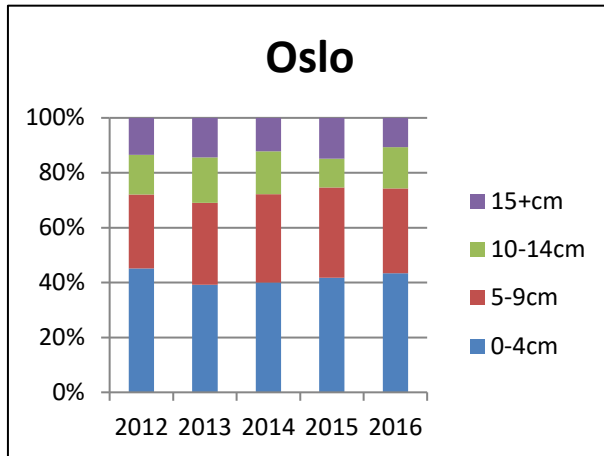
LR = local recurrence; FNA = fine needle aspiration; Virgin = referred with untouched tumor.

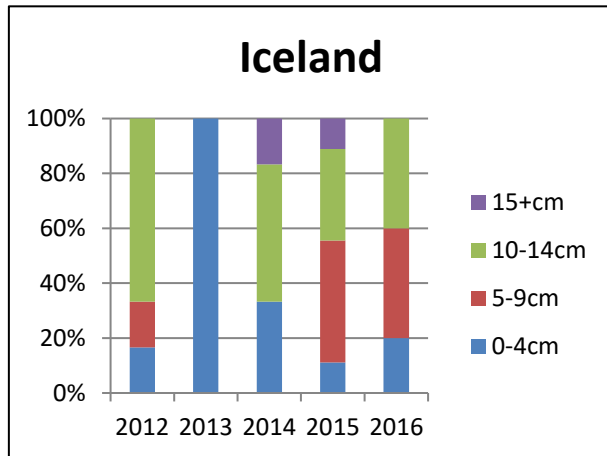
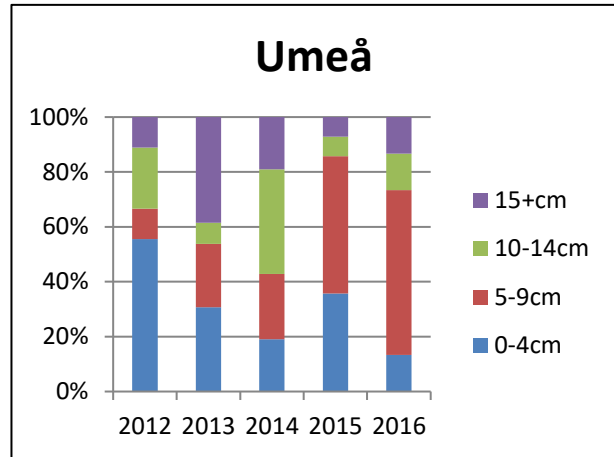
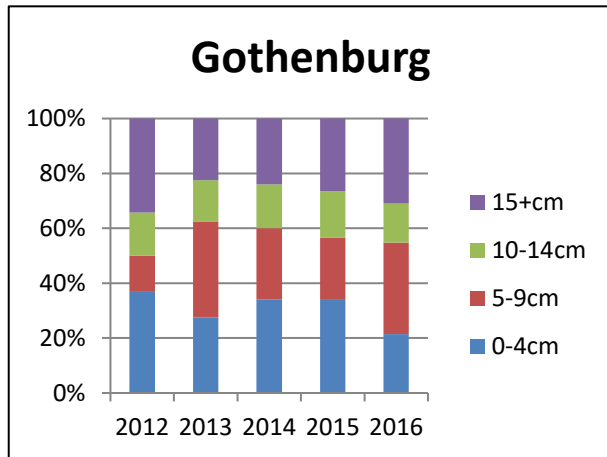




Tumor size

The distribution of tumor sizes over the years is fairly consistent for the larger centers. In general, the larger the center, the smaller the variation in tumor size from year to year.





