FUNCTION AFTER PARTIAL PELVIC RESECTION FOR EWING'S SARCOMA

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Seven patients with Ewing's sarcoma of the pelvis were treated by chemotherapy followed by wide resection of the primary tumour. Although good function after operation is possible, survival in this series reflects the poor prognosis associated with the disease; two patients died, two are alive with local recurrence and metastases and three patients are alive with no evidence of disease.

Ewing's sarcoma is the most common malignant bone tumour of the pelvis in children and adolescents. When compared with Ewing's sarcoma in the extremities, a primary in the pelvis has a particularly poor prognosis. Twenty such cases have been treated at this hospital during the last 25 years with only one long-term survivor; a five-year survival rate has been reported by other authors as ranging from zero to 10% (Dahlin, Coventry and Scanlon 1961; Falk and Alpert 1967; Phillips and Higinbotham 1967; Macintosh, Price and Jeffree 1975; Pritchard et al. 1975; Bacci, Campanacci and Pagani 1978). Historically the mainstay of treatment in Ewing's sarcoma was radiotherapy and, although in most patients this achieved satisfactory local control, death would nonetheless occur within two years from disseminated disease. It was argued that there was, therefore, no role for surgery (Boyer, Brickner and Perry 1967), but ablative surgery has been performed in some centres and has been shown to be associated with improved survival (Pritchard et al. 1975); the overall prognosis, however, remained poor.

With the advent of intensive multi-drug, multi-cycle chemotherapy to control both the primary and the systemic micrometastases, the possibility of treating peripheral lesions improved and, when combined with megavoltage radiotherapy, 60% to 70% of patients may be cured (Kinsella et al. 1984). However, less than a third of patients with primary tumours in the proximal extremity or pelvis are long-term survivors (Rosen 1978).

In order to improve local control in these sites and to reduce the morbidity and associated functional impairment after radiation therapy, there has been a recent trend towards the use of pre-operative chemotherapy (to reduce the size of the pelvic primary) followed by wide excision (Marcove and Rosen 1980; Pritchard 1980).

Although a consideration of survival after this type of management of Ewing's sarcoma is of overriding importance, the principal aim of this paper is to examine the functional result after wide resection of the pelvic primary in a group of children and adolescents treated at the Royal Children's Hospital.

PATIENTS AND METHODS

Nine patients with a mean age of 13.7 years (range 9.3 to 18.3 years) presented between 1978 and 1984 with Ewing's sarcoma of the pelvis. Pre-treatment investigations included a thorough assessment of the primary lesion by conventional radiography, tomography (from 1980 computerised tomography), a radioisotope scan of the whole skeleton using technetium-99m and, finally, a biopsy. In addition to the radio-isotope scan a general assessment of the disease was made by conventional radiography and tomography of the chest, as well as by bone marrow aspiration and a full blood count including ESR and liver function tests.

The treatment protocol consisted of chemotherapy followed by resection of the primary tumour with or without radiotherapy. Before 1981, combination chemotherapy with vincristine, adriamycin and DTIC (dacarbazine) was used (Protocol A; Campbell, Ekert and Waters 1983). Subsequently a modification of the T9 protocol used at the Memorial Sloan-Kettering Institute for Cancer Research was employed (Rosen et al. 1981).

Two patients, each aged 13 years, were excluded from this study. One had metastatic disease at presentation and received chemotherapy alone (T9 protocol) but died after 18 months. The second was not considered for
surgery because of a persistent wound infection caused by *Staphylococcus aureus* after a biopsy performed elsewhere: in this instance, regression of the primary was achieved with the T9 protocol. The patient subsequently received radiotherapy to the ilium (50 Gy) but developed local recurrence and metastases seven months after presentation and died two months later.

Seven patients with no metastases at presentation had wide resection of the primary (Figs 1 to 5). The mean age at operation was 13.4 years and in each case the duration of pre-operative chemotherapy was determined by the response of the primary tumour. One patient (Patient 7) was initially treated elsewhere with a full course of chemotherapy followed by radiotherapy.

The plane of resection was outside the pseudocapsule through muscle, and the pelvis was divided at least 2 cm beyond the margins of the tumour as delineated by radiography and bone scintigraphy. The type of resection and reconstruction was classified according to Enneking and Dunham (1978). We modified this system, adding a Type IIIb to describe resection of the ischium and ischiopubic ramus. Two of the four patients who had a Type I resection had a Type I reconstruction, that is, stabilisation of the neck of the ilium to the ala of the sacrum using threaded Steinmann pins and autogenous bone graft (Patients 1 and 4).

