Surgery of Pelvic Bone Tumors

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Editors

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Surgical Approaches in Pelvic Bone Tumors

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Andrea Angelini, Alberto Crimì, Elisa Pala, and Pietro Ruggieri

1.1 Introduction

Surgical approaches to the pelvis in musculoskeletal oncology are employed primarily for tumor removal and, in recent years for pelvic reconstructions. Because of the constraints posed by pelvic anatomy and tumor volume, standard "traumatological" exposures are often inadequate. Moreover, preoperative biopsy is frequently performed to reach the definitive diagnosis before surgical treatment and biopsy tract must be included with the specimen to avoid local tumor cell seeding [1]. This aspect underlines that also the trocar-needle biopsy should be performed by a surgeon with experience in pelvic resection [2]. The surgical approach for pelvic resections was first described by Enneking in 1978 [3] to achieve the desired surgical objective: the utilitarian pelvic incision. This extended ilioinguinal approach has been described and used for all the primary (benign and malignant) and secondary tumors of the pelvic girdle. It can be exploited partially or completely depending on the tumor malignancy and site as well as it can be extended for wider pelvic resections [4–6]. Since the initial description, various modifications have been proposed by Campanacci, Karakousis, and other authors [7–11]. The main ones are the T-incision, the question mark incision, the vertical posterior extension to the vertebral midline, and the ilioinguinal approach extended to the contralateral pubic ramus. These approaches require an appropriate preoperative planning and surgeon' familiarity with the anatomic relationships of pelvic region [12]. In some cases, a multidisciplinary approach with two different team for resection and reconstructive procedures could be useful under oncologic point of view.

1.2 Preoperative Evaluation

1.2.1 Relative Indications

Several preoperative considerations must be considered before proceeding with internal/external hemipelvectomy. There are some precautions that should be taken into account to avoid intra/postoperative complications.

1. As is true for all areas of medicine, a complete history is crucial to estimate patient's suitability for surgery, estimation of comorbidities, and definition of surgical-related risks. In particular, in oncologic patients, aspects resulting from prior surgery, biopsy tract, radiation therapy, history of infection may significantly influence the choice of surgical procedure and approach. Moreover, depending on the size

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and site of the tumor, all pertinent imaging and pathologic studies should be completed before the final decision to proceed is made.
2. We strongly suggest the use of rectal probe placed into the rectum and fixed to the perineal region. Not only does it allow the surgeon ability to demarcate the rectum during surgery, but also it reduces the risk of wound contamination by fecal material in the immediate postoperative management.
3. Localizing the ureters during a surgical procedure can be a challenging task in patients undergoing pelvic resection. The ureter lies in the interval between the peritoneum and the psoas fascia and may be displaced by large tumors extending medially into the pelvis. A prophylactic Double-J ureteral stent placement few days before surgery may reduce the chance of injury to the ureter or increase the chance that an injury will be recognized immediately [4, 9, 13, 14]. Moreover, a Foley catheter should always be inserted into the bladder.
4. A general anesthetic is usually administered. An arterial catheter is inserted for continuous hemodynamic monitoring, and a central venous catheter is advisable. One or more large-bore peripheral venous catheters are secured in place.
5. Infectious complications following major surgical procedures are a significant source of morbidity and potential mortality [15–18]. Antibiotic prophylaxis is intended for elective procedures in which the incision will be closed in the operating room. Numerous protocols have been designed for pelvic surgery, but usually must be adapted to specific resistance patterns of each hospital environment [18]. Prophylactic antibiotics should be administered shortly before or at bacterial inoculation. This should be done 15–60 min before skin incision. The majority of studies suggest that a single dose is effective but that for lengthy procedures (>3 h) the dose should be repeated at intervals one or two times the half-life of the drug. It has also been suggested that with large blood loss (>1500 mL), a second dose should be given.
6. In addiction to optimization of the patient's cardiopulmonary and general medical health before such massive surgical procedures are undertaken, the strict collaboration with anesthesiologist to alleviate the burden of local disease certainly plays a significant role. As a large amount of blood loss is sometimes encountered in limb salvage procedures for pelvic tumors, it is essential to identify risk factors predicting the possibility of extensive hemorrhage. The differences in patients' general condition, blood clotting ability, surgical team experience as well as speed and volume of blood transfusion may influence brisk hemorrhage. Tang et al. focused a study on this topic, finding that acetabulum or sacrum involvement, a tumor volume greater than 400 cm- and an anticipated operation time of more than 200 min are likely to have a large amount of blood loss [19]. We usually suggest large amounts of transfused blood and platelets should be prepared in such cases. Moreover, anesthesiologist should be intraoperatively updated on the current blood loss to avoid chasing progressively low hemoglobin levels. We usually avoid the use of the Esmarch bandage in patients with oncologic disease, even in case of external hemipelvectomy.

1.2.2 Patient Positioning

Patient positioning and surgical incision depend on the portion of the pelvis and soft tissue to be resected, surgeon taste, and experience. All these positions have pros and cons. The patient can be positioned in supine position (with a bumper on the contralateral side), so when required the table can be tilted. A sandbag beneath the lower thoracic spine of the affected site is useful to roll the patient approximately 45° anteriorly during posterior dissection. In this anterior "floppy-lateral" position, the skin should be prepared from the great toe on the involved site to the level of the xiphoid proximally (including the entire abdomen above the pubic symphysis), and beyond the midline posteriorly.

Lateral decubitus position allows simultaneous unilateral ventral and dorsal exposure of the hemipelvis, with the abdominal organs shifting downward far from the deep surgical plan. The patient is placed with the affected side up and the contralateral iliac crest centered over the point of flexion of the operating room table. Obviously, all bony prominences should be protected as well as the contralateral axilla and upper extremities.

Combined approaches may be performed simultaneously or staged as separate procedures depending on tumor site, type of reconstruction, and patient's comorbidities. When a custom-made prosthesis with spinal fixation is considered for pelvic reconstruction, a consecutive procedure which would allow a change in the patient's position under the same anesthesia is possible. In this case, we prefer a first surgical time in prone position before the second surgical time in supine (with the possibility of tilting the patient 45°) or lateral position. Regardless of the variation chosen, a third surgical time for complete spinopelvic fixation could be required.

1.3 Utilitarian Pelvic Incision

The utilitarian incision provides access to the inner and outer aspects of the innominate bone, the lower part of the abdomen, and the proximal femur. The starting point is the posterior inferior iliac spine, the incision then follows the iliac crest reaching the anterior superior iliac spine (Fig. 1.1a). At this point, it divides in two branches (Fig. 1.1b): the first branch of the incision extends along the inguinal ligament ending at the symphysis pubis, the second branch extends caudally with a gentle curve on the anterior aspect of the thigh for 5–7.6 cm and then bends laterally crossing the femoral shaft just below the greater trochanter following the posterior aspect of the femur and the insertion of the gluteus maximus muscle. In the modified T-shaped approach, the surgical incision is much more laterally in the turning point compared to the utilitarian incision; the distal branch runs straight on the lateral aspect of the thigh and does not turn posteriorly like in the Enneking approach. It was described for the first time by Karakousis in 1989 [20].

Some adjustments are necessary based on the size and position of the tumor: in periacetabular resections the incision is extended on the lateral thigh, in posterior resections the posterior part of the incision can be extended to the spine (with an added vertical incision), in anterior resection (pubic rami resections), the ilioinguinal incision can be extended to the contralateral side or downward facilitating the femoral vessels identification [9, 13, 14, 20, 21].

The preoperative planning of the resection and a correct biopsy technique are pivotal in order to

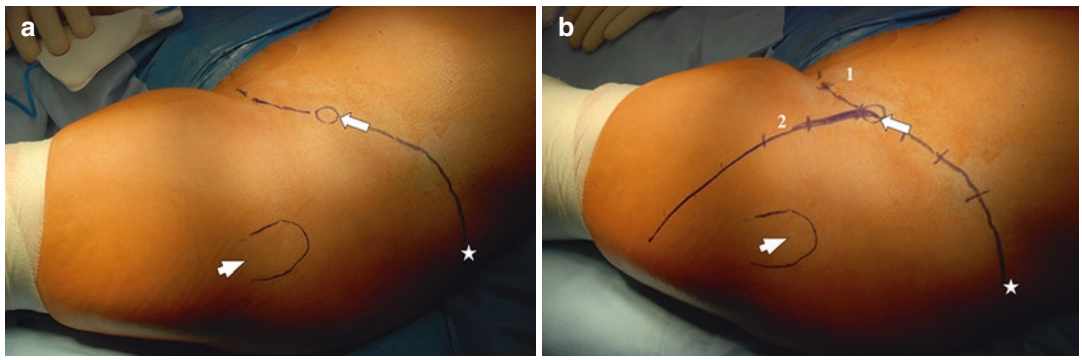


Fig. 1.1 Utilitarian pelvic incision. (**a**) The landmarks are the great trochanter (arrow head), anterior superior iliac spine (white arrow), and symphysis pubic. The starting point is the posterior inferior iliac spine (white star) and the incision follows the iliac crest reaching the anterior superior iliac spine. (**b**) Then it divides in two branches: the first branch extends along the inguinal ligament ending at the symphysis pubis (n. 1) and the second branch extends caudally on the anterior aspect of the thigh and then laterally just below the greater trochanter (n. 2)

avoid the jeopardizing effect on the soft tissue survival and reconstruction. The biopsy has to be performed along the utilitarian pelvic incision because the excision of the biopsy tract to avoid seeding of the tumor cells can bring to an extensive soft tissue damage [14].

1.4 Indications Related to Types of Pelvic Resections

Pelvic resections are classified according to the Musculoskeletal Tumor Society into four types: (1) Type I—iliac resection, (2) Type II—periacetabular resection, (3) Type III—obturator resection, (4) Type IV—resections involving sacrum [13]. Resections combining different portions can be classified and represented with the relative roman numbers, such a resection involving iliac and acetabular areas is called Type I/II resection. When all the three parts of the innominate bone are resected preserving the limb, the procedure is called internal hemipelvectomy (Type I/II/III) [22].

If the resection includes the proximal femur it is defined a Type H resection, divided in: Type H1—resection involving the femoral head, Type H2—resection involving the trochanteric area, Type H3—resection involving the subtrochanteric area [13, 21].

When resection includes the sacrum, the subclassification is categorized in four types: Type 1—resection involving a total sacrectomy, Type 2—resection involving a emisacrectomy, Type 3—resection involving a partial sacrectomy associated with an external hemipelvectomy, Type 4—resection involving a total sacrectomy associated with an external hemipelvectomy [4, 20].

1.4.1 Type I Resection

In order to obtain a Type I resection, only the first portion (most posterior part) of the utilitarian pelvic incision is usually needed. Anteriorly, the lateral attachment of the inguinal ligament is resected together with the external oblique aponeurosis, internal oblique, and transversus abdominis muscles. The anterior osteotomy is performed through the greater sciatic notch or just over the acetabulum (preserving the hip joint), under direct visualization to prevent injuries to the superior gluteal nerve and vessels. The posterior osteotomy is through or near the sacroiliac joint using an osteotome directed from posterior to anterior, with a protection of lumbosacral trunk and sacral roots. The exposure can be implemented by the release of the iliolumbar ligament at the posterior part of the iliac crest. The L5 nerve root should be visualized and preserved because it runs inferior and medial to the ligament [9, 13, 14, 20, 21].

1.4.2 Type II Resection

If the tumor involves the acetabulum (a tumor arising from the acetabulum itself or from the proximal femur and involving the hip joint in the acetabular component), a periacetabular Type II resection is indicated. In contrast to the iliac resection, the internal hemipelvectomy could be performed if an adequate wide resection procedure could be performed sparing the major nerves and preserving a functional limb [3, 21]. The lateral arm of the incision to the thigh is developed through the skin and the subcutaneous tissue, releasing the tensor fascia lata, sartorius muscle, and the straight head of the rectus femoris from their insertions on the iliac crest and anterosuperior iliac spine, respectively. The anterior osteotomy is performed through the anterior column of the acetabulum, the base of the superior pubic ramus. The posterior osteotomy is in the posterior acetabular column or in the ischium. The superior osteotomy is through the greater sciatic notch. If the posterior column is involved, some authors suggest en-bloc removal of the acetabulum and ischium [9, 20].

1.4.3 Type III Resection

Type III pelvic resection requires a medial osteotomy (through the pubic symphysis) and it is the

case where the utilitarian incision should be extended to the contralateral pubic ramus. Another osteotomy should be performed just medial to the acetabulum, avoiding the hip dislocation [21]. In this kind of resection, due to their proximity to the pelvic sidewall, obturator artery, vein, and nerve are usually sacrificed with part of the obturator internus muscle [13, 20, 21]. In order to avoid hernias of the peritoneum, a careful reconstruction of the inguinal floor is required all along the excised part of the pubic ramus. After the excision of the bony part, femoral vessels and spermatic cord should be repositioned deep to the abdominal wall reconstruction [23].

1.4.4 Type IV Resections and Sacrectomies

Type IV resections involve the sacrum. Sacrectomy can be partial or total, combined usually with iliac resections and lower lumbar spine resections [4, 21, 24, 25]. The S2 level is pivotal to define the outcome and surgical approaches to obtain a resection with wide margins. A tumor extending below the S2 level can be treated with a partial sacrectomy (transverse, sagittal, combined) without spino-pelvic reconstruction, with good expected neurologic results related to bladder and bowel function [24–27]. Moreover, a posterior-only approach could be used in selected cases [28]. If the sacroiliac joint is not involved by a sacral tumor (lateral sacral tumor), a sagittal partial sacrectomy is indicated, whereas in case of sacroiliac joint involvement a partial sacrectomy and resection of the posterior part of the ileum (type I, IV resection) should be considered [29]. Sacral midline tumors not involving the sacroiliac joint are treated with a transverse sacrectomy [30, 31]. A total sacrectomy is indicated when an aggressive lesion involves the proximal sacrum with anterior extension (rarely tumor can penetrate the anterior pelvic fascia extending to the rectum and other pelvic organs) [32–36]. In this case, sacral roots are necessarily sacrificed to obtain wide surgical margins and local tumor control [24, 37]. Despite major complications and implicit neurological deficits of this resection technique, patients' survival and tumor control can be achieved with a total sacrectomy [24–27]. If the tumor invades S1, lumbar spine, and pelvis, the proposed surgical approach is a combined staged posterior and anterior approach. The combined approach finds indication in tumors with high vascularization, primary sacral tumors involving S1 or invading the lumbosacral junction [9, 38].

1.5 Deep Surgical Dissection

A large flap of the gluteus maximus is reflected posteriorly in order to give exposure to the greater and lesser sciatic notches, the ischium, and the proximal third of the femur. The flap is based on a line that extends from the most medial portion of the posterior part of the iliac wing to the posterolateral aspect of the thigh [3]. The sciatic nerve is close to the pelvis at the sciatic notch; it is usually not infiltrated and can be isolated and easily separated from the tumor. Iliac muscle, gluteus medius, and gluteus minimus muscles are usually excised in order to obtain wide margins and good coverage of the pelvic tumor (more gluteus medius is not excised more abductor function will be preserved). The superior gluteal artery and vein are sacrificed because the gluteus medius and gluteus minimus are resected with the tumor.

In the anterior branch of the approach (Fig. 1.2a), the inguinal ligament has to be detached from the anterior superior iliac spine and, as well as in ilioinguinal approach, the aponeurosis of the external oblique muscle has to be incised from the superficial inguinal ring to the anterior superior iliac spine (Fig. 1.2b). Spermatic cord in male or the round ligament in female patients should be protected and retracted medially, then the section of the posterior wall of the ilioinguinal canal (fibers of internal oblique and transverse abdominis muscles) is performed under tension. The femoral bundle should be identified between the pubic tubercle and iliac crest, just anterior to the superior pubic ramus (Fig. 1.2c). Inferior epigastric artery and vein should be ligated. The multidisciplinary team

should include the plastic surgeon considering that, if ipsilateral vertical abdominis musculocutaneous flap should be used, the deep inferior epigastric artery should be preserved and protected. The important structures in the area should be identified and protected: the spermatic cord (while round ligament in women can be sacrificed), the femoral vessels (section of the iliopectineal fascia in order to mobilize the vascular bundle), and iliopsoas muscle with the femoral nerve that lies deep inside the muscle [13, 14, 20, 39]. A large vessel loop is placed around the common iliac vessels to assist with their mobilization (Fig. 1.2d). Arising from the medial and lateral aspects of the common femoral artery are the external pudendal and superficial circumflex iliac arteries that could be ligated to allow mobilization of the femoral vessels. Protection of the bladder is required, if pubis ramus osteotomy has to be performed. The pubic symphysis is exposed by detaching the anterior rectus abdominis and pyramidalis muscles from their insertion onto the ipsilateral pubic crest (Fig. 1.2e). The urethra that lies just inferior to the pubic symphysis and separated only by the arcuate ligament, should be protected during osteotomy. These structures are better identified with a Foley catheter inserted. In the following step, the help of a general surgeon is needed at this time to gentle separate the abdominal organs from the pelvic tumor assessing that wide margins are granted.

In the posterior pelvis after anterior part of the sacroiliac joint is identified and going further medially, common iliac vessels should be identified and followed into the pelvis (the same for the inferior vena cava in a right internal hemipelvectomy). A Double-J ureteral stent inserted before surgery in the ureter facilitates its identification as it crosses the common iliac artery, it must be identified and should be retracted medially. The posterior part of the sacroiliac joint should be visualized; the L5 nerve roots come out just below the L5 vertebra's transverse process, where the iliolumbar ligament attaches to the posterior ilium. In pelvic resection type 4, a posterior approach is usually needed for vertebral instrumentation.

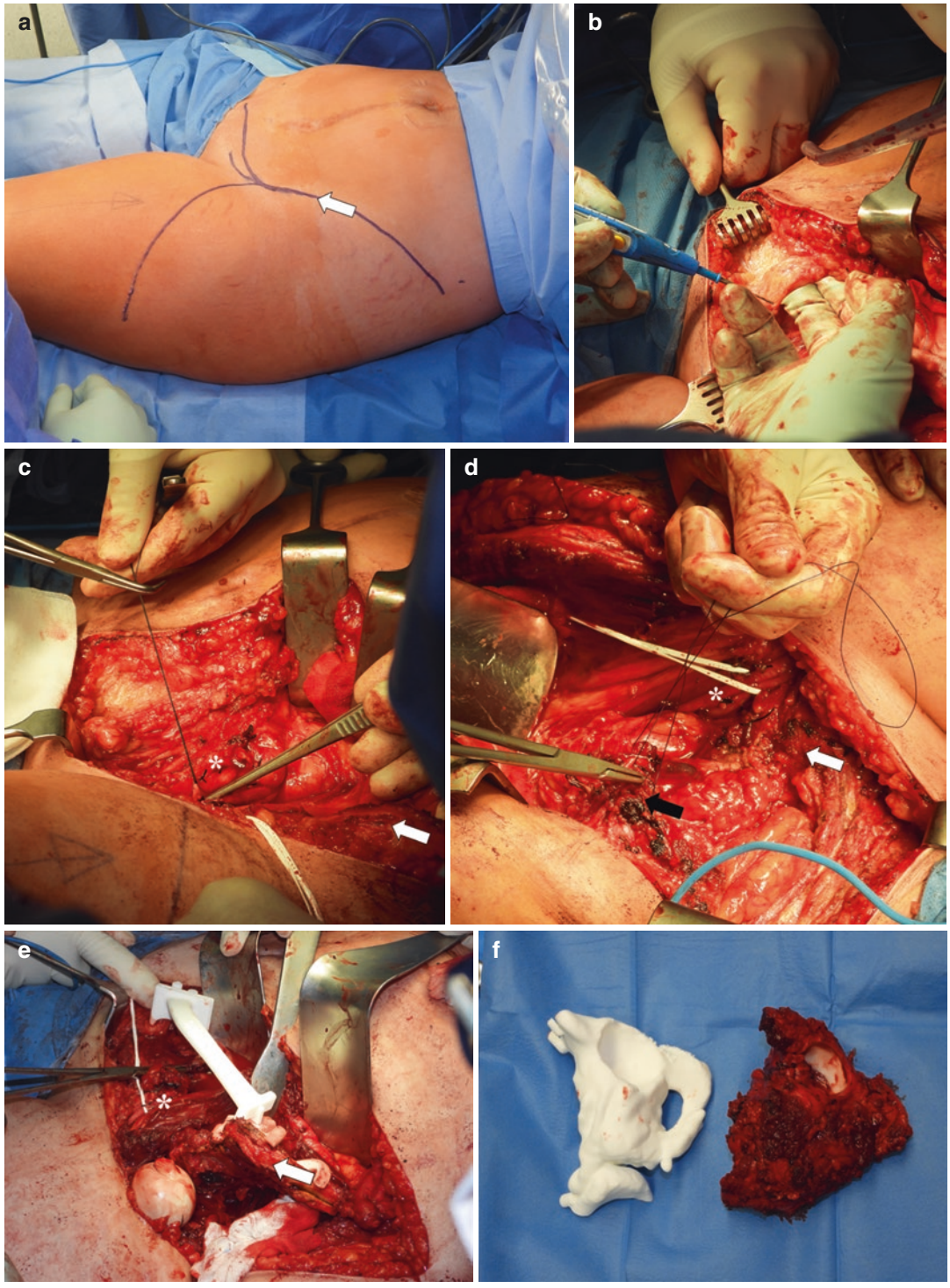
Once the bone cuts have been completed, the pelvis will open, but the sacrospinal and sacropubic ligaments must be resected to release the hemipelvis and make it loose still [13, 14, 20, 39]. The specimen should be compared with preoperative resection planes and margins macroscopically evaluated (Fig. 1.2f). After that, the reconstructive phase can be carried out (Fig. 1.2g–j).

The "reverse question mark" approach is characterized by the absence of the anterior branch of the utilitarian pelvic incision (Fig. 1.3a) and could be used when pubic osteotomy is planned close to the acetabulum. The deep surgical dissection includes the same previously described steps starting from neurovascular identification (Fig. 1.3b). One of the advantages of the supine position is the intra- and inter-observer reproducibility of radiographic measurements, especially when a custom-made resection (Fig. 1.3c) and prosthetic reconstruction are planned (Fig. 1.3d, e).

1.6 Pelvic and Soft Tissue Reconstruction

At today, there are few instances in which a staged approach may be preferable. In most of the cases, a consecutive procedure allows the possibility of

Fig. 1.2 Young patient (12 years-old) with Ewing's sarcoma of the left hemipelvis. (**a**) The skin incision was first drawn follows the utilitarian approach in supine position. (**b**) The inguinal ligament is incised from the anterior superior iliac spine (white arrow), as well as the aponeurosis of the external oblique muscle. (**c**) The femoral bundle has been identified (asterisk) and (**d**) protected with a large vessel loop. The tensor fascia lata, sartorius muscle, and the straight head of the rectus femoris have been released from their insertions on the iliac crest and anterosuperior iliac spine, respectively (black arrow). (**e**) A cutting jig has been positioned on the exposed iliac bone and pubic symphysis to perform correct osteotomies. (**f**) The model of the tumor and the specimen is shown to emphasize the similarity between resection plan and actual margins. (**g**) After tumor removal, it is possible to evaluate bone defect, osteotomy surface of the iliac bone (dashed line), and neurovascular bundle (asterisk). (**h**) In this case, reconstruction has been performed with an iliofemoral coarctation stabilized with a mesh tube (Trevira; Implantcast, Buxtehude, Germany). (**i**) Soft tissue reconstruction and (**j**) reinforcement of the abdominal wall with fascia lata graft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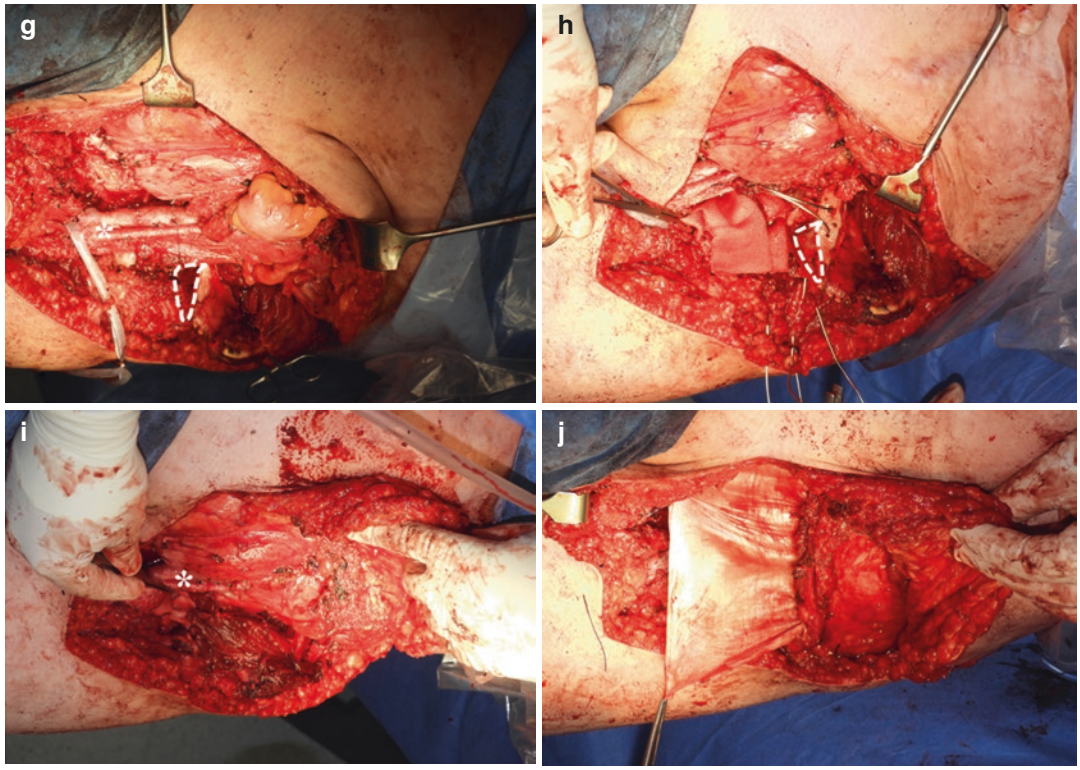

Fig. 1.2 (continued)

an immediate reconstruction, with a better reattachment of the soft tissue. In wide pelvic resections (mainly in combined type including a type II), the reconstruction of soft tissue defect and adequate implant coverage is crucial [40, 41]. Modular prostheses, custom-made 3D-printing prostheses, massive allografts, and other techniques are used for these challenging reconstructions [42–50] and infection remains the main complication [15–18]. In literature, it is widely reported that good soft tissue coverage of the prosthesis is considered one of the most relevant factors associated with implant survival [16, 18, 46].

The intersection point of the cutaneous incisions is at risk of delayed healing and wound necrosis (with finally high risk of periprosthetic), significantly higher if the soft tissue reconstruction of the deep tissues is not adequate. Enneking suggested, if there was not enough tissue to close the wound primarily, to cover important structures with flaps of omentum, it dressed with pigskin and then by skin grafts [3].

Different flap techniques are in use and are available considering the extension and the soft tissue damage during tumor excision [51, 52]. Preoperative CT with contrast study is always mandatory to properly plan the flap.

1.6.1 Rectus Abdominis Musculocutaneous Flap

Local flaps (advancement, rotation, propeller, and transposition flaps) are mainly based on a perforator as a pedicle. The rectus abdominis musculocutaneous (RAM) flap could be used as a muscular or a musculocutaneous flap, and could be realized with a transverse RAM (TRAM) or a vertical RAM (VRAM) based on the orientation of the skin paddle, to fill small defects with exposed vital structures. The VRAM flap is a solution in periacetabular and sacral reconstructions [41, 53–57]. In some cases, if there is a large fascial defect, it can be associated with a

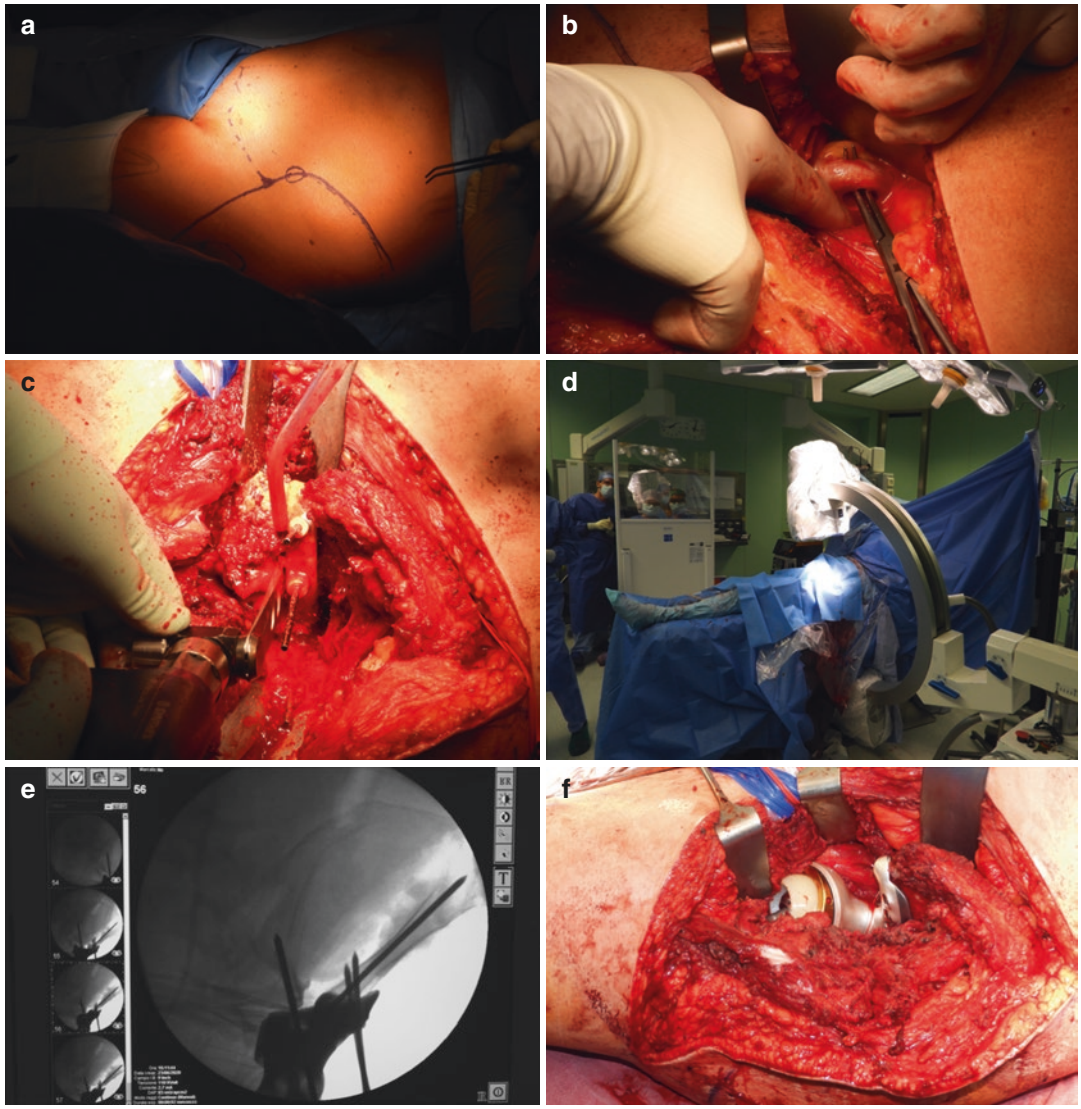

Fig. 1.3 Adult patient (52 years-old male) with an osteosarcoma of the left hemipelvis. (**a**) The "reverse question mark" approach has been drawn on the skin. The classic anterior branch of the utilitarian pelvic incision is dashed medially to the pubic symphysis. (**b**) Identification of the femoral bundle. (**c**) The iliac wing should be accurately prepared to fit with the cutting guide jig. (**d**) The picture shows the classic use of C-arm fluoroscopy in intraoperative orthopedic procedures. (**e**) Image intensification is very useful in the evaluation of bone resection and reconstructive aspects, and it allows greater flexibility with standard radiographic projections. (**f**) Definitive custom-made 3D printed prosthesis implanted before soft tissue reconstruction

synthetic mesh or an acellular dermal matrix. These solutions can be used to repair anterior defects of the donor site, posterior defects or both. The patient is positioned supine, the rectus abdominis muscle is palpated and outlined with a marker, and the flap is designed around the needed skin island (Fig. 1.4a). A midline incision extending from the pubic symphysis to just above the umbilicus is performed. The rectus abdominis muscle is then dissected maintaining intact the anterior portion of the sheath to avoid damaging the vascular perforators (Fig. 1.4b). The harvested rectus flap could be rotated on its pedicle (Fig. 1.4c, d) and tunneled via an intraperitoneal

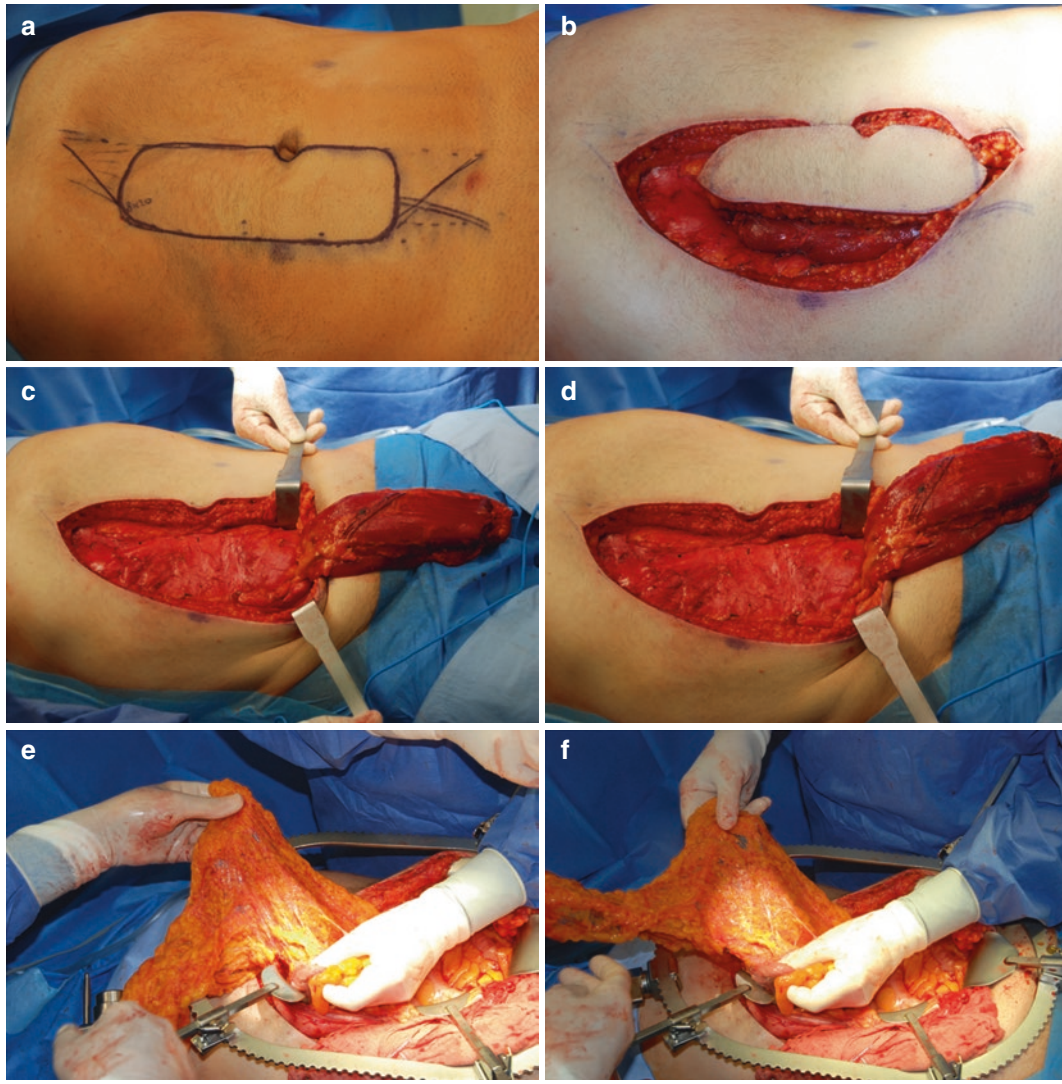

Fig. 1.4 Adult patient (54 years-old) with sacral chordoma. (**a**) Patient in supine position. The skin island is drawn based on the shape of the rectus abdominis muscle and the planned plastic reconstruction. (**b**) The rectus abdominis muscle is then dissected maintaining intact the anterior portion of the sheath to avoid damaging the vascular perforators. (**c**, **d**) The harvested rectus flap can be rotated on its pedicle and temporarily placed intraperitoneally. (**e**, **f**) During the anterior approach for proximal sacral resection, an omental-pedicled flap based on the right gastroepiploic artery is fashioned and used to fill the dead space

route into the pelvis or via an extrapelvic subcutaneous route to support wound closure [56, 57].

1.6.2 Other Flaps

Superior gluteal artery perforator (SGAP) or inferior gluteal artery perforator (IGAP) flaps are fascio-cutaneous flaps usually considered for partial sacral or total sacrectomies [58]. They may eventually include the underneath muscle, even if this myocutaneous technique should be generally avoided because it can lead to severe walking impairment. The use of a pedicled omental flap has been described as a tool of decreasing wound complications reducing the

dead space with a vascularized tissue (Fig. 1.4e, f) [53, 59]. Anterolateral thigh (ALT) flap is a reliable flap that can be used in periacetabular and sacral soft tissue reconstruction in some rare situations, usually to cover perineal or groin soft tissue defects. Tensor fascia latae (TFL) flap is a good flap for the coverage of the trochanteric, periacetabular, perineum, and abdominal wall soft tissue defects. It can be both a muscular or musculocutaneous flap.

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2 The History of Pelvic Tumor Surgery

Peter S. Rose and Franklin H. Sim

2.1 Introduction

The field of pelvic tumor surgery has advanced over the last 125+ years; this progress has been based on advances in several related areas of medicine:

- Improved anesthesia and perioperative care capabilities.
- Greater understanding of sarcomatous disease processes and margins.
- Improved imaging capabilities, particularly the use of computed tomography and magnetic resonance imaging.
- The development of adjuvant chemo- and radio-therapy.
- Expansion of orthopedic resections to locally advanced visceral disease processes.
- Critical examination of patient results and outcomes.

At present, most patients with localized pelvic sarcomas are candidates for curative resection, although high immediate and long-term morbidity remains inherent to these procedures. As well, the majority of patients are candidates for limb salvage operations. Several controversies remain in the selection and management of patients for these aggressive surgeries.

2.2 Early History

The first known attempted hemipelvectomy was by Bilroth in 1891 with a fatal outcome from hemorrhagic shock [1]. A subsequent successful operation (for advanced tuberculosis of the hip) was performed in 1900 by Hogarth-Pringle and is the first reported in the English literature [2]. Kocher described the first limb sparing pelvic excision in the late nineteenth century [3], but Putti provides the first well-documented case of internal hemipelvectomy in 1914 with successful outcome [4].

Speed popularized the term "hemipelvectomy" to describe radical amputation through the pelvis and replace the cumbersome "inter-ilio-abdominal amputation," while Gordon-Taylor referenced the procedure as a "hindquarter amputation" [5, 6]. The modern term "internal hemipelvectomy" to describe limb sparing approaches was first reported by Eilber in 1979 [7], and by analogy amputative resections are often referred to as "external hemipelvectomies" in contemporary practice.

The early twentieth century publications were primarily case reports or small case series which emphasized the surgical anatomy of approaches with relatively little data on patient outcome

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beyond mortality [6, 8]. Initially, operative mortality remained prohibitively high during the first half of the twentieth century. Gordon-Taylor reported operative mortality in 31 of 55 patients (56%) treated with hemipelvectomy for sarcoma or tuberculosis in 1934 and described the procedure as "one of the most colossal mutilations practiced on the human frame" [5, 9]. This operative mortality decreased to 22% in a later report as their team gained experience [10]. The decrease is likely due to a combination of team experience and improved perioperative care.

2.3 Advances in Disease Understanding

The mid-twentieth century brought significant advances in the scientific understanding of sarcomatous disease processes and the treatment of tuberculosis (an early indication for major pelvic resection). This included the establishment of sarcoma diagnostic categories and a tabulation of the natural history of conditions. For example, Dahlin and Henderson enumerated the basic treatment principles of chondrosarcoma in 1956 which remain true for the treatment of pelvic chondrosarcomas to this day [11]:

1. An adequate biopsy specimen for diagnosis should be obtained.
2. The definitive operation that is carried out is performed in such a manner that the biopsy wound will be excluded from the incision and will be removed with the specimen or limb, or both, without being opened again.
3. The tumor itself should be completely excised with a zone of surrounding tissue so that the surgeon does not break into or see the tumor at any time.

Dahlin and Henderson documented the difficult and morbid course of tumor recurrence as justification for aggressive initial treatment. They noted that only 3.4% of patients with inadequate surgical treatment survived or remained disease-free at 10 years, while 41% of patients treated according to these principles remained disease-free, a decade or more after surgery. This work remains one of the first and clearest tabulation of the principles of bone sarcoma resection and the greater than tenfold increase in survival seen with proper treatment.

Similar results accrued in other bone sarcomas and in soft tissue sarcomas to define the strong importance of proper biopsy and en bloc resection techniques in the treatment of sarcomas [12, 13]. Enneking, a pioneering pelvic sarcoma surgeon who helped usher in the modern era of treatment, tabulated and popularized these principles to guide sarcoma surgery in general [14]. The accumulating experience which helped to define disease processes and these principles and the dissemination of them to surgeons helped propel the role of surgery as a part of curative treatment protocols for pelvic neoplasms.

2.4 Imaging Advances

The imaging of pelvic sarcomas remains complex today, even with the variety of advanced imaging modalities available. The first pelvic tumor surgeries were based on plain film radiographs, physical examination, and surgical exploration. Later surgeons used plain film tomograms to better image the bone in combination with catheter angiograms and barium enemas to infer soft tissue extension [15]. Bone scans were incorporated as well but lacked spatial resolution.

The lack of imaging frequently lead to poorly placed biopsies, inadequate margins, and poor outcomes. Enneking's large series published in 1978 (patients operated between 1957 and 1977) revealed that one-third of patients treated with pelvic resections had oncologically inadequate surgeries for these reasons [16]. Tumor recurrence was seen in 100% of patients with inadequate surgeries. The certain morbidity of these procedures and far from certain surgical outcomes naturally tempered the enthusiasm of physicians and patients alike in selecting aggressive management of pelvic sarcomas.

The advent of computed tomography in the 1970s significantly improved the ability to image patients with pelvic tumors [17, 18]. CT imaging

provided surgeons with two primary benefits. First, it allowed much improved anatomic definition of the extent of pelvic sarcomas to define their osseous and soft tissue extension as well as visceral relationships. Second, CT scans of the chest provided improved sensitivity to detect pulmonary metastases compared to chest radiographs or lung tomograms. This second benefit allowed teams to more reliably exclude from surgery patients with established metastatic disease. CT became widely available at regional tumor centers in the early 1980s. In a similar fashion, magnetic resonance imaging provided additional anatomic discrimination of tumor extent and became widely available by 1990 [19]. In the recent two decades, positron emission tomography has similarly increased the ability of physicians to properly stage sarcoma patients [20].

These imaging advances improved the ability of surgeons to assess patients for resectability, decrease inadvertent positive margins, and to avoid morbid surgery on patients with metastatic disease. The current imaging of pelvic sarcomas is center-specific but typically combines CT and MR imaging of the local disease with CT of the chest and bone scan (or potentially PET) for staging.

2.5 Adjuvant Treatments

The three most common bone sarcomas encountered in the pelvic region include chondrosarcoma, osteosarcoma, and Ewing's sarcoma. Chondrosarcoma remains stubbornly resistant to any known adjuvant treatment, with prognosis heavily influenced by grade and surgical margin for patients with localized pelvic tumors [21]. However, dramatic advancements in chemotherapy have improved the prognosis for patients with osteosarcoma and Ewing's sarcoma.

Prior to adjuvant chemotherapy, the survival of clinically localized osteosarcoma was <15% [12]. While specific survival rates for pelvic osteosarcoma in the prechemotherapy era are not reliably recorded, these tumors are known to carry an even worse prognosis than extremity tumors, and it is reasonable to infer that long-term disease-free survival was rarely achieved in these patients.

The advent of doxorubicin-based chemotherapy immediately and dramatically improved the survival of patients with osteosarcoma [22]. These advances provided a meaningful potential for survival for patients with high-grade axial sarcomas and opened the door to consideration of aggressive surgical treatment for what had generally been considered a fatal disease. Parallel advances were made in the treatment of Ewing's sarcoma during this era as well [23].

Simultaneous advances were made in the understanding of the use of radiotherapy for pelvic Ewing's sarcoma [24]. Because of the uncertainties of imaging, margin, and prognosis, the majority of patients with Ewing's sarcoma of the pelvis were treated with radiation therapy for local control. Greater enthusiasm grew for surgical management of pelvic Ewing's tumors (with or without radiation) in the 1980s and 1990s with improved imaging, although this remains a controversial aspect of pelvic sarcoma treatment [25, 26].

2.6 Application to Visceral Diseases

While initially associated with high morbidity, the same conditions which led to advances in pelvic sarcoma surgery provided parallel advances in surgery for pelvic visceral diseases [27]. This allowed for the identification of a subset of patients with locally advanced visceral malignancies and musculoskeletal involvement and no distant metastases; typical examples would be locally advanced primary or recurrent colorectal cancer invading the sacrum or gynecologic malignancy invading the pelvic sidewall or ilium with no distant tumor spread.

Musculoskeletal involvement of visceral malignancies had traditionally been considered a marker of unresectability. However, by combining the advancing understanding of tumor biology and pelvic resection techniques, extended en bloc resections of visceral disease and involved musculoskeletal structures began in the

mid-1980s to provide curative treatment for select patients [28, 29]. While initially limited to patients with modest osseous involvement, expanding experience showed that reasonable oncologic results and survival could be obtained even with extensive resections [30]. At present, extended pelvic exenterations (en bloc resection of the visceral malignancy and associated musculoskeletal structures) are now offered at select cancer centers with reasonable patient morbidity and oncologic outcome. As is seen in virtually all pelvic tumors, margin status is a key determinant of outcome, highlighting the role of aggressive resections in curative intent procedures.

2.7 Collaboration and Examination of Results

A key aspect of surgical and scientific advancement is the collaborative sharing and critical examination of results. In parallel with the development of the field of pelvic sarcoma surgery, several professional organizations formed to improve progress and better evaluate the outcomes of patients with musculoskeletal malignancies. Notable organizations in this field include:

- The Musculoskeletal Tumor Society (MSTS, est. 1977)
- The International Society of Limb Salvage (ISOLS, est. 1981)
- The European Musculoskeletal Oncology Society (EMSOS, est. 1987)
- The Connective Tissue Oncology Society (CTOS, est. 1995)

These multidisciplinary professional organizations have (and continue) to actively advance the practice and understanding of pelvic tumor surgery. A prime example of this is the evaluation system for the systematic evaluation of patient outcomes initiated at the inception of ISOLS in the 1981 meeting. This culminated in the standard MSTS outcome instrument for evaluating the results of musculoskeletal tumor surgery [31] which remains in use to this day.

2.8 Current Practice in Pelvic Tumor Surgery

Modern imaging now allows reliable determination of tumor extent and the overt metastatic status of patients presenting with pelvic malignancies. As well, current practice provides for limb sparing resections in the majority of patients. The common nomenclature for amputative resections is either "external hemipelvectomy" or "hindquarter amputation." Limb sparing resections are termed "internal hemipelvectomies" and classified as outlined by Enneking and Dunham as to involvement of the iliac bone, acetabulum, or pubic region [16]. Clinical outcome assessment is still most commonly performed using the Musculoskeletal Tumor Society rating scale [31], although more generalizable patient reported outcomes are becoming more common.

Time and institutional practice patterns have seen different approaches and shifts in the management of pelvic sarcoma patients. The initial management of these patients focused on tumor removal alone; reconstruction was rarely used and difficult with the techniques available [7]. Recent reports have demonstrated the enduring value of this technique, and it remains a viable surgical option in contemporary practice [32].

However, other centers have demonstrated improved functional results with restoration of femorosacral continuity (anatomic reconstruction or substitution) following limb sparing resection in the pelvis [33]; this is most difficult in resections which remove the acetabulum.

A number of different approaches have (and continue) to be used in these patients. While cemented and reinforced conventional arthroplasty constructs have been reported (commonly referred to as the Harrington technique), they are most commonly used after surgery for periacetabular metastases which typically remove less bone than a primary tumor excision with oncologic margins [34]. Iliofemoral arthrodesis was initially performed for these patients but remained technically challenging with pseudarthroses and modest functional outcomes [35].

Early anatomic reconstruction experience utilized massive pelvic allografts or processed

autograft for reconstruction [36]. These reconstructions were technically demanding and suffered high complication rates. The saddle prosthesis, an adaptation of an implant for massive bone loss after failed or infected hip arthroplasty, was utilized in tumor resections in an attempt to provide a reconstructive option utilizing a modular endoprosthesis [37]. However, greater experience and follow-up have highlighted the limitations of this method, and its use in current practice is rare [38].

Modern techniques of periacetabular reconstruction include modular endoprostheses, custom prostheses, and porous tantalum implants [39, 40]. Each of these techniques has relative advantages and disadvantages based on resection and remaining bone stock as well as center experience and preferences. The use of intraoperative navigation or preprinted custom cutting guides allows precise resections to be made to match prefabricated implants.

Not all pelvic resections are commonly considered for reconstruction. While reconstructions have been reported following pubic resections [41], most centers provide soft tissue reconstructions only for these resections. Controversy exists as to whether resections of the supra-acetabular ilium require reconstruction or not. Some centers advocate for no reconstruction to minimize complications and allow medialization of the hip center to decrease Trendelenberg gait (at the expense of leg length discrepancy) [42]. Other centers have shown good results with reconstruction of these defects [43].

Despite advances on many fronts, there remains a role for external hemipelvectomy/hindquarter amputation in current clinical practice [44]. Patients are currently considered for hemipelvectomy in three primary scenarios:

1. En bloc resection of a tumor would leave a limb with such little function as to make amputation preferable. This primarily occurs when tumor extent would require removal of two or three of the critical elements of limb function (the sciatic nerve, the femoral neurovascular bundle, and the acetabulum).
2. Patients in whom resection will result in a soft tissue defect so large that the wound cannot be closed without the benefit of an amputation flap. With increasing experience with free flap coverage and the use of omentum for closure, this scenario is becoming less common.
3. For salvage of patients who experience tumor recurrence following internal hemipelvectomy.

While many teams have been pessimistic about patient function following external hemipelvectomy, modern prosthetic management can allow single hand-free ambulation for many individuals [45].

2.9 Contemporary Issues in Pelvic Tumor Surgery

Despite the large number of advances made since the first reported attempt at hemipelvectomy in 1890, a number of areas of pelvic tumor surgery remain unresolved. The need for (and if performed method of) bony reconstruction after major pelvic bone resection remains unclear. Reconstruction appears to offer better function at the price of higher complications, but selection and center treatment bias clearly influence these results. True long-term follow-up studies of patients are rare and show an expected decline in function in long-term survivors of their malignancies [46].

The uncertainties of the role and method of reconstruction are magnified in pediatric patients in whom little published literature exists to guide surgeons [47]. Most children undergoing major pelvic surgery have consideration of reconstruction for iliac defects. If the acetabulum is resected, consideration for reconstruction is given in older adolescents; young patients are generally treated with resection arthroplasty. While not strictly tabulated, the authors' clinical experience of this in young patients is generally favorable.

The role of amputation or limb salvage remains controversial. The criteria outlined above represent the classic criteria for hindquarter amputation, but some centers strive to avoid the morbidity of this by offering limb salvage to "borderline" cases. It is not clear which path provides better functional and oncologic outcomes.

Because of the morbidity of surgical resection, many centers try to employ radiotherapy when possible. This is most common in patients with Ewing's sarcoma in whom local control may be achieved with surgery, radiotherapy, or both. The combination of surgery and radiotherapy appears to decrease the risk of local failure [48]. Some studies have suggested improved survival with surgical treatment [25, 26, 49]. However, others have not shown a clear benefit [50]. No studies randomize patients between treatment arms. In addition to Ewing's sarcoma, some groups have attempted to employ high-dose proton-based radiotherapy to achieve local control of otherwise adversely presenting pelvic sarcomas with some success [51]. To date, the authors' personal experience with this for non-Ewing's tumors has been uniformly poor.

The timing of chemotherapy around major pelvic resections is an area of concern. It is established in extremity osteosarcoma, for example, that delays in resumption of chemotherapy after surgery negatively impact survival [52]. Additionally, a prospective randomized trial showed no difference in oncologic outcomes in osteosarcoma treated with immediate surgery followed by chemotherapy compared to a standard regimen of preoperative chemotherapy, surgery, and postoperative chemotherapy [53]. The magnitude (and complication profile) of large pelvic tumor surgeries is such that patients are at high risk to experience significant postoperative delays in chemotherapy resumption. This has led some centers (including the authors') to complete most or all chemotherapy prior to surgical resection in select pelvic sarcoma patients judged to be at high risk for perioperative complications. It must be stated that data regarding this practice are still being gathered, and patients undergoing "front-loading" of chemotherapy are carefully monitored with serial imaging studies for disease response.

Finally, readers should know that the oncologic staging of pelvic sarcomas has recently changed. An analysis by the American Joint Commission on Cancer (AJCC) highlighted the adverse prognosis of axial location on sarcomas. In light of this, the recently released eighth Edition AJCC Staging Manual has incorporated anatomic location in the staging of bone sarcomas (with specific criteria for pelvic tumors) in an attempt to better predict the clinical outcome of these difficult cases [54]. Accumulating data will hopefully demonstrate whether this change has value in clinical care.

2.10 Conclusions

The field of pelvic tumor has undergone a series of advances since the first major pelvic resections were undertaken over a century ago. Modern imaging, improved disease understanding, and adjuvant therapies are the pillars of these advancements. However, the morbidity of these treatments remains formidable and the prognosis guarded. Unfortunately, the words of Gordon-Taylor, a pioneering pelvic tumor surgeon, remain true in this field over a half century after they were written [9]:

> *I still cherish the hope of a golden era of cancer therapy when gross mechanical destruction of disease and cruel mutilation of tissue shall be no more. Unfortunately, these times are not yet.*

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Imaging of Pelvic Bone Tumors

3

Andrea Angelini, Joele Canapeti, Giulia Trovarelli, Joseph Benevenia, and Pietro Ruggieri

3.1 Introduction

Imaging of the pelvis can be a challenging task, especially in the evaluation of tumors and tumor-like lesion. Numerous primary and secondary musculoskeletal tumors may affect pelvic bones and usually many features appear different with the same tumors arising in other parts of the body. Conventional radiographs with multiple views (inlet, outlet, judet, etc.) represent the first screening approach in the evaluation of osseous lesions for most symptomatic patients, with the limit of the low sensitivity in detection and diagnosis. A correlation with age, history, onset, and duration of symptoms is necessary to raise the clinical suspicion. CT and MRI are the most powerful tools used for diagnosis, staging, monitoring therapy, and follow-up.

Despite the varied appearance and overlapping radiological features of pelvic tumors, a correct diagnosis should embrace the radiologic evaluation with histopathology. In this chapter, the imaging characteristics of the most common pelvic tumors will be discussed.

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3.2 Hematologic Malignancies and Secondary Malignant Bone Tumors

3.2.1 Pelvic Involvement of Multiple Myeloma and Plasmacytoma

Multiple myeloma is a malignancy of monoclonal plasma cells that represent the second most prevalent blood malignancy (10%) after non-Hodgkin's lymphoma [1]. Bony involvement is very common and pelvic bones are affected in 6% of the patients [2]. Lytic bone disease is a major feature of multiple myeloma, with multiple "punched-out" lesions with the absence of reactive sclerosis, but sometimes not easily detectable at conventional radiographs (Fig. 3.1a) [3]. Whole-body low-dose CT (LDCT), PET/CT, and MRI have a relevant role in the novel diagnostic criteria for symptomatic multiple myeloma (Fig. 3.1b, c) [4]. PET/CT demonstrates a significant higher sensitivity compared with whole-body X-ray for the detection of osteolytic lesions in multiple myeloma [5] and has an independent prognostic value both at diagnosis and after treatment [6]. Plasmacytoma is a focal, solitary proliferation of plasma cells that seems to be the early stage of a multiple myeloma.

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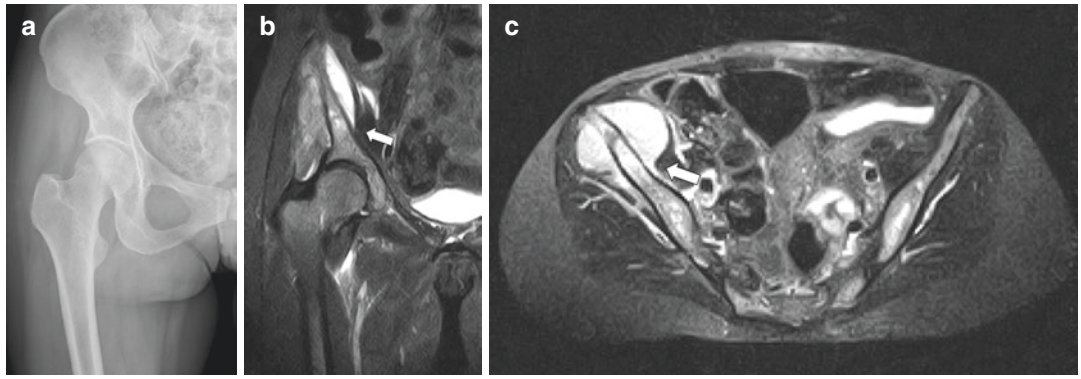

Fig. 3.1 Multiple myeloma in a 54-year old woman. (**a**) Pelvic plain radiograph is not adequate to show the lytic lesion in the right iliac bone. No fracture detected and the hip joint demonstrates mild degenerative changes. (**b**) Coronal and (**c**) Axial T1-weighted fat-saturated MRI of the pelvis obtained after intravenous administration of gadolinium demonstrate multiple areas of diffuse enhancement in the bone marrow of the sacrum and pelvic bones and a soft-tissue mass (white arrow) with symmetric growth outside the right ilium. Note the absence of cortical disruption

3.2.2 Metastases

Metastases are the most common malignant tumors and can derive mainly from breast, lung, prostate, kidney, and thyroid cancers. The incidence of symptomatic bone metastasis affecting the pelvis is increasing because of advances in diagnostic work-up, improvement of overall prognosis under chemo-, immune-, and radiotherapy [7–9]. These secondary tumors should always be considered in the differential diagnosis when aggressive lesions are observed in pelvic bones. The discovery of solitary lesion requires further analysis to exclude other primary tumors [10]. Osseous metastases may be lytic (more common), sclerotic, or mixed mainly based on histologic subtype (Fig. 3.2). Isotope scan, PET/CT, and MRI are very sensitive in detecting bone metastases and are mandatory for a complete staging at time of diagnosis [11, 12].

3.3 Primary Malignant Bone Tumors

3.3.1 Chondrosarcoma

Chondrosarcoma accounts for approximately 20% of malignant bone tumors. It is the third most common primary malignancy of bone after myeloma and osteosarcoma [13], and pelvis is the most common location [14]. Even if rare, chondrosarcomas may also affect sacral bone [15]. It is a malignant cartilaginous matrix-producing lesion with a typical progression from a low to high-grade tumor [16, 17]. They may arise from an enchondroma (central chondrosarcomas) or from an osteochondroma (peripheral chondrosarcomas), but fast growth may suggest a dedifferentiated chondrosarcoma [18]. Previous studies reported several histological parameters including grade, tumor necrosis, mitotic count, and myxoid tumor matrix for predicting the behavior of the tumor and the prognosis for the patients, even if also low-grade tumors should be treated with an aggressive surgical approach [17].

On plane radiographs, primary cartilage tumors in the pelvis should be approached with more caution than those of the extremities, because it is not possible to use the same diagnostic criteria to differentiate enchondromas from low-grade chondrosarcomas. Low-grade chondrosarcoma appears as a destructive lytic lesion with a lobulated contour, well-defined margins, endosteal scalloping, and may have cortical expansion. High-grade tumors are usually diagnosed in stage IIB (based on Enneking classification) with cortical destruction, periosteal reaction, and an associated soft tissue mass (Fig. 3.3a, b). The chondroid matrix can show

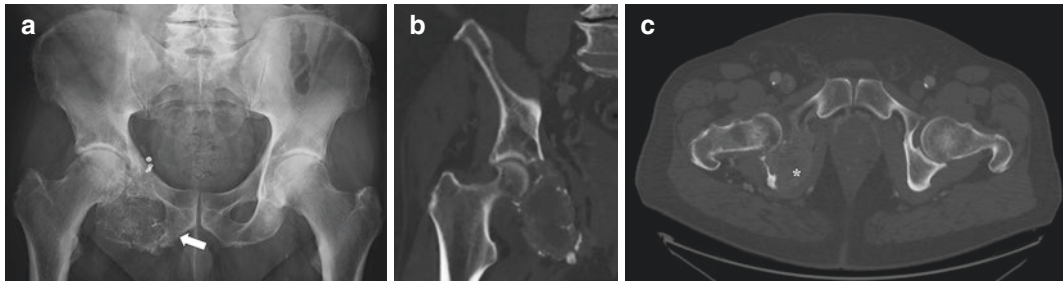

Fig. 3.2 Metastatic renal carcinoma in a 76-year old man. (**a**) Pelvic plain radiograph demonstrating a predominant lytic metastatic lesion (white arrow) in the right ischium. Note the extensive cortical involvement, predisposing it to a pathological fracture. (**b**) Coronal and (**c**) Axial pelvic CT Scan showing involvement of the right acetabular region by the large lytic metastatic lesion. Note the extensive extraosseous involvement (asterisk)

typical "ring and arc" calcifications that are more evident on CT scans, whereas the nonmineralized cartilaginous portion of the tumor has a low density. On MRI, the lobules are iso-hypointense on T1-w images (Fig. 3.3c, d) and have high intensity on T2-w images (Fig. 3.3e, f). The presence of a large lytic aggressive area adjacent to a cartilage tumor may suggest the diagnosis of dedifferentiated chondrosarcoma, especially if matrix mineralization with a bimorphic pattern on CT scan is observed [18, 19].

3.3.2 Ewing Sarcoma

The imaging appearance of Ewing sarcoma of the sacrum and the pelvis is similar to that of the extremities, even if it is variable [20, 21]. The tumor usually fills the bone marrow cavity and destroys the cortex with a moth-eaten and permeative pattern on plain radiographs (Fig. 3.4a) and CT. There is often a soft tissue mass associated to the tumor and a classic sclerotic reaction with a concentric expansion called "onion-skin" appearance may be present. CT scans and MRI play a predominant role in evaluating the soft tissue and bony extension of the lesion, and in evaluation of response to adjuvant therapies and surgical plan [22]. MRI features are nonspecific: the tumor is iso-hypointense on T1-w, with increased signal intensity on T2-w, with variable contrast enhancement (Fig. 3.4b) [23, 24]. PET/CT and dynamic MRI are under evaluation as imaging tools for restaging and tumor response to primary chemotherapy [11, 25].

3.3.3 Osteosarcoma

Conventional osteosarcoma rarely affects the pelvis, with approximately 8% of all the sites, even if it accounts for 22% of all primary pelvic bone malignant tumors [26, 27]. Most of the tumors are secondary, occurring after radiation therapy or in Paget's disease [28–30]. The plain radiographs are usually diagnostic, with an aggressive permeative pattern, combination of radiolucency and radiodensity, cortical disruption, and soft tissue involvement (Fig. 3.5a). On CT, most pelvic osteosarcomas contain "cloud-like" osteoid matrix formation and show the sunray image (stripes of density perpendicular to the cortex) (Fig. 3.5b, c) [27–31]. The telangiectatic osteosarcoma appears as predominantly lytic bone mass with minimal sclerosis on radiographs [27, 32]. MRI shows no specific features with the usual pattern of low T1-w and high T2-w signal, with heterogeneous contrast enhancement (Fig. 3.5d, e) [27]. Sometimes fluid-fluid levels may be present, especially in predominantly lytic lesions [32, 33].

3.4 Benign Tumors

3.4.1 Giant Cell Tumor

Giant cell tumors (GCTs) are benign but locally aggressive tumors that rarely affect the pelvic bones (1.5–6.1% of bone GCTs) [34, 35]. On the other hand, GCTs are the second most frequent primary tumors of the sacrum after chordoma [36, 37]. On radiographs and CT scans, GCTs are

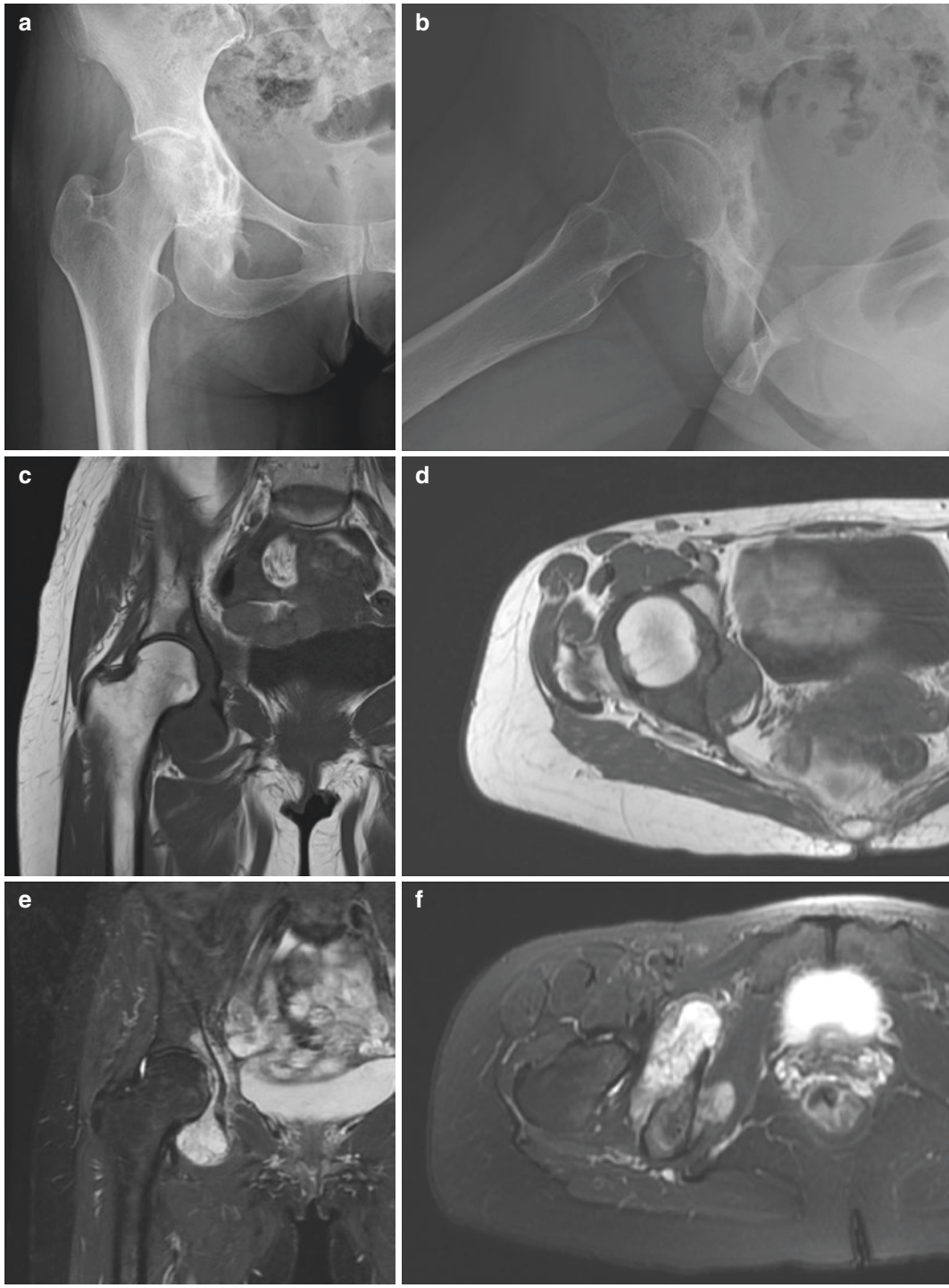

Fig. 3.3 Chondrosarcoma of the pelvis in a 60-year old woman. (**a**) Antero-posterior and (**b**) axial radiographs of the right hip reveal a periacetabular area of lucency with surrounding sclerosis and not well-defined calcifications. (**c**) Coronal and (**d**) Axial T1-weighted MR images show a tumor involving the entire acetabulum and lobular soft tissue extension with iso/hypointense signal. (**e**) Coronal and (**f**) Axial T2-weighted fat-saturated MR images demonstrate a lobulated T2 hyperintense mass compatible with a cartilage tumor

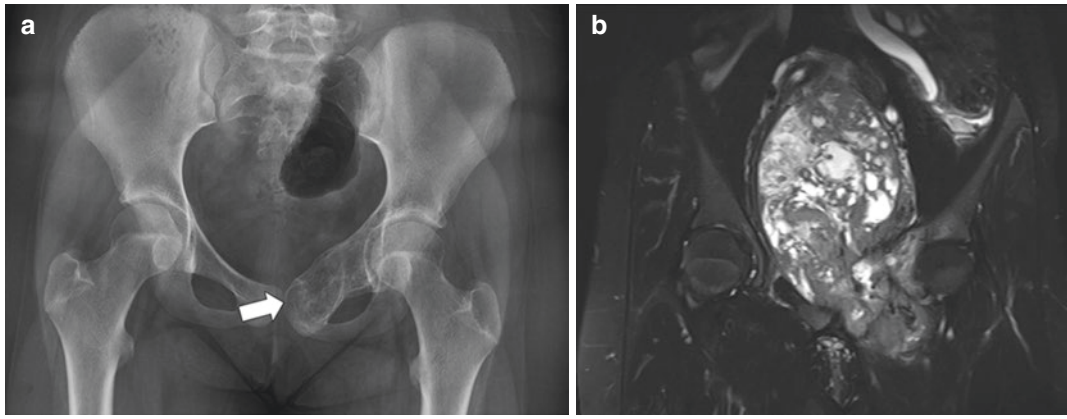

Fig. 3.4 Ewing's sarcoma in a 13-year old female. (**a**) Pelvic plain radiograph shows an expansile bony lesion centered within the left pubic bone (white arrow) up to the acetabular area. The lesion did not demonstrate gross cortical disruption, aggressive periosteal reaction, or an association with an overt soft tissue component. (**b**) Gadolinium-enhanced fat-saturated MRI coronal image reveals an extensive soft tissue mass with necrotic areas and involvement of the adjacent anatomic structures

lytic lesions that appear usually more destructive than typically seen in long bones, often with a soft tissue mass. Cortex is usually destroyed and tumor mass do not present other typical features of extremities GCTs such as sclerotic rim, periostitis, and mineralization [38]. On MRI, GCTs usually demonstrate low-signal on T1, but may have a significant heterogeneity on T2-w and fluid-sensitive sequences due to hemorrhage or necrosis [38]. The presence of fibrous components and hemosiderin gives the predominantly low to intermediate signal on T2, whereas fluid and cystic changes determine the increased T2 signal. Secondary aneurysmal bone cyst may result in prominent areas with extensive fluid-fluid levels [39].

3.4.2 Aneurysmal Bone Cyst

Aneurysmal bone cyst (ABC) is relative rare benign expansile osteolytic bone lesion with blood-filled cystic spaces. Recent studies demonstrated a neoplastic origin in primary ABC in the rearrangement of the TRE17/USP6 locus occurs resulting in TRE17 overexpression [40]. Flat bones are frequently involved and the pelvis is a common site, accounting for up to 50% of cases [41]. On radiographs and CT, ABCs appear as eccentric, well-defined lytic lesions with a thin peripheral rim of sclerosis. Cystic space with fluid-fluid levels and contrast enhancement of the septa are hallmark features of ABCs on CT and MRI images. MRI shows a high signal intensity of the fluid-fluid levels on T1-w sequences with a strong contrast enhancement due to the intense vascularization. Telangiectatic osteosarcomas represent the main differential diagnosis, especially in lesions with aggressive radiographic appearance, cortical destruction, and a soft tissue extension.

3.4.3 Osteochondroma

Osteochondromas are frequently observed in growing skeleton, especially in metaphysis of the long bones, but every bone can be affected. Ilium represents the most common site in the pelvis [42]. The imaging features are characteristic: bony excrescence with well-defined limits (Fig. 3.6a), sessile or pedunculated growth with continuity of the cortex, and medullary canal (Fig. 3.6b, c). In the pelvis, these lesions are found incidentally [43]. Differential diagnosis with peripheral chondrosarcomas is mandatory and mainly based on imaging features. In favor of malignancy are size (>5 cm), thick cartilaginous

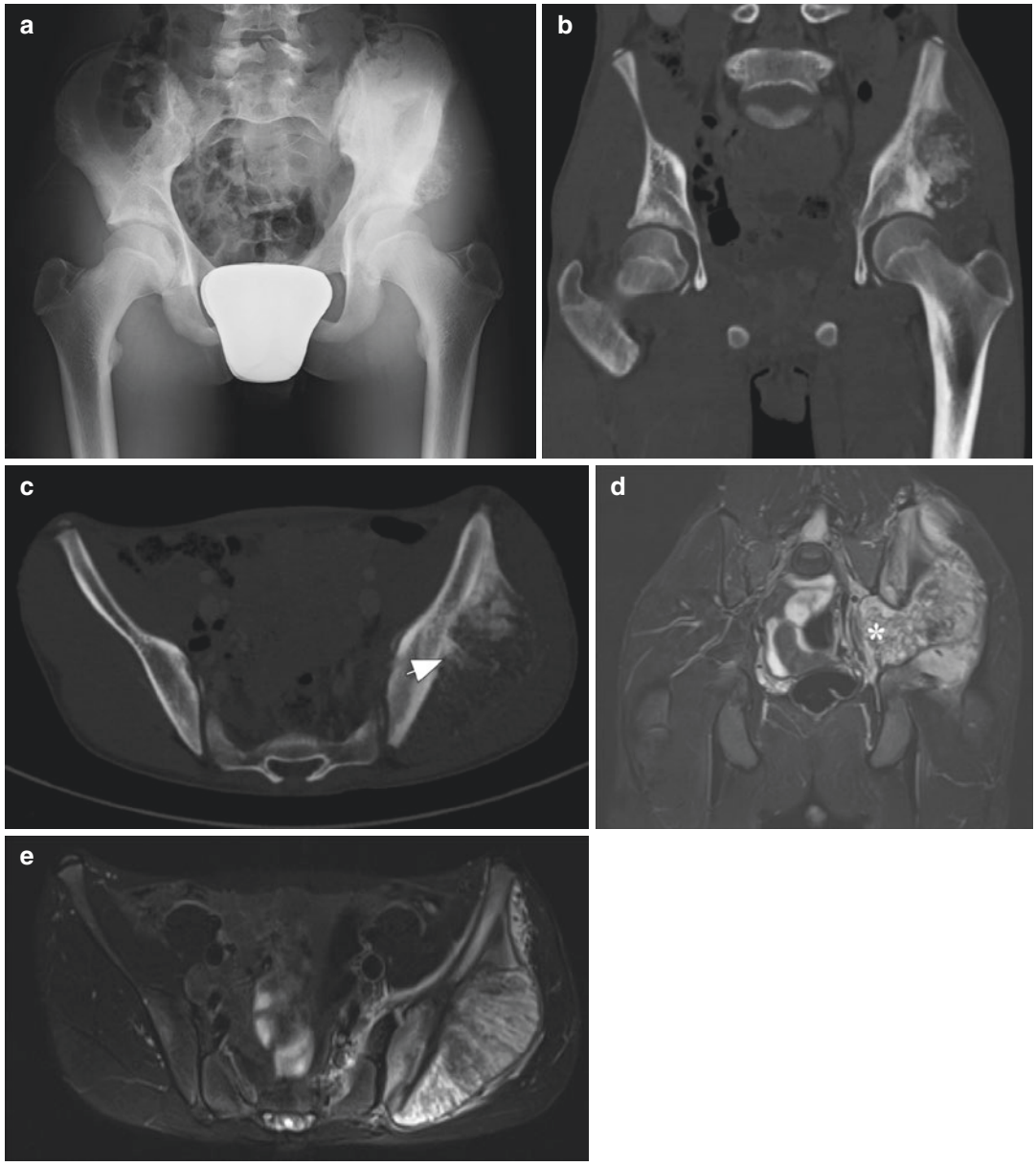

Fig. 3.5 Osteosarcoma in a 15-year old male. (**a**) Pelvic plain radiograph demonstrates an irregularly calcified lesion that involves the left hemipelvis. (**b**) Coronal and (**c**) Axial CT scan show a large associated soft tissue mass with cloudy-like areas of matrix mineralization and stripes of density perpendicular to the cortex (arrowhead). (**d**) Coronal and (**e**) Axial fat suppressed Gd-chelate enhanced MR image show asymmetric soft tissue extension. Tumor is seen to cross the greater sciatic foramen compressing and dislocating the anatomic structures inside the pelvis (asterisk)

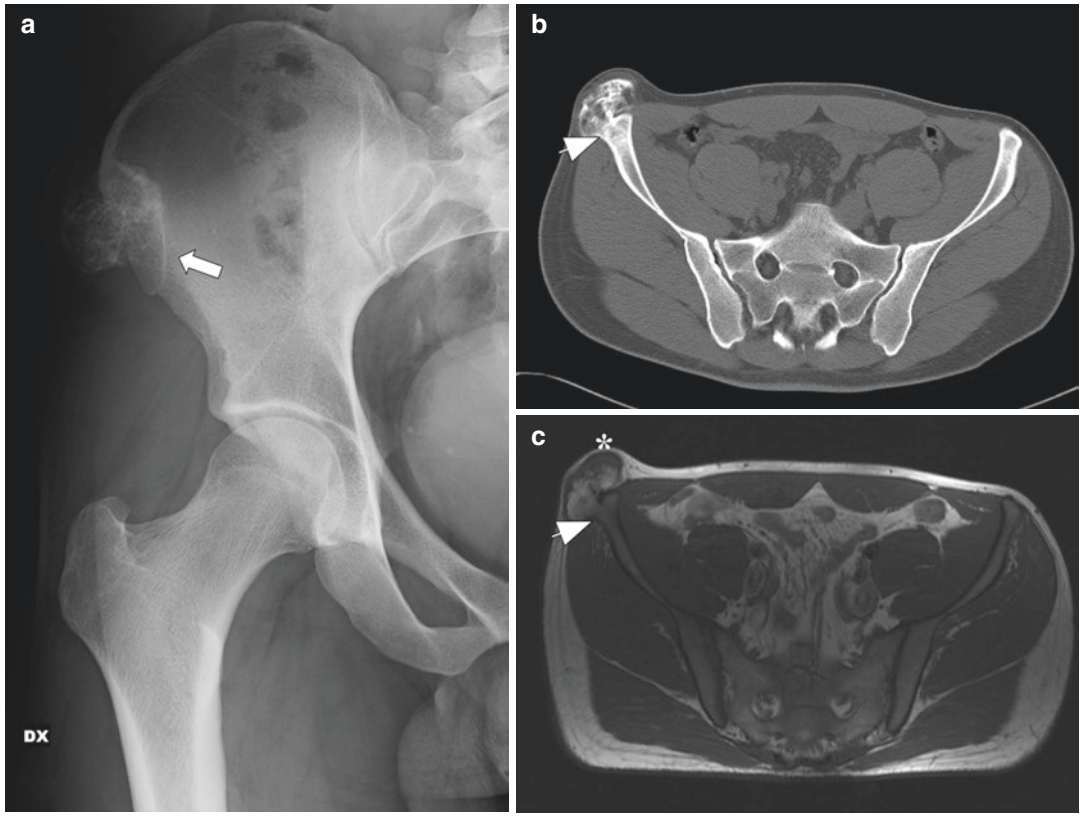

Fig. 3.6 Solitary osteochondroma in a 25-year old man. (**a**) Pelvic plain radiograph demonstrates an irregularly calcified, pedunculated lesion (white arrow) arising from the right iliac crest. (**b**) Axial CT scan shows the peripheral outgrowth with its cortex in continuity with the iliac bone (arrowhead). (**c**) Axial T1-w MRI image of the same lesion demonstrates the continuity of the cortex and medullary portion with the parent bone (arrowhead) and identifies a thin cartilage cap (asterisk)

cap (>2 cm), poorly defined cap, irregular calcifications, and rapid enhancement on dynamic Gd-enhanced MR images (less than 10 s after arterial enhancement) [44].

3.4.4 Fibrous Dysplasia

Fibrous dysplasia is an intramedullary hamartoma commonly observed in proximal femur and in pelvic bones [45]. It may be either monostotic or polyostotic, in the latter case associated with multiple endocrine abnormalities (McCune-Albright's syndrome) or intramuscular mixomas (Mazabraud's syndrome). Usually asymptomatic diagnosed as incidental finding, sometimes may cause pathologic fracture, discontinuous pain, deformity or lower limb discrepancy. The radiologic presentation is the same as in extremities, with well-defined defect rounded by a rind of bone sclerosis, radiolucency with "ground glass" appearance, thin and expanded cortex without periosteal reaction

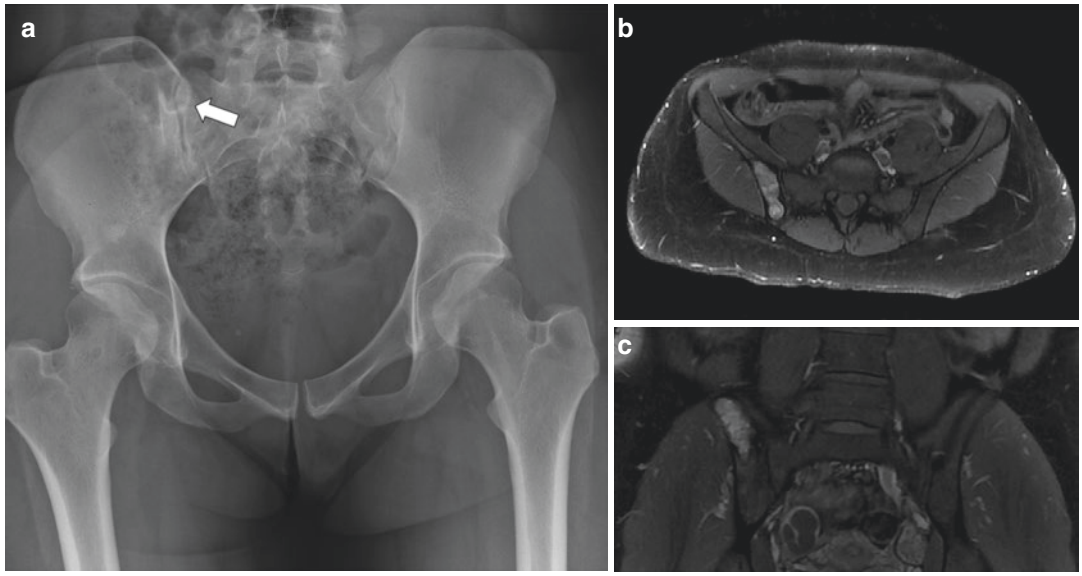

Fig. 3.7 Monostotic fibrous dysplasia in a 30-year old woman. (**a**) Lesions in the right ilium (white arrow) with mixed lysis and sclerosis with peripheral bone sclerosis. (**b**) Axial and (**c**) coronal T2-weighted fat sat image show heterogeneous lesion that is largely hyperintense with hypointense foci within the matrix. All images show the well-defined character and the absence of cystic components

(Fig. 3.7). Cystic cavities and cartilaginous area may be present and easily detectable on MRI, whereas the lesion appears with a fairly homogeneous low-signal in T1-w images.

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Most Common Histological Type of Pelvic Bone Tumors

4

Marilyn M. Bui and Andrew E. Rosenberg

4.1 Introduction

Previous study of 6000 patients had shown that patients with pelvic tumors are usually older, and their tumors are larger relative to patients with tumors in extremities. The majority of tumors in the pelvis are malignant (metastases, myeloma, chondrosarcoma, Ewing sarcoma, osteosarcoma, and malignant fibrous histiocytoma (MFH)/fibrosarcoma) [1]. The most frequent primary bone tumors of pelvis include chondrosarcoma (24%), Ewing sarcoma (16%), osteosarcoma (9%), malignant fibrous histiocytoma (MFH)/fibrosarcoma (5%), Langerhans cell histocytosis (4%), aneurysmal bone cyst (4%), fibrous dysplasia (4%), benign miscellaneous bone tumors (25%), and miscellaneous malignant bone tumor (8%) [1]. Combine with the literature review and authors' institutional experience, the pathology of 14 most common histological type of pelvic bone tumors will be discussed in the order of illustrating the gross and histological features of these tumors, highlighting the ancillary testing of diagnostic, prognostic, and predictive markers, and addressing the collaborative opportunities between pathologists and orthopedic surgeons to improve the quality, safety, and value of patient care.