Bone sarcomas

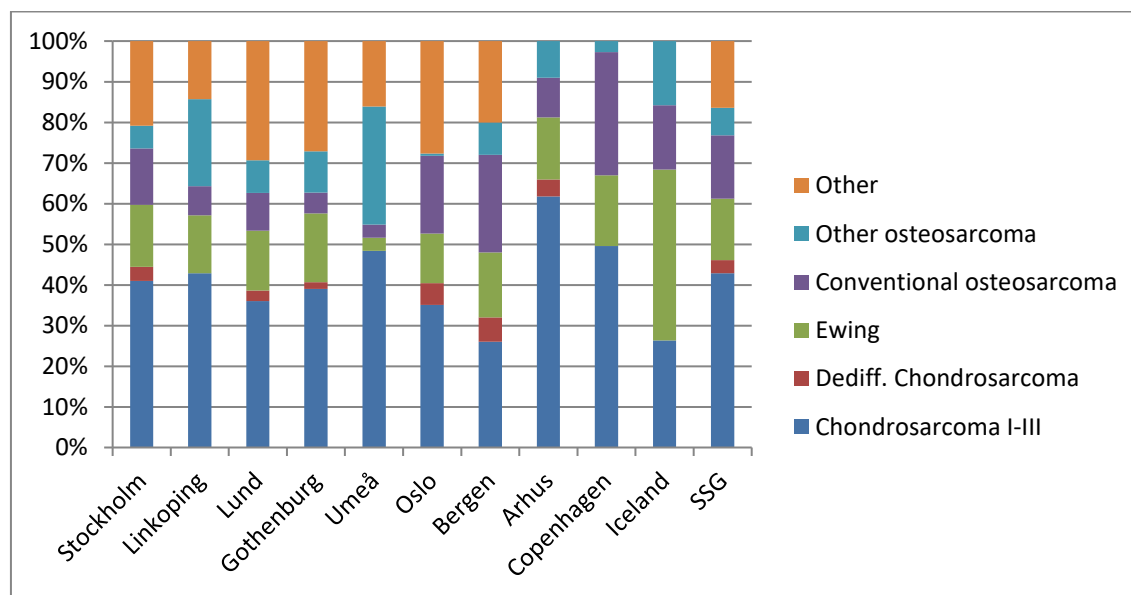
The first part of the report on bone sarcomas (BS) describes some key terms for the entire time period, 2012-16, per center. The data is not presented for each center and year since each group would be too small for analysis.

The data from Sweden does not include head and neck BS.