Hip function in those patients still alive at review was assessed by the system proposed by the International Symposium on Limb Salvage in Musculoskeletal Oncology (Enneking 1987, Tables I and II). Information relating to function in the remaining patients before local recurrence was obtained from the case notes for two patients and from an interview with one.

**RESULTS**

**Disease status (Table III).** Two patients have died (Patients 1 and 2), two are alive with local recurrence and metastases (Patients 5 and 7) and three patients with a median follow-up of 25 months (range 19 to 42 months) are alive with no clinical, radiographic or scintigraphic evidence of local or distant disease. Histological examination of the resected specimens revealed residual viable tumour in Patients 1, 2 and 7 (although the resection was macroscopically wide in all cases, residual viable tumour was demonstrated bordering the osseous plane of resection in Patients 2 and 7; the clearance of resection in these two patients was therefore marginal).

**Table 1.** Functional evaluation of the surgical management of musculoskeletal tumours (proposed by the International Symposium on Limb Salvage in Musculoskeletal Oncology)

<table>
<thead>
<tr>
<th>Rating</th>
<th>Movement*</th>
<th>Pain</th>
<th>Stability and type of deformity</th>
<th>Strength†</th>
<th>Patient's response/rating</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>180°</td>
<td>None</td>
<td>Negative Trendelenburg or 0°–5° adduction or flexion contracture or 0–1 cm shortening</td>
<td>5 kg at ankle for 20 seconds</td>
<td>Enthused/unrestricted</td>
<td>None</td>
</tr>
<tr>
<td>Good</td>
<td>120° to 180°</td>
<td>Modest or intermittent pain, not disabling (only salicylate or equivalent usage)</td>
<td>Compensated Trendelenburg (without stick) or 5°–10° adduction or flexion contracture or 1–2 cm shortening</td>
<td>2.5 kg at ankle for 10 seconds</td>
<td>Likes/minor restrictions</td>
<td>Minor (not disabling)</td>
</tr>
<tr>
<td>Fair</td>
<td>60° to 120°</td>
<td>Moderate, not continuous, disabling when present (intermittent narcotic or equivalent usage)</td>
<td>Compensated Trendelenburg with stick or 10°–20° adduction or flexion contracture or 2–4 cm shortening</td>
<td>Can only overcome gravity</td>
<td>Accepts/partial disability</td>
<td>Major (partially disabling)</td>
</tr>
<tr>
<td>Poor</td>
<td>0° to 60°</td>
<td>Severe, continuous disabling pain (continuous narcotic usage)</td>
<td>Uncompensated Trendelenburg or 20° adduction or flexion contracture or shortening greater than 4 cm or habitual crutch</td>
<td>Cannot overcome gravity</td>
<td>Dislikes/total disability</td>
<td>Major (causing failure)</td>
</tr>
</tbody>
</table>

* Combined flexion, abduction and rotation of the lower extremity.
† Able to maintain lower extremity in maximum abduction while lying on side or maximum flexion while supine.
Assessment of function (Table IV). Movement of hip. Of the six patients for whom range of movement data were obtained, all had a combined range in three planes greater than 180° (mean 240°, range 185° to 290°). There were no contractures, and the restriction of movement varied in different planes.

Pain. Four of the seven patients had no pain in the months after operation. Three experienced modest intermittent pain associated with exercise in the region of the affected hip but this was not disabling. Patients 1, 2, 5 and 7 subsequently developed pain of varying intensity associated with local recurrence of the tumour. Stability and deformity. The four patients who had a Type I resection, which in each case included resection of iliacus and a generous cuff of hip extensor and abductor muscles, had a positive Trendelenburg sign and shortening ranging from 2 cm to 5 cm. Stabilisation by a Type I reconstruction did not prevent shortening. The greatest shortening (5 cm) occurred in Patient 3 who had no reconstruction. There was no instability or shortening if the pelvic ring was disrupted anteriorly (Patient 5) or if only the ischiium was excised; these patients, however, complained of tilting when sitting or riding a bicycle due to the absence of the ischiium.