Before getting into the details of each tumor type, few updates are worth mentioning. For malignant bone tumor staging, the American Joint Committee on Cancer (AJCC) eighth edition published in 2017 is generally used [2]. The updates relevant to pelvic bone tumor include: (1) Pelvis has a separate and distinct TNM classification but not a separate stage grouping. See Table 4.1 [2]. (2) Multiple myeloma and primary malignant lymphoma not staged using this staging system but rather the plasma cell disorders and lymphoma staging system. (3) Stage III is reserved for grade 2 (G2) and grade 3 (G3). (4) Grade 4 (G4) has been eliminated. Grade 1 (G1) is for low grade while G2 and G3 are for high grade. For the definition of the histological tumor types, currently we are using the WHO Classification of Tumors of Soft Tissue and Bone fourth edition where malignant fibrous histocytoma (MFH) is replaced by undifferentiated high-grade pleomorphic sarcoma and separated from fibrosarcoma which is a distinct entity.

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Table 4.1 AJCC Staging Version 8 for pelvis bone tumors

Primary tumor	Criteria
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor confined to one pelvis segment with no extraosseous extension
T1a	Tumor ≤8 cm in greatest dimension
T1b	Tumor >8 cm in greatest dimension
T2	Tumor confined to one pelvis segment with extraosseous extension or two segments without extraosseous extension
T2a	Tumor ≤8 cm in greatest dimension
T2b	Tumor >8 cm in greatest dimension
T3	Tumor spanning two pelvic segments with extraosseous extension
T3a	Tumor ≤8 cm in greatest dimension
T3b	Tumor >8 cm in greatest dimension
Regional lymph nodes	
NX	Lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Regional lymph node metastasis
Distant metastasis	
M0	No distant metastasis
M1	Distant metastasis
M1a	Lung
M1b	Other distant sites

4.2 Chondrosarcoma

Chondrosarcomas are a heterogeneous group, which includes the primary central, secondary central, periosteal, dedifferentiated, mesenchymal, and clear cell variants. The distinguishing hallmark of chondrosarcoma is the tumor cells producing cartilaginous matrix. The most common primary central chondrosarcoma is conventional type. Grossly the tumor has the appearance of hyaline cartilage. The histological criteria used for diagnosis include high cellularity, permeation of cortical and/or medullary bone, dysplastic chondrocytes, myxoid matrix or chodroid matrix liquefaction, necrosis, and increased mitotic activity [3]. The histological grade is the single most important prognostic factor of local recurrence and metastasis. The grading criteria for grade I to III based on the cellularity, nuclear features, and mitosis and the prognosis are summarized in Table 4.2. Myxoid change is seen in grade II and III tumors.

Table 4.2 Conventional Chondrosarcoma grading

Grade	Histology	Prognosis
I	Moderately cellular, mildly atypical nuclei without visible nucleoli under low magnification, no mitosis	Locally aggressive, rare metastasis, good prognosis
II	More cellular and greater degree of nuclear atypia than grade I, nucleoli can be found under high magnification, mitoses are seen	Poor prognosis
III	Hypercellular, markedly atypical nuclei with hyperchromasia, irregular nuclear contour, and enlargement, prominent nucleoli easily visible under low magnification, frequent mitosis including atypical figures	Worst prognosis

Conventional chondrosarcoma can undergo dedifferentiation and give rise to dedifferentiated chondrosarcoma variant. Grossly the demarcation of the cartilaginous and noncartilaginous components is readily recognizable. The dedifferentiate component is typically a high-grade undifferentiated sarcoma. Histology of a dedifferentiated chondrosarcoma is illustrated in Fig. 4.1a, b. The prognosis of this tumor is dismal. Heterozygous mutations of the isocitrate dehydrogenase 1 and 2 genes (*IDH1* and *IDH2*) are found in conventional chondrosarcoma as well as dedifferentiated chondrosarcoma (including the noncartilaginous component) [4]. This biomarker is potentially useful to distinguish a chondrosarcoma from a chondroblastic osteosarcoma. PB1 pathway (TP53) mutation is also a common event in both tumor types [4]. The features of rare variants of chondrosarcoma are summarized in Table 4.3.

4.3 Ewing Sarcoma

Ewing sarcoma is a family of tumor with neuroectodermal origin. It is a high-grade malignancy with small, blue, and round tumors cells

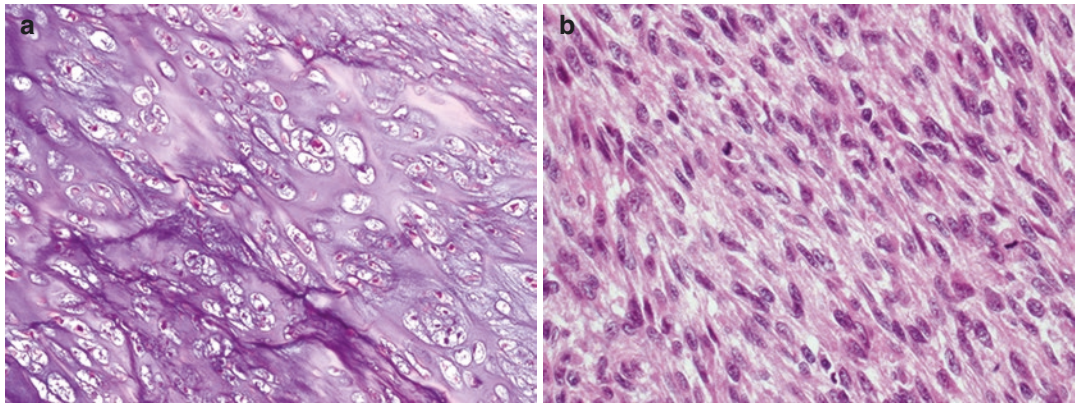

Fig. 4.1 Dedifferentiated chondrosarcoma. (**a**) Conventional low-grade chondrosarcoma component with cartilaginous matrix. (**b**) Dedifferentiated chondrosarcoma component composed of high-grade malignant spindle cells without cartilaginous matrix

Table 4.3 Summary of histological variants of chondrosarcoma

Tumor type	Component	Prognosis
Conventional	Chondrosarcoma Grade I Grade II Grade III	Depends on grade
Dedifferentiated	Low-grade conventional chondrosarcoma plus high-grade dedifferentiated sarcoma or osteosarcoma	Poor
Mesenchymal	Low-grade conventional chondrosarcoma plus poorly differentiated malignant small round cells	Poor
Clear cell	Clear cells or chondroblastoma-like cells	Depends on grade

showing pathognomonic molecular signatures which are pathogenesis drivers, including approximately 85% harboring a somatic chromosomal translocation t(11;22)(q24;q12) resulted in *EWSR1-FLI1* gene fusion [5]. This and other genes involved in Ewing sarcoma are summarized in Table 4.4. Molecular testing for the signature gene and products are useful in confirming the diagnosis. Molecular markers, such as TP53, telomerase expression, or CDKN2A loss, have shown prognostic significance [6].

Table 4.4 Summary of Ewing sarcoma gene fusions

Translocation	Genes involved
t(11;22)(q24;q12)	*EWSR1-FLI1*
t(21;22)(q22;q12)	*EWSR1-ERG*
t(2;22)(q33;q12)	*EWSR1-FEV*
t(7;22)(p22;q12)	*EWSR1-ETV1*
t(17;22)(q12;q12)	*EWSR1-E1AF*
inv(22)(q12q;12)	*EWSR1-ZSG*
t(16;21)(p11;q22)	*FUS-ERG*

Recently, a rare group of Ewing-like sarcoma is recognized and classified as undifferentiated round cell sarcoma by its genomic distinction. These tumors exhibit t(4;19)(q35;q13) or t(10;19)(q26;q13) with *CIC-DUX4* gene fusion or inv(X)(p11.4p11.22) with *BCOR-CCNB3* fusion. Their clinical behavior is also different from classic Ewing sarcoma [7, 8].

Grossly, the tumor has tan-gray cut surface without osteoid or cartilaginous matrix. A classic Ewing sarcoma is composed of hypercellular small round cells with scant cytoplasm and round nuclei arranged in a dyscohesive pattern. Neuroectodermal differentiation can be seen with tumor cells forming rosette-like structures. This small blue round cell pattern resembles lymphoma, osteosarcoma, rhabdomyosarcoma, and poorly differentiated carcinoma or neuroendocrine carcinoma. However, the cytoplasm of Ewing sarcoma appears clear and contains glycogen, which is stained positive by periodic acid-

Schiff (PAS). Immunohistochemical stain pattern of Ewing sarcoma includes positive vimentin, CD99 (membranous), Keratin (aberrantly expressed in 30% cases), neuroendocrine markers (aberrant expression sometimes), FLI-1, ERG, and NKX2.2 [9]. Comparing to lymphoma, Ewing sarcoma lacks the lymphoglandular bodies which are cytoplasmic debris of lymphoma. Osteosarcoma, especially the small cell variant, produces osteoid and lacks the Ewing sarcoma translocation. Rhabdomyosarcomas are immunoreactive to desmin, myogenin, and/or myoD1. Metastatic carcinomas are immunoreactive to cytokeratin. However, the differential diagnosis between a Ewing sarcoma with aberrant cytokeratin and/or neuroendocrine marker expression relies on the other immunostain markers listed above or molecular confirmation. A nonclassic Ewing sarcoma, the tumor cells are larger with more pleomorphic nuclei and prominent nucleoli. A radiological, gross, and histological illustration of Ewing sarcoma is in Fig. 4.2a–e.

Intraoperative evaluation of pathologic fracture of pelvis caused by Ewing sarcoma can be challenging, especially when there is no prior biopsy was performed. In our hands, touch prep cytology has been routinely used intraoperatively in conjunction to frozen section to facilitate a preliminary diagnosis of Ewing sarcoma and triage the tissue for molecular testing using air-dried slides [10]. In addition, we validated an antibody PRKCB which is a member of protein kinase C multigene family encoding serine/threonine kinases in our laboratory. This biomarker has 98% sensitivity and 96% specificity in detecting *EWSR1-FLI1* rearrangement, hence serves as a rapid and economic surrogate diagnostic marker for Ewing sarcoma. These quality improvement

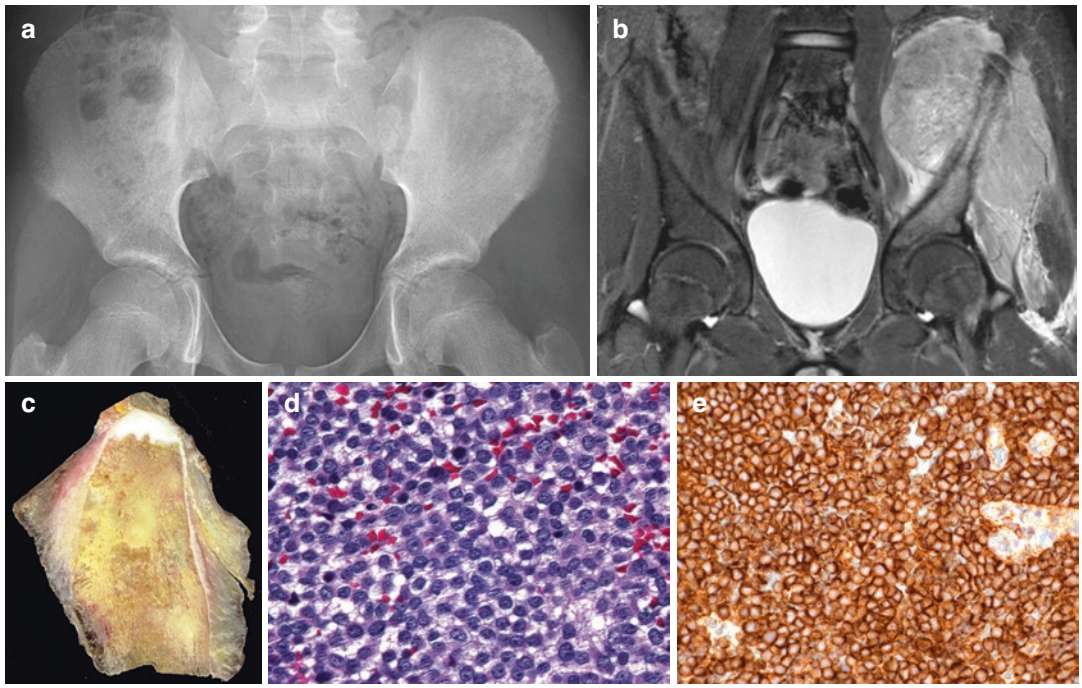

Fig. 4.2 Ewings Sarcoma. (**a**) Frontal radiograph of pelvis shows poorly defined lucent mass involving the left ilium. (**b**) Coronal stir-weighted MR image shows a large tumor arising in the medullary cavity and transgressing the cortices and forming large extra and intra pelvic soft tissue masses. (**c**) Coronal section through the ilium post chemotherapy shows that the bone is distorted by a necrotic yellow mass. The iliac crest is composed of white appearing cartilage as the patient is a child. (**d**) The tumor is composed of uniform, primitive appearing round cells that have fine chromatin and small nucleoli, and clear to eosinophilic cytoplasm. (**e**) Tumor cells show strong membranous staining for CD99 by immunohistochemistry

projects initiated by pathologists greatly improve the delivery of the care of Ewing sarcoma patients in our institution.

4.4 Osteosarcoma

Osteosarcomas are a heterogeneous group, which includes the primary central, secondary central, and surface of the bone, conventional, telangiectatic, and small cell variants. The distinguishing hallmark of osteosarcoma is the tumor cells producing osteoid matrix. The most common primary central osteosarcoma is conventional type which includes osteoblastic, chondroblastic, and fibroblastic variants. Osteoblastic osteosarcomas have a predominantly osteoid matrix, which can be thick or thin and branching. Chondroblastic osteosarcomas have a predominant chondroid matrix. Fibroblastic osteosarcomas produce only minimal amounts of osteoid and have high-grade spindled cell architecture. Telangiectatic osteosarcoma is characterized by having large blood-filled spaces, which are usually separated by thin septa. Although prognosis is thought to be similar to conventional osteosarcomas, they are much more sensitive to chemotherapy. Small cell osteosarcoma produces variable amounts of osteoid, and morphologically resembles Ewing sarcoma, but lacks the t(11;22) translocation. The characteristics of primary central osteosarcoma are summarized in Table 4.5. Surface osteosarcoma very rarely affects the pelvic bone. Grossly the conventional osteosarcoma shows hard tab-white cut surface, typically extends into the soft tissue. Histology of osteosarcoma is illustrated in Fig. 4.3a–d.

Table 4.5 Characteristics of histological variants of primary central osteosarcoma

Tumor type	Component	Prognosis
Conventional	High-grade sarcoma with osteoid formation Osteoblastic (76–80%) Chondroblastic (10–13%) Fibroblastic (10%)	High-grade tumor. Subtype does not differ in prognosis and therapy
Telangiectatic	High-grade osteosarcoma with characteristic blood lakes and spaces	Similar to conventional type
Giant cell-rich	High-grade osteosarcoma with abundant osteoclast-like giant cells	Similar to conventional type
Small cell	High-grade osteosarcoma with characteristic small tumor cells	Slightly worse prognosis than conventional type
Low-grade central	Low-grade osteosarcoma	Excellent prognosis

The pathologists' primary role is to make a definitive diagnosis of osteosarcoma and accurately classify and grade the tumor on preoperative biopsy samples. High-grade osteosarcoma is typically treated with neoadjuvant chemotherapy. The pathologists' second role is to evaluate the therapy response which is critically important for prognosis. Osteosarcomas with greater than 90% tumor necrosis (less than 10% viable tumor cells) are considered good responders and have better overall and disease-free survival [11]. The sampling of osteosarcoma includes cross-sectioning the central and largest slice of the tumor. The tumor slice is further divided into 1 cm × 1 cm slices and prepared for histologic examination. As a part of the therapy changes, tumor necrosis is documented in the pathology report, which is reversely related to the percentage of viable tumor cells as an independent prognostic of osteosarcoma.

Osteosarcomas are typically immunoreactive to CD99 which is a sensitive but not a specific marker. Osteocalcin is useful for highlighting osteoid. Recurrent amplifications at 1q21-23 and 17p are commonly seen, and comparative genomic hybridization analysis has revealed frequent chromosomal gains, such as the gain of 8q23, seen in about half of osteosarcomas [12]. CDK4 with or without MDM2 is commonly amplified in aggressive osteosarcomas. Patients with hereditary retinoblastoma (RB) and Li

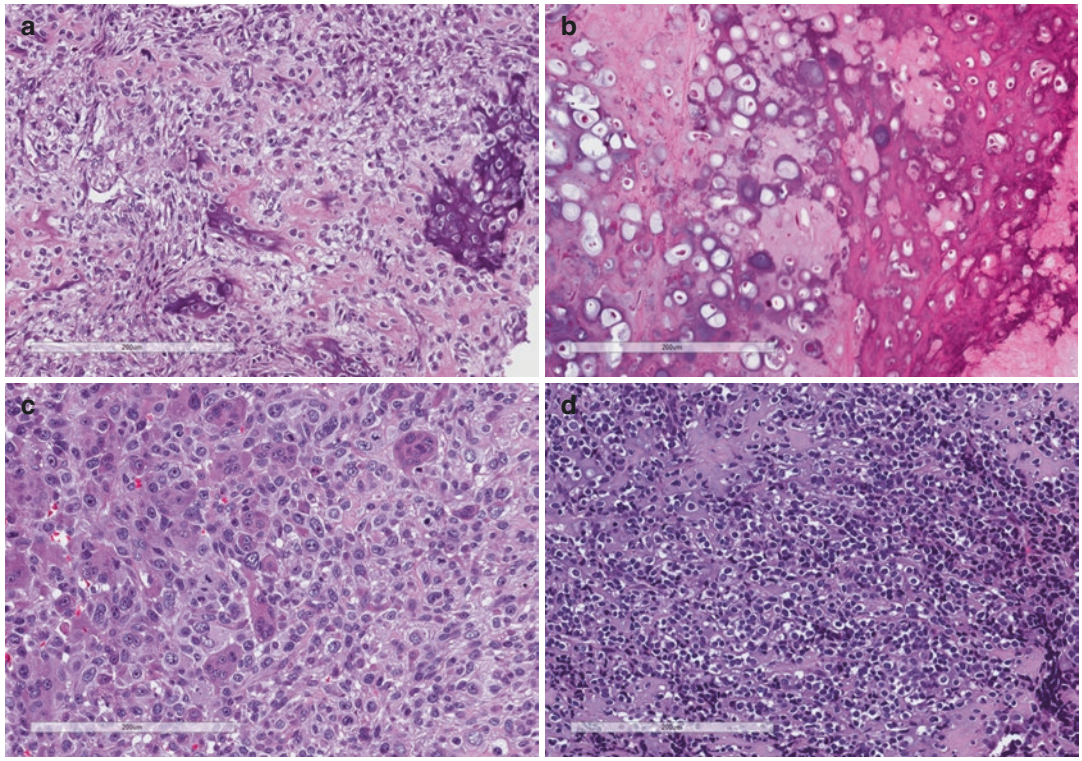

Fig. 4.3 Osteosarcoma. (**a**) Osteoblastic osteosarcoma showing malignant tumor cells producing osteoid matrix. (**b**) Chondroblastic osteosarcoma showing malignant cartilage. (**c**) Giant cell-rich osteosarcoma showing multinucleated giant cells. (**d**) Small cell osteosarcoma showing blue round tumor cells producing osteoid matrix

Fraumeni syndrome have an increased risk of developing osteosarcomas. RB1 alterations have also been seen in up to 40% of sporadic osteosarcomas, while TP53 alterations have been seen in up to 35% of osteosarcomas. Many genetic aberrations have been found in high frequency, some of which may offer prognostic value [13]. Osteosarcoma of pelvis, in the setting of Paget disease and radiation associated are of particularly unfavorable outcomes [14].

4.5 Multiple Myeloma/Plasma Cell Myeloma

Plasma cell myeloma commonly occurs in pelvis either as a primary tumor or as a part of multiple myeloma. For the patients with prior history of plasma cell myeloma, the diagnosis of this tumor in pelvic specimen is straight forward. The classic histology of myeloma includes round tumor cells have eccentrically located nuclei with abundant cytoplasm. With the increased tumor grade from well-differentiated, to moderately differentiated and to poorly differentiated myeloma, the tumor exhibits increased nuclear size, nuclear pleomorphism, prominent nucleoli, mitotic activity, and necrosis. The high-grade features resemble diffuse large B-cell lymphoma. However, myeloma cells are immunoreactive to CD38, CD138 (syndecan-1), and MUM1 with monoclonality of either kappa or lambda chain (kappa or lambda chain restriction). Flow cytometry is ideal for fresh tissue to confirm myeloma diagnosis.

On the other hand, intraoperative evaluation of pathologic fracture of pelvis caused by undiagnosed myeloma can be challenging. In our institution, touch prep cytology has been routinely used intraoperatively in conjunction to frozen section to facilitate a rapid diagnosis of myeloma [10]. The touch prep smears prepared from fresh

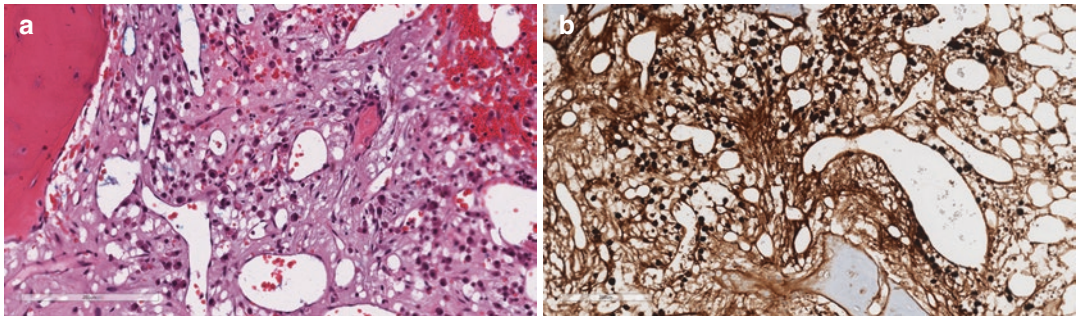

Fig. 4.4 Myeloma. (**a**) Neoplastic plasma cells display eccentrically located nuclei and abundant cytoplasm. (**b**) Tumor cells showing lambda chain-restriction by in situ hybridization

tissue are ideal to show the characteristic clock-face like nuclei and perinuclear hof of neoplastic plasma cells which are diagnostic of this tumor. Fresh tissue can then be triaged for flow cytometry study for confirmation. This practice has proven to be most accurate and efficient in managing this type of patients. The histology of myeloma is illustrated in Fig. 4.4a, b.

Genetically plasma cell myeloma has two distinct groups. One group (40%) harbors a balanced reciprocal translocation of the immunoglobin heavy-chain locus (*IGH*) with different partner genes including *FGFR3/MMSET* on 4p16.3, *CCND3* on 6p21, *CCND1* on 11q13, *MAF* on 16223, and *MAFB* on 20q11. Other group (60%) is hyperdiploidy with polysomes 3, 5, 7, 9, 11, 15, 19, and 21. *MYC* on 8q24 is associated with this group. The tumor prognosis is associated with multiple genetic markers [15].

4.6 Metastatic Carcinoma

Metastatic disease from carcinoma is common in pelvis. In the 4431 metastatic lesions registered in the archive of the Rizzoli institute, 833 (18.8%) were found to occur in the pelvic region including 559 (12.6%) are located in the ilium, 80 (1.8%) in the ischium, and 53 (1.2%) in the pubis [16]. The primary sites of the carcinomas include lung, breast, prostate, kidney, head and neck, and gastrointestinal tract. When a primary tumor is present, the diagnosis of metastatic disease is achieved by comparing the histology of pelvic lesion with the primary disease. However, when a primary site unknown or without a primary, histomorphology in conjunction with pertinent ancillary testing including immunohistochemistry are used to render a definitive diagnosis.

Intraoperative evaluation of pathologic fracture of pelvis caused by undiagnosed metastatic carcinoma can be challenging. "Epithelioid malignancy" diagnosis is not adequate to guide the optimal patient care in this type of clinical situation. In our institution, touch prep cytology has been routinely used intraoperatively in conjunction to frozen section to facilitate a rapid diagnosis [10]. The touch prep smears prepared from fresh tissue avoid of crush artifact, which is commonly seen in frozen section slides. The epithelial nuclei, glandular formation, intracytoplastic mucin, and squamous cytoplasm provide unequivocal evidence for the diagnosis of metastatic carcinoma intraoperatively. Accurate and immediate diagnosis of metastatic carcinoma will guide the surgeon to fix the pathologic fracture in the manner which is totally different from the fixation of a sarcoma caused pathological fracture.

4.7 Undifferentiated High-Grade Pleomorphic Sarcoma

Undifferentiated pleomorphic sarcoma, is a contemporary concept to include a group of high-grade sarcomas, has no identifiable line of differentiation when analyzed by current technologies [13]. Its histology is variable and may show different morphologic patterns composed

of spindle cells, pleomorphic cells, epithelioid cells, round cells, and multinucleated giant cells. Mitotic activity is typically prominent with atypical mitotic figures. Tumor necrosis can be seen. Due to the lack of consistent and identifiable diagnostic biomarkers, this group of tumor remains a diagnosis of exclusion. This is an aggressive malignancy with frequent metastases. Tumor necrosis in response to neoadjuvant chemotherapy provides important prognostic information. Further studies are warranted to reveal the prognostic and predictive biomarkers of this tumor.

4.8 Fibrosarcoma

Fibrosarcoma is a very specific diagnosis to include the tumor composed of intermediate- to high-grade fibroblastic spindle cells, which have littler genetic or molecular information [13]. Grossly, the tumor is firm and tan-white. The classic histology exhibits spindle cells devoid of significant pleomorphism arranged in "herringbone" pattern. There is no bone, cartilage or other line of differentiation other than fibroblastic. It is challenging to make a definite diagnosis of this tumor on limited biopsy specimen. Because this is a diagnosis of exclusion, the examination of the resected specimen is warranted. When a tumor has marked cytological atypia and storiform growth pattern, it should be classified as undifferentiated high-grade pleomorphic sarcoma. The prognosis depends on the patient's age, tumor grade, and stage.

4.9 Chondroma/Enchondroma

This is a benign hyaline cartilaginous tumor. Histologically it is hypocellular, avascular, with prominent hyaline cartilage matrix, and arranged in a multinodular architectural pattern. The chondrocytes are bland with no mitotic activity or necrosis. Heterozygous somatic mutations of *IDH1* and *IDH2* have been frequently identified in enchondromas as well as chondrosarcomas showing the genetic linkage of these two entities.

4.10 Langerhans Cell Histiocytosis/Eosinophilic Granuloma

Although this tumor most frequently involves the ribs, it also involves the pelvis as the primary site. This is a clonal neoplastic-like disease composed of Langerhans cells, which are specialized histiocytes with nuclear grooves (reniform nuclei), and admixed with inflammatory cells including prominent eosinophilia. The hallmark Langerhans cells are immunoreactive to CD1a, CD207/Langerin, and S-100, while negative for CD45. The identification of Langerhans cells are the key for diagnosis [3]. The prognosis for patient with monostotic or limited polystotic diseases is good.

4.11 Desmoplastic Fibroma

This is very rare, benign, and locally aggressive spindle cell tumor of fibroblastic origin. The histological features include bland spindle cells with abundant collagenous stroma, resemble desmoid tumor of the soft tissue. One main differential diagnosis is low-grade central osteosarcoma. The latter is typically positive for MDM2 amplification.

4.12 Aneurysmal Bone Cyst

Aneurysmal bone cyst is a benign tumor. Grossly it is well-defined and composed of blood-filled cysts, which lack specific cell-lining and consist of a wall of spindle cells with scattered osteoclast-type multinucleated giant cells. The neoplastic cells are spindle, which can be indistinguishable from reactive fibroblasts and myofibroblasts. However, the tumor cells show *USP6* rearrangement in 70% of the primary aneurysmal bone cyst, not the secondary ones [17]. The spindle cells are bland and lack of mitotic activity. Reactive woven bone may be seen with osteoblasts rimming. The main differential diagnosis is telangiectatic osteosarcoma, which is characterized by having large blood-filled spaces, but malignant tumor cells with osteoid formation. Solid variant of aneurysmal bone cyst may be diagnostically challenging.

However, *USP6* rearrangement testing can be used to confirm the diagnosis.

4.13 Giant Cell Tumor

Giant cell tumor of bone is a benign but locally aggressive and recurrent tumor. The tumor is composed of numerous characteristic giant cells which are large and osteoclast-like. These cells are the background cells reactive to the true neoplastic cells. The neoplastic cells are mononuclear primitive mesenchymal stromal cells expressing receptor activator for NF-κB ligand (RANKL), the master regulator of osteoclast differentiation. Macrophages and osteoclasts express RANK. The interaction between the neoplastic mononuclear stromal cells and marcophages/osteoclasts by a RANKL-dependent mechanism via the stimulation of macrophage-colony stimulation factor (MCSF) results in neoplastic proliferation and induces osteoclast formation. During this process, tumor-associated macrophage-like osteoclast precursors, which are also mononuclear cells, are recruited by tumoral stromal cells to participate in osteoclast differentiation and activation. Because osteoclast formation is the major consequence of giant cell tumor, inhibition of osteoclast formation and activity is the key for therapeutic approach. For example, bisphosphonate inhibits osteoclast-mediated resorption of bone/osetolysis and anti-RANKL antibody targets the RANKL-dependent mechanism of giant cell formation.

Osteoprotegerin (OPG) is a soluble decoy receptor that is produced by osteoblasts to inhibit osteoclast differentiation through its binding to RANKL, which prevents RANK binding. OPG expression reflects a protective mechanism of the skeleton to compensate increased bone resorption. Bone remodeling is mainly controlled by the balance of RANKL/OPG. Osteoprotegerin ligand (OPGL), also named receptor activator of RANKL is also expressed in the stroma-like tumor cells of GCTB. The ratio of OPGL/OPG by tumor cells may contribute to the degree of osteoclastogenesis and bone resorption [18].

Grossly the tumor is red-brown with hemorrhage and yellow areas reflect lipid laden macrophage-rich areas. Histologically, the tumor is composed of numerous multinucleated giant cells and scattered mononuclear cells that are round or spindle. Because H3-3A (H3F3A) gene mutation is common in giant cell tumor of bone (95%) and immunohistochemical study of H3.3 G34W is a reliable surrogate marker for this mutation. Immunostain of H3.3 G34W is useful in confirming the diagnosis of giant cell tumor of bone when other morphological differential diagnoses are considered [19]. Histological illustration of giant cell tumor of bone is in Fig. 4.5a, b. Lipid laden or hemosiderin laden macrophages are also present. The tumor is mainly solid and may contain cystic areas. Secondary aneurysmal bone cyst component is seen in 10% of giant cell tumor. The tumor may be mitotically active; however, a benign giant cell tumor typically does not have atypical mitosis or significant nuclear

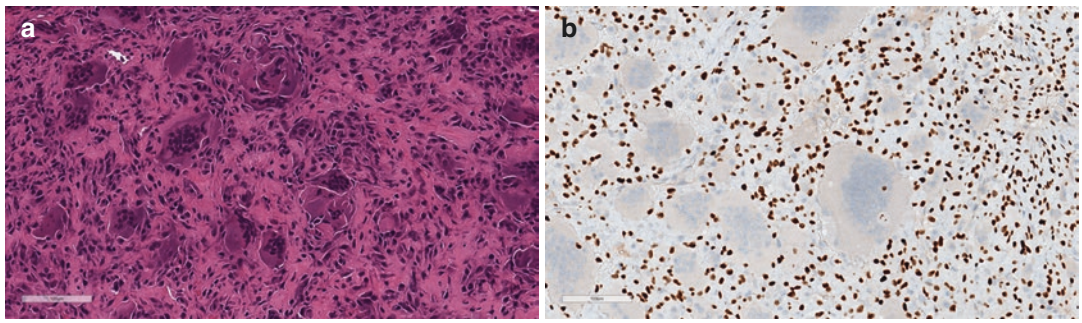

Fig. 4.5 Giant cell tumor of bone. (**a**) Tumor cells are mononuclear. The multinucleated giant cells are nonneoplastic. (**b**) The monocular tumor cells are highlighted by H3.3 G34W immunohistochemistry which is a reliable surrogate marker for underling molecular pathology

atypia. The latter is associated with a malignant transformation.

4.14 Fibrous Dysplasia

This is a benign fibro-osseous lesion that is caused by postzygotic activating missense mutations in the GNAS gene on 20q13 [20]. This mutation and fibrous dysplasia are also associated with McCune-Albright syndrome. Histologically, the tumor is composed of bland spindle fibroblastic cells admixed with irregular bony spicules without osteoblastic rimming. The irregular bony spicules have often been described as "alphabet soup" or "Chinese characters" in configuration [3]. The prognosis of monostotic fibrous dysplasia is excellent. Malignant transformation is exceptionally rare.

4.15 Osteoid Osteoma

This is benign bone-forming tumor. The nidus of the tumor consists of a combination of osteoid and woven bone surrounded by osteoblasts. The nidus is vascular-rich given the appearance of granulation tissue surrounded by sclerotic bone. The diagnosis of osteoid osteoma is typically straightforward with clinical and radiological correlation.

4.16 Conclusion

Pelvic bone tumors are a diverse group of benign, intermediate, and malignant neoplasm. Their histological type is designated based on their closest histological resemble of the normal mesenchymal tissue. Ancillary testing such as immunohistochemistry, cytogenetics, and molecular techniques have greatly improved the pathologic diagnosis of these tumors. Biomarkers that provide prognostic and predictive information are limited. Intraoperative touch prep cytology can be used to distinguish metastatic carcinoma and plasmacytoma in conjunction with frozen section, which provides valuable information for immediate management of pathologist fracture caused by these tumors.

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Benign Pelvic Bone Tumors

5

John E. Mullinax and G. Douglas Letson

5.1 Introduction

The primary diagnostic dichotomy for patients with bone tumors is separating a benign tumor from one that is malignant. Determination of a benign or indolent process from one that is malignant with potential for local or distant spread is crucial to the care of the patient. For those lesions deemed to be benign, the treatment decision then becomes a balance of intervention with observation. This balance is particularly important for the benign bone lesions that arise in the pelvis due to the anatomical constraints which often lead to increased morbidity with intervention relative to other sites of disease.

Classification of bone tumors at all sites is based on the Enneking classification. First published in 1980, this construct serves to delineate benign and malignant bone lesions based on clinical, radiographic, and pathologic characteristics [1]. Using a combination of the radiographic appearance, anatomic extent of the tumor, and the histologic grade, this system was the first to classify bone tumors based on their malignant potential, or lack thereof. In this system, those tumors considered to be benign or indolent were indicated by Arabic numerals (1, 2, and 3) as opposed to the malignant tumors which were indicated by Roman numerals (I, II, and III). The utility of this classification system is largely based on the easily ascertained variables for each lesion that quickly guide the intervention required.

The radiographic interpretation of bone lesions is fundamental and attempts to describe the rate of growth for each lesion. Benign or indolent lesions are considered those with either no growth or very slow growth pattern within the bone. The rate of growth in balance with the rate of osteoblastic activity yields hallmark findings on radiographs. The malignant potential of bone lesions was first described by Lodwick in 1980 using radiographic findings, followed by Madewell in 1981 using pathologic correlates [2, 3]. Later, Carraciolo et al. combined the findings of both classification systems with a separate dataset [4]. Termed the Modified Lodwick-Madewell classification system, the authors describe six separate categories of radiographic findings that correlate with, to an escalating degree, the malignant potential.

Apart from the imaging findings associated with the bone lesion of interest, the histologic diagnosis is paramount to understanding the behavior of each tumor. The spectrum of specific diagnoses is wide but well described by the World Health Organization Classification system. Using this pathologic categorical description, bone tumors are placed in groups based on the presumed tissue of origin followed by the degree

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of malignancy. Diagnoses are considered benign, low grade, or high grade depending on their rate of growth and probability of metastasis.

The purpose of this chapter is to focus on the evaluation and management of benign bone tumors that arise with the pelvis. Using the classification systems above, the scope is defined as tumors that are considered benign by the clinical Enneking classification, the radiographic Modified Lodwick-Madewell classification, and the pathologic WHO classification. Attention will be focused on the degree of intervention necessary for these tumors with the treatment philosophy as a balance between morbidity and need for elimination of the lesion.

5.2 Initial Evaluation of the Patient

The primary presentation of the patient with a pelvic bone tumor is generally considered either symptomatic or asymptomatic. For those with symptoms, a diagnostic strategy is focused by the presentation whether it is weakness, pain, restricted range of motion, or visceral compression (i.e., bladder or bowel dysfunction). In these cases, the bone tumor may be a coincidental finding or one that is directly contributing to the constellation of symptoms. Patients with asymptomatic bone tumors of the pelvis are often discovered during an imaging modality performed for an unrelated reason. The two scenarios are important to distinguish based on the threshold degree for intervention as the asymptomatic patient has a much higher likelihood of morbidity relative to their preoperative state.

The clinical exam of the patient with a pelvic bone tumor is of fundamental importance to delineate an effective and cost-effective diagnostic strategy. Primary focus should be on focal weakness of muscle groups, range of motion, and neurovascular deficits. The pelvic anatomy is such that assessment of visceral function is also an important factor for clinical exam. Impairment of bowel, bladder, or sexual function may suggest compression of the viscera directly or neurologic compression. The vascular supply of the pelvic viscera and lower extremities courses through the pelvis and therefore a comprehensive examination should evaluate any lower extremity symptoms (i.e., claudication or rest pain) that might be secondary to arterial compromise. Additionally, in the lower extremities, unilateral edema is an important finding as it may suggest pelvic venous compromise with or without an element of thrombosis.

The diagnostic imaging modalities chosen for evaluation should be driven by the clinical exam but there are some general caveats. In those patients suspected to have a bone tumor within the pelvis, a plain radiograph combined with a contrast-enhanced MRI is preferred. The findings on plain radiograph will allow for discrimination of malignant potential and the contrast-enhanced MRI will allow for assessment of the peritumoral soft tissue. Utilization of axial imaging without plain radiograph will not allow for comprehensive evaluation of a pelvic bone lesion. In those cases with visceral compromise suspected by clinical exam, contrast-enhanced CT scan is also recommended to assess for external compression. A detailed gastroenterological or urologic exam should also be considered for those with bowel and bladder dysfunction, respectively, as symptoms may be coincidental rather than caused by the pelvic bone tumor.

In addition to radiographs and axial imaging, functional imaging is important for many bone lesions. Positron-emission tomography (PET) combined with CT imaging is helpful to describe the metabolic activity of a bone lesion. PET/CT imaging can be most useful in the patient with a history of malignancy and new pelvic bone lesion. Low metabolic activity would suggest a separate, sporadic process while increased metabolic activity would suggest possible metastatic disease from the historical primary malignancy. Another functional imaging modality is bone scintigraphy, or "bone scan." Using radiolabeled methylene diphosphonates, the degree of bone turnover and perfusion is assessed which can aid in the interpretation of the complementary imaging modalities [5].

At the conclusion of the diagnostic evaluation, including the clinical exam and imaging modalities, patients found to have a benign bone lesion

should be categorized as either symptomatic or asymptomatic. Intervention should be considered more strongly for the former and the focus of intervention should be on improvement of symptoms. Compressive symptoms are more easily addressed with surgical intervention while pain symptoms are less likely to have a durable response. In the asymptomatic patient, surgical intervention should be contemplated relative to the Ennenking classification with an approach that escalates with higher grade. Observation of grade 1 lesions is an optimal approach while curettage is appropriate for Grade 2 lesions. The addition of a surgical adjuvant to curettage such as methyl methacrylate or liquid nitrogen (cryotherapy) should be considered for Grade 3 lesions. Within these two groups of patients (Grade 2 and 3), the specific histologic diagnosis of the benign bone tumor is the most important factor that impacts the decision to operate. The remainder of this chapter focuses on the surgical approach relative to the different subtype diagnoses.

5.3 Chondrogenic Tumors

5.3.1 General Considerations for Cartilage Tumors of the Pelvis

Benign cartilaginous tumors of the pelvis are rare. Clinical suspicion of chondrosarcoma should be high with any bone lesion of the pelvis that suggests cartilaginous origin on imaging. At a minimum, a cartilaginous lesion of the pelvis with benign radiographic appearance warrants interval imaging follow-up. Any change in the characteristics of the lesion should warrant resection rather than curettage to avoid an intralesional procedure on a malignant cartilaginous lesion which can have disastrous oncologic effects. In a similar manner, percutaneous biopsy of bone lesion in the pelvis with features consistent of cartilage origin should generally be avoided. The gross consistency of chondrosarcoma is often semisolid and thus perforation of the tumor with biopsy needle can lead to local contamination of the tissue planes. If percutaneous biopsy must be undertaken, the procedure should be done in concert with the treating surgeon so that the surgical approach is considered relative to the planned biopsy tract.

5.3.2 Enchondroma

A common benign cartilaginous tumor arising within the medullary canal of long bones is an enchondroma. These tumors can be radiographically well-demarcated in the small bones of the hand but can also present with ill-defined borders in long bones. These are most often discovered incidentally on plain radiographs. Follow-up imaging with MRI characteristically demonstrates a hyperintense mass on T2-weighted imaging sequences.

As most are discovered incidentally, most do not need any therapy. Indications for intervention include symptoms that can be attributed directly to the lesion or any features of malignancy. Radiographic features of malignancy would include permeative appearance in the surrounding bone, periosteal reaction, or adjacent soft tissue mass. Biopsy is not generally required given the characteristic imaging findings but if tissue is obtained, pathologic findings concerning for malignancy would include nuclear atypia or increased myxoid component among a hypercellular hyaline cartilage stroma.

5.3.3 Chondroblastoma

Typically involving the epiphysis, this benign cartilaginous tumor is a rare entity arising from within the long bones. They typically present with benign radiographic appearance (sclerotic borders) but may also have a periosteal reaction. The gross appearance is consistent with the infiltrate of mononuclear cells characteristic to this tumor. The degree of atypia is variable on pathologic analysis of these lesions and interpretation of the histologic findings relative to the radiographic findings is helpful in confirming the diagnosis.

Chondroblastoma is often a symptomatic lesion and thus intervention is preferred. One approach to treatment is an intraoperative biopsy to confirm the diagnosis followed by curettage and bone graft. Limiting the dissection of tissue planes on approach to the lesion should be a focus of operative planning and thus, in the event the lesion is found to be malignant, proper resection can be undertaken without a wide field of contamination. This can be accomplished using a minimally invasive, percutaneous technique under fluoroscopic guidance (Fig. 5.1). In addition to curettage, others have described intraoperative adjuvant therapy such as cryosurgery with good functional outcomes and low recurrence rate [6].

5.3.4 Chondromyxoid Fibroma

Unlike the prior chondrogenic tumors discussed, this benign cartilaginous neoplasm most commonly presents with pain and can be found in the iliac wing. The radiographic appearance demonstrates a scalloped lesion, often with sclerotic noncontiguous borders. As most are symptomatic, intervention is preferred for these lesions. Resection is indicated for these lesions as there is significant overlap with chondrosarcoma based on the imaging characteristics and even pathologic assessment of small tissue quantity obtained with biopsy (Fig. 5.2). Understanding this risk of an underlying malignancy, resection of an intact tumor is preferred to curettage.

5.3.5 Osteochondroma

Arising as outgrowth from the bone, osteochondromas are the most common benign bone lesion. These lesions are considered a benign cartilaginous tumor with continuity to the cortex and medullary canal. These lesions characteristically arise pointing away from a joint and have a pathognomonic cartilaginous cap. Also termed exostoses, they may arise as a single lesion or in multiple lesions. Approximately 15% of patients with multiple lesions have an underlying germline mutation in the tumor suppressor genes EXT 1 or 2 which results in the syndrome known as hereditary multiple exostosis [7].

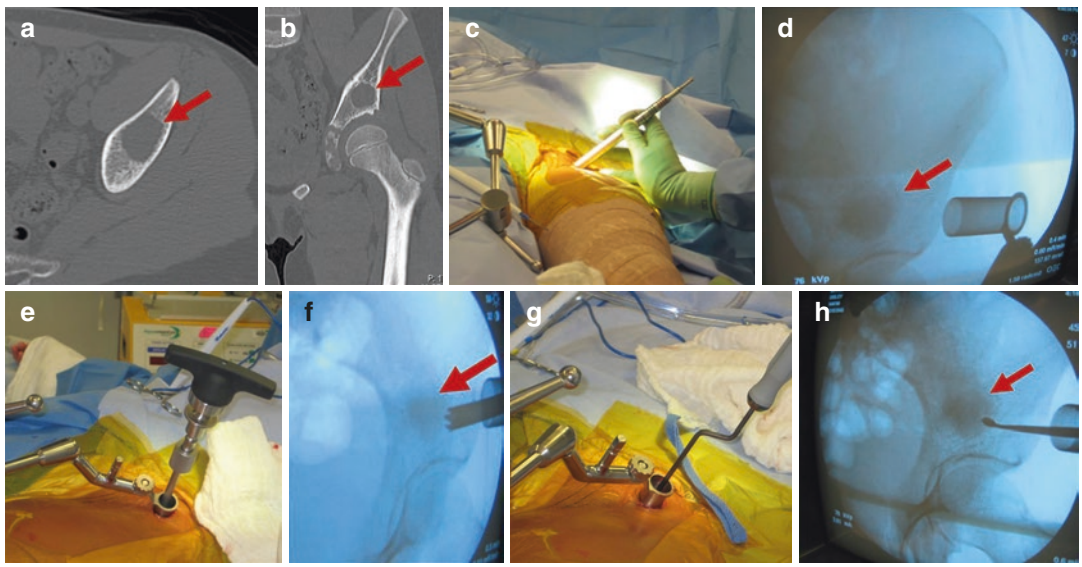

Fig. 5.1 Percutaneous curettage of pelvic chondroblastoma. CT demonstrates a pelvic lesion with benign appearance in the axial (**a**) and coronal (**b**) plane. Intraoperative placement of a canula under fluoroscopic guidance (**c**, **d**) allows for dissection through the soft tissue with minimal displacement of normal anatomic planes. Biopsy is undertaken in a coaxial fashion under fluoroscopic guidance to prevent contamination of surrounding soft tissue (**e**, **f**). Once diagnosis of benign lesion is confirmed, curettage is performed through the percutaneous access canula (**g**, **h**). (Red arrow = bone lesion)

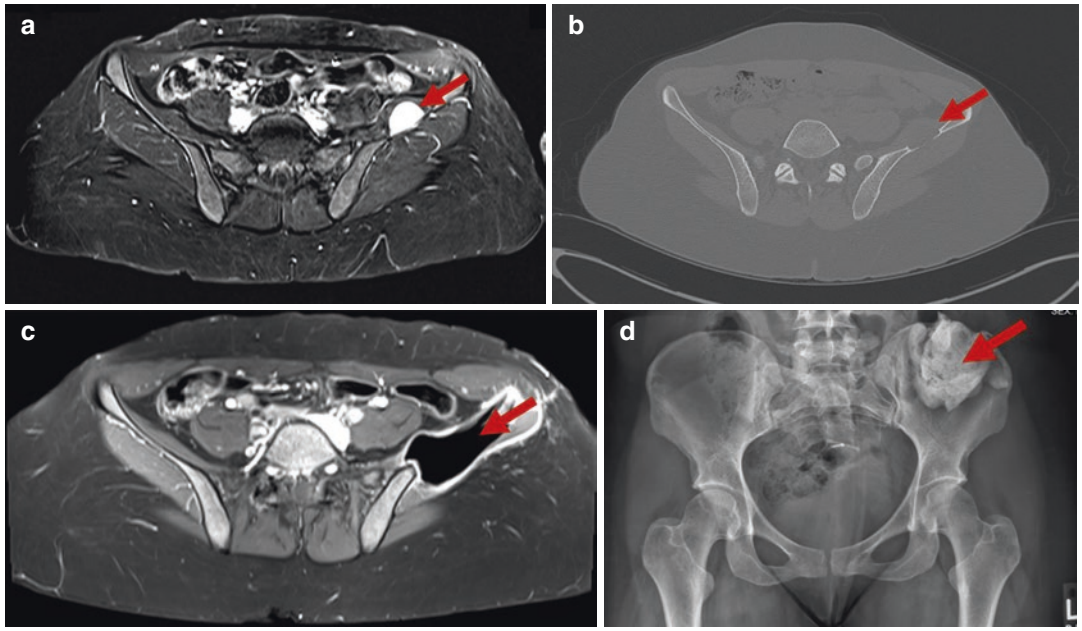

Fig. 5.2 Surgical management of pelvic chondromyxoid fibroma. Preoperative images demonstrate a lesion within the left iliac wing on MRI (**a**) and CT scan (**b**). Following resection with placement of bone cement, the postoperative MRI (**c**) and radiograph (**d**) demonstrate resolution of the lesion

Intervention is reserved for symptomatic lesions and those that harbor features concerning for malignancy on imaging. The cartilaginous cap, a hallmark feature of this lesion, should be <2 cm. Features that would raise concern for a malignancy rather than benign lesion include, rapid growth of the lesion, a cartilage cap >2 cm, or loss of corticomedullary continuity. These findings warrant biopsy before planned excision. Once the diagnosis is confirmed as an osteochondroma rather than a malignant bone lesion, the removal of these lesions does not require an extensive resection but rather excision of the pedunculated lesion from the bone involved. Care should be taken to ensure the lesion is removed entirely, all the way to the base.

5.4 Osteogenic Tumors

5.4.1 Osteoid Osteoma

Arising within the diaphysis or metaphysis of the long bones, osteoid osteomas demonstrate a characteristic finding on plain radiographs. The central, lucent nidus surrounded by a sclerotic peripheral zone is readily identified on plain radiographs and confirmed with axial imaging in the form of a CT scan. These lesions are typically found in males between 10 and 20 years old and are, by definition, <2 cm in size. The clinical scenario most often involves pain at night which is often relieved by aspirin. Intervention is not generally necessary for these patients but, when required, can be in the form of curettage or simple excision. More recently, a percutaneous technique of radiofrequency ablation (RFA) has been advocated [8, 9]. Technical constraints of this approach limit RFA to lesions <2 cm and thus osteoblastomas (below) are not approached in this manner. When the latter is performed, pathologic confirmation of the diagnosis involves histologic identification of the nidus.

5.4.2 Osteoblastoma

Compared to an osteoid osteoma, an osteoblastoma has the same underlying histologic findings with the primary clinical difference being the size

of the lesion. While an osteoid osteoma must be <2 cm in size, an osteoblastoma will be larger in maximal diameter. Additional differences include the predilection of osteoblastomas to occur within the spine and the lack of a clear nocturnal pain cycle as is classic for osteoid osteomas. The indications for treatment are not different, though, due to the size of the lesion, morbidity can be higher. Planning the surgical resection with respect to the preoperative symptoms is important caveat to treating these lesions.

5.5 Giant Cell Tumor

With the alternative osteogenic tumors arising from an over proliferation of osteoblasts, the giant cell tumor (GCT) of bone is one characterized by an overproliferation of osteoclasts. The underlying biology is centered on the disruption of normal bone remodeling physiology where the osteoclasts proliferate in response to Receptor activator of nuclear factor kappa-B ligand (RANKL) which is physiologically secreted by osteoblasts. In GCT, the neoplastic stromal cells secrete RANKL without negative feedback, thereby driving the tumor growth.

The majority of GCT have an indolent, even benign clinical course but up to 5% may develop distant metastatic disease, primarily in the lung [10]. For those with localized disease, there have been several interventions described. Extended curettage is the preferred approach and can produce excellent long-term results (Fig. 5.3). Several adjuvant treatments are described to reduce local recurrence rates such as methyl methacrylate [11]. Radiation therapy has also been described in the adjuvant setting but long-term follow-up has not demonstrated local control benefit [12].

Others advocate the use of liquid nitrogen as a surgical adjuvant [13]. Complete resection is associated with a recurrence rate <20% but this approach, especially in the pelvis, must be considered relative to the proposed deficit associated with the resection. Owing to the unique pathophysiology of the RANKL-axis in the development of these tumors, neoadjuvant treatment with denosumab has been advocated to decrease the size of the tumor and therefore the extent of resection. Denosumab is a monoclonal antibody which binds RANKL, thereby interrupting the positive stimulation signal between the neoplastic stromal and osteoclasts within the tumor [14, 15]. Preoperative treatment with denosumab is of primary importance for lesions with a large soft tissue component as bone formation at the margins can assist with resection and limit intraoperative morbidity. Conversely, lesions with a planned intralesional (curettage) approach should not receive preoperative denosumab as this treatment is associated with bone formation at the reactive zone, thereby precluding complete curettage. Finally, radiation has been described for the treatment of large, locally advanced lesions but concern with subsequent development of radiation-associated sarcoma limits the broad use of this approach.

5.6 Cystic Disease of the Bone

5.6.1 Unicameral Bone Cyst (UBC)

Simple cystic lesion within the bone most commonly arises in the adolescent and young adult population with 80% arising within the second decade of life. Termed unicameral bone cysts (UBC), these lesions most often present in the long bones with predilection for the proximal humerus and femur. Clinical presentation can be either incidental or within the context of long bone fracture. In those with the latter, spontaneous resolution can result in 15% of cases owing to the bone remodeling associated with fracture repair. In those with incidental findings, the decision to intervene is primarily one of impending fracture risk.

Distinguishing these lesions from more complicated cystic disease such as aneurysmal bone cyst (ABC) is important prior to treatment. Following identification on plain radiograph, CT scan is helpful to determine the thickness of the cyst wall and consequent fracture risk. Intervention on UBC should address the impending fracture risk without imparting undue morbidity. For this reason, excision or en bloc

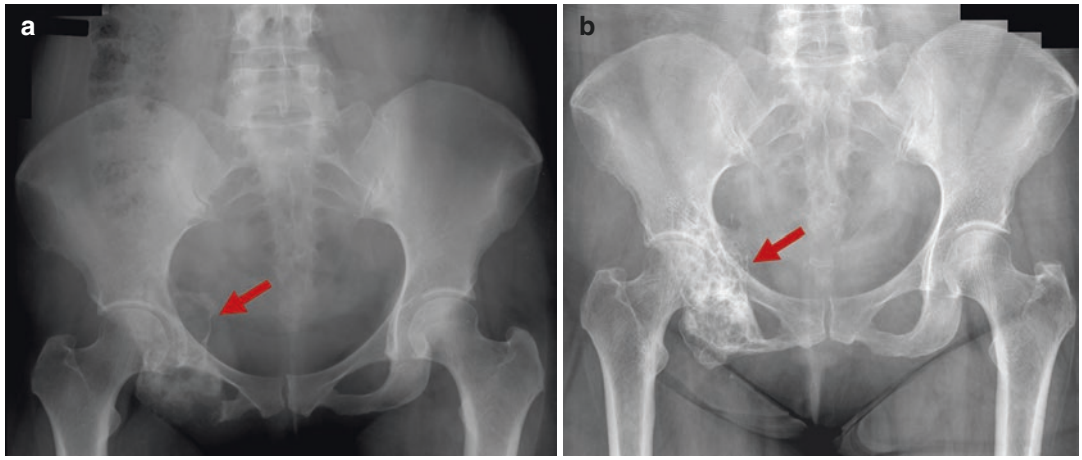

Fig. 5.3 Giant cell tumor long-term follow-up. Treatment of a pelvic giant cell tumor (red arrow) with curettage and adjuvant radiation therapy (**a**). Long-term follow-up with stability and remodeling evident on radiograph 24 years after the index procedure (**b**)

resection is not indicated but rather intervention should address the cystic cavity to allow physiologic bone remodeling. Following aspiration to confirm the diagnosis, injection of methylprednisolone acetate has been suggested as a low risk, even percutaneous, intervention. This procedure may also be repeated for those lesions that do not respond to a single injection [16]. More recently, this approach has been supplanted by curettage with bone graft for pelvic UBC. Given that the UBC will resolve once growth plates close, treatment should only be considered in the young (<10 years) patient while observation is preferred in the adolescent patient.

5.6.2 Aneurysmal Bone Cyst (ABC)

A more complex cystic lesion within the bone is the aneurysmal bone cyst (ABC). These lesions, rather than simple cystic lesions, are comprised of multiloculated regions of abnormal angiogenesis within a larger area of bone destruction. Like UBC, these lesions most commonly arise within the long bones but a larger percent (9%) arise within the pelvis. In addition, while UBCs are more commonly restricted to the bone, ABC can have a large soft tissue component emanating from the primary lesion within the bone. In the pelvis, this soft tissue component can generate significant symptoms related to compression of neurovascular structures, bladder, or bowel.

The diagnosis of ABC must be confirmed, apart from a malignant lesion, prior to intervention. Approximately 30% of ABC are described as "secondary" and related to prior trauma or associated adjacent malignancy. Identification of a possible underlying malignant process associated with the ABC is important to avoid an intralesional procedure with resulting tumor dissemination. For the primary ABC, the primary pathologic diagnostic dilemma is distinguishing these lesions from telangiectatic osteosarcoma (TO) with the latter requiring multimodal therapy and surgical intervention with oncologic intent. Histologic differences associated with TO include atypical cells with increased mitoses. Molecular pathology is helpful to discriminate the two diagnoses as well with ABC containing a characteristic gene rearrangement in the USP6 gene [17].

Unlike the focus on intracystic treatment as with UBC, the only treatment with guaranteed complete resolution is wide resection. Often, the size of these lesions precludes complete resection due to the associated postoperative deficits in the setting of a benign process. Intralesional therapy, if undertaken should not be with methylpredniso-

lone as in the case of UBC but rather with ethanol or doxycycline [18, 19]. If surgical intervention is required or contemplated, a strong consideration should be given to preoperative embolization, especially for pelvic lesions to mitigate the intraoperative blood loss which has frequently encountered with these lesions.

5.7 Hemangioma

Benign vascular tumors arising within the bone are a common finding in the adult population with approximately 10% of patients found to have a vertebral body hemangioma on imaging performed for another reason [20]. Imaging is characteristic with plain radiograph demonstrating pathognomonic mineralization in the form of phleboliths within the larger lesion. On cross-sectional imaging, this mineralization is again evident in a "polka dot" pattern. Often, the imaging findings are diagnostic, and no biopsy is needed for these lesions which have universally benign radiographic appearance.

Bone hemangiomata are most often incidental findings without symptoms and treatment is not required for these lesions. Serial imaging over a period of 2 years is warranted to ensure stability. Large vertebral lesions may require intervention due to compression at spinal nerve roots or when impending vertebral body fracture is of concern. In these cases, curettage with stabilization is preferred to resection. Symptomatic lesions in the long bones can be treated with curettage or embolization, the latter most easily repeated for nonresolving lesions.

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Chondrosarcoma of the Pelvis

6

Gerhard M. Hobusch and Reinhard Windhager

6.1 Epidemiology, Presentation, Diagnosis

CS is a rare disease with an estimated incidence of 3.4–5/1,000,000/year according to Netherlands cancer registry from 1989 to 2013 including 2186 patients, the American SEER database from 1973 to 2003 including 2890 patients or the Vienna Bone, and soft tissue tumor registry from 1965 to 2019 including 395 patients with CS [1, 2]. According to musculoskeletal-oncologic high-volume centers, one quarter of all CSs occur in the pelvis, thereby around 33–80% in iliac wing, 13–50% in the acetabular region, and each 5–17% in the pubis as well as 5% in the adjacent sacrum [3, 4]. There is a male dominance of 55% and the tumor appears mainly in adults and only seldom in children and young adults [5].

CS in the pelvis grow deep inside and can therefore stay undetected and symptomless for a long time, thus becoming huge masses [6]. Especially in low-grade tumors, gradually increasing pain is the most common presenting symptom [7]. CS can be frequently linked to compression of pelvic organs as bladder, prostate, or bowel. Olivieri et al. presented a single case of advanced CS with even urinary obstruction by the tumor [8]. Depending on localization, the regular digestive function is impaired and voiding disorders and sexual dysfunction can occur (see Fig. 6.4). The huge size and anatomic involvement of neurovascular abdominal as well as urological structures at the time of diagnosis can create big challenges to both limb-salvage as well as to ablative surgery [6, 9, 10]. A diagnostic preoperative CT- or ultrasound-guided biopsy is mandatory, even though preoperative histological grading should be interpreted with great caution in the pelvis, although it is reliable in the CS of the long bones. The concordance between the preoperative biopsy and the final pathological analysis in terms of histological grade in pelvic CS with 36% is much lower than in long-bone CS with 83% [11]. Independent of bioptic results, a wide resection is recommended in all cases of pelvic masses because of unreliable prediction from biopsy and preoperative imaging [12]. All surgical steps as the resection as well as the reconstruction should be planned as well as executed by a multidisciplinary team.

6.2 Imaging

Both plane radiographs as well as layered imaging in terms of MRI and CT with facultative 3-D reconstructions are necessary in the management of diagnosis and increasingly in the treatment of pelvic CS. Apart from imaging-differences

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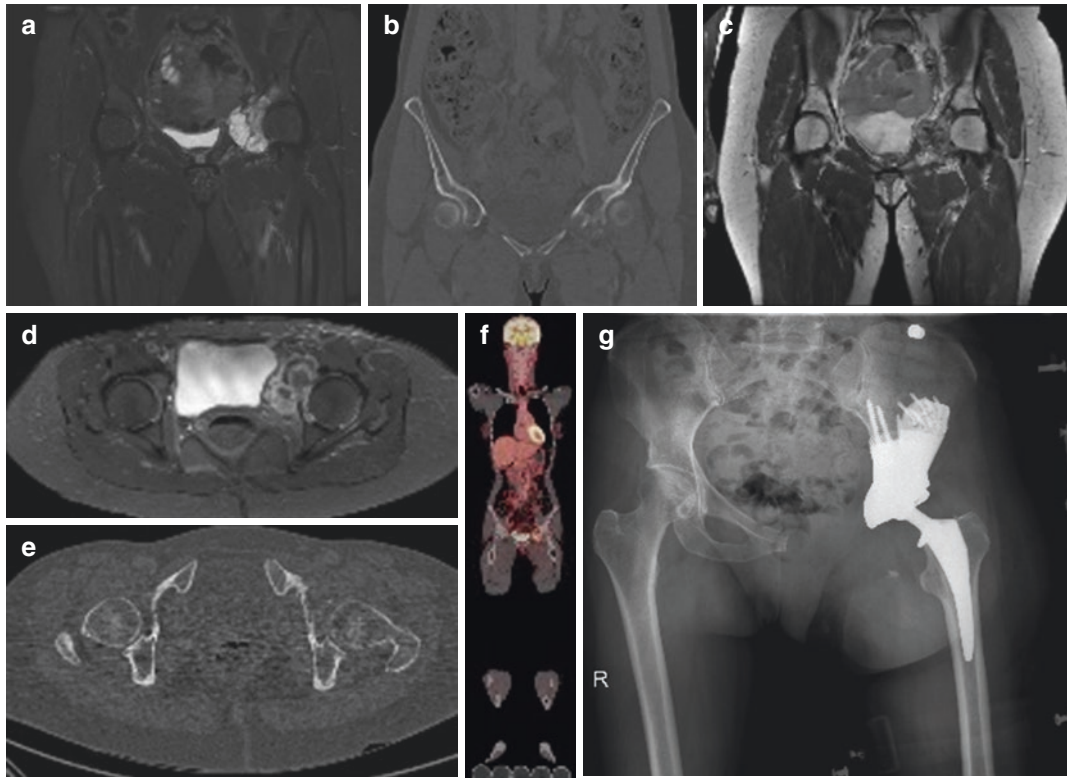

Fig. 6.1 Pre- and postoperative radiologic series of a 50-year-old female patient with acetabular chondrosarcoma G3 and after II + III resection of the left hemipelvis and reconstruction with a custom-made pelvis prosthesis. (**a**, **c**) Preoperative coronar pelvic MRI (STIR and T1 TSE Dotarem), (**b**, **e**) preoperative coronar and transversal CT, (**d**) preoperative transversal MRI (T1 TSE Dotarem), (**f**) FDG PET-CT whole body, (**g**) postoperative ap pelvic X-ray with custom-made pelvis prosthesis. X-rays ap of a 51-year-old patient with a 17-year follow-up after II + III resection of the right hemipelvis and reconstruction with a custom-made pelvis prosthesis after pelvic CS G2

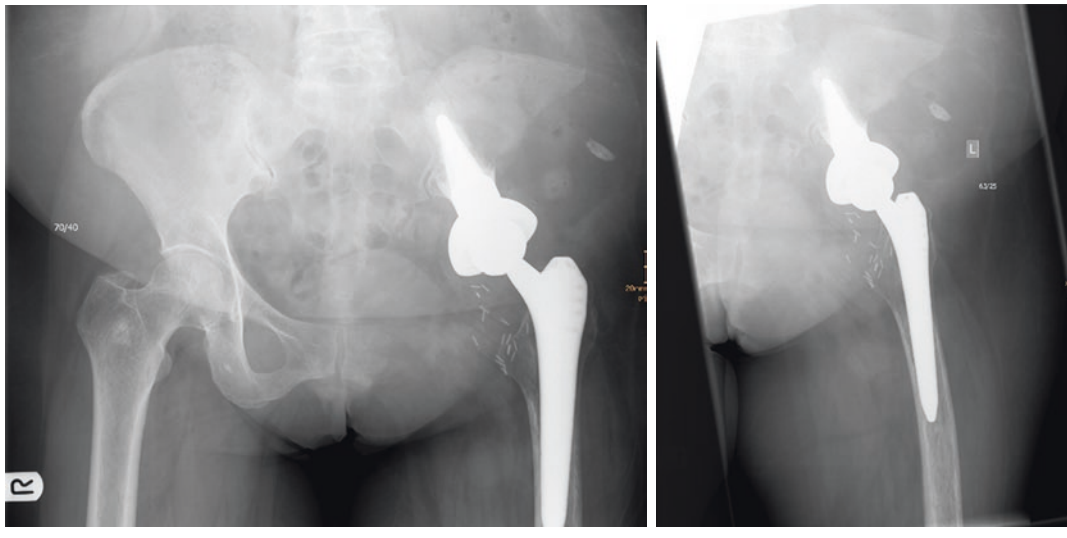

Fig. 6.2 X-rays ap of a 58-year-old female patient with a 6-months follow-up after G3 dedifferentiated CS, type 2–3 resection left hemipelvis and reconstruction with a stemmed acetabular cup prosthesis

between subtypes of chondrogenic lesions, there are common radiological diagnostic features like the typical chondroid matrix-calcification (ring-and-arc-sign) and lobular-type architecture and endosteal scalloping [13]. Along with these mixed lytic and sclerotic patterns, more aggressive patterns of bone destruction like moth-eaten and permeative lysis can be seen [13]. Brenner et al. argued a higher standardizes uptake value (SUV) in CS patients developing recurrent or metastatic disease suggesting that as a preoperative staging tool, FDG-PET may provide improved outcome prediction in combination with histopathologic findings [14]. Imaging is becoming increasingly important in the surgical planning as well as perioperatively as CT or fused preoperative MRI and CT images provide computer-assisted navigation in surgical resections to reduce the risk of intralesional resection and to avoid too-wide margins to achieve better functional outcomes. Gerbers et al. demonstrated a successful hip-joint saving surgery of a patient suffering from a periacetabular CS by optimizing surgical margins with computer navigation [15, 16].

6.3 Pathology

In the pelvis, the conventional central CS is the most dominating subtype in about 47–80% and arises in normal bone. It can be distinguished from peripheral (secondary) subtypes in 30% that occur as a malignant transformation of bone surface-based osteochondroma or enchondroma. The risk of malignant transformation in a solitary lesion is estimated at up to 1% in comparison to multiple lesions at up to 5%. A further distinction can be made between rare subtypes like the dedifferenciated central and peripheral CS in 1.4–15% and considerably rarer the periosteal, clear cell, and mesenchymal CS, in all together 2% of pelvic CS [2, 3, 6, 17]. Grading of CS is basically determined by cellularity, size and shape, and number of nuclei and mitotic figures and although grades 2 und 3 can be well distinguished, it is sometimes hard in differentiating a benign cartilaginous lesion from a well-differentiated grade 1 CS. On the other hand, different proportions of low grade (18–26%), grade 2 (37–61%), and grade 3 (8–22%) [4, 10, 18] tumors among different published case series may indicate a large variance in various regions in the world or difficulties in tumor grading.

6.4 Surgical Treatment

According to the literature, hind quarter amputation, denoting the unilateral pelvic leg amputation, is done in 13–52% of the CS patients in different tumor centers [4, 6, 17, 19, 20]. Limb-salvage in pelvic CS patients is considered the gold standard nowadays and is supported by the fact that limb-savage surgery is not a risk factor for low survival [21]. Furthermore, functional outcome shows better results after inner hemipelvectomy/limb salvage with MSTS93 between 61.4% and 86.6% than after external hemipelvectomy with MSTS93 score of 20–37.6% in several case series [10, 20]. Due to the predilection sites of pelvic CS, inner hemipelvectomies can be performed often instead of forequarter amputation. The Enneking–Dunn classification for internal hemipelvectomies divides the pelvic bone into three sections and the portion of the sacrum adjacent to the ilium as the fourth section and thus describes the anatomic regions of resection [22].

Type I (iliosacral) resections depend on whether the continuity of the pelvic ring can be preserved (Type IA versus Type IB). In type IB resections, there is agreement not to reconstruct the defect; however, even in Type IA resections, Beadel et al. promote a less mandatory restoration of the pelvic ring after iliosacral resections due to good clinical results in a case control study with mixed sarcoma. The authors report similar functional scores like TESS 72% (55–100) in the non-reconstruction compared with TESS 68% (59–86) in the reconstruction group. Furthermore, the chronic pain medication in the reconstruction group was higher, the mean surgery time was 25% shorter (463:613 min), and the mean blood loss was 30% less (4325:6250 ml) in the non-reconstruction group. Authors conclude that complications were more common after recon-

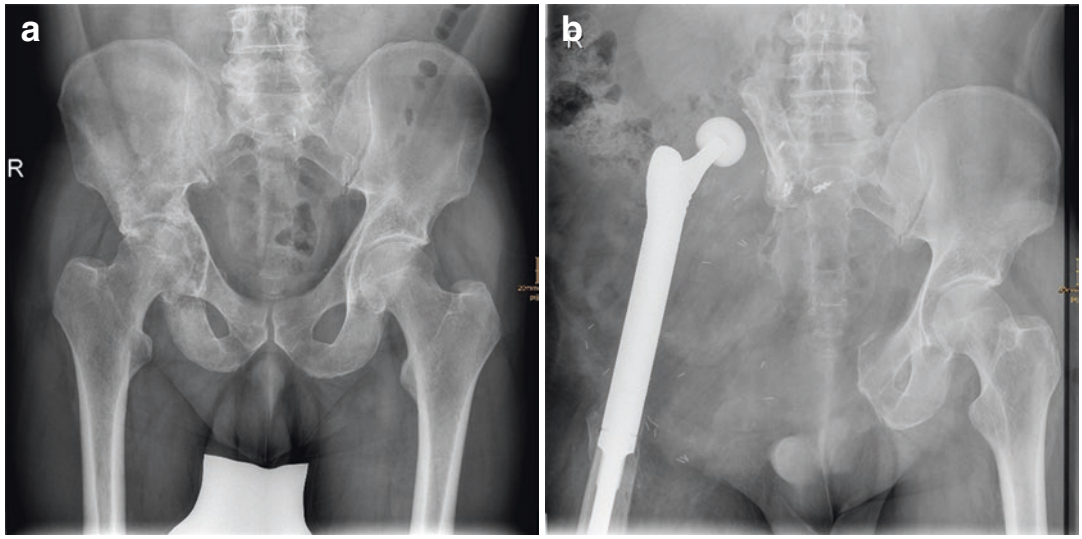

Fig. 6.3 Preoperative (**a**) and 4-years follow-up (**b**) X-rays (ap) of a 68-year-old patient after dedifferentiated G3 CS of the right hemipelvis after I–IV resection and type IIb hip-transposition with proximal femur GMRS® tumorendoprosthesis, LARS® Ligament and Fixation with Mitek®-ancor

struction surgery [23]. On the other hand, several authors consider iliosacral arthrodesis by biological (fibula autograft/allograft) or composite osteosynthesis (using polyaxial screws and rods in the sacrum and ischium/pubis) a good option when the acetabulum can be maintained and in case a stabilization of the pelvic ring is desired [19, 24]. Type II periacetabular resections leave a bone defect that can be bridged by many different reconstruction methods or left without reconstruction or being left in a situation with a iliofemoral or iliosacral pseudoarthrosis or a hip transposition/flail hip [19, 25, 26]. The later, described by Gebert et al., is considered to be an adequate surgical intervention without reconstruction of pelvic continuity. It can be achieved by a turnaround of the inferior part of the acetabulum and fixation with screws to the sacrum or iliac bone (type I), or by an artificial ligament fixation of either the femoral head (type IIa) or a proximal endoprosthetic femoral replacement to the sacrum (type IIb) (see Fig. 6.3) [25]. Hip transposition, according to Puri et al., provides good MSTS93 functional results of 73 points apart from limp shortening and impaired gait. Therefore, this technique emphasizes that the reconstruction of the pelvic continuity is not necessary to preserve hip mobility [27]. Pring et al. and Puri et al. described iliofemoral and ischiofemoral arthrodesis after CS resection, a way to achieve a strong bone union between pelvic bone and femur of limb-salvage without restoration of the pelvic ring. Both of them describe good final MSTS93 results of 73 points with this technique [19, 26, 27].

Ever since limb-salvage is the primary surgical objective, an anatomic reconstruction is the overriding wish. Large pelvic allografts, sole allografts, or composite allografts in conjunction with a metal joint replacement as well as megaprostheses for pelvic reconstruction are used to reach this goal, despite a background of high infectious and mechanical failure rates. Competing risk analysis by Puchner et al. identified endoprosthetic reconstruction after pelvic sarcoma resection as the most significant factor for the first major complication in terms of the ISOLS failure mode classification with a HR 4.9 (2.2–9.8) [28]. Future studies will tell us, if the use of personalized pelvic implants, improvements in implant surface-bone congruency as well as the use of patient-specific instruments and navigation for a better planning and to accomplish shorter operation times will be able

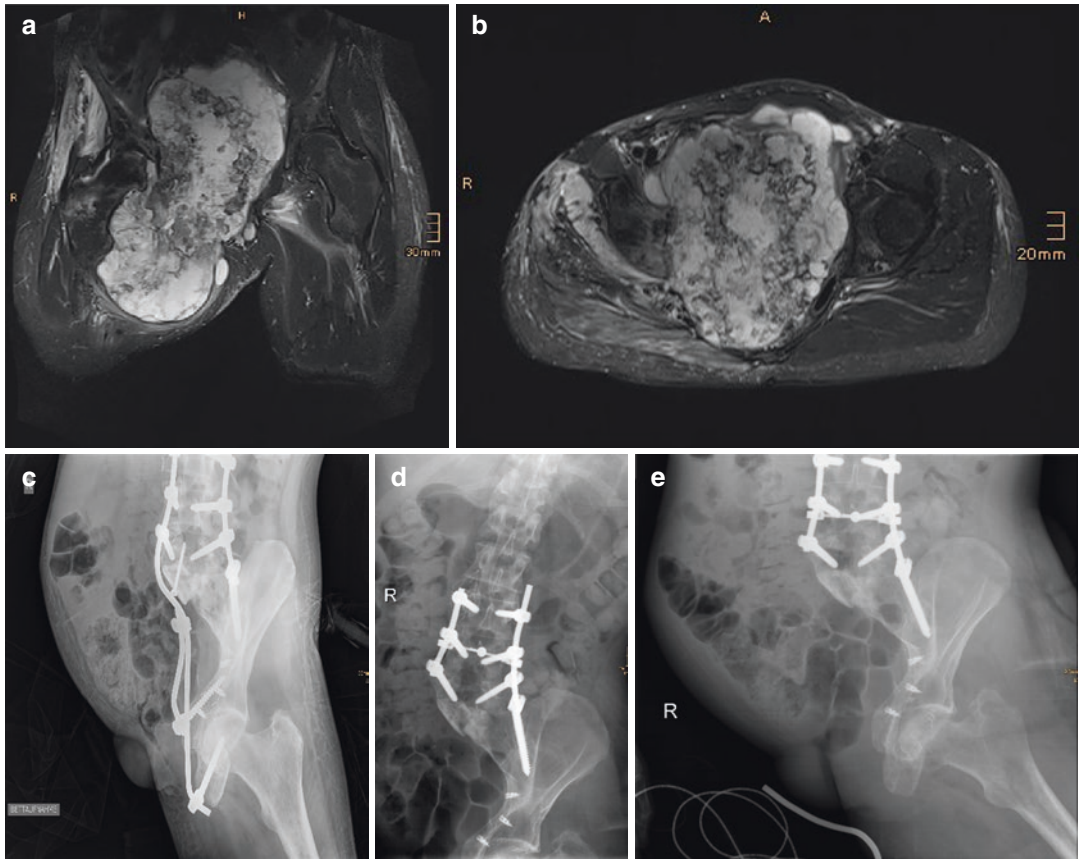

Fig. 6.4 MRI of a permagna CS G2 of the right hemipelvis crossing the middleline (**a**, **b**). X-rays of a postoperative situs after external hemipelvectomy (extended type1–4 resection) and ventral stabilization. Iliospinal fusion with Titanium rods and screws of the contralateral pelvis. X-rays ap and lateral (**c**, **d**) and after partial removal of hardware after fusion due to infection (**e**)

to reduce infection rates and implant survival. Jaiswal et al. sums up data about 98 patients (>50% CS patients) after resection and reconstruction with custom-made pelvic endoprostheses. After endoprosthetic reconstruction of periacetabular tumors, infection rates are high, ranging between 10% and 30% [29]. Despite this fact, the infection rates in CS-only literature seem to be lower compared with mixed-sarcoma cohorts of pelvic reconstruction [6, 20]. High dislocation rates of 40% in the beginning of endoprosthetic reconstructions were reduced down to a rate of 20% due to efforts in introduction of larger femoral head [29]. Although endoprosthetic reconstruction provides good functional results in patients without complications measured by Toronto-extremity salvage score (TESS) of 70.3% (see Fig. 6.1), functional results change for the worse when complications occurred to a TESS of 37.1%. High complication rates concerning infection in 30% and dislocation in 15% were also reported after reconstruction after Type II and II/III resection by stemmed acetabular pedestal cups (see Fig. 6.2) [30, 31]. Competing risk analysis revealed a surgical revision rate of 48% after 10 years [31]. High rates of aseptic loosening could suggest impaired bone quality after CS for still unknown reasons for this mechanical problem [32]. Still, young patients with the explicit demand for anatomic reconstruction might give a good indication for stemmed acetabular cups.

Apart from small case series, saddle prostheses were no longer applied in musculoskeletal oncology for their considerable morbidity and complications such as dislocation as well as heterotopic ossification, vertical migration, and infection leading to MSTS functional outcome of 50.8. Infection occurred in 8 of 18 CS patients (44%) [33] (Figs. 6.3 and 6.4).

An alternative to metal implants in anatomic reconstructions are allografts. Campanacci et al. concludes in a case series of pelvic massive allograft reconstruction that patients with CS might benefit most in comparison to other primary malignancies considering the potential risk of infection and necrosis and a high general morbidity rate of massive allograft, due to less interference with neoadjuvant treatment side effects. In fact, CS patients in this cohort had a better allograft survival and low allograft-necrosis than patients with other malignancies [34]. In line with this statement, Langlais et al. specify that massive allograft reconstructions should be done in patients younger than 60 years, in good general health condition and physically active because it is a demanding procedure [35]. Functional outcomes in periacetabular reconstructions with allografts are good and sometimes excellent with MSTS93 of 61–67% [34, 35].

In type III (ischiopubic) resections, reconstructions are usually not mandatory due to persistent pelvic continuity.

To conclude, the surgeon's choice ought to be made under consideration of individual patients' attributes, functional requirements, and personal wishes [27].

6.5 Radiotherapy and Chemotherapy

CS are considered as chemo- and radiation-resistant tumors, therefore being deployed only very sporadically in cases after recurrent tumor or marginal resection borders [36]. The low vascularity of the tumors as well as the extracellular matrix of hyaline and restricted drug penetrance might be factors that count for chemoresistance [37, 38]. The low mitotic fraction might also be the reason for radio-resistance. Due to limited efficiency of currently available drugs, research focuses on the understanding of molecular pathomechanisms to develop molecularly targeted therapies [39].

6.6 Oncologic Outcome

Patients with CS are treated primarily by aggressive surgical resection. Five-years and 10-years overall survival in pelvic CS range between 55–92% [4, 6, 10, 17, 35, 40] and 45–88% [4, 10, 35, 40] with significant differences between GI-III of 100/50/26% of survival with the lowest survival of 48% in patients suffering from mesenchymal and 10–24% in patients suffering from dedifferentiating CS [2, 41]. Recurrent tumors occur in 19–45% of pelvic CS patients [4, 6, 19, 20, 40, 42, 43], the type of surgery may not be a reason for it [40]. Furthermore, several authors report a higher survival to death in Grade I in comparison to Grade II or III CS [4, 10, 17, 18]. Deloin et al. reported a significant higher proportion of metastasis between low- and high-grade CS patients and a consecutively lower survival in high-grade CS. According to Mavrogenis et al., patients with metastasis are sixfold more likely to be high-grade than low-grade CS. Many different factors like periacetabular location, pathologic fracture and tumor size are associated with low survival [12, 17, 18, 44]. Surgical margins, conferring to Stevenson et al., determine local recurrence in all tumor-grades, but local recurrence itself affects disease-specific survival only in grade II and III CS [12]. Competing risk analysis revealed recurrent tumor as the most important risk factors for survival of CS patients with hazard ratios of 5.5 and 6.07, respectively, which emphasizes the importance of wide resections as a mainstay in CS surgery [4, 12]. Important to know is that even in recurrent cases of CS, aggressive surgical intervention can lead to long-term survival in about 50% of the patients [45].

Conflict of Interest Statement No benefits have been or will be received from a commercial party related directed or indirectly to the subject matter of this article.

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Osteosarcoma of the Pelvis

7

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7.1 Introduction

Primary malignancies of bone are rare and account for less than 1% of all malignancies. Osteosarcoma is the most common primary bone tumor and it can occur anywhere in the axial and appendicular skeleton [1]. Survival today, with the use of multimodal chemotherapy and surgery, is 60–70% at 5 years. Osteosarcoma of the pelvis, however, accounts for less than 10% of all osteosarcomas and survival is far worse, ranging from 20% to 40% at 5 years [2].

This stark contrast in survival can be attributed to many factors. Patients with pelvic osteosarcoma often present with larger tumors and up to 50% of these patients present with metastasis at diagnosis [2]. They often have a delay in diagnosis or misdiagnosis and present with longer duration of symptoms. Symptoms are frequently associated with vague pain often mimicking other benign conditions [3]. Surgical management of these tumors is quite challenging. There is poor tumor compartmentalization within the pelvis and achieving a margin can be difficult, given tumor size and proximity to vital structures.

Lastly, the histologic subtypes found in pelvic osteosarcoma also seem to contribute to overall poor survival. A higher percentage of these osteosarcomas are chondroblastic when compared with appendicular osteosarcomas, a histologic subtype that responds poorly to chemotherapy. Secondary osteosarcomas also make up one-third of pelvic osteosarcomas, another histologic subtype with an inherently poor prognosis [4].

7.2 Histology/Pathophysiology

Osteosarcoma is an osteoid-producing neoplastic tumor of mesenchymal origin. In the pelvis, osteosarcoma most commonly presents as intramedullary or conventional high-grade osteosarcoma between ages 15 and 30. Intramedullary osteosarcomas are further classified into subtypes differentiated by histologic and radiographic appearance including conventional (osteoblastic, chondroblastic, fibroblastic), telangiectatic, small cell, giant cell rich, and osteoblastoma-like. Less common are surface-based pelvic osteosarcoma subtypes including periosteal, parosteal, and high-grade surface osteosarcomas.

Studies show that the most common pelvic subtype is osteogenic osteosarcoma, making up about 70% of cases [5]. Parry et al. reported on the largest series of primary pelvic osteosarcoma and found that the chondroblastic variant was most common, with worse 5-year survival, as previously mentioned [2].

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A significant proportion of pelvic osteosarcomas are secondary and affect patients who are 60–80 years of age. These secondary osteosarcomas arise in the setting of prior pelvic radiation or from Paget's Disease. The reasons for prior pelvic radiation in these patients can include genitourinary or lower intestinal carcinomas, most commonly prostate cancer in men and cervical/anorectal cancer in women. Paget's Osteosarcoma has significantly poorer prognosis with some series showing 0% 5-year survival [2]. This type of secondary osteosarcoma is typically both radiotherapy and chemotherapy resistant and poor survival is seen given patients advanced age and comorbidities [6].