Totals per center 2012-2016

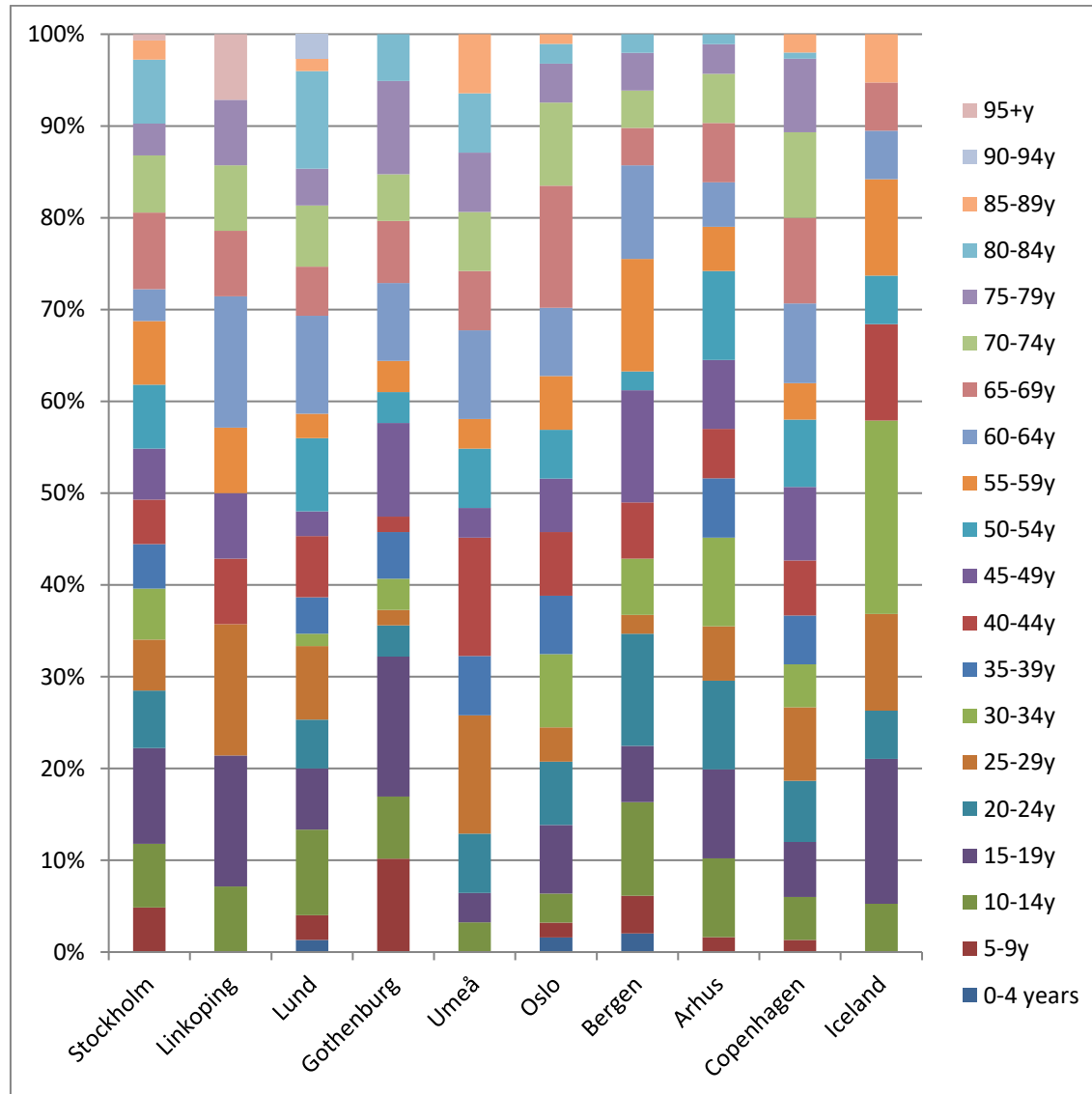
Histotypes

Bone sarcomas (BS) are classified according to the WHO Classification of Tumours of Soft Tissue and Bone. World Health Organization classification of tumours, 4th edition. Data on type of bone sarcoma per center for the larger tumor types is presented below. Rare entities are presented together as "Other".



Age at diagnosis

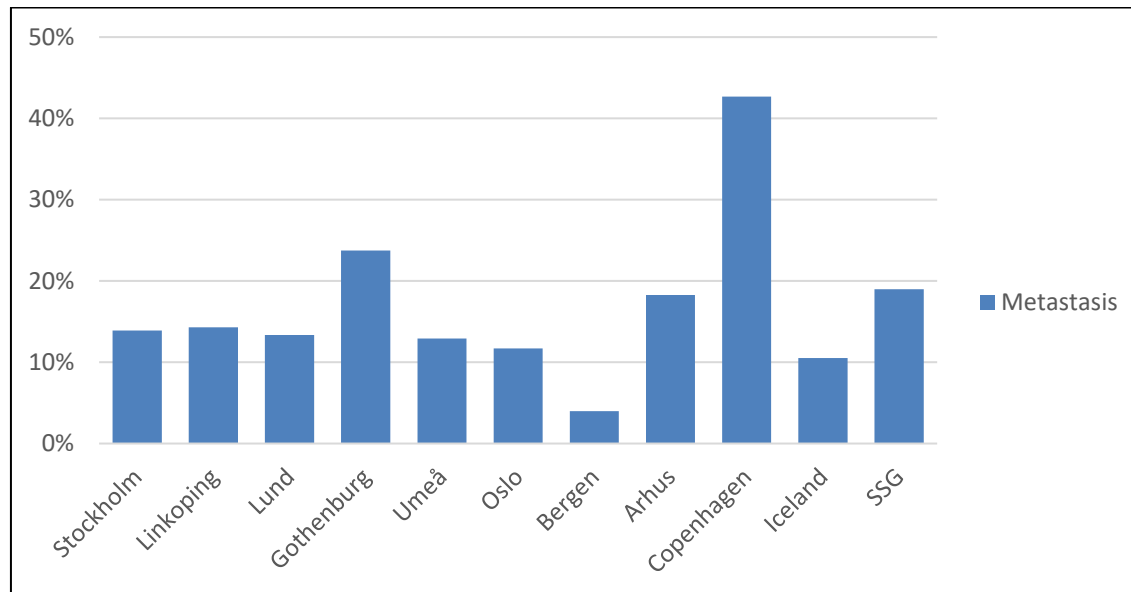
The age distribution on patients diagnosed with BS differs from that of those affected by STS. BS is more common among children and adolescents which is demonstrated below. In general, osteosarcoma and Ewing sarcoma are more common among children and adolescents while chondrosarcoma constitutes most of the cases in the later incidence peak around 60-80 years of age.