Strength. Abductor weakness (as indicated by a positive

### Table II. Overall rating system

<table>
<thead>
<tr>
<th>Rating</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>Five of the six primary factors (see Table I) must rate as excellent; the sixth may be good, fair or poor</td>
</tr>
<tr>
<td>Good</td>
<td>Five of the six primary factors good or better; the sixth fair or poor</td>
</tr>
<tr>
<td>Fair</td>
<td>Five of the six primary factors fair or better</td>
</tr>
<tr>
<td>Poor</td>
<td>Two or more of the primary factors poor</td>
</tr>
</tbody>
</table>

### Table III. Results of treatment in seven patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)/sex</th>
<th>Type of resection</th>
<th>Chemotherapy</th>
<th>Radiotherapy</th>
<th>Disease status (months after operation)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Pre-op</td>
<td>Postop</td>
<td>Pre-op</td>
</tr>
<tr>
<td>1</td>
<td>12 M</td>
<td>I</td>
<td>A*</td>
<td>A</td>
<td>55 Gy</td>
</tr>
<tr>
<td>2</td>
<td>14 F</td>
<td>IIIb</td>
<td>A, T9</td>
<td>T9</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>12 M</td>
<td>I</td>
<td>T9†</td>
<td>T9</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>9 F</td>
<td>III</td>
<td>T9</td>
<td>T9</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>13 M</td>
<td>III</td>
<td>T9</td>
<td>T9</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>17 M</td>
<td>IIIb</td>
<td>T9</td>
<td>T9</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>18 F</td>
<td>I</td>
<td>T9</td>
<td></td>
<td>60 Gy</td>
</tr>
</tbody>
</table>

* Protocol A. Campbell et al. 1983 (see text)
† T9. Rosen et al. 1981 (see text)
DWD, died with disease
AWD, alive with disease
NED, no evidence of disease

### Table IV. Assessment of function (see Tables I and II)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Functional assessment</th>
<th>Return to sport</th>
<th>Overall rating</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Movement</td>
<td>Pain</td>
<td>Stability/deformity</td>
</tr>
<tr>
<td>1*</td>
<td>Excellent</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td>2*</td>
<td>Excellent</td>
<td>Excellent</td>
<td>Excellent</td>
</tr>
<tr>
<td>3</td>
<td>Excellent</td>
<td>Good</td>
<td>Poor</td>
</tr>
<tr>
<td>4</td>
<td>Excellent</td>
<td>Good</td>
<td>Fair</td>
</tr>
<tr>
<td>5</td>
<td>Excellent</td>
<td>Excellent</td>
<td>Excellent</td>
</tr>
<tr>
<td>6</td>
<td>Excellent</td>
<td>Excellent</td>
<td>Excellent</td>
</tr>
<tr>
<td>7†</td>
<td>Not known</td>
<td>Excellent</td>
<td>Good</td>
</tr>
</tbody>
</table>

* Obtained from case notes
† Interviewed

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Trendelenburg sign) was confirmed when power was
tested objectively in Patients 3 and 4. The case history
stated that Patient 1 was unable to straight leg-raise and
had a positive Trendelenburg sign. Although the hip in
Patient 5 was considered to be stable, weakness in
flexion, abduction and adduction was demonstrated. The
power of the hip abductors and flexors was normal in
only one patient (Patient 6) who had had a Type IIIb
resection.

**Patient's response and subsequent activities.** No patient
was enthusiastic about the result of surgery with respect
to function but accepted the varying degrees of deficit.
No information concerning return to sporting activities
was available for Patient 2, and Patient 7 had an early
recurrence which prevented a return to anything other
than activities of daily living. Details of return to sport
were available for five patients: these showed that, even
in the presence of instability and shortening, a rapid
return to some sporting activity was possible and could
be maintained even with the subsequent development of
marked shortening in the affected leg.

**Complications.** The only operative complication occurred
in Patient 3 with the division of the lateral cutaneous
nerve of the thigh. A superficial wound infection
developed during chemotherapy in Patient 6 six weeks
after operation; this rapidly resolved with antibiotic
therapy. All the remaining wounds healed without
complications.

**Overall rating.** This could be made only for the four
patients who were examined, and there were three good
results and one poor. But for the late wound infection in
Patient 6 (which resolved without further morbidity) the
result would have been excellent; the other two good
results and the one poor result were in patients in whom
the pelvic ring had been disrupted by resection of the
tumour. The poor result in Patient 3 was a consequence
of the 5 cm of shortening and weak hip abductors;
nonetheless, his function with respect to activities of
daily living and ability to play badminton did not differ
greatly from Patient 6.