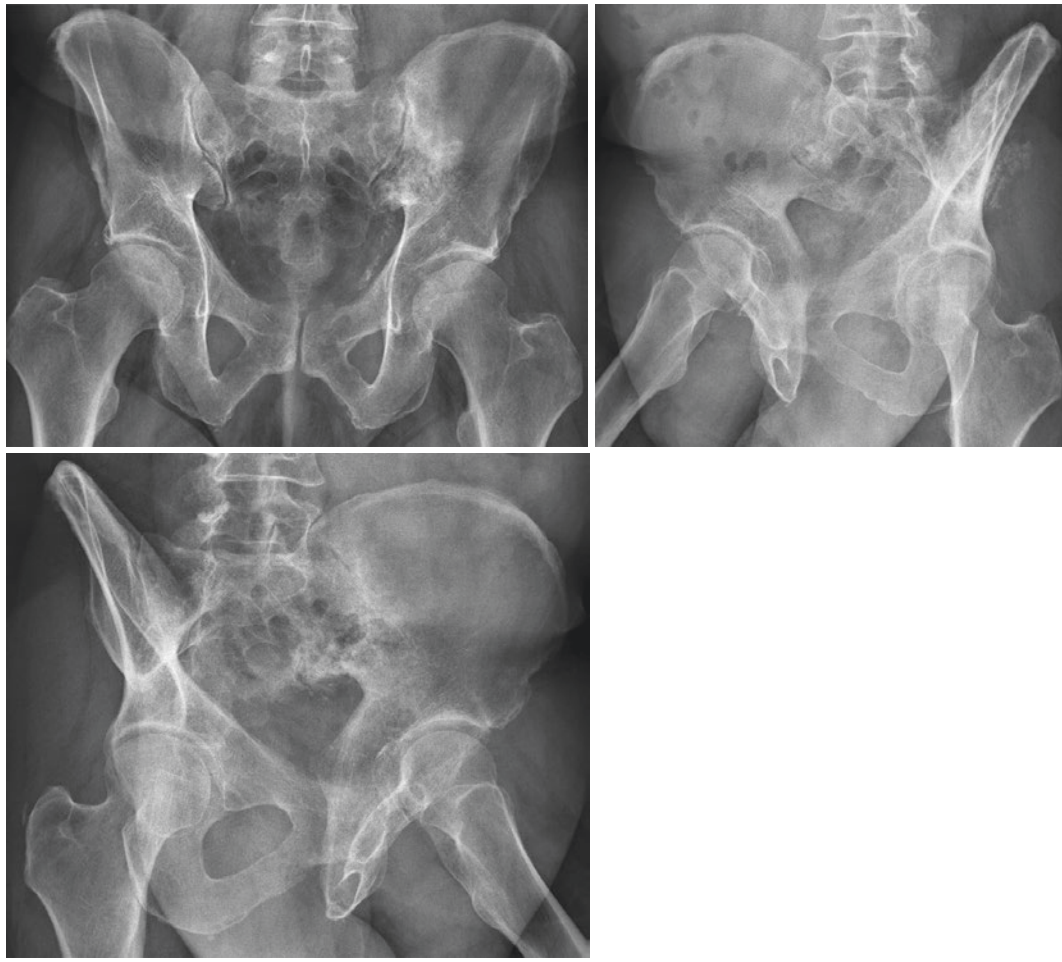
7.3 Imaging Osteosarcoma of the Pelvis

Appropriate imaging is critical for the diagnosis, staging, and subsequent surgical management of osteosarcoma of the pelvis. Multimodal imaging is obtained, and each facet of the workup provides unique and important data.

While advanced imaging has enhanced the surgeon's ability to visualize a tumor and its relationship to vital structures and surrounding soft tissues, plain radiographs remain the first line. Workup typically consists of an AP Pelvis radiograph, Judet views and inlet/outlet X-rays can also prove useful when trying to understanding the anatomy of the tumor (Figs. 7.1–7.3). Radiographs of conventional osteoblastic osteosarcoma reveal a mixed radiolucent and sclerotic lesion with a distorted trabecular pattern. The boundaries of these aggressive malignancies are usually ill-defined. Rapid growth may lead to elevated periosteum, creating a Codman's triangle, and new bone growth under the elevated periosteum can lead to a sunburst pattern. A key finding of osteosarcoma is osteoid production, which is commonly described as amorphous and ill-defined with an inability to determine where the lesion starts and stops. Patients commonly present with large soft tissue masses expanding from the bone, and osteoid production is often seen throughout the soft tissue mass on radiographs. Plain X-rays are also important in the postoperative setting for purposes of surveillance.

Axial imaging for pelvic osteosarcoma is essential and allows the surgeon the ability to define the local extent of the tumor. CT scan of the tumor provides information in regard to the bony anatomy and ossification/mineralization of the soft tissues. It helps delineate the osteoid formation and can show the cortical destruction caused by the malignancy (Fig. 7.4). MRI has been shown to be the most sensitive study to determine tumor boundaries when preparing for surgical resection (Fig. 7.5) [7]. MRI with and without gadolinium contrast has supplanted CT as the imaging modality of choice for many surgeons. MRI allows visualization of the intramedullary extent of the disease in addition to defining the soft tissue expansion of the tumor. Osteoid matrix demonstrates hypointensity on both T1 and T2 weighted images. MRI can also clarify the relationship of the tumor to critical neurovascular and visceral structures in order to determine the feasibility of surgical resection. The entire pelvis is imaged to assess for skip lesions, which is important in both surgical planning and is an important negative prognostic indicator. MRI of the pelvis will show the hip joint and femoral heads to check for intra-articular involvement (Fig. 7.6). The absence of a hip joint effusion has been shown to have a high negative predictive value, but the presence of an effusion carries a low sensitivity [8]. The sacroiliac joint should also be closely examined, trans-articular extension can occur and happens more frequently in osteosarcoma and chondrosarcoma than Ewing's sarcoma [9]. When osteosarcoma does contaminate the SI joint, it has been shown to preferentially cross at the intraosseous ligamentous portion in the posterior joint as opposed to the articular cartilage anteriorly [10], so this should be carefully scrutinized for disease.

In addition to delineating the local extent of the tumor, advances in MRI technology may



Figs. 7.1–7.3 AP Pelvis and Judet radiographs of a 58-year-old male with left sided hip pain. Obtaining radiographs is an important first step in the diagnosis of pelvic osteosarcoma. These radiographs depict a poorly defined, destructive lesion of the posterior ilium with patchy extraosseous mineralization. Obturator oblique imaging reveals the ossified soft tissue component is along the posterior ilium. The iliac oblique radiograph shows the mixed sclerotic and lytic lesion involving of the caudal aspect of the posterior ilium with expansion towards the sciatic notch

allow evaluation of the response to neoadjuvant therapy. Dynamic contrast-enhanced (DCE) MRI depicts the microvascularity to the tumor, while apparent diffusion coefficient (ADC) quantifies the changing tumoral water content in response to treatment. Magnetic resonance spectroscopy (MRS) detects cell turnover activity and may act as a stand-in for biologic aggressiveness and also reveal the cellular response to chemotherapy [11]. The roles of these MRI sequences in the treatment of osteosarcoma are still under investigation. Care must be taken when applying these advanced modalities because the higher water content of chondroblastic osteosarcoma can be misinterpreted for necrosis [12].

Patients must be assessed for metastatic disease once the diagnosis of pelvic osteosarcoma is suspected. The most frequent site of metastasis is

the lungs, and a non-contrast CT scan has been shown to be more sensitive than chest X-rays in detecting pulmonary nodules [13].

Other imaging modalities to evaluate for metastases include Tc-99 whole-body bone scintigraphy and 18-Fluorodeoxy-glucose Positron emission tomography (18FDG-PET) combined with whole body CT. Tc-99 bone scan remains the standard of care for staging and is a low-cost means of examining the entire skeleton (Fig. 7.7). The Children's Oncology Group imaging guidelines for children with osteosarcoma states that bone scintigraphy is required for staging, while performing a single photon emission computer tomography (SPECT) in conjunction with the bone scan is a recommendation. The group recommends the use of FDG-PET for osteosarcoma but clearly states this is not a requirement [14]. FDG is a radiotracer that is metabolized in highly active tissue (Fig. 7.8). A PET-CT can therefore quantify metabolic activity of a tumor and possibly show responses to chemotherapy in addition to revealing distant metastasis. The ability of PET scans to distinguish neoplastic pulmonary nodules from benign masses has also proven useful, both in initial staging and during surveillance [15]. Another utility of PET lies in detecting local recurrence and separating return of malignancy versus benign postoperative changes [16].

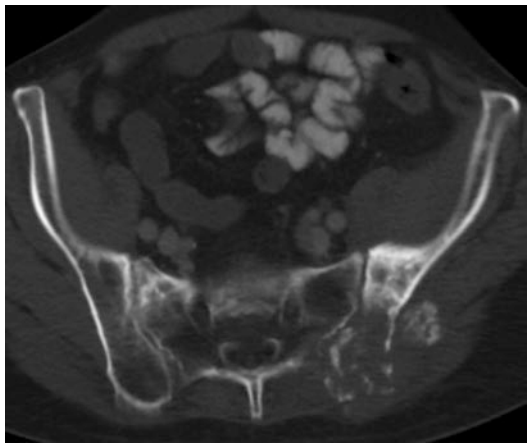
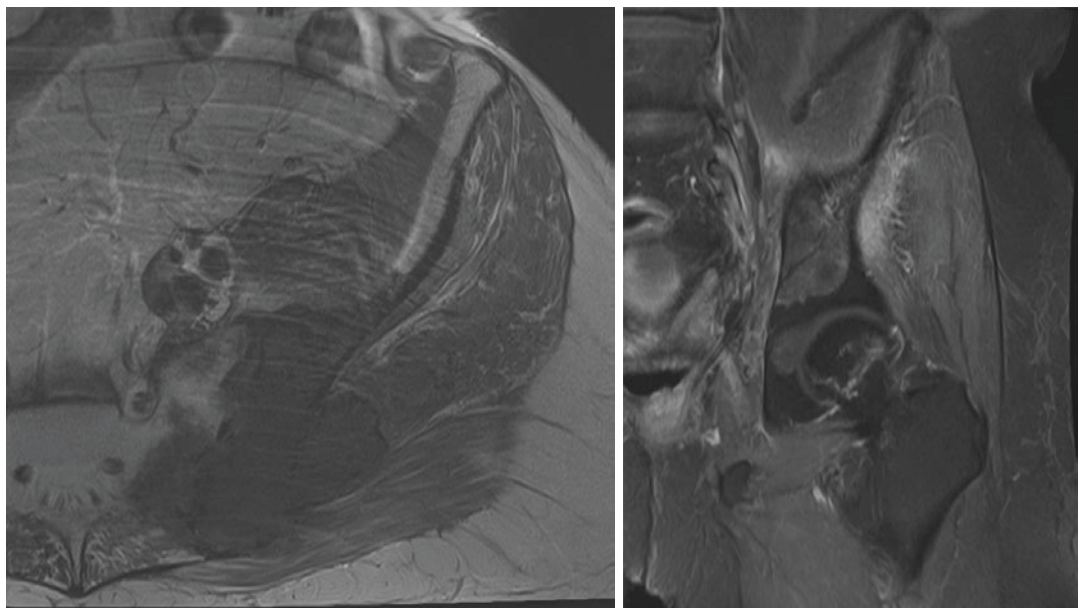


Fig. 7.4 CT scan of the pelvis shows the destructive nature of the osteosarcoma with obliterated cortices along the posterior ilium and loss of the trabecular architecture. Osteoid production in the soft tissue mass is also well visualized on CT imaging



Figs. 7.5 and 7.6 On axial T1 MRI with gadolinium contrast, this osteosarcoma is hypointense and demonstrates contrast enhancement. The tumor involves the gluteus medius posteriorly, but the anterior neurovascular structures are free from involvement. There is intraosseous expansion to the posterior SI joint and posterior S2 and S3 neuroforamen. The coronal MRI shows the tumor is extraarticular and did not invade the hip joint

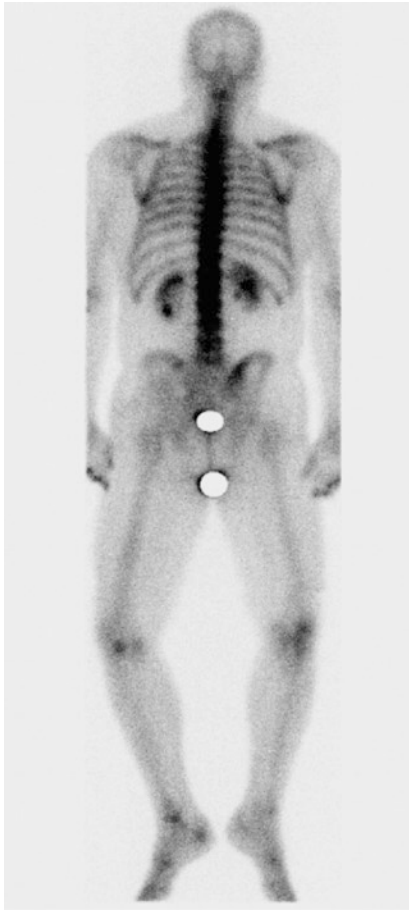


Fig. 7.7 Nuclear medicine bone scan is used to evaluate for local and distant metastases. This patient has an isolated tumor of the left posterior ilium

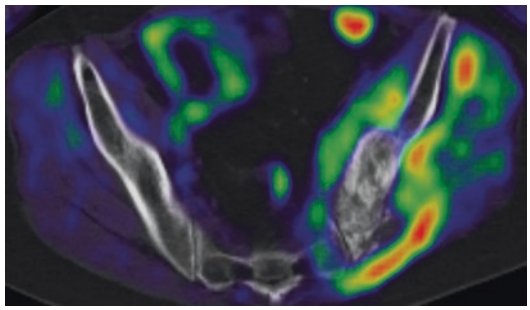


Fig. 7.8 PET-CT shows uptake of 2-deoxy-fluoro-D-glucose in the metabolically active malignancy. The red coloring depicts the areas highest avidity, demonstrated here along the posterior periphery of the tumor

7.4 Medical Treatment for Pelvic Osteosarcoma

Until the application of chemotherapy, the survival rate of osteosarcoma was below 20%. Patients were commonly treated with ablative surgeries such as limb amputation or hemipelvectomy, but despite local control via tumor removal, survival rates failed to improve. In the 1970s, methotrexate was used to treat osteosarcoma following its success with leukemia patients. Various trials of systemic chemotherapeutic agents through this period demonstrated improved survival and reduced metastatic burden. Each individual drug demonstrates a unique response rate in treating osteosarcoma, most commonly around 30–40%. The scientific community quickly recognized the benefit of combining chemotherapeutic agents. Randomized, prospective trials in the 1980s clearly demonstrated superiority of adjuvant multimodal chemotherapy over surgery alone [17, 18].

The development of neoadjuvant chemotherapy prior to surgery developed as a means of preventing metastasis during the months required to fabricate custom implants for joint reconstruction [19]. Concerns regarding the delayed delivery of chemotherapy with this approach were put to rest with a prospective trial showing no difference between neoadjuvant chemotherapy followed by surgery versus immediate surgery and adjuvant chemotherapy [20]. A benefit of neoadjuvant treatment is that biologic response can be assessed by evaluation of the extent of tumor necrosis at the time of surgical resection.

Most treatment protocols call for 10 weeks of neoadjuvant chemotherapy followed by surgical tumor resection, followed by 20 weeks of adjuvant chemotherapy. The most common used agents today are methotrexate, doxorubicin, and cisplatin, owing the acronym MAP to usage of drug trade names (Adriamycin® [Bedford Laboratories; Bedford, OH] Platinol® [Bristol-Myers Squibb; Princeton, NJ]). Several other drugs have also proven effective against osteosarcoma including Ifosfamide, etoposide, cyclophosphamide, vincristine, and bleomycin. Ifosfamide and etoposide have been shown to be

especially useful in treating recurrent and metastatic disease. Unfortunately, the addition of Ifosfamide and etoposide upfront to MAP neoadjuvant protocols has not been shown to reduce the rates of local recurrence or metastasis [21].

With current multimodal treatment, the survival rate most frequently quoted is 70% for osteosarcoma. This often-cited rate applies to localized disease of the extremities in which wide surgical margins are achieved [22]. However, osteosarcoma of the pelvis conveys a much poorer prognosis. Regardless of disease location, the statistics for survival have not changed significantly in the past three decades. Stagnation in curative rates and dismal outcomes for pelvic and metastatic disease have inspired efforts to find new means of treating osteosarcoma.

There are several exciting therapies emerging in the treatment of osteosarcoma, most of which do not involve archetypal cytotoxic drugs. Most strategies of these novel therapeutics rely on advances in knowledge regarding how osteosarcoma avoids detection and destruction by the patient's immune system. One such example is muramyl tripeptide-phosphatidyl ethanolamine (MTP-PE, or mifamurtide), which is an analog of a bacterial cell wall and causes the activation of monocytes and macrophages to become tumoricidal. Mifamurtide is currently approved for clinical use in Europe but not in the United States [23].

The development of monoclonal antibodies to osteosarcoma cell surface markers may also lead to tumor destruction. There have been several antibody designs to attack osteosarcoma cell lines, including bispecific T-cell engagers (BiTE) antibodies, which bring tumor cells in close proximity to T-cells that can then be activated and directly cause cellular destruction. Antibodies have also been designed to selectively deliver cytotoxic chemicals to tumor cells, such as vedotin [24]. Vaccines consisting of tumor cells have also been developed. Dendritic cells detect the tumor antigens in the vaccines and prime T cells to create cytotoxic T lymphocytes. Oncolytic viruses that will only reproduce within tumor cells are also being developed [25]. Another division of emerging therapies is adoptive cell therapy, in which T cells are engineered to detect and destroy tumor cells that have downregulated certain proteins in order to avoid recognition by the immune system. Chimeric antigen receptor T cells (CAR-Ts) and Tumor-infiltrating lymphocytes (TILs) are two such examples of adoptive cell immunity.

7.5 Surgical Treatment

7.5.1 General Principles

En bloc resection with a wide margin is the recommended surgical treatment for osteosarcoma. Patients with pelvic osteosarcomas that present with smaller tumors are more likely to have a limb sparing resection or an internal hemipelvectomy, whereas larger tumors are more likely to require limb sacrificing resection or an external hemipelvectomy [26, 27].

Due to location, proximity to viscera and neurovascular structures and large size at presentation, osteosarcoma of the pelvis is a challenge to treat surgically. This challenge leads to prolonged surgical time, high rate of complications, and high blood loss. The goals of surgical treatment of osteosarcoma of the pelvis is adequate local oncologic control and to optimize function for quality of life.

7.5.2 Effect of Margins

Parry et al. showed that poorer outcomes resulted in patients with pelvic osteosarcoma that had an intralesional margin [2]. They found a local recurrence rate of 17% in patients with wide margins, 32% for marginal margins, and 44% for intralesional margins.

Other authors have found that despite achieving wide margins in pelvic osteosarcoma surgery, the local recurrence rate is still high. Donati et al. showed in a study of 60 patients with primary high-grade pelvic osteosarcoma that although they obtained wide margins in 18 of the cases, 8 patients had local recurrence [28]. Similar outcomes have also been shown in

a series of primary pelvic sarcomas by Shin et al. [29]

7.5.3 Limb Sacrificing Resection/ External Hemipelvectomy

External hemipelvectomy or hindquarter amputation is surgical resection of the osteosarcoma-involved pelvis with sacrifice of the lower limb. External hemipelvectomy should be considered in cases where a functional limb salvage cannot be performed. This procedure is not recommended in cases of widespread metastatic disease that is unresponsive to chemotherapy, except for palliation in patients with intractable pain.

This procedure is classically performed with a posterior fasciocutaneous or myofascial-cutaneous flap with the gluteus maximus for wound coverage. External hemipelvectomy can also be performed with an anterior myofascial-cutaneous flap with the quadriceps musculature or a filet flap of the lower leg musculature on a vascular pedicle maintaining the external iliac vessels in the pelvis, superficial femoral vessels, and popliteal vessels as it courses through the lower extremity to supply the flap.

7.5.4 Limb Sparing Resection/ Internal Hemipelvectomy

Internal hemipelvectomy is the surgical resection of pelvic osteosarcoma and can include the sacrum, ilium, ischium, and/or pubis, while sparing a functional lower extremity. Advances in the diagnostic and therapeutic regimens as well as implementation of a multidisciplinary team approach to treating pelvic osteosarcoma have paved the path to limb salvage.

Appropriate patient selection and planning of surgical resection with wide margins in an internal hemipelvectomy can provide a good functional outcome. Salunke et al. showed in their series of 23 patients with pelvic bone sarcomas treated with internal hemipelvectomy that there was an 83% 5-year survival rate and a good mean functional MSTS score [30]. They did show a correlation of higher tumor necrosis response to chemotherapy linked with a better prognosis.

Internal hemipelvectomy should not be considered in cases where sciatic nerve roots must be sacrificed.

7.5.5 Computer Navigation

Limb sparing pelvic tumor resection can be a difficult operation due to the complexity of pelvic anatomy. Computer navigation has recently become available for improving the precision of pelvic tumor resection (Fig. 7.9). Computer-navigated pelvic resection is a novel technology, and therefore it has not been widely adopted, but reports have been published on its effectiveness. Laitinen et al. concluded that navigation-assisted surgery for posterior ilium and sacral tumor resection improves oncological outcome and safety for the patient since navigation reduced the rate of intralesional margins [31]. Abraham et al. showed that in a series of 23 patients undergoing navigated resection of pelvic and sacral tumors that they were able to perform resection with negative margins in 21 of the patients [32]. Wong et al. showed that in simulated pelvic tumor resections, patient-specific instrumentation is as accurate as navigation, but with shorter bone resection time [33].

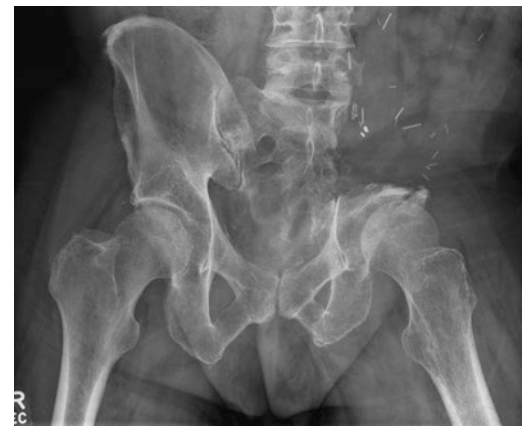


Fig. 7.9 Post-operative radiograph reveals a modified hemipelvectomy with preservation of the hip joint and resection of the sacroiliac joint and sacral ala

7.6 Summary

Osteosarcoma of the pelvis continues to be one of the most challenging diseases treated by orthopedic oncologists. Treatment goals are the same in all osteosarcomas. However, there are significant negative factors associated with this entity including increased local recurrence rates, decreased survival, and overall worse function and morbidity after surgical management.

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Ewing's Sarcoma of the Pelvis

8

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8.1 Introduction

Although rare, Ewing's sarcoma (ES) is the second most frequent primary bone malignancy after osteosarcoma [1, 2]. ES is part of a spectrum of neoplastic diseases known as the Ewing's Family of Tumors (EFT) [3]. In addition to ES, this group includes peripheral neuroectodermal tumor (PNET), extraosseous ES (EES), atypical ES, and malignant small-cell tumors of the thoracopulmonary region (Askin tumor) [4–8]. Common histologic and immunohistochemical features in addition to non-random chromosomal features and similar responses to radiation and chemotherapy suggest that this group shares a common cell of origin [9]. Increasing evidence suggests that this is specifically a mesenchymal progenitor cell [10], though this remains debated. Overall outcomes of patients with EFT have dramatically improved with optimization of multimodal therapies [11]. However, pelvic EFT has been associated with inferior outcomes compared with other skeletal sites as a result of a higher rate of metastatic disease at presentation in addition to nearby radiosensitive organs limiting the ability to achieve local control via surgical resection or radiation without excessive morbidity [12–17].

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8.2 Histology

EFT are high-grade malignancies characterized histologically by sheets of small, round blue cells with scant cytoplasm (Fig. 8.1a–c). These primitive, undifferentiated cells do not resemble mesodermal tissue in contrast to those of the other primary bone malignancies osteosarcoma and chondrosarcoma, which demonstrate histologic features of bone and cartilage, respectively [18–20]. The histologic features of ES may be distinguished from PNET, as the latter is characterized by neural differentiation identified on light microscopy and by a neural phenotype on immunohistochemical staining [21]. Sensitive immunohistochemical markers for EFT include positive vimentin, FLI-1, NKX2.2, and CD99 (Fig. 8.2), also known as MIC2 [22].

8.3 Molecular Pathogenesis

The pathologic molecular alteration of EFT is the reciprocal translocation between chromosomes 11 and 22, t(11;22)(q24;q12), which results in the fusion gene *EWSR1-FLI1* [23–28]. This fusion gene is present in 90–95% of cases, while similar variant translocations and the respective fusion genes they have produced, such as *EWSR-ERG*, make up the remainder [23, 29]. These fusion genes can be detected using fluorescence in situ hybridization (FISH) or reverse transcrip-

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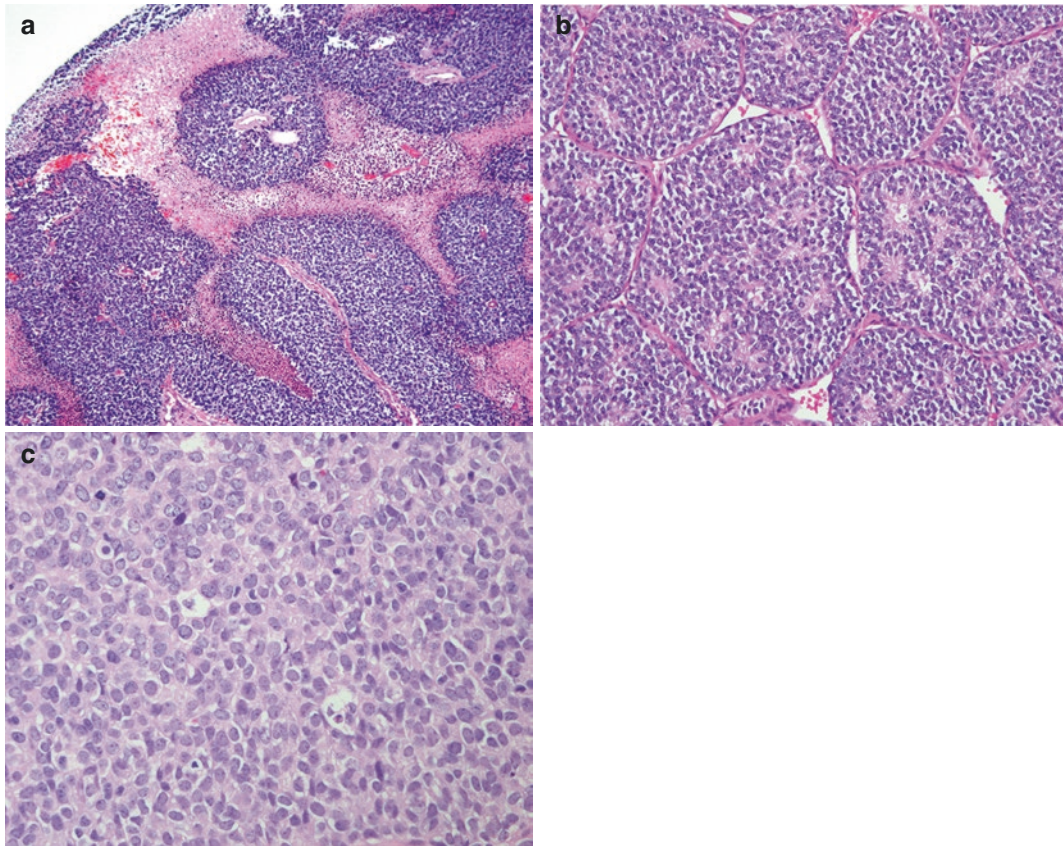

Fig. 8.1 (**a**) H&E staining at 100× shows small, round blue cells with geographic necrosis (pseudo-rosettes). (**b**) H&E staining at 200× demonstrates typical rosette formations, with sheets of cells divided into lobules by thin fibrous strands. (**c**) H&E staining at 400× demonstrates a uniform sheet of small, round blue cells

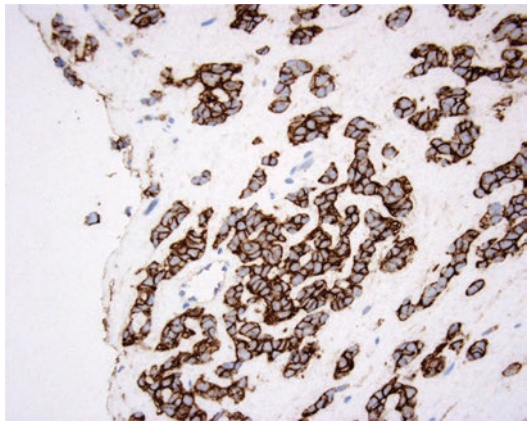

Fig. 8.2 CD99 immunohistochemistry at 400× demonstrates a typical CD99 positive membranous staining pattern

tase polymerase chain reaction (RT-PCR) for molecular diagnosis of EFT with high levels of sensitivity and specificity [30].

There are two likely mechanisms for the contribution of these chimeric proteins to neoplastic transformation [24, 27, 31, 32]: by influencing the transcription of (1) genes normally regulated by native FLI or EWS protein; and (2) genes different from those normally regulated by native FLI or EWS proteins. Acting as transcriptional activators, the chimeric fusion proteins drive oncogenesis by deregulation of cell signaling, apoptosis, telomerase activity, proliferation, and metastasis [33]. The gene affected by the fusion proteins may serve as a target for future molecular-based therapy [34].

8.4 Epidemiology

Primary EFT may present throughout the axial and appendicular skeleton [35]. Based on data from 975 patients from the European Intergroup Cooperative Ewing Sarcoma Studies (EI-CESS) trials [36, 37], the pelvis is the primary site in 25% of ES cases [12, 38, 39]. According to the National Cancer Institute's Surveillance, Epidemiology, and End Results Program (SEER), there is a dramatic variation in race across patients with ES, with white children having an approximate sixfold higher incidence rate than black children [40].

The peak incidence of EFT is in the second decade, while 30% of patients presenting before the age of 10 and an additional 30% of patients after the age of 20 [41]. EFTs are rare after the third decade [42]. There is significant racial and gender variation in rates of EFT [40]. Caucasian and male patients are disproportionately affected and EFT are extremely rare in black or Asian patients [40].

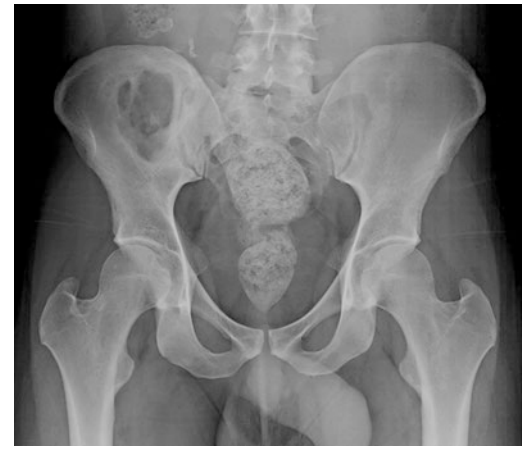

Fig. 8.3 Antero-posterior (AP) X-ray of the pelvis demonstrating a large destructive lesion in the right iliac wing with internal calcifications consistent with Ewing's sarcoma

8.5 Imaging

The radiographic appearance of ES is variable, but is classically described as "moth-eaten," demonstrating an aggressive poorly marginated destructive lesion (Fig. 8.3) [43]. These tumors fill the bone marrow cavity and destroy the cortex. Common features include permeative lesions with associated periosteal reaction and sclerosis. This radiographic sclerosis is correlated with histologic sclerosis. While 80% of cases have an associated soft tissue mass, soft tissue calcification is rare and is found in only 10% of cases [44]. The tumor may expand the cortex and displace the overlying periosteum, forming the characteristic clinical sign of Codman's triangle—though this is more readily appreciated on long bone lesions rather than pelvic tumors. In addition, most tumors demonstrate a laminated "onion skin" periosteal reaction [43]. Cross-sectional imaging is essential for preoperative planning in order to define tumor extent and evaluate for involvement of adjacent structures. Computed tomography (CT) allows for accurate delineation of cortical destruction (Fig. 8.4). Magnetic resonance imaging (MRI) is preferred for definition of tumor size and tumor relation to fascial planes, blood vessels, nerves, and pelvic organs (Fig. 8.5a–c). EFT may present with either a hot or cold bone scan. Positron emission tomography (PET) is useful for staging but has not been shown to be predictive of chemotherapy responsiveness [45–47].

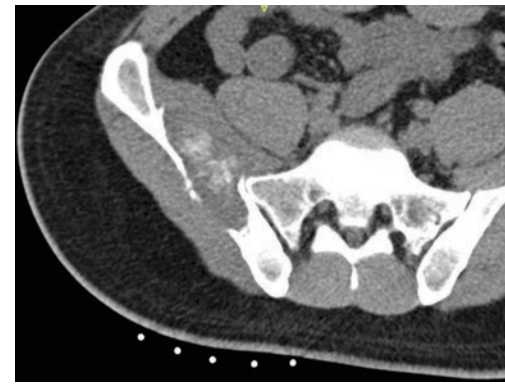

Fig. 8.4 Axial CT scan taken during image-guided biopsy of a patient with Ewing's sarcoma demonstrating a lytic lesion in the right iliac wing with cortical disruption and internal calcifications

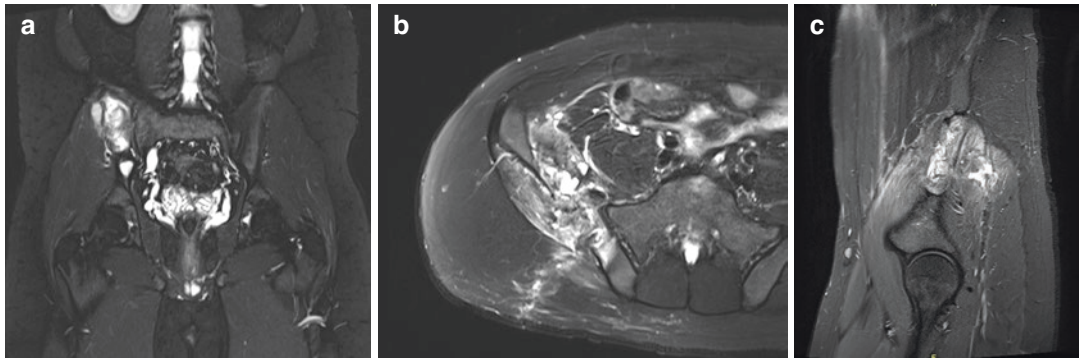

Fig. 8.5 Coronal STIR (**a**), axial T2 (**b**), and sagittal T1 (**c**) MRI of the pelvis in a patient with Ewing's sarcoma demonstrating a heterogeneous mass arising from the right iliac bone with a large, bi-cortical soft tissue component and surrounding inflammatory zone

8.6 Staging

There is no staging system specific to EFT, but the commonly utilized tumor, node, metastases (TNM) system may be applied. Chest radiographs, chest/abdomen/pelvis CT scans, whole-body technetium bone scans, and PET scans are standard imaging modalities to detect metastases [45–50]. In general, EFT patients are staged as having either localized or metastatic disease, which is the strongest predictor of prognosis [51]. Minimal metastatic disease in the peripheral blood or bone marrow detected with PCR but not detected on traditional imaging has been reported in as high as 30% of patients with otherwise localized disease [52]. The prognostic significance of molecularly detectable minimal disseminated disease (MRD) remains controversial but may play a role in explaining why some patients with localized disease have poor outcomes [53, 54].

8.7 Prognostic Factors

The presence of metastatic disease at presentation is the most important prognostic factor for EFT [55, 56]. Five-year survival for patients with localized disease is approximately 70%, compared with 33% in patients with metastatic disease at presentation [57]. The most common sites of metastatic disease are the lung and bone/bone marrow (most commonly spine), while lymph node, liver, and brain metastases are rare. Among patients with clinical metastases, there is a trend for improved survival among those with lung metastases compared with those with bone metastases [58–60]. Patients with pelvic EFT are significantly more likely to present with metastatic disease compared with those with a different primary site (25% vs. 16%) [38]. Other risk factors associated with clinically evident metastatic disease at the time of diagnosis include elevated lactic dehydrogenase (LDH), fever, and an interval between symptom onset and diagnosis of less than 3 months [58, 61–63]. Other strong prognostic factors for poor overall survival include tumor volume over 200 ml, older age, low socioeconomic status, and a histologic response of less than 100% to chemotherapy [64–67]. There has been moderate but conflicting evidence regarding the prognostic significance of achieving negative, disease-free tumor margins during surgical resection on event-free survival [65, 68].

8.8 Biopsy

As with other bone and soft tissue malignancies, pathologic biopsy is central to diagnosis. To avoid compromising potential future limb salvage surgery, surgeons must be consulted to plan the site of biopsy [69]. CT-guided core needle biopsy is

the most common mode of tissue sampling; however, open biopsy may be required if there is a high rate of tissue necrosis to reduce the effect of sampling error and to obtain sufficient tissue for multiple immunohistochemical tests [69].

8.9 Treatment

A multidisciplinary approach in treating patients with EFT is essential [11, 70–72]. EFT is a systemic disease, and patients treated with local therapy alone have high relapse rates [73]. Chemotherapy can eradicate metastatic deposits if initiated when tumor burden is low and is therefore critical in the treatment of EFT [71]. Current standard treatment regimens for localized EFT consist of multiagent neoadjuvant chemotherapy followed by local treatment with surgical resection and/or radiation therapy, followed by additional postoperative adjuvant chemotherapy [11, 74–77]. The treatment strategy for patients with metastatic disease is similar to that of localized disease as multimodal therapy with chemotherapy, radiation therapy, and surgical resection has the potential for cure, and can prolong progression-free survival and relieve pain [75, 78]. In particular, the decision to pursue surgical resection in patients with metastatic disease must be carefully weighed against the impact of a long period of time off chemotherapy [55].

8.10 Chemotherapy

The chemotherapy treatment strategy for EFT has been optimized from the efforts of multiple collaborative national and international trials [79–81]. Multi-neoadjuvant chemotherapy increases the likelihood of local control by reducing tumor burden prior to surgery [82–84]. In many cases, the soft tissue component of the tumor may disappear completely [44, 85]. The traditional agents used in therapy include vincristine, doxorubicin, cyclophosphamide, dactinomycin, and ifosfamide, with some regimens also including etoposide [86–91].

The timing of chemotherapy in relation to local control has been identified as an important factor in the management of EFT [92–95]. Results from the National Cancer Database demonstrated improved survival when patients initiated local therapy by week 15 of induction chemotherapy. As a result, most current protocols advocate for 12 weeks of induction chemotherapy followed by immediate local control with surgery or radiation therapy [92].

8.11 Surgery

Surgical resection is regarded as superior to radiation therapy for local control and performed whenever a marginal or wide resection is feasible [96–98]. While there has been no randomized prospective study to determine which is superior, retrospective series including a 956 patient cohort from the Children's Oncology Group (COG) have suggested higher failure rates in patients treated with radiation therapy alone as compared with surgery alone [97, 99]. Anatomic location within the pelvis plays a key role in determining the approach for local control [12–16]. Lesions of the iliac wing, ischium, or pubis are more amenable to resection than lesions involving the acetabulum, sacroiliac joint, or sacrum [100, 101]. Surgical planning should be based on the MRI scan after induction chemotherapy, which can be dramatically different from the pre-chemotherapy MRI scan [102, 103]. Internal hemipelvectomy with en bloc excision of the affected hemipelvis is the most common approach for surgical treatment of patients with pelvic EFT [13, 104, 105]. Preserving the extremity improves functional and psychological outcomes while producing equivalent oncologic results to external hemipelvectomy (hindquarter amputation) [15, 102]. Depending on the clinical situation, surgeon, and patient preference, resection may or may not be accompanied by reconstruction with allograft or endoprosthesis. For tumors deemed unresectable following induction therapy, patients should be referred for definitive radiation therapy rather than attempting debulking surgery [14].

8.12 Radiation Therapy

The radiosensitivity of ES was noted in James Ewing's original description of the tumor in 1921. EFTs are radiosensitive but radiotherapy alone is insufficient to prevent local recurrence [106–108]. Radiation therapy is indicated for primary local control if the tumor is deemed unresectable, or can be used postoperatively in the setting of marginal resection [14, 100, 109–113]. Recommended doses range from 55 Gray (Gy) for most primary sites of disease to 45 Gy for microscopic disease [76, 107, 114]. Complications of radiation therapy include radiation-induced sarcomas, skin fibrosis, edema, wound healing problems, pathologic fracture, exacerbation of chemotherapy-induced myelosuppression, and growth deformity or arrest [110, 115–119]. The use of proton beam therapy rather than conventional photon therapy has recently been advocated as a way to reduce these side effects, especially in the pelvis where toxicity to the spinal canal, intestine, rectum, bladder, and femoral head must be mitigated [74, 100, 120–122]. There is concern that proton beam therapy may be associated with neutron scatter radiation and a risk for secondary malignancies, warranting future investigation [119]. Intensity-modulated radiation therapy (IMRT) [123–125] and three-dimensional conformal radiation therapy (3CD-CRT) [124] are newer strategies that are utilized to improve accuracy, spare normal tissue, and reduce complications [126]. Data guiding the role of adjuvant radiation therapy in the setting of surgery for patients with inadequate margins is conflicting [127]. The Euro-E.W.I.N.G. group reported on 599 patients and found a reduction in local recurrence in patients treated with adjuvant radiation therapy [128], while the Cooperative Ewing Sarcoma Study (CESS) trials [37, 114, 115, 129] demonstrated no difference.

8.13 Surveillance and Recurrence

Local recurrence is a poor prognostic sign and can occur with or without metastases [12, 130, 131]. Local recurrences can be treated with an approach similar to that of a primary lesion but must be tailored to the patient on an individualized basis, as the lower chance of disease eradication may guide treatment toward palliation [130, 132, 133]. Salvage chemotherapeutic treatment protocols may include irinotecan, gemcitabine/docetaxel, temozolomide, or aerosol rubitecan [84, 134]. Anti-insulin-like growth factor-1 receptor (IGF-1R) antibodies have also been used to treat patients with relapsed EFT and have demonstrated short-term, but not durable, benefit [135–137]. Late recurrence up to 10 years after treatment has been observed for EFT, making surveillance crucial [138, 139]. Surveillance guidelines are offered by the National Comprehensive Cancer Network (NCCN) and include 3-month interval visits for the first 2 years followed by 4–6-month interval visits for the next 3 years.

8.14 Case Presentations

8.14.1 Case 1

A 19-year-old male initially sought care for left hip and leg pain for 2 months. Pain was atraumatic in onset and had been gradually worsening since it began. Pain was worse with weight-bearing, had failed to respond to anti-inflammatory medications, and was accompanied by subjective weakness and loss of function. The patient also reported waking up multiple times a night with pain. His past medical and surgical history were unremarkable. His family history was positive for breast cancer in his maternal grandmother. He presented to the emergency department, where exam was notable only for pain with left hip range of motion and mild swelling about the left gluteal region.

Initial X-ray imaging demonstrated a subtle lytic lesion within the left iliac wing (Fig. 8.6) that prompted further imaging and laboratory workup. Labs were notable for a moderately elevated c-reactive protein and pro-calcitonin but were otherwise normal. CT (Fig. 8.7a, b) and MRI (Fig. 8.8a, b) were notable for a heterogeneous mass arising from the left iliac bone with a

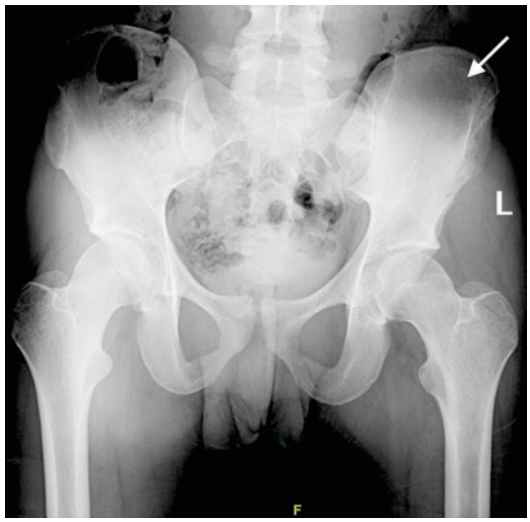

Fig. 8.6 Initial antero-posterior (AP) X-ray of the pelvis in a patient presenting with left hip and leg pain demonstrates a subtle lytic lesion within the left iliac wing

large associated soft tissue component. He underwent CT-guided biopsy of the lesion, with histology demonstrating sheets of small, round blue cells consistent with Ewing's sarcoma (Fig. 8.9a). These cells stained positive for both CD99 (membranous staining) and NKX2.2 (nuclear staining) (Fig. 8.9b). Fluorescence in situ hybridization (FISH) analysis was positive for the characteristic EWSR1 rearrangement.

The patient underwent staging with a positron emission tomography (PET) scan, on which the large iliac wing mass demonstrated intense metabolic activity (Fig. 8.10). In addition, the PET scan revealed diffuse osseous metastatic disease to the right humerus, spine, pelvis, and left proximal femur, as well as possible metastatic pelvic lymphadenopathy. Given the findings of widely metastatic Ewing's sarcoma, he was started on chemotherapy consisting of vincristine, doxoru-

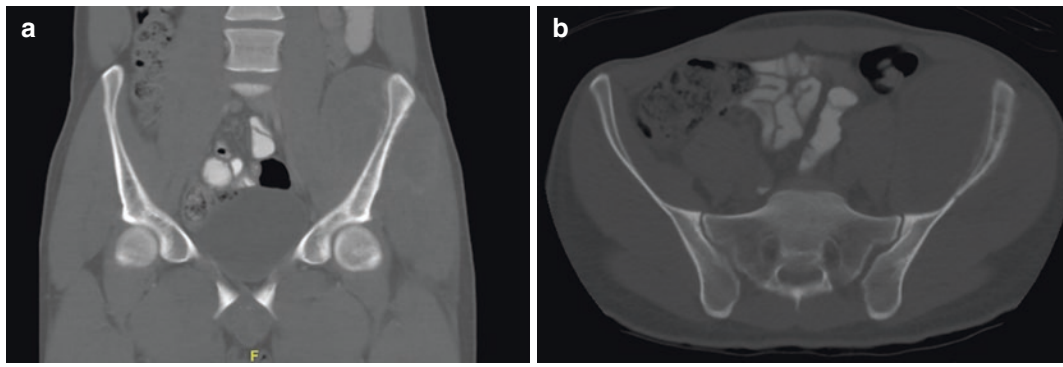

Fig. 8.7 Coronal (**a**) and axial (**b**) CT scan demonstrates irregularity of the left iliac wing with associated periosteal reaction ("onion skin" appearance) and a large soft tissue component

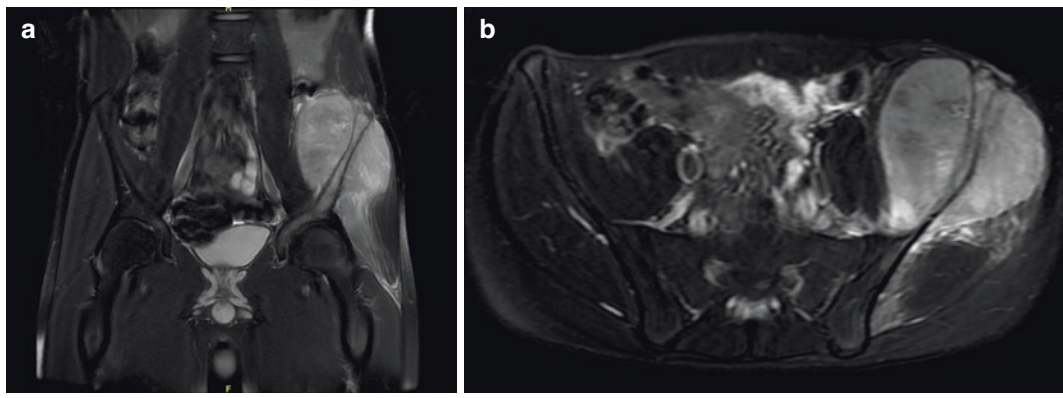

Fig. 8.8 Coronal (**a**) and axial (**b**) T2-weighted MRI shows a large mass arising from the left iliac bone, with a bicortical soft tissue component extending into the iliacus and gluteal muscles

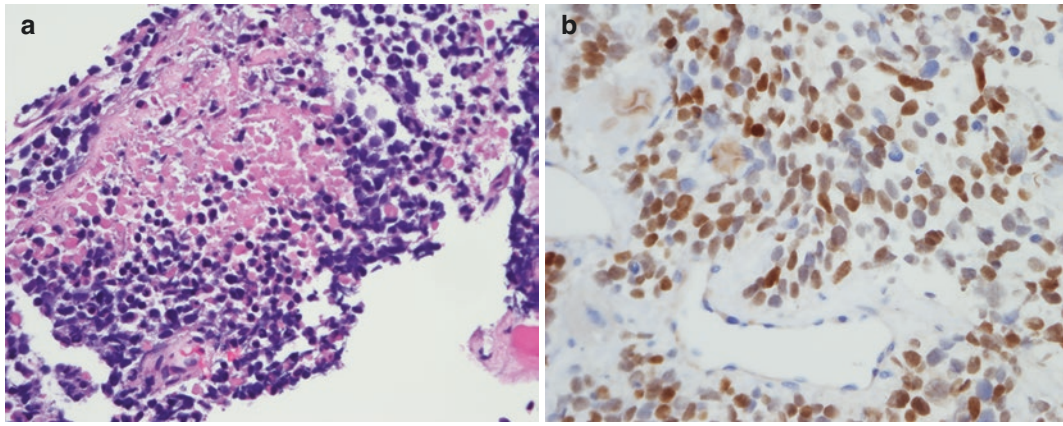

Fig. 8.9 Histology of a patient with Ewing's sarcoma that demonstrates (**a**) sheets of small, round blue cells on H&E stain and (**b**) positive nuclear staining for NKX2.2

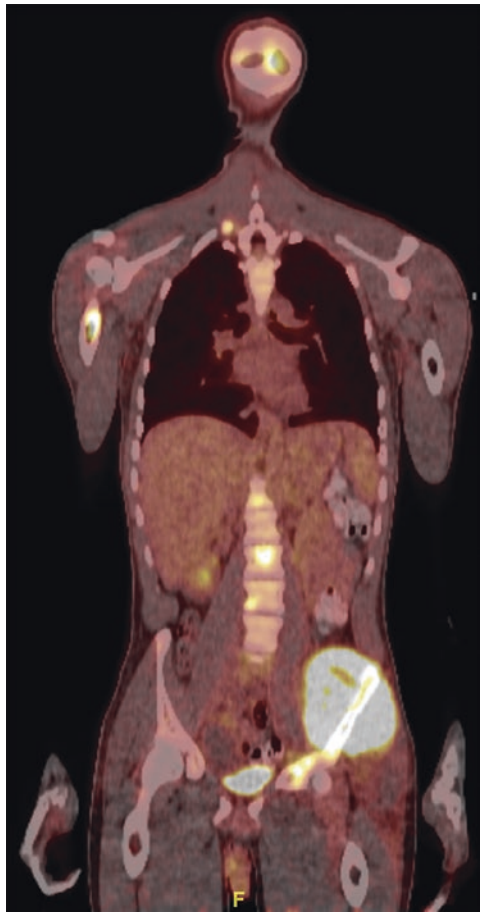

Fig. 8.10 Positron emission tomography (PET) scan of a patient with metastatic Ewing's sarcoma demonstrates a large left pelvic lesion with high metabolic activity, as well as metastases to the right proximal humerus and spine

bicin, and cyclophosphamide. Surgical planning was postponed until response to chemotherapy was able to be assessed.

8.14.2 Case 2

A 2-year-old previously healthy female presented with 5 days of limping. Her parents denied any history of witnessed trauma. They noted she has been progressively inverting her right foot and elevating her right hip while walking. The limp had not improved with ibuprofen. She had no complaints of pain, and no history of recent illness, fevers, or chills. Upon presentation, she exhibited a right-sided antalgic gait, but had full painless right hip range of motion, no focal tenderness, and an otherwise normal exam. Laboratory analysis including complete blood count, electrolytes, and inflammatory markers (erythrocyte sedimentation rate and c-reactive protein) was normal.

Initial X-ray imaging demonstrated a poorly defined lesion within the right ilium, superior to the acetabular dome, with a wide zone of transition and no periosteal reaction (Fig. 8.11). Given these findings, she underwent CT scan (Fig. 8.12a, b) and MRI (Fig. 8.13a, b), which revealed a large, permeative lesion in the right iliac wing with a surrounding superficial and deep soft tissue component. CT-guided biopsy

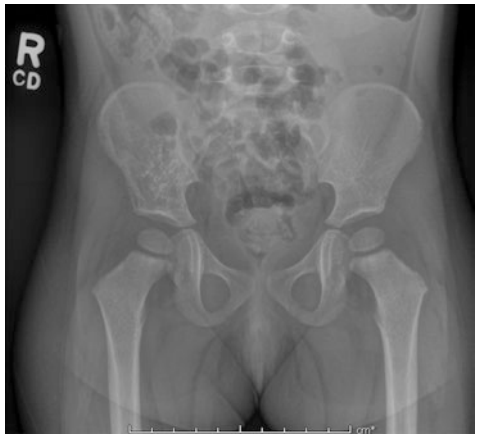

Fig. 8.11 Antero-posterior X-ray of the pelvis in a 2-year-old female presenting with a limp demonstrates a poorly defined, permeative lesion within the right ilium, superior to the acetabular dome, with a wide zone of transition and internal calcifications

of the mass was performed. Histology demonstrated clusters and sheets of small, round blue cells with high nuclear-to-cytoplasmic ratios, round nuclei, and irregular borders. These cells stained positive for CD99 in a diffuse, membranous pattern. FISH analysis was positive for the ESWR1 rearrangement, solidifying a suspected diagnosis of Ewing's sarcoma of the pelvis.

She underwent staging, including a CT scan of her chest, abdomen, and pelvis, which was negative for metastasis, and PET scan, which revealed diffusely increased uptake in the right iliac wing, but no evidence of hypermetabolic lymphadenopathy or metastatic disease.

The treatment strategy for this patient consisted of neoadjuvant chemotherapy, surgical resection, and consolidation chemotherapy. She

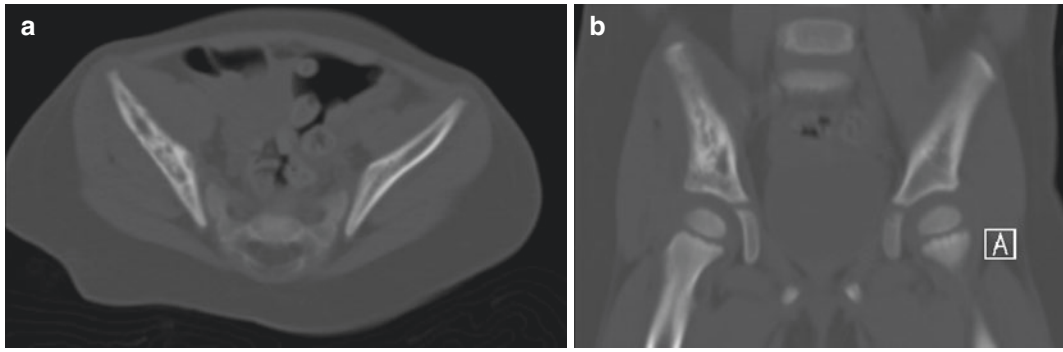

Fig. 8.12 Axial (**a**) and coronal (**b**) CT scan shows a permeative lytic lesion of the right iliac wing with internal dystrophic calcifications and faint periosteal reaction

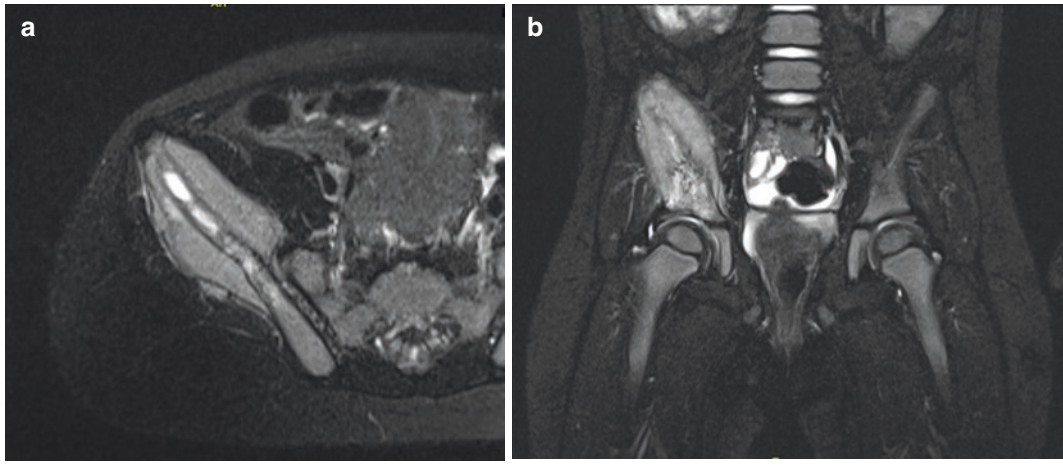

Fig. 8.13 Axial T2-weighted (**a**) and coronal STIR (**b**) MRI reveals a mass of the right iliac wing extending to the anterior sacroiliac joint and through the acetabular dome, with a large superficial and deep soft tissue component

was treated with 6 cycles of vincristine, doxorubicin, and cyclophosphamide with excellent response and reduction of her primary tumor size. She then underwent right internal hemipelvectomy of zones 1 and 2, followed by an additional 11 rounds of chemotherapy. Surgical pathology demonstrated Ewing's sarcoma with >99% tumor necrosis and negative margins.

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9 Pelvic Metastases: Diagnosis and Treatment

Eduardo J. Ortiz-Cruz, Manuel Peleteiro-Pensado, Irene Barrientos-Ruiz, and Raquel Ortiz-Hernando

9.1 Introduction

Metastatic bone disease (MBD) to the pelvis is a challenging problem that affects the patient's life and Quality of Life (QoL) and the orthopedic surgeons increasingly face it. Due to the relatively large dimension of the pelvic cavity, tumors at that location could reach considerable size before symptoms appear, like a pain, pathologic fractures, and limit the ability to ambulate independently [1].

The optimal treatment of bone metastasis may be complex and demand multimodality treatment strategies to achieve optimal outcomes. Some locations of metastases within the pelvis have no significant impact on pelvic stability and function (e.g., ilium and pubis), but tumors located on the posterior ilium may carry a risk to lumbosacral integrity; tumors of the acetabulum may impair the hip function and the weight-bearing; due to the high mechanical loads and this is the location that more of the pelvic surgery indications are made [1].

Metastatic involvement of the pelvis is common, second only to axial involvement. Certainly, 833 (18.8%) of all 4431 metastatic lesions documented in the archive of the Rizzoli Institute were found to occur in the pelvic region: 12.6% are located in the ilium, 1.8% in the ischium, and 1.2% in the pubis [2].

9.2 Diagnosis

Detection of bone metastases is essential for accurate staging and optimal treatment. The objectives of imaging are to recognize sites of metastasis and to consider involved locations in which complications are likely, such as pathologic fracture. Imaging is also used to monitor the biopsy, if believed indispensable.

There are four situations at diagnosis:

1. **Patient with no previous history of cancer.**

   The assessment of a patient with no previous history of cancer has to be similar that the evaluation for solitary symptomatic bone lesion. National Comprehensive Cancer Network (NCCN) guidelines [3] suggest complete a workup for potential bone metastasis for symptomatic bone lesion associated with abnormal radiograph in patients over 40 years of age.

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2. **Patients with a past history of remission cancer, it does not matter how long ago the cancer happened.**

   Most of these patients require biopsy confirmation. If this event is the first evidence of recurrence or the disease progression, the staging assessment must include a contrast-enhanced CT of the chest, abdomen, and pelvis for visceral organ evaluation, a bone scan or FDG-PET/CT may be also indicated.
3. **Patients with known history of cancer at the present time, with known metastases**.

   These types of patients who were found to have other visceral metastasis on restaging evaluation, presumptive diagnosis with imaging modalities may be sufficient for a reasonable diagnosis of bone metastasis and the biopsy might be not indicated.
4. **Patients with a history of cancer at the present time and without known metastases.**

   Generally, the diagnostic staging evaluation should precede a biopsy of suspicious bone lesions, particularly if a pathologic or impending pathologic fracture is present or suspected.

9.3 Treatment Planning

There are mostly three types of treatment: nonoperative, minimally invasive procedures, and surgery. The selected procedure should offer an adequate treatment to the patient to achieve the best possible QoL, while avoiding under or overtreatment. The factors associated with poor QoL include loss of limb function, bedridden, and pathologic fracture.

The treatment depends on the patient's symptoms, prognosis, patient class [4], histologic type, site of the metastasis, amount of bone loss, and functional performance such as the Eastern Cooperative Oncology Group (ECOG) Performance Status scales [5] and family aims.

The main indications for surgery include patients with compromised skeletal stability, pain that not responding to other modalities of treatment or some solitary bone metastases [1, 4, 6], and prognoses. Surgery on the pelvic bone is a challenging procedure both for the patient and surgeon [7], and that is why a clear understanding of all potential varieties of treatment, beginning from nonsurgical therapies to surgeries options [1, 4, 8].

9.3.1 Nonoperative Treatment

The radiation therapy is effective in providing palliation and relief of painful bone metastasis, with a global pain response rate as 60%. Therefore, the external irradiation is the standard nonoperative care for patients with localized bone pain and palliation [9].

Lesions of the hemipelvis not directly involving the hip joint as avulsion fractures of the anterior superior/inferior iliac spines, iliac crest, and superior/inferior pubic rami can generally be treated with external beam radiation and modification of weight-bearing.

9.3.2 Minimally Invasive Palliative Procedures

Patients who have intractable pain at a site of previously irradiated and may not be eligible for supplementary radiation and neither for surgery, the interventional radiology adds to the armamentarium an effective palliative treatment of their symptoms [10]. The most frequent techniques are radiofrequency-ablation, microwave tumors ablation, cryoablation, and cementoplasty, which could be used in combination with the previous techniques [11–14].

9.3.3 Surgical Treatment

The decision to proceed with surgery could be difficult for the surgeon and patient too, because the risks of surgery may outweigh the proposed benefits of improved pain and function. It is indispensable to identify the next six key subjects.

9.3.3.1 Types of Pelvis Resection

According to Enneking classification that is based on the resected region of the pelvis. Type I (ilium); type II (periacetabular region); zone III, pubis and ischium. When resection of the posterior ilium with the sacral wing is classified as either an extended type I or type IV resection [15]. When the MBD is located at periacetabular region (type II), most of the nonoperative treatments such as radiotherapy, chemotherapy, hormonal therapy, and bisphosphonates are insufficient to reduce pain and restore ambulation and this is the zone that is more frequent to indicate surgery (Fig. 9.1a, b).

9.3.3.2 Patients' Classes

It is critical to individualize the type of patient who requires a surgical treatment. Capanna and Campanacci [4] introduced a protocol in long bone metastases that provide an aim to look for a proper treatment and it is adapted for pelvic metastases too. The patients are divided into four classes: (1) solitary lesion with good prognosis; (2) pathologic fracture; (3) impending fracture; and (4) other lesions.

All patients included in Classes 1, 2, and 3 should have been referral to oncology orthopedic surgeon for surgical treatment and have to be evaluated by the medical oncologist and medical radiotherapist for the assessment of neo or adjuvant treatments. Most of the class 4 patients are treated conservatively by chemotherapy, hormonotherapy, and/or radiation therapy.

9.3.3.3 Zones At Risk for Mechanical Failure

According to Muller and Capanna [16], type 2 parallels to the nonarticular part of long bones (humerus, femur, and tibia) and those are the lesions with a high risk for mechanical failure due to progressive destruction of the hip joint and they have a surgery indication (Fig. 9.2a–d). ZONES 1 and 3 are comparable to non-weight-bearing and expendable bones of the extremity (clavicle, sternum, and fibula) and they do not compromise the mechanical stability of the pelvic ring (Fig. 9.3a–d) and they do not need bone reconstruction after the resection.

9.3.3.4 Metastatic Acetabular Classification

It is important to keep in mind the classification described by Harrington, in order to select the best type of surgical management of acetabulum [17].

Periacetabular reconstruction for class II and class III lesions require restoration of structural integrity of the medial wall, superior dome, and lateral wall to adjust the acetabular components. Harrington [17] described good results with no loosening of implants by using a protrusio ring with or without mesh in class II and adding

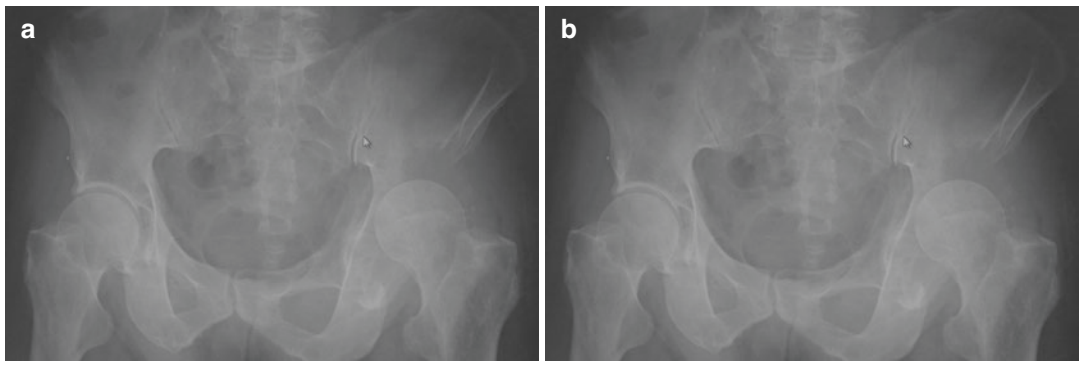

Fig. 9.1 (**a**) Anteroposterior (AP) radiograph of the pelvis that shows an osteolytic lesion at left zone 2, in a 70-year-old male patient with a lung cancer (class 3 patient according to Muller and Cappana [16]). (**b**) Same patient and destructive progression of the lytic lesion at zone 2, after nonoperative treatments, at this point is changed a class 2 patient

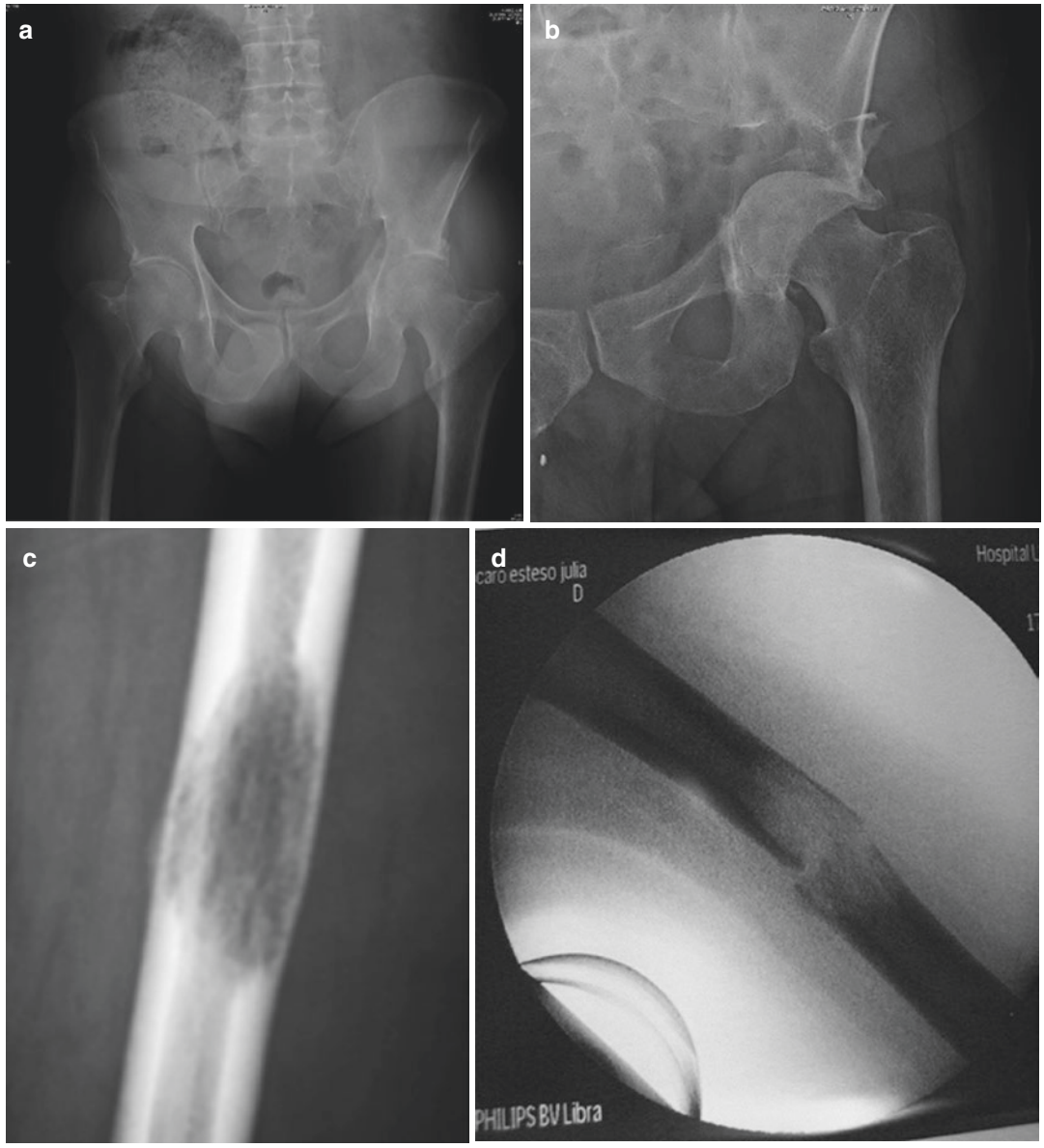

Fig. 9.2 (**a**) AP radiograph of the pelvis that shows an osteolytic lesion zone 2, in 75-year-old female patient, with metastatic thyroid cancer (class 3 patient). (**b**) Same patient, with tumor progression and demonstrates a predictable femoral head protrusio (class 2) that probably had to be restored it before happened. (**c**) Conventional radiograph of metastatic osteolytic lesion in femoral diaphysis, with a high risk of pathologic fracture, that parallels to the lesion at zone 2 of pelvis, as displayed in **a**. (**d**) AP view of the same lesion of the **c**, currently with pathologic fracture, that parallels to the lesion at zone 2 of pelvis with femoral head protrusion as **b**

threaded-Steinmann pins in class III lesions. In spite of the limited life expectancy of patients, reconstruction with anti-protrusio cages, screws, and cemented hip replacement is beneficial for improving their QoL.

Modifications of the technique have been proposed. Using retrograde screws placed through the protrusio cage to transmit weight loads to structurally intact bone, which follows the principles outlined by Harrington [18, 19].

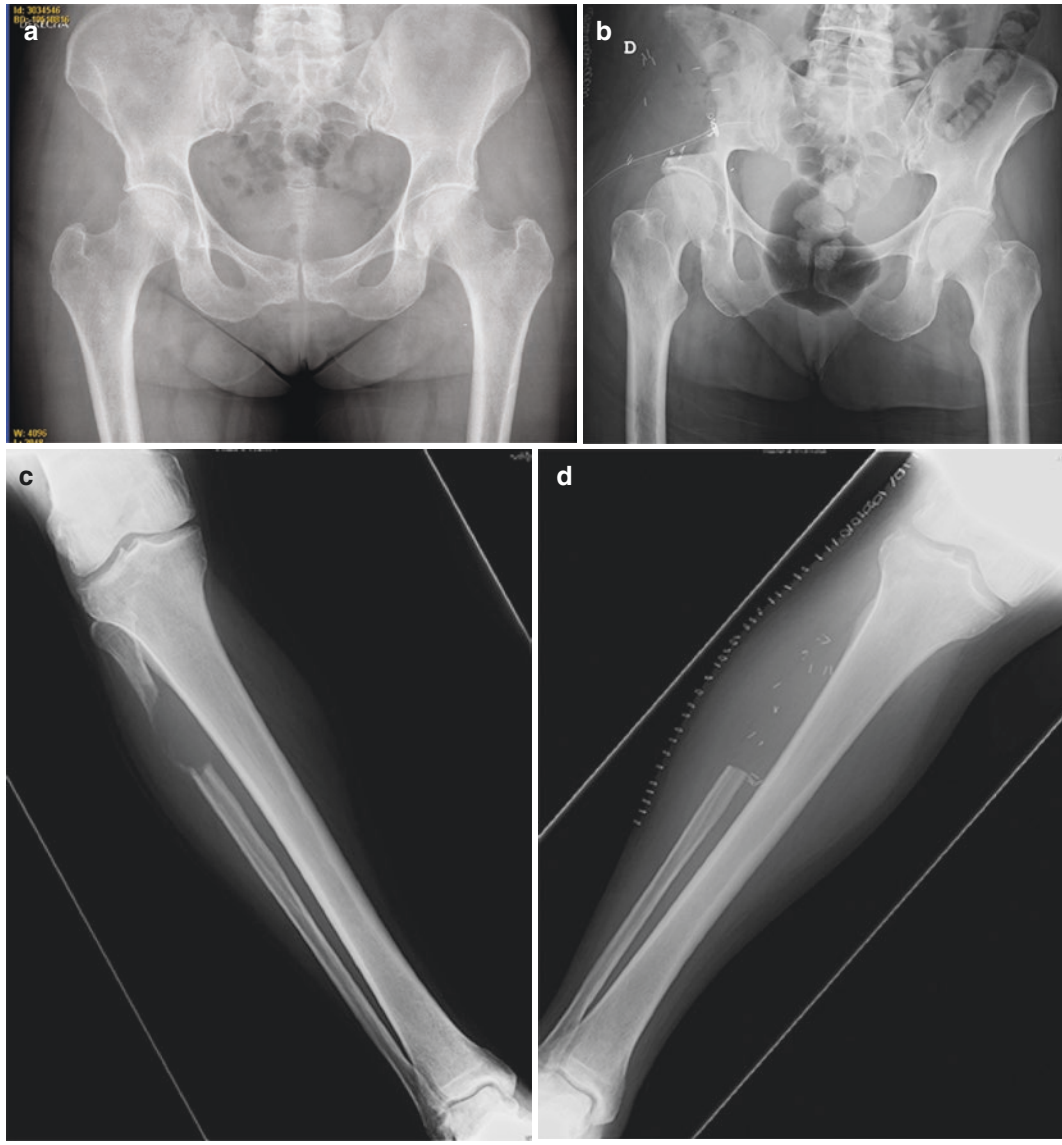

Fig. 9.3 (**a**) Osteolytic infiltrative lesion at zone 1 (supra-acetabular) in a 63-year-old female patient with solitary bone metastasis of follicular thyroid carcinoma (class 3). (**b**) Identical patient, after wide tumor resection and not pelvic reconstruction, due that there is no compromise of the mechanical stability of the pelvic ring. (**c**) AP view of the tibia and fibula that shows an osteolytic lesion at proximal end of the fibula (expandable bone) in a 65-year-old male with lung cancer, which parallels with a zone 1 or 3 of the pelvis, which are possible to resect them without reconstruction. (**d**) Radiograph after wide tumor resection without reconstruction of the proximal fibula; that parallels with wide tumoral resection of the zones 1 and 3 of the pelvis, as shown in **b**

9.3.3.5 Patient Prognosis

The estimation of survival will assist to select the appropriate treatment indication. A systematic review of the literature made by Wood and coworkers [20] in patients with MBD to long bones and/or pelvis, treated surgically and revising the clinical studies which reported pain relief, function outcomes, perioperative complications, and mortality, they found an advantage for surgical management; however, there is significant risk of morbidity and mortality that had to be considered.

There are tools which estimate the likelihood of survival after surgery for patients with skeletal metastases, and some of them are located in the web and easy to check (www.pathfx.org) [21], and they help for surgical decision-making.

9.3.3.6 Type of Surgery

Pelvic metastases are treated either with tumor intralesional resection and not reconstruction, intralesional resection, and reconstruction or wide resection and reconstruction or not.

Respect which type of resection has to be indicated, there are few data available to compare the outcome of wide resection and intralesional resection for pelvic metastases. The study of Pietro Ruggieri and coworkers [22] evaluated the role of intralesional or marginal resection and compared with wide resection; they did not find difference in survival to death between wide resection and intralesional resection even in patients with solitary metastases. Nevertheless, the wide margin and reconstruction could be appropriate for patient with solitary metastases, in order to attempt to increase the survival [16]. The Interdisciplinary Consensus on the Management of Bone Metastases from Renal Cell Carcinoma [23] supports the wide resection essentially if the metastases are from thyroid or renal carcinomas. Preoperative embolization of these tumors is strongly suggested to reduce intraoperative blood loss.

Periacetabular Defects

Larger lesions can be treated by tumor intralesional resection (curettage) and cement packing and if a solitary metastases lesion, as mentioned before, it could be indicated to get wide margin.

Large lesions with impending or completed acetabular fractures may require surgery, with the goal of constructing a durable hip joint to offer pain relief and allow immediate weight-bearing. This kind of reconstruction requires a construct that effectively bypasses the acetabulum, mainly with total hip arthroplasty with some combination of pelvic or acetabular hardware and cement.

Indications for Surgical Reconstruction of the Acetabulum

1. Lesions that compromise the load transfer from the lower limb to spine are as follows:
   (a) Periacetabular weight-bearing dome compromised by tumor.
   (b) Fracture of the medial or weight-bearing dome of the acetabulum.
   (c) Posterior Ilium lesions not involving the acetabulum can be treated by intralesional resection and cement augmentation.
   (d) Acetabular lesions that are contained (with an intact medial wall) can be reconstructed by a cemented arthroplasty. Protrusio acetabular cups compensate for deficiencies of the medial wall, while cement and pin fixation [18, 19] with modified Harrington methods can be used effectively to reconstruct large defects in the acetabular column and dome.

Reconstruction of the Acetabulum: Options

Metal cage like device, fixed in place with a combination of screws and cement. Into this cage is placed a constrained hip cup or preferable double mobility cup to prevent dislocation of the total hip replacement [24]. A long-stem femoral component is often used, not just to complete the total hip reconstruction, but also to prevent against pathologic fracture of the femur in the case of disease progression. Nevertheless, long cemented femoral stems may lead to adverse events, for instance, hypotension or desaturation that are supposed to be secondary to embolic phenomena, but this event is still in study [25, 26]. Cemented components are generally chosen in the context of metastatic bone disease since the associated use of radiation therapy will limit the degree of bone ingrowth with uncemented prostheses.

(a) Minor lytic lesions when the cortical bone is undamaged, it is a good solution to perform PMMA-augmentation with the use of vertebroplasty kits. Bone cement increases the resistance of the acetabulum and allows full loading of the affected limb (Fig. 9.4).

(b) In case the acetabulum cannot be reconstructed, but significant iliac crest is available, stemmed acetabular implants (inverted ice cream cone prosthesis or pedestal cup) let anchorage of the acetabular shell into the posterior ilium with the stem [27] (Fig. 9.5a, b). Alternatively, a customs acetabular or pelvic prosthesis may be used. As these resections and reconstructions are massive and involve significant blood loss and risk of complications, it is important to have determined preoperatively that the benefits outweigh the risks.

(c) In cases where there are no further reconstruction options available, a flail limb reconstruction by removing the femoral head and debulking of the tumor can be performed; resection arthroplasty may be considered in patients thought to be too great a risk for reconstructive surgery although this procedure generally will preclude return to ambulation in this fragile population and pain relief is variable (Fig. 9.6a, b) [28].

(d) Hindquarter amputation is a last measure reserved for cases of tumor fungating through skin, non-suppressive deep infections, or uncontrollable pain. Although, these procedures have a high rate of complications, and a death rate of about 50% within 12 months [29], the patient's life expectancy and the improvement of the quality of life produced could justify the surgical risks.

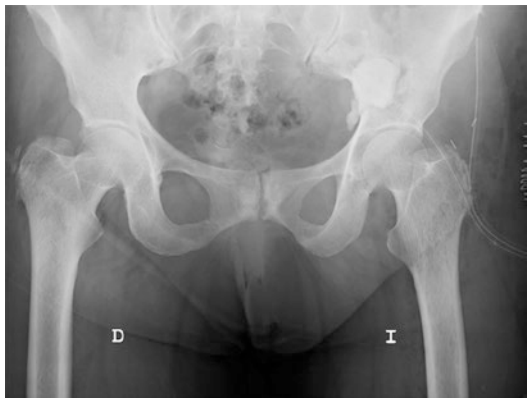

Fig. 9.4 AP view of the pelvis with an osteolytic lesion of supra-acetabular region with percutaneous acetabuloplasty with PMMA

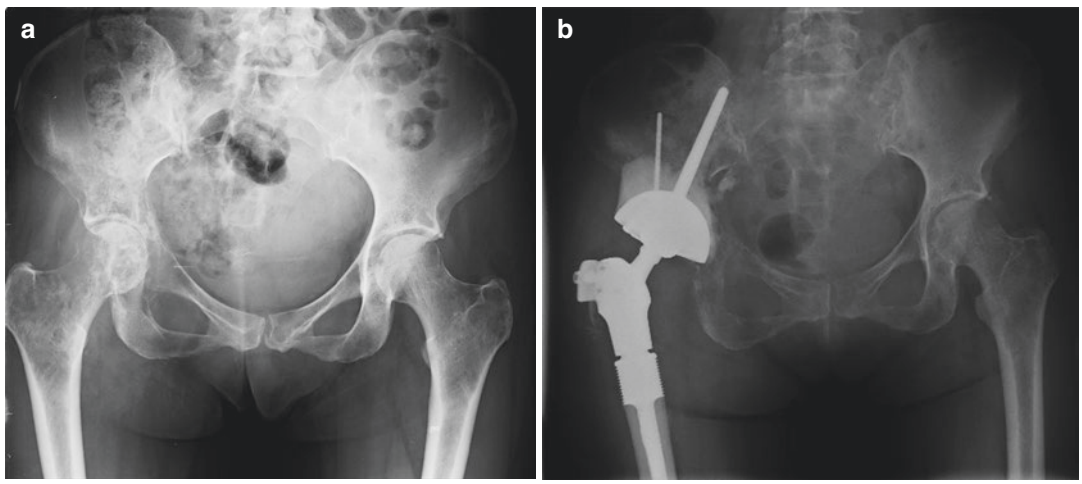

Fig. 9.5 (**a**) Bone metastases from breast carcinoma in a female patient of 45 years old, with severe pain and capable of all self-care, but unable to carry out any work activities. The radiograph demonstrated destruction of the superior, lateral, and medial walls of the right acetabulum (zone 2) and impending femoral of the femoral neck. She is ECOG 2 and class 3 patient; surgery was indicated after tumor board committee. (**b**) Intralesional resection zone II and wide resection of proximal femur. The reconstruction was doing by inverted ice cream cone prostheses (Ice-Cream Cone-style implant (Coned; Stanmore Worldwide Ltd, Elmstree, UK)) with PMMA filling the gap of the acetabulum, dual mobility, no constrained cup, and cemented proximal femur tumoral prostheses

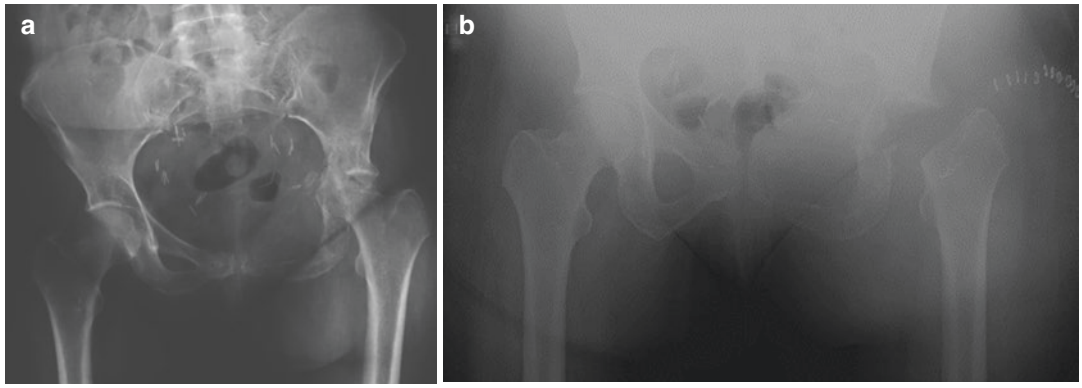

Fig. 9.6 (**a**, **b**) The X-rays shows an acetabular protrusion of the 59-year-old female patient affected by bladder Carcinoma and multiples bone and visceral metastases. She has severe left hip pain. She has a pathologic fracture involving a posterior column and pubic osteolisis. Nonsurgical pretreatments have failed to relieve severe pain. Consequently, the surgical indication to dismiss the pain was a resection arthroplasty after tumor board and consent by the patient and relatives

9.4 Conclusions

Pelvis metastases is a challenging problem that affects the quality of life. Individual treatment should be based on rational guidelines; however, the outcomes of these procedures depend on an interdisciplinary decision on treatment by an entire multidisciplinary team. You need to know the prognoses, class of patient, and types of "surgical" local treatment.

The future of the treatment of metastatic bone disease is stimulating. We hope with time, these complex reconstructions will be addressed more easily with more standard and predictable implant constructs.

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Demolitive Surgery for Pelvic Bone Tumors

10

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10.1 Introduction (History)

The need for external hemipelvectomy, like other amputations for musculoskeletal tumors, has decreased in frequency since the advent of limb salvage in the 1970s [1–3]. The amputative hemipelvectomy is a procedure that involves the removal of all or part of the hemipelvis along with ipsilateral lower extremity. This procedure results in dramatic functional impairments with increased energy and oxygen requirements for locomotion [4]. Medical comorbidities and psychosocial issues regarding body image add to the challenge faced by these patients. While the true incidence of external hemipelvectomy in the United States is not known, it is estimated to be about one per one million each year [5]. Overall, external hemipelvectomies make up 0.5% of lower extremity amputations.