Metastasis at diagnosis

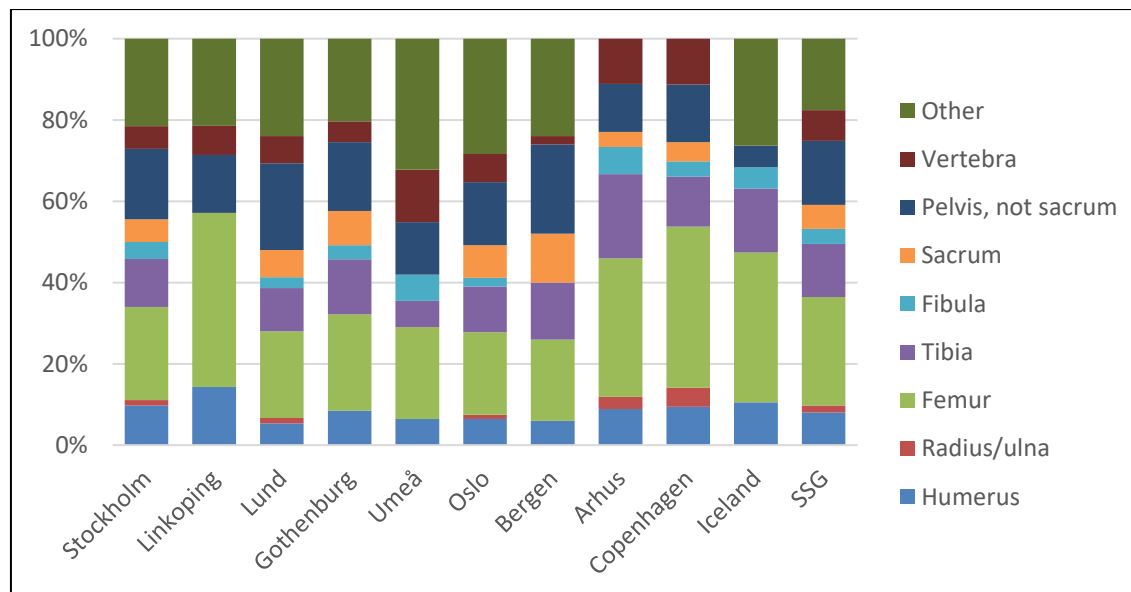
The rate of metastasis at diagnosis is a poor prognostic marker in BS. Overall, every fifth patient present with metastasis. The higher rate reported from Copenhagen merits further evaluation and of it persists over time.

Please note that the y-axis ends at 50%.



Tumor site

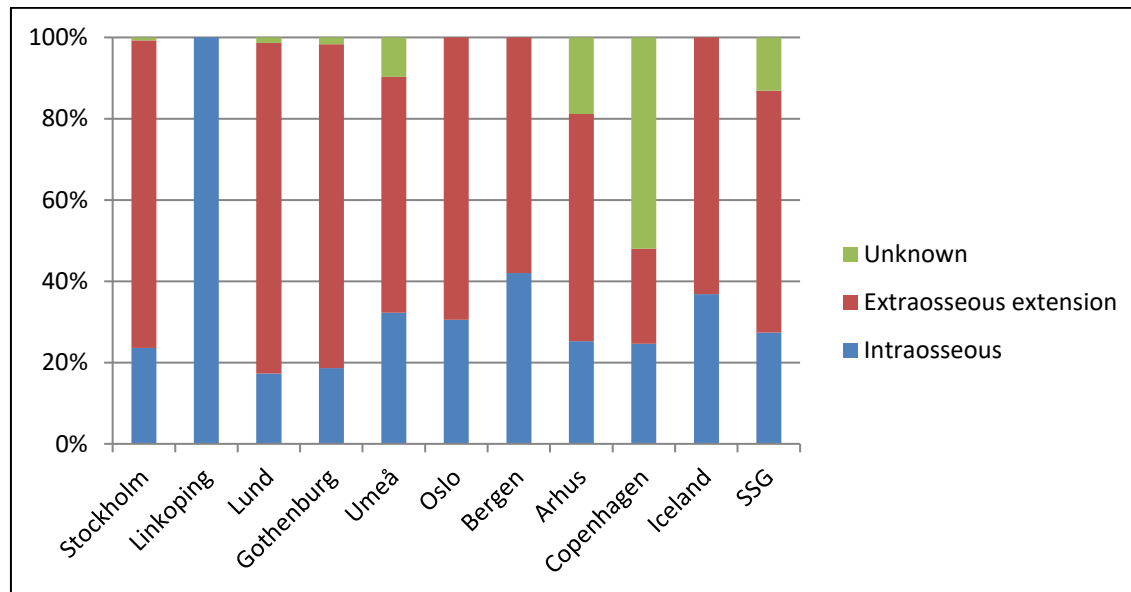
Most BS develop in the femur and the pelvis. "Other" includes several smaller bones, e.g. ribs, sternum and bones of the hand and feet.