**DISCUSSION**

Any discussion of the functional results of a surgical
procedure performed as part of the management of a
malignant tumour must also take into account the effect
of such a procedure on the subsequent behaviour of the
disease. With regard to the treatment of connective
tissue sarcoma in the region of the hip Enneking (1966)
stated that, when surgically feasible, radical resection is
preferred to hemipelvectomy if the prognosis will not be
compromised and a satisfactory functional result can be
expected. Wide partial excision has become an accepta-
ble procedure for specific aggressive benign and
malignant lesions in the innominate bone, thus avoiding
the emotional trauma and functional impairment asso-
ciated with hemipelvectomy (McLaughlin et al. 1975;
Erikson and Hjelmstedt 1976; Steel 1978; Enneking and

There is, however, little detailed information about
function after such procedures.

This study has shown that good function with
minimal morbidity may be retained after partial resec-
tion of the pelvis in children and adolescents. A return to
sporting activities may be achieved, although at a
reduced level of competitiveness. The only comparable
series is that reported by Enneking and Dunham (1978)
who found that those who had a Type I resection and
reconstruction had almost normal function, no more
than 2 cm of shortening and no impairment of stability
and movement of the hip. All the Type I resections in this
study had some degree of residual instability and
weakness. Reconstruction of the pelvic ring did not
necessarily prevent shortening. Should these patients
survive, leg-length inequality may require corrective
surgery. The adductor weakness demonstrated is likely
to be due to the disruption of the origin of these muscles
when a generous cuff of muscle is resected with the ilium.
The three patients who had a Type III or IIIb resection
had normal stability and either no weakness or minimal
weakness, principally of the adductors.

The major criticisms of the rating system used is that
each of the parameters carries equal weight in the final
rating: impairment of one or two parameters may
therefore result in an overall rating which does not
necessarily reflect the true residual function of the hip.
However, the principal advantage of the assessment
system is that it may become possible to compare the
functional results of this procedure between centres.

The fate of the patients in this study is not
encouraging. Four of the seven patients relapsed, two
have died and two who had what would currently be
accepted as adequate chemotherapy are alive with
disease but with a poor prognosis. This reflects the
general view that, although chemotherapy has consider-
ably improved survival in patients with peripheral
lesions, the effect on pelvic tumours has been
disappointing.

The rationale for the resection of pelvic Ewing's
sarcoma is based upon two considerations. The first is
the presence of residual tumour reported by Telles,
Rabson and Pomeroy (1978) who in a post-mortem study
demonstrated residual tumour in 13 out of 20 primary
pelvic tumours treated by radiotherapy and various
combinations of chemotherapeutic agents. The second is
an attempt to prevent the considerable morbidity and
residual functional impairment associated with radical
radiotherapy (Lewis, Marcove and Rosen 1977). Ewing's
sarcomas of the pelvis are frequently larger (with greater
soft-tissue extension) than peripheral lesions, and an
inverse relationship between the size of the primary and
survival has been reported (Marcus and Million 1984;
Evans et al. 1985). Surgery is usually deferred until a
satisfactory response to chemotherapy and regression in
tumour size has been observed, thus making wide partial resection feasible; but, even with careful pre-operative planning, the margins of resection may not be clear of tumour, as demonstrated in two cases in this study. There is recent evidence from the Memorial Sloan-Kettering Institute for Cancer Research that local control and disease-free survival may be improving by a combination of pre-operative chemotherapy and wide resection followed by lower doses of radiotherapy than usually employed in primary treatment (Li et al. 1983; Ong et al. 1984).

In conclusion, good function may be retained after resection of part of the pelvis in children and adolescents, even if this involves disruption of the pelvic ring. However, the combination of chemotherapy and wide resection of primary Ewing's sarcoma does not appear to greatly influence or improve the prognosis which continues to be poor.

In the light of recent information from the Memorial Sloan-Kettering Institute for Cancer Research and from our observation that local recurrence may develop at a site several centimetres from that of the primary and margin of resection, we recommend that these patients, in addition to chemotherapy, should receive radiotherapy (30 Gy to 40 Gy) to the entire hemipelvis after operation. This dose of irradiation should minimise the morbidity associated with radical radiotherapy.

REFERENCES