The first report of this procedure in the literature was in 1889, which was performed by Theodor Billroth, who was unsuccessful in his attempt after the patient died from shock (Fig. 10.1) [5]. In 1895, Caciopoli, Jaboulay, and Girard [6, 7], all considered contemporaries of Billroth, were credited for the first hemipelvectomy performed with the patient surviving. By 1909, Ransohoff [8] was recognized as being the first surgeon in the United States to complete an external hemipelvectomy [5, 9].

In 1932, Kellogg Speed [10] is reported in the literature as the first to have coined the term "hemipelvectomy." Gordon-Taylor performed the procedure on a cohort of 41 patients, where the perioperative mortality was 61%. In 1934, he observed a perioperative mortality rate of 59.5%, with the most common causes of death being shock and hemorrhage [11, 12]. In 1948, Robert Wise reported an innovative operative technique used to control hemorrhage by safely ligating the external iliac vessels during a hemipelvectomy (Fig. 10.2) [13, 14]. During the subsequent two decades, progress was made in these procedures, with postoperative mortality rates reduced to 22% and a 21% survival rate at 5 years [11, 12, 15, 16]. Hemipelvic amputations were largely replaced by limb-salvage procedures using allografts and prosthetic composites pioneered by William Enneking and Henry Mankin during the 1960–1970s [1, 2, 17].

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Fig. 10.1 Theodor Billroth. Prussian-born Austrian surgeon (and amateur musician). Reported to be the first to attempt a hemipelvic amputation. (*Courtesy of Bert & Judith van der Waal van Dijk*)

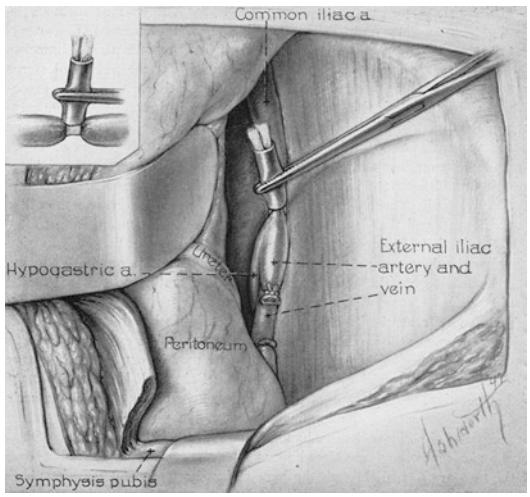

Fig. 10.2 Early hemostatic technique. Drawing of temporary control of the common iliac artery during a hemipelvectomy procedure, using controlling tape placed around the common iliac artery. (*Courtesy of Wolters Kluwer Health, Inc. Robert A. Wise, Control of the Common Iliac Artery during Sacro-Iliiac Disarticulation (Hemipelvectomy), Annals of Surgery 1948*)

10.2 Indication (Bone, Soft Tissue, Nerve, or Vascular)

Anatomical considerations for limb-salvage versus amputative are dependent on the three major structures: the periacetabular bone, femoral vessels, and sciatic nerve. A relative indication for amputative (external) hemipelvectomy has been in cases where two of the three anatomical structures must be resected to achieve an adequate margin [5, 18]. Other indications are for aggressive and malignant lower limb tumors of bone and soft-tissue that cannot be managed with lower level ablative procedures such as hip disarticulation or high above-knee amputation. Other non-oncologic conditions may necessitate hemipelvectomy including extensive infection, trauma-related problems, and complications of pelvic and hip surgery. Decision-making is primarily based upon pathologic analysis and associated margins. If function after a limb-salvage is no better than amputation, a hemipelvectomy should be considered. From this point forward, the use of the term "hemipelvectomy" in this chapter will describe an amputative procedure.

10.3 Preoperative

Staging is necessary prior to consideration of surgical options. Computed tomography (CT) and magnetic resonance imaging (MRI) imaging are essential to determine the anatomic extent of tumor resection. The goal of surgery is an adequate (R0 or R1) margin [19]. Although angiography was utilized in the past, it has been supplanted by CT angiography (CTA) and MR angiography (MRA). With CT data from the bony pelvis, an individualized three-dimensional pelvic model can be used for preoperative planning. The biopsy should be positioned in line with the incision location for the hemipelvectomy. In cases where needle biopsy is not diagnostic, an open biopsy is neccessary. It is important to avoid contamination of the retroperitoneum during the biopsy. When neoadjuvant treatment is completed, the tumor is restaged to assess the plan for surgical margins. Imaging with MRI is necessary in three planes (axial, coronal, and sagittal), and at least T1 (fat sequences) and T2 (fat sup-

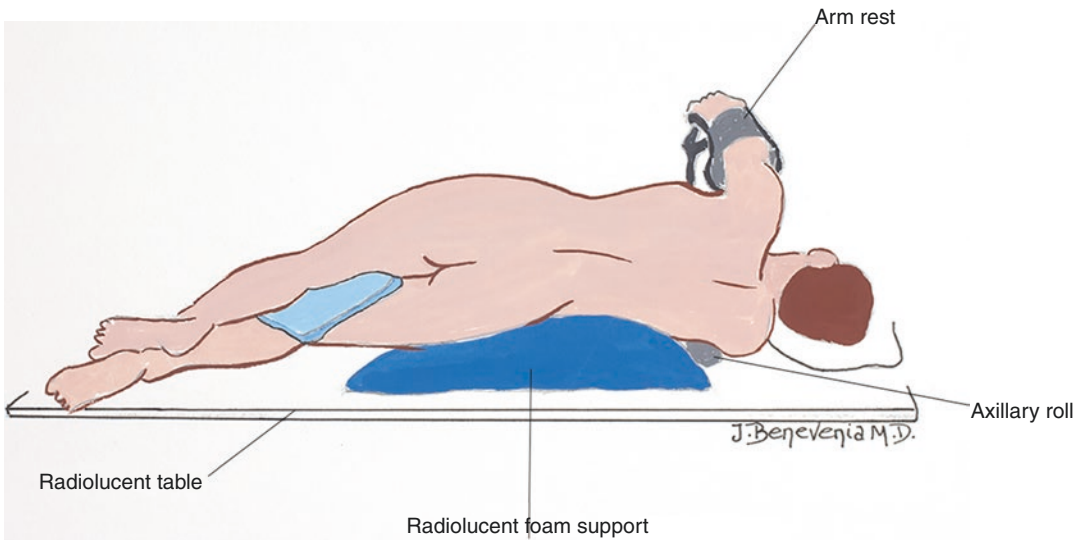

Fig. 10.3 Lateral decubitus position. The positioning of the patient in the later decubitus position is used for the standard hemipelvectomy and subsequent hemipelvectomy subtypes. The patient is placed on a radiolucent table, for imaging purposes, with a bump under the patient's lateral side and axilla. In the upper extremity is secured in an arm rest

pressed) is necessary. Contrast with gadolinium is usually helpful to define tumor extent, especially in recurrence and after prior surgeries.

10.4 Perioperative Considerations

A multidisciplinary approach may include vascular surgery, general surgery, musculoskeletal oncology, urology, and plastic surgery. In cases of visceral involvement, a patient may need a staged diverting colostomy and ureteral stents. Preoperative medical, oncologic, cardiac, and anesthetic evaluations are necessary. Team composition includes surgical technologists, radiological technologist, and proper instrumentation. Use of a Foley catheter before beginning the procedure. Scheduling a postoperative bed in the step-down or intensive care unit.

10.5 Amputative Hemipelvectomy Types

10.5.1 Standard Hemipelvectomy

The following sections will be dedicated to the surgical technique for a standard hemipelvectomy: Insert of a Foley catheter to initiate. The patient is then placed in a lateral decubitus position with the involved side up on a radiolucent table, for use of fluoroscopy (Fig. 10.3). The patient is supported on the table to best facilitate anterior and posterior dissection by modifying table tilt. These initial instructions are applicable to all hemipelvectomy subtypes.

Standard hemipelvectomy involves the disarticulation of the ipsilateral pelvis from the sacroiliac joint to the pubic symphysis and removal of the ipsilateral lower extremity (Fig. 10.4). This requires the division of the pelvic vasculature (Fig. 10.5). A posterior myocutaneous or fasciocutaneous flap from the gluteal region is utilized to cover the resulting defect. This procedure is reserved for tumors originating from within the pelvis, as well as high-grade pelvic tumors located in the anterior and lateral aspect of the pelvis and thigh (Fig. 10.6). This amputative procedure is performed in patients where limb salvage cannot be done safely.

10.5.1.1 Surgical Technique

- **Step 1**: An anterior dissection is performed first; the incision (Fig. 10.7) extends from approximately 5 cm above the anterior supe-

rior iliac spine to the pubic tubercle. Deepen the incision through the tensor fascia, external oblique aponeurosis, and internal oblique and transversalis musculature. Followed by retraction of the spermatic cord medially. Expose the iliac fossa by blunt dissection. Elevate the parietal peritoneum off the iliac vessels, permitting it to fall inferiorly with the viscera. Ligation of the inferior epigastric vessels follows. Release the rectus muscle and sheath from the pubis. Once the iliac vessels are identified, retract the ureter medially, and ligate and divide the common iliac artery and vein. Putting lateral traction on the iliac artery and vein and ligate and divide their branches to the sacrum, bladder, and rectum separating the bladder and rectum from the pelvic sidewall and exposing the sacral nerve roots. In cases requiring further exposure, divide the symphysis pubis and sacroiliac joint before this dissection. The anterior wound is then packed with warm, moist gauze.

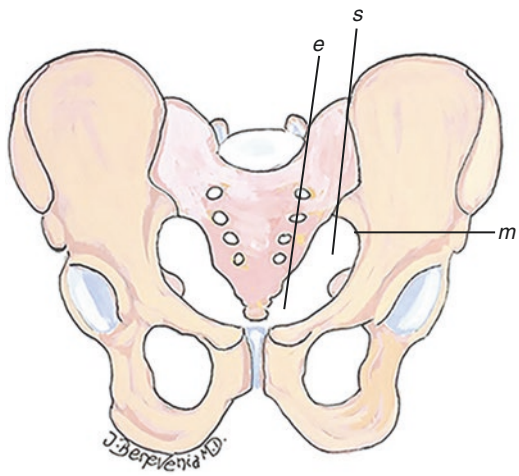

Fig. 10.4 Hemipelvectomy anatomy. Illustration of the differing osteotomies for standard hemipelvectomy and subtypes: Standard (s), Modified (m), and Extended (e)

- **Step 2**: Make a posterior skin incision (Fig. 10.7), extending approximately from 5 cm above the anterior superior iliac spine, going over the anterior aspect of the greater trochanter, parallel to the gluteal fold around the thigh, connecting with the inferior end of the anterior incision. The posterior flap is raised by dissecting the gluteal fascia directly off the gluteus maximus (Fig. 10.8). Include

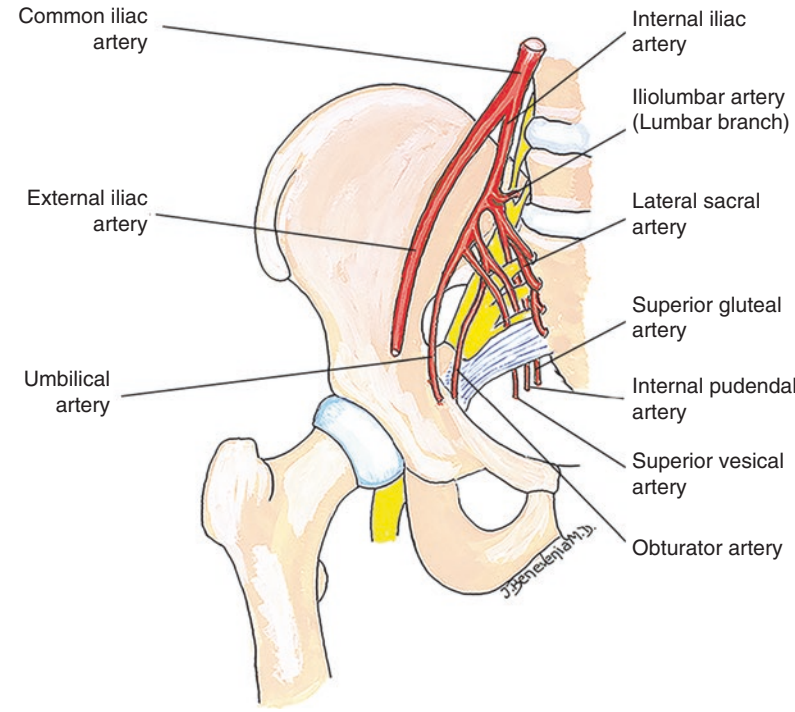

Fig. 10.5 Hemipelvectomy pelvic vascular anatomy

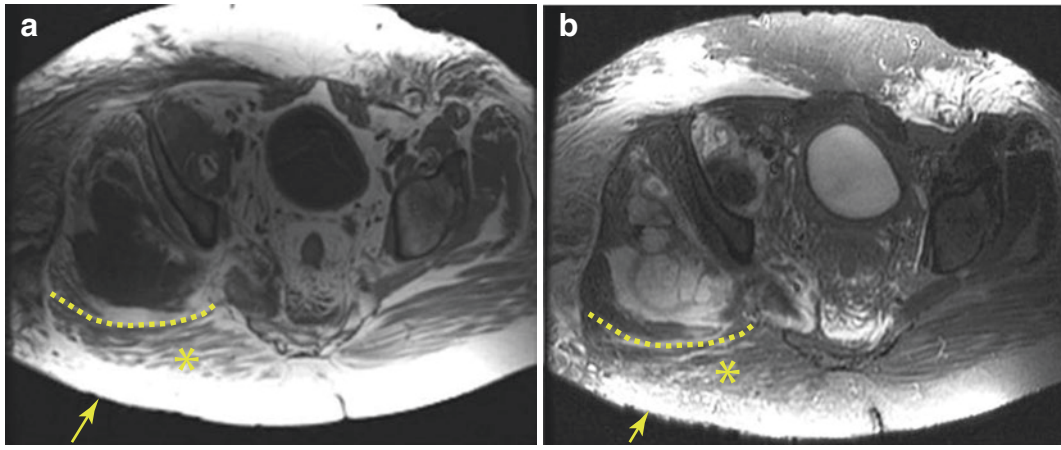

Fig. 10.6 Posterior flap hemipelvectomy case. A 51-year-old male patient with chondrosarcoma (high-grade) of the right pelvis. Presented with 4–5 months of significant right lower limb pain, following the sciatic nerve distribution, and an enlarging right pelvic mass. Imaging showed a large tumor that begins from the most proximal portion of the right ilium involving the posterior soft tissue of the gluteus medius and gluteus minimus, with preservation of the gluteus maximus. The tumor was found near the right greater sciatic notch with notable peritumoral edema. A decision was made to perform an external hemipelvectomy. (**a**) Magnetic resonance imaging (MRI) T1 (left) showing preservation of the gluteus maximus (asterisks) and skin (arrows) for posterior flap wound closure. (**b**) MRI T2 (right) showing chondrosarcoma on the anterolateral side of the right pelvis

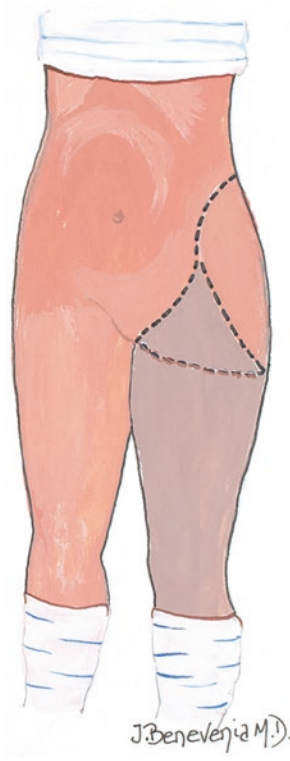

Fig. 10.7 Posterior flap incision. The incision begins at the iliac crest, extending across the anterior superior iliac spine, posteriorly across the gluteal crease, and anteriorly along the pubis in the groin to complete the incision

the fascia with the flap. When possible, include the medial portion of the gluteus maximus with the flap. Superiorly elevate the flap off the iliac crest. Divide the external oblique, sacrospinalis, latissimus dorsi, and quadratus lumborum from the crest of the ilium. Reflect the gluteus maximus from the sacrotuberous ligament, coccyx, and sacrum. Divide the iliopsoas muscle; genitofemoral, obturator, and femoral nerves; and lumbosacral nerve trunk at the level of the iliac crest.

Abduct the hip, placing tension on the soft tissues around the symphysis pubis. Pass a long right-angle clamp around the symphysis and divide it with a scalpel. The sacral nerve roots are divided preserving the nervi erigentes. Reflect the iliacus muscle laterally, exposing the anterior aspect of the sacroiliac joint. Divide the joint anteriorly with a scalpel or osteotome and divide the iliolumbar ligament. Place considerable traction on the extremity, separating the pelvic sidewall from the viscera. Proceeding from anterior to posterior, divide the following from the pelvic sidewall: urogenital diaphragm, pubococcygeus, ischiococcygeus, iliococcygeus, piriformis, sacrotuberous ligament, and sacrospinous ligaments. These structures are all

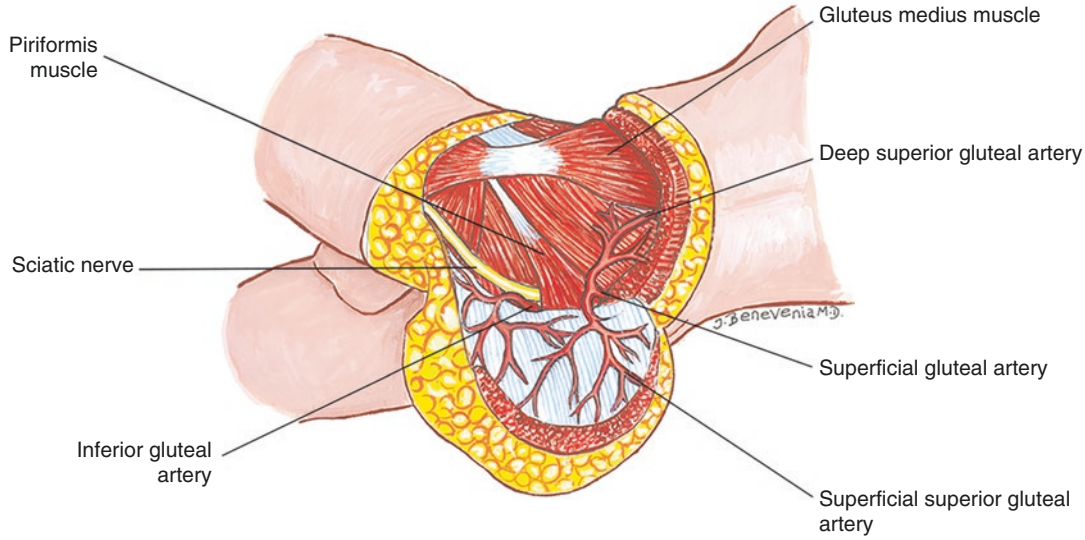

Fig. 10.8 Posterior flap dissection

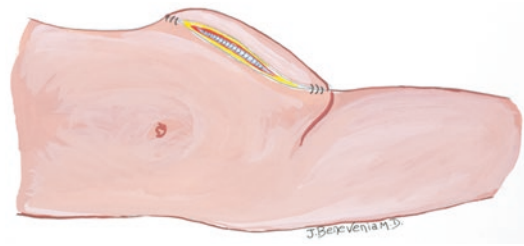

Fig. 10.9 Closure of posterior flap. Gluteal fascia is sutured to the fascia of the abdominal wall, with suction drains (not shown) placed prior to skin closure

divided under tension. Move the extremity anteriorly and divide the posterior aspect of the sacroiliac joint to complete the dissection.

Closure: Place suction drains in the wound and suture the gluteal fascia to the fascia of the abdominal wall. Close the skin (Fig. 10.9). Regarding postoperative care, drains, urinary catheters, and vacuum sponge dressings are used as per surgeon preference. Drains are discontinued when output volume is <50 cc per 12-h shift.

10.5.2 Anterior Flap Hemipelvectomy

Anterior flap hemipelvectomy is reserved for tumors located in the gluteal region and proximal section of the posterior thigh [20]. The resulting operative defect involving the gluteal region, hemipelvis, and lower extremity is then enveloped using an anterior flap made from the quadriceps femoris muscles, subcutaneous layers, and skin. The superficial femoral artery is spared to supply the anterior myocutaneous flap (Fig. 10.12, 10.13, 10.14, 10.15, and 10.16).

10.5.2.1 Surgical Technique

The patient is placed in the lateral decubitus position with the operated side up. Secure the patient to the table so that it can be tilted to facilitate the anterior and posterior dissections (Fig. 10.6). The skin from toes to rib cages is prepared and drape the extremity free. A mark is then made for the skin incision that should include the length and width of the anterior flap that will adequately cover the posterior defect. An incision is then made superiorly across the iliac crest to the mid-lateral point, around the buttock just lateral to the anus, and the mid-medial point of the thigh. The incision is taken down the thigh a distance adequate to cover the posterior defect, across the front of the thigh to the mid-lateral point, and superiorly to join the superior incision (Fig. 10.10).

- **Step 1**: Posterior dissection is done first. Skin margins are preserved, at least 3 cm from the

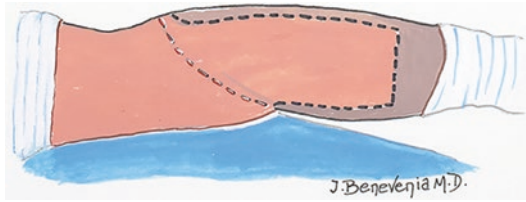

Fig. 10.10 Anterior Flap incision (right). The incision is made going superiorly across the iliac crest to the mid-lateral point, around the buttock just lateral to the anus, and to the mid-medial point of the thigh. The incision is then brought down the thigh a distance that is adequate to cover the posterior defect, across the front of the thigh to the mid-lateral point, and superiorly to join the superior incision

anus. The gluteus maximus and sacrospinalis are detached from the sacrum; followed by the external oblique, sacrospinalis, latissimus dorsi, and quadratus lumborum muscles from the iliac crest. Flex the hip and place the tissues in the region of the gluteal fold under tension. Detach the origins of the gluteus maximus from the coccyx and sacrotuberous ligament. Bluntly dissect laterally to the rectum into the ischiorectal fossa. Maneuver to the front of the patient and deepen the anterior incision at the linking point of the middle and distal thirds of the thigh through the quadriceps to the femur. Continue the dissection laterally in a cephalad direction to the anterior superior spine severing the vastus lateralis from the femur; separate the tensor fascia femoris from its fascia including the tumor.

- **Step 2:** Medially dissect, starting at Hunter's canal, and ligate and divide the superficial femoral vessels. Track the vessels superiorly to the inguinal ligament, dividing and ligating multiple small branches to the adductor muscles. Apply upward traction on the myocutaneous flap and detach the vastus medialis muscle and intermedius from the femur. Ligate and divide the deep femoral vessels at their origin from the common femoral artery and vein (Fig. 10.11). Separate the myocutaneous flap from the pelvis by releasing the abdominal muscles from the iliac crest, the sartorius from the anterior superior spine, the rectus femoris from the anterior inferior spine, and the rectus abdominis from the pubis. Retract the flap medially and dissect along the femoral nerve into the pelvis to expose the iliac vessels. Separate the symphysis pubis while protecting the bladder and urethra. Ligate and divide the internal iliac vessels at their origin from the common iliac. While applying medial traction on the bladder and rectus, divide the visceral branches of the internal iliac vessels. Divide the psoas muscle as it joins the iliacus muscle and divide the underlying obturator nerve, but protect the femoral nerve going into the flap. Separate the lumbosacral nerve and the sacral nerve roots. Apply traction on the pelvic diaphragm by elevating the extremity and divide the urogenital diaphragm, levator ani, and piriformis near the pelvis. Separate the sacroiliac joint and the iliolumbar ligament and remove the tumor.

Closure: Turn the quadriceps flap onto the posterior defect and suture to the abdominal wall, sacrospinalis, sacrum, and pelvic diaphragm, with a suction drain placed prior to skin closure (Fig. 10.17). For postoperative care, the patient may ambulate when comfort and stability permits.

10.5.3 Extended Hemipelvectomy

Under certain circumstances, a standard external hemipelvectomy is not sufficient to prevent a tumor from spreading. In instances where there is tumor involvement of the ilium or ischium that extends up to the sacroiliac joint (SI) and invades through the cartilage, performing a standard hemipelvectomy, in this case, would lead to possible tumor spillage and dissemination. Therefore, a more proximal transection of bone through the sacral foramina, known as an extended hemipelvectomy, can prevent these potentially fatal complications from occurring. Buttock tumors located within the gluteus muscles and some soft tissue sarcomas that can extend along nerve sheaths can also be managed with an extended

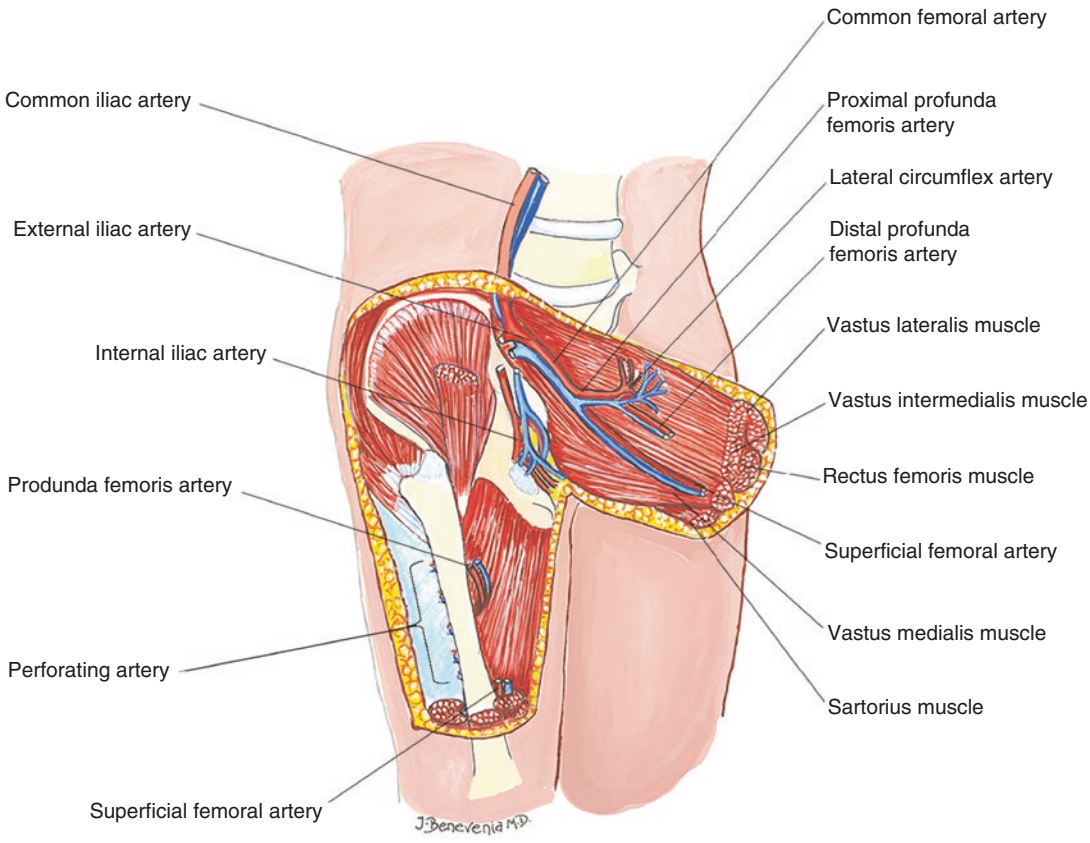

Fig. 10.11 Myocutaneous anterior flap anatomy with preservation of the superficial femoral vessels, deep femoral vessels, and quadricep musculature

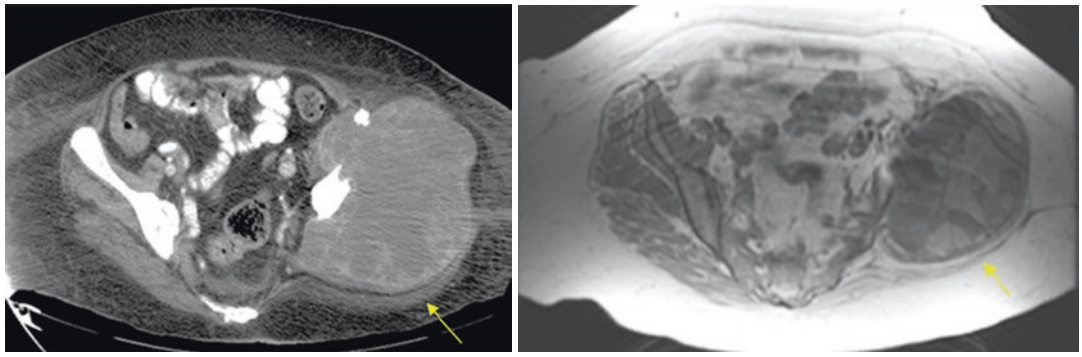

Fig. 10.12 Anterior flap hemipelvectomy. Case 1—42-year-old female patient with a history of osteosarcoma in the left femur with previous surgical attempts at limb salvage, who presented with persistent left lower extremity pain and enlarging mass. Imaging demonstrated recurrent left-sided pelvic osteosarcoma with destruction of the left hemipelvis, extending posteriorly to the gluteal region. CT Pelvis Axial (top) shows destruction of the left iliac wing. MRI T1 axial (bottom) shows thinning of the gluteus maximus (yellow arrows). Based upon the anatomic extent of the recurrence, anterior flap hemipelvectomy was performed

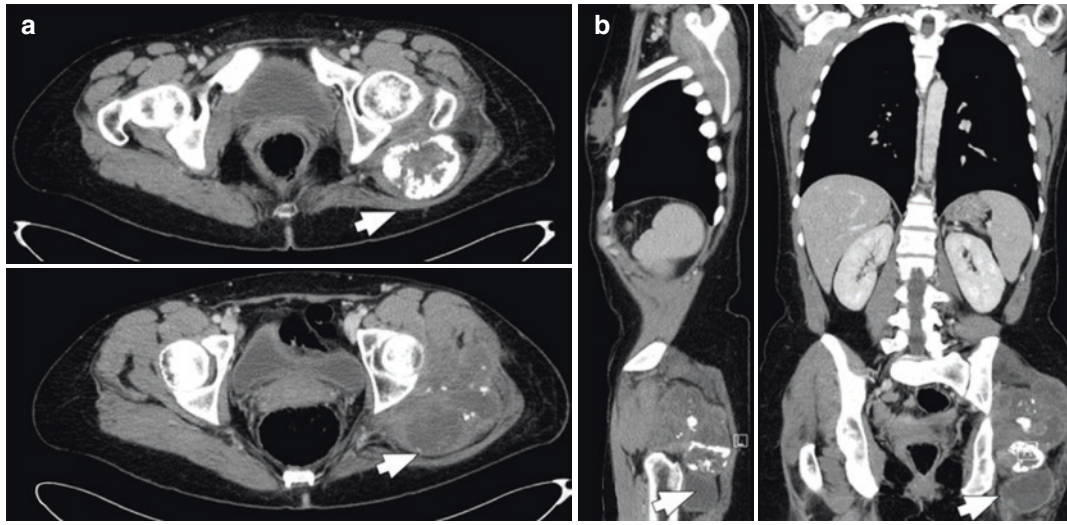

Fig. 10.13 Anterior flap hemipelvectomy Case 2. Thirty-five-year-old female patient with synovial sarcoma of the left hemipelvis. (**a**) Axial CT scan shows extensive involvement of the soft tissue in the posterior region of the left hemipelvis. The synovial sarcoma (white arrow) appears as soft tissue mass with a slightly higher density than muscle and with the easily detectable calcifications. (**b**) Sagittal and coronal CT images of the pelvis better demonstrate the tumor extension from the proximal ilium to the subtrochanteric area, including the sciatic nerve

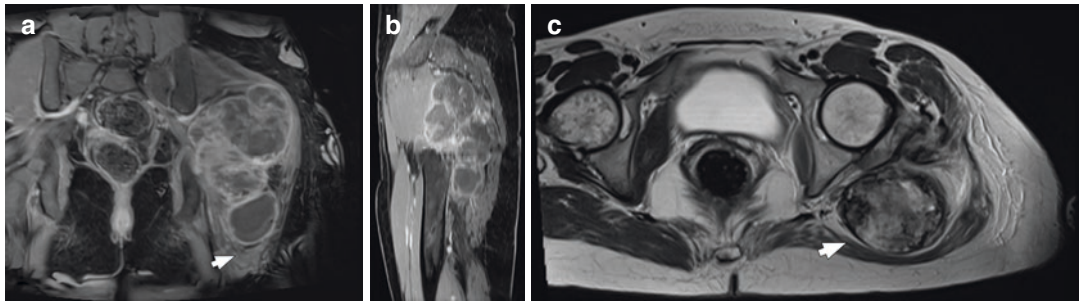

Fig. 10.14 Anterior flap hemipelvectomy Case 2 cont. Thirty-five-year-old female patient with synovial sarcoma of the left hemipelvis. A subsequent MRI demonstrates with coronal (**a**) sagittal (**b**), and axial (**c**) images demonstrate the marked inhomogeneity, enhancement, and septation. Pathology confirmed a synovial sarcoma with typical SYT-SSX chromosomal translocation

hemipelvectomy. This procedure includes the removal of the hemipelvis along with additional structures not limited to the ipsilateral sacrum, lumbar spine, and/or contralateral pelvis (Fig. 10.18).

10.5.3.1 Surgical Technique

- **Step 1**: In most circumstances, bone transection is done through the sacral foramina; a lumbo-sacral laminectomy with nerve root ligation is often necessary. If an anterior flap is being utilized, the medial skin incision should be made over the mid-sacral spines, allowing for visualization of the dorsal sacral foramina. If a posterior flap is being utilized, this flap must be dissected back to the mid-sacral spines to allow for accurate localization of the dorsal foramina. In the anterior aspect, all of the branches of the internal iliac artery overlying the sacral nerve roots must be very

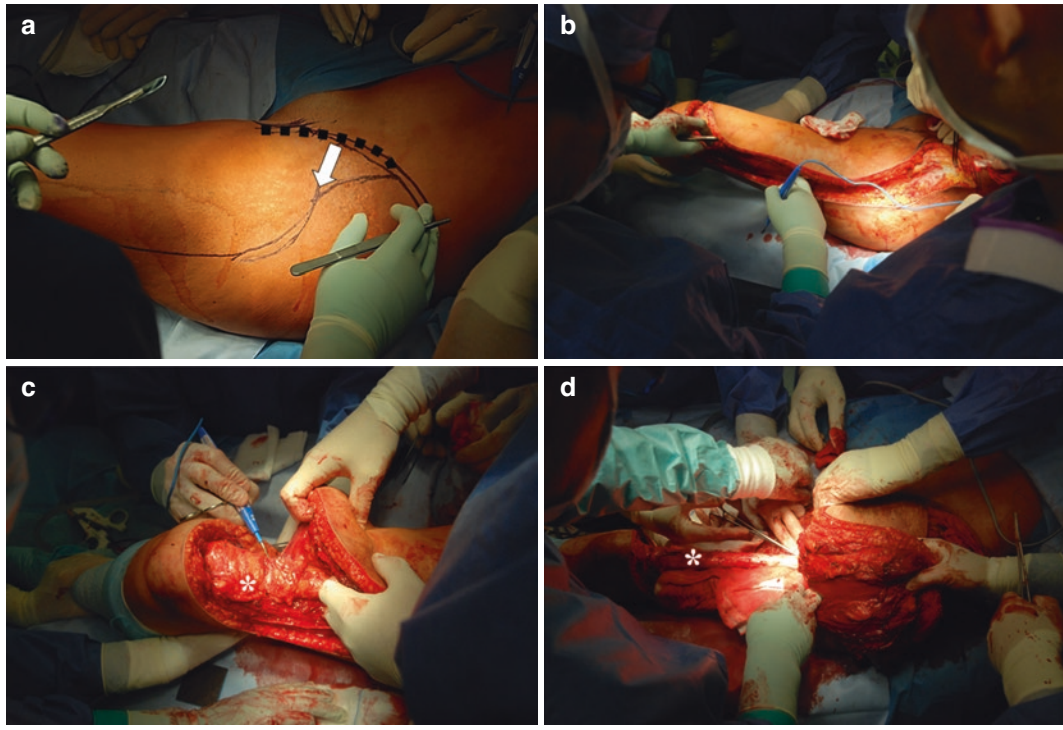

Fig. 10.15 Anterior flap hemipelvectomy Case 2 cont. Thirty-five-year-old female patient with synovial sarcoma of the left hemipelvis. Surgery consists of an external hemipelvectomy with partial resection of the ilium and atypical anterior thigh flap considering the extensive involvement of the gluteus. The landmarks are the great trochanter, anterior superior iliac spine. The starting point is the posterior inferior iliac spine and the incision follows the iliac crest reaching the anterior superior iliac spine. Then, it extends caudally on the anterior aspect of the thigh and then laterally just below the greater trochanter (white arrow). (**a**) Vascularization of the flap is based on the major vascular pedicle of the pelvis and extremity vessels (dot black line). (**b**) Lateral incision of the myocutaneous flap. (**c**) Release of the quadriceps femoris muscle from the femur (asterisk). (**d**) During this phase, care must be taken not to separate muscle bundles of the myocutaneous flap from the overlying skin

carefully dissected. Failure to secure these vessels may lead to substantial blood loss when the nerve roots and sacrum are being transected.

- **Step 2**: Once the vessels are secured, move to the posterior aspect of the patient. Beginning at the tip of the coccyx, an osteotome is now utilized to divide the coccyx and sacrum in a plane that divides the sacral foramina. The initial course of the osteotome should be parallel to the mid-sacral spines. Then, the surgeon should reach around the coccyx with their left hand to find the S-5 neural foramina within the sacrum, and then work superiorly, progressing from one foramina to the next. The surgeon should be holding the osteotome in their right hand and allow the assistant to drive the osteotome through the bone with the mallet. The surgeon's left hand will have located the next highest foramina and will guide the direction of bony transection. The bone at the upper portion of the sacrum is fragile, and therefore the surgeon must be careful to not accidentally fracture through the bone during osteotomy. Finally, the lumbosacral ligament is divided, and the specimen is released.

10.5.4 Modified Hemipelvectomy

The modified hemipelvectomy involves resection of the hemipelvis through the ilium for tumors of the lower pelvis sparing a portion of the upper bony pelvis (Fig. 10.19). The wing of the ilium is

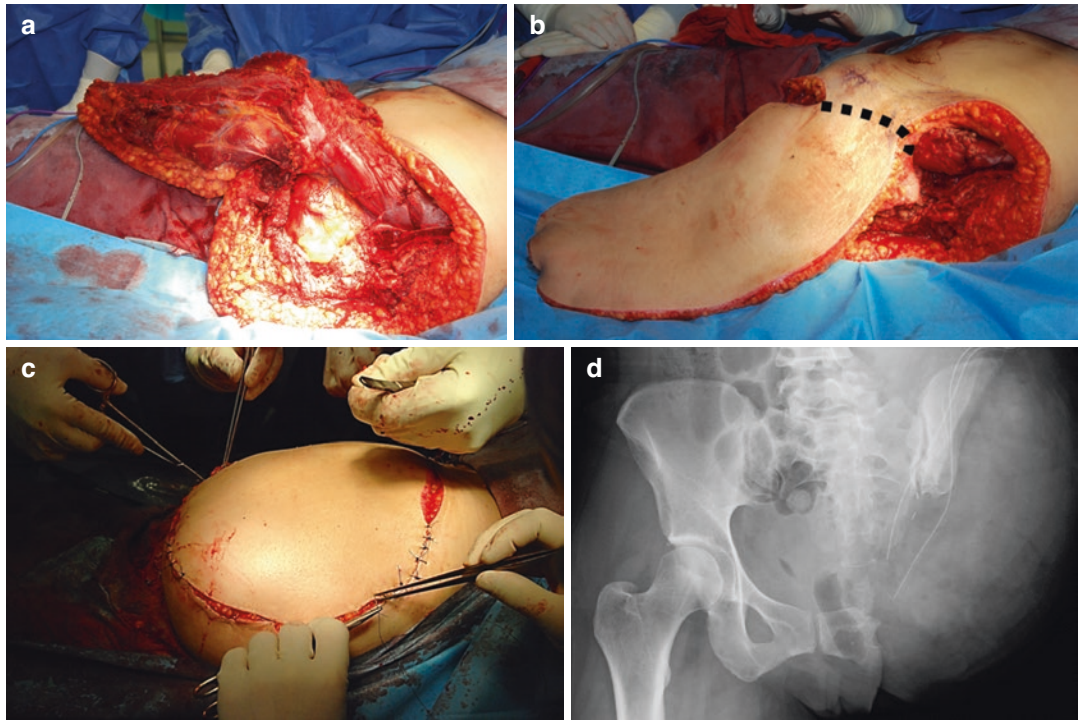

Fig. 10.16 Anterior flap hemipelvectomy Case 2 cont. Thirty-five-year-old female patient with synovial sarcoma of the left hemipelvis. (**a**) Surgical field after tumor removal. (**b**) The myocutaneous flap is well vascularized (dot black line) and the skin is adequate for coverage of operative defect. (**c**) Closure, the myocutaneous flap is folded posteriorly and bleeding points are secured. (**d**) Postoperative X-ray shows the partial resection of the ilium and the pubis (modified hemipelvectomy)

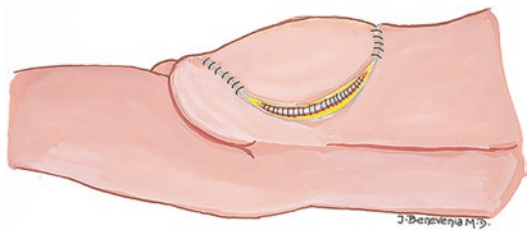

Fig. 10.17 Closure of anterior flap (right). The quadriceps flap is turned onto the posterior defect for the closure of the wound, finished by suturing the quadriceps to the abdominal wall, sacrospinalis, sacrum, and pelvic diaphragm, with the use of drains, urinary catheters, and vacuum sponges similar to posterior flap

the most commonly spared structure; however, the spared portion depends on the location of the bony pelvic tumor. The most common indication for a modified hemipelvectomy is large soft tissue sarcomas located high in the medial thigh. The modified hemipelvectomy may add a cosmetic and functional aspect. Sparing of the wing of the ilium allows the patient to maintain a normal waistline, leading to clothes feeling and fitting better for the patient. The remaining ilium may serve as a "post" for a prosthesis. Technical advantages are also present, as this procedure spares the superior and inferior gluteal vessels

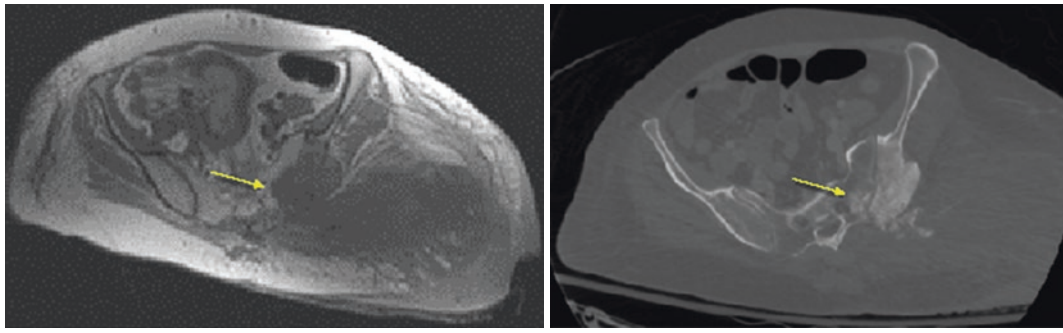

Fig. 10.18 Extended hemipelvectomy case. A 28-year-old female with left pelvic osteosarcoma (right). Presented to the ICU with a fungating large soft tissue mass at the left pelvis. Imaging showed tumor replacement of the posterior aspect of the ilium, extension through the SI joint, and a large soft tissue mass expanding through the gluteal musculature and lateral thigh. MRI T1 (top) and CT (bottom) axials shows tumor involvement of the sacroiliac joint (SI) (arrow) with penetration of gluteal structures. Due to extensive tumor involvement at the hemipelvis and sacrum, as well as tumor crossing the midline, a decision was made to perform an extended hemipelvectomy

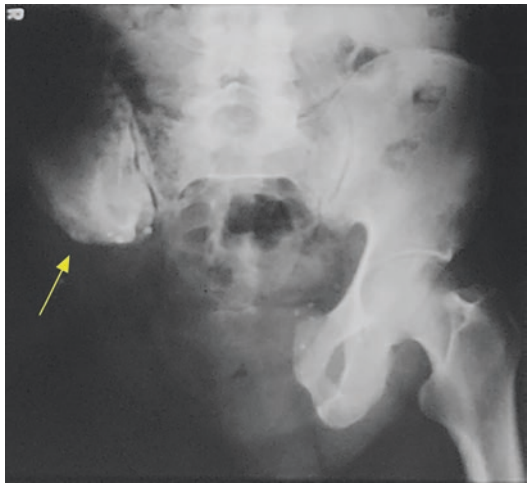

Fig. 10.19 Modified hemipelvectomy. Postoperative AP radiography of a patient with a right pelvic sarcoma that underwent a modified hemipelvectomy, with preservation of a portion of ilium (yellow arrow)

along with the posterior portion of the gluteus maximus muscle, improving the vascularity of the long posterior skin flap used to cover the resulting wound.

10.5.4.1 Surgical Technique

- **Step 1**: The incision and early dissection for a modified hemipelvectomy are identical to the steps described above for a standard hemipelvectomy, except less skin is incised at the level of the anterior superior iliac spine and a longer posterior skin flap is created for closure. Dissection of the posterior skin flap is considered complete when an imaginary line can be drawn between the anterior superior iliac spine and the tip of the coccyx. After this step, shift focus to the anterior aspect of the patient and perform blunt dissection to expose the common iliac artery and vein. The ipsilateral rectus muscle should be released from the superior pubic ramus, then the pubic symphysis is divided. The greater sciatic foramen should be identified from both the anterior and posterior aspects. All of the gluteus muscles should then be divided with electrocautery between the anterior superior iliac spine and the posterior aspect of the greater sciatic foramen. The iliacus muscle and psoas tendon should be divided on the anterior to expose the pelvis. Next, the sartorius muscle is identified arising from the anterior superior iliac spine and is dissected through its tendon.
- **Step 2**: Now with the pelvis exposed, a Gigli saw is used to divide the pelvis from the sciatic foramen to the section of bone between the origin of the rectus femoris and sartorius muscles. The superior vesical, obturator, and inferior vesical vessels are severed. Since the superior and inferior gluteal vessels are preserved, the gluteus muscles maintain perfu-

sion. The sciatic nerve is now divided at the same level that the muscles were divided. Strong upward traction is placed on the extremity to transect the urogenital diaphragm and levator ani muscle to create a free plane above the urethra, bladder, and rectum. Drainage and closure of the wound are similar to the steps described above for a standard hemipelvectomy. It is important to conserve the skin over the anterior superior iliac spine as increased tension over the bony prominence may lead to delayed wound healing.

10.5.5 Compound Hemipelvectomy

In certain cases, pelvic tumors can extend to the rectum, female adnexal region, bladder, and other viscera (Fig. 10.20). Resection of abdominal and/or pelvic viscera along with amputation of the hemipelvis may be indicated in such circumstances.

Patients best suited for this procedure are those with locally aggressive pelvic tumors that are of low-grade malignancy. Compound hemipelvectomy tends to carry an increased risk of postoperative infection due to the resection of the abdominal or pelvic viscera.

A multidisciplinary approach and consults of a variety of services may be warranted given the nature of this subtype.

10.6 Soft-Tissue Coverage

Flap sectional should be considered preoperatively for proper planning. Surgical site closure can be accomplished with an anterior or posterior myocutaneous flap depending on the location of the pelvic tumor. While myocutaneous hemipel-

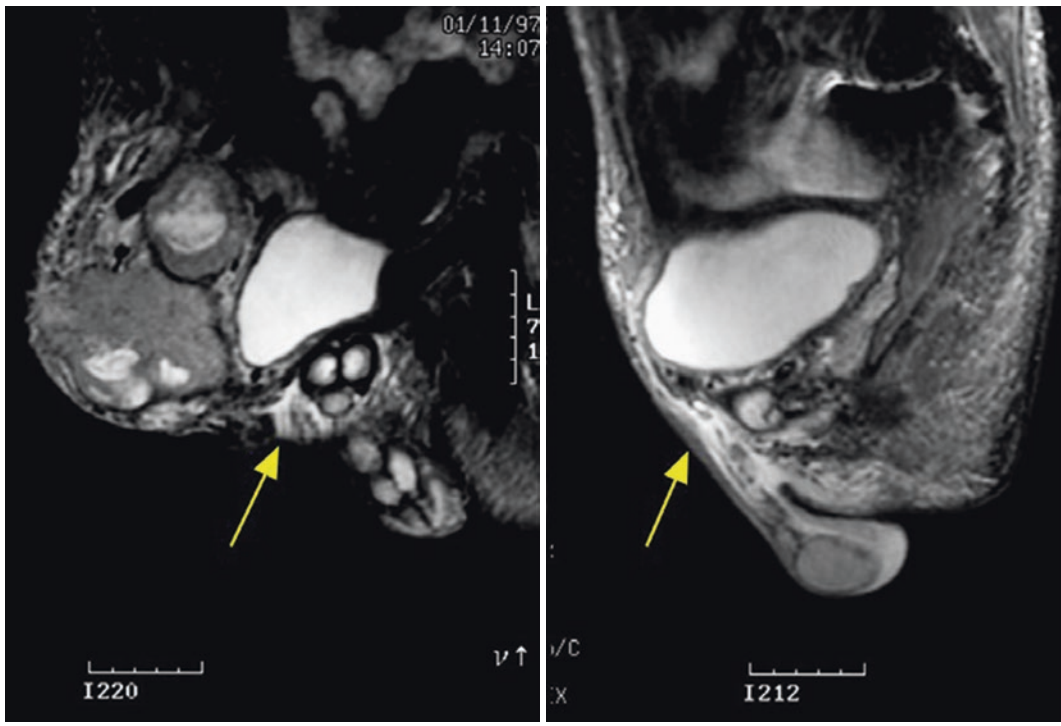

Fig. 10.20 Compound hemipelvectomy case. Fifty-three-year-old male patient with undifferentiated pleomorphic sarcoma (UPS). Presented with 5 months of right knee and hip pain. Imaging demonstrated tumor involvement in the right hemipelvis, right lower extremity. MRI T2 Coronal (left) and Sagittal (right) imaging show tumor involvement of spermatic cord (yellow arrows). Given the extent of tumor involvement to the spermatic cord, a compound hemipelvectomy was performed

vectomy flaps are adequate for closure, cases do have a high probability of wound morbidity after soft-tissue reconstruction [21]. The involvement of plastic surgeons in cases that would include the use of a free flap closure is prudent and sometimes required. The availability and consultation of a plastic surgery service are dependent on institution and surgeon preference. In a study by Apfelstaedt et al., there was no statistical difference between flap failure and ligation of the common iliac artery compared with ligation of the external iliac artery only. Increased operative time and perioperative complications can also lead to an increase in fap necrosis and infection. The best option for reconstructive faps is the use of the amputated tissue (free fillet faps) [21–23]

10.7 Complications

The rate of associated morbidities regarding external hemipelvectomy is high. While literature related to the complications following hemipelvectomy is scarce, rates are estimated to be from 20% to 50% [5, 24]. Given the high incidence of postoperative complications, in a review of 136 patients who underwent external hemipelvectomy, Kiiski et al. [25] observed postoperative 30-day mortality of less than 1%. Senchenkov et al. [26] discovered that the complexity of the case and increased time in the operating room were factors associated with increased rates of postoperative complications. Flap necrosis and wound infection are considered to be the most common complications in both internal and external hemipelvectomy. Flap necrosis typically requires operative debridement and tissue coverage, often delaying the patient's rehabilitation process.

Many patients have significant phantom pain in the early postoperative course. Residual limb spasm has been reported to occur more commonly than phantom pain and may present weeks or even months after the procedure; it is most common after traumatic hemipelvectomy. In their review of 160 external hemipelvectomies, Senchenkov et al. reported a morbidity rate of 54%, including intraoperative genitourinary (18%) and gastrointestinal injuries (3%) [23, 25, 27, 28]. Regarding quality of life measures, a study by Beck et al. observed that patient's overall quality of life parameters was comparable between groups undergoing both external and internal hemipelvectomy. Beck would also observe those who underwent an external hemipelvectomy had an increase in experienced pain severity and were less independent in bladder function [29].

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Principles of Pelvic Surgery

11

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11.1 Introduction

Hemipelvectomies and pelvic resections are infrequently performed procedures, mainly performed for primary malignancies of bone and soft tissue. These surgeries may also be performed for metastatic disease, infection, severe trauma, and palliation. A hemipelvectomy, or hindquarter amputation, involves resecting the entire hemipelvis and ipsilateral extremity, and prior to the 1970s, pelvic tumors were often treated with hindquarter amputations. In 1978, Enneking and Dunham [1] described a large series of pelvic resections classifying them by the segment of the innominate bone resected. This widely accepted classification, and its basis forms the foundation of the subsequent decades of research on pelvic resections or modified internal hemipelvectomies. It is the finding of multiple centers from around the globe, that with appropriate patient selection, advances in imaging, adjuvant treatments, and contemporary resection and reconstructive techniques, limb salvage surgery for pelvic sarcoma can be performed safely with reasonable outcomes.

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11.2 Epidemiology

Primary bone sarcomas make up about 0.2% of all cancers with 15–20% of those being located in the pelvis [2–4]. The most common malignant primary bone sarcomas encountered are osteosarcoma (35%), chondrosarcoma (30%), and Ewing's sarcoma (16%) [2]. Similarly, they are the most common primary bone sarcomas found in the pelvis. However, 45% of Ewing's sarcoma cases are located in the pelvis compared to only 5% of osteosarcomas found in the pelvis. Benign bone tumors that occur in the pelvis that often require surgical treatment include giant cell tumor of bone, osteoblastoma, and aneurysmal bone cyst. Primary soft tissue sarcomas as a group are more common than primary bone sarcomas and account for 1.0% of all cancers with 5% being located in the pelvis [3–5]. The same surgical principles used in the management of pelvic primary bone sarcoma can be applied to soft tissue sarcomas involving adjacent pelvic bony structures.

11.3 Surgical Indications

Surgical extirpation is the cornerstone of sarcoma management and when the sarcoma involves the innominate bone, surgical resection is often indicated if it is technically feasible. Pelvic sarcomas often present when they are already advanced

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owing to the significant capacity of the pelvis to accommodate a growing tumor prior to it becoming symptomatic to the patient. This delay in detection translates into tumors that are large and in close proximity to viscera and neurovascular structures. Therefore, surgical treatment becomes difficult.

The goal of surgical management is to remove the tumor en bloc and with negative surgical margins. Where preservation of the limb is possible with the least effect on the patient's function, limb salvage surgery is preferred [6]. If the extent of tumor involvement in the pelvis makes resection with negative margins not feasible, this would be an indication for an external hemipelvectomy or hindquarter amputation.

Some patients' pelvic bone tumors may be managed with radiotherapy or other modes of surgical intervention. An example is Ewing's sarcoma, which is generally sensitive to chemotherapy and radiation therapy. In such patients where resecting the pelvic tumor would cause significant morbidity, definitive radiation therapy is an option that has been reported to provide similar local control rates and acceptable complications [7, 8]. However, this is debatable as some studies have documented a benefit to surgical management of pelvic Ewing's sarcoma [9–11]. Other minimally invasive options have also been described for metastasis, including CT-guided ethanol and thermal ablation, cryoablation, radiofrequency ablation, and percutaneous acetabuloplasty with cement [12].

Other malignancies that may affect the pelvis include lymphoma and myeloma, where the mainstay treatments for those patients are chemotherapy, radiation therapy, and bisphosphonates. However, if patients have sustained or are at risk for a pathologic fracture, surgical management such as curettage, cementation, and joint reconstruction may be performed [3, 12].

There has been an overall decrease in mortality from cancer with the advances in systemic treatment, with mortality rates in the USA decreasing for men and women by 1.8% and 1.4%, respectively [13]. Metastatic disease affecting the pelvis is significantly more prevalent than pelvic primary bone or soft tissue sarcomas. Similarly to lymphoma and myeloma, pelvic metastatic disease is usually first treated with nonoperative management. However, studies have reported improved survival and lower morbidity with pelvic resection compared to curettage in certain patient populations [12]. These included patients with a solitary metastatic lesion, primary carcinoma with good prognosis, and a long time interval between diagnosis of primary and metastasis [12, 14]. Management of metastatic disease has been discussed in greater detail in an earlier chapter.

11.4 Surgical Planning

Management of pelvic tumors requires a multidisciplinary approach from both medical and surgical subspecialties. These include radiology, radiation oncology, pathology, medical oncology, orthopedic oncology, urology, vascular surgery, surgical oncology, colorectal surgery, neurosurgery, gynecology, and plastic surgery. Medical oncologists and radiation oncologists manage adjuvant therapy regimens and treatment timing. Radiologists are able to assess tumor extent and help plan/perform the biopsy. Depending on structures involved, other surgical subspecialties may be required intraoperatively for successful and safe tumor resection.

Preoperative laboratory and imaging workup are essential to guiding treatment in pelvic malignancies. Laboratory studies including complete blood count with differential, electrolyte panel, and serum and urine protein electrophoresis are helpful for diagnosing pancytopenia, life-threatening hypercalcemia, and multiple myeloma. Imaging studies should start with plain radiographs of the pelvis to get a complete evaluation of pelvic involvement, to identify the pattern of bone formation in the tumors, and serve as a baseline for future surveillance. Advanced imaging studies including a contrasted MRI are crucial for determining the extent of local disease, the involvement of major neurovascular structures and viscera, and the feasibility of limb salvage surgery. A CT of the pelvis is useful to evaluate the amount of bone destruction and is

often complementary to findings seen on MRI. If the extent of vascular involvement is unclear from a contrasted MRI, formal MR or CT angiography may be useful. When a primary bone sarcoma is suspected, staging studies include a CT chest and bone scan. If a soft tissue sarcoma is suspected, CT chest for staging is required. Should the pelvic lesion be suspected to be metastatic in nature, a CT chest, abdomen, and pelvis is also performed to identify a visceral primary lesion. In multiple myeloma patients, a skeletal survey should be performed (CT or plain X-ray) to evaluate for other sites of disease.

Once all appropriate imaging has been performed, the biopsy can be planned. The goals of the biopsy are to obtain an adequate amount of tissue that results in a definitive diagnosis without compromising the planned resection. The biopsy is a fundamental part of the workup and can cause significant morbidity if done inappropriately. Inappropriate or inadequate biopsy can lead to a change in treatment plan, making wide resection and limb salvage surgery no longer feasible [6, 15]. Percutaneous core needle biopsies (with or without CT guidance) and open biopsy techniques are performed. If a CT-guided biopsy is planned, there must be a discussion between the treating surgeon and the radiologist performing the biopsy about the planned approach for resection in order to prevent contamination of uninvolved compartments. Care must also be taken not to contaminate the retroperitoneum. If an incisional biopsy is performed, the biopsy should be made in line with the planned surgical incision. This is usually along the iliac crest. Intraoperative fluoroscopy may be necessary for tumors without a soft tissue component and contained within the bone to ensure lesional tissue is collected. Once enough tissue has been obtained, meticulous hemostasis must be achieved to prevent hematoma formation as this may extend the area of tumor contamination. Lastly, careful wound closure is important to minimize wound complications, especially if neoadjuvant chemotherapy or radiation therapy is anticipated [15].

Following neoadjuvant treatment (chemotherapy or radiation therapy), restaging studies should be performed. These studies typically include local imaging (plain X-rays, MRI, and/or CT) and metastatic imaging studies (CT chest, bone scan). Where the extent of the tumor raises the concern for visceral involvement, the necessary consultations with urologists, gynecologic oncologists, and/or colorectal surgeons to coordinate concurrent visceral resection is recommended.

Timing of surgery is usually about 4 weeks after finishing the last cycle of chemotherapy and similar timing after neoadjuvant radiation therapy to allow cell counts and overlying skin to recover. In patients who received neoadjuvant chemotherapy, absolute neutrophil count (ANC) and platelet count need to recover appropriately prior to surgery. Five hundred to 1000 cells/μL and 50,000 cells/μL are considered appropriate ANC and platelet levels, respectively [16]. If cell counts have not recovered in time for surgery, the patient may benefit from another cycle of chemotherapy prior to resection.

11.5 Types of Resection

Where limb salvage and a curative resection are the indicated, resection of the tumor with negative margins is the goal. Anatomic considerations determine the technical feasibility of performing this with preservation of vital structures.

In 1978, Enneking and Dunham [1] described pelvic resections based on the part of the innominate bone excised and surrounding soft tissue. This widely accepted classification of pelvic resections and its subsequent modifications are based on the bony anatomy of the innominate bone. This classification provides a framework for appreciating how the level of bony resection is accompanied by the requirement for dissection of certain soft tissue structures to permit sufficient mobilization of the bony segment for resection (Fig. 11.1).

A type I excision involves resection of the ilium and type IA included the buttock muscles and sciatic nerve, if necessary (Fig. 11.2). A type II excision is a periacetabular resection involving resection of the acetabulum while preserving the femoral head, whereas a type IIA resection

involves en bloc extra-articular resection of the hip joint, i.e., the acetabulum and femoral head. A type III excision is a pubic resection, and a type IIIA resection includes resection of the femoral neurovascular bundle and surrounding muscles, sparing the hip joint. Depending on the location and size of the tumor, these resections can be combined; e.g., a type II/III resection involves removing the acetabulum and ischium (Fig. 11.3). A hemipelvectomy would be classified as type I/II/III. Additions to this classification system include sacral and femoral head resections. Sacral resections are classified as a type IV resection. Pelvic resections that include the femoral head are designated H and subdivided into three types: (1) femoral head, (2) peritrochanteric, and (3) subtrochanteric [17]. Spinopelvic resections have also been classified into four types based on the how much of the sacrum is resected and if the sacral resections are combined with an external hemipelvectomy [18].

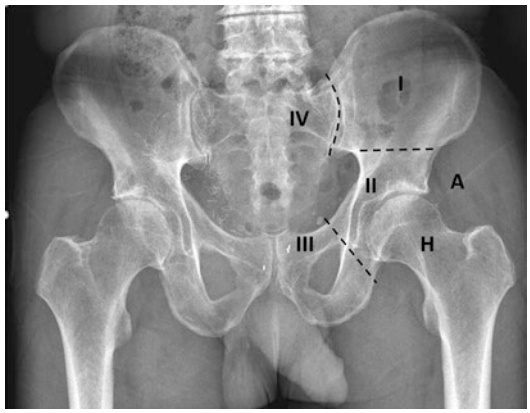

Fig. 11.1 Classification of pelvic resections

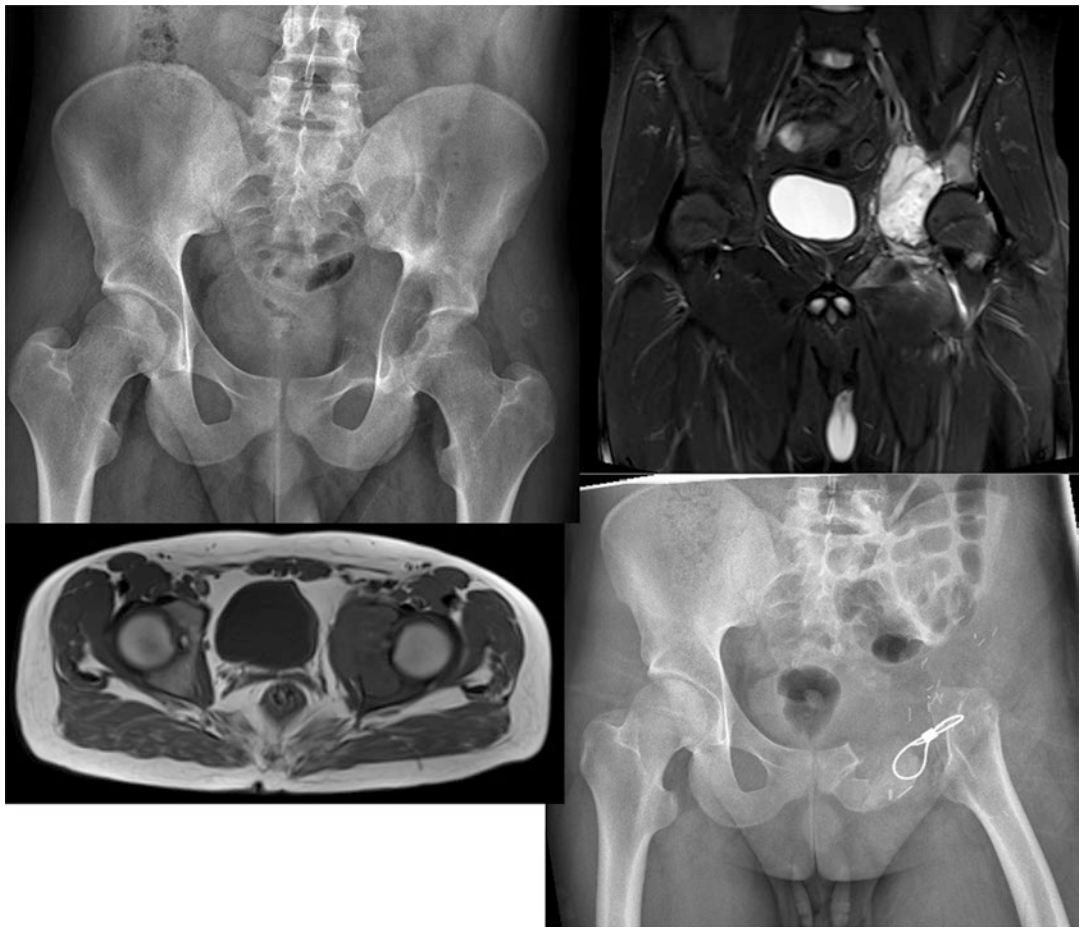

Fig. 11.2 Type I resection for Ewing's sarcoma with allograft reconstruction

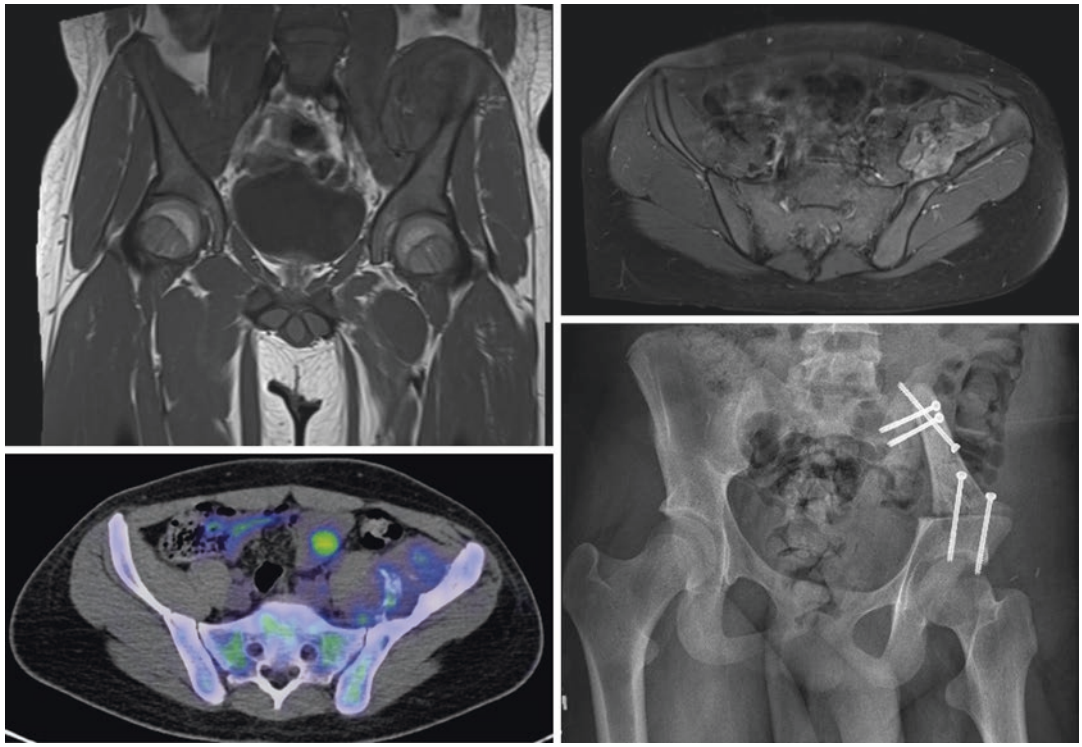

Fig. 11.3 Type II/III H1 resection without reconstruction performed

11.6 Anatomic Considerations

The anatomy of the pelvis is complex, and there are important visceral, neurovascular, and bone and soft tissue structures that are prone to tumor involvement or surgical injury and must be considered when performing surgical resections. The pelvic ring is stabilized by the pubic symphysis anteriorly and the sacroiliac joint, sacrospinous, and sacrotuberous ligaments posteriorly. The abdominal aorta bifurcates into the common iliac arteries usually at the level of the fourth lumbar vertebra. The internal and external iliac vessels bifurcate anterior to the sacroiliac joint. The internal iliac vessels exit the pelvis through the greater sciatic notch, terminating as the superior and inferior gluteal vessels. These vessels perfuse the gluteal muscles and preservation of the internal iliac artery, and its branches are important for maintaining their vascular supply. The external iliac artery exits the pelvis medial to the iliopsoas tendon and deep to the inguinal ligament to become the femoral artery. The ureters cross over at the bifurcation of the internal and external iliac vessels, traveling from lateral to medial to enter the bladder.

The gluteus medius and minimus originate from the outer table of the ilium and insert onto the greater trochanter. These muscles usually provide a margin for tumors arising from the ilium with soft tissue extension laterally. The iliacus originates from the iliac fossa and joins the psoas major to form the iliopsoas tendon that inserts on the lesser trochanter. Similarly, the iliacus usually provides a medial margin for tumors arising from the ilium with soft tissue extension medially. The adductor muscles, gracilis, pelvic floor muscles, and hamstrings originate from different areas of the pubis and ischium and may need to be released depending on the tumor's location. The femoral nerve can be found running between the iliacus and psoas major before exiting the pelvis with the femoral vessels. The sciatic nerve exits the greater sciatic notch anterior to the piriformis.

In male patients, the spermatic cord running in the inguinal canal is important to identify and protect. Other structures including the prostate, bladder, and corpus of the penis may either be involved or in close proximity to the tumor and should be protected. Pelvic resection can affect male sexual function by damage to the corpus origin or through neurologic damage. In female patients, the ovaries, uterus, and vagina may be involved or in close proximity to the tumor. The urethra runs just inferior to the pubic symphysis and must be retracted if a pubic symphyseal cut is required. The rectum may also be involved depending on the tumor's location. Any prior radiation therapy to the pelvic area increases fibrosis and adhesions and increases the risk to visceral damage.

11.7 Surgical Approaches

A utilitarian extended ilioinguinal approach is commonly used for pelvic resection surgery. It permits access to the pubic symphysis anteriorly, the sacroiliac joint and sacrum posteriorly, as well as the hip and proximal femur distally. Depending on the location of the tumor, modifications to this incision may be necessary. For type I resections, only the posterior limb of the extended ilioinguinal approach may be required. In type II resections, a lateral limb down the lateral thigh is required and can be achieved by curving the distal and medial end of the incision back toward the lateral aspect of the femur. Alternatives to this include a T-incision [19] and the modified Ollier transtrochanteric approach [20, 21]. If the resection is to remain extra-articular, the joint capsule is kept intact and the proximal femur is osteotomized at the appropriate level. If the resection is intra-articular, the capsule is circumferentially incised to allow femoral head dislocation. Type III resections may require extending the anterior limb over to the contralateral side of the pubic symphysis. Anterior, posterior, or combined approaches may be needed for type IV resections depending on location, size, and presacral soft tissue extension [16, 22–24]. With all resections, care must be taken not to violate the periosteum when dissecting and releasing muscles from the pelvis near the tumor. A more extensive discussion of approaches for pelvic resection is included in an earlier chapter.

11.8 External Hemipelvectomy/ Hindquarter Amputation

Limb salvage surgery for sarcoma may be precluded by extensive invasion of critical structures by tumor. The indications for an external hemipelvectomy or a hindquarter amputation are mostly related to how advanced a disease is locally and the extent to which adjacent neurovascular structures are invaded. Should obtaining clear surgical margins be deemed unfeasible with limb salvage surgery in a patient without systemic disease, hindquarter amputation should be considered. Neural structures at the level of the pelvis in particular do not lend themselves well to reconstruction, owing to the distance from the level of injury at the time of transection to the sensory and motor target end organs in the lower extremity. Loss of muscular control or loss of nerve function can be compensated for with the use of external orthoses and is not an absolute contraindication to limb salvage. Major artery reconstruction when indicated adds to the complexity of the surgery but with venous or endoprosthetic grafting can be associated with acceptable limb salvage rate [25]. When resection of major nerves is paired with an unstable skeleton, external hemipelvectomy should be considered as the resulting lower extremity may be insensate, weak, or flail and unstable. Living with such a limb may be more burdensome and disabling to a patient than living without it. The three structures that should be considering in the decision making are the periacetabular portion of the pelvis, the sciatic nerve, and the femoral nerve. Where two of these three structures require resection, external hemipelvectomy should be seriously considered [3, 16].

When an external hemipelvectomy is indicated, the two most common conventional approaches are the posterior flap hemipelvectomy or the anterior flap hemipelvectomy.

A posterior flap hemipelvectomy involves the following components: (1) Ilioinguinal approach to explore the retroperitoneal space and for release of the anterior abdominal wall muscles, mobilization, ligation, and transection of structures including the iliac vessels. (2) Perineal dissection involving a posterior extension of the medial end of the ilioinguinal approach toward the posterior aspect of the thigh and the region of the horizontal gluteal crease. This permits access to the pubic symphysis for their disarticulation or the medial ends of the pubic rami for their transection. (3) Raising the posterior flap which typically extends from the iliotibial band laterally, connecting to the ilioinguinal incision superiorly and coursing posteriorly along the horizontal gluteal fold. This flap may be a fasciocutaneous flap in the classical hemipelvectomy/hindquarter amputation where its perfusion is based of fasciocutaneous perforators from its base. However, where the gluteus maximus is not involved by disease, preservation of this muscle and the superior gluteal artery with it results in a more robust flap for closure. (4) Mobilization and division of the pelvic floor muscles to complete the detachment of the pelvis inferiorly. (5) Final mobilization and amputation of the hemipelvis. A classical hemipelvectomy involves disarticulation of the sacroiliac joint. In a modified hemipelvectomy, the innominate bone is osteotomized through the sciatic notch, while in an extended hemipelvectomy, the osteotomy is through the sacrum.

An anterior flap hemipelvectomy is indicated for the management of sarcoma when the disease extends posteriorly and precludes the preservation of the posterior flap. Prior surgery with contamination of the posterior flap by sarcoma is also an indication for this approach. Nononcologic indication for this procedure includes extensive sacral or trochanteric decubitus ulcers and osteomyelitis such as in paraplegic patients. The anterior flap is a myocutaneous flap that is perfused by the femoral artery and includes muscles of the anterior compartment of the thigh and the overlying skin. The good vascularity of this flap is regarded as an advantage, and the muscle bulk is useful in occluding dead space on wound closure. Dissection of the anterior flap involves medial and lateral incisions along the borders of the quadriceps muscles and raising the anterior compartments of the thigh as a flap off the femur. The superficial femoral artery is ligated distally as it courses from the anterior compartment through the adductor hiatus to the popliteal fossa. The adductor compartment may also be included if a larger flap is desired or if part of the anterior compartment of the thigh has to be removed.

11.9 Complications

Pelvic tumor resection surgery is complex and often prolonged and is associated with a significant risk of complications that patients should be duly counseled on. The mortality in the perioperative period is an important risk, and studies document rates of perioperative mortality from 0% to 10% [16, 26–30]. This illustrates that with good perioperative care, such surgeries can be performed safely. However, apart from the risk of mortality, the complication rate for pelvic tumor resection surgery is quite significant. Studies vary in their reported rates which range from 15% to 68% [26–29, 31–37]. Since complications occur at such a high rate, they should be anticipated and prevented when possible and dealt with as they occur.

These complications may be divided into the intraoperative complications, complications diagnosed in the early postoperative period, those diagnosed in the intermediate postoperative period, and late complications.

Intraoperative complications are related to the frequently prolonged nature of these surgeries, the risk of sudden blood loss during certain portions of the procedure, and inadvertent injury to pelvic viscera. The prolonged surgeries put patients at risk of pressure sores, deep vein thrombosis, and respiratory compromise. Patient with significant cardiovascular comorbidities should be closely monitored owing to the potential for significant blood loss throughout the course of the surgery as well as the risk of sudden brisk blood loss. The extensive nature of some pelvic tumors requires mobilization of pelvic viscera, and this puts patients at risk of injury to

these pelvic viscera. Identification of injury to pelvic viscera is crucial if these injuries are to be managed appropriately and to permit appropriate consultation with other surgical subspecialties if indicated to address these complications. The pelvic viscera or vessels are at particular risk of injury in type III resections [29]. Due to these risks, high-risk anesthesia teams are needed, and patients should be adequately resuscitated and arrangements should be made for adequate amounts of blood products to be available. Typically, patients are taken care of in monitored intensive care units postoperatively.

In the early postoperative period, wound complications are of significant concern and neurologic deficits resulting from surgery are often apparent. Flap necrosis, wound dehiscence, and soft tissue infections are common especially considering the large size of these wounds, the risk of bacterial translocation from pelvic viscera, and the challenges in maintaining the hygiene of portions of the wounds that are adjacent to the perineum. Increased rate of wound complications and flap necrosis have been associated with longer surgical times; the extent of resection and ligation of the common iliac artery is also associated with higher rates of flap necrosis [31]. Neurologic deficits are also a common complication as manipulation and mobilization of nerves are required to perform pelvic resection, while many of these deficits may improve with time others do not improve owing to the severity of the injury [29, 38]. Resections involving the ilium involve mobilizing the psoas muscle and femoral nerve, resection around the sciatic notch may involve significant manipulation of the sciatic nerve, and resection involving the sacrum frequently involves significant nerve root manipulation or sacrifice [39]. Reconstructive procedures may also result in an increase in the tension across the nerves and result in injury. Nerve injuries may also involve the contralateral side which may be related to surgical manipulation, prolonged surgery, or ischemia.

While superficial infections and wound dehiscence are often diagnosed in the early postoperative period, deep infections may only be diagnosed at a later stage or after failure of antibiotic therapy to manage what appears at first to be a superficial infection. Studies have also found an association of reconstruction with a higher rate of complications [33, 40, 41]. While antibiotic therapy may be adequate for treating some cases of soft tissue infection, severe superficial infections and deep infection frequently require repeat surgery. Deep infection in the setting of endoprosthetic or allograft reconstruction may also involve removal of the components and allograft used in the reconstruction. The development of wound complications such as infection or wound dehiscence may preclude the initiation or restarting of systemic cytotoxic chemotherapy. It is thus imperative that the indication for reconstruction be weighed against the increased risk of complications and the impact that this may have on a patient's subsequent systemic treatment.

Complications that are diagnosed late include abdominal hernia and complications related to failure of reconstructive procedures. Abdominal hernia is also a known complication owing to the need to detach the insertions of the abdominal wall musculature, and abdominal wall reconstruction may be indicated where there is a significant defect [31, 42, 43]. Prosthetic reconstructions may also fail after the early postoperative period, and these include periprosthetic fractures, implant fracture, dislocation, and loosening.

Lastly, there is the risk of local recurrence. Local recurrence rates vary significantly in published studies from 9% to 45%, while it is clear that intralesional margins result in poorer outcomes, several large studies have not shown differences in recurrence rates in marginal and wide resections [19, 28, 33, 34, 36, 44–48]. Where local recurrence or deep infection arises and limb preservation is not feasible, a secondary hindquarter amputation may be indicated, and studies report a rate of 8–12% of patients requiring this [34, 37, 49].

11.10 Conclusion

Pelvic resection for oncologic disease requires meticulous planning, multidisciplinary teams, and careful execution owing to the complex anatomy, challenging exposure, and often advanced nature of these tumors on presentation. The

understanding gained from decades of research on the topic has improved the safety and outcomes of this type of surgery.

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Innovative Techniques in Pelvic Reconstructions

12

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12.1 Introduction

Significant advancements have been made in pelvic reconstruction since the first documented pelvic resections were attempted at the turn of the twentieth century. The first documented nonfatal pelvic resection was for sarcoma performed in 1895 by Girard of Berne, and in 1935, Sir Gordon Gordon-Taylor of Britain called such resections as "one of the most colossal mutilations practiced on the human frame" [1, 2]. Since that time, the knowledge base of pelvic anatomy, advancements in technique and cross-sectional imaging, and the rapid sophistication of metallurgy and implant development have made this once morbid procedure now safe and effective.

Pelvic resections can be left flail or be reconstructed to maximize function. In order to achieve an adequate reconstruction, innovative techniques have been proposed with various use of autograft, allograft, and custom implants that span from the spine to the femur. Primary musculoskeletal tumors, metastatic lesions, trauma, and infection of the pelvis are among the indications for this relatively uncommon procedure. The scope of this discussion will focus on the indications, relevant anatomy, and innovations related to spinopelvic, sacral, bulk allograft, and proximal femoral reconstructions.

12.2 Indications

Pelvic resections including hemipelvectomies occur relatively rarely with rough estimates approximating 1 per one million persons annually [3]. Enneking and Dunham classified pelvic resections in relation to sarcoma of the innominate bone that failed treatment by medical means [4]. A variety of oncologic processes can indicate a pelvic resection and reconstruction. Pelvic primary bone tumors compose 15–20% of all primary bone tumors. Furthermore, chondrosarcoma, osteosarcoma, and Ewing's sarcoma compose 50–80% of all pelvic bone tumors [5, 6]. Ewing's sarcoma and osteosarcoma are most highly prevalent in the adolescent or young adult population, whereas chondrosarcoma most often presents in the fourth to seventh decades of life [5]. Chordoma, fibrosarcoma, Langerhans cells histiocytosis, aneurysmal bone cyst, giant cell tumor, and fibrous dysplasia can also necessitate pelvic resection although with less frequency than those oncologic process aforementioned. Metastatic disease to the pelvis can originate from the breast, lung, prostate, kidney, and thyroid; however, many of these lesions can be managed with radiation or chemotherapy with a minority of metastatic lesions indicating pelvic

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resection [7, 8]. Infection, complications from arthroplasty, and trauma can also require pelvic resection, albeit at a relatively lower frequency.

12.3 Classification of Pelvic Resection

Pelvic resections vary widely in location and size, and therefore, pelvic reconstructions can vary. To organize these pelvic resections and the necessity and types of reconstructions, it is helpful to understand the Enneking and Dunham classification of pelvic resections. The Enneking and Dunham classification is based on the specific anatomic locations of resection. In brief, resections of the ilium are Type 1, resections of the periacetabular region are Type 2, resections of the pubic rami or obturator rings are Type 3, and resections of the sacrum are Type 4, which have subtypes depending on resection of adjacent anatomy [4]. Reconstruction can be considered in resection types that disrupt the pelvic ring, although leaving a patient flail is often a viable alternative. For example, within Type 1 resections of the ilium, reconstruction may be favored if the pelvic ring is disrupted as in the case of complete ilium resection but not necessary in cases of partial resection with an intact pelvic ring. Extensive resections can disrupt the continuity of the pelvic ring and the ability of the pelvis to support the continuity between the lower extremity and axial skeleton.

The overarching goal for pelvic reconstruction is to provide pelvic girdle support for maximal function. The function of the pelvis is to provide continuity between the lower extremity and the axial skeleton to allow for locomotion in addition to protecting the pelvic contents and providing muscular attachments for the torso and lower extremities. Although pelvic resection without reconstruction should be considered, the functional outcomes of resection without reconstruction are often unacceptable to patient and surgeon in light of modern techniques and implant design. However, reconstruction must be considered carefully with a full assessment of the complication profile as reconstruction following a pelvic resection significantly increases the morbidity to the patient. Therefore, in principle, the indication for pelvic reconstruction is any patient who has undergone destabilizing pelvic resection for the aforementioned pathologies that is willing and able to withstand the extensive surgery and rehabilitation intended to maximize function.

12.4 Spinopelvic Reconstruction

Pelvic resections that extend to the sacrum or lumbar spine often require spinopelvic reconstruction to reestablish the relationship between the pelvic and axial skeleton. Depending on the size and extent of resection, the reconstruction construct can vary widely. Instrumentation to aid in these reconstructions also varies with the employment of pedicle screws, plates and screws, rods, allograft, autograft, etc., and there exists very limited literature and no standard of care with regard to these constructs. The following are illustrative cases and insights into spinopelvic reconstructions by the authors of this chapter.