Tumor location

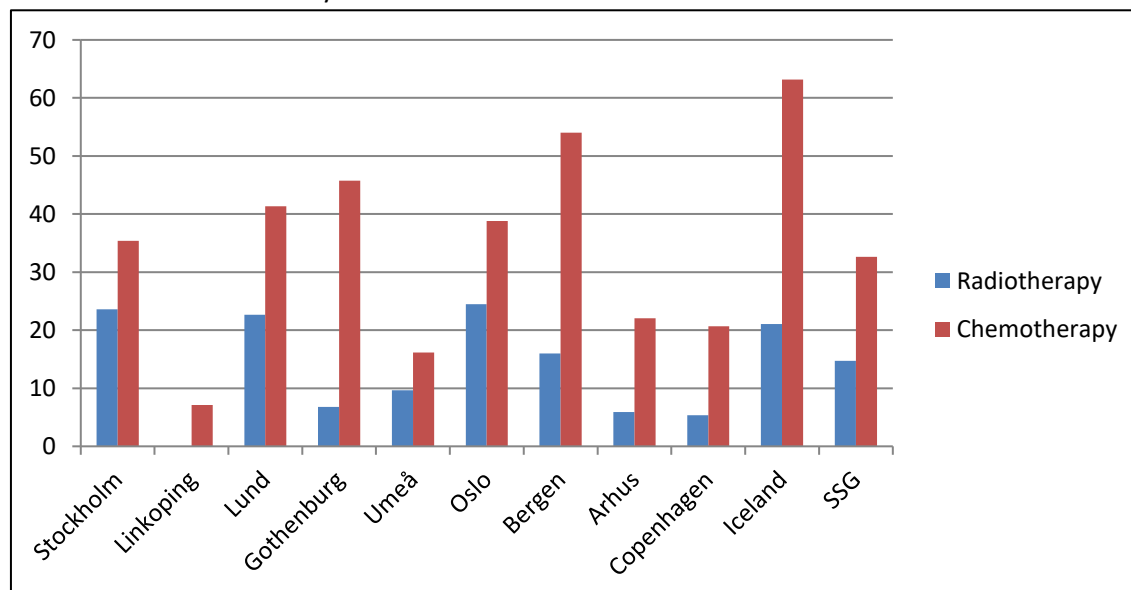
Just short of one third of all BS cases present with intraosseous spread only. The remaining two thirds have an extraosseous extension at diagnosis.

The data on tumor location is recorded differently in the different countries, hence the larger fraction of unknown tumor location in the data from Denmark.



Adjuvant chemo- or radiotherapy

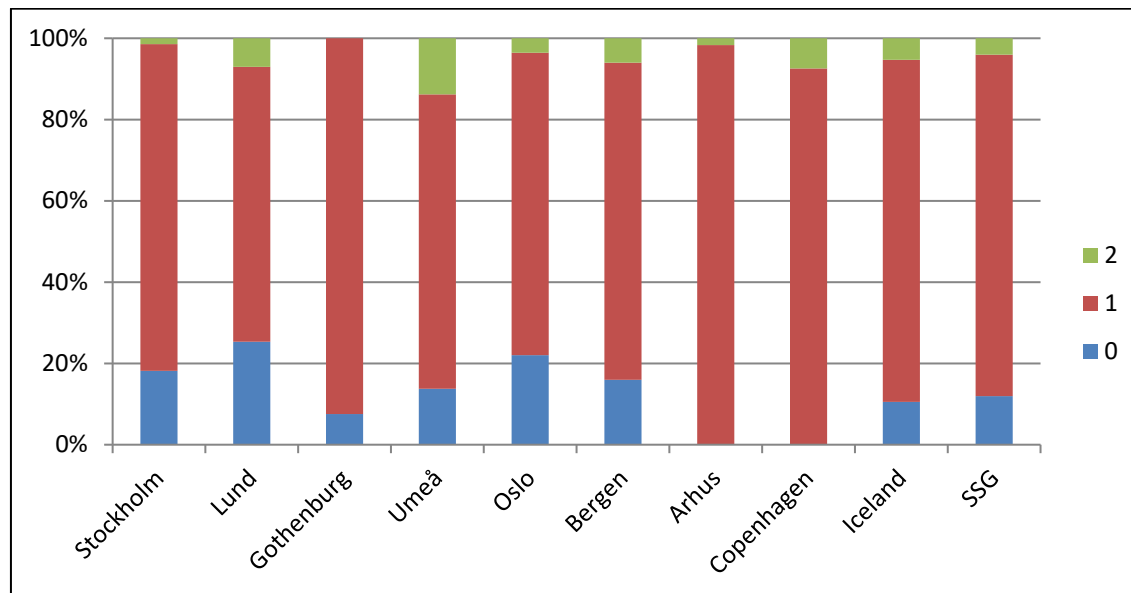
Patients presenting with Ewing sarcoma or osteosarcoma are treated with chemotherapy within or according to study protocols. For patients with chondrosarcoma adjuvant therapy is not regularly used due to limited treatment response. Data from Århus and Copenhagen includes grade 1 chondrosarcomas and giant cell tumors of the bone, entities not treated with chemotherapy which explains their lower rate. Please note that the y-axis ends at 70%.