12.4.1 Case 1

The patient is a 33-year-old male who initially presented with back and hip pain with subsequent biopsy-proven grade 3 chondroblastoma of the pelvis without metastatic disease. The patient underwent neoadjuvant chemotherapy with 90% tumor necrosis prior to referral to a tertiary orthopedic oncology service. Advanced cross-sectional imaging of the tumor revealed an expansive lesion about the left lower lumbar spine, sacrum, and ilium with posterior soft tissue extension (Fig. 12.1). The operation to resect and reconstruct the spinopelvic defect was undertaken in two stages.

12.4.1.1 Stage 1

First, the patient was positioned prone and a posterior longitudinal midline incision with extension over the left gluteus was utilized to access the lumbar spine and sacrum. Pedicle screws were placed at the L2/L3 levels bilaterally and L4/L5 levels on the right; additionally, rods were placed

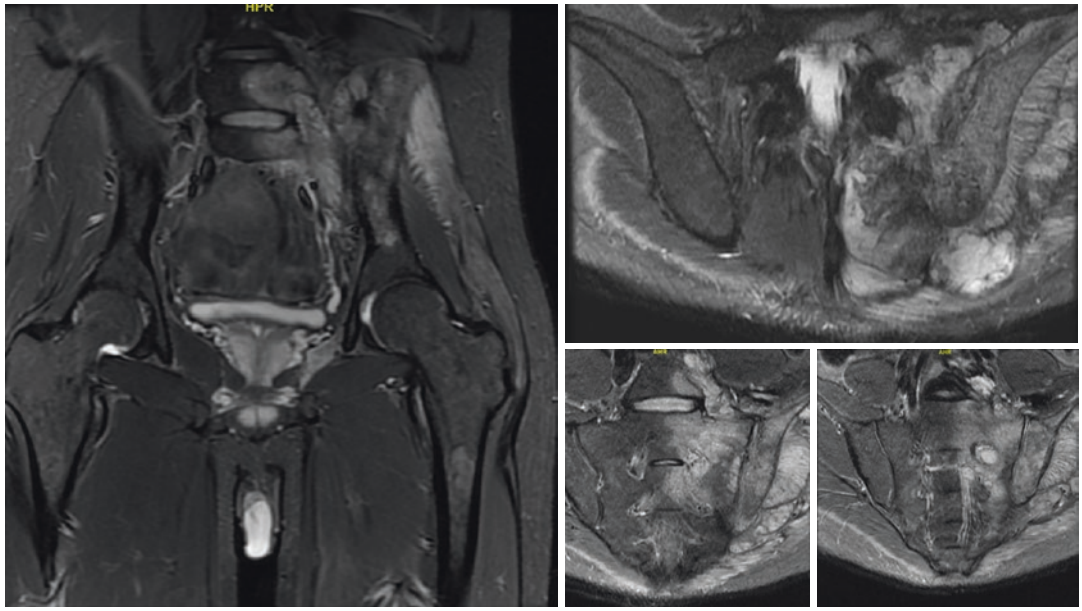

Fig. 12.1 Grade 3 chondroblastoma of the pelvis in a 33-year-old male. Magnetic resonance cross-sectional imaging show expansive lesion involving the lumbar spine, sacrum, and ilium

to enable spinopelvic fixation in the second stage of the surgery. Laminectomy and ligation of the nerve roots on the left lower lumbar and sacral levels were completed as well as posterior osteotomy cuts through margin negative pelvis with a diamond tip burred under navigation guidance. Dissection of the tumor to achieve negative margins were carried out from the posterior incision.

12.4.1.2 Stage 2

In stage 2 of the operation performed 3 days later, the patient was positioned supine. Vascular surgery performed an anterior approach to the lower lumbar spine and sacrum with mobilization of the great vessels which required ligation of the left internal iliac artery and left common iliac vein. The osteotomy cuts were completed from the anterior approach under navigation guidance and the tumor, hemisacrum, and hemi-ilium were resected in an en bloc fashion while preserving acetabulum. Plastic surgery then performed a vascularized fibular autograft to span the lumbar spine to acetabulum adjacent to a humeral shaft allograft for structural support. Multiple pedicle screws into the remaining pelvis were placed and connected to the rods placed during the first stage to further reconstruct the resected hemipelvis (Fig. 12.2). A vastus lateralis rotational muscle flap was mobilized anteriorly to provide coverage over the hardware within the pelvis. Toe-touch weight-bearing restric-

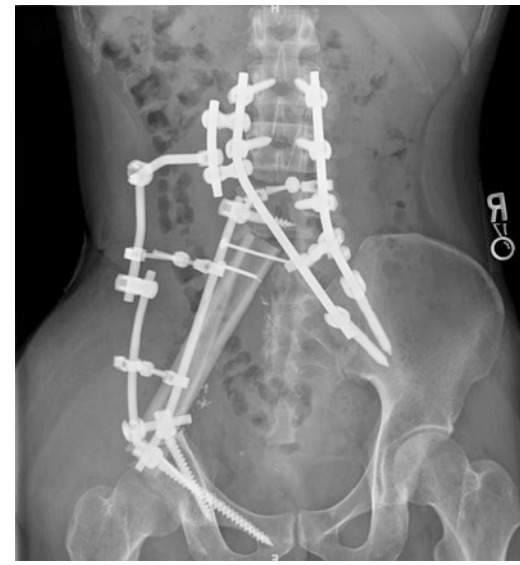

Fig. 12.2 Same patient. Postoperative radiograph shows the spinopelvic reconstruction with multiple pedicled screws and rods associated with vascularized fibular autograft and a humeral shaft allograft for bone support

tions were in place for 8 weeks. At last follow-up, the patient was ambulating independently with a walker within his house. At 6 months follow-up, the patient has remained recurrence-free.

12.5 Sacral Reconstruction

The indications for sacral reconstruction most frequently include chordoma, chondrosarcoma, giant cell tumor, and osteosarcoma [9]. This relatively rare reconstruction can be addressed with various techniques, but the principle of maintaining pelvic ring continuity remains paramount. The use of autograft, allograft, titanium bars, plates, and custom 3-D printed implants have all been documented in the literature [9–12]. Sacrectomy and reconstruction carry a significant morbidity with a neurologic deficit expected in most reconstructions in addition to a high infection and wound-healing complication rate. Furthermore, nerve ligation at the sacral level can affect bladder, bowel, and sexual function, and it is our preference to preserve at least one side of the sacrum in an attempt to maximize function. Based on the current literature, there is no consensus on the best reconstruction method, but anterior spinal column fixation in conjunction with posterior instrumentation may be required to minimize the risk of hardware failure.

12.5.1 Case 2

The patient is a 32-year-old male with right leg sciatica with calf atrophy for several months who initially attempted nonoperative measures. MRI revealed a large tumoral lesion centered about the R sacral ala extending into the S1 and S2 neural foramen and into the pelvis displacing the bladder (Fig. 12.3). Subsequent biopsy demonstrated grade 1 chondrosarcoma, and staging revealed no metastatic disease.

12.5.1.1 Stage 1

The patient was positioned supine for an anterior approach to the sacrum by vascular surgery. The great vessels were mobilized, and the right internal iliac artery and vein were ligated to gain access to the sacrum. The tumor was dissected away from the bowel and peritoneum. Utilizing navigation, osteotomies with a diamond-tipped burr were performed in a longitudinal fashion along the sacrum and L5–S1 disk on the right side with preservation of the left-sided hemisacrum. The left hemisacrum was preserved in order to maximize the patient's postoperative bowel, bladder, and sexual function.

12.5.1.2 Stage 2

Two days later, the patient was positioned prone and a midline lumbar incision with lateral exten-

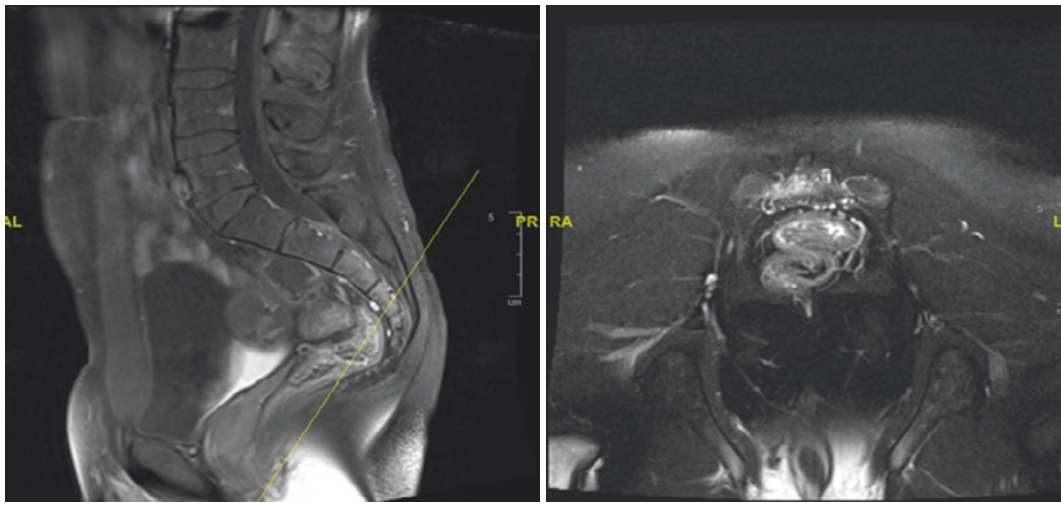

Fig. 12.3 Grade 1 chondrosarcoma of the sacrum in a 32-year-old male. Sagittal and axial MRI show the tumor extension into the proximal sacrum (S1 and S2 neural foramen)

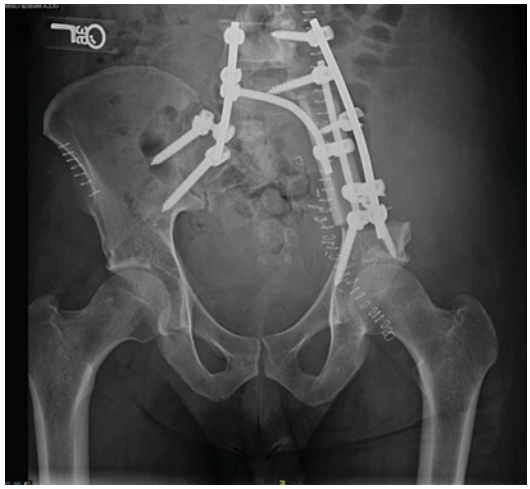

Fig. 12.4 Same patient. Postoperative radiograph shows the resection performed with navigation (longitudinal split of the sacrum and acetabular sparing) and reconstruction with pedicled screws, rods, and allograft

sion over the gluteus maximus toward the midportion of the right thigh was undertaken. The sciatic nerve was dissected proximally to the sciatic notch. Laminectomy of the lumbosacral junction was completed, and utilizing navigation, posterior osteotomy to join the anterior osteotomy made in stage 1 was completed. The sacrum was split longitudinally and then to the right at the L5–S1 disk space. The ilium was freed of muscular attachments on the left, and a supraacetabular osteotomy was completed. The left hemisacrum, left iliac wing, and tumor were then resected en bloc. A humeral shaft allograft was placed from the low lumbar spine to the supraacetabular pelvis. Pedicle screws were placed into the lumbar spine, left ilium, and supraacetabular pelvis on the right side. Multiple rods and connecters were utilized to reconstruct the pelvic ring (Fig. 12.4). The patient is ambulatory with preserved bowel and bowel function at 20-month follow-up.

12.5.2 Case 3

The patient is a 50-year-old male who presented with low back pain and difficulty with bowel movements. Advanced imaging revealed large expansile tumor lesion emanating from the left lumbosacral junction which extended both posterior and anterior to the ilium (Fig. 12.5). Subsequent biopsy revealed a grade 1 chondrosarcoma.

12.5.2.1 Operation

A midline longitudinal incision through the lumbar spine which extended over the gluteus to the left lateral thigh was utilized to access the lumbar spine and tumor. The tumor was carefully dissected away from the lamina of the low lumbar spine, and the nerve roots were meticulously dissected away from the tumor. Laminectomy was completed in the lower lumbar spine and sacrum. Utilizing navigation, a diamond tip burr was utilized for the near total sacrectomy. Posteriorly based iliac osteotomies were completed, and the tumor was dissected from the pelvic contents and resected en bloc. A humeral allograft was utilized to span the defect from right sacral remnant to left iliac wing to reconstruct the pelvic ring. Pedicle screws, iliac bolts, and multiple rods were utilized to reconstruct and support the spinopelvic junction (Fig. 12.6). The patient has partial paresis in the sciatic distribution of the left lower extremity and continues his rehabilitation 9-month follow-up.

12.6 Pelvic Allograft Reconstruction

Wide resection of primary malignant tumors of the pelvis coupled with limb-sparing surgery when possible is the preferred treatment of our group. Although there is no unanimous opinion regarding the ideal reconstruction method, pelvic allograft is a viable option for reconstruction. An advantage of allograft reconstruction is that it can be shaped and sized in order to match the postresection defect. Allograft use also provides the advantage of patient bone stock preservation [13]. Furthermore, use of an allograft may allow the patient to avoid a flail hip or arthrodesis. Pelvic allograft accommodates anatomic reconstruction of pelvic bony architecture as well as allows for multiple hip preservation options [14]. Patients report a high rate of functionality with

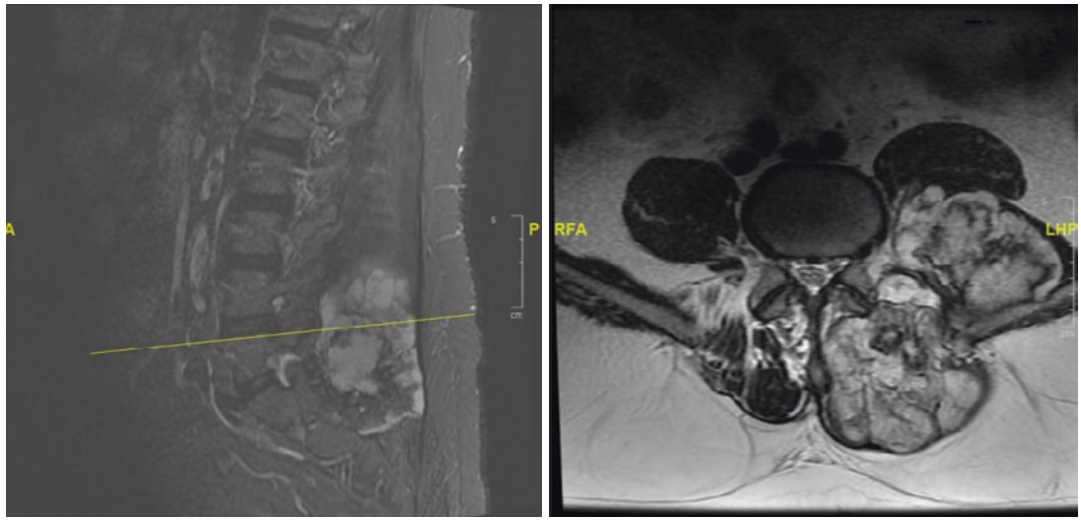

Fig. 12.5 Grade 1 chondrosarcoma of the sacrum with posterior soft tissue extension in a 50-year-old male. Sagittal and axial MRI show the typical characteristics and growth pattern of the tumor

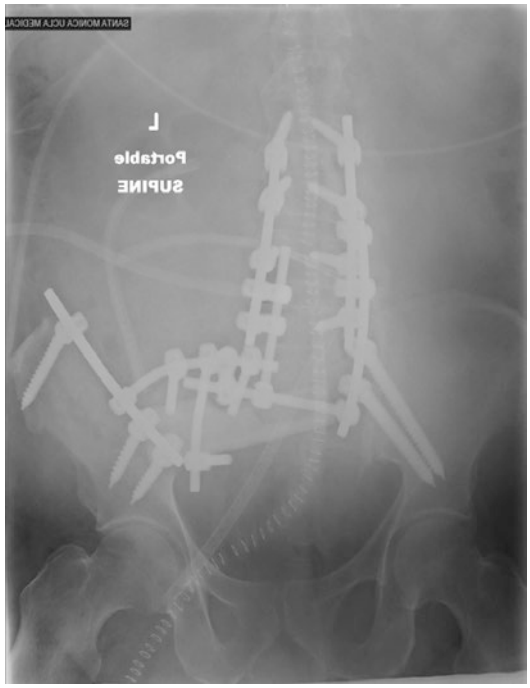

Fig. 12.6 Same patient. En bloc tumor resection required total sacrectomy extended to the ilium. Postoperative radiograph shows pedicle screws, iliac bolts, and multiple rods that were utilized to reconstruct and support the spinopelvic junction

regard to pain and ambulation [14, 15]. Children and adolescents achieve substantially increased functionality compared to adults [14].

Yet, as is the case with most pelvis reconstructions options, pelvic allograft reconstructions are associated with significant rates of infection and mechanical failure. Infection rates who underwent allograft reconstruction after pelvic resection or internal hemipelvectomy range from 12.5% to 20%, respectively [14, 16]. Given the high baseline risk of infection, history of persistent infection or immunosuppressed state should preclude use of allograft [17]. Sciatic and/or femoral nerve palsies can occur in as many as 25% of patients; however, this is most common in the setting of periacetabular resections. Most of these palsies are complete and partially resolve [14]. Hip instability is a known complication of allograft reconstruction. Patients must be followed for fracture and nonunion in the setting of allograft use or irradiated bone. In a series of 24 patients who underwent pelvic allograft reconstruction after tumor resection, 12% of the cohort developed nonunion. Two of the three nonunions in the series occurred at the site of fixation of the allograft to the ilium. Rates of allograft fracture have been reported to range between 0% and 21% [14, 18, 19]. Due to the many possible complications that the patient must be monitored for long-term, the social situation of the patient must be considered before opting for allograft reconstruction.

12.6.1 Case 4

The patient is a 57-year-old male with history of right pelvic liposarcoma status post chemoradiation, resection, and right total hip arthroplasty presenting with worsening right hip pain and difficulty ambulating. Physical exam was notable for mild tenderness to palpation over the right iliac wing.

MRI of the right hip demonstrated marrow replacement destruction of the iliac crest without associated soft tissue mass and pathologic fracture of the right ilium, suggestive of osteoradionecrosis. Radiation-induced sarcoma was suspected given the patient's history of radiation in the area. CT of the abdomen and pelvis demonstrated diffuse sclerotic appearance of the right iliac wing concerning for neoplastic infiltration (Fig. 12.7). Furthermore, PET scan revealed avidity in the right iliac wing. Subsequent CT-guided biopsy was diagnostic for radiation-associated sarcoma. The patient was scheduled to undergo resection of the pelvic sarcoma in a staged manner.

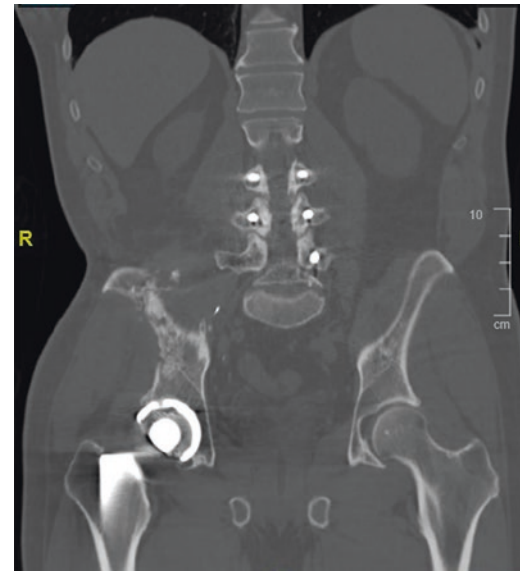

Fig. 12.7 Radiation-induced sarcoma of the right hemipelvis in a 57-year-old male, with a total hip arthroplasty

12.6.1.1 Stage 1

In the first stage of this staged resection, the patient was positioned prone and a posterior incision was made longitudinally in line with the lumbar spine; then, dissection was carried out to the right toward the right buttock and sciatic nerve. With guidance from intraoperative navigation, an osteotomy was made just medial to the right sacroiliac joint. Pedicle screws were placed at L3 and L4 as well as two large iliac bolts in anticipation of the spinopelvic reconstruction. An L5 osteotomy was performed. Further bone wax was placed in the defect, and rods were used to span the defect, positioned strategically for subsequent anterior reconstruction (Fig. 12.8).

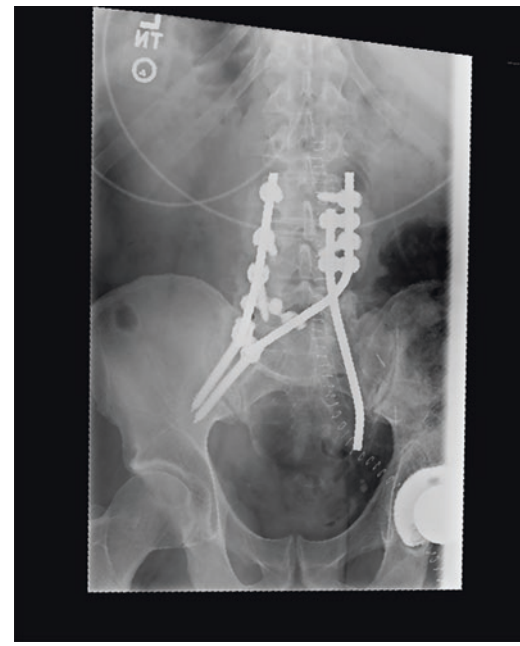

Fig. 12.8 Same patient. First surgical stage with posterior approach. Pedicle screws were placed at L3 and L4 as well as two large iliac bolts in anticipation of the spinopelvic reconstruction

12.6.1.2 Stage 2

Five days following stage 1, the patient underwent the second stage. The patient was positioned in lateral decubitus, and a curvilinear incision was made extending from the posterior superior iliac spine toward the anterior iliac spine and toward the knee. The tumor was situated within the ilium extending out from the inner and outer tables of the pelvis. Tissue along the iliac crest and the hip abductors was detached with the mass.

Using intraoperative navigation, pelvic osteotomies were performed. The osteotomy was created from the superior ramus and then through the inferior ramus toward the ischium, distal to the prior acetabular cup from the prior hip reconstruction. Further dissection was performed proximally along the anterior sacrum, and an osteotomy was completed in line with the prior posterior osteotomy in the first stage. The tumor was resected en bloc and sent to pathology for analysis. Negative margins were obtained.

A pelvic allograft was fashioned to fit the defect from the right sacroiliac joint to the distal right ischium. The plastic surgery service had concurrently harvested a free vascularized fibular graft with its associated vascular pedicle. The graft was fit and placed on the inner aspect of the allograft, held in place with screws and abutting the pelvic graft. Finally, the hip capsule was entered, and the proximal femoral component was removed. Proximal femoral replacement was performed with a bipolar prosthesis into the allograft. The reconstruction defects were covered with a vastus lateralis flap. Final reconstruction is shown in Fig. 12.9.

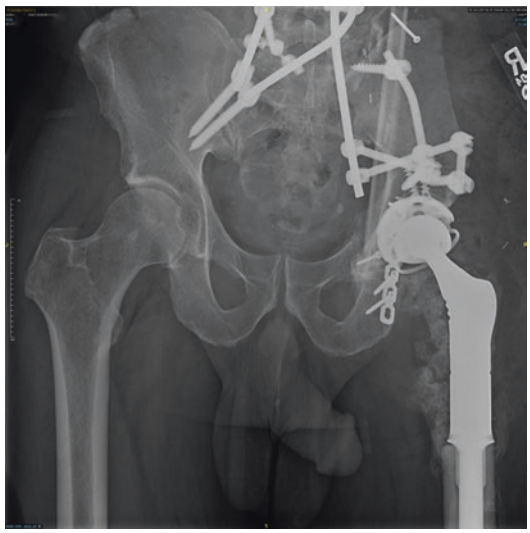

Fig. 12.9 Same patient. Final reconstruction using a pelvic allograft

12.7 Proximal Femoral Reconstruction

Wide resection of a tumor in the periacetabular region requires en bloc resection of proximal femur with subsequent complex reconstruction. Periacetabular resection without reconstruction will likely result in instability. Reconstruction can comprise of allograft arthrodesis, intercalary allografts, endoprostheses, and allograft–endoprosthesis composites. Although allograft techniques have an advantage of possible bony host-to-allograft incorporation, they also come with the significant risks of nonunion, fracture, and infection. Endoprostheses, on the other hand, are technically simpler reconstructions and provided the added benefit of shorter time to weight-bearing [20–23]. While endoprostheses share some of disadvantages of allografts, they also possess the risk of instability [13].

In a series of 137 patients who underwent proximal femoral reconstruction, a difference in outcomes was found to exist between osteoarticular allografts and all other allograft reconstructions. Osteoarticular allografts were significantly less likely to have a positive result than allograft–endoprosthesis composites. Development of osteoarthritis in patients with osteoarticular allografts led to a 39% rate of total joint replacement [13]. Other studies in other anatomic sites confirm that patients with allograft-endoprosthetic reconstruction have superior functional results compared to osteoarticular allografts [24–26]. Nonetheless, infection remains a serious complication regardless of reconstruction type. Wide resection, multiple surgeries, avascular allograft, and neoadjuvant chemoradiation all lead to a high baseline likelihood of infection in these oncologic surgeries [13]. Massive alloprosthetic reconstruction using allograft bone combined with arthroplasty may preserve limb length and maximize function, particularly in young patients [18, 27].

12.7.1 Case 5

The patient is a 37-year-old male with prior diagnosis of right pelvis undifferentiated pleomorphic sarcoma status post-neoadjuvant chemoradiation

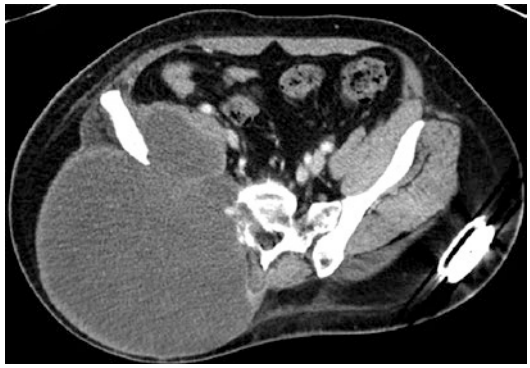

Fig. 12.10 Huge sarcoma of the hemipelvis in a 37-year-old male. Axial CT scan shows the aggressiveness of the tumor with soft tissue extension within the pelvis and in the posterior area

with worsening right hip pain and right lower extremity weakness with symptoms consistent with sciatica. The patient was found to have a large fungating soft tissue mass consistent with known sarcoma.

CT of the pelvis demonstrated a large soft tissue mass centered about and infiltrating the R iliac crest, with extension across the right sacroiliac joint and into the sacrum. The lesion involves the right S2, S3, and S4 neural foramina. Within the pelvis, it abuts the posterior sciatic nerve before it enters the sciatic foramen (Fig. 12.10). The patient was scheduled for staged resection of the pelvic mass.

12.7.1.1 Stage 1

The patient was positioned prone, and a midline incision was made from the mid-lumbar spine to the coccyx. The incision was ellipsed over the right soft tissue mass. Pedicle screws were placed bilaterally at L4 and L5. Decompression was performed from L4/5 to the distal sacrum and coccyx.

Using intraoperative navigation, an osteotomy was carried through the distal sacrum toward the left side. The piriformis was identified, and blunt dissection was performed over the anterior aspect of the sacrum. Further dissection was carried out around the mass on the right side through the gluteus maximus muscle primarily. One dural defect was noted in an area of scar tissue adherent to the dura and concerning for tumor. All frozen sections obtained were negative for tumor, including paraspinal musculature margins. The mass was kept intact without violating the capsule. The right-sided sacral nerve roots were ligated with silk ligatures and sacrificed. A construct was placed connecting the left L4 and L5 pedicle screws to the left iliac bolts. This was connected to the right L4 and L5 pedicle screws via cross connectors. Closure was performed, and the patient was transferred to the intensive care unit postoperatively.

12.7.1.2 Stage 2

Two days after the first stage, the patient returned to the operating room for the second stage. A curvilinear incision was made from the mid-back extending along the iliac crest to the distal thigh. A second limb of the incision provided anterior exposure of the distal vastus lateralis tendon, which was cut distally. The dissection was carried from distally to proximally. This was difficult to perform due to scar tissue from prior radiation therapy. The tumor was adherent to the sciatic nerve, and it was thus difficult to obtain a wide margin in this area. Nonetheless, margins were negative for tumor in frozen sections.

After completion of soft tissue dissection around the tumor, osteotomies were performed with the aid of intraoperative navigation. The femoral neck was cut with an oscillating saw in order to better expose the acetabulum prior to its osteotomy. An osteotomy then was made through the acetabulum to partially reflect the tumor mass in order to better expose the sciatic nerve and tumor pseudocapsule interface. Further dissection was carried out releasing further in the proximal sacrum as well as distal L5. The prior osteotomy was connected through where the sacral osteotomy was performed in stage 1. The tumor was removed en bloc.

After the wound was irrigated thoroughly, attention was turned to reconstruction. An allograft pelvis was sized to fit into the large osseous defect. The pelvic graft was anchored in place with pedicle screws and wedged into the remaining acetabulum. With the aid of intraoperative navigation, the graft was reamed sequentially in order to reconstruct the acetabulum. A

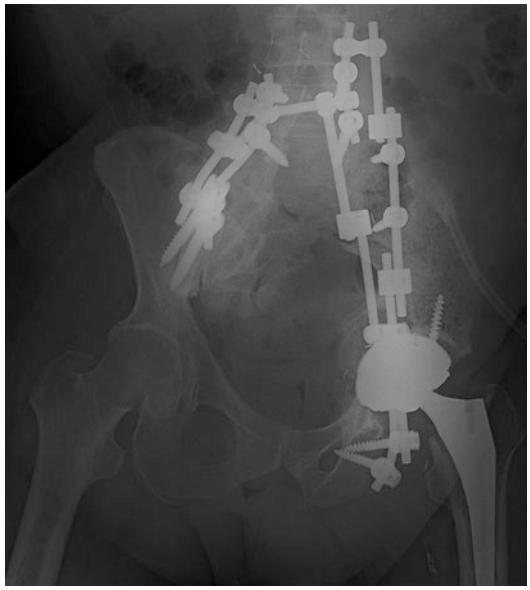

Fig. 12.11 Same patient. Final reconstruction after en bloc resection

hemispherical shell cup was inserted and anchored with screws into both native and allograft pelvis. The proximal femur was broached and reamed. A modular mobile bearing construct was employed. A constraining liner was not chosen since anchorage into the pelvis would be more than 50% allograft bone. Further, screws were placed into the allograft and host bone. The sacral hardware was joined to the construct. An A-like frame was constructed to allow for good stability. Final structural reconstruction is shown in Fig. 12.11.

With the plastic surgery service, soft tissue reconstruction was performed using a pedicled vastus lateralis flap. A lateral hamstring flap was also reflected into the defect, and the defect was closed completely. The patient was transferred postoperatively to the intensive care unit.

12.8 Intraoperative Navigation

One of the greatest challenges when resecting malignant sacropelvic tumors is achieving negative tumor margins. The complex anatomy of this region makes adequate resection difficult. Furthermore, neoadjuvant radiation may make defining soft tissue planes even more challenging due to distortion of normal anatomy by scar. Local recurrence rates are alarmingly common in cases of marginal resection and nearly 100% after intralesional resection; it is thus of the utmost importance to achieve negative margins. Standard resection techniques have been found to result in intralesional resection rate of 29% [28]. Even the most experienced surgeons have difficulty replicating a proposed osteotomy more than 50% of the time in a sawbones model [29].

Navigation-assisted resection may be useful in this situation, although it may add time and cost to an already complex and expensive procedure. Computer-aided navigation has become increasingly used in musculoskeletal tumor surgery [30, 31]. Navigation technology has been used with success in other surgical disciplines including neurosurgery, urology, spinal surgery, otolaryngology, orthopedic trauma, and arthroplasty [32]. Navigation requires preoperative imaging that is then integrated by software to help develop a preoperative plan. CT provides bony detail, MRI provides soft tissue detail, and PET-CT provides excellent discrimination between tumor and nontumor tissue. Intraoperative navigation is based on overlying the preoperative imaging onto fixed anatomic landmarks (e.g., anterior superior iliac spine) to provide proposed bone cuts.

In a series of 24 patients with primary tumors of the pelvis or sacrum, computer-aided navigation resulted in negative bony margins in all patients and negative soft tissue margins in 91% of cases [33]. Similarly, in a series of 31 patients with pelvic tumors, there was a reduction in intralesional resection rates from 29% to 8.7% using intraoperative navigation [34].

12.9 Conclusion

With advances in our understanding of anatomy, imaging, surgical technique, and implant development, pelvic reconstruction in the setting of oncologic resection has evolved from a morbid procedure to one that is relatively safe and effective. The Enneking and Dunham classification organizes the breadth of pelvic resections into

specific anatomic locations of resection; reconstruction may be favored in the setting of pelvic ring disruption. The goal of pelvic reconstruction is to provide continuity between the lower extremity and the axial skeleton in order to maximize function.

Spinopelvic reconstruction is often required in the setting of pelvic resections that extend to the sacrum or lumbar spine. Sacral reconstruction is relatively rare and generally performed after sacrectomy for chordoma, chondrosarcoma, giant cell tumor, or osteosarcoma. Wide resection of a tumor in the periacetabular region requires en bloc resection of proximal femur with subsequent complex proximal femoral resection. Bony reconstruction can be performed with pelvic allograft or autograft with employment of pedicle screws, plates, screws, and/or rods. Intraoperative navigation technology can be used to provide real-time intraoperative feedback with increased likelihood of negative bony margins. Finally, soft tissue coverage of the resulting spinopelvic defect is of the utmost importance to avoid wound-related complications. Rotational flaps such as the pedicled vastus lateralis flap may provide adequate coverage.

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Navigation in Pelvic Surgery

13

David M. Joyce

13.1 Introduction

Computer navigation for bony pelvic oncology cases was introduced as a guidance tool in the early 2000s [1, 2]. Prior to this, the only option for intraoperative guidance in pelvic bone resection of tumors was 2-dimensional (2D) fluoroscopy typically used in fracture fixation cases. This limited options for resection types and affected reconstructive options. Navigation began its incorporation into oncology because those with experience in pelvic oncologic surgery felt a frustration with the results of the standard resection used for surgery [3–10]. Malignant tumors in the pelvis that involve pelvic bone often have large soft tissue components that compromise visualization [11]. Computer navigation was specifically introduced in the pelvis to help in identifying the complete complex 3-dimensional extent of the malignant neoplasm involving both soft tissue and bone and safely assist in executing a local wide excision [12]. Navigation has improved the visualization of the surgical field through a virtual 3D reconstruction allowing for precise osteotomy resection levels [11, 13]. This section is dedicated to advanced imaging (computed tomography (CT) and magnetic resonance imaging (MRI)) computer navigation of the bony pelvis in pelvic oncology.

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13.2 Why Pelvic Computer Navigation

Prior to computer navigation the thought of limb salvage, maintained function and decreased morbidity were almost impossible without sacrificing one of the former to obtain adequate surgical margins [14]. The complex 3-dimensional nature of the pelvis often prevents direct line of sight, with no way to place a retractor to prevent inadvertent tumor violation. The difficulty is only magnified by structural alterations caused by tumor, neoadjuvant treatment, and rarity contributing to lack of experience on part of surgeon in conventional pelvic surgery which led to larger bone resections with surgeons opting for the more morbid procedure out of the need to obtain the best oncologic margins and hence the best oncologic outcome with little concern for function [14]. Looking back at pelvic oncology experience involving sarcoma resections in the pelvis, there are higher positive margin rates and higher local recurrence rates compared to sarcomas in the extremities confirming the difficulty in obtaining negative margins in pelvic sarcoma surgery [2, 15] which is partly due to inherent complexity of the pelvis. The pelvic complexity and added inaccuracy with free-hand cuts have led to positive surgical margins in the majority of cases, with local recurrence approaching rates of 70–80% when using conventional methods involving fluoroscopy, visual, and tactile references [3–6, 11, 16–18]. In practice, surgeons performing conventional pelvic surgery have to plan at least a 2 cm margins in order to be assured no tumor viola-

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tion [8]. The purpose of computer navigation in the musculoskeletal oncology setting is to plan and assist in executing a local wide excision, to reduce and eliminate the rate of intralesional resection with preservation of vital structures as well as mechanical structures. Navigation allows for accurate identification of the local bony anatomy and define the extent of the tumor which helps the surgeon better understand the relationship between the tumor and native structures in a distorted environment [19–23]. This technology aids a surgeon by identifying critical structures, improve margins, and preserve structure and theoretically function [24]. It allows for real-time visual feedback that can be seen on a monitor intraoperatively and augments and enhances the surgeon's tactile perception of the surgical field. Primary pelvic tumors often have an intraosseous component making it nearly impossible to appreciate extent of tumor infiltration intraoperatively [23]. In conventional pelvic resection, infiltration of the tumor within the bone marrow cannot be seen intraoperatively and requires a reliance of the surgeon's interpretation of preoperative imaging on MRI and CT and then properly correlates to bony landmarks intraoperatively on the patient's pelvis during surgery. This relationship between what is seen on preoperative imaging and what is identified in the patient can lead to errors that translate into positive margins in the noncomputer-assisted surgery. Navigation allows a surgeon to appreciate the extent of the tumor infiltration in the bone on the computer navigation monitor and know that their tool is correctly located on the planned resection plane avoiding tumor. One of the main goals of computer navigation is to help eliminate positive margins hopefully leading to more disease-free patients. The other benefit the navigation is the accuracy of the cuts because with accurate 3D navigation you now have a very precise cut that allows the surgeon to take advantage of precision 3D printing (Fig. 13.1) now being used in both models and custom 3D printed anatomic reconstructions for use in the pelvis (Fig. 13.2). Improved accuracy through this modality is one of the primary benefits and the

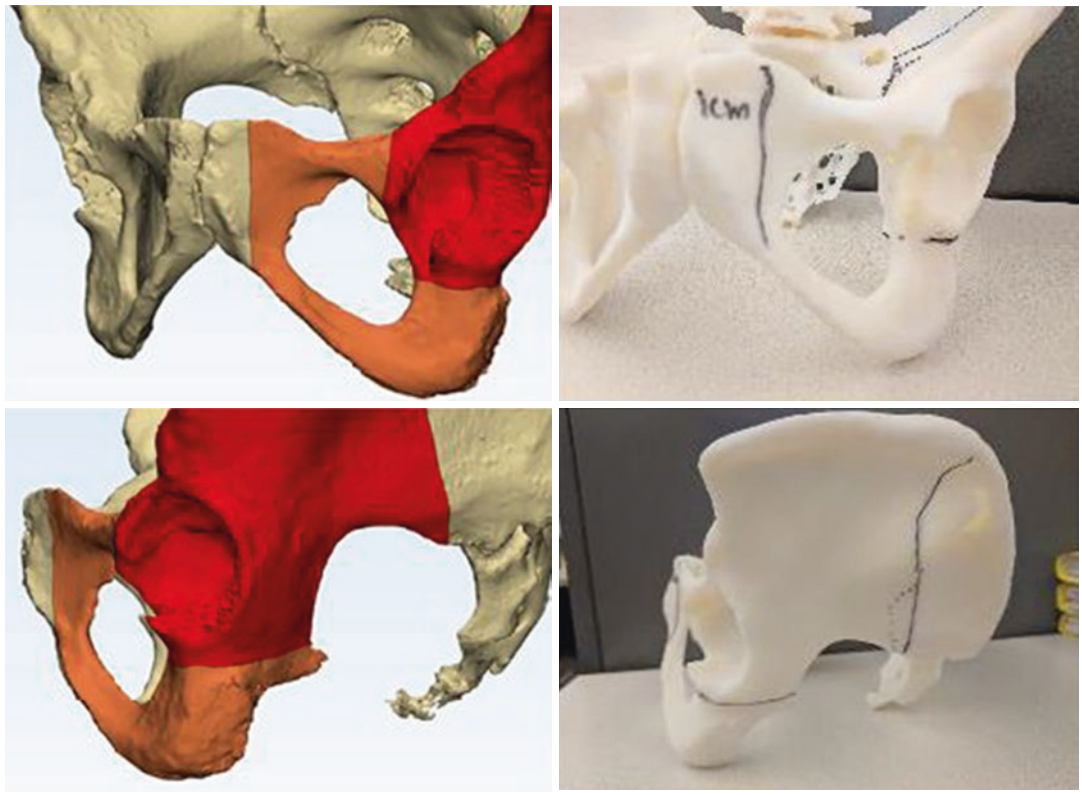

Fig. 13.1 3D virtual printing used for planning after resection lines are placed on 3D printed model

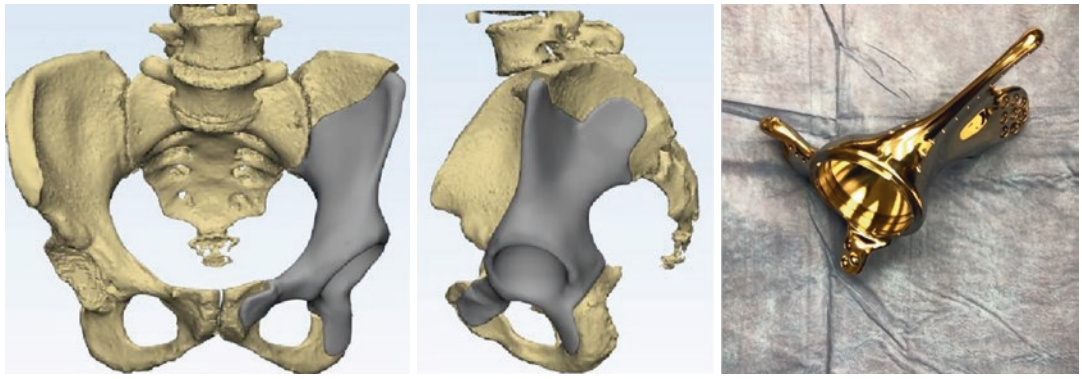

Fig. 13.2 Custom 3D printed anatomic reconstruction

ability to get closer to the tumor if needed to save critical structures without compromising margins. While pelvic oncology surgery first principle is to remove all the tumor with a negative margin, using computer navigation one can remove less normal tissue which aids in reconstruction without increasing the risk of inadvertent tumor violation [21].

13.3 History of Navigation

While computer navigational surgery in pelvic oncology is relatively new, computer-assisted surgery has been used in other surgical fields such as neurosurgery for cranial tumor resections and biopsies for at least the last 25 years [25, 26]. Several different computer navigation systems exist in the commercial world consisting of imageless navigation, fluoroscopic navigation, and advanced imaging navigation [27]. Adoption of computer navigation in reconstructive surgery was born out of a need for more accurate component placement and most navigation was developed for spine, trauma or hip and knee surgery for accuracy of hardware placement within 1 mm of the desired location [1, 26, 28–33]. In 1997, it was described in use for periacetabular osteotomies and thus may have served as the foundation for using it in pelvic oncology [26, 33]. Possibly, the first use of computer-navigated chisels for a pelvic tumor was reported in 2004 to perform an osteotomy for a sacral tumor [2].

Fig. 13.3 Optical Tracking System that can be moved around to improve "line of sight"

13.4 Computer Navigation Process Overview

There are required components to computer navigation in the setting of the pelvis [22, 34] including an intraoperative computer platform loaded with computer navigation software and tracking system. A patient's tracker or dynamic reference base (DRB), which can either consist of optical trackers (consisting of three OCD cameras (Figs. 13.3 and 13.4)) or electromagnetic trackers can track the instruments relative to the patient in space [35–39]. An intraoperative registration process in which the patient's preoperative imaging and patient's intraoperative position is matched and creates a referencing system that allows the patient and tools to be tracked in space with respect to each other. The registration

Fig. 13.4 Patient Tracker or Dynamic Reference Base (DRB) attached to iliac wing and covered with sterile bag

process can involve intraoperative CT, 3D fluoroscopic, imageless, kinematic, bone morphing, or "pair matching" with bony landmarks or fiducials, surface mapping, and ultrasound referencing [27, 40, 41]. Navigation of the pelvis cannot be performed with imageless referencing because this mode uses the pelvic plane and bony landmarks for reference, and soft tissue on the pelvis and tumor render this method fairly inaccurate [42–44] requiring all pelvic oncology navigation be based on advanced imaging such as MRI and CT [34].

13.5 Preoperative Planning

The use of computer navigation starts with obtaining advanced 3D imaging of the pelvis and tumor in question. At a minimum, a CT scan of the affected bone must be obtained with 0.5–2 mm cuts, continuous with no overlap that covers the entire compartment, for the pelvis that includes both innominate bones and sacrum [2, 20, 22, 23, 45]. Understanding how best to adequately identify appropriate margins on imaging will dictate which imaging is needed preoperatively. CT is ideal for cortical bone resection planning, but MRI is essential to determine marrow infiltration and true bony involvement [46, 47] and provides the best method for defining marrow involvement for bone malignancy and soft tissue malignancy when planning for resection margins [29]. MRI and CT (if cortical destruction) to a point have a high sensitivity and specificity for bony tumor infiltration, but margins are difficult to identify intraoperatively, and intralesional resection is very much possible in the pelvis [7, 19] (Fig. 13.5). A surgeon can measure on MRI the marrow infiltration and then recreate this measurement on the CT used in computer navigation as long as a similar slice thickness is used or it can be augmented or "fused" with a MRI for the benefit of identifying marrow or periosteal edema then uploaded to the navigation system [23, 29, 46]. In general, when performing surgery in the pelvis using computer navigation, the surgeon may only use the CT imaging, although some will use a CT-MRI fusion [12, 18, 29, 46]. Some users feel the CT-MRI fusion is beneficial to the surgical procedure by being able to identify marrow margins or margins outside peritumoral edema [29, 34, 46]. While one does not have to take advantage of CT-MRI fusion, this modality is probably the best way to look at soft tissue when using computer navigation. CT-MRI fusion allows for determining the extent of tumor resection planes based off the bony involvement seen on the navigated software [48]. Currently, surgeons not using navigation rely on their interpretation of tumor location on CT and MRI obtained preoperatively and then attempt to translate that interpretation into pelvic landmarks during the intraoperative resection leading to significant inaccuracies in tumor resection [20, 23]. Planning for resection can be done on the computer navigation system prior to surgery or can be used as a way to augment the visual representation of the surgeon. If the surgeon elects to plan resection planes prior to surgery, this virtual planning comes at the time when a CT and/or MRI scan is imported into the navigational software prior to

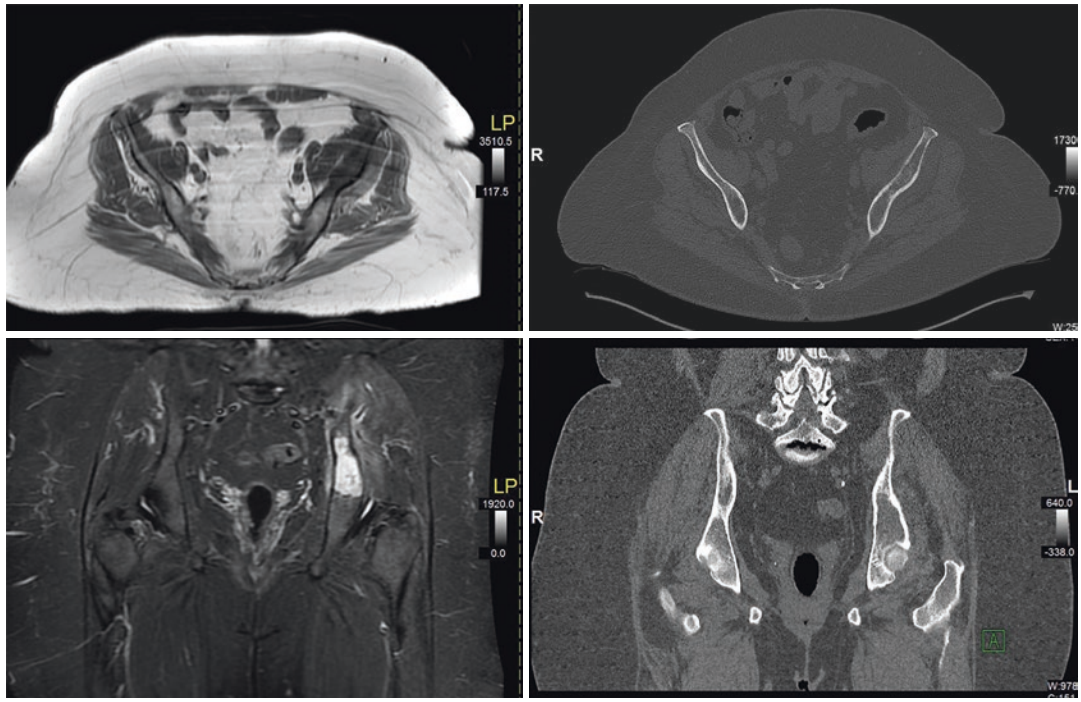

Fig. 13.5 MRI vs. CT scan for identifying marrow infiltration, CT when compared to MRI under appreciates the extent of tumor in the intraosseous bone

surgery and the surgeon elects planes to determine the starting point and vector of the intended plane for future osteotomy or resection plane (Fig. 13.6). One can create planes that represent resection levels and osteotomy sites. One can also paint the tumor and provide the user with collision warnings to minimize that chance of getting into tumor (Fig. 13.7). One can create multiple planes to allow for cuts than are multiplanar (Fig. 13.8).

13.6 Intraoperative Computer Navigation

The intraoperative process consists of tracker or dynamic reference base (DRB) placement on the patient, registration, and then resection. Registration of the patient to the system occurs after the patient is asleep and draped into a sterile field and can occur either at the beginning prior to major surgical incision or at the time prior to bone resection after bone exposure is obtained. The choice based on the technology available to surgeons with the options includes surface matching, paired-point matching with or without fiducial marker, or creating a intraoperative CT scan of a segment of bone to match the preoperative imaging. During computer-assisted surgery, the surgeon must appreciate the importance of DRB placement. Certain types of DRBs can have issues with line of sight such as optical trackers, and it is important to think about surgeon's approach and patient's positioning when using navigated tools [49]. Some navigation systems use electromagnetic trackers for the DRB and the instrument and will not have the same line of sight problem that optical trackers with infrared sensors do; however, they can be prone to electromagnetic interference [36–39, 50]. No tracking system is perfect, and each has their inherent problems [37, 50–52]. However, one study was able to minimize registration error, in the setting of fiducial marker use, by placing the DRB as far away from the fiducial markers and resec-

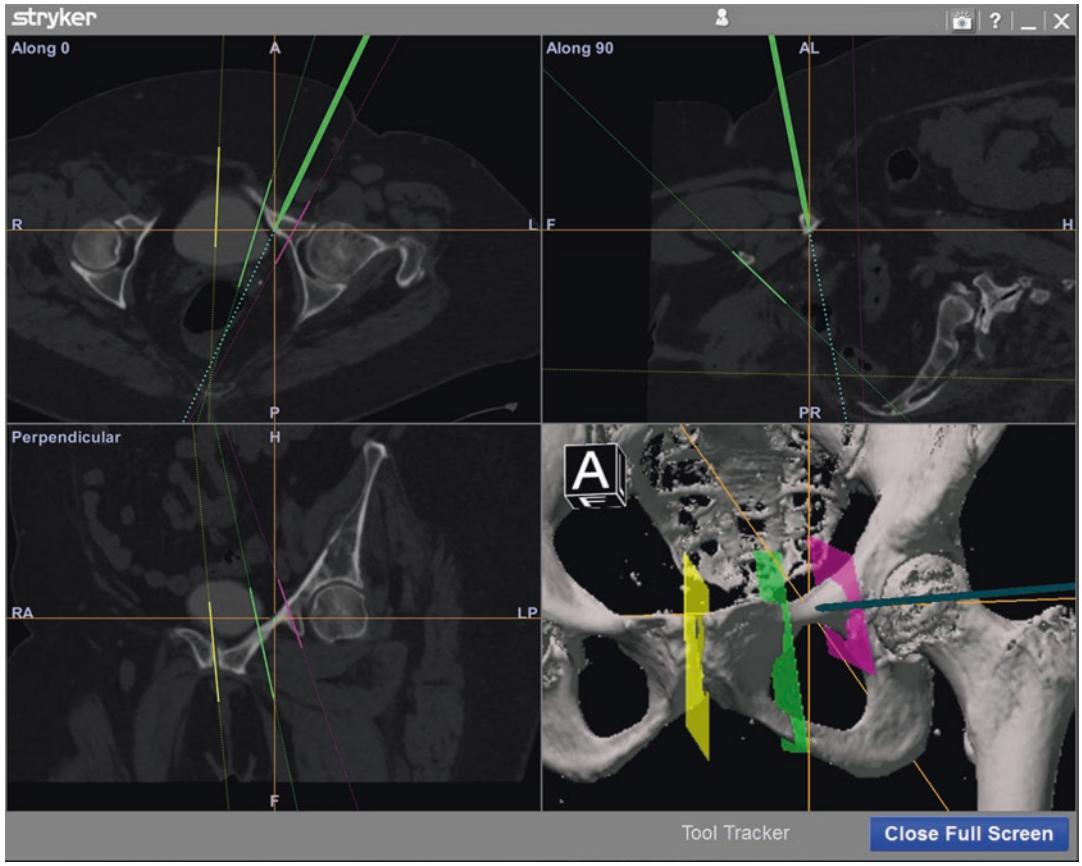

Fig. 13.6 Three planes were created in the virtual planning area to identify the pubic symphysis (yellow), intended resection plane (green) and tumor margin (pink)

tion zone as possible [53]. The DRB can be placed far enough away from the surgical site so to minimize possible disturbances during surgery to the tracker and system; i.e., leaning on the tracker with a retractor or hand, the surgeon should place it in a relatively safe but visualized area. Some authors have recommended the tracker be placed on the contralateral side of the pelvis and when surface matching to create a large enough exposure to allow for all the working area of bone to be included in the sampling surface area [53]. While other authors have expressed concern that the further the tracker is placed away from the resection plane the more room there is for play in the system through stretching and stressing soft tissue connections in the pelvis [51, 54]. Pins are placed into a stable part of the pelvis away from the surgical site to minimize interference and compromise to the registration [53]. One can place pins for the tracker in the same side iliac wing as long as the pins our outside the peritumoral edema zone and outside the resection zone [55]. Only solid pins with good fixation should be accepted because any change in tracker position will affect your accuracy. The DRB must be connected to the bone of an intact pelvic ring for the planned resection or osteotomy because ligament resection can allow bones to move independently and will affect the accuracy of the bone interface. Placing three pins better stabilizes the tracker on the subjects body [56]. Tracker pins that loosen corrupt the registration accuracy and the registration process will be required to be performed again [2].

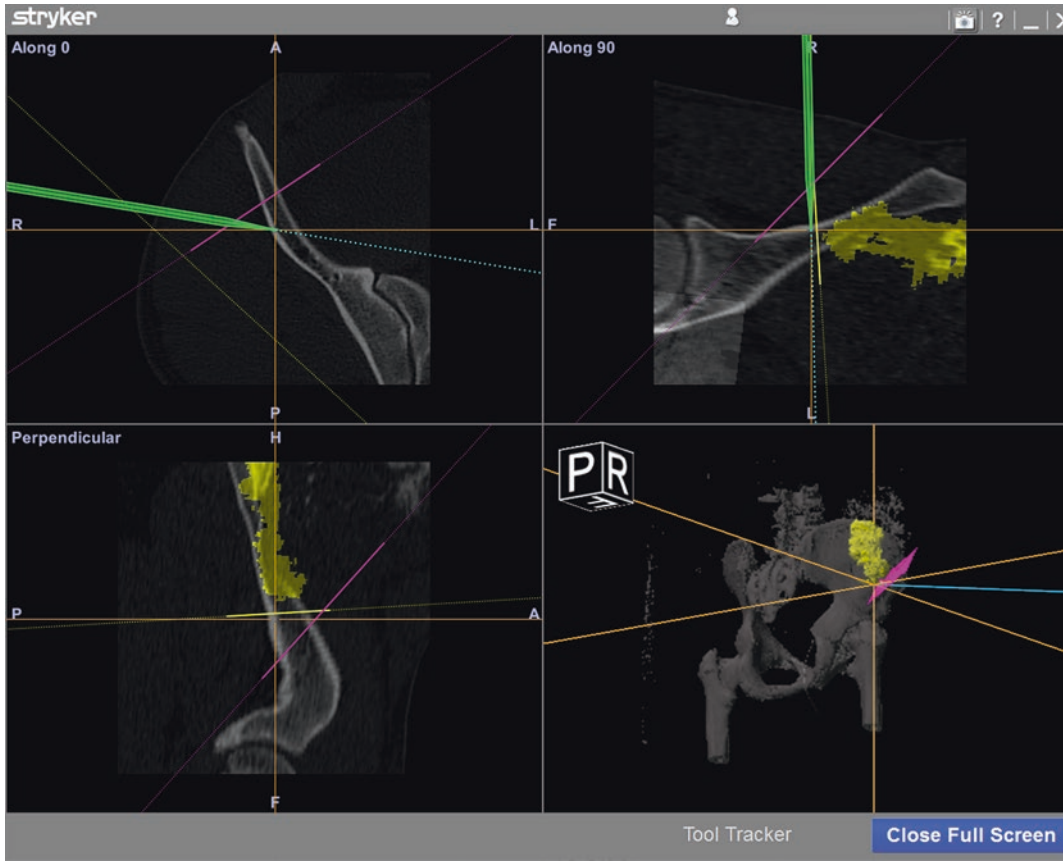

Fig. 13.7 Tumor painted in yellow

13.7 Registration

All advanced imaging-based navigation systems require a registration process prior to navigated surgical resection [57]. Registration is the most significant and error-prone step in navigation due to the fact the surgeon variation can be introduced into the system [58]. This occurs after a DRB has been placed in a position that will be stable throughout surgery [2, 24]. This can be done several different ways and is mostly based on the type of computer navigation system available. If no advanced intraoperative imaging is available registration, then paired-point matching is primarily used and requires a minimum of four points or "paired points" that are chosen on the patient's preoperative MRI or CT image and then identified on patient's bony anatomy [22, 24, 25, 27, 34, 48]. This requires the surgeon to accurately identify both on the CT or MRI image and on the patient's pelvis the corresponding points. Often, the pubic symphysis, ASIS, AIIS, pubic tubercle, and PSIS are used for patient to image matching and confirmation [20, 53]. However, there are specific bony landmarks often that cannot be identified or palpated due to patient positioning. Surface mapping has also been used where one can select 50–65 points on the bony pelvis to register the patient [24]. The difficulty with surface mapping is soft tissue components such as cartilage, ligament, and soft tissue which will interfere due to pliability; thus, bone cortex is used due to its consistency and ease of ability to be defined on a CT scan [48]. MRI is not typically used for surface registration due to the system having difficulty identifying cortex on MRI, and

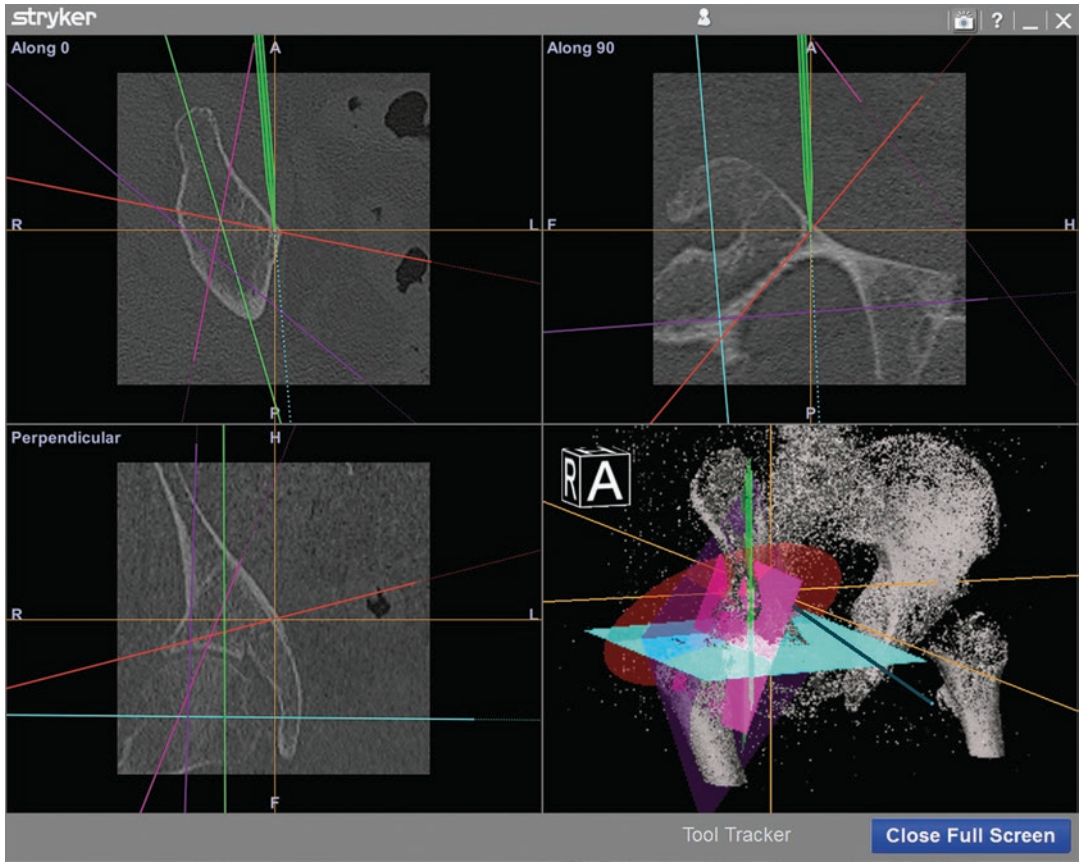

Fig. 13.8 Multiplanar cut with all planes identified at once

thus, CT is the preferred modality when using surface mapping [12, 29]. Both of these processes have inaccuracies of the registration, thus translating into errors in resection location; thus, there was a demand for decreasing registration error, i.e., the difference between the picked point on the preoperative CT and the correct point in the patient. Patients with significant body habitus or BMI [22] will limit exposure and make it difficult to accurately identify bony landmarks for paired-point matching. Planned registration landmarks whether surface features or fiducials should not involve osseous features that may be included in the resection [29], and when using either paired-point or surface matching, the bone and/or the fiducials must be adequately exposed to perform the registration.

Fiducial markers or small implants such as a screw or pin were added to the computer navigation process as a way for quicker and more reliably accurate paired-point registration with minimization of the registration error, i.e., the difference between the preoperative imaging and the actual patient bony anatomy. Fiducials allow for better paired-point registration than would be obtained with bony landmarks alone. These implants serve as fiducials for pair-point matching during patient registration at the time of surgical resection [2]. Fiducial markers can be placed in the pelvis, but an additional surgery to place these markers must occur prior to the preoperative CT [45, 57]. Fiducial positioning is chosen based on ease of access during surgery such as the iliac crests, ASIS, AIIS, and posterior iliac spines [2, 18]. No more than four fiducial markers are needed for an accuracy of 1.5 mm [59], but they need to be accessible in both the prone and supine position if the patient needs two

approaches. With CT, small titanium fiducial markers such as a Kirschner wire [12] or screw are the preferred implant as stainless steel will have too much metal artifact to identify a 1 mm point of registration. Titanium screws or K-wires chosen as fiducial markers do not allow for the MRI to be used in preoperative imaging for navigation. Bioabsorbable fiducial markers are required when MRI is used in paired-point registration in order to obtain a registration error of <1 mm [18] because of metal artifact. Bioabsorbable nonmetallic 1.5-mm pins placed beyond the tumor resection can act as fiducials so that CT-MRI fusion and MRI images alone can be used for patient registration [29]. When using fiducial markers, the slice thickness of either the preoperative MRI or CT scan must be less than the width of the fiducial; otherwise, the possibility exists that the fiducial marker will be missed on the scan [29]. Due to the possibility of needing to flip the patient in the case of some surgeries involving parts of the sacrum, fiducial markers (1.8-mm titanium K-wires) can limit the need for a second intraoperative image acquisition that allow them to quickly reregister the patient after replacing the patient tracker [2]. When placing fiducials, they should be placed in solid bone because placing them erroneously in soft tissue would lead to significant registration errors and an inaccurate navigated tool.

Registration with intraoperative advanced imaging through either a 3 D fluoroscopy C-arm (Fig. 13.9a, b) CT scan machine or O-arm intraoperatively (Fig. 13.9c–f) to create a CT image that can be matched with the preoperative imaging [45, 60–63] to make the registration process more successfully consistent with minimal registration error. Image to image registration using intraoperative advanced imaging such as 3D fluoroscopy or intraoperative CT [45] offers the benefit of not requiring fiducial markers along with a preresection surgery and can have improved registration accuracy over pair-point matching using bony landmarks. These systems can allow the surgeon to manually match and then use automatic matching done by the navigational software to fine-tune the process (Fig. 13.10). The image fusion process is not without its own contribution to registration errors because merging is still done visually by the surgeon leading to potential error even with the best processes [29, 64].

13.8 Registration Accuracy

Registration error is a way to determine accuracy of the "match" can be reported in some systems based on the method used to match the patient to the preoperative CT scan or CT/MRI fusion scan [29, 46]. Registration error can be calculated by the navigation software and gives an indication of the mismatch and provides a value of difference between the point picked on the preoperative advanced image on the intraoperative monitor and the corresponding location on patient's bony. It can help the surgeon decide if they need to perform registration again depending on the difference seen. A registration error of <1 mm is the goal, but some will accept below 2 mm [22, 24, 46, 48, 65, 66]. Most would advocate for a CT or MRI scan slice thickness of 0.5–1 mm in order to obtain registration errors less than 2 mm. For paired-point and surface matching, a registration error of greater than 2 mm will not infrequently be obtained which can be due to soft tissue such as thick cartilage, tendinous, or ligamentous insertion being in the way of a bony landmark leading to an inaccurate registration [22]. Even with fiducial markers, placing the navigational tool inside a screw head (variability of 1–2 mm) or at the tip of a K-wire can introduce error by not selecting the correct point on the preoperative imaging and then by not matching that location well enough intraoperatively. Depending on the software used, the surgeon has options to improve the accuracy of the registration through several different processes. Surface matching originally intended as a way to avoid a second surgery (preresection fiducial surgery placement) is a way to improve the accuracy of the registration [21, 41, 57]. The navigational probe can be used to select in continuous succession a minimum of 30 points but often up to 100 points on the patient's exposed pelvis boney surface [22, 24]. If the registration error is unacceptable even with attempts at refinement, the surgeon will have to start over again by locating

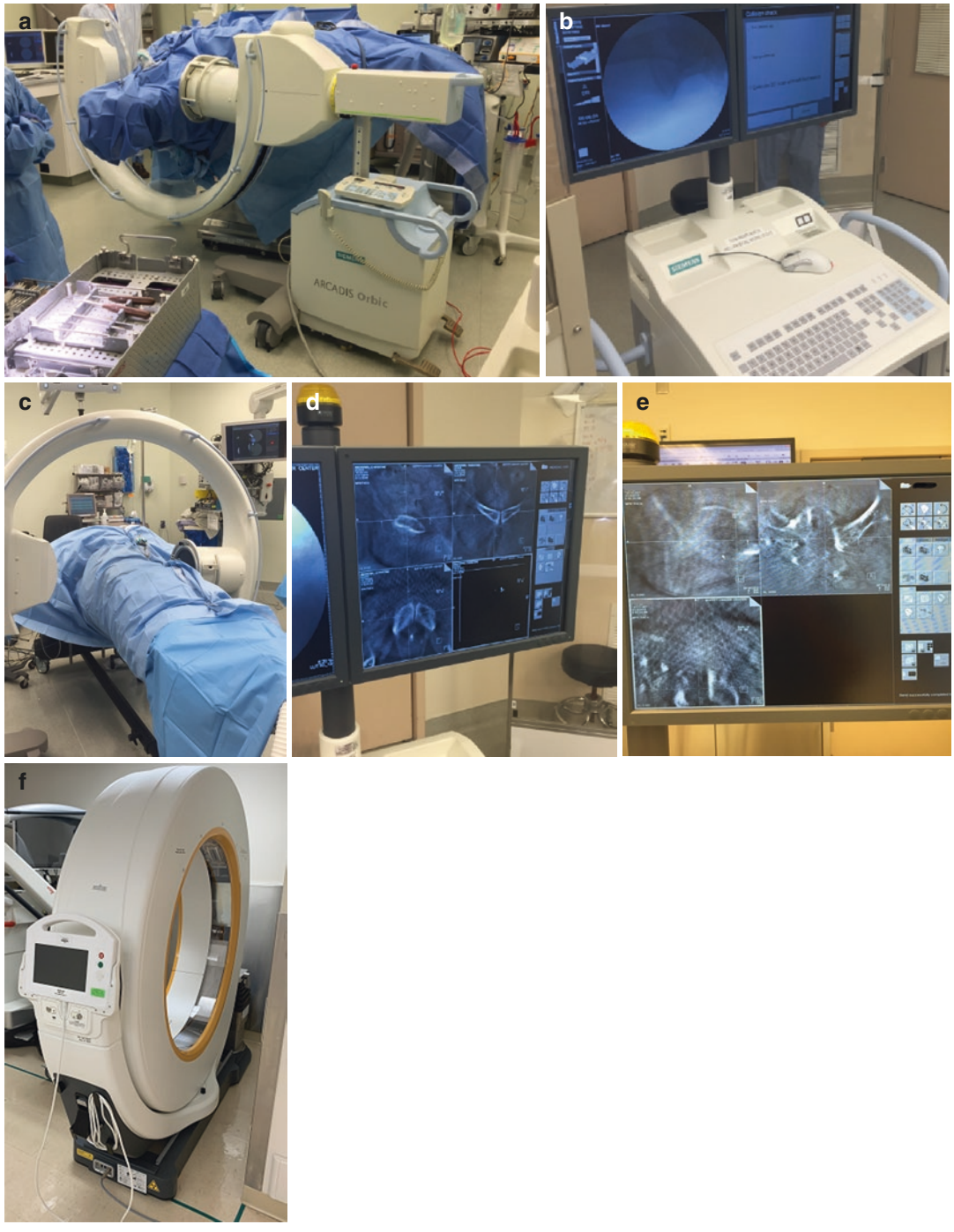

Fig. 13.9 (**a**) 3D fluoroscopy unit used to create intraoperative CT like image. (**b**) CT like image created by 3D fluoroscopy unit for import into navigation system. (**c**) Intraoperative CT scanner. (**d**, **e**) Intraoperative image acquisition. (**f**) A particular of a CT scan machine

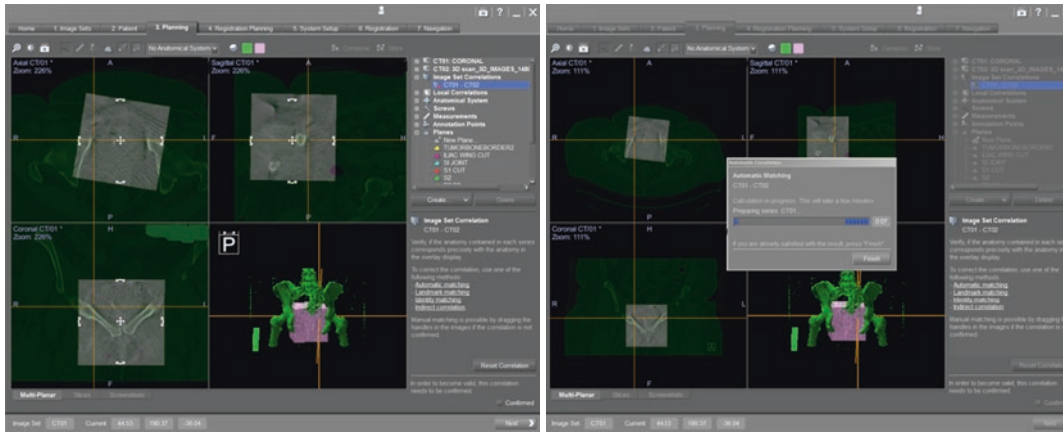

Fig. 13.10 Process of "fusing" or matching the preoperative CT scan with the intraoperative pseudo CT image created by the 3D fluoroscopy machine

points after confirming in the navigation system he has picked his correct landmarks on the preoperative scan assuming that metal scatter is not interfering with identifying the point. One may not be able to obtain a less than 1 mm registration error, and one will have to accept a 2 mm error. Rarely does one have to abandon navigation.

13.9 Computer-Navigated Resection

Once registration is complete, and most of the surgical dissection is performed navigated resection can be attempted. Custom tracker connectors allow about any type of tool to be navigated as long as it can be calibrated registered to the patient with a tracker, so they can be tracked in space (Fig. 13.11). Several different tools can be used for navigation and can be navigated and include diathermy device [22, 24], ostetomes [67], chisels [1, 2, 33], drills [68, 69], burrs [18, 46], screwdriver [67], and oscillating saws [20, 24, 34, 70]. One thing to be aware is tool localization may differ between the display of the instrument on the monitor and what is seen in the operative field because tool registration can degrade with striking a tool or using a vibrating instrument can cause loosening of the instrument tracker causing inaccuracies over time [71].

Fig. 13.11 Navigated 1/4 in. osteotome

The navigated saw and navigated osteotome are some of the most common tools used. The navigated saw with a thinner, smaller blade allows one to use it in smaller exposures with limited excursion, but the instability due to vibrations with increased flexibility creates instability and produces potential error in the

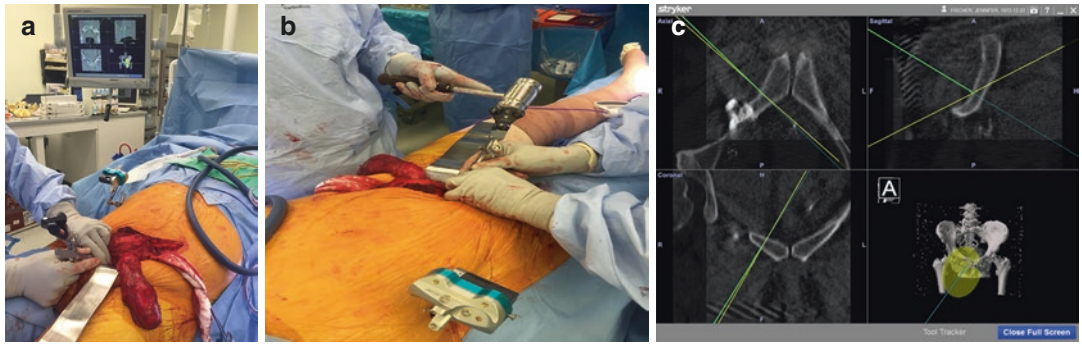

Fig. 13.12 (**a**) Navigated ½ in. osteotome for completion of cut. (**b**) Navigated osteotome for completion of cut. (**c**) Planned cuts

planned resections [72] as well as injury to tissue or disruption of tumor due to oscillations [24]. A slight push by the surgeon, the smaller saw blade allows significant flexion of several millimeters that can cause an inaccurate positional reading of the saw blade [23, 70]. Even with these drawbacks with the saw, there have been significant improvements for angle of cut and location of the cut plane (2.8 mm) when using a navigated saw compared to the free-hand process (5.7 mm) [20]. The other navigational tool at hand is the osteotome that allows the user to have nonflexible tool that gives reliable depth and trajectory feedback via the computer navigational screen to the surgeon (Fig. 13.12a, b). The drawback to the osteotome is that using it can cause unwanted fractures in bone. Both the saw and the osteotome are ideal for uniplanar cuts, but when a multiplanar three-dimensional cut is needed, another method is used. The surgeon can use the navigational pointer to identify these "way-ward points" [12] in a multiplanar cut and mark them with cautery or a sterile marker [11, 23, 48, 68]. At this point a drill or a burr can be used to create several holes along the planned cut can then be completed with either an osteotome or burr at the discretion of the surgeon. A burr has benefit over a drill, in that it can be used to thin cortex on the far side before coming into contact with tumor or a critical structure that may exist on the opposite side (Fig. 13.13). Disruption of the ring by ligament or bone sectioning can potentially disturb the accuracy of the spatial relationship and corrupt the registration by introducing a certain amount of uncertainty into the system for the next cut [18]. This is particularly important because cutting sacroiliac and symphyseal ligaments in the pelvis can create subtle mobility that can affect the correct location of the bone cut. An inappropriately timed osteotomy introduces error into the navigation system by leading to inaccurate second osteotomy [22].

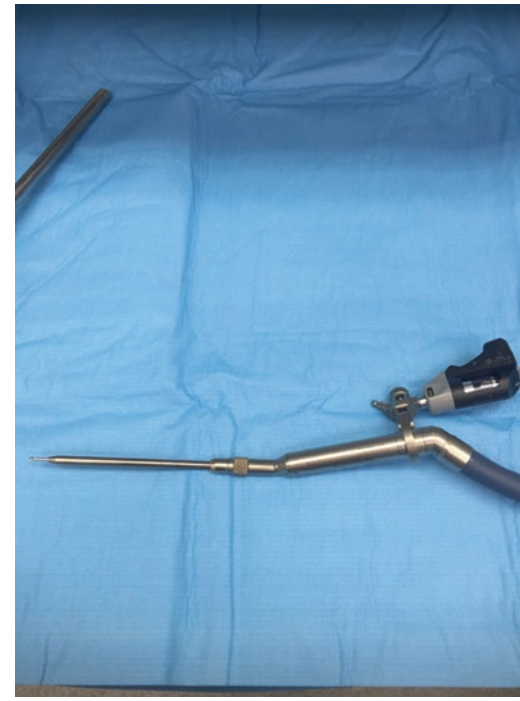

Fig. 13.13 Navigated burr 1.5 mm head

13.10 Topics Unique to Computer Navigation

One of the drawbacks to navigation is the added time to surgery, and with added surgical time, complications rates can increase. Tracker insertion and registration increases surgical time between 15 and 47 min but, after the learning curve, can often decrease from an average of 30 min to 20 min after the surgeon has performed more navigated surgeries [22]. Relatively speaking, this added time in pelvic surgery is probably insignificant given 30 min of delayed surgical time may only represent 6% of surgical time in an 8-h case. Although additional operating time is needed for navigation set up; planning and defining the resection plane on preoperatively obtained images can reduce the overall surgical time since the osteotomy is already defined on the navigation screen, thus negating the need for multiple fluoroscopic images [48]. Time is then saved during surgery by not having to bring in C-arm or X-ray for multiple orthogonal images to determine the correct resection level if that can even be appreciated on fluoroscopic images. Using navigation, one can eliminate the need for a C-arm to come in and out of the surgical field, thereby decreasing the risk of inadvertent contamination as well as limiting the radiation exposure to the surgical staff [40, 73, 74]. Navigation often can allow surgery to be performed through one approach which could negate the need for flipping the patient. In the situation or having to flip back and forth, bony landmarks can be identified in other areas of the pelvis that can then be marked on the computer navigation software that can be used to reestablish the registration [24]. Fiducial markers can make things easier or needed if the patient is needed to be flipped from supine to prone; not all the time can the surgery be performed only through a posterior approach [8, 49]. Fiducial markers can improve and may negate the need for another image acquisition spin need be performed, but rather the fiducial markers can be marked as rescue points in order to alleviate the need for another spin.

13.11 Visualization, Accuracy, and Margin Benefits

The overall benefit to navigated surgery is the reproducibility of the surgical resection. In a study of 28 patients with 61 osteotomies using paired-point registration, the quantitative difference between the planned osteotomies and performed osteotomies was 2.52 ± 2.32 mm for all patients and 2.82 ± 2.01 for the pelvis [23, 75]. An ex vivo experimental study showed the accuracy of performed osteotomy planes with respect to the planned planes in the pelvis was significantly improved by almost 9 mm using a navigated saw, averaging 2.8 mm compared to 11.2 mm for the freehand saw ($p < 0.001$), and no intralesional tumor resections were executed compared to 22% ($N = 5$) intralesional violations in the freehand group ($N = 23$) [70]. This showed by choosing a desired safe margin of 10 mm that the maximum difference achieved between the cut and desired margin was 6.5 mm for the navigated saw cut compared to the 13 mm conventional cut [70]. In a separate laboratory study involving intraoperative CT registration and navigated saw, the navigated sawbones' entry cuts were within 1.4 ± 1 mm and exit cuts 1.9 ± 1.2 mm from the intended osteotomy plane and were significantly different ($p < =0.01$) to nonnavigated 2.8 ± 4.9 mm entry cut and 3.5 ± 4.6 mm exit cut in a pelvic bone model [72]. The navigated saw accuracy was evaluated on a cadaver and produced similar differences in intended and executed osteotomies which showed a navigated entry cut of 1.5 mm ± 0.9 mm and navigated exit cut of 2.1 ± 1.5 mm [72]. Navigation theoretically allows a 95% certainty of avoiding a positive margin as long as the osteotomy is planned no closer than 5 mm of the tumor [72]. Using conventional techniques, there is only a 52% probability of achieving a 1 cm margin in a triplane-simulated tumor model of the pelvis due to the complex 3-D geometry [15]. Another experimental test comparing navigated vs. nonnavigated saw cuts with a nonnavigated saw had a 22% intralesional violation rate compared to 0%

in the navigated modality [60]. One study looking at navigation in the pelvis reduced the intralesional rate to 8.7% (n = 2) with clear bone margins in all cases (n = 31) with a 13% local recurrence rate compared to the traditional method where intralesional rate and local recurrence rate were found to be 29% and 27%, respectively [24].

13.12 Tissue Sparing and Reconstruction

Surgeons using computer navigation believe it allows for more complex resections and reconstructions than are possible with conventional surgery allowing preservation of sacral nerve root controlling bladder and bowel (42% of time), resect unresectable tumors (13% of the time), and avoidance of hindquarter amputation (10% of the time) [24]. Navigation can allow for preservation of the complete or partial joint making reconstructions less complex because more bone can be saved without compromising margins [76] as a free-hand navigated saw improves cutting accuracy [72, 77]. Even a navigated tool's bone loss related to the saw blade thickness or other resection tool can be adjusted for on computer navigation planning by shifting the planned resection planes by 1.5 mm [20]. In fact, most resection planes can be adjusted preoperatively based on known resection width tools such as the osteotome (0.6 mm) and oscillating saw blade (1.25 mm) which produced a loss of bone width of 2 mm due to oscillation [72]. The precision and reproducibility of navigation can allow surgeons to resect the tumor and then use the navigation software to plan for an allograft piece of pelvis to reconstruct allowing for functional limb salvage with a joint reconstruction [78–81]. Computer navigation osteotomies help with matching to surface contact area of the allograft to the host bone that can minimize nonunion rates [79]. Navigation, with it's better accuracy and precision is now used to save bone and produce joint-sparing bone cuts [60, 76] (Fig. 13.14). The

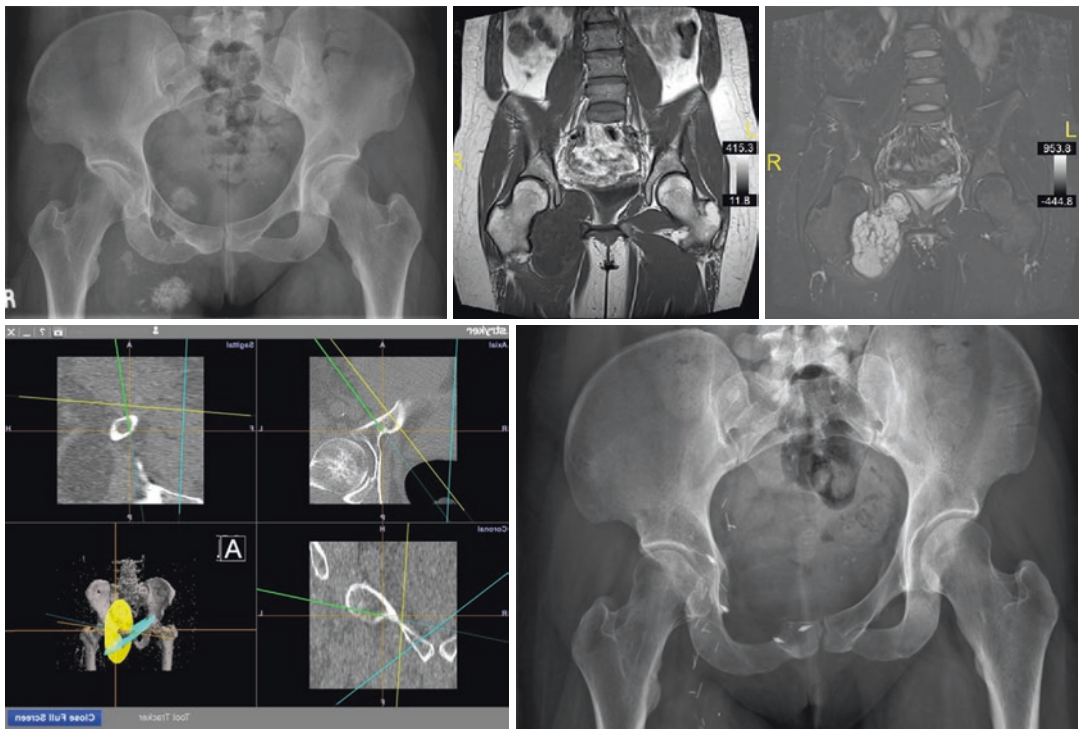

Fig. 13.14 Examples of joint and pelvic ring preserving cut of the hip joint for a chondrosarcoma

system can also be used to produce pelvic ring sparing cuts that will prevent the ring from being disrupted. In the era of 3-D printing, custom prosthesis can now be created that eliminate the need to modify an allograft in order maximize contact area (Fig. 13.15). In order for the custom prosthesis to fit precisely, only computer navigation osteotomies can consistently provide enough accuracy. The precise cuts and virtual model can be loaded to the system to allow for prosthesis matching osteotomies (Fig. 13.16). Theoretically, this can minimize reconstruction and wound exposure time as well as having better reconstruction to host contact that can reduce complications and failures. Computer navigation does not only have to be used for resection and reconstruction of malignant tumors; its use in benign tumors has been described [13] as there is benefit to using navigation for a en bloc resection over the choice of using curettage in an effort to minimize recurrence and not affect structural integrity of the pelvis [13, 82]. Computer navigation using an O-arm has been used to treat benign tumors and hematologic tumors tumor ablation and kyphoplasty in benign and malignant nonprimary bone tumors [83].

While navigation may be useful in achieving negative margins, there is some that feel that with some of the difficulty in learning to use navigation that navigation may not routinely be needed for all

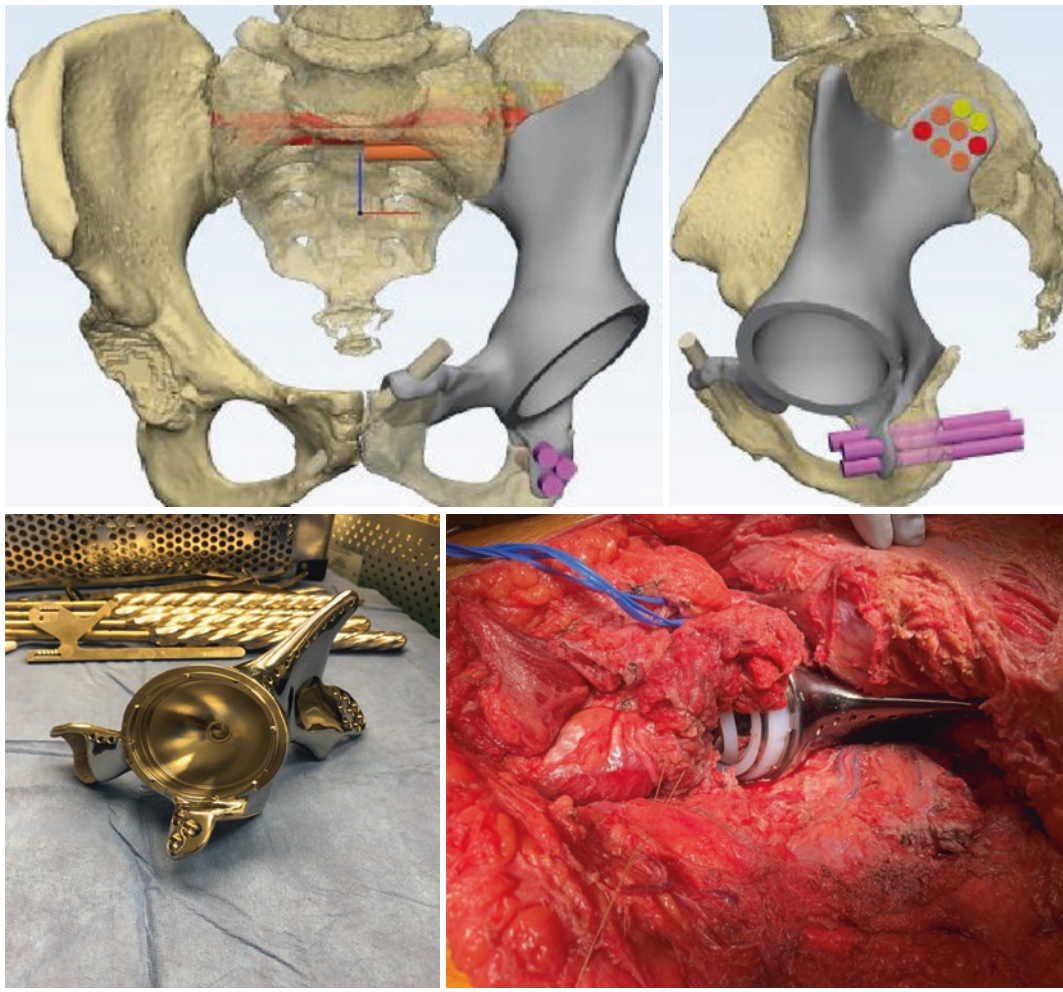

Fig. 13.15 3D planning and printing for navigated resection and reconstruction of the pelvic sarcoma

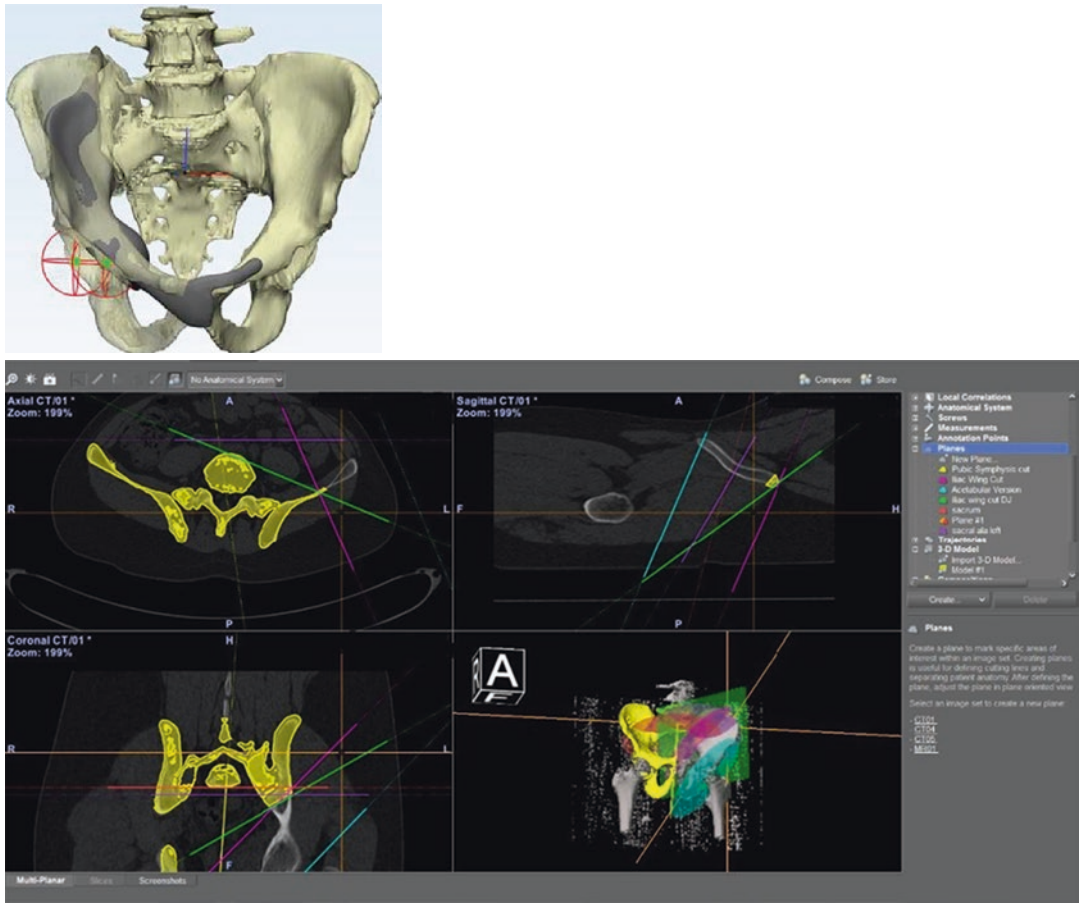

Fig. 13.16 Two 3D virtual models with one showing the ability to plan for the new hip center and the other model (yellow) being loaded to the intraoperative navigational software to assist with planning osteotomies

oncologic resections in bone [55, 84]. Pelvic surgery is difficult, and adding navigation only helps if one is familiar with the system. One must understand the limitation of the navigation system one is using as well as having knowledge of accuracy of the system [27, 50–52, 85]. Doing 1 or 2 pelvic cases a year does not allow the surgeon to become familiar enough with using pelvic navigation in this setting. Surgeons are often frustrated with need for multiple steps needed to make one complete resection with computer navigation. Computer navigation is a learned process and skill that takes relatively large numbers of cases to master. This is not a once a year tool. As with any technology unless you routinely use it, you would not see the benefit it provides. Centers that do high volume of computer navigation cases can offer their patients more accurate surgical resection and reconstructions. Computer navigation has increased the precision and accuracy of pelvic resections leading to preservation pelvic structures with the intention of not compromising oncologic outcomes in terms of margins and recurrence.

13.13 Computer Navigation Summary

The overall benefit to navigated surgery is the accuracy and reproducibility of the surgical osteotomy. It is difficult to achieve negative margins in pelvic surgery [8] as tumors involved in the pelvis have a higher prevalence of positive margins [9] leading to higher recurrence rates and poorer outcomes.

Pelvic three-dimensional anatomy is difficult to understand and master conceptually when trying to resect tumors [14, 72] making it difficult to get tumor-free margin leading to higher local recurrence [2, 21, 86, 87]. Without navigation, performing the planned resection with precision and accuracy is very difficult to achieve with conventional resections in the pelvis [15, 70]. Computer-navigated surgery can increase precision of the osteotomies for tumor resection [1, 2, 12, 18, 48]. Clinical studies have shown that navigated tools have assisted in attaining negative margins in malignant pelvic tumors [1, 18, 20, 34, 68, 86]. The surgeon can plan the desired margin with respect to the tumor on the navigational software and then use it to assist during resections, maximizing accuracy and precision and minimizing sacrifice of critical structure [88]. Where computer navigation excels is the ability to "see beyond walls" because it is hard to know what lies on the other side of the pelvic cut. Navigation literally shows the surgeon where the instrument is heading in real time and how close the tool is coming to tumor on the other side. It can help the surgeon minimize the chances of positive bone margins. It can also help the surgeon perform resections that make reconstruction easier and help maintain function. The future of computer navigation surgery will likely include the benefit of robotic assistive devices and patient-specific instrumentation [86]. Computer navigation currently is a passive system which only provides information or feedback, while the future computer-navigated robotic-assisted surgeries will be performed with a more active role that physically guides and limits the surgeon from straying outside of predetermined resection planes [89]. The benefit to robotic-assisted surgery involves still maintaining control of saws and osteotomes while minimizing the effect of tool vibration and fatigue on part of the surgeons hand that navigation alone cannot currently provide [90]. During surgery, computer navigation offers a lot of benefits; however, the system is not full proof. Navigation does not guarantee the margin will be clear because the incorrect margin or resection level is surgeon chosen on the preoperative imaging, but the system does provide accuracy, consistency, and reproducibility with the bony resection plane [24].

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Spinopelvic Fixation After Sacrectomy

14

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14.1 Introduction

Obtaining solid arthrodesis of the lumbosacral region of the spine continues to be a challenge for spine surgeons. Various spinal pathologies require a spinopelvic fixation including adult deformities with coronal or sagittal malalignment, neuromuscular scoliosis with pelvic obliquity, high-grade spondylolisthesis, and lumbosacral tumors (primary or secondary) [1–3]. The latter represents actually the most challenging indication for a spinopelvic fixation, due to the special anatomical (large vessels, bladder, bowel) and biomechanical characteristics of the lumbosacral region. From a biomechanical point, the sacrum–sacroiliac joint–ilium complex cannot be discussed separately, as the vertical load from the axial skeleton is transferred to the lower limbs via this area; thereby, an intact sacropelvic region is one of the key points of the human upright posture and walking ability. Tumors in this region alone as well as their surgical resection can significantly influence the biomechanics of the sacropelvic complex resulting in failure of axial load transmission. Therefore, to perform an oncologically and functionally optimal surgery, both the anatomical and biomechanical consequences must be carefully considered during preoperative planning [4].

En bloc resection of tumors in the lumbosacral region with procedures such as total sacrectomy or L5 spondylectomy is typically indicated for patients with locally invasive primary sacral tumors such as chordomas, sarcomas, chondrosarcomas, or giant cell tumors. In contrast, this strategy has been applied in limited cases to metastatic diseases, mainly due to recent advances in adjuvant treatment in surgical oncology [5]. Additionally, treatment of metastatic tumors in the lumbar spine near the lumbosacral junction often requires pelvic fixation even with separation surgery procedures to allow for adequate stabilization of the lumbosacral junction, as these patients often have poor bone quality [5].

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Therefore, standardizing universal surgical procedures and techniques is a copious attempt for spinal and orthopedic surgeons.

14.2 Indications

Total sacrectomy is indicated for primary sacral malignancies. More rarely, sacrectomy is indicated for patients with primary or recurrent pelvic visceral tumors (most commonly colorectal carcinoma with sacral involvement by direct extension) and no evidence of metastatic or nodal disease. The techniques hereby described may also be adapted to intralesional treatment of benign tumors such as osteoblastoma and aneurysmal bone cyst. Last, some carefully selected benign aggressive sacral tumors may also be considered for en bloc resection, particularly if small or recurrent [6].

A surgical procedure of a much larger scale is the amputative sacral resection that extends into the pelvis and may be necessary in the following instances: tumor epicenter within the ilium but disease extends across the sacroiliac joint, or tumor epicenter in the sacrum but removal requires resection of the femoral nerve as well as the lumbosacral trunk or the lumbosacral trunk and the hip joint articulation. In these cases, the ultimate function of the limb is so poor that external hemipelvectomy in conjunction with sacral resection allows the maximal oncologic margin to be obtained and provides for healthy and robust flap coverage from the limb [6].

On the other side, the presence of disseminated malignancy is a strong relative contraindication for sacrectomy. The procedure is of such magnitude and generally entails deliberate neurological defects with frequent loss of bowel, bladder, sexual, and potentially lower extremity function that it is usually inappropriate to pursue without curative intent. Patients with tumor thrombus in the iliac veins or vena cava by sarcoma predictably have a rapid development of metastatic disease and demise [6]; evidence of the above on preoperative imaging can prompt catheter-directed biopsy, while its finding at time of surgery prompts abortion of resection [6]. The inability to obtain a tumor-free margin of resection is similarly a relative contraindication. The medical status of the patients also is important; patients receiving chemotherapy frequently require alterations in their chemotherapy schedules to allow for surgery of this magnitude. All patients are subject to an intense preoperative medical evaluation including a dobutamine stress echocardiogram for (a) anyone with known cardiovascular disease; (b) men above age of 40 years; or (c) women above age of 50 years [6].

Special consideration has been made regarding the use of spinopelvic fixation with dual iliac screws. These constructs are necessary mostly if (a) total sacrectomy is performed where the whole sacroiliac joint is removed on both sides [7], (b) partial sacrectomy is performed involving more than 50% of sacroiliac joint on each side, or partial sacrectomy involving less than one half of the sacrum but with one-side sacroiliac joint resection [8], and (c) in palliative fixation cases for unstable destructive lumbosacral metastatic lesions where pedicle screw anchorage in the sacrum is extremely poor [1, 9].

14.3 Classification

There is no standard classification of these procedures. Mayo Clinic has proposed the following classification, where resections could be divided into five types based on the extent of the lumbosacral resection and the need for an associated external hemipelvectomy [10, 11]. They are as follows: Type IA resection—total sacrectomy; Type IB resection—subtotal sacrectomy above the S1 foramen; Type IC resection—subtotal sacrectomy below the S1 foramen (the SI joints are not disrupted here, and a reconstruction is not typically performed); Type II resection—hemisacrectomy with or without partial lumbar excision, and iliac wing resection; Type III resection—external hemipelvectomy with hemisacrectomy with or without partial lumbar excision; Type IV resection—external hemipelvectomy with total sacrectomy with or without lumbar excision; and Type V resection—hemicorporectomy-type procedures (Fig. 14.1).

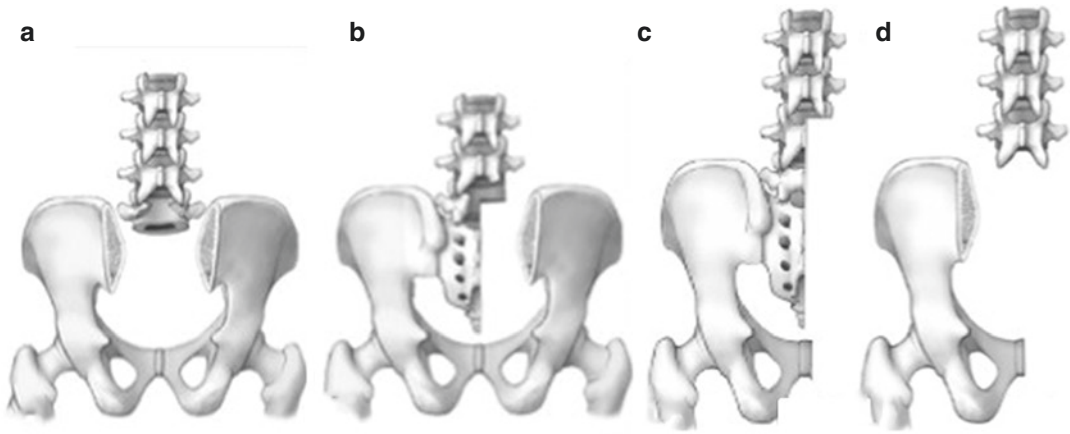

Fig. 14.1 Mayo classification of spinopelvic resections: (**a**) total sacral resection (total sacrectomy), (**b**) hemisacral resection (partial sacrectomy), (**c**) hemisacral and lower lumbar spine resection (partial sacrectomy and extended external hemipelvectomy), and (**d**) total sacral and lower lumbar spine resection (total sacrectomy and [extended] external hemipelvectomy)

14.4 Techniques and Implants

Generally, there are three components in spinopelvic surgery, spinopelvic fixation, posterior pelvic ring fixation, and anterior spinal column support [5]. The infrequency of these procedures does not allow for an established gold-standard technique, as the various instrumentation techniques are scattered across the literature in case reports and small case series; however, it was implied that incorporation of anterior spinal column fixation in reconstructing the spinopelvic junction may lead to improved outcomes with lower rates of hardware failure and other surgical complications including blood loss [12].

14.4.1 Spinopelvic Fixation

The goal of the surgical stabilization after an extended oncological procedure such as sacrectomy is to restore the load-bearing structures from the lumbar spine to the remaining pelvis. There are several techniques for spinopelvic instrumentation that were originally described for spinal deformity or trauma surgery. These include sacral sublaminar wires and hooks, S1 tricortical screws, Galveston rod technique (direct implantation of L rods into the iliac crests), intrasacral rods, transiliac bars, iliac screws, and S2-alar-iliac screws [5]. Some of the earliest reported techniques used Harrington rods [13, 14], or hooks and CD rods connected to transiliac bars [15]. The Galveston technique for spinopelvic fixation was initially described by Allen and Ferguson for use in scoliosis surgery in 1982 [16]. It was later modified for use in spinopelvic reconstruction after sacrectomy [17, 18]. Since then, various modifications have been proposed and the direct implantation of rods into iliac crests has been practically abandoned. In the modified Galveston technique, the rods are fixed to the pelvis with iliac screws in the caudal part of the system. Further modifications include the multiple rod–screw techniques such as the four-rod reconstruction or double-rod double iliac screw reconstruction [19]. The use of one single U-shaped rod anchored with iliac screws—the closed-loop technique—was published in 2009 providing a more harmonic stress distribution along the whole construct [20].

Iliac screws represent a modified version of the Galveston technique having three times more biomechanical strength than the Galveston technique, and at the same time, they are applicable in every case, unless a hemi- or partial pelvectomy is required [5]. In fact, one of the main indications

for the use of dual iliac screws is the need to perform partial or total sacrectomy in order to have a solid basis at the bottom of the spinopelvic construct. In addition, the dual iliac screw techniques may be used in palliative fixation of metastatic lumbosacral lesions with extremely poor sacral bone quality [1]. Several biomechanical cadaveric studies have evaluated theses fixation techniques. In the setting of total sacrectomy, Mindea et al. [7] showed that the double-rod double iliac screw technique provided the most rigid fixation, followed by the single-rod double iliac screw fixation, in comparison with single-rod single iliac screw or modified Galveston technique. Yu et al. [21] showed that dual iliac screws, when all inserted in the lower iliac column, exhibited higher compressive and torsional stiffness not only when compared to single iliac screws (short and long) but also to dual iliac screws where two screws are inserted in the lower iliac column and two screws in the upper iliac column.

In terms of selecting iliac screw length, according to the biomechanical study of Zheng et al. [22], short iliac screws (7 mm in diameter and 70 mm in length) are susceptible to loosening after cyclic loading. Bone cement augmentation of short screws has shown a significant increase in the fixation strength of short screws to an extent similar to that of long iliac screws (7 mm in diameter and 120 mm in length). Therefore, given the potential complications of long screw breach, short iliac screw fixation with augmentation with bone cement may be a viable option for lumbopelvic reconstruction, although much larger screw diameters are currently available and more commonly used [5]. According to biomechanical analyses, it cannot be argued that a stronger construct with multiple rods and screws increases the rigidity of the construct. However, it should be also kept in mind that more metal implants increase the risk of wound healing problems; therefore, increased caution is required [23].

14.4.2 Posterior Pelvic Ring Fixation

Techniques for posterior pelvic ring reconstruction include allografts (femur or tibia) with screw fixation to bilateral iliac, transiliac bars, and cages [5]. With the triangular frame reconstruction, the pulled down L5 vertebral body is affixed to the bilateral ilium with sacral rods. The pelvis is also connected to the spinal rods with a second sacral rod [4]. Murakami et al. [24] showed in their in vitro and in silico biomechanical analyses that there was less stress concentration on the implants with this technique; however, excessive stress occurred in the iliac bones that could be associated with loosening of the sacral rods. Gallia et al. [25] published a challenging technique known as the Johns Hopkins University (JHU) technique; a modified Galveston technique was used, where a transiliac bar was inserted through the iliac crests, and single iliac screws were implanted and linked with a horizontal rod. The spinal rods were attached to the transiliac bar with L connectors, and the transiliac bar, the horizontal rod, and a horizontal connector between the vertical spinal rods were connected with one other using vertical connectors. Last, a femoral allograft was placed horizontally, between the two iliac crests bridging the defect.

14.4.3 Anterior Spinal Column Support

The importance of anterior spinal column support in lumbopelvic reconstruction after total sacrectomy has been discussed extensively. In 2005, Dickey et al. [26] published the use of bilateral fibular grafts. The fibular grafts are placed between the L5 vertebra and the bilateral iliopectineal area, and this technique can be combined with the posterior stabilization techniques. In vitro and in silico biomechanical study showed that with the help of these combined systems, greater rigidity can be achieved; however, the increase of the morbidity from the extension of the surgery has to be also considered [4]. A cadaveric biomechanical study by Cheng et al. [27] evaluated the following four constructs: sacral rod reconstruction; bilateral fibular flap reconstruction; four-rod reconstruction; and improved compound reconstruction (a combination of the previous methods). Among these,

improved compound reconstruction that utilized the sacral rod and the fibular triangular construct in the anterior approach produced optimal structural stability after total sacrectomy. Similarly, Clark et al. [28] examined the biomechanical strength of three constructs: femoral strut allograft reconstruction, where a femoral allograft was placed between iliac and secured with bone screws; L5–iliac cage strut reconstruction, where two titanium cages were placed obliquely, each wedged between the inferior L5 endplate and the iliac bone; and S1 body replacement with expandable cage reconstruction, in which a rod was placed from the inferior L5 endplate and fixed to a transiliac bar and a 22-mm expandable cage was placed between the L5 endplate and the transiliac bar. They concluded that the latter technique provided the most biomechanically stable structure.

- *Type 1 and 2 resections*
- Resections at or below the level of the S2 neuroforamen are generally resected through a posterior approach unless there is involvement of pelvic visceral or vascular structures. Given the need to obtain an oncologic margin, this generally implies lesions at or below the S2/3 vestigial disk [6].
- Lesions cephalad to this level or involving pelvic structures are treated first with anterior mobilization of pelvic structures, vessel ligation, and unicortical anterior sacral osteotomy. The use of pedicle flaps is encouraged for facilitating wound healing. A pedicled myocutaneous rectus abdominis flap can be harvested in this stage and tucked into the abdomen with the anterior procedure. Tumor resection is then completed through a posterior approach, and the rectus flap is pulled through the abdomen and rotated to assist in wound closure and reconstruction of the posterior abdominal wall. The posterior approach can be performed 48 h later, unless the rectum is devascularized and requires resection with the tumor specimen [6].
- Resections cephalad to the S1 neuroforamen require spinopelvic reconstruction. Fibula autografts or allografts can be used additionally to posterior spinal instrumentation. Pedicle screw instrumentation is performed in usually the remaining three to four vertebral body sites (Fig. 14.2a, b). Prior instrumentation, appropriate changes to the surgical table should be made to restore lumbar lordosis. Pedicle screws are placed aggressively to extend to the anterior cortex or even bicortically. Usually, after the sacrum is removed, a hand can be placed ventral to the spine to feel the pedicle screws as they come through to allow for safe bicortical placement. Screws are placed in the remaining ilium, ideally with the double iliac screw techniques. "Docking sites" are placed for fibula strut grafts in the supra-acetabular region. A burr is used to place these from behind. If the level of iliac resection prohibits this, the ischium is usually an appropriate site for docking stations as well. Once this is done, fibula strut grafts are placed as described by Dickey et al. [26], in a "cathedral fashion"; struts are placed in the supra-acetabular region and then end in the last remaining vertebral segment. Appropriate rods are placed after the strut grafts are positioned, and compression is achieved across these to lock the fibula grafts in. If the patient has undergone prior pelvic radiation, consideration is given to using vascularized fibular grafts. This significantly extends the operative time and may require staging to a further day [6, 11].
- *Type 3 and 4 resections*
- Partial and total sacrectomies in conjunction with external hemipelvectomy represent the amputative sacrectomies. These procedures are performed in one stage. Patients undergoing Type 3 resections are considered for an instrumented spinopelvic arthrodesis to the remaining limb if more than 50% of the lumbosacral articulation is resected. The instrumentation can be performed 48 h after the amputation to allow time for final margins to be ascertained and to minimize the physiologic impact on the patient. In Type 4 resections, the resection is performed in a single stage, and the spinopelvic instrumentation between the remaining lumbar spine and

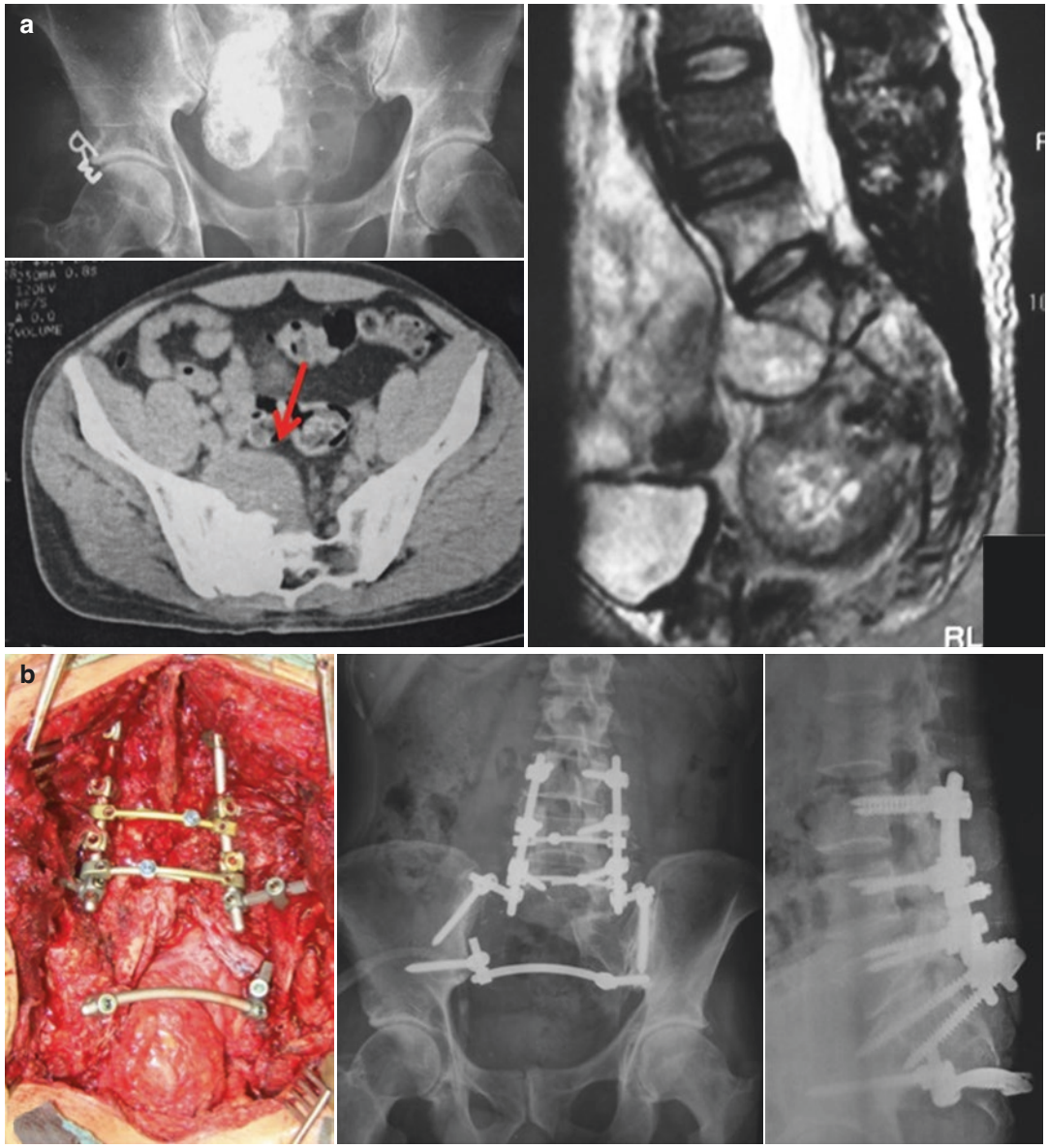

Fig. 14.2 (**a**) A 35-year-old man with a sacropelvic chondrosarcoma. (**b**) Type II resection and spinopelvic reconstruction were done without evidence of local recurrence at 8-year follow-up

remaining limb is in a second stage. Therefore, in Type 4 resections, the tumor-free portion of the amputated femur is ideally stored sterilely in a liquid nitrogen freezer until this second stage of the surgery [29].

- The need for an instrumented spinopelvic reconstruction after a Type 3 resection is controversial. In our experience, if the majority of the lumbosacral articulation is resected, patients likely benefit from instrumented fusion across the spinopelvic junction. This is generally performed in a second stage approximately 48 h after the index surgical procedure. It is usually simple to reopen the wound (and probably advantageous to wash out the inevitable degree of hematoma which develops). Reconstruction is performed using spinopelvic instrumentation from L4 through

the ilium on the retained side. There is usually excellent exposure to perform a discectomy of the remaining disk at the L5–S1 segment and provide an anterior interbody graft at this junction. Depending on the vascular mobilization achieved in the index procedure or desired in the secondary procedure, similar anterior lumbar interbody fusion can be performed at the L4–5 level as well [29].

- In the case of a Type 4 resection, it is necessary to provide reconstruction between the remaining lumbar spine and remaining hemipelvis and limb. Because of the very large magnitude of the oncologic resection, these procedures are staged at least 48 h and oftentimes longer after the index procedure, once the patient has physiologically recovered appropriately. Key aspects of the reconstruction of a Type 4 procedure include centralizing the remaining hemipelvis and limb under the lumbar spine as well as providing a robust autograft strut between the lowest remaining vertebral body and the hemipelvis. At the time of the index resection, a portion of the femur of the amputated limb that is largely free of tumor is saved sterilely in a liquid nitrogen freezer. This provides a strut graft to bridge the gap between the remaining lumbar spine and pelvis on the retained side. Pedicle screw instrumentation is performed into at least the lowest three segments of the lumbar spine on the remaining side. Screw fixation is obtained in the bone stock of the remaining ilium avoiding the hip joint. In performing the reconstruction after a Type 4 resection, two key factors are involved. First, the pelvis should be externally rotated centralizing the remaining lumbar spine over the remaining pelvis such that the patient's center of gravity is relatively uniform. Second, a foraminotomy of the lowest one or two lumbar segments remaining should be performed, in order to avoid too much traction on the lumbar nerve roots to the remaining leg from the previous maneuver. Once instrumentation is in place, the femoral autograft from the resected limb is used as a strut graft between the supra-acetabular pelvis and the remaining lumbar spine. Rods and screws allow for fixation and compression across this graft. An alloderm or similar membrane can be prophylactically used to sequester the abdominal contents away from the instrumentation. Similar to the Type 3 resection, the anterior thigh flap is inserted to close the soft tissue defect. As Type 4 resections commonly involve resection of the anus and genital structures, the amount of skin defect may require the full aspect of skin from the quadriceps flap [29].

14.5 Results

Oncologic results are most favorable when complete resection of the tumor is obtained. It is best illustrated by the data of Fuchs et al. [30], reporting the operative management of sacral chordoma. In a series of 52 patients undergoing surgery, complete survival was seen in all patients in whom a wide margin was achieved at the time of surgery. In contrast, the majority of patients with less than a wide margin resection succumb to disease. Results of more aggressive tumors depended heavily upon the response to chemotherapy. Regarding neurologic function after major sacrectomy, preservation of bilateral S2 nerve roots and a unilateral S3 nerve root or unilateral S2, S3, and S4 nerve roots is required for predictable maintenance of bowel and bladder function [31–33]. In those patients undergoing major spinopelvic reconstruction, a study of 45 patients [chondrosarcoma (n = 11); other sarcomas (n = 11); osteosarcoma (n = 9); chordoma (n = 6); locally invasive carcinoma (n = 5); and others (n = 3)] at mean 38-month follow-up has shown that 28 were living and 17 were deceased; 22 of 28 surviving patients were disease-free and 19 of surviving patients were independent in their activities of daily living; 20 patients required early operation for wound healing; and 16 of these 20 patients had a deep infection; in the patients requiring reoperation, a mean of three reoperations was necessary; 4 patients in this cohort have been revised for instrumentation failure [10]. These results pertain to very large resec-

tions, which disrupt spinopelvic continuity; much fewer complications and more favorable results are seen with lesser sacral resections provided appropriate margins are obtained [34].

The current literature has not focused on pseudarthrosis in the setting of lumbopelvic reconstruction, and the nonunion rate cannot be assessed. Likewise, there is limited data on the mechanical failure rate. In a systematic review by Bederman et al. [12], it was shown that instrumentation failure was evident in 16.1% of patients (5 of 31 patients). Although there was no statistically significant difference, patients without anterior column support tended to have high mechanical failure rates (17.4% vs. 12.5%). This was also shown recently by Tang et al. [35]; in their study, 63 patients who underwent spinopelvic reconstruction following total sacrectomy were studied. Postoperative mechanical failure of the fixation occurred in 25% of patients, and the factors associated with this failure were: single-rod instrumentation with single or double iliac screws; posterior fixation without anterior augmentation; and female gender.

This evidence is suggestive of the potential benefit of adding anterior column support to spinopelvic reconstruction after total sacrectomy, but since extensive instrumentation both anteriorly and posteriorly requires more operative time and more sophisticated techniques, the potential complications must be thoroughly discussed with the patients [5].

14.6 Future Perspectives

So far, there have not been any methods/implants available for total or partial SI joint replacement; thus, all kinds of stabilization are far from the natural biomechanics. Current stabilization techniques try to ensure a stable fixation between the lumbar spine and the pelvis with metal or combined systems [4]. Recently, investigators from China reported the use of a 3D-printed sacral endoprosthesis after total en bloc sacrectomy [36, 37]. In their series, the authors compared the reconstruction with 3D-printed prosthesis (10 patients) to combined reconstruction, including anterior spinal column fixation (14 patients), and spinopelvic fixation alone (8 patients). Compared to the other two groups, the endoprosthesis group had significantly better spinopelvic stability and implant survival with no greater intraoperative hemorrhage or perioperative complications. Authors found also radiological evidence of implant osseointegration at a mean of 7.2 months. However, the study's retrospective design, the small sample size and short follow-up period (mean 22.1 months), and the fact that some patients from the 3D implant group underwent supplemental reconstruction at the time of surgery led to inevitable selection bias that cannot be ignored. Nevertheless, further research and development of novel materials could be the future answer to the treatment of these extremely complex and challenging cases.

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15 Reconstruction vs. No Reconstruction for Pelvic Resections

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15.1 Introduction

Approximately 5–10% of all primary bone tumors are located in the pelvis and pose a challenging problem for tumor orthopedic surgeons [1]. The most common types of pelvic sarcomas are chondrosarcoma, Ewing's sarcoma, and osteosarcoma, while chordoma is the most common primary bone tumor in the sacrum [2]. Although pelvic bone metastases are not rare, most metastatic tumors in the pelvis are managed with radiation and pain palliation, and they do not require surgical treatment. Due to significant difficulties in terms of diagnosis and surgical treatment of pelvic tumors, the survival rate for these patients is much lower compared to malignancies in the extremities [2]. Diagnosis of these lesions on a clinical base is extremely difficult since due to their deep location most pelvic tumors are impalpable, while they can substantially extend without inducing local symptoms. Moreover, the radiographic findings in early stages are not diagnostic, so these tumors are usually detectable on radiographs when they have already invaded an extensive part of the pelvis and have resulted in significant bone destruction or sclerosis. Subsequently, initial detection of pelvic tumors is usually made with advanced imaging including computer tomography (CT), magnetic resonance (MR) imaging, and bone scintigraphy.

As a result of the late diagnosis, the operation for adequate oncologic resection of pelvic tumors with safe margins may be exceptionally difficult. The late diagnosis and wide spread of these tumors at the time of initial detection, the highly demanding procedures for complete resection of pelvic tumors, and the high rate of complications following these procedures are some of the causative factors for the poor prognosis for these patients.

15.2 Examination and Imaging

The main symptom associated with pelvic tumors is a vague, often poorly localized pain around the pelvis that may also radiate downward to the limbs due to compression, irritation, and/or encasement of the peripheral nerve roots and major vessels. A palpable mass that is the second most common symptom of all tumors is absent in most cases, especially during the early stages. These tumors may become palpable only when they have increased in size substantially.

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Radiographs are not diagnostic in most early cases since radiographs have a low sensitivity for pelvic tumors in early stages, especially for the inexperienced physician. In patients with indicative symptoms for a pelvic tumor or when radiographic signs of malignancy are suspected on simple radiographs, further imaging with MR imaging and CT should proceed. Although a complete imaging evaluation of a lesion can confirm or exclude malignancy with enough certainty, bone biopsy is always mandatory.

15.3 Biopsy

Biopsy must always follow the imaging evaluation of a suspicious pelvic lesion in order to confirm diagnosis and identify the histological type of the lesion. Due to high risk for contamination of the surrounding tissue with cancer cells during biopsy, the procedure must be performed only by experienced oncologic surgeons. Unfortunately, several incidents have been described in which an improperly performed biopsy by unexperienced surgeons resulted in inoperability of a tumor that was initially resectable. Contamination of the retroperitoneum with cancer cells during biopsy is a detrimental event that can alter and/or worsen patients' survival [3]. The success of a biopsy depends mostly on the accurate targeting of the involved area and the proper placement of the biopsy tract at this area; therefore, an imaging-guided closed biopsy technique is strongly recommended.

Biopsy can be performed under CT guidance using a 3–6-mm gauge needle (core biopsy technique with a tru-cut® needle). Tumors in the iliac wing can be easily accessed through the iliac crest, whereby the tip of the biopsy needle should follow a subperiosteal route. The biopsy specimen should additionally include a thin layer of the soft tissue that covers the medial aspect of the iliac wing [4]. When a needle biopsy is not diagnostic, we recommend to repeat the closed biopsy technique with imaging guidance and cooperation with the interventional radiologist; if the repeat closed biopsy technique is nondiagnostic, an open biopsy is indicated. For lesions located around the ischium, an open biopsy is usually performed through a Ludloff approach, while for tumors in the pubic rami, an open biopsy can be performed through an anterior approach medially to the neurovascular bundle. Regardless of the biopsy method, the route for accessing the tumor during biopsy must be the same with the route of the surgical approach that will be used for the final resection because the biopsy tract (either closed or open) should be excised with the final tumor specimen.

15.4 Preoperative Management

A multidisciplinary approach is required, as necessary for any malignancy. The multidisciplinary team may include urologists, vascular surgeons, colorectal surgeons, orthopedic surgeons, neurosurgeons, plastic surgeons, and spine surgeons [3–8]. After histological confirmation of the tumor, a surgical staging protocol is recommended using comprehensive imaging evaluation; lung CT, abdomen CT, and a whole-body bone scintigraphy are mandatory in order to investigate whether bone, lung, or intra-abdominal metastases are present (Table 15.1). Moreover, a sonography of the inguinal and para-aortal lymph nodes is helpful for detection of cancerous infiltration as this adverse event can significantly alter the surgical plan and the overall management of the patients. Preoperative evaluation may also

Table 15.1 Imaging studies included in staging protocol for pelvic malignancies

Imaging study	Investigation
Lung CT	Lung metastases
Abdomen CT	Abdominal metastases
Whole-body bone scintigraphy	Remote bone metastases
Sonography of regional lymph nodes	Inguinal and para-aortal lymph nodes infiltration
MR angiography or intravenous pyelography (IVP)	Infiltration of iliac vessels
Pyeloureterogram or cystoscopy	Ureteral or cyst cancerous invasion
Rectoscopy	Rectal cancerous invasion

include MR angiography or intravenous pyelography (IVP) to assess possible invasion of the iliac vessels, to evaluate the anatomic relationship of the tumor with these vessels, and to check the vascular supply of soft tissue flaps that may be used for wound closure. Based on the site and the extent of the lesion, further studies may also be required such as pyeloureterogram, cystoscopy, and rectoscopy. In case of ureteral involvement, consultation by an urologist is necessary, and an ureter stent or a pigtail catheter may be required. Regarding the preoperative laboratory evaluation of patients undergoing tumor resections, 500–1000 cells/ml for absolute neutrophil count and 50,000 cells/ml for platelet count are considered relatively safe cutoff values for an adequate postoperative immunologic response and coagulation of the patients [1]. Due to the significantly increased risk for venous thromboembolism in these patients, several tertiary tumor centers now routinely insert a vena cava filter in all patients undergoing major pelvic resections [9].

Based on the histology of the tumor, neoadjuvant chemotherapy or radiation therapy may be recommended since most pelvic sarcomas such as Ewing sarcomas are sensitive to neoadjuvant treatments for tumor response, facilitation of tumor resection, and improvement of patients' survival. In certain cases, some inoperable tumors may even become operable after neoadjuvant chemotherapy. However, there are types of sarcoma such as chondrosarcomas that are not sensitive to (neo-)adjuvant chemotherapy or radiotherapy. A safety hold-off period of 2–5 weeks after the last chemotherapy session and of 4–5 weeks after the last radiotherapy session is usually recommended for the surgical procedure to be implemented [6]. Restaging after the neoadjuvant therapy should be performed. This new evaluation will set the final margins in three dimensions (sagittal, coronal, and frontal) and six planes (proximal, distal, anterior, posterior, lateral, and medial) for the proper resection of the tumor [3, 4, 6–8, 10]. At this point, using novel technologies, a 3D pelvic model can be designed based on the restaging imaging. This approach allows reevaluation of the planning of resection and the possible reconstruction of the resulting bone defect [11].

15.5 Surgery and Resection Types

Wide resection (resection margins outside the reactive zone of the tumor in healthy tissue) is the recommended type of oncologic resection for pelvic sarcomas. There is a general consent that for a wide resection a healthy surrounding osseous area of approximately 2–3 cm must be included in the resected specimen, while when the tumor invades the surrounding soft tissue, a more extensive area of normal soft tissue of up to 5 cm is required. In general, small pelvic tumors are addressed with partial (limited) pelvic resections, while larger tumors are managed with (1) external hemipelvectomy with hindquarter amputation, (2) internal hemipelvectomy with preservation of the limb, (3) sacrectomy for sacral tumors, and (4) extended hemipelvectomy (external hemipelvectomy with additional resection of the lower lumbar spine). Depending on the resected section of the pelvis and whether this section involves a weight-bearing or moving element of the pelvis, the resulted bone defect may be reconstructed or not.

The techniques for reconstruction of pelvic bone defects are broadly classified into biological procedures such as bone grafting, and nonbiological methods such as endoprosthetic reconstructions. Before the 1970s, almost all tumors involving critical parts of the pelvis were addressed with external hemipelvectomy and amputation. Nowadays, due to the significant advances in the fields of bioengineering, imaging techniques and adjuvant treatments internal hemipelvectomy with some forms of pelvic reconstruction are considered a favorable option without compromising the survival of the patients [1]. Regardless of the method, it is of great significance to ensure a sufficient musculature for wound closure and reconstruction coverage. Although the advances in microsurgical flaps over the past decades have significantly decreased the rate of wound healing complications, these complications with their devastating repercussions are still very common [12–17]. The treatment plan regarding the preoperative management, type of resection, and subsequent reconstruction of the bone defect should be based

Table 15.2 Enneking and Dunham classification for pelvis resections

Resection type	Pelvic region
Type I	Iliac wing
Type II	Periacetabular area
Type III	Pubic rami
Type IV	Sacrum

Femoral resections are designated as follows: H1 for femoral head resections, H2 for peritrochanteric area resections, H3 for proximal femoral resections

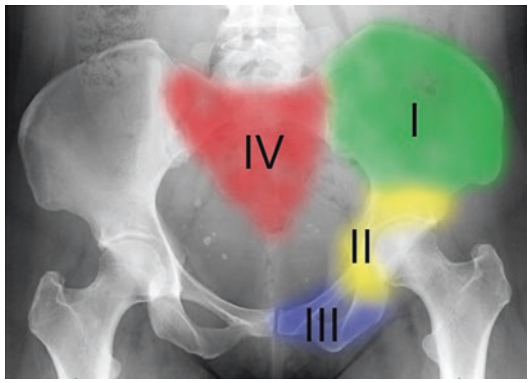

Fig. 15.1 Enneking and Dunham classification of pelvic resections

on consideration of the anatomic location and extent of the tumor, the age and comorbidities of the patients, and the overall prognosis of the malignancy [7, 18, 19].

The most widespread and efficient surgical classification system for pelvic resections is the Enneking and Dunham classification by which pelvic resections are classified into four types (Table 15.2) [20]; Type I resection involves the ilium, Type II the periacetabular area, Type III the pubic rami, and Type IV the sacrum (Fig. 15.1). Several combinations of these types can be made, and a pelvic resection that includes more than one type is defined by the combined number of the individual types (e.g., Type II–III resection). The resection that includes the largest part of the hemipelvis (Type I–II–III) is called hemipelvectomy [21–23]. When part of the femur is also resected, the resection is designated as Type H and is further subclassified into three types: Type H1 when the femoral head is resected, Type H2 when the pertrochanteric area is resected, and Type H3 when the proximal femur is resected [4, 8]. Last, when the tumor not only involves the posterior part of the pelvis and sacrum but also extends to the lower lumbar spine, a more extensive procedure called extended hemipelvectomy is required. This procedure includes external hemipelvectomy and resection of ilium, sacrum, and part of the lower lumbar spine. Certain details about the surgical approach, the extent of resection, and the methods for reconstruction of the bone defects must be set out based on the Enneking and Dunham classification.

15.6 External Hemipelvectomy Versus Limb-Salvage

The decision about lower limb amputation versus limb-salvage surgery is complex, and surgeons are called to decide whether an adequate wide resection is feasible without ending up with a severely compromised lower limb. When surgeons decide to proceed with limb-salvage surgery, the postoperative functional results should always be aimed to be superior to those of an external hemipelvectomy [21, 24–27]. Although there are several well-defined prerequisites that must be fulfilled for a limb-salvage procedure, in many cases, fulfillment of all these criteria is debatable. The main contraindications for a limb-salvage procedure include (1) recurrence of the tumor after a previous limb-salvage procedure unless a wide resection can be definitely achieved and the overall status of the patients allows for a new limb-sparing procedure, (2) extension of the tumor in the sacral foramens and involvement of the nerve roots, (3) extension of the tumor in sciatic notch and involvement of the sciatic nerve, and (4) infiltration of iliac vessels resulting in nonrepairable vascular lesions (Table 15.3). Although femoral involvement or extensive infiltration of the psoas muscle is not considered contraindications for internal hemipelvectomy, the pros and cons of such a major procedure with a significantly compromised lower limb afterward must be considered with caution. According to O'Connor and Sim the acetabulum, the sciatic nerve and the femoral neurovascular bundle are

Table 15.3 Indications for external hemipelvectomy

Indications
Extension of the tumor in the sacral foramens and involvement of the nerve roots
Extension of the tumor in sciatic notch and involvement of the sciatic nerve
Infiltration of iliac vessels resulting in nonrepairable vascular lesions
Recurrence of the tumor after a previous limb-salvage procedure (unless a wide resection can be definitely achieved and the overall status of the patients allows for a new limb-sparing procedure)

three major anatomic elements of the pelvis, and when two of these three must be resected, external hemipelvectomy should be considered [28].

15.6.1 Type I Resections: Os Ilium

Tumors in the iliac wing can be accessed through the posterior part of the ilioinguinal approach; a simpler approach through an incision along the iliac crest is usually adequate for smaller tumors in the iliac crest. The resection of these tumors is achieved through two osteotomies, one proximal and one distal. The distal iliac osteotomy is at the supra-acetabular level and can be carried out through the sciatic notch, while the proximal iliac osteotomy is usually adjacent to the iliosacral joint. On the proximal osteotomy site, the iliolumbar ligament that marks the position of the L5 root (the root is located inferior and medially to the ligament) must be identified and usually needs to be cut for adequate exposure. In some cases, the proximal extent of the tumor is too close to the sacroiliac joint, and a wide resection can only be achieved through disarticulation of the sacroiliac joint [3, 4, 6–8]. The subsequent reconstruction of the bone defect following Type I resections depends on the integrity of pelvic girdle (Table 15.4).

15.6.1.1 Partial Resection of the Ilium Without Pelvic Ring Disruption

When iliac resection is only partial and does not disrupt the pelvic continuity as with tumors located in the iliac crest, reconstruction is not required. Similar to these tumors, more distally located tumors that are small in size and can be adequately resected without disrupting the continuity of the iliac wing can be also solely resected.

Table 15.4 Reconstruction of bone defects after Type I resections

Type I resection	Reconstruction
Partial iliac resection without pelvic ring disruption 1. Iliac crest tumors 2. Small tumors of the iliac wing	Resection without reconstruction
Complete iliac resection with pelvic ring disruption	
1. Tumors involving the largest part of the ilium 2. Tumors extending to sacroiliac joint	Osseous reconstruction with autograft or allograft Sacroiliac arthrodesis with grafts

15.6.1.2 Complete Resection of the Ilium with Pelvic Ring Disruption

Bone defects after pelvic resections that involve the largest part of the supra-acetabular ilium or extend to the sacroiliac joint destabilize the pelvic ring and usually lead to certain functional deficits. In most of these cases, bone defects are managed with some form of bone reconstruction or arthrodesis (sacroiliac arthrodesis) to support the two sides of the resection (ilium–ilium or sacrum–ilium) and to provide adequate pelvic ring stability. This can be achieved with grafts, metallic implants, or a combination of both. Usually, a tibial or a fibular autograft is harvested and placed at the resection site in order to bridge the defect. The one side of the graft is fixed at the supra-acetabular osteotomy site, while the other at the proximal iliac osteotomy or at the sacral osteotomy. In case of a very large bone defect, massive allografts can also be used.

The functional outcomes after these reconstructive procedures are reported to be excellent, while complication rates are low. Although reconstruction for these defects is recommended by most authors, Beadel et al. compared the results with and without reconstruction of bone defects after iliosacral resections (4 and 12

Table 15.5 Reconstruction of bone defects after Type II and Type II–III resections

Reconstruction	Techniques
Nonreconstruction	1. External hemipelvectomy 2. Iliofemoral arthrodesis 3. Resection arthroplasty 4. Hip transposition technique
Reconstruction	1. Biological reconstruction (a) Bulk pelvic allograft (b) Autoclaved or irradiated pelvic autograft 2. Endoprosthetic reconstruction (a) Saddle prosthesis (b) Stemmed cup Reverse ice-cream cone implant Pedestal cup (c) Custom-made implant (d) Allograft/prosthetic composite

patients, respectively) and suggested that reconstruction to restore pelvic stability after sacroiliac resections is not always necessary [29]. Interestingly, in this case–control study, although the functional scores (Toronto Extremity Salvage Score and Musculoskeletal Tumor Society), the rates of local recurrence, and survival were similar between the groups, patients without reconstruction had a lesser need for walking supports, required less pain medications, and were more likely to return to work [29]. Therefore, any reconstruction should be considered with caution.

15.6.2 Type II Resections: Periacetabular

For tumors located around the acetabulum with or without extension to the proximal femur, a Type II pelvic resection including the acetabulum is warranted. This type of resection is a highly demanding procedure, and good functional outcomes necessitate restoration of the native hip biomechanics. The hip center of rotation and the force transmission along the pelvic ring must be restored so that the weight-bearing pattern throughout the gait cycle is not altered [20]. For this type of resection, three osteotomies are performed. The superior osteotomy at the supraacetabular area is usually performed through the greater sciatic notch, the anterior osteotomy is performed at the anterior acetabular column around the base of the upper pubic rami, and the posterior osteotomy is at the proximal part of the posterior acetabular column or more distally at the ischium. Both reconstruction and nonreconstruction options, and biological methods, and endoprosthetic options are available for the management of the periacetabular bone defects (Table 15.5).

15.6.2.1 Nonreconstruction Methods: Iliofemoral Arthrodesis

Iliofemoral arthrodesis was a very popular method for Type II or Type I–II resections during the previous decades. By this type of arthrodesis, the stability of the pelvic ring is maintained, while the patients preserve a certain degree of a painless limb function [30]. The disadvantages of iliofemoral arthrodesis include loss of joint function, constant use of gait support, limb length discrepancy, and long consolidation times [31, 32]. The functional results of this operation are significantly better when the proximal iliac osteotomy is close to the native acetabulum so that limb shortening is minimized and successful fusion is more likely. However, the attempt to maintain a large part of the ilium may compromise safe resections margins and result in an increased risk for local recurrence. For the fusion between the proximal femur and the remaining pelvis, a combination of hardware can be used including plates, cerclage wires, and cables [33–35]. Moreover, the fusion can be augmented with bone grafts that are fixed along the fusion site.

Commonly used autografts include the iliac crest graft and the free fibula graft, either avascular or vascularized. Although the use of the free vascularized fibula graft increases the complexity of the procedure and requires microsurgical skills, the reported union time with this technique is significantly decreased and stability of the pelvic ring is enhanced [36–39]. In cases that part of the pubis or the ischium needs to be resected as well (e.g., Type II–III resections), additional fusion of the femur to the ischium or to the pubic bone is recommended [8, 40]. Last, another fusion technique that can be used in selected cases is the tibia–hindfoot rotationplasty with calcaneopelvic arthrodesis. This technique involves partial resection of the pelvis, resection of femur, and preservation of the lower limb below the knee. The remaining lower limb is rotated 180°, the midfoot and forefoot are resected, and the calcaneus is fixed to the pelvic osteotomy site [35, 41].

There is a large body of evidence regarding the results of iliofemoral arthrodesis. One of the first large studies for this technique included 60 patients with iliofemoral fusions. The results in this study were disappointing since pseudarthrosis occurred in 50% of the patients [28]. In another study by Fuchs et al. enrolling 32 patients who treated by this method, solid fusion was done in 21 patients, while primary pseudarthrosis with adaption of the femur to the ilium was done in 5 patients [42]. In the group of patients with primary iliofemoral arthrodesis, failed fusion and pseudarthrosis were observed in 14% of the cases. In terms of functional results, patients with primary pseudarthroses reported significantly lower values in the Musculoskeletal Tumor Society Score (MSTS) than those with solid fusions. All patients in this study regardless of the success of the fusion reported mild low back pain, probably due to altered forces on their lumbar spine. The average leg length discrepancy (LLD) in this series was 4.8 cm that was compensated with shoe lifts. The authors concluded that iliofemoral arthrodesis should be preferred in young patients with more demanding functional activities, while primary pseudarthrosis must be performed in older patients. In a smaller study evaluating the results of this method, the authors reported satisfactory results regarding LLD (average, 2 cm) and postoperative functional status [39]. In another study, the total cost of three different methods (external hemipelvectomy with use orthotic device, internal hemipelvectomy with endoprosthetic reconstruction, and iliofemoral arthrodesis) for the management of bone defects after oncologic pelvic resections was compared; iliofemoral arthrodesis was clearly associated with a significantly lower cost compared to the other two methods [43].

15.6.2.2 Nonreconstruction Methods: Resection Arthroplasty

Similar to the Girdlestone procedure, the method of resection arthroplasty (or flail hip technique) after oncologic pelvic resections is an alternative option that was also very popular in the early days of pelvic tumor surgery and is gaining support again [44]. As with any limb-salvage procedure, this method is indicated only when wide resection is possible without significant compromise of the lower limb function. According to this technique, the involved acetabular area is solely resected resulting in a flail hip without a supporting pelvic articular surface. The aim of this surgery is to establish a fibrous union between the remaining ilium and the femur as opposed to arthrodesis in which an osseous union is aimed. As expected, this surgery is associated with significantly limb shortening which can be addressed with either shoe blocks in mild cases or distraction osteogenesis in more severe cases. Even though unassisted ambulation has been reported for patients treated in this fashion, supporting walking aids are usually required [45]. The functional results of resection arthroplasty are greatly depended on the extent of the resected ilium [46]. Although this method is not very common today, the low rates of complications in terms of wound healing problems and mechanical failures have led to a renaissance of this method during the past years, especially in severely compromised patients.

Schwartz et al. evaluated the capacity for independent walking and the functional outcomes after resection arthroplasty in eight patients, with a 2.9 years minimum follow-up

[45]. The authors reported excellent results; at the last follow-up, seven of the eight patients were free of pain and able to walk without supports, while the overall mean MSTS score was 73.3% (range, 53.3–80.0%).

15.6.2.3 Nonreconstruction Methods: Hip Transposition Technique

In the hip transposition technique, the hip joint is transferred cranially to the level of the proximal osteotomy. The inferior part of the acetabulum (in cases that it can be preserved) is rotated 90° and fixed with soft tissue or textile implants at the proximal osteotomy site in order to form a pseudo-joint. If the acetabulum is completely resected, the femoral head can be wrapped into an artificial capsule that is attached with bone anchors to the remaining ilium [47]. Even though this technique has fairly good functional results, limb shortening as with any nonreconstruction technique is a significant drawback [48]. Distraction osteogenesis for the management of the resulting LLD has been recommended by many surgeons [49]. Revision surgery after the hip transposition technique is rarely reported, and the rates of postoperative complications are significantly lower compared to more advanced methods for reconstruction of bone defects such as the endoprosthetic reconstruction and the use of structural allografts [50, 51]. Hillman et al. reported the outcomes of hip transposition in 17 patients after Type II, Type I–II, Type II–III, and Type I–II–III resections [50]. LLD was evident in four patients. Although three of these patients (18%) had postoperative skin problems, the overall complication rate was very low. The authors highlighted that no incidence of deep infection or local recurrence occurred in this series.

15.6.2.4 Reconstruction Methods: Graft Implantation

Biological reconstruction of the bone defect refers to application of a structural bone graft for substitution of the resected bone segment. This graft may be either the resected pelvic segment reimplanted as an autograft or a pelvic massive allograft [10, 52–58]. Since allografts can be shaped intraoperatively to copy the complex geometry of the corresponding pelvic defect, in theory they can be used to reconstruct the normal pelvic anatomy after almost any type of resection. In contrast, a prerequisite for the use of the resected bone as an autograft is tumor denaturation that can be achieved through several techniques including autoclave, pasteurization, freezing, and extracorporeal irradiation. Following the process of tumor denaturation with these techniques, the graft is reimplanted to fit into the defect. This method has certain advantages compared to allografts such as the easy accessibility of the graft since bone bank is not required, and the optimum fit of the graft into the defect [59–61]. Moreover, there is no need for immunosuppression (which decreases the potential for bone healing), while the risk of infection is also lower [58, 62, 63]. Compared to nonreconstruction methods, biological reconstruction has the obvious advantage of preservation of the hip joint function, while compared to endoprostheses, reattachment of the surrounding musculature is easier [64]. However, grafts have been linked with high rates of serious complications such as infections or mechanical complications such as failure of consolidation [2, 10, 56].

In a large study of 945 patients investigating the factors influencing graft consolidation, the nonunion rate was reported 17.3% [65]. A significant risk factor for failure of consolidation was the suboptimal fit of the graft into the defect and the subsequent large interfragmentary gaps. The overall infection rate of allografts ranged from 10% to 33% [28, 66]. Some authors recommend impregnation of the grafts with antibiotics such as rifampicin in order to reduce infectious complications [67]. Dellove et al. in their study using structural bone allografts after periacetabular oncologic resections in 24 patients reported a high rate of complications (11 patients, 46%), requiring a revision surgery in all cases [68]. Regarding the functional outcomes in this study, authors reported an average MSTS score of 73%, but only half of the patients were capable for independent walking without crutches. The authors also highlighted that the functional outcomes were clearly better for the younger patients (average MSTS score 82% in patients <20 years

old compared to average MSTS score 65% in the elderly) [68]. In an another study of 18 patients with irradiated autografts after periacetabular pelvic resections, the authors reported good functional results with an average MSTS score of 73% and an average TESS of 71% [69]. The authors also reported that three revision surgeries were performed in three patients with deep infections; although graft union was difficult to identified, there was no evidence of nonunion that required a revision surgery [69].

15.6.2.5 Reconstruction Methods: Endoprostheses

Endoprostheses for periacetabular reconstructions are the first option in most practices with many different types of endoprostheses developed over the last decades [57, 70–78]. A solid and stable fixation of the prosthesis into the remaining pelvis is a prerequisite for successful outcomes. Also, in order to increase hip joint stability and decrease dislocation rates, large femoral head sizes have been recommended, while the peripelvic musculature including the gluteal fascia must be preserved and reattached to femur when possible [8]. Until the development of a pseudocapsule around the prosthetic joint, artificial ligaments can also be used to augment stability and musculature reattachment, although their efficacy is questioned. Last, for the restoration of the native hip biomechanics the final position of the acetabular cup must be symmetrical with the opposite site in terms of height, lateral distance, and orientation [79].

Saddle Prosthesis

Saddle prosthesis is one of the first implants that was developed for reconstruction after Type II and Type II–III pelvic resections. These implants were initially developed and used for reconstruction of large pelvic defects after failed total hip arthroplasties or after resections in infected reconstructions [73]. By the late 1990s though, there was already a sufficient body of evidence regarding the use of these implants after oncologic pelvic resections [72]. For the insertion of this implant, a notch is created in the remaining ilium and the proximal part of the prosthesis that simulates a saddle hinges over this notch [70]. Initial results of this endoprosthesis in terms of mechanical complications such as loosening, hip dislocation, and intraoperative or postoperative fractures were very discouraging. A major cause for these poor results was the final eccentric position of the artificial joint. The high rates of complications led to many modifications of these implants; newer implants had the advantage of modularity, offering more options in terms of restoration of the native hip biomechanics [80].

Aboulafia et al. studied the outcomes of saddle prostheses in 17 patients with pelvic tumors [70]. Functional results in their study were reported to be excellent or good in 12 patients and fair or poor in 5 patients. Dislocation rate was high (53%), whereas other complications included wound healing problems and infections. Implant loosening and cranial migration of the prosthesis were also frequently reported in the long term. Some of the critical points for improved functional outcomes for these implants were preservation of a sufficient iliac notch and a proper selection of the implant length. In this way, the muscle tension of the iliopsoas and the hip abductors is restored and the stability between the pelvis and femur is increased. In line with this, poor iliac bone stock and suboptimal status of the psoas and the hip abductor muscles should be considered contraindications for the use of these implants.

Custom-Made Pelvic Prosthesis

Due to the high rates of infection following the use of allograft reconstructions and the need for improved functional results, modular custom-made endoprotheses were widely distributed in the middle 1990s for reconstruction of periacetabular bone defects [2, 50, 78]. For the generation of such implants, a preoperative CT of the pelvis is required, and a 3D pelvic model is formed onto which the resection margins are simulated and marked and a modular custom-made pelvic endoprosthesis is subsequently manufactured. The theoretical advantage of this method is that since the endoprosthesis matches exactly to the defect and replaces with high precision the resected bone segment, the native hip function is suppos-

edly completely restored. To achieve that though, the intraoperative resection must be copied as much as possible to the predesigned resection on the pelvic model. Despite the initial enthusiasm and the excellent functional results, the long-term outcomes were not as expected as these bulk implants were associated with certain complications including loosening, migration, and implant failure. Moreover, the large dead space that was created after insertion of these implants resulted in high rates of hematoma formation and postoperative infection. The high rates of these complications led in the late 2000s to a significant decline in the use of these implants [2]. However, due to the precise preoperative planning of the resection margins and the extensive pelvic resection that is required for these implants, these modular endoprostheses were associated with decreased rates of local tumor recurrence [59].

There are many series about custom-made prostheses after oncologic pelvic resections [71, 74]. Hillman et al. reported the outcomes and complications of several reconstructive techniques after pelvic resections [50]. In 16 patients, a custom-made prosthesis was inserted and the complication rate was significantly high (62.5%). The infection rate was 38%, while local recurrence occurred in 3 patients, and 1 patient presented with wound healing problems. The authors noted that 30 additional procedures were required in these patients (average 1.9 revision operations per patient).

Stemmed Acetabular Prostheses (Cups)

Although saddle prostheses and custom-made implants have been used for many years in pelvic tumor surgery, due to the high complication rates and the high cost of these implants, there was a growing skepticism about their use [81, 82]. To overcome these concerns, a different type of endoprostheses that was initially developed and used in revision surgery after failed THA gained ground over the past years [75, 83]. These acetabular prostheses were called stemmed cups and are composed of an inferior shell and a large superior stem that is fixed proximally into the intramedullary space of the ilium aiming to the posterior superior iliac spine or the sacroiliac joint. Since the structure of the iliac isthmus is similar to that of a long bone with a thick outer cortex and a dense trabecular medulla, the rationale for the use of these implants is that their stem that is inserted into the ilium will provide the required rigid fixation of the implant to the remaining pelvis. The two main types of stemmed cups are the reversed "ice-cream cone-style" prostheses and the pedestal cups [84, 85]. Although the main principles involved in the design of these two implants are similar, the newer types of pedestal cups such as the LUMiC® endoprosthesis are more versatile regarding their stem length, orientation, and diameter of the cup. These endoprostheses require only a small cranial part of the ilium, while the proper orientation of the stem restores load transmission along the iliosacral joint; therefore, the anatomical and biomechanical continuity between the spine and the lower limb is undisrupted. There are several other advantages that are associated with these implants such as the fact that restoration of the continuity of the pelvic ring is not necessary since the weight-bearing load is transferred to the socket and the high versatility regarding the stem length and orientation of the cup. This versatility provides many options for restoration of the native hip center of rotation and limb length. Moreover, due to the small size of these implants, the resulted dead space is substantially decreased, and soft tissue coverage is more easily achieved, if necessary. Last, these implants are commercially available in a wide variety of stem length and cup orientation; thus, there is no need for preoperative time for planning the construction and manufacturing as with custom-made implants. Secondary to these features, there is also a significant decrease in complications such as hematoma formation and infection [83]. However, despite the precise restoration of the native hip biomechanics, the functional results of these implants widely vary.

Pedestal cups were initially developed for revision surgery in patients with failed THA. There are only few studied in the literature regarding the use of these cups after oncologic periacetabular resections [80, 86–88]. In the largest study of 48 patients with pelvic tumors and a 6-year follow-up, Hipfl et al. [86] reported the

outcomes of a pedestal cup (Schoellner cup; Zimmer Biomet Inc., Warsaw, Indiana) for oncologic pelvic reconstruction. The complication rate in this study was high, including deep infection in 17% of patients, hip dislocation in 15% of patients, and aseptic loosening in 6% of patients. Also, the survival rate of these cups at 1- and 5-year follow-up was 72% and 51%, respectively. Bus et al. also evaluated the same pedestal cup (Schoellner cup; Zimmer Biomet Inc., Warsaw, Indiana) after oncologic pelvic resections in 19 patients with a mean follow-up of 7.9 years [84]. Although the 5-year failure rate was similar to that of Hipfl et al. (50%), there were no revisions for mechanical reasons and all revision cases were due to periprosthetic infections. Complications rate was also high in another study by Bus et al. that evaluated the LUMiC® pedestal cup for periacetabular reconstruction after oncologic resections [89]. In a total of 47 patients, 30% experienced at least one complication. Particularly, infections occurred in 13 patients (28%), while 10 patients (22%) had at least one dislocation. The authors highlighted that when dual mobility cups were used the dislocation rate dropped to 4%. The cumulative failure incidences for the LUMiC® endoprosthesis in that study at 2 and 5 years were 19.4% and 26.5%, respectively [89], much lower than those reported in the study of Hipfl et al. [86].

There are also limited data regarding the use of reversed ice-cream cones-style implants for periacetabular reconstruction after oncologic pelvic resections [90, 91]. The newer types of ice-cream cone implants are basically modifications of the McMinn implant (LINK, Hamburg, Germany) that as the pedestal cups was initially developed for reconstruction of bone defects after failed THA. Barriento-Ruiz et al. in a recent study reported the outcomes of two ice-cream cones-style implants (Coned®, Stanmore Worldwide Ltd, Elstree, UK; and Socincer®, Gijon, Spain) in ten patients after Type II pelvic resections with a median follow-up of 3 years [85]. Infections occurred in four patients (40%), two of which were superficial wound infections and the two were deep infections, and dislocation occurred in one patient (10%) during the follow-up. Interestingly, the authors reported that none of the ten patients required revision surgery for implant removal; thus, the failure incidence at 2 years was 0% [85]. In another study by Fisher et al. ice-cream cones-style implants were inserted in 27 patients after periacetabular oncologic resections [92]. Compared to the previous study, while dislocation rate was almost similar (14.8%), the infection rate was significantly lower (11.1%). During the follow-up (mean, 39 months, range, 18–80 months), only 1 patient required implant removal due to loosening. In another recent study enrolling 24 patients with periacetabular oncologic resections who underwent reconstruction with an ice-cream cone-style implant, at least one complication occurred in 58% of the patients [93]. These complications included deep infection (17%), dislocation (18%), and mechanical failures (8%). At 5-year follow-up, implants survival in this study was 75%.

Allograft/Prosthetic Composites

Another option for reconstruction of periacetabular defects is the use of a composite structure including a conventional acetabular prosthesis surrounded by a bone graft. The theoretical advantage of this technique is that the additional use of the graft will allow for reconstruction of larger bone defects, while the conventional prosthesis will restore hip range of motion resulting in good functional results. The bone graft that is attached to the prosthesis can be either the resected bone segment used as an autoclaved or irradiated autograft or a bank bone allograft [52, 94–100]. Autografts have the advantage of the optimum fit to the defect, while they are easily accessible if the oncologic center is properly equipped and a bone bank is available. Although this method yields good functional results, the use of an autograft or an allograft has in long term all these complications that are associated with bone grafts such as infection, nonunion, loosening, and fracture. The process for tumoral necrosis such as irradiation and heating for the autografts compromises the bone quality of the grafts that are major risk factors for graft fracture and consolidation failure [97]. The reported infection rates after these operations range from

15% to 50% [50, 52, 54]. The wide range in this rate is probably due to the varying extent of soft tissue and bone resection and the subsequently varying extent of the resulting dead space.

15.6.3 Type I–II Resections

Tumors that involve not only the acetabular/supra-acetabular area but also extend to the largest part of ilium (resection Type I–II) are common, and their management requires cautious preoperative planning and a more ablative procedure. A critical factor in terms of managing the bone defects is whether an adequate cranial part of the ilium is preserved (Table 15.6). The nonreconstruction options for Type I–II resections are similar to those for Type II resections including iliofemoral arthrodesis (or sacrofemoral arthrodesis), resection arthroplasty, and hip transposition. When an adequate cranial part of the ilium is preserved, the reconstruction options are also similar to those described for Type II resections including biological reconstructions and endoprosthetic reconstructions with saddle prostheses, stemmed cups, and custom-made prostheses. If the cranial part of the ilium is not adequate, endoprosthetic reconstruction is possible only with custom-made prostheses, while structural bone grafts (either a structural allograft or an autograft) are an alternative option [10, 12]. Allografts can be used for reconstruction following almost any type of pelvic resections, and although they have the advantage of good functional results and restoration of native hip biomechanics (at least during the initial postoperative period), the high rates of complications such as loosening, infection, and fractures remain an unsolved problem. Therefore, due to their high complication rates and their limited availability, structural pelvic allografts for such extensive defects are not very popular.

Table 15.6 Reconstruction of bone defects after Type I–II and Type I–II–III resections

Reconstruction	Techniques
Nonreconstruction	1. External hemipelvectomy 2. Iliofemoral or sacrofemoral arthrodesis 3. Resection arthroplasty 4. Hip transposition technique
Reconstruction	
1. Biological reconstruction	1. Bulk pelvic allograft 2. Autoclaved or irradiated pelvic autograft
2. Endoprosthetic reconstruction (a) Adequate preservation of cranial iliac part	1. Saddle prosthesis 2. Stemmed cup Reverse ice-cream cone implant Pedestal cup 3. Custom-made implant
(b) Inadequate preservation of cranial iliac part	Custom-made implant

15.6.4 Type III Resections: Ischiopubic Rami

Type III resection is required for tumors around the pubic and ischial rami. For this type of resection, two pelvic osteotomies are performed. The medial level of resection is through the pubic symphysis or in some cases through the opposite pubic rami. The lateral osteotomy is performed medially to the acetabulum. In some cases, the obturator nerve with the neighboring obturator vessels must be sacrificed due to their close proximity to the tumor. Tumors of the upper or lower pubic ramus can be solely resected without further management of the bone defects since this type of resection does not disrupt the native pelvic ring stability. An important part of this procedure is the cautious reconstruction of the soft tissue envelope in order to prevent herniation of the bladder or intestines through the defect. Reconstruction of the inguinal floor is mandatory to prevent peritoneal herniation [101, 102]. Augmentation of the soft tissue reconstruction with a synthetic mesh or fascia lata allograft is commonly performed.

15.6.5 Type II–III Resections

When tumors involve the upper or lower pubic rami with additional extension to the acetabulum, a more extended approach compared to a Type II resection is required. While the medial level of the resection is similar to that of a Type II resec-

tion, the lateral resection must be extended proximally to the supra-acetabular level (Type II–III). The resulting bone defect after this type of resection can be addressed with any of the methods that have been described for Type II resections.

15.6.6 Type I–II–III–IV Resections

In some cases, the tumor extends to include not only the largest part of hemipelvis, but part of the sacrum as well. In these cases, complete resection of the hemipelvis must be performed either as an external or an internal hemipelvectomy. The indications for external hemipelvectomy and lower limb amputation versus internal hemipelvectomy have already been discussed previously.

The main difference compared to Type I–II or Type I–II–III resections is that there is no remaining proximal iliac wing; therefore, the reconstruction and nonreconstruction methods that are available for the management of the resulted bone defect are similar to those of Type I–II resections when no adequate cranial part of the ilium is preserved. Nonreconstruction options include resection arthroplasty, sacrofemoral arthrodesis, or a hip transposition, while reconstructive options include custom-made implants or allografts. Allografts have the advantage of reattachment of the pelvic musculature to the graft, which in theory leads to improved postoperative functional results. Custom-made prostheses are mega-implants that replace the patients' hemipelvis and are associated with high rates of loosening and migration. Spontaneous periprosthetic heterotopic ossification around these implants can develop that is associated with lower rates of loosening and dislocation. In most cases, an external hemipelvectomy is performed at the index surgery or after local tumor recurrence or complicated reconstructions for these extensive tumors.

15.6.7 Type IV Resections: Sacrum

The surgical approach for a sacrectomy can be either anterior or posterior, while for large proximal tumors that extend to the lumbopelvic junction or for tumors with extensive presacral soft tissue invasion, a combined approach may be required. From the anterior approach, the tumors can be accessed either through a transabdominal route or a retroperitoneal route. Depending on the tumor's extension into the sacrum, sacrectomies can be either partial or total (Table 15.7) [8, 22, 23, 103]. Partial resections may be transverse, sagittal, or a combination. Lateral sacral tumors are addressed with a sagittal partial sacrectomy, whereas midline tumors are addressed with a transverse partial sacrectomy. For tumors located below the S2 level, a partial sacrectomy is usually adequate for wide resection of the tumor. In such cases, the lumbosacral joint is preserved, while bladder and bowel function are usually not affected [104, 105]. Conversely, for tumors that develop around the proximal part of the sacrum with anterior expansion, total sacrectomy is required. In total sacrectomies, the adjacent nerve roots are sacrificed in order to achieve a wide-margin resection. These operations are associated with severe neurological dysfunction and a high rate of wound complications, although the use of the transpelvic vertical rectus abdominis myocutaneous flap has significantly decreased the latter [106–112].

Resections for tumors below the S1 level do not result in pelvic ring and lumbosacral instability since the sacroiliac joints and the lumbosacral junction are left intact. On the other hand, sacrectomies that require S1 resection disrupt the lumbosacral and iliosacral junctions, lead to instability, and necessitate some form of spino-

Table 15.7 Reconstruction of bone defects after Type IV resections

Type IV resection	Reconstruction
Partial sacrectomy <50% of the sacroiliac joint on each side (e.g., tumors below S1)	Sole resection
Total sacrectomy	
1. Tumors involving the largest part of sacrum 2. Proximal sacral tumors	Resection and spinopelvic stabilization Resection without stabilization ("spine on a biological sling")

pelvic stabilization [113–122]. Without stabilization, after a total sacrectomy, the lumbar spine usually migrates downward and remains between the iliac bones. The inferiorly migrated lumbar spine is maintained at this new position by a "biological sling" that is formed by the surrounding musculature and the developing scar tissue. Although patients commonly describe back or leg pain, walking is possible with the use of a brace [119, 120, 123]. Therefore, some authors do not recommend bone reconstruction after total sacrectomies since the postoperative ambulatory status is acceptable and the rate of complications such as surgical site infections is significantly lower compared to more complex skeletal reconstructions [120–122]. However, according to most authors, the indications for lumbopelvic stabilization include a total sacrectomy and a partial sacrectomy that involves at least 50% of the sacroiliac joint on each side [124–126].

Several different techniques using variable types of implants such as plates, screws, wires, and bars have been described for spinopelvic stabilization after major spinopelvic resections. In cases of partial sacrectomies, spinopelvic fusion can be achieved with a combination of bars or screws into the sacrum and rods (Zimmer, Inc., Warsaw, Indiana) with pedicle screws into the lumbar vertebrae. Otherwise, in cases of total sacrectomies, a commonly used technique for spinopelvic fusion is the Luque–Galveston technique, in which the pelvis is engaged into the fusion with bars or screws into the iliac bones bilaterally. Many modifications of spinopelvic fusion have been developed over the past years [113, 127, 128]. Although the newer implants may achieve a more rigid fixation to the pelvis, the issue of a robust proximal lumbar fixation has still not been solved. Currently, the pedicle screw–rod construct is the most common instrumentation system for spinopelvic fixation since they are easily inserted and provide a more rigid fixation compared to other implants such as hooks or wires. As a general consent, at least the three lower spinal segments must be included in the fusion.

15.7 Extended Hemipelvectomy

When pelvic tumors expand to the lower lumbar vertebrae, an extended hemipelvectomy with additional resection of the lower lumbar spine is required. Since this surgery leads to spinopelvic dissociation, the bone defect after an internal extended hemipelvectomy should be reconstructed [124, 129]. Many authors recommend that internal extended hemipelvectomy must be preserved only for patients with localized disease, while it should not be performed in cases when prior surgery in the same area has been performed due to the increased risk for local recurrence and complications [8, 129]. Additionally, given the high rate of failure and the increased morbidity that is associated with extended internal hemipelvectomy, this surgery is not indicated for compromised patients or for patients with metastatic bone disease.

Although the oncologic outcomes after internal or external (extended) hemipelvectomy do not significantly differ, the postoperative function of the lower limbs after internal extended hemipelvectomy is very poor due to resection of the lumbosacral plexus [129]. In reality, amputation of the lower limb (extended external hemipelvectomy) is preferred in almost all cases and internal extended hemipelvectomy is very rarely performed [22, 23]. After an extended external hemipelvectomy, a spinopelvic fusion for preservation of the function of the contralateral limb is required. Part of the resected femur of the amputated limb such as the femoral condyles can be used as a strut autograft for augmentation of this fusion [22].

15.8 Conclusions

Whether an external hemipelvectomy or a partial pelvic resection should be performed is a decision that should be made by the surgeon based on several factors such as the underlying diagnosis, the patients' medical status, the tumor's characteristics, and the correlated expected survival. It has been proven that with proper patient selection and proper surgical technique internal hemipel-

vectomies do not carry a higher risk for recurrence compared to external hemipelvectomy, with similar survival rates [19, 130]. On the other hand, external hemipelvectomy and amputation have a lower incidence of complications and a faster recovery time compared to partial pelvic resections. Also, even though amputees experience certain limitations in daily activities such as walking, rising and sitting down or climbing chairs, they can have a relatively high level of activities [131]. Reconstruction of the bone defect must be considered on a case-by-case basis as it is a complex issue with no strong guidelines. The available reconstruction techniques provide better functional results over non-reconstruction since hip and lower limb functions are preserved, but at the expense of a higher complication rate [3, 5–7, 10, 70, 132].

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16 The Hip Transposition as a Reconstructive Technique After Pelvic Resection

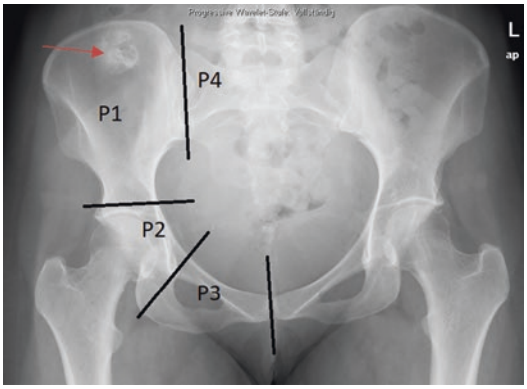
Timo Lübben and Georg Gosheger

16.1 Introduction

Resection of pelvic tumors is one of the most challenging surgical procedures in tumor surgery. Wide resection following the definition of Enneking [1] is to be considered standard of care for most malignancies. Limb salvage can be achieved in most of the cases of pelvic bone tumors without narrowing the margins of the resection [2]. While endoaprosthetic replacement and allograft/autograft reconstruction suffer severe postoperative problems due to infection and poor soft tissue coverage [3–9], hip transposition, although far from perfect, can be considered a functionally good and long-lasting surgical alternative [10].

16.2 Indication

Hip transposition can be the procedure of choice for Enneking type P2-, P1-2, P1-3, and P1-4 resection of the pelvis. Alternative operative procedures, i.e., endoprosthetic replacement, in the case of P2-/P2-3-resection should be discussed.



ap x-ray; arrow mark → parosteal osteosarcoma of the iliac bone

16.3 Contraindications

The contraindications for hip transposition are the same as for internal hemipelvectomy.

Absolute

Tumors crossing the middle of the sacral bone (when wide resection is not possible) and metastatic disease (specialties (i.e., single late metastasis of renal cell carcinoma) excluded)

Relative

Bad general condition, bad soft tissue coverage (especially involvement of the gluteus maximus muscle), involvement of the sciatic nerve, and involvement of the femoral nerve/the femoral artery [11]. Age above 65 years is associated with higher rate of complications [12].

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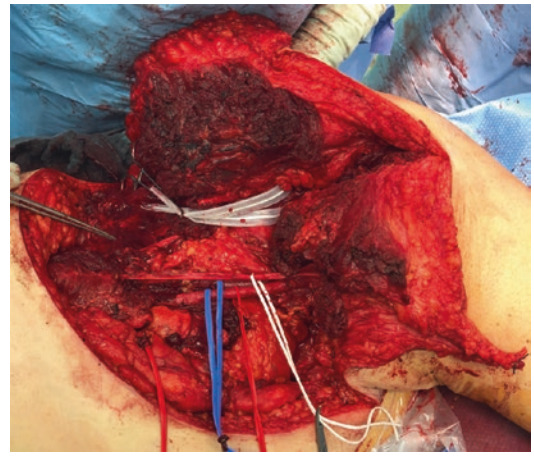
16.4 Preparation

Performing an internal hemipelvectomy is challenging and needs careful preoperative planning and preparation!

We recommend:

1. MRI-based planning of the tumor resection and soft tissue closure (MRI not older than 4 weeks/matched navigation if possible).
2. Use Enneking's approach! For extraarticular resection of the proximal femur, a second cut from the SIAS following the tensor fascia lata to the lateral thigh (lateral part of Judet's approach) can be helpful.
3. Two skilled surgeons/surgical teams performing the operation (simultaneous ventral and dorsal approach) help to shorten intraoperative time.
4. If possible use preoperative epidural catheter for intra- and postoperative pain therapy.
5. Urinary catheter placement (green colored) and ureteral stenting (ipsi- or bilateral) for easy palpation of ureter, urethra, and bladder.
6. Positioning of the patient in lateral position for easy unfolding and through maximizing space between ribs and pelvis.
7. Intraoperatively preserve the vessels feeding the gluteus maximus flap whenever possible; alternatively think about performing a rectus abdominis flap/free flaps or if not possible external hemipelvectomy.