Number of surgeries

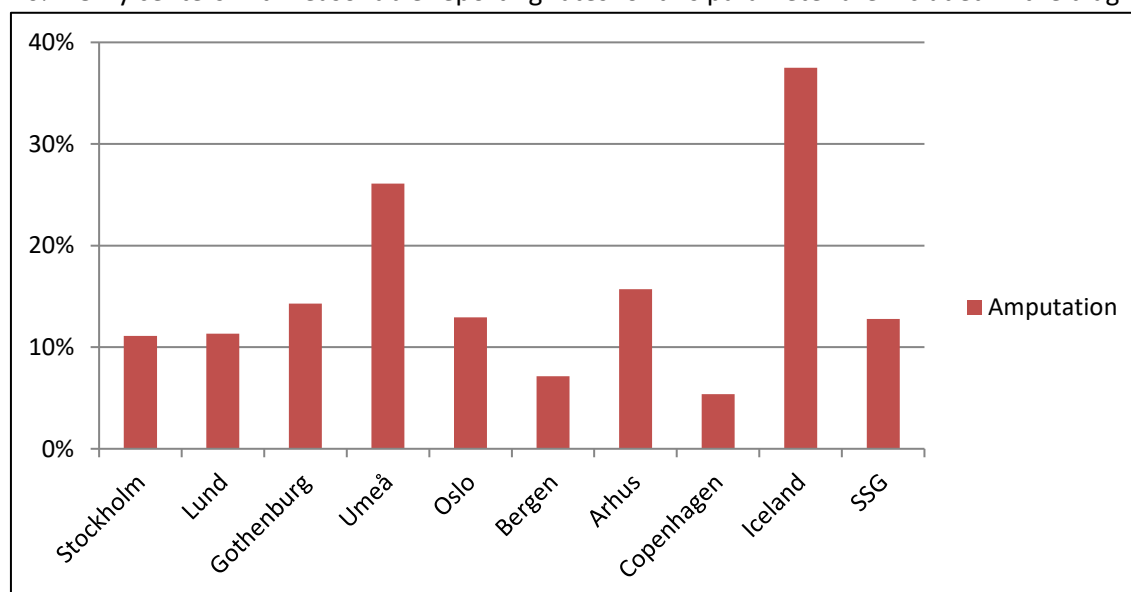
The vast majority of patients with BS undergo one surgery due to primary tumor. The numbers are similar at all sarcoma centers. However, all patients in Arhus and Copenhagen undergo surgery whereas 10-25% of patients at other centers do not. If this reflects different modes of data recording requires further analysis in the future.

Only centers with reasonable reporting rates for this parameter are included in the diagram.



Type of operation (final surgery)