HINT: No false ambitions! Good soft tissue coverage doesn't help in the case of tumor contamination!!!! WIDE RESECTION FIRST!

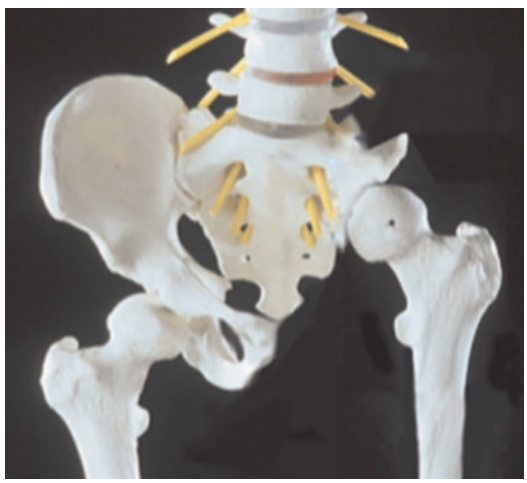


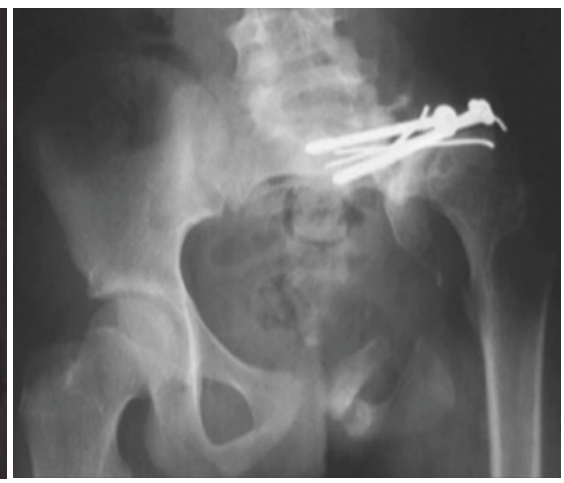
situs after internal extraarticular hemipelvectomy P1-4: red loops, A. iliaca externa and femoralis; blue loop, V. iliaca externa; white loop, N. femoralis; and strap sciatic nerve.

16.5 Performing Hip Transposition

Three types of hip transpositions are described:

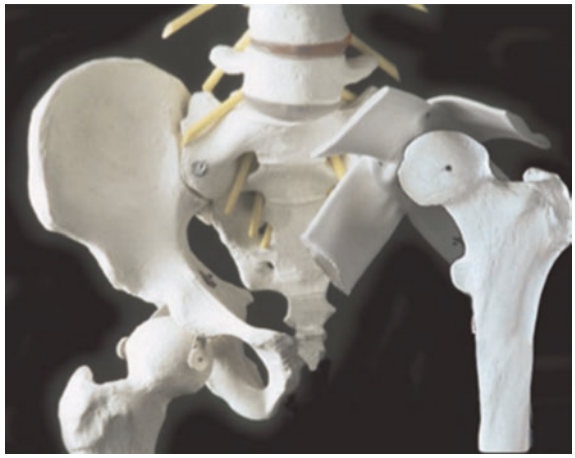
While Type 1 is in need of osteosynthesis (with additional complications like pseudarthrosis or screw displacements),

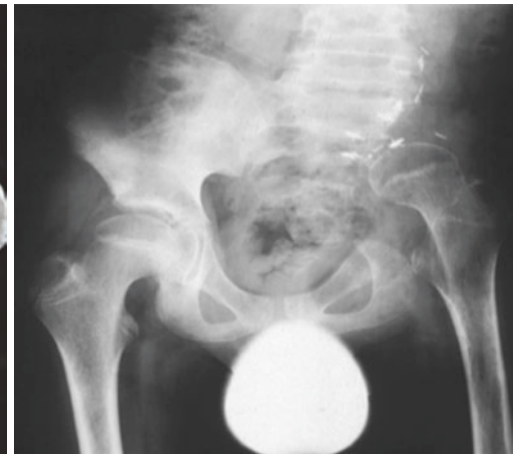




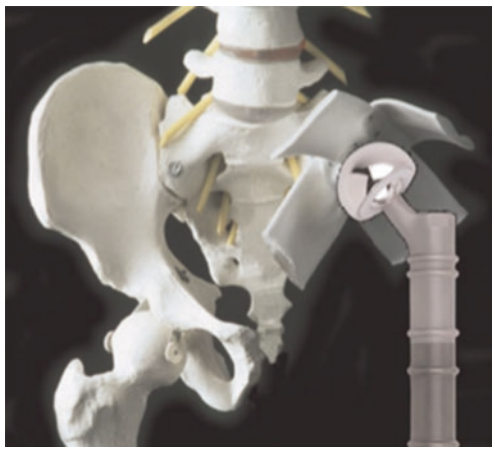
types 2a and 2b are easy to perform.

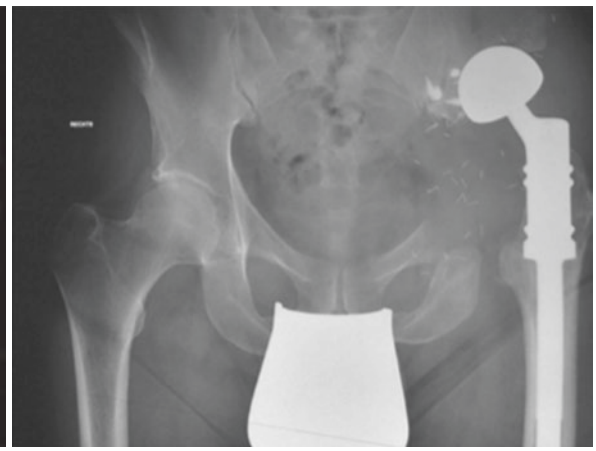
Relevant in clinical terms are Type 2a: hip transposition after intraarticular resection of the hip joint.





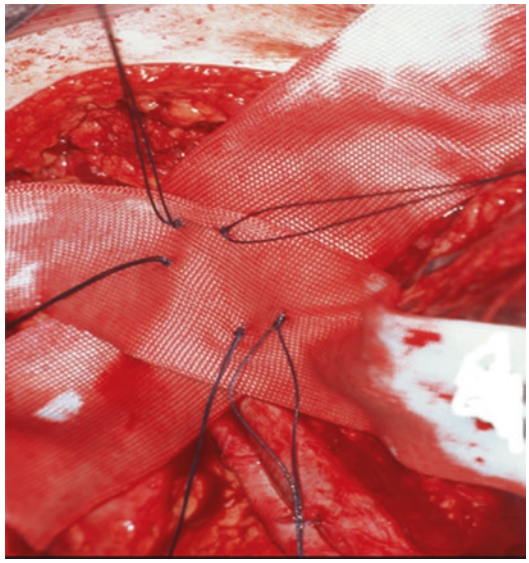
and 2b: hip transposition after extraarticular resection of the hip joint.



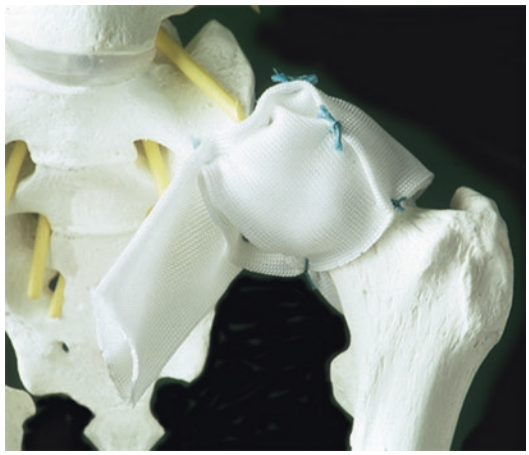


Type 2a:

1. try to narrow the space between the femoral head and the remaining bone, when this is possible proceed, otherwise perform soft tissue release (necessary mostly of the adductor muscles),
2. place 3–5 bone anchors in the remaining bone of the ileum or sacrum (hint: not all kinds of anchors do the job [13]) and fix the attachment tube at the site of optimal fixation,



3. attach the tube on the femoral head with non-resorbable sutures,



Rö einfügen

4. attach remaining muscles to the attachment tube or to the bone for narrowing dead space; HINT: The psoas muscle can be transferred to the trochanter major for better abduction,
5. minimum 2 deep drains,
6. Flip gluteus maximus flap to the front and close musculature and fascia.
7. Close wound, the use of clamps or sutures can be discussed [14], and hard evidence for the use of skin sutures for pelvic wounds has not yet been established.

Type 2 b hip transposition

After the resection of the proximal femur, proceed implanting a proximal femur replacement. In standard care, we use Implantcast MUTARS prox. For femur replacement (silver coated for reduction of deep infections [15–17]/reconstruction length 80 mm, 100 mm, and then every 10 mm) and ic-bipolar head (HINT: use small size, i.e., 44 mm)/CAVE in the case of an allergic reaction to nickel/chrome, etc., ions switch to a full ceramic bipolar head (i.e., Mathys Bionit 2). When finished implanting and testing the endoprosthesis, proceed as in 2a reconstructions.

HINT: While leg length discrepancy is fixed in Type 2a reconstructions, you can adapt the length of the proximal femur replacement in type 2b reconstructions depending on the soft tissue coverage!

postoperative care

- at days 1–5, prolonged antibiotic prophylaxis/antibiotic therapy (second generation cephalosporin) is administered.
- immobilization without splint for 2–6 weeks/alternative procedures like external stabilization via fixateur externe are possible [18].
- partial weight bearing for 3 months after the operation.
- restrictions in movement depending on the soft tissue coverage and wound healing,

complications

- prolonged wound healing/wound necrosis → wait for 5–7 days till revision operation for minimizing the numbers of revisions; close monitoring of CRP and leukocyte count.
- infection → don't hesitate to revise and debride; when used, change endoprosthetic material; remove attachment tube early.
- persistent infection → complete removal of all endoprosthetic material, depending on the entity of the tumor change procedure to flail hip +− vacuum foam or spacer.
- HINT: Don't postpone relevant postoperative chemotherapy for more than 6 weeks.
- complete flap necrosis → shorten leg, change flap, use free flap, or change to external hemipelvectomy,

follow-up

- depending on the entity of the tumor, in general, every 3 months for 2 years, every 6 month for 5 years, and then every year,

late follow-up

- In the case of severe leg length discrepancy more than 5 years after the primary operation, discuss secondary leg lengthening, i.e., via intramedullary nail or external fixateur [19].

16.6 Short summary

While the operative procedure of the hip transposition is fairly easy to perform after successful internal hemipelvectomy, postoperative complications are to be expected. Pelvic resections and reconstruction remain to be among the most challenging operative interventions and should be performed at specialized centers of care.

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3D Printed Reconstructions

17

Andrea Angelini, Daniel Kotrych,
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17.1 Introduction

Limb salvage surgery has been shown to be feasible and effective for most of the primary and secondary pelvic bone tumors [1–4]. The surgical accuracy improved significantly in recent years, thanks to the advances in surgical techniques, biomedical engineering, tumor segmentation on imaging, preoperative planning, and intraoperative tools for surgical margins [5–6]. The increasing interest in the development of 3D printing technology is based on the possibility to improve precision surgery and to realize a personalized custom implant on patient's anatomy [7–11]. In fact, 3D printing techniques have been widely used in numerous fields of orthopedic surgery and musculoskeletal oncology: bone tumor resection and functional reconstruction [12–19], primary and revision arthroplasty [14, 20–23], spinal surgery [13, 24–26], management of complex bone fractures [27–31], and treatment of infective complications [32]. A biocompatible and tailored 3D-printed titanium implant is the final result of a multistep process that begins with the imaging data acquisition and the fabrication of bone models. Of course, each step needs to be monitored and carried out in a structured and precise manner [33, 34]. In the following paragraphs, we will get into the details of each aspect of 3D-printing technology.

17.2 Anatomical Models and Surgical Plan

The first step in the use of 3D-printing technology is the "segmentation" process whereby imaging data (digital imaging and communications in medicine—DICOM) are purchases and converted into a digital 3D-model. This process is particularly relevant in patients with pelvic bone tumors because of the complex anatomy of the pelvis and the need of accurate visualization of tumor extension. Usually, a CT scan of the pelvis with 1 mm cut every 1 mm step is required to have a voxel size of 1 mm square that defines the level of accuracy of the plan. A virtual 3D model may be realized and analyzed by a multidisciplinary team that involves surgeons and engineers, with higher information compared to a multiplanar CT image alone (Fig. 17.1) [35]. In the virtual 3D planning, it is possible to add

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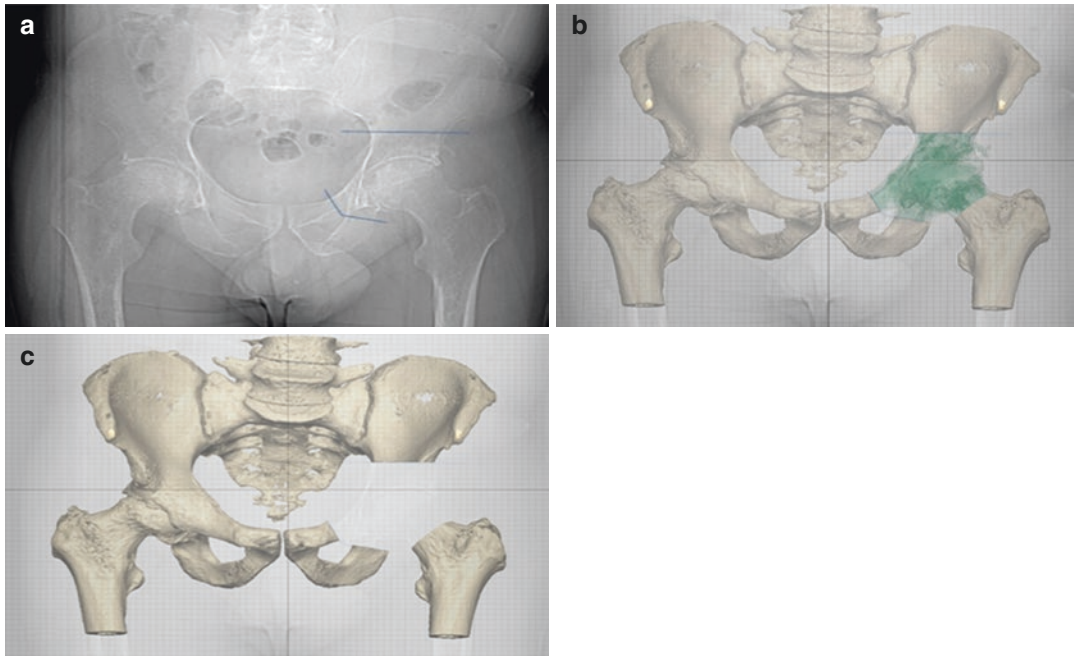

Fig. 17.1 Osteosarcoma of the pelvis in a 52-year-old man. (**a**) Pelvic plain radiographs with the resection levels annotated directly by the surgeon. The pictures are then transferred to engineers for the analysis after segmentation process; (**b**) creation of a digital 3D model with tumor highlighted (green color); (**c**) The expected bone defect can be easily analyzed on the virtual 3D model

anatomic landmarks and resection planes and to visualize specifically the tumor volume (Fig. 17.2a–c). These aspects should be transferred from the surgeon to the engineers considering the surgical approach (based on tumor site), the extent of soft tissue involvement, and close critical anatomic structures. All of these elements should be visualized during surgery for a safe resection (Fig. 17.2d, e). These models can be printed with materials fulfilling biocompatibility standards in a 1:1 scale, so they can be sterilized and brought to the surgical field (Fig. 17.3a) [36] or in plastic transparent material for academic/education aims. 3D printed models improve the surgeon's understanding of the surgical challenge and can be used to explain surgical procedure to the patient (Fig. 17.3b, c), to test the printed surgical tools, and as simulation to improve safety in real-life surgery (Fig. 17.3d). These models are an effective tool for finalizing surgical planning, also considering the crucial role of resection margins as prognostic factor in most of the malignant bone tumors [37–42]. The authors believe that there is a strong difference in preparing the surgical plan for oncologic disease compared to other complex fractures or revision surgery. The main aspect is the possibility of relevant changes in tumor volume from the imaging acquisition to the surgical procedure. It is important to take into account the worst case scenario with a tumor growth not respondent to preoperative chemotherapy or other neoadjuvant procedures, thereby considering a planned bone cut to be safe this occurrence [43].

17.3 Patient-Specific Tools and Guides

Three-dimensional printing is a simple way to obtain patient-specific instrumentation (PSI) that enables the surgeon to follow and realize the surgical plan based on preoperative imaging.

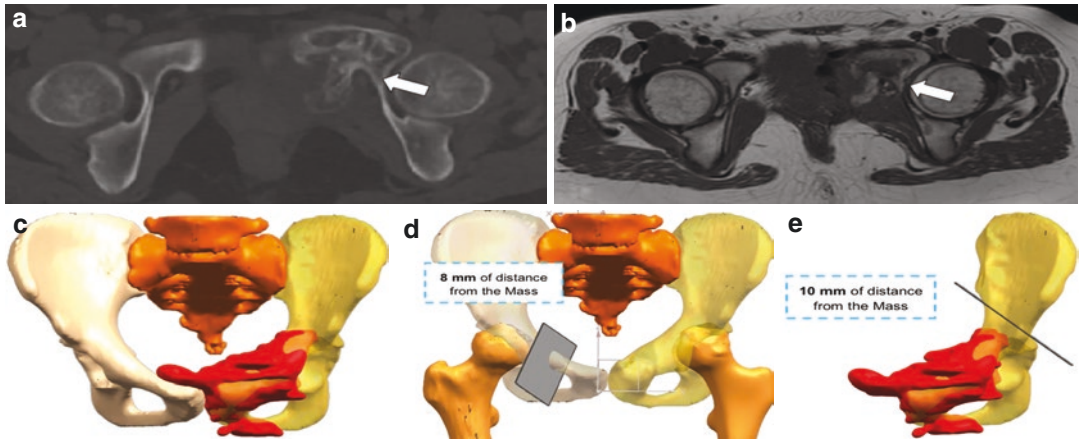

Fig. 17.2 Ewing's sarcoma of the pelvis in a 13-year-old girl. (**a**) Axial CT scan with 1 mm cut every 1 mm step and (**b**) axial T1-weighted MR images were obtained after preoperative neoadjuvant chemotherapy for the evaluation of tumor volume. The tumor (white arrow) is highlighted by the surgeon on both examinations. (**c**) In the virtual 3D planning (using web-based platform Promade, Lima Corporate Medical Systems, Villanova San Daniele del Friuli, Italy), it is possible to visualize the tumor volume (red area) and the involvement of the contralateral ischiopubic branches. (**d**, **e**) Resection planes have been added considering the surgical approach, the extent of soft tissue involvement, and safe resection margins

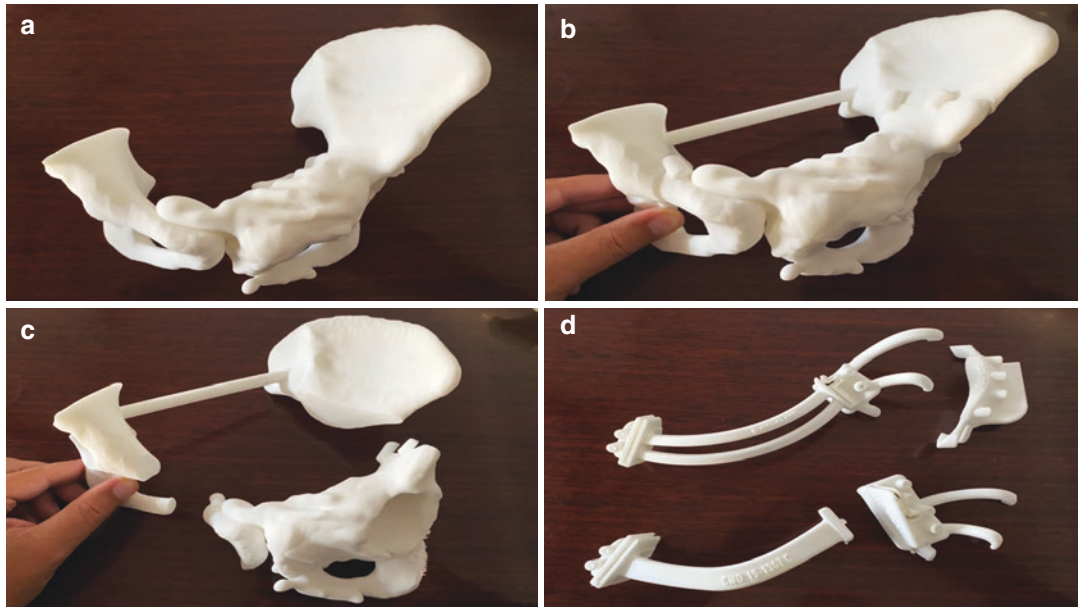

Fig. 17.3 Same patient: Ewing's sarcoma of the pelvis in a 13-year-old girl. (**a**) The 1:1-sized 3D printed model of the entire hemipelvis allows the surgeon to accurately appreciate the anatomy and (**b**, **c**) the surgical planning with detachable parts. (**d**) 3D printed cutting jigs. (Implant designed with Promade, Lima Corporate Medical Systems, Villanova San Daniele del Friuli, Italy)

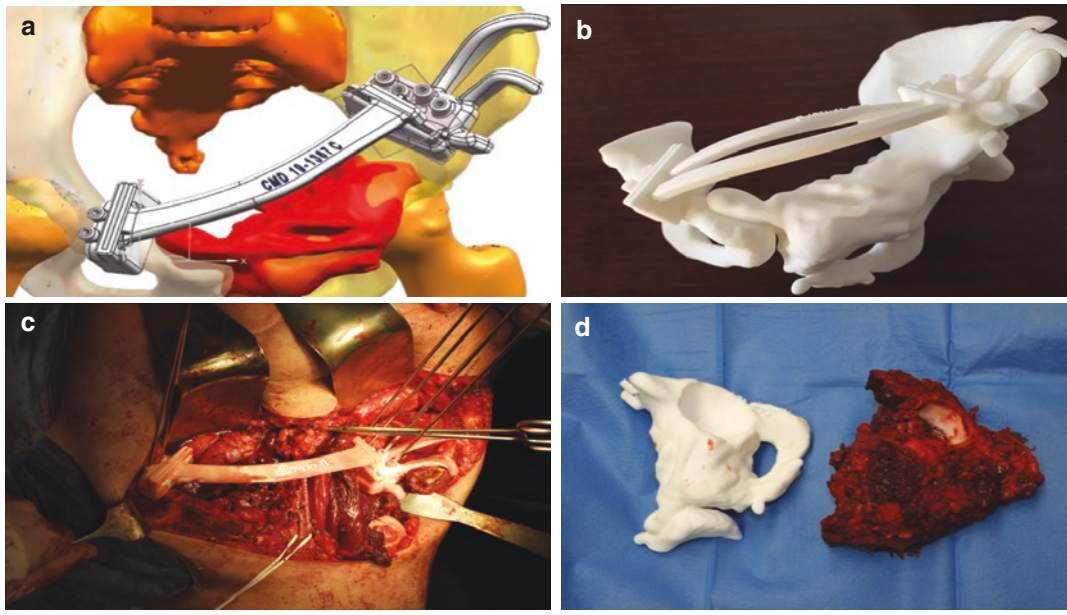

Fig. 17.4 Same patient: Ewing's sarcoma of the pelvis in a 13-year-old girl. (**a**) Preoperative virtual planning with custom cutting jigs and (**b**) 3D-printed patient-specific instrumentations available for intraoperative use. (**c**) The jigs are fixed to the bone with K-wires. (**d**) Intraoperative photograph showing the use of 3-dimensional model of the specimen and the correspondent resected tumor (Implant designed with Promade, Lima Corporate Medical Systems, Villanova San Daniele del Friuli, Italy)

Standard cutting jigs are widely used in primary and revision arthroplasty, whereas their use is precluded in musculoskeletal oncology. The main objectives of using 3D printed PSI are to increase the accuracy of tumor resection in the pelvis and the precision of implant placement [11, 44]. In fact, custom-made prostheses usually need the absolute precision in performing the multiplanar osteotomies [45], and more than one cut planes are necessary for the tumor resection (Fig. 17.4a). There are specific tools designed for bone resection (Fig. 17.4b), and other customized drill guides are studied for improving screw trajectories maximizing implant stability. The cutting jigs reduce the errors derived from the freehand use of oscillating saw blades, uncorrect directions, and contamination due to tumor margin violation. The jigs should be fixed to the bone with one or more K-wires after correct alignment with host bone to reduce further errors related to vibration of the saw blade or guide displacement during bone cut (Fig. 17.4c). Drill-guided PSI is used to guide drill holes that are planned for a specific trajectory and can be mounted on the definitive implant for bone fixation. The intraoperative availability of sterilized 3D-printed models helps the surgeon with correct orientation, especially when they present detachable parts to show the position of cutting PSI (Fig. 17.4d).

Nowaday, we think that the 3D-printed PSIs should be considered a less expensive, easier, and comparable alternative to computer navigation for challenging tumor resections. Some Authors [46] specifically analyzed this topic, showing a clinically acceptable accuracy of 2.62 mm vs 3.6 mm at the resection planes comparing PSI guides with computer navigation. The relative difficulty compared to computer navigation is to make absolutely bare the target bony surface, which will perfectly align with the PSI footprint. In the authors' experience, the application of PSI is valuable in most of the cases after a detailed preoperative planning together with dedicated engineers. A wide surgical exposure is important because correct guide placement and accessibility with different cutting tools (osteotome or

oscillating saw) provide safe resection margins without compromising neurovascular structures.

The ability to improve accuracy in preoperative planning and in real-life surgery using 3D models and 3D printed PSI may strongly influence the surgical, oncological, and functional outcome.

17.4 3D-Printed Prostheses in Pelvic Reconstruction

In recent years, the improvement of 3D printing technology has reached high levels, up to the point of being able to produce prosthetic implants. These customized prostheses may be used as an innovative alternative to different biologic and prosthetic reconstructive strategies [1, 9, 47–50]. However, few studied with limited case series reported results at early/mid-term follow-up in pelvic reconstructions [12, 14, 15, 51, 52]. We recently reported our experience in a relatively large series (41 cases) treated with custom-designed 3D-printed prostheses in different oncologic and nononcologic settings [15].

There are some concepts that should be considered in the implant design of a 3D-printed pelvic prosthesis: (1) how to fill the bone defect; (2) how to assess the correct shape and areas with porous surface structure; and (3) how to obtain a stable fixation at long term; (4) how to optimize the soft tissue reattachment, the osseointegration between prosthesis and host bone, and its coverage with vascularized tissue.

17.4.1 How to Fill the Bone Defect

Historically, allograft prosthetic composite has been widely used to reconstruct large bone defects in the pelvis, despite the relatively high complication rate [53, 54]. With the use of 3D printed technology, it is possible to print custom implants in titanium metal with increasingly popularity and low reduction of cost and time. The industrial application of this process alloy implants is that they are manufactured by metal laser sintering (DMLS) or electron beam melting (EBM) technologies, with adequate internal porous structures that could be considerably valid alternatives to allografts in terms of mechanical scaffold. Based on the preoperative imaging studies, it is possible to determine exactly the bone defect. Many prosthetic models may be design based on the unaffected site, but we usually suggest a peripheral downsize of 1 mm in order to achieve optimal fit of the implant and adequate soft tissue coverage, with attention for any miscalculation (too small or too short implant).

17.4.2 Shape and Structure

The challenge in the use of custom implants in pelvic reconstructions is achieving a perfect fit considering the distribution of forces during weight-bearing activities at physiological status. Looking the literature, a wide spectrum of implant designs has been reported, with different shapes and concepts [12, 14, 15, 51, 52, 55, 56]. Custom-made 3D printed prostheses have specific indications in the reconstruction of periacetabular area, which represents the most demanding site for anatomy and hip joint function. Dai et al. reported their experience in ten patients with different designs and fixation on the remaining ilium (or the sacrum), pubic rami (same side or other side), and the ischial rami [56]. Wang et al. specifically evaluated the outcomes in 11 periacetabular 3D printed implants with similar reconstructions, searching with different strategies a complete restoration of the pelvic ring to obtain satisfactory functional results [19]. We do not emphasize a strict anatomical pelvic ring reconstruction [14, 15]. In our experience, a 3D-printed custom-made prosthesis can be safely used when a good and stable interface bone/implant could be obtained in residual part of the ilium or sacrum (Fig. 17.5a, b). The hip joint can be therefore replaced with "conventional" modular cups (Fig. 17.5c), as well as the proximal femur (Fig. 17.5d). Usually, the bone-contacting surfaces should be realized with a porous structure to facilitate the bone ingrowth as proven by in vitro and in vivo studies [7]. The metal 3D printing allows the production of

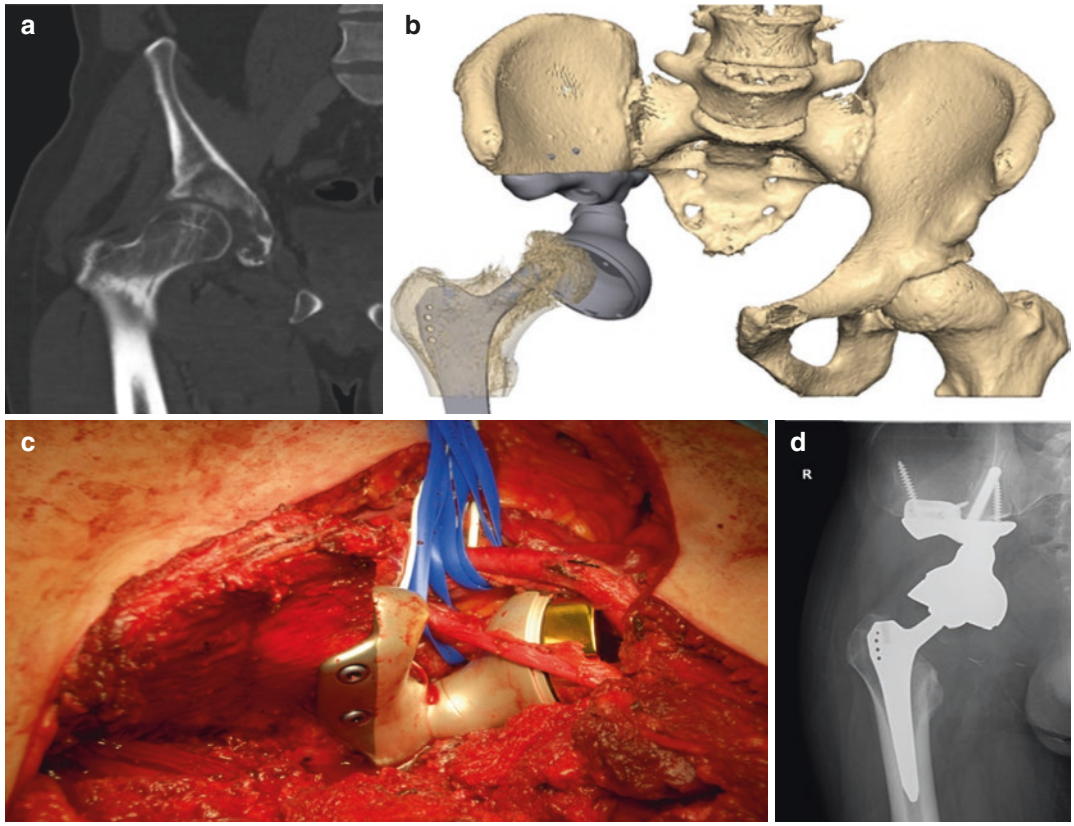

Fig. 17.5 Chondrosarcoma gr. 2 of the right acetabular area in a 38-year-old male treated with type II-III resection. (**a**) Preoperative CT scan and (**b**) surgical planning on virtual 3D model. (**c**) intraoperative photograph showing the definitive implant of a custom-made 3D-printed prosthesis with "conventional" modular cup. (**d**) Postoperative radiograph shows our philosophy of not pursuing the complete restoration of the pelvic ring (Implant designed with C-Fit 3D®, Implantcast Ltd., Buxtehude, Germany)

implants with complex shapes, alternating porous surfaces (implant/bone interface) and smooth surfaces and minimizing local friction to overlying important soft tissues. On the other hand, the weight of the implant and stress forces concentration can be optimized working on the customizable texture of the internal structure.

17.4.3 Long-Term Fixation

The long-term mechanical strength of a 3D printed pelvic prostheses is not guaranteed considering the limits of recent clinical use and mid-term follow-up of the largest published series [12, 14, 15, 51]. The EBM or SLM technology alloys a successive layering of melted titanium according to a computer-aided design (CAD) model, so there is the possibility to create a porous surface with ingrowth bone characteristics [7, 8, 57]. The perfect fit between host bone and prosthesis is a primary requirement for immediate stability and long-term fixation. Wong et al. described the use of printed guide plates for intraoperative precise resection and implant installation [46]. In our experience, 3D-printed PSIs are routinely used and are considered the most effective tools for guided resection and reconstruction. During the last few years, we used different strategies for primary fixation to host bone, which today should be considered in combination: long cancellous screws, short cortical screws, press-fit porous stems, and small hooks for stabilization (Fig. 17.6). In the design

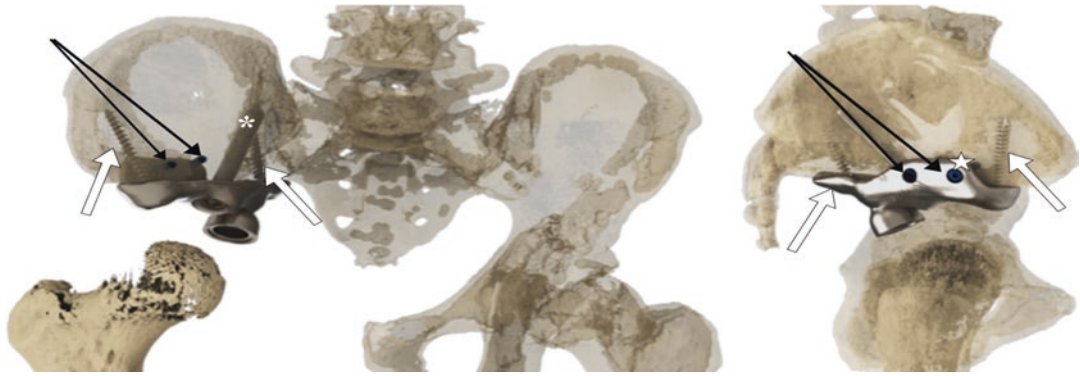

Fig. 17.6 Same patient: **Chondrosarcoma gr. Two of the right acetabular area in a 38-year-old male.** The transparent virtual 3D model shows the strategies for primary fixation to host bone: long cancellous screws with 6.5 mm of diameter (white arrows), short cortical screws with 4.5 mm of diameter (black arrows), press-fit porous stems (asterisks), and hook for stabilization (white star). (Implant designed with C-Fit 3D®, Implantcast Ltd., Buxtehude, Germany)

phase of the custom implant, we suggest to plan counter screws as safety locking to avoid screw mobilization. When resection involves the proximal part of the ilium, sacroiliac joint, or the sacrum, there is a need of further stabilization of the implant for adequate loading transfer and balance. In these cases, we suggest a posterior pedicle screw-rod fixation connected to tulip-head polyaxial screws directly designed on the custom prosthesis, to add stability to the spinopelvic continuity.

17.4.4 Soft Tissue Reattachment and Prosthetic Coverage

The soft tissue coverage of the prosthesis with well-vascularized muscles is one of the most important aspects to avoid wound dehiscence or deep infection [53, 58–60]. The porous surface can be realized not only in bone/implant interface but also in some areas (i.e., outer part of the ilium) to increase the friction and adherence of the residual soft tissues to the prosthesis. In some cases, we designed specific large holes to guarantee a fixation of muscles/tendons through the prosthesis [15], as well as reported by other Authors [61]. Different flaps can be used by the reconstructive surgeon to plan a specific solution for implant coverage and wound closure [62, 63]. The rectus abdominis musculocutaneous flap is one of the most adaptable flaps for both periacetabular and sacral reconstructions.

17.5 Advantages of 3D Printing Technology in Reconstruction for Pelvic Defect

Advantages of 3D-printed pelvic prostheses are listed in the following:

- The shape and design of the 3D-printed pelvic prostheses were based on thin-layer CT scans converted into a digital 3D-model. Thanks to the multidisciplinary collaboration between engineers and surgeons, a more accurate resection with PSIs may allow an improvement of oncologic outcome. Moreover, preoperative 3D printed models of the anatomy and tumor site may help to visualize, predict, and better understand the surgical challenge.
- The optimization of 3D printed custom-jigs guides for intraoperative resection is safe and cheap compared to intraoperative navigation in our experience, considering the limits of the latter reported in literature [16, 41]. This aspect influences the operation time and intraoperative implant exposure, reducing consequently the risk of infectious complications.

- The possibility to produce anatomic implants with a wide freedom in terms of shapes and structure allows a stable reconstruction and a good restoration of function. The reported functional results evaluated with MSTS score at a mid-term follow-up ranged from 63% to 85% [12, 14, 15, 51, 52, 56].
- The bony ingrowth in the porous structure of the prosthesis may reduce mechanical failures at long-term follow-up that usually occurs due to osteolysis or nonunion at the junction in the reconstructions with APC [1, 64, 65].
- There are potential developments in 3D-printed technology looking at newer materials to decrease the risks of infection and improve bone ingrowth.

17.6 Conclusions

The use of custom-made 3D-printed prostheses represents today a well-established method for reconstruction of large pelvic bone defects, which will become increasingly accessible in the next decades. These implants have the potential objectives to improve oncologic and functional outcomes in patients with pelvic bone tumors, with acceptable complication rate and satisfactory safety.

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Particle Radiotherapy

18

Petra Georg and Eugen Boris Hug

18.1 Background

Particles used in the radiotherapy are energetic protons or positive ions (e.g., carbon ions). Particle therapy has been introduced to increase cure rates and/or decrease side effects for patients who need radiotherapy as part of their cancer treatment. The most common type of particle therapy is proton therapy. The second type routinely used is treatment with carbon ions. For protons and carbon ions, the dose increases, while the particle penetrates the tissue and loses energy continuously. Hence, the dose increases with increasing thickness up to the Bragg peak, where the maximum energy deposits and it occurs near the end of the particle's range. Both proton and carbon ion therapies exhibit a defined Bragg peak in the body, so they deliver their maximum lethal dosage at or near the tumor.

Beyond the Bragg peak, the dose drops to zero (for protons) or almost zero (for heavier ions) (Fig. 18.1).

The advantage of this energy deposition profile is that less energy is deposited into the healthy tissue surrounding the target tissue. Protons and carbon ions cause damage of the DNA of tissue cells, ultimately causing their death. Because of their reduced ability to repair damaged DNA, cancerous cells are particularly vulnerable to this damage. From a radiation biology standpoint, there is considerable rationale to support the use of carbon ion beams in treating cancer patients. Carbon ions are heavier than protons and so provide a higher relative biological effectiveness (RBE), which increases with depth to reach the maximum at the end of the beam's range. Thus, the RBE of a carbon ion beam increases as the ions advance deeper into the tumor-lying region. Carbon ion radiotherapy provides the highest linear energy transfer (LET) of any currently

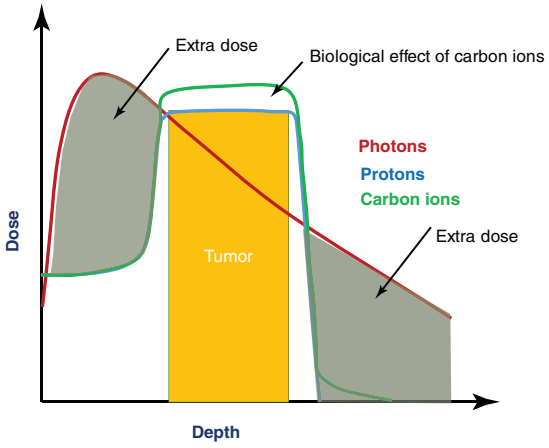

Fig. 18.1 Dose depth curve for photons, protons and carbon ions

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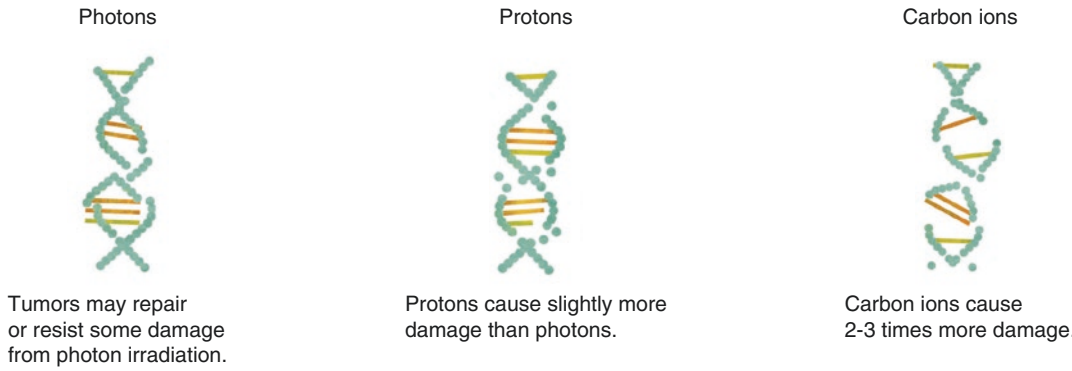

Fig. 18.2 Biological differencies between photons, protons and carbon ions

available form of clinical radiation. This high energy delivery to the tumor results in many double-strand DNA breaks, which are very difficult for the tumor to repair (Fig. 18.2). In comparison, conventional radiation produces principally single strand DNA breaks, which allow many of the tumor cells to survive.

There are clear advantages to treat otherwise intractable hypoxic and radioresistant cancers with carbon ions, while opening the door for substantially hypofractionated treatment of normal and radiosensitive disease with protons. The higher outright cell mortality produced by carbon ion radiotherapy may also provide a clearer antigen signature to stimulate the patient's immune system. This might be beneficial in improving the disease outcome.

As far as pelvic bone tumors are difficult to manage, particle therapy may widen the scope of treatment possibilities with the aim of improving disease control in this patient population.

18.1.1 Chordoma and Chondrosarcoma

18.1.1.1 Rationale for Particle Therapy

Chordoma is a rare (1–4% of all malignant bone tumors), slowly growing tumor arising from cellular remnants of the notochord, anywhere along the spine. Sacrum is the predominant location of this tumor in the pelvis. Chondrosarcomas are relatively rare bone tumors. Their predominant location in the pelvis is also in the sacrum, but they can also be located primarily in the pelvic ring. Both tumor entities are characterized by slow, but locally aggressive and invasive growth patterns leading to large tumor volumes. The management of these tumors is challenging, because they lie in close vicinity to critical structures, like spinal cord, nerve roots, or bowel structures. These anatomic structures often limit surgical access and resectability, as well as the delivery of high radiation doses. Nonetheless, surgery remains the initial standard treatment and postoperative high-dose radiotherapy is frequently recommended. The major problem is the insufficient local control, due to limited possibility of high dose application because of normal tissue tolerance. Large randomized prospective studies comparing photons with protons and/or carbon ions are missing to date. This might be due to the fact that particle therapy is presently not easily available and that these subgroups of bone tumors have a comparatively low incidence rate, so systematic randomized trials are difficult to perform [1].

18.1.1.2 Clinical Evidence of Particle Therapy

By using protons or carbon ions, the local control rates can be improved to 70–100% in patients with extracranial chordomas [2–12]. Focusing on the sacral chordoma, there are reports using proton or carbon treatment.

18.1.1.3 Carbon Ion Radiotherapy

Japanese data are focusing on definitive carbon ion radiotherapy (CIRT).

At National Institute of Radiological Sciences in Chiba in Japan (NIRS), carbon ion therapy is used since 20 years. Imai et al. reported their results on local control and side effects. In the initial report on phase I and II clinical trials in 38 patients, the 5-year local control and survival rate was 89% and 86% in 38 patients [2]. Two patients (5%) developed severe skin toxicity after treatment, 2 of them required skin grafting, and 15 patients (16%) had neuropathic pain that impacted their quality of life. In the next report on 95 patients, the authors showed 5-year local control and survival rates of 88% and 86%, 2 patients had late grade 3 skin, and 2 patients had grade 4 late skin and soft tissue complication; 90% of patients remained ambulatory, and 50% needed no pain medication [3].

In their extended patient population of 188 patients, their report local control was 77.2% and overall survival was 81.1% at 5 years. There were grade 3 toxicity of the peripheral nerves in 6 patients and grade 4 toxicity of the skin in 2 patients [4].

The second institution using carbon ions in the treatment of sacral chordoma is the Heidelberg Ion Therapy center. In 2014, they started a randomized phase II trial of hypofractionated proton versus carbon ion radiation therapy in patients with sacrococcygeal chordoma—the ISAC trial protocol. This is the only one randomized study comparing protons and carbon ions using the same dose and fractionation Scheme (22 fractions with 3 Gy RBE per fraction). The aim of this study is to confirm the toxicity results of the Japanese data and to compare them with the toxicity analysis of proton therapy given in the same fractionation [5]. Their preliminary results using carbon ion therapy in a patient cohort of 56 patients show 79% local control and 100% overall survival at 2 years. The 2- and 3-year local control probability was 76% and 53%, respectively. However, in this cohort, 15 patients were treated for recurrent tumors. Local control was significantly better in the naive patients with 85% at 2 years vs 47% in recurrent patients. A total of 23 patients were irradiated with carbon ions in combination with photon IMRT, while 33 received carbon ion therapy only. No grade 3 or higher toxicity occurred after radiation treatment. Five patients showed a decrease in pain after treatment [6].

At Hyogo Ion Beam Medical Center (HIBMC) in Japan, protons or carbon ions were used for treating patients with sacral chordomas, and they report 94% local control and 83% overall survival at 3 years in 23 patients. In 9 patients (39%), they observed grade 3 or higher late toxicity. The most frequent event was dermatitis [grade 4 in five patients (22%)], followed by neuropathies, including motor disorder, sensory disorder/pain, and urinary retention [grade 3 in four patients (17%)] [7].

Clinical outcome comparison between CIRT and surgery for sacral chordoma was performed by Nishida et al. [8]. Although the patient population was very limited, 10 patients were treated with surgery and 7 patients with CIRT. The local recurrence-free survival rate at 5 years was 62.5% for the surgery and 100% for the carbon ion RT group. Urinary anorectal function worsened in 6 patients (60%) of the surgical group and remained unchanged in all patients treated with carbon ions. Postoperative wound complications requiring surgical treatment occurred in 3 patients (30%) of the surgical group and in 1 patient (14%) after CIRT. The functional outcome evaluated using the Muskuloskeletal Tumor Society scoring system (MSTS) was 55% in the surgery group and 75% in the carbon ion group, and carbon ion group had significantly higher scores in the emotional acceptance than did the surgery group [8].

A direct comparison of clinical and functional outcomes between surgery and CIRT for pelvic chondrosarcoma was published only in one report from Osaka Medical Center. They compared 24 patients who underwent surgery with 7 patients treated by CIRT, which was performed at NIRS and HIBMC. The surgery was performed with reconstruction in 13 patients, and 11 patients have resection only. The 5-year local control and overall survival rates were 68% and 72%. Despite the limited number of patients treated with CIRT, there were no significant differences in survival or local control between these patients and surgical treated patients. The functional outcome was

measured with Musculoskeletal Tumor Society (MSTS) functional score available in 21 patients. The mean MSTS score was 59%, and the mean score in CIRT patients (n = 7) was 72.6% vs 49.6% in patients treated with surgery (n = 14), showing better outcomes in CIRT patients. Complication developed in 9 patients after surgery, and seven patients developed deep wound infection at a median follow-up of 6 months. All seven patients had periacetabular tumors and were treated with prosthetic pelvic reconstruction. To reduce the infection, three patients needed removal of the implant, three needed wound debridement, and one needed a flap reconstruction. Implant dislocation was seen in 2 patients, and pulmonary thrombosis in one patient. One patient exhibited nerve palsy, but eventually recovered. Complications developed in 5 patients after CIRT. Five pelvic compression fractures and four cases of avascular femoral head necrosis were seen in 5 patients. One patient needed a unilateral hip replacement because of femoral neck fracture [9].

18.1.1.4 Proton Radiotherapy

At Massachusetts General Hospital in Boston, the standard treatment for sacral tumors is surgery. Within a phase II trial, DeLaney et al. reported their results on 50 patients with tumors in the spine and sacrum. There were 29 chordomas, 14 chondrosarcomas, and 7 other histologies. The majority of patients had surgery, and 13 patients had biopsy only (26%). Only 26 patients had primary location of the disease in the sacrum. Patients with sacral tumor received preoperative radiotherapy to reduce risk of seeding during surgery, and after surgery, patients were treated up to 70.2 Gy RBE, if resections were R0 and up to 77.4 Gy RBE if R0 was not achieved. A mixture of photons and protons was used. Local control and overall survival for the whole group were 78% and 87% at 5 years. Radiation complications grade 3 or more developed in 6 patients (two neuropathies, one erectile dysfunction, one rectal bleeding, and two sacral insufficiency fractures). No spinal cord injuries were seen. Grade 3 sacral neuropathies and erectile dysfunction occurred at doses of 77.1–77.4 Gy RBE to central sacral chordomas, where spinal canal/sacral nerve sparing was not possible because of tumor location [10]. In their updated results with 7.3 years of median follow-up, the 5- and 8-year actuarial local control rates for the whole group were 81% and 74%, and for primary tumors, the rates were higher with 95% and 85%, because local recurrence was less common for primary tumors (11%) than for recurrent tumors (50%). The overall survival was 84% and 65% at 5 and 8 years. The actuarial risk for late grade 3 and 4 toxicity was 13% at 8 years. No late neurological toxicities were found with radiation doses 72.0 Gy RBE, but 3 sacral neuropathies developed after doses of 76.6–77.4 Gy RBE [11]. In the updated analysis of tumor response, Kabolizadeh et al. evaluated 40 patients with unresected chordoma, treated with definitive photon/proton radiation therapy. Tumor location in the sacrum was in 27 patients. With a median follow-up of 50.3 months, the 5-year local control and overall survival rates were 85.1% and 81.9%. Additionally, the authors evaluated volumetric response of the total target volume using MRI and CT imaging. They found that significant volumetric reduction of the total target volume presented at a median follow-up of 18 months, followed by further gradual reduction throughout the rest of the follow-up period. Volumetric analysis was more reliable and reproducible in comparison with the modified RECIST [12].

The impact of tumor volume on overall survival was also described by the Boston group in a retrospective analysis of 24 patients with newly diagnosed, previously untreated spinal chordomas (only core biopsy, no prior incision or resection). At a median follow-up of 56 months, overall survival was 91% and local control was 81.5%. Tumor volume more than 500 cm^3 was correlated with worse survival (50% at 56 months). Long-term side effects included 8 sacral insufficiency fractures, 1 secondary malignancy, 1 foot drop, 1 erectile dysfunction, 1 perineal numbness, 2 worsening urinary/fecal incontinence, and 4 grade 2 rectal bleedings [13].

Authors from Paul Scherrer Institute (PSI) reported their initial experience with spot

scanning-based technology using protons in 40 chordoma patients. Published results are not focused on sacrum, but reporting mobile spine and sacrum together, 11 patients had tumor located in sacrum. Twenty one patients underwent macroscopically complete surgical resection (no GTV left); the remaining 19 were treated with macroscopic disease, 31 patients were treated with protons only, 9 with a mix of photons and protons. Thirty-seven patients (93%) received >70 Gy RBE (range, 59.4–75.2). Local control was 62% and overall survival was 80% at 5 years. In patients without metal stabilization device (part of this may be explained by the dosimetric uncertainties due to metal implant artifacts and part may be due to a correlation between need of surgical stabilization and bigger and more infiltrating tumors), the local control was 100%.In sacral patients, there was only one case of late grade 3 toxicity with subcutaneous fistula requiring multiple surgical debridement [14]. For the first time here, the negative effect of metal (e.g., titanium) implant material used for reconstruction/spinal stabilization on the local control was described. The reasons may be multifactorial, the authors describe the dosimetric uncertainties, because they may impact the range calculation for particle therapy, but it also includes the uncertainty in precise delineation of target and organs at risk (especially in the spinal canal) because of the imaging artifacts. If a debulking surgery is planned, the possibility of substituting metal implants with carbon fiber devices should be considered to enable radiation with curative intent.

In the clinical outcome paper on 26 pediatric patients (mean age at time of proton therapy was 13.2 years) with chordoma and chondrosarcoma treated at PSI, they reported local control rates of 81% for chordoma and 80% for chondrosarcoma at 5 years and the corresponding overall survival rates were 89% for chordoma and 75% for chondrosarcoma, with no high-grade late toxicities observed. However, the majority of patients had skull base location of the tumors (17 patients), 8 patients had spinal tumor location, and only one patient had sacral chordoma [15].

In a large retrospective cohort, authors from Hyogo Ion Beam Medical center evaluated proton beam therapy results in 96 patients with bone sarcomas. Seventy two patients (75%) had chordoma, 20 patients (20.8%) had chondrosarcoma, and four patients (7.2%) had osteosarcoma. Patients received a median total dose of 70 Gy RBE. The most frequent location was skull base in 68 patients (70.8%) and sacral spine only in 13 patients (13.5%), 12 patients with sacral tumors had chordoma, and only 2 patients of 13 patients with sacral tumors underwent surgical resection. The authors did not report data of sacral tumors separately, but they reported the outcome data of patients with tumors of the spine (including 8 patients with tumor location in cervical spine, 5 in lumbar spine, 2 with lumbosacral spine, and 13 with sacral tumors, total 28 patients). The 5-year local control and overall survival rates were 55.6% and 70.7%. Late grade 3 and higher toxicities occurred in 9 patients (9.4%), and the noncerebral toxicities were musculoskeletal and connective tissue disorders in 3 and necrosis in 2 patients [16].

All data on carbon ion or proton radiotherapy suggest good local control and functional outcomes, suggesting a good alternative for surgery or even avoiding surgery. The Italian Sarcoma Group initiated in 2016 a randomized and observational study on surgery versus definitive radiation therapy in primary localized disease (sacral chordoma, SACRO Study). This study is aimed at estimating the effectiveness of definitive radiotherapy as compared to standard surgical treatment for patients with primary sacral chordoma who are candidates to a complete en bloc resection, in terms of relapse-free survival (RFS). The radiotherapy options used in this study are proton or carbon ion radiotherapy. The secondary objectives are to estimate the efficacy, activity, safety, and quality of life for definitive radiotherapy as compared to standard surgery, as well as to identify radiological and pathological characteristics that might be used as predictors of relapse-free survival, progression-free survival, and overall survival (*ClinicalTrials.gov identifier (NCT number): NCT02986516).*

18.1.2 Ewing Sarcoma

18.1.2.1 Rationale for Particle Therapy

Ewing's sarcomas are highly sensitive to radiation therapy. Their locations often involve bones, which are not so easily resectable, where radiotherapy is well-established treatment. Radiotherapy is used in the postoperative setting for patients with close or positive resection margins and sometimes in the setting of a poor or slow clinical response to neoadjuvant chemotherapy as an additive neoadjuvant treatment. Radiation is typically used instead of surgery for children with unresectable tumors or in cases in which surgery would result in severe, mutilating morbidity. Nevertheless, surgery plays a substantial role in the management of these tumors due to several reports where not only local but also distant failure occurred more frequently in patients treated with radiotherapy only than in those treated with surgery or surgery–radiotherapy [17–19]. However, definitive radiotherapy causes side effects as a result of the volume of normal tissue also irradiated. Due to the steep dose profile, particle therapy offers better sparing of normal tissue, therefore reducing the toxicity.

18.1.2.2 Clinical Evidence of Particle Therapy

A majority of reviewed reports on particle therapy for Ewing sarcoma focus on the use of protons. This has two major reasons; first of all, the accessibility of proton therapy is much broader, because there are simply more proton only centers than carbon ion therapy centers. The other reason is also the radiosensitivity of this histology. Carbon ions offer stronger biological effects because of their high LET, but this is the strongest argument in their use in radioresistant tumors such as osteosarcomas.

18.1.2.3 Carbon Ion Radiotherapy

The only report about the use of carbon ion radiotherapy in Ewing sarcomas is coming from the NIRS experience. They reported on five unresectable patients: 3 patients had tumors located in the pelvis, and two patients in the spine. After carbon ion radiotherapy, 2 patients showed tumor shrinkage. No severe acute toxicity was observed. One patient with spinal tumor developed local failure, and the tumors located in the pelvis were controlled. But all pelvic tumor patients developed distant metastases. So the authors concluded that local control is favorable, but distant control is unsatisfactory [20].

18.1.2.4 Proton Radiotherapy

Report from Paul Scherrer Institute on 38 pediatric patients (median age 9.9 years) with Ewing sarcoma treated with pencil beam scanning proton therapy shows 81.5% local control and 85% overall survival at 5 years. The majority of these patients had the primary tumor location in the axial/pelvic site (27 pts., 71.7%). The outcome data in this patient subcohort were 75.1% local control and 80.6% overall survival at 5 years. All local recurrences developed in the irradiation field and in nonextremity locations. Two patients developed late grade 3 toxicity (kyphoscoliosis in 1 pt. and endocrine dysfunction in 1 pt) [21].

At Massachusetts General Hospital, Rombi et al. evaluated the outcome of 30 pediatric Ewing sarcoma patients treated with proton therapy. The 3-year local control and overall survival rates were 86% and 89%, respectively. The only severe late toxicities were hematological malignancies associated with the use of topoisomerase and anthracycline. However, only 4 patients in this cohort had pelvic tumor mass and 2 patients had lumbosacral spine tumors [22].

18.1.3 Osteosarcoma

18.1.3.1 Rationale for Particle Therapy

Osteosarcoma is the most common primary bone malignancy in children and adolescents and is very radioresistant [23]. Neoadjuvant chemotherapy, followed by surgical resection, and further adjuvant chemotherapy is the typical treatment approach for high-grade osteosarcomas [24]. While this approach yields an acceptable overall survival for resectable cases, outcomes remain poor for unresectable osteosarcomas such as those in the pelvis or trunk [25].

Carbon ion radiotherapy (CIRT) has stronger biological effects and more conformal dose distribution compared to photon- and proton-based therapies [26, 27].

18.1.3.2 Clinical Evidence of Particle Therapy

Surgery remains the standard for the treatment of osteosarcoma; however, similar to Ewing sarcoma, osteosarcoma also develops in bones, where a radical surgery without major morbidity is not possible. Osteosarcomas are well known as radioresistant tumors, so the rationale of using carbon ion therapy is obvious.

18.1.3.3 Carbon Ion Radiotherapy

First report on efficacy and safety of carbon ion radiotherapy in bone and soft tissue sarcomas comes from NIRS from a phase I/II dose escalation study. Fifty-seven patients with 64 sites of bone and soft tissue sarcomas not suitable for resection received carbon ion radiotherapy. Tumors involved the spine or paraspinal soft tissues in 19 patients, pelvis in 32 patients, and extremities in six patients. Bone sarcomas had 41 patients with osteosarcoma [15], chordoma [11], chondrosarcoma [6], primitive neuroectodermal tumor (PNET) [5], malignant fibrous histiocytoma (MFH) [1], and other histologies [3], and 16 patients had soft tissue sarcomas. The total dose ranged from 52.8 to 73.6 Gy (RBE) and was administered in 16 fixed fractions over 4 weeks. The median tumor size was 559 cm^3 (range: 20–2290 cm^3). The local control rates were 88% and 73% at 1 year and 3 years of follow-up, respectively. The 1- and 3-year overall survival rates were 82% and 46%, respectively. Looking on the bone and cartilage manifestations, local control could be reached in 11/15 osteosarcoma cases, in 12/13 chordoma manifestations, and 5/7 chondrosarcoma lesions [28].

The next experience with CIRT in the treatment of unresectable sarcoma concentrated on spinal sarcomas only. Matsumoto et al. evaluated the outcome in 47 patients with 48 medically unresectable spinal sarcomas included in phase I/II and phase II clinical trials for bone and soft tissue sarcomas. Five-year local control and overall survival rates were 79% and 52%. Sacral tumors were excluded, the major histology was osteosarcoma in 13 patients and chondrosarcoma in 13 patients, and other histologies were chordoma [9], malignant fibrous histiocytoma (MFH) [7], Ewing sarcoma, [2] and others [4]. One patient had grade 3 and one patient grade 4 late skin toxicity with skin ulcer requiring grafts. Vertebral body compression occurred in 7 patients. Twenty-two of the surviving 28 patients who had primary tumors remained ambulatory without supportive devices, so the authors concluded that CIRT was both effective and safe for the treatment of patients with unresectable spinal sarcoma [29].

First experience on pediatric unresectable osteosarcoma presented results on 26 patients (median age 16 years) with inoperable osteosarcoma of the trunk (24 pelvic, 1 mediastinal, and 1 paravertebral). Median CIRT dose was 70.4 Gy RBE delivered in 16 fractions. Local control was 69.9% and 62.9%, and overall survival was 50.0% and 41.7% at 3 and 5 years, respectively. Grade 3–4 adverse events excluding fractures of affected bone were observed in 4 cases. There was one case of grade 3 skin toxicity, one case of grade 4 skin toxicity, and 2 cases of neurologic dysfunction due to nerve injury. There was 1 case of grade 4 bone toxicity in which the sacrum (S1) was involved with the disease and developed a fracture after CIRT [30].

At Heidelberg Ion Therapy Center, a nonrandomized therapy trial to determine the safety and efficacy of heavy ion radiotherapy in patients with nonresectable osteosarcoma in children older than 6 years was started in 2010. Desired target dose is 60–66 Gy RBE with 45 Gy RBE proton therapy and a carbon ion boost of 15–21 Gy RBE. The primary objectives are the determination of feasibility and toxicity of proton therapy and heavy ion therapy using carbon ions boost. Secondary objectives are tumor response, disease-free survival and overall survival. The aim is to improve outcome for patients with nonresectable osteosarcoma [31].

The experience from HIBMC showed very good results in 91 patients with nonmetastatic unresectable or incompletely resected pelvic bone and soft tissue sarcomas of the pelvis. The particle therapy with protons was performed in

52 patients, and carbon ion therapy was performed in 39 patients. The histologic type was chordoma in 53 patients, chondrosarcoma in 14, osteosarcoma in 10, malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma in 5, and others in 9 patients. The 3-year local control and overall survival rates were 92% and 83%. Late grade 3 or higher toxicity was observed in 23 patients (25%). Late grade 3 toxicities involved peripheral nerves in 6 patients, pain in 5, bone in 2, genitourinary tract in 2, muscle in 2, skin in 2, and the vascular system in 1 patient. Late grade 4 toxicities involved skin in 9 patients [32].

18.1.3.4 Proton Radiotherapy

At Massachusetts General Hospital, unresectable or incompletely resected osteosarcoma is treated with proton therapy or with combination of photon and proton therapy. The initial report on 55 patients with a median age of 29 years (2–76) showed local control 82% and 72% at 3 and 5 years, and the five-year OS was 67%. The extent of surgical resection did not correlate with outcome. Grade 3–4 late toxicity was seen in 30.1% of patients. One patient died from treatment-associated acute lymphocytic leukemia, and one from secondary carcinoma of the maxilla [33].

18.2 Summary

Bone tumors involving the pelvis are in the majority of cases chordomas, chondrosarcomas, Ewing sarcomas, and osteosarcomas. For other histologies, the evidence of particle therapy is very limited and it can be recommended in selected cases, i.e., in pediatric patients and young adults when reduction of integral dose delivered to healthy tissue is needed in order to avoid late complications or secondary radiation-induced malignancies. Other examples are when dose escalation is aimed for or in previously irradiated cases. For the histologies mentioned above, there is evidence for the use of proton as well as carbon ion therapy in combination with surgery or as an exclusive local treatment option. According to the literature, proton treatment is used more in pediatric patients and in combination with surgery. Here, local control rates up to 70–85% at 3–5 years can be reached. If surgery is not possible, the reports on definitive carbon treatment are increasing with reported local control rates of 70–94% at 3–5 years (Table 18.1). All together, the data presented suggest that the use of particle therapy is reasonable and offers a good therapeutic alternative, which can avoid, i.e., mutilating surgery in many cases.

However, clear recommendation to select between the indication for protons or carbon ion cannot be given based on the reported data. Randomized trials comparing protons vs carbon ions in bone tumors are limited to the ISAC protocol for sacrococcygeal chordoma [5]. However, in cases when surgery is possible without major mutilation, a combination with proton treatment especially if gross tumor resection was performed is indicated. In cases of inoperability or major mutilation, definitive carbon treatment offers a valuable treatment choice (Table 18.1).

18.3 Conclusion

The management of pelvic bone tumors remains very challenging. The main reason for that is the limited radical resectability, so additive treatment modalities should be used. Even in cases where resectability can be reached, impaired functional outcome can be expected. In cases of implant reconstruction, deep wound infection is still a major problem.

Particle therapy opens the window of new therapeutic options aiming at improving not only the "classical" outcome parameters like local control and survival but also functional outcomes.

In the case of sacral chordoma, the recommendation for using carbon ion or proton radiotherapy for definitive treatment after biopsy only or in patients who refused surgery was postulated by the Chordoma Global Consensus Group [34]. Also, in cases of local-regional recurrent chordoma, particle therapy should be considered [35]. ESMO–PaedCan–EURACAN Clinical Practice Guidelines recommend also new techniques (e.g., proton and carbon ion beam RT), particularly for unresectable primary osteosarcoma and chondrosarcoma [36].

Table 18.1 Treatment outcomes of particle radiotherapy for pelvic bone tumors

Author	Diagnosis	Patients (n)	Particle	Local control	Overall survival
Imai et al. (2010) [2]	Chordoma	38	Carbon ion	89% (5y)	86% (5y)
Imai et al. (2011) [3]	Chordoma	95	Carbon ion	88% (5y)	86% (5y)
Imai et al. (2016) [4]	Chordoma	188	Carbon ion	77.2% (5y)	81.1% (5y)
Uhl et al. (2015) [5]	Chordoma	23 33	Photon/carbon ion Carbon ion	79% (2y)	100% (2y)
Mima et al. (2014) [6]	Chordoma	23	Carbon ion	94% (3y)	83% (3y)
DeLaney et al. (2019) [11]	Chordoma, chondrosarcoma, and others	50	Photon/proton± surgery	78% (5y)	87% (5y)
DeLaney et al. (2014) [12]	Chordoma, chondrosarcoma, and others	50	Photon/proton± Surgery	74% (8y)	65% (8y)
Kabolizadeh et al. (2017) [12]	Chordoma	40	Proton/proton± Surgery	85.1% (5y)	81.9% (5y)
Chen et al. (2013) [13]	Chordoma (spine)	24	Proton	81.5% (5y)	91% 85y)
Staab et al. (2011) [14]	Chordoma	40	Proton ± surgery	62% (5y)	80% (5y)
Rombi et al. (2013) [15]	Chordoma, Chondrosarcoma, and pediatrics (skull base, spine, sacrum)	26	Proton ± surgery	81% (5y) 80% (5y)	89% (5y) 75% (5y)
Demizu et al. (2017) [16]	Chordoma, Chondrosarcoma, and osteosarcoma (skull base, spine, and sacrum)	72 20 4	Proton ± surgery	55.6% (5y)	70.7% (5y)
Weber et al. (2017) [21]	Ewing sarcoma Axial and pelvic	38 27	Proton ± surgery	81.5% (5y) 75.1% (5y)	85% (5y) 80.6% (5y)
Rombi et al. (2012) [22]	Ewing sarcoma	30	Proton ± surgery	86% (3y)	89% (3y)
Kamada et al. (2002) [28]	Osteosarcoma, Chordoma, Chondrosarcoma, and PNET[a], MFH[b], and other, Soft tissue sarcoma	15 11 6 5 1 3 16	Carbon ion	73% (3y)	46% (3y)
Matsumoto et al. (2013) [29]	Osteosarcoma, Chondrosarcoma, Chordoma, MFH[b], Ewing sarcoma, and Other	13 13 9 7 2 4	Carbon ion	79% (5y)	52% (5y)
Mohamad et al. (2018) [30]	Osteosarcoma Pelvis [24], trunk	26	Carbon ion	69.9% (3y) 62.9% (5y)	50% (3y) 41.7% (5y)
Demizu et al. (2017) [32]	Chordoma, Chondrosarcoma Osteosarcoma MFH[b] Other	53 14 10 5 9	Proton (52) Carbon ion (39)	92% (3y)	83% (3y)
Ciernik et al. (2011) [33]	Osteosarcoma	55	Proton or photon/ proton + surgery	82% (3y) 72% (5y)	67% (5y)

[a]*PNET* Primitive neuroectodermal tumor
[b]*MFH* Malignant fibrous histiocytoma

There are no randomized trials directly comparing protons and carbon ions published so far. One randomized trial is currently recruiting patients for sacral chordoma [5].The limited reported evidence shows no difference between using protons vs carbons. However, looking into biological behavior of these particles, carbons should be considered in radioresistant tumor histologies and in large nonresectable tumor volumes, where a big tumor burden has to be managed.

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Chemotherapy for Pelvic Bone Tumors

19

Antonella Brunello and Vittorina Zagonel

19.1 Introduction

The vast majority of pelvic bone tumors are secondary, due to metastatic spread of cancer of other primary sites and hematopoietic neoplasms such as multiple myeloma or solitary plasmacytoma of bone.

Primary bone tumors are rare, accounting for less than 0.2% of malignant neoplasms as reported in the EUROCARE (European Cancer Registry-based study on survival and care of cancer patients) database [1].

Medical treatment of metastatic bone tumors varies widely according to primary tumor type and may be highly effective especially for endocrine-sensitive tumors (i.e., breast cancer and prostate cancer). Multidisciplinary discussion must be therefore undertaken before making definitive decisions.

In many cases, treatment of bone metastases from solid tumors or myeloma includes the use of antiresorptive bone agents (i.e., zoledronic acid and denosumab), beside antitumor-directed therapy.

Both zoledronic acid and denosumab are generally administered monthly, but for patients with tumors which have good prognosis (i.e., endocrine-sensitive breast cancer with bone metastases only) there is evidence supporting less-intensive schedules [2, 3].

Primary malignant bone tumors are chondrosarcoma, osteosarcoma, Ewing sarcoma, high grade sarcomas, and chordomas. Giant cell tumors, which are locally aggressive benign tumors that may rarely metastasize, are also among primary tumors of bone, which may localize in the pelvis.

Given the rarity of sarcoma, it is of utmost importance to have initial diagnosis and treatment guided by a specialized multidisciplinary tumor board, and recent evidence suggests that such an approach is associated with improved survival [4].

19.2 Osteosarcoma

Osteosarcoma is the most common type of bone sarcoma in children and young adults, along with Ewing sarcoma, but it can also occur in older subjects. In children and young adults, osteosarcoma is most frequently located in the extremity, with an increasing proportion of axial localization with age.

Chemotherapy has been recognized as essential for the treatment of high-grade osteosarcoma since the pivotal study by Link and colleagues [5]. Indeed, before the introduction of systemic therapy, cure rates for osteosarcoma were less than 20%, even among patients who presented

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with no detectable metastatic disease. Since that time, evidence has mounted on the presence of micrometastatic disease at the time of diagnosis in most patients, providing the background for the role of systemic therapy in achieving cure. The introduction of combination chemotherapy improved the probability for cure to rates as high as 70% [5]. Cytotoxic drugs, which have been shown to be active in osteosarcoma, are Doxorubicin, Ifosfamide, Methotrexate, and Cisplatin [6], yielding objective response rates, when used as single agents, of 43%, 33%, 32%, and 26%, respectively.

Conventionally, surgery of the primary tumor is performed after preoperative chemotherapy, an approach pioneered in the 1970s by Rosen and colleagues [7]. Although no survival advantage of neoadjuvant compared to post-operative chemotherapy has been demonstrated, and surgery is rarely influenced, obtaining symptom improvement, time available for surgical planning, and availability of the primary tumor for assessment of histologic response to preoperative chemotherapy have made this approach a standard of care for the majority of patients. In specific cases, such as when diagnostic uncertainty exists or when demolitive surgery cannot be avoided, then immediate resection could be suggested.

When osteosarcoma is treated in the neoadjuvant setting (that is, before surgery), necrosis in the primary tumor can be assessed at the time of definitive surgical resection. Chemotherapy-induced necrosis strongly correlates with event-free survival (EFS) and overall survival (OS) [8], with high necrosis in the primary tumor correlating with lower probability of recurrence and death. Radiological response likely underestimates real antineoplastic activity, as osteosarcoma may present with substantial necrosis following chemotherapy without change in dimensions due to the osteoid matrix produced by the tumor.

Chemotherapy regimens based on a combination of high-dose methotrexate, doxorubicin, and cisplatin (the so-called "MAP" regimen) have been shown to provide 3-year EFS roughly around 70% [9, 10], with differences in outcome strongly correlated with the histological response in terms of necrosis.

Based on these observations, several trials have investigated the possibility of improving outcomes for patients with tumors exhibiting low necrosis following initial chemotherapy. In the European and American Osteosarcoma Study Group (EURAMOS) trial, patients received neoadjuvant therapy with the MAP regimen for 10 weeks and were randomized after definitive surgery to different adjuvant treatments according to necrosis at definitive surgery. Patients with good histologic response were randomized to receive either continuation of MAP or MAP with the addition of interferon-alfa, whereas patients with poor histologic response were randomized to receive either continuation of MAP or MAP with the addition of high-dose ifosfamide and etoposide [11]. The addition of interferon to MAP did not improve the risk of events for patients with higher necrosis [10]. The addition of high-dose ifosfamide and etoposide to MAP did not improve the risk of events for patients with less necrosis [12].

Since the first pivotal studies on neoadjuvant chemotherapy, very little progress has been made with regard to further improvement of survival rates obtained with the MAP regimen.

The Intergroup Study 0133 studied the role of liposomal muramyl tripeptide (MTP), a derivative of the Bacillus Calmette-Guérin cell wall, which stimulates macrophages' response against tumor cells, in an adjuvant randomized trial, which showed that the addition of MTP to standard chemotherapy provided a trend toward improved EFS and a statistically significant improvement in overall survival [9].

In light of the results of this trial, MTP has been approved for use in combination with chemotherapy in the treatment of localized osteosarcoma for patients age 2–30 years in several Countries.

Pelvic primary site is associated with a more dismal prognosis compared to patients with primary osteosarcoma of the extremities, with a 5-year survival of about 30% compared to 5-year survival for localized extremity osteosarcoma of approximately 70%, [8, 13]. The poor survival of patients with pelvic osteosarcoma is multifactorial, including large tumor volume at presentation,

more problematic surgery due to higher morbidity, difficulty in achieving adequate surgical margins, higher likelihood of metastatic disease at presentation, inferior necrosis after preoperative chemotherapy, and presence of macroscopic tumor emboli in the large regional vessels [14–19].

With recurrent osteosarcoma, chemotherapy options are quite limited. The combination of ifosfamide and etoposide appears to be one of the most active regimens for these patients [20, 21].

The combination of gemcitabine and docetaxel has also been investigated for patients with recurrent osteosarcoma [22] with findings that indicate only modest activity.

Newer agents are being studied. Currently, for relapsed osteosarcoma, there is evidence of activity of antiangiogenetic agents, and data from phase II trials with sorafenib, also in combination with everolimus, and regorafenib have been recently published [23–26], with median progression-free survival times of about 4 months.

19.3 Ewing Sarcoma

Before the introduction of systemic chemotherapy, Ewing sarcoma had a cure rate of less than 10%, even among patients who presented with localized disease [27]. The outcomes for patients with Ewing sarcoma have dramatically improved since then, with as many as 70% of patients presenting with localized disease achieving long-term event-free survival (EFS) with the use of multiagent chemotherapy [28–30].

Survival rates fall down to 30–50% if patients present with lung metastases and to less than 20% when patients present with metastases to distant bones or bone marrow [28].

Being generally highly sensitive to chemotherapy, Ewing sarcoma treatment involves primary chemotherapy before local treatment.

Cytotoxics with documented activity, which are widely used in treatment protocols for Ewing Sarcoma, are Doxorubicin, Ifosfamide, Cyclophosphamide, Etoposide, Vincristine, and D-Actinomycin.

Similar to osteosarcoma, the percentage of necrosis postchemotherapy is associated with prognosis, with higher necrosis associated with better outcomes.

The Intergroup trial INT-0091conducted by the Pediatric Oncology Group and the Children's Cancer Group demonstrated that the addition of ifosfamide and etoposide to cyclophosphamide, doxorubicin, and vincristine significantly improved outcomes for patients with localized Ewing sarcoma [28]. Among patients with localized disease, patients randomized to cyclophosphamide, doxorubicin, and vincristine alternating with ifosfamide and etoposide had a statistically significant better 5-year EFS compared to patients randomized to cyclophosphamide, doxorubicin, and vincristine alone (69% and 54%, respectively). The addition of ifosfamide to etoposide did not improve outcomes for patients with metastatic disease at initial presentation, though [31].

In contrast to other bone and soft tissue tumors, Ewing sarcoma outcomes seem to improve with dose intensification.

A single-center study from the Memorial Sloan Kettering Cancer Center reported a high rate of EFS with the use of very high-dose alkylating agent therapy given over a shorter duration of just 21 weeks [32]. In a Children's Oncology Group trial, dose intensification was obtained by shortening the interval between chemotherapy cycles [30], with the administration of the usual five-drug combination every 2 weeks for 28 weeks, achieving better outcomes compared to the five-drug combination administered every 3 weeks for 42 weeks. Patients randomized to the interval-compressed arm had a significantly greater 5-year EFS (73% versus 65% for patients randomized to the standard arm).

In the Euro-E.W.I.N.G.99 trial, patients with localized Ewing sarcoma were randomized between high-dose chemotherapy with busulfan and melphalan or standard chemotherapy (vincristine, dactinomycin, and ifosfamide, seven courses) if they were at high risk for relapse (either poor histologic response after receiving six courses of chemotherapy with vincristine, ifosfamide, doxorubicin, and etoposide or with tumor volume at diagnosis ≥200 mL if unresected, or initially resected, or resected after

radiotherapy) [33]. In an intent-to-treat analysis, the 3- and 8-year EFS was 69% and 60.7% with high-dose chemotherapy versus 56.7% and 47.1% with standard chemotherapy, respectively. Overall survival (OS) also favored high-dose chemotherapy, with 3- and 8-year OS compared to standard chemotherapy of 78% vs 64.5% and 72.2% vs 55.6%, respectively.

Patients with recurrent Ewing sarcoma have a number of systemic therapy options. Historically, patients used to be retreated with chemotherapy combinations used as part of initial therapy, with some responses and durable remissions reported [34]. There is evidence suggesting activity of high doses of ifosfamide (15 g/m^2) in patients with recurrent Ewing sarcoma who were previously treated with lower doses as part of initial therapy [35].

Currently, patients with recurrent Ewing sarcoma are candidates for clinical trials of novel agents or may be treated with a number of salvage chemotherapy regimens with documented activity in this setting.

Campothecin-based regimens are currently among the most active available chemotherapy regimens for patients with relapsed Ewing. The combination of topotecan with cyclophosphamide has shown activity in this population [36–38]. The combination of irinotecan and temozolomide has also shown activity in patients with relapsed Ewing sarcoma [39–42].

Also, the combination of gemcitabine with docetaxel has shown some activity in patients with recurrent Ewing sarcoma [43].

The first interim results of the currently ongoing rEECur trial, assessing these regimens prospectively in a randomized manner, have recently been reported [44]. In this trial, patients with refractory or recurrent Ewing sarcoma were randomized to receive either topotecan and cyclophosphamide, irinotecan and temolozomide, and gemcitabine and docetaxel or high-dose ifosfamide. Patients randomized in the gemcitabine and docetaxel arm had 11.5% response rate, median progression-free survival of 3 months, and median OS 13.7 months. After assessing the probabilities that overall response and progression-free survival were better for gemcitabine and docetaxel than for each other arm, all comparisons favored the other arms, and the gemcitabine and docetaxel arm was dropped, with recruitment continuing in the remaining arms.

Several targeted agents have been studied, such as mammalian Target Of Rapamicin (mTOR) inhibitors [45] or Insulin-like Growth Factor-1 inhibitors [46], with no one proving enough activity to warrant approval. Furthermore, despite understanding the critical role of EWSR1 fusion oncogenes in the pathogenesis of Ewing sarcoma, strategies to target EWSR1 fusion oncogenes and oncoproteins have so far been difficult to develop.

One of the strongest prognostic factors associated with a poor outcome of patients with localized Ewing sarcoma is primary tumor located in the pelvis. Half of the cases of Ewing sarcoma arise in the extremity bones, followed by pelvis, ribs, and vertebrae [47]. The most common sites within the pelvis are the iliac bones followed by the pubis [48]. The higher incidence of primary pelvic sites for Ewing sarcoma in adults is one of the possible reasons for poorer outcomes in older subjects compared to younger ones. Also, since metastatic disease is the most important prognostic factor for Ewing sarcoma, the worse prognosis of tumors localized in the pelvis is at least partly related to a higher percentage of metastatic cases at diagnosis [49–52].

The optimal strategy of addressing local therapy in pelvic disease with surgery, radiation therapy, or both has been long investigated, yet remains highly controversial [53].

Indeed, the time interval between chemotherapy initiation and definitive local therapy impact event-free survival, with longer intervals being associated with unfavorable outcomes [54].

In pelvic primary localization, the timing of local therapy commonly relies on multidisciplinary assessment, being dependent on tumor size, specific location, and response to neoadjuvant chemotherapy. As a result, the decision as to when to offer local therapy for large pelvic tumors often involves a thorough multidisciplinary discussion among medical, radiation, and orthopedic oncologists. In patients with chemo-

responsive disease, whether the maximal benefit of chemotherapy should be pursued to facilitate the best possible surgical resection or radiation of the smallest tumor volume is still matter of debate.

On the one hand, continuing chemotherapy to maximum response could be helpful to reduce total tumor volume and would avoid the risk of an interruption with local therapy, given the fact that recovery from surgery for axial tumors is often prolonged, making it difficult to timely resume chemotherapy. Moreover, the delivery of concomitant radiation therapy and chemotherapy can be sometimes difficult, especially in older patients. On the other hand, delaying local therapy may not be beneficial. In some cases, radiation therapy in the absence of surgery could provide sufficient local control of Ewing sarcoma, particularly those arising in the sacrum, which displays a better prognosis than Ewing sarcoma of the hip bones [55], probably at least in part due to smaller tumor volumes in the sacrum than in the hip bones. So far, no randomized controlled trials have directly compared radiation and surgery.

Despite historical data of two to threefold increased local failure and decreased survival in primary Ewing sarcoma of the pelvis compared to other sites, more recent series report local and distant control rates for pelvic Ewing's sarcoma that approach those of nonpelvic [53, 56–59] and this may be due to optimizing chemotherapy doses and schedules with improved supportive therapy, better radiation techniques, and more aggressive surgery.

19.4 Chondrosarcoma

Chondrosarcoma is the most frequent bone sarcoma of adulthood, and it is primary located in the extremities and the axial skeleton, with the pelvis representing the most common location.

Typically, chondrosarcomas are low-grade, locally aggressive, nonmetastasizing tumors (grade I), but in some cases, chondrosarcoma can be high grade (grades II–III) [60]. Chondrosarcoma of the pelvis is often high grade and is almost twice as likely to present with metastatic disease at presentation compared with high-grade chondrosarcoma of the extremities [61]. Globally, high-grade chondrosarcoma of pelvic bones is associated with the highest rates of local recurrence and worst prognosis after surgical treatment [62].

The majority of chondrosarcomas display conventional subtype features, but some other subtypes have been described, namely, a mesenchymal and clear cell chondrosarcoma. Moreover, conventional chondrosarcoma can 'dedifferentiate' into a highly aggressive form, which is the dedifferentiated chondrosarcoma, typically yielding a very poor prognosis [63, 64].

In chondrosarcoma, unlike osteosarcoma and Ewing's sarcoma, chemotherapy and radiation therapy have not been proven to be effective neither as adjuvant treatment nor for distant control, and therefore, surgery is the primary treatment.

Inoperable, locally advanced and metastatic high-grade chondrosarcomas have a poor prognosis [65], and this may also be related to the inefficacy of chemotherapy in conventional chondrosarcoma.

However, there is some evidence for chemosensitivity of mesenchymal chondrosarcoma, thus suggesting a role for adjuvant or neoadjuvant therapy with Ewing-type regimens [66, 67].

Dedifferentiated chondrosarcoma is commonly treated as a high-grade bone sarcoma, with systemic and local therapies that need to be adapted to patient's age [65, 68]. Among available drugs, beside anthracyclines, ifosfamide, and cisplatin, some activity of gemcitabine in combination with docetaxel has been reported [69].

19.5 Chordoma

Chordoma is a very rare mesenchymal neoplasm arising from embryonic remnants of the notochord in axial skeleton, with a reported yearly incidence of approximately 0.08/100,000 people [70, 71], and it affects predominantly the mobile spine and the sacrum in older adults [72].

Conventional chordoma is a low-grade, locally invasive malignancy. Immunohistochemistry nuclear positivity for Brachyury is the diagnostic hallmark [73].

Although typically slow-growing, the natural history of chordoma is marked by a high tendency toward local recurrence, with reported local failure in approximately 40–50% of patients undergoing surgery [74].