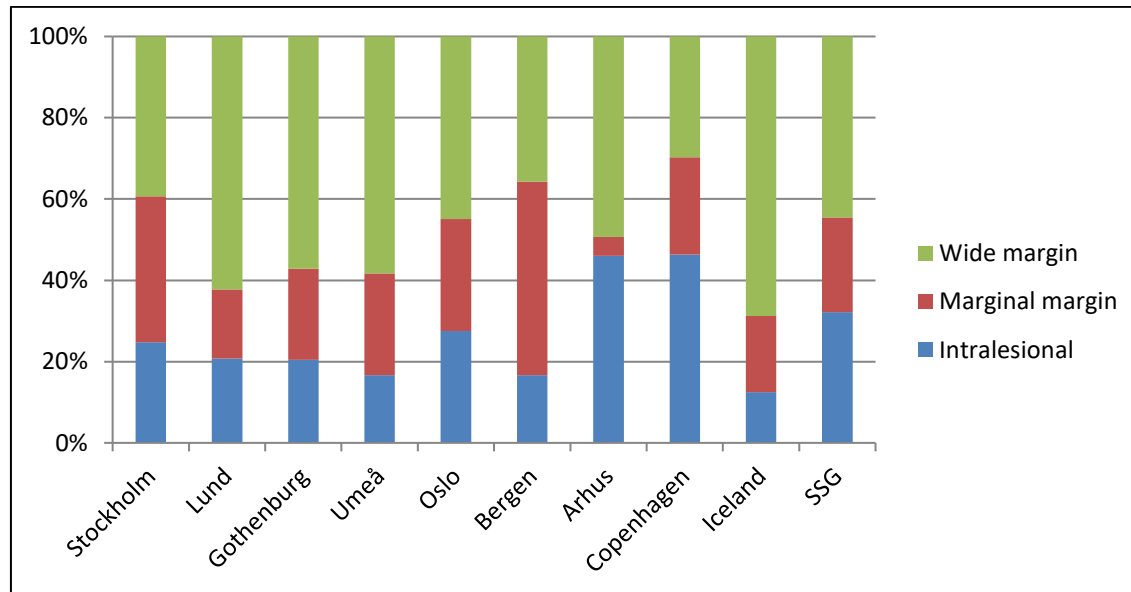
The type of surgery varies between the different centers. This is probably due to few patients undergoing amputation per center per year why it has to be monitored over time. Please note that the y-axis ends at 40%. Only centers with reasonable reporting rates for this parameter are included in the diagram.



Surgical margin

The reported final surgical margin differs between the sarcoma centers. This should be correlated to tumor size, site and whether or not all patients undergo surgery. When comparing the number of surgeries reported above to this parameter the increased rate of intralesional surgical margins in Arhus and Copenhagen probably reflects that grade 1 chondrosarcomas and giant cell tumors of the bone are included in their data.

Only centers with reasonable reporting rates for this parameter are included in the diagram.



Data per center and year

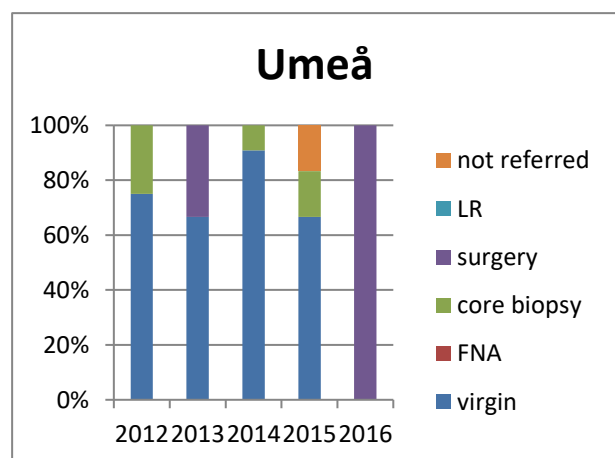
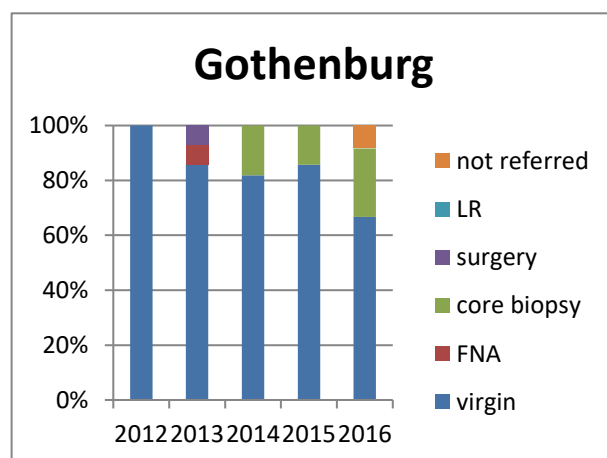
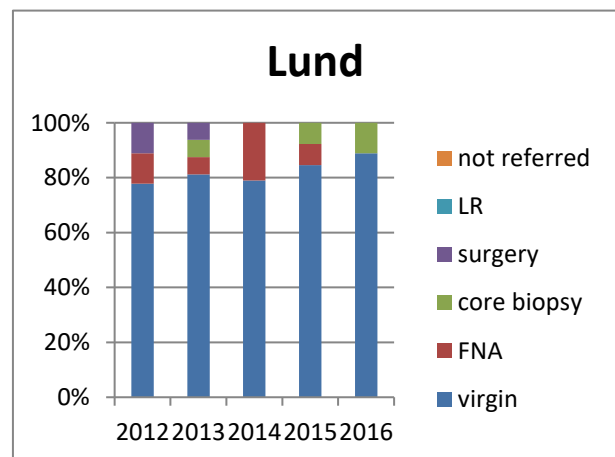
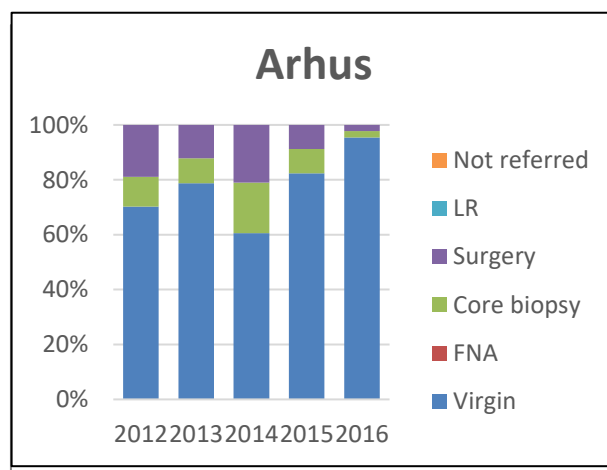
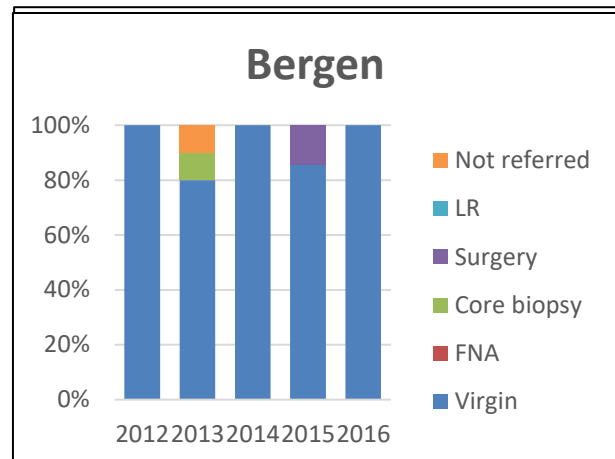
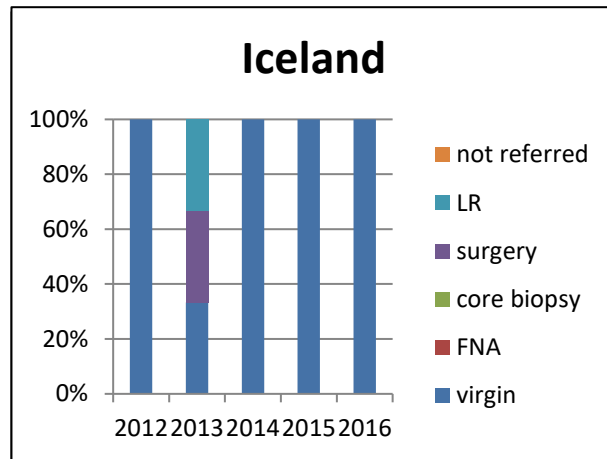
Number of cases

| | <i>2012</i> | <i>2013</i> | <i>2014</i> | <i>2015</i> | <i>2016</i> | <i>Total</i> |
|-------------------|-------------|-------------|-------------|-------------|-------------|--------------|
| <i>Oslo</i> | 33 | 25 | 36 | 43 | 51 | 188 |
| <i>Bergen</i> | 12 | 10 | 11 | 7 | 10 | 50 |
| <i>Stockholm</i> | 29 | 23 | 25 | 41 | 26 | 144 |
| <i>Linköping</i> | 1 | | | 7 | 6 | 14 |
| <i>Lund</i> | 18 | 16 | 19 | 13 | 9 | 75 |
| <i>Gothenburg</i> | 13 | 14 | 12 | 8 | 12 | 59 |
| <i>Umeå</i> | 4 | 6 | 12 | 7 | 2 | 31 |
| <i>Iceland</i> | 6 | 3 | 3 | 5 | 2 | 19 |
| <i>Copenhagen</i> | 26 | 34 | 38 | 17 | 35 | 150 |
| <i>Arhus</i> | 37 | 33 | 38 | 34 | 44 | 186 |
| <i>Total</i> | 179 | 164 | 194 | 182 | 197 | 916 |

Referral pattern

The referral pattern for bone sarcomas shows a high level of virgin referrals, i.e. that patients are referred before any invasive procedure which is in accordance with guidelines.

LR = local recurrence; FNA = fine needle aspiration; Virgin = referred with untouched tumor.



Tumor size

The distribution of tumor sizes over the years is fairly consistent for the larger centers. In general, the larger the center the smaller the variation in tumor size from year to year.

No data regarding tumor size of BS is available from Iceland.