Dedifferentiated chordomas account for less than 5% of all cases, with brachyury expression that can be lost, and behave more aggressively than the conventional counterpart. Approximately 30% of patients with chordoma will develop metastases, usually late in the natural history of the disease and mostly after local recurrence.

The mainstay of treatment for chordoma is surgery and/or radiation therapy. When a local relapse occurs, it carries very poor survival rates and local control is hardly achieved [70].

Chemotherapy for advanced chordoma is generally inactive, yet there is some evidence of activity of tyrosin-chinase inhibitors, such as imatinib and sorafenib, which can provide stabilization of disease and nondimensional tumor responses, and studies of epidermal growth factor receptor and mTOR inhibitors are ongoing [75–77].

19.6 High-Grade Sarcomas of Bone

High-grade spindle/pleomorphic sarcomas constitute a heterogeneous group of primary malignant bone tumors that do not fulfill the histological criteria for a diagnosis of osteosarcoma, chondrosarcoma, or Ewing sarcoma [78].

Vascular sarcomas, namely, epithelioid hemangioendothelioma and angiosarcoma, may occasionally arise in the bone. Angiosarcoma is highly aggressive, with a poor prognosis, whereas epithelioid hemangioendothelioma has an intermediate biological behavior. Vascular tumors of bone may present as unifocal or multifocal disease [79].

Given the rarity of these forms, the evidence is limited regarding treatment and outcomes. For patients with angiosarcoma of bone, improvements in survival rates may be obtained with the use of adjuvant chemotherapy based on cisplatin, doxorubicin, and ifosfamide, but there are reports of the efficacy of taxanes as well [80].

For epithelioid hemangioendothelioma, which is advanced or metastatic and not amenable to local treatment, sirolimus has been reported to display some activity [81].

19.7 Giant Cell Tumors of Bone

Giant cell tumors are benign, locally aggressive, and rarely metastasizing intramedullary bone tumors composed of mononuclear cells and osteoclast-like multinucleated giant cells, with a variable and unpredictable potential for aggressive growth [82]. Giant cell tumor of bone can localize in the pelvic bone although this is not very common, with the acetabular area being the most frequent site [83].

Giant cell tumor cells typically present with a high expression of Receptor Activator of Nuclear factor Kappa-B (RANK)/RANK-ligand (RANKL), and Denosumab, a RANKL inhibitor, has been successfully used to treat unresectable or metastatic disease [84].

Denosumab may provide an option for treatment of initially locally advanced tumors when used as neoadjuvant therapy to facilitate complete surgical resection or avoid mutilating surgery [85].

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Role of Plastic Surgery in the Treatment of Pelvic Tumors

20

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Primary pelvic tumors are rare, and the principal goal of the surgical treatment is to obtain a free margin resection. Although the majority of patients can be treated with limb-salvaging internal hemipelvectomies [1, 2], hindquarter amputations and hip disarticulation are still performed. In order to extirpate completely the tumor, the reconstructive surgeon may be faced to large soft-tissue defects. A concomitant one-step autologous reconstruction, mostly using myocutaneous or fasciocutaneous flaps, has been proved to lead to better results, thanks to the well-vascularized tissues, which are used to cover defects, grafts, and implants or just to obliterate the dead space. Moreover, these flaps can be hard to be performed in a second time, especially when radiotherapy or infections compromise the donor site.

The reconstruction of periacetabular and sacral defects, after pelvic tumor resection, can be classified among the most challenging procedures in orthopedic oncology and plastic reconstructive surgery. This scenario, in fact, is charged by a high rate of complications and dissatisfying mechanical and nonmechanical results: infections and wound dehiscences are common, especially when associated with radiotherapy. Nevertheless the poor cosmetic, physical and psychological outcomes, and soft-tissue reconstructions can improve the quality of life of the patients. Over years, these techniques allowed even more aggressive resections.

The purpose of this chapter is to provide an overview of the most used reconstructive techniques following bone tumor resection in the pelvic district. The hardest part to create a reconstructive algorithm is that each patient needs a tailored solution, due to the high complexity and variability of these cases.

20.1 Flap Planning

The reconstructive surgeon must be involved in the preoperative multidisciplinary meetings in order to plan the possible solutions. Even though, in any case, no risks must be taken not to completely extirpate the tumor, the multidisciplinary team approach can better plan collateral procedures, such as colostomy or ileostomy, leaving open many reconstructive techniques. Imaging exams must be acquired and collectively discussed. The presence of vascular pedicles must be detailed, usually in our practice with an

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Angio-CT, and the operative plan should be agreed and proposed to the patient. For sacral resection, we usually prefer to place ureteral stents that can help their identification during the tumor resection. Patient positioning depends mostly on the location of the tumor. We advise to mark the skin in the preoperative setting. Usually, we start with the patient in the supine position for periacetabular tumor and in the prone position for the tumor of the sacrum, except for those involving S2 or higher levels where we prefer to perform an anterior abdominal exploration and a posterior resection. The eventual changing of the position of the patient can be needed for the flap dissection.

In our experience, flaps using tissue from rectus abdominis muscle or from the gluteal area allow the best coverage for sacral resections, providing a moderate quantity of tissue to fill the dead space. Other solutions might be the omental flap or the combination of the previous techniques.

20.2 Rectus Abdominis Musculocutaneous Flap

The rectus Abdominis Musculocutaneous flap is type III according to the Mathes and Nahai classification. It can be used as a muscular or as a musculocutaneous flap, allowing several types of reconstructions. It can be named differently based on the orientation of its skin paddle: transverse rectus abdominis musculocutaneous (TRAM) flap or vertical rectus abdominis musculocutaneous (VRAM) flap. It is supported by two main vessels: the deep inferior epigastric artery (DIEA), which is the principal one, and the superior epigastric artery. The rectus abdominis muscle measures about 30 x 10 cm and can be used as pedicled or microsurgical flap. The principal pedicle, the DIEA, measures 14–18 cm in length and has a caliber of about 3–4 mm. It arises from the external femoral artery and travels superomedially in the extraperitoneal tissue piercing the transversalis fascia. After leaving several and constant branches, it enters the rectus sheath passing anterior to the arcuate line. Normally, it divides into two or three major vessels that run cranially into the muscle belly, originating several muscular or musculocutaneous perforators. These vessels, which are usually bigger when originate from the medial raw, supply the skin paddle, which must be placed over them. Among our institute, an angio-TC, if not yet performed, is usually requested to investigate the abdominal wall vessels, because several surgical procedures can have damaged deep inferior epigastric vessels, such as colostomy, appendectomy, etc. The main venous drainage is granted by the deep inferior epigastric vein, which is connected to the external iliac vein.

The VRAM is one of the principal solutions for both periacetabular (Fig. 20.1) and sacral (Fig. 20.2) reconstructions. The orientation of the skin paddle is decided based mainly on the extension and on the location of the skin defect. In the case of periacetabular resections or sacral low resections, when the patient can be placed in the supine position for all the time, a simultaneous flap elevation can be performed by secondary equipment [3]. If a change of position is mandatory, the flap elevation can be performed prior to the abdominal exploration, if needed, or after the posterior resection, if the entire demolitive time can be completed in the prone position. Usually, the rectus sheath is approached medially, far from the skin paddle in order not to damage skin perforators. The muscle can be easily separated from the posterior fascia, but care must be taken to handle the pedicle, which usually lies on the posterior surface of the muscle belly. If flap elevation is performed prior to turning the patient, the authors advise to leave the pubis insertion attached, in order to avoid pedicle twisting and to put the flap in a plastic bag and leaving it where it can be clearly retrieved and managed from the posterior approach.

In the case of large defects of the fascia, it can be reconstructed placing a synthetic mesh or an acellular dermal matrix. These solutions can be used to repair anterior defects of the donor site, posterior defects, or both. These repairs should

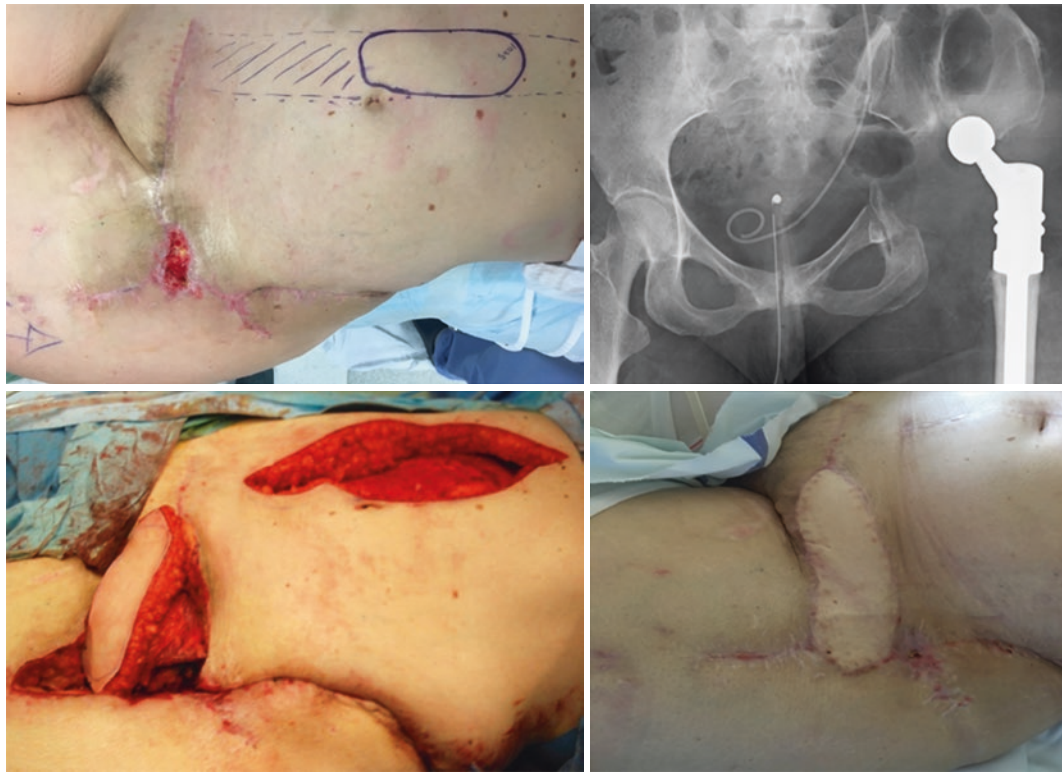

Fig. 20.1 Delayed wound healing in a patient previously treated for aseptic loosening of acetabular component after sarcoma resection, undergone a revision with custom-made prosthesis and soft tissue reconstruction with a pedicled VRAM flap

be tension-free. Even though both alternatives have been described, many Authors still prefer the synthetic options, especially to avoid long-term hernias or bulging, which are costly and hard to treat. Care must be taken leaving a gap posterior to make the pedicle passing through. It is very important that the mesh doesn't decubitate on the vessels and its course is regular and without twistings or kinkings. Finally, the insetting of the flap must be tension-free to avoid necrosis, flap loss, or wound cracking during the change of position.

Advancement SGAP (superior gluteal artery perforator) or IGAP (inferior gluteal artery perforator) Flaps.

The advancement SGAP flaps are fasciocutaneous flaps that can eventually include the underneath muscle. The myocutaneous technique should be considered only in paraplegic patients; otherwise, a severe dysfunctional deambulation is caused. In this case, the flap is considered a type III Mathes–Nahai classification. Usually, they are proposed for partial or total sacrectomy (Fig. 20.3). Even though it has been described, the microsurgical employments are rare. These flaps are irrorated by several perforator vessels coming from the superior and the inferior gluteal arteries. The superior gluteal artery is the largest branch of the internal iliac artery, which is the posterior division. It originates above from the upper border of the piriformis muscle, dividing soon into a superficial and a deep branch. The deep one runs vascularizating the gluteus medius and the iliac bone, while the superficial one the upper portion

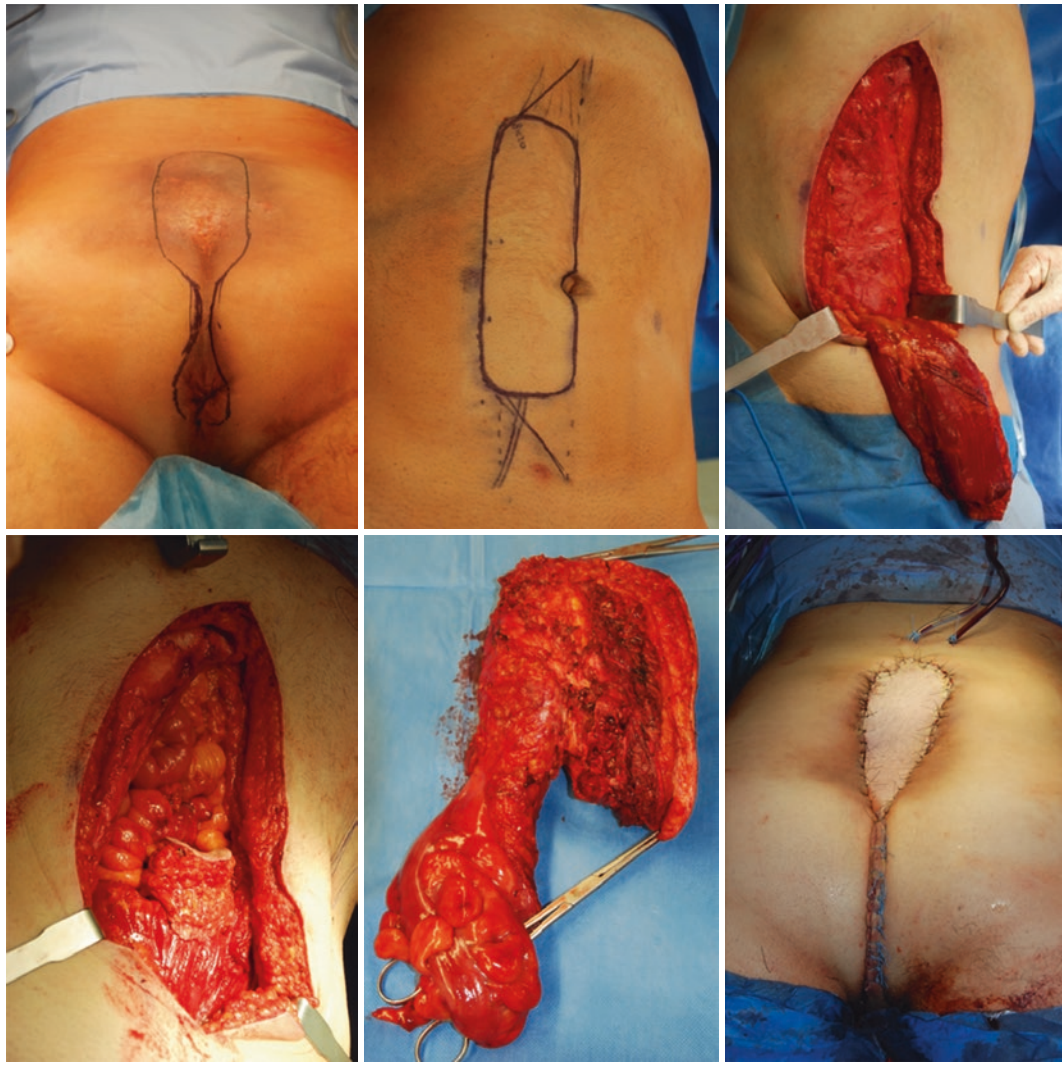

Fig. 20.2 Sacrectomy for chordoma resection, undergone colostomy surgery and repaired with a pedicled VRAM flap

of the anatomic area. From this last branch, our perforators of interest usually originate.

The inferior gluteal artery (IGA) represents the terminal branch of the anterior division of the internal iliac artery. This artery travels with the greater sciatic nerve through the greater sciatic foramen, and then, it pierces the sacral fascia toward the surface. Normally, under the inferior portion of the gluteus maximus, perforators are given off to supply the overlying fat and skin (Fig. 20.3). These last vessels run more oblique than those originated from the superior gluteal artery. The mean length of the completed dissected pedicles is 5–7 cm for the superior gluteal flap and 7–10 cm for the inferior gluteal flap, but normally a minimal subfascial dissection is required, avoiding risks. When longer movements are necessaries, we prefer to use other techniques, usually the VRAM flap, also because they're most likely associated with the need of filling the dead space. The venous drainage accompanies the arterial system.

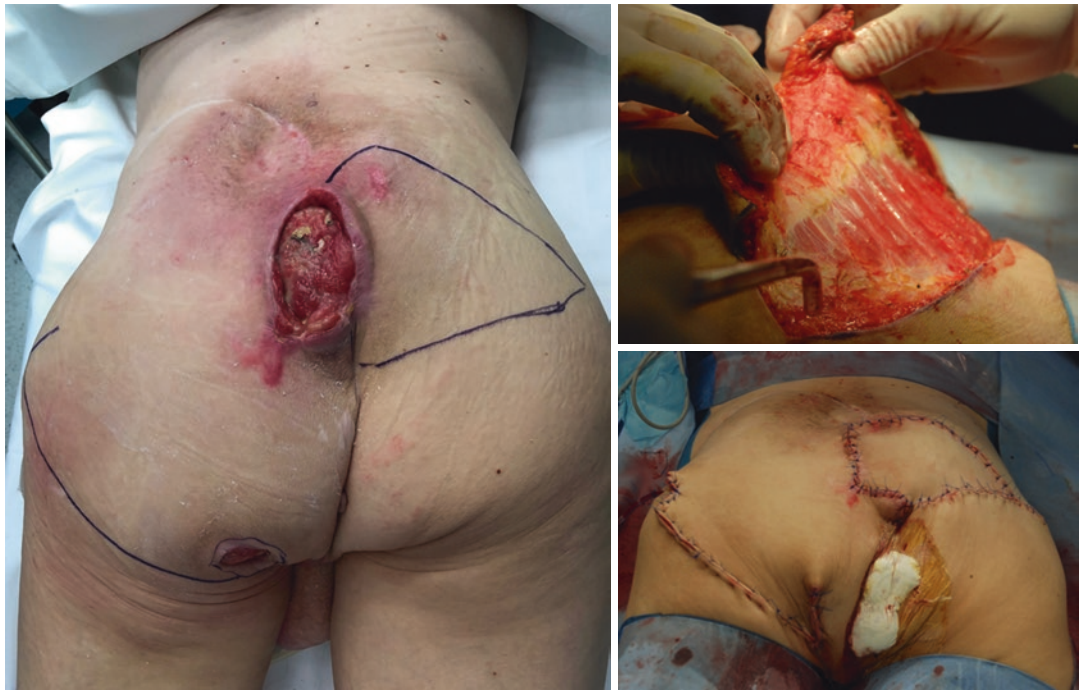

Fig. 20.3 Decubitus sacral and ischial ulcer in unresectable chondrosarcoma, repaired with advancement fasciocutaneous SGAP flap for sacral region and IGAP flap for the ischial region

20.3 Anterolateral Thigh (ALT) Flap

The anterolateral thigh (ALT) flap is a well-known and reliable flap. Although it is a workhouse technique, in sacral and periacetabular reconstructions, it can be performed in very rare situations, mainly using it pedicled to cover groin or perineal defects. The pedicle originates from the lateral circumflex femoral artery (LCFA). This branch of the profunda femoris artery originates 8–10 cm inferior to the anterior superior iliac spine, and it divides into three branches: ascending, transverse, and descending. The descending one runs on the medial edge of the vastus lateralis giving off several perforator vessels to the fascia and the overlying fat and skin. Most of these perforators have an intramuscular path, while few can present a septal one. The skin island is generally taken centrally over the chosen perforator, with a variable dimension based on the defect size. Although a larger width than 8 cm may not be closed primarily, greater dimensions have been described. The pedicle length is around 11 cm with a caliber of 2.1 mm. The venous drainage is granted by a comitantes system, which finally drains in the greater saphenous vein. This flap can be raised as fasciocutaneous, fascial, composite, including a portion of the rectus femoris muscle, or chimeric with a part of the vastus lateralis muscle. Preoperative Angio–CT scan is mandatory to properly plan the flap, especially in these patients where there can have much damage of this vascular system. The flap is raised medially to laterally, in the subfascial plan, looking for the chosen perforator. Once identified, the perforator is carefully dissected through the muscle to its origin from the descending branch of the LCFA (Fig. 20.4). Depending on the necessity of the pedicle length, the rising of the LCFA can be continued proximally.

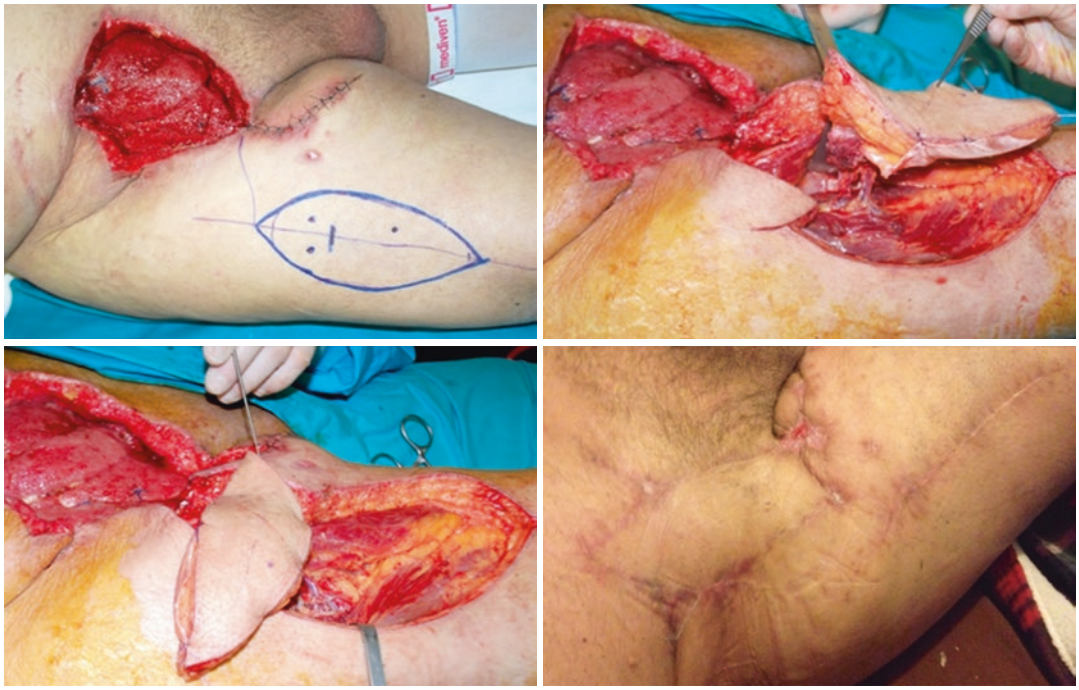

Fig. 20.4 Wound dehiscence after sarcoma and lymphatic resection, repaired with a pedicled ALT flap

20.4 Tensor Fascia Latae (TFL) Flap

The tensor fascia latae (TFL) flap is a type I Mathes Nahai. It can be considered a reliable flap for the coverage of the trochanteric, periacetabular, perineum, and abdominal wall as pedicled, and it has also been employed as microsurgical flap. It can rise both as muscular or musculocutaneous flap. This muscle originates from the external border of the iliac crest between the sartorius and the gluteus medius with a tendon 5 cm wide. It descends as a band, inserted onto the iliotibial band. The pedicle is represented by the transverse branch of the LCFA, while the descending has been already described and the ascending branch travels to the gluteus minimus muscle. The transverse branch divides into three branches before entering the TFL muscle at a point around 8 – 10 cm inferior to the anterior superior iliac spine. It has a length of 4 – 6 cm and a caliber of 2 – 3 mm. A small fragment of iliac crest bone can be included if needed, preserving little branches that reach it. Venous drainage is usually granted by the comitantes system. The skin island can be 20 cm long and 10 cm wide, even though only a width of 8 cm consents a primary closure of the donor site.

20.5 Disarticulation and Hemipelvectomy

Disarticulation of the hip is the surgical removal of the entire lower limb through the hip joint, while the hemipelvectomy requires the ablation of the entire or a portion of the iliac bone [4].

Disarticulations or Hemipelvectomies are extremely mutilating procedures, but sometimes they are still required to obtain a radical resection. Standard techniques are Boyd's method, and the posterior flap is described by Slocum. Alternative options are represented by the anterior flap, or "anomalous flap", or adapted modified techniques. First, inguinal or iliac lymph nodes can be removed or not depending on the preoperative planning. Boyd described an anatomic strategy, which is still the basic procedure. It is oriented to the transection of the muscles at

their origin to minimize the blood loss and to provide a well-padded stump to grant a weight-bearing surface for prosthesis.

Slocum's technique employs a posteromedial flap to cover the stump, which can be used for both hip disarticulation and hemipelvectomy. This flap is about 10 cm long or more, in order to gently cover the stump without tension. It is important to ligate the femoral vessels to make them falling above the inguinal ligament and to make the two branches of the obturator nerve retract, to avoid the pressure areas. The tensor fascia latae is divided at the level of the greater trochanter, where the muscle reaches it. The gluteus maximus is divided at the distal end to the posterior skin flap. This flap, containing this muscle, is finally rotated anteriorly to cover the stump. Although Boyd's technique is considered the basic technique, among our Institute, we prefer Slocum's technique to better provide a useful a nonpainful stump.

The standard hemipelvectomy is generally performed drawing a posterior or a gluteal flap to cover the defect. Standard hemipelvectomy disarticulates the symphysis pubis and sacroiliac joint, while in extended hemipelvectomy, the resection line passes through the sacrum and in conservative hemipelvectomy through the ilium above the acetabulum, leaving the iliac crest. Finally, internal hemipelvectomy is defined as a limb-sparing procedure [5].

Anterior flap disarticulation or hemipelvectomy is used for lesions of the buttock or posterior proximal thigh. It employs a quadriceps myocutaneous flap based on the superficial femoral artery [6].

20.6 Postoperative Management

Normally, these patients require at least one night in the ICU. We started using Air Fluidized Therapy beds, Clinitron©, especially for those patients undergone to a sacral resection, for not to let the weight bearing area on the flap. Obviously, the fluid resuscitation is mandatory to maintain an adequate perfusion and the choice to use hydrocolloids or blood product depends on the postoperative blood tests. Physical and rehabilitative programs are scheduled early in the postoperative time in order to make the patient sitting and deambulating soon.

20.7 Complications

Pelvic reconstruction surgery is usually associated with a high ratio of complications [1, 7]. Despite the use of the Clinitron© bed, the greatest number of them is related to the skin dehiscence, due to a problem in the surgical wound healing process [1, 8]. It is essential to properly and early treat this skin issues, mainly because they can contraindicate the start of adjuvant therapies (such as chemotherapy), which are essential in most of the cases. This need makes the wound healing care an absolute priority for the patient. To prevent wound dehiscences or infections, among our institute, we usually place one or multiple drains in order to avoid hematoma or seroma formations. We strongly believe that these are the first step through a surgical failure, due to the pression that an accumulation of fluids can place on the skin margins and on the flap itself. This process can lead to a reduction of the blood support. According to the oncologic principles, we place the drains exit on the skin near to the surgical wound, allowing an eventually subsequent en bloc resection. Speaking of irradiated wounds, large flaps, or sacral and posterior resection in general, we prefer to keep drains longer than what literature generally advises (less than 30 cc per day).

The incidence of a flap loss is low and depends mostly on the area and the type of flap used. Speaking of the sacrum, these problems regard mostly the V–Y advancement flaps, which are less likely to obliterate the dead space, allowing the hematoma or seroma formations. In a recent study, [9] it has been found that the use of acellular dermal matrix to reconstruct the posterior abdominal wall reduces the risk of posterior bowel herniations, bowel obstructions, and fistula formations. Due to these reasons, for posterior reconstructions, the Authors suggest to use gluteal flaps only for smaller defects, performing a VRAM flap plus dermal matrix for any greater demolition, especially for combined anterior and posterior approaches [1].

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21

Anesthesia for Pelvic Bone Cancer Surgery: From Risk Evaluation to Postoperative Course

Gian Mario Parise, Bianca Ferrarese, Alessandro Graziano, Manuela Funes, Francesco Ambrosio, and Paolo Navalesi

Orthopedic oncologic surgeries are challenging for the anesthesiologist. When planning anesthesia, numerous factors must be considered. Pelvic oncologic surgeries vary in length and complexity. Thus, a well-formulated anesthetic plan created by effective communication between the anesthesiologist and the surgeon is essential to ensure optimal patient outcomes [1, 2].

Preoperative assessment of the patient for anesthesia begins 2–3 weeks prior to the scheduled surgery date. Patients are seen by the anesthesiologist and they must receive a thorough preoperative evaluation to elucidate significant comorbidities or side effects secondary to chemotherapy and radiation [1, 2]. The anesthetic preoperative consultation should include full blood examination including baseline hemoglobin level, hematocrit, platelet count, iron and coagulation function (prothrombin time (PT) with international normalized ratio (INR), and activated partial thromboplastin time (aPTT)). Preoperative evaluation should also include ECG and chest radiograph. Based on the patient's medical history, physical exam, and test results, the anesthesiologist may also require further cardiac evaluation (echocardiography, stress testing) and/or pulmonary evaluation (spirometry, arterial blood gas). In this phase, postoperative ICU stay should be discussed according to the type of surgery and the patient's medical history.

The management of patients undergoing pelvic tumor surgeries is particularly complicated because of the possibility of massive hemorrhage [1–6]. According to studies, pelvic tumor surgeries show an intraoperative blood loss ranging from 2500 to 5000 mL [2, 4]. Although, in most circumstances, the administration of blood and/or blood products can be used to effectively correct hemoglobin concentrations and coagulation function, there is a growing body of evidence that shows the potential adverse effects of allogeneic blood product administration. These adverse effects include the transmission of infectious diseases, immunosuppression, transfusion-related acute lung injury, transfusion reactions, and they can also be associated with decreased tumor-free survival [2, 5–9]. Moreover, patients with comorbidities or those who have undergone chemotherapy and radiotherapy may often have significant anemia and thrombocytopenia. Advances in anesthesia, surgery, and transfusion medicine over the past decade have led to the development of "patient blood management," a multimodal, evidence-based preoperative and perioperative strategy, aimed to minimize the need for red blood cell transfusions, consisting of treating anemia and reducing perioperative blood loss [8, 10]. Depending on the cause and degree of

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anemia, the urgency of the procedure, and the anticipated blood loss, patients are given a specific therapy in order to improve their hemoglobin level. Individuals with iron deficiency or iron deficiency anemia should be treated with iron, allowing adequate time for effect of treatment before surgery (typically 2–4 weeks for correction). Intravenous (IV) iron is an option if less than 4–6 weeks are available, and for patients with poor tolerance or an inadequate response to oral iron. For individuals with anemia of chronic disease/inflammation, we typically administer preoperative EPO (together with supplemental iron to avoid functional iron deficiency) [8].

The table below shows patient blood management according to our procedure (Table 21.1).

A study also shows that preoperative sucrosomial iron supplementation at least 4 weeks prior to an elective surgery in non-anemic patients limits the drop in postoperative Hb levels, resulting in higher postoperative hemoglobin, quicker postsurgical recovery, shorter hospitalization, and decreased surgery-related costs [10].

Table 21.1 Preoperative patient blood management

1	LONG PREOPERATIVE TIME (4 WEEKS) • **low serum iron and low ferritin** (<30 ug/L) Sucrosomial Iron 4 tabs/day p.o. (2 tabs in the morning + 2 tabs in the evening) for 30 days • **low serum iron and normal ferritin** (>30 ug/L) Sucrosomial Iron 4 tabs/day p.o. for 30 days + EPO 40000 IU s.c./week • **Normal serum iron and high ferritin** (>100 ug/L) EPO 40000 IU s.c./week
2	MEDIUM PREOPERATIVE TIME (2–3 WEEKS) • **low serum iron and low ferritin** (<30 ug/L) Ferric Carboxymaltose 1000 mg i.v. (check Hb e ferritin after 10–12 days) possible repetition of ferric Carboxymaltose 1000 mg ev • **low serum iron and normal ferritin** (>30 ug/L) Ferric Carboxymaltose 1000 mg i.v. + EPO 40000 IU s.c./week • **Normal serum iron and high ferritin** (>100 ug/L) EPO 40000 IU s.c./week
3	SHORT PREOPERATIVE TIME (1 WEEK) • **low serum iron and low ferritin** Ferric Carboxymaltose 1000 mg i.v. + EPO 40000 IU s.c. × 2 • **Normal serum iron and high ferritin** (>100 ug/L) EPO 40000 IU s.c. × 2

For patients requiring urgent or emergency surgery, it may not be possible to correct all factors adversely affecting the baseline hemoglobin level and defects affecting hemostasis, and transfusions may be needed. In the preoperative period, patient preferences and acceptance or refusal to receive various blood components should be discussed, and related consents and advanced directives should be obtained and documented.

Recommended preoperative evaluation also includes a pain control plan. Oncologic patients often have pain prior to surgery and they often receive significant amounts of drugs and opioids to control it. The anesthesiologist needs an accurate idea of the patient's level of pain, opioid tolerance, and requirements [2]. Patient preparation should include adjustments of preoperative medications to avoid withdrawal effect, treatment to reduce preoperative pain/anxiety, and beginning of preoperative treatment as part of a multimodal pain control plan, which is vital to successful postoperative pain management.

To improve surgical outcomes, adequate patient hydration and nutrition should be achieved. Preoperative fluid management should guarantee a near-zero fluid balance. Studies show that optimization of the preoperative metabolism by operating a patient under the influence of insulin after giving a carbohydrate load preoperatively (such as 100 g oral carbohydrate drink) results in attenuation of glucagon release, complete abolition of cortisol release, which is thus far less catabolic in the postoperative period [11].

About preoperative fasting, the American Society of Anesthesiologists (ASA) recommends to minimize starvation: patients should stop solids 8–6 h and liquids 2 h before a procedure [12].

In extensive oncologic surgeries, bowel preparation can be useful. Laxatives should be given 2 days prior to the surgery and adequate intravenous hydration and nutrition should be ensured.

These preoperative interventions (along with patient and family education and counseling, preoperative cessation of smoking, drinking, prophy-

laxis of infection and thromboembolic events, and normothermia) are part of standard care pathways known as Enhanced Recovery After Surgery (ERAS) protocols. The aims of the ERAS programs are to reduce surgery-related morbidity, standardize and optimize perioperative medical care, decrease the length of hospital stay, and facilitate the patient's return to normal life [13].

On the scheduled surgery date, the patient is taken into the operating room. After his identification, a blood pressure cuff, ECG leads, and a pulse oximeter are applied to monitor his non-invasive blood pressure, electrical activity of the heart, and oxygen saturation. A large peripheral intravenous (IV) line is placed to administer premedication, drugs, fluids, and possible blood products.

The aims of premedication are reduction of anxiety and pain, enhancing the hypnotic effects of general anesthesia, promotion of retrograde amnesia, and reduction of vagal reflexes.

A great number of anesthetic techniques can be used. The epidural or spinal+epidural technique in combination with general anesthesia (GA) is usually the best choice [1, 2].

The level of the spine at which the epidural catheter is placed should be discussed with the orthopedic surgeon according to the location and size of the tumor, type and extension of the surgery, and patient positioning. Epidural catheters are usually inserted at T11 to T12 or T12 to L1 interspaces, prior to induction of general anesthesia, and they are secured in place to enable both additional intraoperative drug infusion and postoperative analgesia.

The administration of the anesthetic through the epidural catheter aims at obtaining analgesia up to dermatomes T2–T3, generating both systemic and cardiac blockade of the sympathetic nervous system, which causes vasodilatation and blocks the cardiac accelerator fibers, leading to hypotension and superior rate control [14].

According to the literature, intraoperative hypotensive epidural anesthesia may reduce bleeding and transfusion, improve the quality of the operative field, and shorten operative times [5, 6, 8, 14].

After epidural catheter placement, general anesthesia is induced, and the patient is intubated and connected to the ventilator. The anesthetics generally used are: Propofol, Midazolam, or Ketamine to reach unconsciousness, Fentanyl or Ketamine for analgesia, Rocuronium or Cisatracurium for neuromuscular block. The maintenance of general anesthesia can be achieved by inhalation of a volatile anesthetic agent (Desflurane, Sevoflurane) or by total intravenous anesthesia (TIVA) or TIVA target-controlled infusion (TCI). Low-dose Remifentanil (a short-acting synthetic opioid analgesic drug) is given intraoperatively by continuous intravenous infusion to relieve pain.

Placing an arterial catheter for continuous blood pressure monitoring and sampling, and acquiring a central venous line (internal jugular vein or subclavian vein) are imperative prior to surgical incision [2]. The central line placement should always be checked via chest X-ray, and the catheter tip position should be close to the border of the superior vena cava and the right atrium. A central venous catheter is fundamental for fluids and blood products infusion, infusion of vasoactive and inotropic agents, and monitoring the central venous pressure. Vasopressors and inotropes are often used to maintain an adequate central venous pressure and venous return and to optimize the heart rate, especially when a spinal or epidural anesthetic is used. Hypotension may cause hypoperfusion, thus blood pressure must be monitored closely, especially in those patients suffering from cardiovascular, pulmonary, or renal diseases. Studies have shown that an intraoperative mean arterial blood pressure value of 60 mmHg is enough to ensure optimal organ perfusion, although this target should be higher in hypertensive patients and those with cardiovascular comorbidities [7, 14].

Invasive blood pressure monitoring with arterial line is often associated with hemodynamic monitoring (MostCare®, Picco®) based on the analysis of the peripheral arterial waveform, which provides the measurement of the main hemodynamic parameters, thanks to specific algorithms.

A major complication of pelvic oncologic surgeries is massive blood loss [1–6]. Large tumors, requiring extensive resection with a

close proximity to vascular structures, will be more likely to cause a larger blood loss. The internal iliac vessels are more likely to be troublesome when tumors are situated posteriorly in the pelvis. Anterior pelvic tumors that require dissection near the bladder neck are problematic because of the perivesical venous plexus that often bleeds heavily or continuously. Bone cuts expose the bleeding bone and can provide a sustained source of hemorrhage. Renal cell metastatic tumors are exceptional in being highly vascular and are associated with massive blood loss during surgeries [1]. Therefore, controlled hypotensive epidural anesthesia is very helpful in aiming to decrease the intraoperative blood loss and red blood cell transfusion requirements [5, 6, 8, 14]. A restrictive red blood cell transfusion threshold of 8.0 g/dL is recommended for patients undergoing oncologic orthopedic surgery or when the blood loss is greater than 20% of the total blood volume. To manage the high flow of blood and fluids required, rapid infusion devices are often helpful. Autologous transfusions do not improve long-term outcomes over exogenous donor red blood cell transfusions [2]. Unfortunately, cell salvage cannot be used as it may increase the risk of spreading tumor cells systemically, although there is some promising research on the effectiveness of filtration and irradiation to reduce the tumor load of salvaged blood [1].

Acute Normovolemic Hemodilution (ANH) is another technique used to decrease the influence of perioperative transfusions. It is a blood conservation technique that involves the removal of blood from a patient shortly after the induction of anesthesia, with maintenance of normovolemia using a crystalloid and/or colloid replacement fluid. The blood withdrawn from the patient is frequently kept at room temperature, to be returned to the patient later in the procedure, no longer than 8 h after collection. This action has shown some efficacy in reducing allogeneic blood transfusions in cardiac and miscellaneous procedures, though not with orthopedic surgeries. This technique is also often associated with significantly higher intraoperative fluid and vasopressor requirements [2].

As with any prolonged surgery, careful attention needs to be given to prevent peripheral nerve compression. Poor patient positioning may result in devastating outcomes [2, 14]. Patients must be placed in anatomic positions with pressure points padded to prevent compression or stretch-related injuries. When a patient is placed in the supine position, if the upper extremities are abducted, they should remain supinated and padded at no more than 90 degrees. For a patient in the lateral decubitus or prone position, the neck should be maintained in a midline position, and there should be no excessive pressure on the eyes and ears. The abdomen should not be compressed because excessive pressure may compromise ventilation and decrease venous return from the lower extremities [2].

Pelvic tumor surgeries may be complicated by intraoperative hypothermia [14]. Patient heat loss is primarily due to a significant exposure of the skin and internal viscera to the operating room air. Forced air warming devices and fluid warmers can be adopted to prevent this complication. A low body temperature can cause and worsen acidosis and coagulopathy, and this combination can be lethal (known also as the trauma triad of death) [7].

A life-threatening complication is cardiovascular collapse, due to acute right heart failure secondary to massive fat embolism, which can occur during the reconstruction phase. Reaming and pressurizing of the femoral canal may release emboli into the circulation, although pulmonary embolism is less common in comparison to hip and knee arthroplasty.

Preemptive management involves avoiding hypovolemia and fluid overload, increasing the inspired oxygen concentration, minimizing the use of vasodilators, and starting the administration of vasopressors (dopamine, norepinephrine, dobutamine).

Transoesophageal echocardiography is not recommended, and the anesthesiologist should pay attention to clinical signs that include desaturation, hypoxemia, hypocapnia, hypotension, and increased pulmonary shunt [1]. If cardiovascular collapse occurs, the team should start cardiopulmonary resuscitation.

At the end of the procedure, patients, who had severe complications or undergo extensive procedures or have severe comorbidities, should be kept electively intubated and admitted to the Intensive Care Unit for postoperative ventilation, fluid and electrolyte management, and correction of coagulopathy. Otherwise, neuromuscular relaxation is antagonized (using sugammadex or neostigmine + atropine), maintenance agents are discontinued, and the patient is allowed to awake. Extubation is performed when spontaneous breathing is re-established and full recovery of neuromuscular activity is achieved.

Afterward, the patient is typically taken to the Post Anesthesia Care Unit (PACU) for an immediate postoperative follow-up, which includes airway management and oxygen administration, monitoring the vital signs (heart rate, blood pressure, temperature, and respiratory rate), managing postoperative pain, treating postoperative nausea and vomiting (PONV), treating postanesthetic shivering, and monitoring the surgical sites for excessive bleeding. Anesthesiologists and nurse anesthetists should also prepare and teach the use of patient-controlled units, and additional intravenous and/or epidural infusions could be administered. The patient remains in the PACU for 3–4 h to ensure recognition of possible late-onset pain or sedation. Then, the patient is transferred to the Orthopedic Oncology Unit. The arterial line is usually removed, and the central venous line should be maintained.

Most frequent postoperative complications are symptomatic anemia, postoperative pain, wound infections, urinary tract infections, and deep venous thrombosis [1, 2] (Table 21.2).

Table 21.2 Postoperative complications

Anemia
Postoperative nausea and vomiting
Excessive postoperative pain
Cardiovascular system
Myocardial infarction
Congestive cardiac failure
Atrial fibrillation
Angina
Wound
Superficial wound infection
Deep wound infection
Wound dehiscence
Wound hematoma
Urinary tract
Urinary tract infection
Acute urinary retention
Vascular system
Deep venous thrombosis
Venous thromboembolic disease
Respiratory tract
Pneumonia
Respiratory failure
Symptomatic electrolyte abnormality
Neurological
Peripheral nerve injury
Postoperative paralysis
Cerebrovascular accident
Death

Blood loss, Hb levels, and coagulation function should be monitored daily. Anemia is a very common complication. The decision criteria for blood transfusion should not be a static value, and the clinical status of the patient should be taken into account; however, one of the most important factors to be considered is the preoperative hemoglobin baseline. If necessary, blood transfusion and fluid therapy should be administered via the central venous line.

Postoperative nausea and vomiting (*PONV*) is often observed. The use of opioids, inhalation of volatile anesthetic agents, and/or specific patient factors are involved in this pathogenesis. A variety of pharmacologic agents are available to reduce the incidence of PONV, such as serotonin (5HT3) inhibitors (e.g., ondansetron); metoclopramide, droperidol, which act primarily through dopaminergic antagonism; dexamethasone [2].

In these patients, postoperative pain management is difficult and challenging, due to the substantial area and site of the surgery [1, 2]. Oncologic orthopedic patients characteristically suffer varying degrees of pain or discomfort long before operation, and this might cause worse postoperative pain, as mentioned earlier. Adequate postoperative pain control is crucial and complex because poorly managed pain can contribute to the development of short- and long-term postoperative complications including atelectasis, pneumonia, ileus, delayed ambulation, and postoperative persistent pain. Pain management

should be targeted to: (a) improve functional outcomes and reduced in-hospital length of stay, (b) reduce the inflammatory and stress response associated with surgery, (c) minimize the risk of persistent postsurgical pain development, and (d) facilitate the return of patients to the next planned oncological therapy [15].

There are several approaches to control the postoperative pain: **epidural analgesia** (EA) using either continuous epidural administration (CEA) and/or patient-controlled epidural (PCEA) techniques, or **intravenous analgesia** (IVA) using either continuous intravenous infusion and/or intravenous patient-controlled analgesia (IV-PCA). The method used is dependent on the technique chosen for anesthesia.

Patients who have an epidural catheter placed (CEA) can be managed with a continuous infusion of a local anesthetic (ropivacaine or levobupivacaine) in combination with an opioid (sufentanyl) (Table 21.3).

A patient-controlled analgesia device may be attached to the epidural line (PCEA). Patient-controlled techniques allow patients to self-administer small boluses of analgesics, thus providing better titration and enhancing responsiveness to their analgesic requirements. The device is programmed by the anesthesiologist to deliver a preset number of analgesic boluses per day, whenever the patient activates it. Each analgesic bolus is followed by a preset lockout period to avoid analgesic overdose. A patient-controlled epidural analgesia (PCEA) usually also consists of a continuous background infusion of a combination of a local anesthetic (ropivacaine or levobupivacaine) and an opioid (sufentanyl). Of note, during the patients' stay in the hospital, additional epidural boluses could be administered manually by the attending anesthetist upon the patient's demand. Usually the infusion is commenced in the ICU or PACU once the patient has become hemodynamically stable. The duration typically ranges from 2 to 6 days, based on the patient's pain. A careful postoperative monitoring of the epidural analgesia should be performed to detect an early onset of neurological complications. When epidural analgesia is no longer needed, the antithrombotic agent is withheld for 12–18 h before the catheter can be removed; and sensory and motor integrity should be reevaluated 6 h after the removal of the catheter.

Table 21.3 Epidural analgesia

Epidural analgesia	
Local anesthetic (from []% to []%)	+ Opioid (from []% to []%)
Ropivacaine (0.125% to 0.2%)	Sufentanyl 0.02%–0.04%
Or levobupivacaine (0.0625% to 0.125%)	

When the epidural catheter could not be placed, patients are given intravenous analgesia: a continuous infusion of analgesic (usually opioids such as morphine) or intravenous patient-controlled techniques (IV-PCA). In this case, the patient can self-administer analgesic preset boluses (morphine) via a patient-controlled analgesia pump. As for PCEA, the device is preset by the anesthesiologist and the analgesic bolus is always followed by a lockout period.

When possible, epidural analgesia should be always preferred. Studies demonstrated a clear antinociceptive superiority of the epidural over the intravenous methods for pain control [2, 16]. Besides, epidural infusion or PCEA often show a lower rate of side effects compared with IVA or IV-PCA. Some of the epidural analgesia benefits include excellent pain control despite the decrease in the total amount of opioids, with minimal respiratory depression, decreased somnolence, and early ambulation, although pruritus, nausea, urine retention, and rare neuroaxial disturbances are still described.

Another novel drug delivery system that is approved for use in 33 European countries but not yet in the United States is a sufentanil (opioid) sublingual tablet system (see Fig. 21.1). This system does not require an IV line. It is a handheld, preprogrammed, noninvasive, patient-activated device that delivers sufentanil 15-mcg microtablets on demand. Numerous studies and randomized, placebo-controlled trials have demonstrated its superiority compared with morphine delivery by IV PCA [17–19].

Fig. 21.1 Sufentanyl sublingual tablet system

Accordingly, the management of postoperative pain proves difficult and often requires a multimodal approach to achieve success. Parenteral opioids alone rarely provide adequate analgesia; the addition of non-opioid strategies (such as acetaminophen, NSAIDs, NMDA receptor antagonists (ketamine), gabapentinoids (pregabalin or gabapentin), β-blockers, α2-adrenergic agonists) is frequently beneficial and should be considered as pharmacological modalities that can be combined to regional or systemic analgesia techniques. However, non-steroidal anti-inflammatory agents are often avoided due to the problem of coagulopathy.

Multimodal analgesia should be considered as the standard of care modality with the goal of reducing the surgical stress and opioid consumption. It is a key component of enhanced-recovery pathways, improving short-term postoperative outcomes not only by shortening the length of stay in the hospital and accelerating the quality of return to "normal life" but also optimizing the conditions in which patients can return to their next intended oncological therapy [15].

The anesthesiologist, surgeon, and physiatrist should work together to minimize the orthopedic oncologic patient's postoperative discomfort.

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22 The Importance of a Multidisciplinary Approach to Pelvic Tumours

Andreas Leithner, Marko Bergovec, and Dimosthenis Andreou

In 1969, more than 50 years ago, the Apollo 11 mission successfully reached the moon, an event that has been internationally celebrated (Fig. 22.1). One might ask, "What does the moon landing have to do with pelvic tumours?" Multidisciplinarity! Such a success has only been possible due to a multidisciplinary, highly specialized team [1]. Another moon-related example is the NASA-team building game, where you are a member of a space crew and where you have to decide which items are the most important ones to successfully reach your 200-mile away space station. A team of people discussing options usually gets results closer to the NASA experts' solution than each of the individuals alone. Teamwork in critical situations like space missions is often the key to success [2].

Orthopaedic surgeons have been known to be highly intelligent [3], but even they cannot successfully treat primary pelvic sarcomas alone. As highlighted in the previous chapters, the multidisciplinary treatment of these tumours is of utmost importance: chemotherapy has been shown to be a prerequisite for survival in patients with osteosarcomas and Ewing sarcomas, and radiotherapy can improve the outcome in at least some patients with Ewing sarcoma, while none of us would ever perform surgery for pelvic chondrosarcoma without adequate imaging. The 2018 ESMO-EURACAN guidelines for soft tissue and visceral

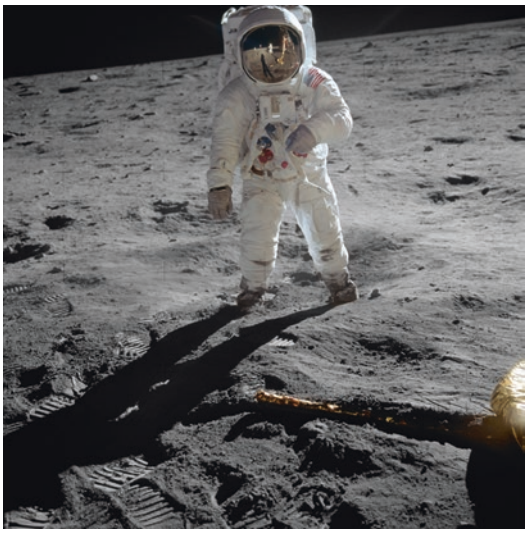

Fig. 22.1 Buzz Aldrin poses on the Moon [concerning licencing—picture taken from wikipedia.org—"This file is in the public domain in the United States because it was solely created by NASA. NASA copyright policy states that "NASA material is not protected by copyright unless noted". (See Template:PD-USGov, NASA copyright policy page or JPL Image Use Policy)"]

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sarcomas therefore state: “A multidisciplinary approach is, therefore, mandatory in all cases, involving pathologists, radiologists, surgeons, radiation therapists, medical oncologists and paediatric oncologists, as well as nuclear medicine specialists and organ-based specialists, as applicable” [4]. Similarly, the 2018 ESMO–PaedCan–EURACAN guidelines for bone sarcomas are focused on multidisciplinarity, stating for e.g. “Samples must be interpreted by an experienced bone sarcoma pathologist, in collaboration with the radiologist, and discussed in a multidisciplinary team” or “All new cases of bone tumours should be formally discussed in a multidisciplinary team at a bone sarcoma reference centre with the radiologist, the pathologist, the surgeon, the radiation oncologist and the medical and/or paediatric oncologist” [5].

We all know the importance of multidisciplinary treatment concepts; nevertheless, in some cases, we tend to forget. These are mostly patients with symptomatic metastases and benign or locally aggressive/rarely metastasizing pelvic tumours, who are sometimes not discussed in interdisciplinary tumour boards, as the indication for surgical treatment seems clear to the orthopaedic oncologist and other treatment options are not considered. But, even in patients with primary sarcomas undergoing multidisciplinary therapy, the optimal sequence of the planned treatment modalities can be unclear, especially if new findings render a reassessment of the necessary initial treatment plan.

22.1 Pathologists Need Clinical and Radiological Information

A 49-year-old woman was primarily admitted with multiple osteolytic tumours (Fig. 22.2a) for further surgical treatment after external curettage of one of those lesions and the external histological diagnosis of giant cell tumour (Fig. 22.2b), a diagnosis confirmed by a second histopathological investigation accomplished by a bone tumour specialist, who did not have access to the patient’s radiographs. Consequently, the patient was transferred to our tumour centre. To exclude the differential diagnosis of brown tumours due to hyperparathyroidism, we determined the parathormon level in serum, which was exorbitantly high at 922 pg/ml. Further investigations confirmed a parathyroid adenoma (Fig. 22.2c). After its resection serum levels of parathormon decreased but the patient developed a hungry bone syndrome, despite a prophylactic treatment with a high-dose calcium substitution. After 1 year of therapy calcium and CrossLaps values returned to normal levels and the radiologic controls showed bone consolidation (Fig. 22.2d). The patient was symptomless after 2 years of follow-up. The take home message of this case is that a pathologist has to have radiological and clinical information (e.g. multiple lesions) to reach the correct diagnosis.

22.2 Surgery Is Not Always the Best Option for Bone Metastases

Whereas it is clear that surgery will not be beneficial in some patients i.e. with multiple osteoblastic prostate cancer metastases (Fig. 22.3), intralesional curettage and stabilization with Steinmann pins may be necessary in other patients with i.e. symptomatic osteolytic pelvic metastases of a renal cell carcinoma, allowing immediate full weight-bearing and improving quality of life (Fig. 22.4). This procedure has been shown to lead to excellent results in selected patients. As a result of good experiences with surgeries like the Harrington procedure, many surgeons, when asked at conferences and courses, proposed similar surgical techniques in a case of a large supraacetabular defect in a 59-year old female patient with multiple endometrial carcinoma metastases (Fig. 22.5a). A multidisciplinary case discussion involving gynaecologists, medical and radiation oncologists, orthopaedic surgeons, radiologists and pathologists, however, came to the conclusion that surgery is not the treatment of choice. At 10 months of follow-up, after local radiotherapy and antihormonal therapy with an aromatase inhibitor, the patient was

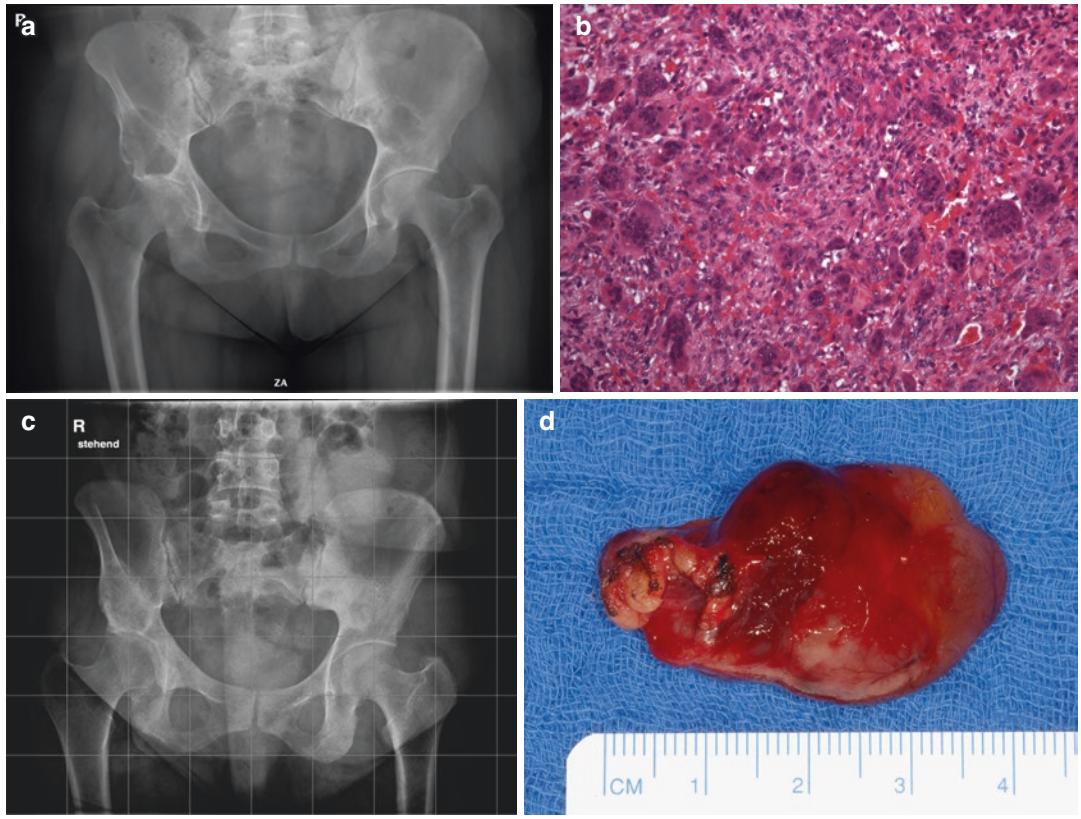

Fig. 22.2 Female, 49y, (**a**) a pelvis x-ray showing multiple osteolytic lesions; (**b**) the histology presents multiple multinucleate giant cells; (**c**) surgical specimen of the parathyroid adenoma; (**d**) complete radiological healing of the osteolytic lesions 2 years after parathyroid resection – without any pelvic surgery necessary

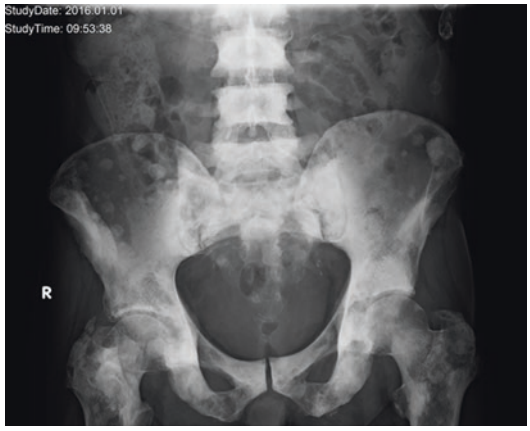

Fig. 22.3 Male, 59 years, multiple prostate carcinoma metastases in all parts of the skeleton

pain-free under full weight-bearing and had a good quality of life without having to undergo any surgery.

22.3 The Optimal Local Treatment Modality in Patients with Pelvic Ewing Sarcoma Is Still a Matter of Debate

No randomized controlled trials comparing the outcome of different local treatment modalities in patients with pelvic Ewing sarcomas are available. As a result, treatment strategies are greatly influenced by the sometimes conflicting evidence of smaller, retrospective analyses and the philosophy of the respective study groups. Definitive radiotherapy tends to be more often utilized in patients treated in the United States, whereas European Cooperative Study Groups consider the results of surgical treatment with or without radiotherapy to be superior, at least in patients with localized disease. However, the results of

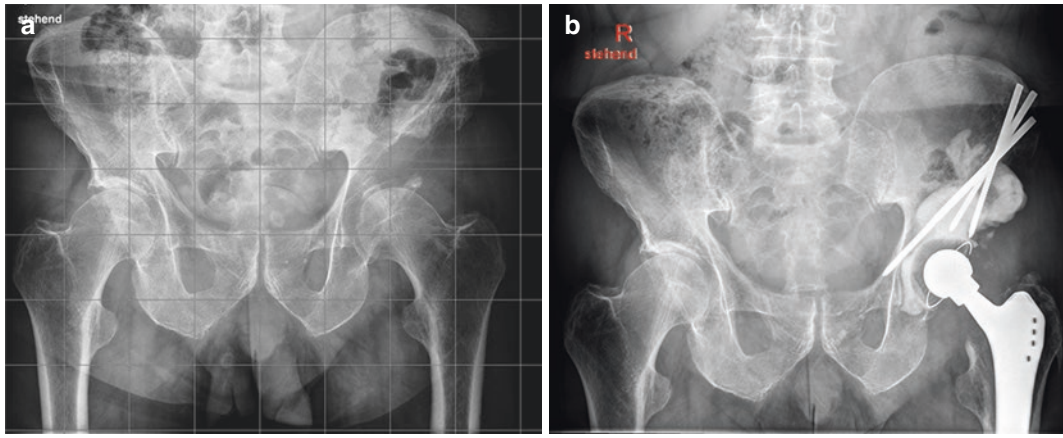

Fig. 22.4 Male, 75 years, multiple renal cell carcinoma metastases with (**a**) a painful disability due to the left supraacetabular osteolysis; (**b**) stable situation with full weight-bearing 10 months after a Harrington procedure following preoperative embolization

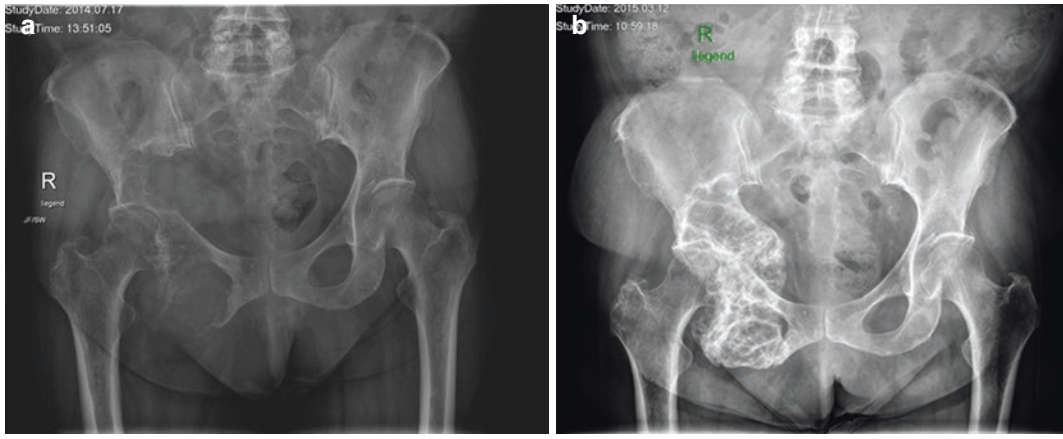

Fig. 22.5 Female, 59 years, multiple metastases of endometrial cancer, (**a**) with a large osteolytic lesion on the right supraacetabular site, (**b**) 10 months after local radiotherapy and antihormonal therapy

surgical treatment appear to depend on the tumour localization in the pelvis, as the Scandinavian Sarcoma Group recently demonstrated that definitive radiotherapy appears to be adequate for patients with sacral tumour localization [6]. A recent report of the Euro-EWING99 consortium verified this finding and additionally showed that patients with localized non-sacral Ewing sarcomas had a significantly better prognosis following surgery and additional radiotherapy, compared with surgery alone [7], raising the question of when radiotherapy should ideally be performed. Proponents of preoperative radiotherapy point out that it is associated with a lower toxicity and fewer long-term complications, compared to postoperative radiotherapy. On the other hand, a recent study demonstrated conclusively that the prognosis of patients with a poor response to neoadjuvant chemotherapy alone could be significantly improved with adjuvant high-dose chemotherapy—a highly toxic treatment that cannot be recommended for all patients [8]. Unfortunately, the histological response to treatment cannot be assessed in patients undergoing both preoperative chemotherapy and preoperative radiotherapy – as a result, the optimal

adjuvant therapy in these patients remains unclear. Therefore, no blanket recommendation for pre- or postoperative radiotherapy can be supported at this point, and the optimal treatment can only be determined after interdisciplinary discussion on a case-to-case basis.

22.4 "Real-Life" Multidisciplinary Treatment Looks Different Than on Paper

Multidisciplinary treatment protocols for patients with osteosarcoma and Ewing sarcoma typically stipulate that surgery is performed as soon as haematological recovery can be expected and that postoperative chemotherapy should commence 7–14 days after the surgical treatment. In reality, though patients tend to experience delays when moving from one discipline to another, especially if medical and surgical treatments take place in different hospitals. Additionally, patients with pelvic tumours undergoing surgery are at a high risk for postoperative complications, which may further delay the adjuvant treatment. However, emerging evidence suggests that such delays may have a negative impact on patients' prognosis. An as yet unpublished analysis of data of the Euro-EWING99 trial demonstrated that patients with localized disease and an interval between surgery and adjuvant chemotherapy longer than 16 days had significantly poorer overall and event-free survival probabilities compared with patients with shorter intervals [9]. This finding creates a challenge for treating physicians – surgeons do not want to endanger their reconstructions or risk of patients developing septic complications by clearing them to receive chemotherapy in cases of delayed wound healing, but they also do not want to have a negative influence on their patients' prognosis. A collective interdisciplinary decision on when to proceed with the adjuvant treatment can help address this challenge.

22.5 Multidisciplinary Treatment Plans Are Not Set in Stone

One of the most common pitfalls in the multidisciplinary treatment of patients with pelvic bone sarcomas is the lack of flexibility that is sometimes shown after multidisciplinary treatment plans have been decided on the following initial interdisciplinary case discussion at a tumour board, as the involved physicians may sometimes lose sight of the whole picture and only focus on their part of the plan. For example, a 42-year-old man presented with a localized pelvic leiomyosarcoma of bone affecting the cranial part of the ilium bone and the adjacent sacral bone (Fig. 22.6a). The interdisciplinary tumour board recommended neoadjuvant chemotherapy, followed by surgical treatment involving a type I/IV resection of the ilium and the affected ipsilateral sacrum and adjuvant chemotherapy. The MRI report after preoperative chemotherapy showed a partial tumour response and the patient was referred for surgical treatment, which was

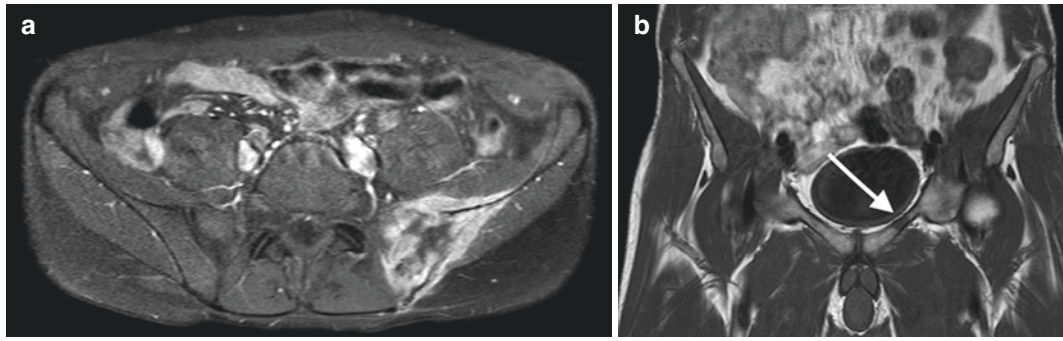

Fig. 22.6 Male, 42 years, with a leiomyosarcoma of the right ilium bone (**a**), and a new skip metastasis (**b**) in the pubic bone under treatment

accordingly planned. However, the musculoskeletal radiologist presenting the MRI in the preoperative tumour board also noted a new, previously undescribed lesion in the ipsilateral pubic bone, which had been presumably missed by the previous physicians who focused on the extension of the local tumour (Fig. 22.6b). Biopsy confirmed the diagnosis of a skip metastasis, and further two cycles of chemotherapy were applied to rule out further disease progression prior to surgical treatment, which now involved a complete interval hemipelvectomy.

In conclusion, a multidisciplinary approach to pelvic tumours is strongly advised. Otherwise, to apply the space flight comparison again, you might not end on the moon but in the marshland beside the space centre.

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Functional Rehabilitation of Pelvic Resection and Reconstruction

23

Stefano Masiero, Giacomo Magro, Mariarosa Avenia, and Francesca Caneva

23.1 Introduction

When we talk about rehabilitation, we mean "a set of measures that assist individuals, who experience or are likely to experience disability, to achieve and maintain optimum functioning in interaction with their environments" (WHO, 2011).

The purpose of rehabilitation interventions is to maintain or restore clinical stability, regain independence to perform everyday activities, and promote reintegration and social involvement.

Rehabilitation measures are aimed at achieving broad outcomes: prevention of the loss of function, slowing the rate of loss of function, improvement or restoration of function, compensation for lost function, and maintenance of current function. Interventions are performed according to an "individual rehabilitation project", designed on the basis of the patient's needs and recovery potential, the patient's and his family's preferences and the resources available.

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The rehabilitation project is the reference for every intervention carried out by the rehabilitation team: it defines the objectives in the short, medium and long terms, the expected times, the global and specific outcomes, the expectations and priorities of the patient, takes into account the global needs and preferences of the patient, of his impairments, disabilities, and above all, residual and recoverable abilities.

Some of the professionals who could be involved in the rehabilitation project are physiatrists, orthopedics, nurses, healthcare assistants, physiotherapists, speech therapists, psychologists, neuropsychologists: they form a multidisciplinary team only focused on the patient.

This kind of multidisciplinary approach is even more useful when dealing with a complex patient, like a cancer patient**.**

When patients receive a cancer diagnosis, they face different challenges, both physical and psychological. The lives of these people are totally upset in a short time, the condition of independence and autonomy in daily activities is lost, their social and family position changes, motor disabilities prevent them from performing simple actions, such as washing and being tidied up, eating, performing postural steps, transferring from the bed to the chair and vice versa, climbing stairs, and walking. All of this leads to a drastic reduction in the quality of life, also in relation to the patient's demand of functionality.

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The increase in cancer survivors determines an increase in the demand for functionality: it implies not only the correction of deformity or disability, but also their prevention and sometimes the need for episodic rehabilitation for life.

During the clinical assessment, the physiatrist evaluates organ functions and disabilities, physical, cognitive and behavioral impairments, social participation and quality of life, family and social environment surrounding the patient.

As a result of the medical examination, the rehabilitation specialist can prescribe motor rehabilitation, physical therapies in support of the motor and functional rehabilitation, treatment of pain syndromes, training in the use of prostheses, orthotics (hip orthosis and reclining wheelchair, pelvic-thigh brace) and assistive technologies, rehabilitation with the aid of robotized assistance, rehabilitation of organ functions and activities aimed at achieving autonomy.

Moreover, the educational and informative activities carried out by an interprofessional team are of great importance such as: training of the patient and their family on how to manage disabling issues and how to use prostheses, orthotics and assistive technologies, regular informational meetings with the patient and their family, and involvement of the patient and their family in the development and updating of the rehabilitation path and programs, giving information and advice to social workers, teachers, co-workers and anyone who could be involved in the management of the disabling issues and the condition of the patient.

23.2 Rehabilitation in Musculoskeletal Oncologic Disease

Patients with bone cancer can present general problems common to all cancers, such as damage from chemotherapy, radiation damage, disuse, asthenia and psychological problems; on the other hand, organ-specific problems due to the direct involvement of vascular, nerve, bone and muscle structures can be seen. A large pelvic surgery, in addition to bone resection and implantation of mechanical prostheses, often involves the partial or complete removal of the stabilizing muscles of the pelvis, the flexor-extensors and the abductors of the thigh. The resection of the head insertions of these muscles will clearly result in a loss or a lack of function in the following phases.

The functional complications most often observed after a pelvis resection are diverse and include: presence of moderate-strong pain especially in the first post-operative days, edema of soft tissue that can affect the ipsilateral lower limb, reduction of the range of joint movement (ROM) in flexion-extension and abduction of the ipsilateral limb, a decrease in the muscle strength (ilio-psoas, quadriceps, gluteus) also correlated to the period of time spent in unloading before surgery, poor motor control (alteration of proprioception) with the presence of compensatory mechanisms, a discrepancy in the limb length (heterometry due to prosthesis length) and neuropathic pain, which in turn can affect daily activities [1].

The factors that influence rehabilitation in this type of patient are the complexity of surgery (surgical demolition, tumor extension, etc.), the type of prosthesis, the residual structures to be recovered, the functional demands and the quality of life expected.

Radiation therapy for bone cancer can cause stiffness and less ability to move joints, as well as loss of muscle strength. A physical therapist can teach exercises to help keep joints and muscles healthy and to work properly (also before radiation therapy) and it may be necessary for a long time after the end of the treatment.

Finally, the treatment of the surgical wound should not be overlooked: surgical wounds in these patients are often more extensive than those associated with traditional prostheses due to the highly invasive surgery required, so manual debridement may be required [2].Indeed, oncological resection requires large resections, which also includes a portion of the soft tissue cover not involved as a surgical margin.

Furthermore, it may be necessary to remove or repair the neuro-vascular bundle nearby, so a complete assessment of the neuro-motor loss

would be needed to plan dynamic strength training and external support requirements. Partial or complete loss of the joint capsule and dynamic stabilizers of the hip joint during tumor resection can leave the hip joint vulnerable to dislocation. This can be enhanced with certain combination of movements, if these joint movements are allowed beyond a certain limit. This restriction depends largely on the surgical approach. The posterolateral approach is more common in the limb salvage surgery (LSS) of this site. Hip rotations, particularly internal rotation, flexion greater than 60° and adduction of the hip joint should be prevented up to 6 weeks. These movement limitations could be achieved using the hip abduction pillow/bracing and the de-rotation splint.

Before patients are discharged from the hospital, it becomes imperative to train them regarding the transfer from the bed in the supine position to standing (to lie supine and to sit on a chair/comfortable in the initial phase of rehabilitation). Knee joint mobilization should be initiated early from the edge of the bed, with the hip joint well supported or laterally with the cushions between the legs. Any restriction of the knee joint interval would adversely affect the overall function since the function of the hip joint of the ipsilateral leg has already been impaired. From a biomechanical viewpoint, lateral pelvic stability is provided by a hip abductor [3]. The reconstruction of abductor muscles, which provide hip joint stability, has been reported as a key to achieve a better functional outcome after proximal femur resection [4]. On the other hand, the pelvic site of the bony attachment of the abductor muscles is typically resected without abductor reconstruction in patients undergoing pelvic tumor resection.

Using pelvic-thigh brace, particularly indicated in prosthetic surgery, helps to treat postsurgical condition, thanks to the immobilization due to the thigh support. This peculiar brace presents a greater prolongation on the medial part of the thigh, which, by resting on the medial condyle of the femur, prevents unfavorable rotations of the hip joint. This gives great security to the patient, stabilizes the hip, limits mobility and allows the bipedal station and early intercourse. In addition, it allows different and progressive adjustments of flexion/extension and abduction/adduction of the hip, giving greater safety to the patient and allowing an early ambulation.

To achieve a truly complete recovery and effective motor control, the patient needs to reestablish an effective proprioceptive sensitivity. The computerized proprioceptive platform (Fig. 23.1) is an absolutely current and technologically advanced tool for rehabilitation. It is a pivoting platform (the degrees of which can be decided on the basis of the patient's condition) connected to a computer that allows certain movements to be performed in order to fully recover the proprioception, based on a personalized training program.

The patient can move in an orthostatic position or sit according to the adopted strategy and objectives, breech or mono-breech based on the type of injury and at the current stage of rehabilitation. The computer screen shows a very important visual feedback for the patient's true

Fig. 23.1 The computerized proprioceptive platform is a pivoting platform connected to a computer that allows certain movements to be performed in order to fully recover the proprioception

Fig. 23.2 Treadmills equipped with digital mirror able to detect every single angular movement of the runner

perception. Only in this way does he really realize what kind of movement, error and speed of correction execution he is performing.

It is now scientifically consolidated that the control of our reflection helps us to improve our movement through the "continuous feedback system" method. The patient experiences a great advantage in reconstructing his motor map, thanks to the help of the reflected image with progressive stimulations.

Nowadays, there are particular treadmills equipped with a 3D camera (Fig. 23.2) that transform the classic mirror in to a digital mirror, which has the ability to detect every single angular movement of the runner, with precision and reliability. Patients can immerse themselves in virtual environments and get a real-time stimulating feedback, both of postural and symmetry type of the supports on the ground.

The latest studies [3], with the incorporation of objective and validated measures of the function, indicated that the patient undergoing LSS obtained higher scores than those with amputations [4].

This suggests the need to examine the post-surgical functional outcomes and plan a personalized treatment in order to provide the best functional results and maximum achievable independence [5, 6].

23.3 Assessment of Functional Status

The variability of the factors involved and the need to customize the rehabilitation treatment is the basis of the rehabilitation process in oncologic patients.

It is, therefore, necessary to try to define a common line of therapeutic intervention, use the most accurate possible assessment scales, which include measures of strength, mobility, general state of psycho-physical health and the quality of life of the patients.

Different tools have been employed to assess the functional status of patients, the most validated in literature are:

- Range of Motion (ROM): measured with a simple hand-held goniometer to obtain both passive and active ranges of motion, [3] provides a measure of joint mobility. It may indicate the presence of partial or complete stiffness (Fig. 23.3).
- Strength can be measured under both isometric and isokinetic conditions by utilizing isokinetic dynamometers [7]; strength measurements of the unaffected limb may be used as a reference.

Fig. 23.3 Physical evaluation of ROM and strength

- Subjective functionality scale such as: the Musculoskeletal Tumor Society scoring system (MSTS) [8]. It is a disease-specific instrument to determine the physical and mental health for patients with extremity sarcoma, which is used to evaluate six items, including pain, function, emotional acceptance, use of any external support, walking ability, and gait alteration
- In addition, the Toronto Extremity Salvage Score (TESS) is widely used for the functional assessment of patients following surgery for musculoskeletal tumors.
- Health status survey short form-36 (SF-36v2) is also frequently used as a measure of health status. It consists of eight sections that evaluate vitality, physical functioning, bodily pain, general health perceptions, physical role functioning, emotional role functioning, social role functioning, and mental health.
- FMA (Functional Mobility assessment) is a self-report outcomes tool designed to measure the effectiveness of wheeled mobility and seating interventions for patients with disabilities.
- Energy expenditure testing to assess differences between groups and provide a global measure of functional performance for comparing a patient's status before and after an intervention:
  - -Gait analysis is used to assess and treat individuals with conditions affecting their ability to walk; it can be used to measure velocity, cadence, ROM, single-limb support time, swing and stance times, and double limb support time, and if it is associated with a dynamic electromyography, we can study patterns of muscle activity during gait.
  - -Oxygen consumption during gait provides an objective measure of gait performance. Individual gait efficiency may be calculated quantitatively by measuring the oxygen consumed per unit of distance travelled per kilogram of body mass.

23.4 Rehabilitation Protocols in Pelvic Resection

Although LSS for malignant bone tumors is considered the treatment of choice, rehabilitation guidelines for this kind of patients have yet to be formally established. Detailed guidelines for this patient population are stratified by the anatomical position, the type of prosthesis, the width of resection, in order to make the rehabilitation protocols applicable and reproducible. With regard to the pelvic and proximal reconstruction of the femur, it can be differentiated according to the involvement of the acetabulum or its exclusion in the periacetabular areas and in the non-acetabular areas (iliac bone, ischium, pubis and sacrum).The protocol also varies according to the type of reconstruction performed, which can be: with prosthesis, with bone graft, with prosthesis and graft [9].

23.5 Rehabilitation Program

The rehabilitation program should be designed to address:

1. Pain. Since cancer patients generally experience multiple concurrent pain syndromes, adequate pain control is an absolute requisite for successful rehabilitation. Pain control might require the integrated use of anticancer treatments, agents from multiple analgesic classes, interventional techniques, topical agents, manual approaches, and modalities.

The unique disease context, in which cancer pain develops, distinguishes it from many other pain-associated diagnoses managed by physiatrists. The majority of cancer pain is due to tumor effects, and for this reason, disease-modifying, anticancer therapy plays a critical role in pain management. For example, radiotherapy often offers a definitive and effective means of controlling pain associated with symptomatic and uncomplicated bone metastases. Bone metastases occur in 60–84% of patients with solid tumors. Pain intensity does not correlate with the number, size, or location of bone metastases. Bone pain is particularly relevant to physiatrists because recruiting muscles that act on or loading affected structures can precipitate severe pain [10].

Pharmacological and instrumental physical therapy to manage pain can include:

(a) Drugs: paracetamol, NSAIDS, opioids, antidepressants, anticonvulsants.
(b) Transcutaneous Electrical Nerve Stimulation(TENS) with rectangular currents pulsed from 30–150 μs at a low frequency (10–150 Hz);
(c) Low level laser therapy (LLLT) with analgesic and decontracting effects;
(d) Massotherapy for the reduction of muscle contraction and stasis edema. This should be applied only 1 month after the end of chemotherapy treatment, because there is a risk of promoting the spread in the blood or in the lymphatic circulation of metastatic components.

2. ROM recovery, muscle tone, mobilization and toning from the immediate post-operative phase, first in discharge and then in treatment, postural and behavioral hygiene, compensatory strategies, and use of guardians.
3. Contrast of Cancer Related Fatigue syndrome (CRF). The National Comprehensive Cancer Network defines CRF as "an unusual, persistent, subjective sense of tiredness related to cancer or cancer treatment that interferes with usual functioning". This is an integral part of the patient's symptoms, disproportionate to the degree of activity and not reducible with sleep or rest.

It is known to be the most common symptom experienced by cancer patients. It affects almost 75% of these patients and 60% of them consider it more disabling than pain. CRF is especially associated with chemotherapy and drug therapy cycles and a quite aggressive incidence. In fact, a majority of patients in active treatment rate their fatigue as "severe" or 7 or more on an 11-point numerical rating scale.

4. Education of the patient and caregiver to provide continuity of assistance. Learning of basic nursing techniques and functional rehabilitation (postural steps, assisted walking, and maintenance exercises).

23.6 Rehabilitation Phases

The rehabilitation program is distinguished by three phases:

1. Pre-operative: absolute proscription of the load from the moment of diagnosis, education in walking with brachial sticks. Rehabilitation even before starting primary cancer therapy and surgery, such as crutch muscles strengthening, could be of great benefit in the post-treatment functional outcome.
2. Early post-operative: simple isometric contractions with the operated limb, contralateral limb mobilization for vasomotor and antithromboembolic purposes, and diaphragmatic respiratory rehabilitation. Slow and concentric exercises are preferred to stimulate the slow type I fibers, which undergo a greater volumetric and functional reduction from rest (up to 30% after 5 weeks). Walking with overflow load.
3. Advanced post-operative: granting of the progressive load and, if applicable, complete; the use of aids to stimulate walking and the patient's "fear of fracture"; co-contraction exercises of the antagonist muscles of the lower limb promote stability and load transfer; proprioceptive exercises; muscle strengthening exercises. Two daily sessions (Table 23.1).

Table 23.1 Example of rehabilitation protocol in pelvic or proximal femoral resection

1 Step (week 1–4)	2 steps (week 5–8)	3 step (week 9–12)
• Extension braceactive knee and ankle joint mobilization and active limb contralateral • Quadriceps isometric reinforcement and ipsilateral sural triceps for maintaining the tone and the trophism • Distal mobilization exercises of the treated limb to prevent venous and lymphatic pooling. • Self-mobilization and strengthening exercises of the body districts not involved in the surgery	• Release of the brace at 45° from the 30th day, then at 60° on the 45th–50th day, • Mobilization. Passive and active assisted hip flexion extension under the pain threshold • Stimulation with electrotherapy, preparatory to walking. • Gradual replacement of the walking frame by elbow crutches: Gait showed slight abduction of the hip (which gradually regressed)	• 60th day, brace unblocked at 90° and progressive load • From 90th day the brace is removed and the full load is granted • Muscle reinforcement with concentric exercises and in isometric seal • Aerobic exercises • Functional exercises in preparation for return home, including stairs training, re-education in the activities of daily living and the postural movements involved in entering and leaving a car.

23.7 Conclusion

The rehabilitation of orthopedic oncology patients needs a global management by a multidisciplinary and expert team that defines a project and a rehabilitation program based on their specific characteristics.

Despite the fact that early physical rehabilitation is the key to achieve good functional outcome and quality of life after LSS, rehabilitation techniques following LSS are largely neither tested nor documented in detail [11].

Most of the resection and oncological reconstruction varies from one individual to another even in a particular site and requires a personalized rehabilitation protocol to design an individual rehabilitation program.

In a recent paper, Shehadeh et al. [12] reported that, following a standardized rehabilitation protocol produced an improved functional outcome, even though their conclusion is based on a small observational study with heterogeneous population who received different types of LSS for different anatomical sites.

Although limb savage surgery for primary malignant tumors has improved because of a significant progress in surgical techniques and endoprosthetic design and manufacture, without optimal peri and postoperative physical rehabilitation, achieving the desired quality of life may not be feasible.

Vincent S. Paramanandam et al. [2] tried to establish some fundamental concepts on which rehabilitative treatment should be based for this type of patients, first of all, the presence of a rehabilitation team of cooperating professionals for the whole period of hospitalization (protocols last more or less 60 days).

Another essential concept is training patients and their caregivers on how to use specific aids (hip orthosis and reclining wheelchair, and pelvic-thigh brace) to provide support to the limb.

New technologies, such as computerized proprioceptive platforms or the treadmill associated with virtual reality, can greatly help patient in motor recovery, making him an active part of the rehabilitation process and setting short-term goals that transmit confidence in his own means.

The role of the physiatrist, in addition to coordinating the multidisciplinary team, is to guide the patient through a long and difficult path and a physical and psychological challenge. This also includes the assessment of the new impairments, the attempt to recover the normal motor function, and in many cases, the acceptance of the disability with the learning of adaptation strategies.

It is essential for rehabilitation team to understand the real needs of the patient and set realistic goals, in order to achieve the highest possible quality of life.

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